Sanjeev Sabharwal
Editor

Pediatric Lower Limb Deformities

Principles and Techniques of Management

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To all children with limb deformities and their caregivers.

Foreword

"There is only one child in the world and the Child's name is All Children." Thus did the poet Carl Sandburg succinctly describe the universal appeal of children and humankind's concern for child welfare, health, and happiness. The most successful charitable organizations, such as Easter Seals (originally, the National Society for Crippled Children) and the March of Dimes (originally, the National Foundation for Infantile Paralysis), continue to focus fundraising and service projects on children with birth defects and developmental and acquired deformities and disorders.

Likewise, major service organizations and clubs, including the Shriners and the Scottish Rite Freemasons, point to their respective children's hospitals with pride and a sense of accomplishment. All of these efforts, which occupy so much time and energy, are directed towards a single goal: helping clinicians and researchers cure or relieve those conditions that prevent a child from reaching his or her full potential. In this regard, there is no higher calling that a human being could possibly undertake than being a frontline soldier in the battle against childhood diseases, disorders, and deformities.

Up until the mid-twentieth century, surgery focused on correcting pediatric congenital, developmental, and acquired musculoskeletal defects and deformities had a limited capacity to obtain full restoration of function. Often times, amputation proved the most practical means of achieving maximal functional capacity for a child with certain limb deficiencies. Correction of angular long bone deformities, for example, involved wedge resection of osseous tissue that would certainly improve the angulation but left the youngster with the need for clunky shoes or braces (or both) to get around.

In 1951, Prof. G. A. Ilizarov discovered how to create new osseous tissue in a widening distraction gap. He worked in far-off Siberia, alone at first, away from the prying eyes of Soviet colleagues, giving him a chance to learn by trial and error the parameters of his evolving methodology. More than 30 years elapsed before surgeons in Western countries learned of Ilizarov's discoveries. By then, techniques and instrumentation had been perfected, basic science research was completed, and a massive Medical Center had been constructed in Kurgan, USSR, all to provide care to children and adults with musculoskeletal conditions never before thought treatable.

As a result, the Methods of Ilizarov first appeared in Western medicine as a mature system of treatment, capable of achieving stunning results. In a sense, the situation resembled the appearance of Nike Athena in Greek mythology, who sprung as a fully grown adult from the head of Zeus, armor-clad and battle-ready.

In the 25 years since the introduction of Ilizarov's therapeutic strategies into Western medicine, ingenious surgeons have found ways to combine Ilizarov's distraction osteogenesis with well-established principles of deformity correction, growth rate prediction, and soft tissue releases to yield new and ever-evolving paradigms to deal most effectively with conditions that interfere with a child's full participation in life's activities.

Although these combined surgical techniques are reported piecemeal in journal articles and at open meetings, there has not been, until now, a textbook updating the current principles of pediatric deformity correction in a comprehensive way. The volume you are holding in your hands was specifically designed to overcome such a deficiency. Dr. Sabharwal has prevailed upon leading practitioners of these modern strategies to write chapters in their fields of interest

and clinical research. As a result, this book is a remarkable compendium of primary source material that will aid clinicians around the world in treating lower limb deformities of childhood.

My only regret is that visionary founders of the charitable organizations and service clubs who dedicated so much effort to establish children's hospitals and child-centered research and support enterprises are not alive today to see what has become of their dreams and hard work. They would be amazed, that's for sure!

Orange, CA, USA Stuart A. Green, MD

Preface

In titling his 1741 book, Nicolas Andry coined the phrase *Orthopédie* from two Greek words: *orthos*, "straight, correct," and *paiedeia*, "rearing of children." While the field of orthopedics has branched further into a number of subspecialties affecting various body parts in the young and old, Andry's original illustration of a straight stake tied to a crooked sapling has withstood the test of time. However, despite the ubiquitous presence of lower limb deformities in children globally (albeit with varied etiologies), a textbook devoted to the treatment of the "crooked child" is sorely missing from the current literature.

This unique text is primarily intended for orthopedic surgeons and trainees worldwide who have an interest in pediatric lower limb deformities. This book is not meant to be a "how to apply an external fixator to the tibia" type of manual, but rather a broad-based text highlighting both general principles and specific strategies for managing the entire spectrum of pediatric lower limb deformities, applying to circumstances of various etiology and resource availability. The authors of the 32 chapters are well-known leaders in the field of pediatric lower limb deformities, and I am truly indebted to every one of them and their coauthors for their excellent contributions. I invited these contributors not only on the basis of their expertise in the field but also in light of the diversity of their working environments and unique challenges that they face when treating children with limb deformities.

This book is divided into five parts, although each chapter can serve as a standalone guide for the clinician dealing with a specific patient. Part I highlights the general principles and techniques, including patient evaluation, decision making, and various surgical methods for deformity correction. Part II deals with related concepts, including management of soft tissue contractures, amputation, and working in resource-challenged environments. Part III includes lower limb deformities associated with specific metabolic, neuromuscular, and tumor-related conditions, as well as skeletal dysplasias. Part IV covers specific congenital and developmental disorders of the lower extremity. Finally, Part V explores various sequelae and complications associated with lower extremity deformities in the growing child. All authors were encouraged to incorporate relevant figures, tables, and highlight boxes to clearly deliver their message to you, the reader. While I did spend many hours editing and making suggestions to each of the corresponding authors, my role here was simply that of a facilitator.

When asked why I chose the field of pediatric orthopedics, I often respond by saying, "It just feels right to make a child's crooked leg straight," perhaps not too far off from what Nicholas Andry had in mind more than 250 years ago. While that may be too simplistic of an answer, I do hope that this book will resonate with you and help guide your encounters with these children and their caretakers.

Newark, NJ, USA

Sanjeev Sabharwal, MD, MPH

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This work would not have been possible without the professional relationships that were developed over the years with members of the Limb Lengthening and Reconstruction Society (LLRS), Pediatric Orthopedic Society of North America (POSNA), and Pediatric Orthopedic Society of India (POSI). I am thankful to the residents, staff, and faculty—in particular, the late Fred Behrens, M.D.—of the Department of Orthopedics at the Rutgers, New Jersey Medical School, who have taught me the value of patience and persistence. The constant support of the Springer publishing team, especially Diane Lamsback and Kristopher Spring, was vital in making this book a reality.

I am grateful to my parents, grandparents, mentors, and students for helping me recognize the importance of integrity and hard work. Thanks to my dear wife, Ranjit, who for the past 30 years took care of essentially everything so that I could pursue an academic career in pediatric orthopedics. I deeply appreciate our three children, Samir, Simran, and Sabhyta, for keeping me honest and grounded.

Contents

Part I General Principles and Techniques

1 Etiology of Lower Limb Deformity 3
Viral V. Jain, Sarah Zawodny, and James McCarthy

2 Clinical Evaluation Including Imaging 15
Joseph J. Gugenheim

3 Decision Making in Lower Extremity Deformity Correction 37
Mark L. Miller and J. Eric Gordon

4 Growth Modulation for Angular and Length Correction 51
Peter M. Stevens

5 Physeal Bar Excision 67
Karl E. Rathjen and Anthony I. Riccio

6 Acute Deformity Correction Using an Osteotomy 79
Vrisha Madhuri and Sangeet Gangadharan

7 Gradual Deformity Correction 105
David S. Feldman, Adam M. Kurland, and Abdel Majid Sheikh Taha

8 Hybrid Techniques for Limb Length and Deformity Correction 121
Mark T. Dahl and Chang-Wug Oh

9 Motorized Intramedullary Lengthening, an Emerging Technology for Limb Length and Deformity Correction 131
Mark T. Dahl

Part II Related Concepts and Management Options

10 Biomechanically Based Clinical Decision Making in Pediatric Foot and Ankle Surgery 153
Jon R. Davids

11 Pediatric Joint Contractures 163
Christopher Iobst

12 Physical Therapy During Limb Lengthening and Deformity Correction: Principles and Techniques 181
Anil Bhave, Erin Baker, and Mary Campbell

13 Amputation and Prosthetic Management: Amputation as a Reconstructive Option 199
John A. Herring

14 Working in Resource-Challenged Environments 215
Scott C. Nelson and Hugh G. Watts

Part III Underlying Conditions

15 Metabolic Disorders 231
Mehmet Kocaoglu, I. Levent Eralp, and F. Erkal Bilen

16 Osteogenesis Imperfecta 255
François R. Fassier

17 Lower Limb Deformity in Neuromuscular Disorders: Pathophysiology, Assessment, Goals, and Principles of Management 267
Unni G. Narayanan

18 Arthrogryposis 297
Reggie Hamdy and Noémi Dahan-Oliel

19 Limb Lengthening and Deformity Correction in Patients with Skeletal Dysplasias 313
Mihir M. Thacker, Ellen Dean Davis, Colleen P. Ditro, and William Mackenzie

20 Lower Extremity Benign Bone Lesions and Related Conditions 333
Lori Karol

21 Management of Juxtaphyseal Malignant Bone Tumors Around the Knee Joint: New Concepts in Limb-Sparing Surgery 349
Hidenori Matsubara and Hiroyuki Tsuchiya

Part IV Congenital and Developmental Disorders

22 Congenital Femoral Deficiency Reconstruction and Lengthening Surgery 361
Dror Paley, David Y. Chong, and Daniel E. Prince

23 Fibular Hemimelia: Principles and Techniques of Management 427
John E. Herzenberg, Lior Shabtai, and Shawn C. Standard

24 Tibial Hemimelia 455
Dror Paley and David Y. Chong

25 Congenital Pseudarthrosis of the Tibia 483
Corinna C. Franklin and Richard S. Davidson

26 Congenital Posteromedial Bowing of the Tibia 495
Benjamin Joseph, Hitesh Shah, and N.D. Siddesh

27 Controversies in Blount's Disease 503
John G. Birch

Part V Sequelae and Complications

28 Methods to Enhance Bone Formation in Distraction Osteogenesis 519
Hae-Ryong Song, Dong Hoon Lee, Seung-Ju Kim, and Ashok Kumar Ramanathan

29 Residual Deformities of the Hip 535
Shawn C. Standard and Daniel K. Ruggles

30 Posttraumatic Lower Limb Deformities in Children 569
Ashok N. Johari, Sandeep A. Patwardhan, and Taral Vishanji Nagda

31 Postinfectious Deformities of the Lower Limb 587
In Ho Choi

32 Iatrogenic Deformities 603
Austin T. Fragomen and S. Robert Rozbruch

Index 621

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Part I

General Principles and Techniques

Etiology of Lower Limb Deformity

1

Viral V. Jain, Sarah Zawodny, and James McCarthy

Introduction

Lower limb deformities can consist of any combination of length inequality, angular deformity, and/or asymmetric girth. While the list of possible etiologies is quite extensive, categorizing them can help simplify diagnosis and treatment. Many investigators have broadly classified lower limb deformity into congenital and acquired, with some including a developmental group. This chapter focuses on the etiologies that are most clinically relevant to the orthopedic surgeons.

We have divided the causes of pediatric lower limb deformities into four broad categories: namely underlying conditions, congenital, developmental, and acquired deformities from sequelae of disease or complications (Table 1.1). Most of these disorders are discussed in greater details in the subsequent chapters.

Underlying Generalized Conditions

Metabolic Disorders

Certain metabolic disorders and endocrinopathies can affect the maturation of growth plate chondrocytes by the alteration of normal regulatory signals or by disturbance of the functional matrix components necessary for new bone formation. Some endocrine disorders such as hypothyroidism and growth hormone deficiency [1] and metabolic disorders including renal osteodystrophy [2] can mechanically weaken the physis, predisposing to pathologic conditions such as slipped capital femoral epiphysis (SCFE). These same disorders can render the chondrocytes more susceptible to compressive forces, leading to angular deformities.

Rickets

Rickets is a clinical manifestation of defective mineralization of the long bone physes due to inadequate levels of calcium or phosphate in children. Osteomalacia, the defective mineralization of osteoid at cortical and trabecular surfaces, is seen in both children and adults. There are many different forms of rickets that result in very similar radiographic and clinical manifestations (Table 1.2). Patients may have decreased longitudinal growth and angular deformities [3] (Fig. 1.1).

Renal Osteodystrophy

Renal osteodystrophy is the combination of rickets and hyperparathyroidism that occurs secondary to end-stage renal disease. The findings of rickets are explained by the failing kidney being unable to produce a sufficient amount of 1, 25-dihydroxyvitamin D3. Additionally, the dysfunction of the tubular system decreases clearance of phosphate and causes secondary hyperparathyroidism, which manifests as brown tumors and subperiosteal erosions [3].

The deformity that results from these metabolic disorders depends on the stage along the physiologic varus/valgus spectrum that the child is at the age of onset of the disorder. The onset of nutritional rickets and inherited disorders is generally seen during infancy and therefore they produce genu varum. On the other hand, renal osteodystrophy usually causes genu valgum because acquired renal failure occurs later when children are more likely in the physiologic valgus age group [4].

Genetic Disorders

Osteogenesis Imperfecta

Osteogenesis imperfecta is caused by genetic mutations that affect the quantity or quality of collagen produced. Many distinct mutations have been identified and can thus create a

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Table 1.1 General categorization of the types of lower limb deformities in children based on etiology

Underlying conditions	Metabolic: rickets, renal osteodystrophy
	Genetic disorders: osteogenesis imperfecta, neurofibromatosis
	Neuromuscular: cerebral palsy, Charcot-Marie-Tooth arthrogryposis
	Skeletal dysplasias
	Benign tumors: fibrous dysplasia, Ollier's, multiple hereditary exostoses
	Malignant tumors
	Inflammation: rheumatoid arthridities, hemophilic arthropathy
	Vascular anomalies
Congenital	Congenital femoral deficiency (proximal focal femoral deficiency)
	Congenital fibular deficiency (fibular hemimelia)
	Congenital tibial deficiency (tibial hemimelia)
	Tibial dysplasia: congenital pseudarthrosis, posteromedial bowing
	Hemihypertrophy
	Amniotic band syndrome
	Congenital knee dislocation
Developmental	Genu varum
	Blount's disease
	Genu valgum
Acquired: sequelae and complications	Residual hip deformity: developmental dysplasia of the hip, slipped capital femoral epiphysis, Legg-Calve-Perthes, avascular necrosis
	Posttraumatic: Cozen fracture
	Post-infectious
	Iatrogenic

Table 1.2 Rickets and related disorders

Type	Cause
Nutritional	Vitamin D deficiency
	Dietary calcium deficiency
	Combination of both
Gastrointestinal	Poor absorption due to disease (gluten sensitivity, Crohn's/ ulcerative colitis, short gut)
End-organ insensitivity	Insensitivity to vitamin D_3
Vitamin D dependent	1 alpha hydroxylase deficiency
X-linked hypophosphatemia	PHEX gene defect, wasting of phosphate in kidney
Renal tubule abnormality	Kidneys waste many molecules, particularly phosphate
Hypophosphatasia	Alkaline phosphatase deficiency (gene mutation on chromosome 1)
Renal osteodystrophy	Renal failure

wide spectrum of phenotypic manifestations. Those who are more severely affected have extensive bony fragility that leads to numerous fractures, short stature, and angular deformities of the long bones and the spine [5].



Fig. 1.1 Genu valgum in a 7-year old-female. The etiology was nutritional rickets. Image courtesy Dr. Hitesh Shah

Neurofibromatosis

Neurofibromatoses (NF) are a group of genetic multisystem disorders involving products of all three germ lines: neuroectoderm, mesoderm, and endoderm. The most common and orthopedically relevant form is neurofibromatosis type 1 (NF1) previously known as von Recklinghausen disease, which involves predominantly peripheral nervous system although it involves cells of mesodermal origin as well.

NF1 is one of the most common autosomal dominant disorders with a prevalence rate of 1:4,000 [6]. It occurs almost equally in all ethnic groups. Approximately 50 % of all cases of NF1 are due to new mutations. The NF1 gene mutations have variable expression—the clinical manifestations of NF1 range from subclinical to severe. However, it has nearly 100 % penetrance in adult individuals. Thus, an adult carrying the mutation will eventually exhibit some clinical feature of the disease. The NF1 gene is located on the long arm of chromosome 17 (17q) and of relatively large size (350,000 base pairs). This may explain the large rate of new mutation associated with this disorder. The gene product neurofibromin acts as a tumor-suppressor gene [7].

Several distinguishing clinical features of NF1 help to make the diagnosis of the condition (Fig. 1.2; Table 1.3).

There are a number of clinical issues associated with NF1 that are important from an orthopedic standpoint. These include tibial dysplasia and hemihypertrophy leading to leg length discrepancies and limb deformities, which are discussed later in the chapter.

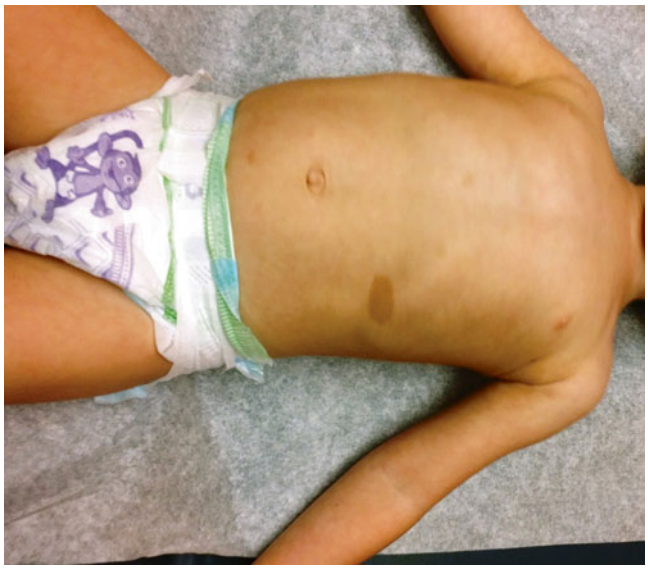


Fig. 1.2 A 3-year-old female patient with neurofibromatosis type 1 (NF1). Note the café au lait spots on the abdomen. Café au lait spots are the most easily noticeable feature of NF1

Table 1.3 Diagnostic criteria of neurofibromatosis type 1

More than six café au lait spots measuring at least 15 mm in adults and 5 mm in children
Two or more neurofibromas of any type or one plexiform neurofibroma
Freckling in the axillary or inguinal regions
Optic glioma
Two or more Lisch nodules (i.e., iris hamartomas)
A distinctive bony lesion, such as sphenoid wing dysplasia, or thinning of the cortex of a long bone with or without pseudarthrosis
A first-degree relative with NF-1, as suggested by these criteria

Another important manifestation of orthopedic interest is development of spinal deformities. Spinal deformities are the most common orthopedic manifestation of NF1. True incidence of spinal deformities in NF1 is not known. It is quoted as from 2 to 36 % in the literature [8, 9]. In the NF clinic at our institution, it is 23 % [6]. Traditionally, two forms of spinal deformities are described in NF1: dystrophic and non-dystrophic. Non-dystrophic curves are similar to idiopathic curves with some exceptions. The dystrophic changes in NF1 include rib penciling, vertebral scalloping, vertebral wedging, spindling of transverse processes, and enlarged intervertebral foramina.

The spinal deformities tend to develop early in the life; therefore all preadolescent children with NF1 should be evaluated by scoliosis screening or the Adam's forward-bend test to rule out the presence of a spinal deformity.



Fig. 1.3 A 17-year-old male patient with Charcot-Marie-Tooth disease. Note the cavus deformity of the foot bilaterally

Neuromuscular Disorders

Cerebral Palsy

Depending on the distribution and severity of their underlying spasticity, children with cerebral palsy can have a wide array of rotational, angular deformities and joint contractures. Excessive femoral anteversion is quite common in spastic diplegia and is associated with increased tone in the medial hamstrings and adductors. Femoral anteversion can also be accompanied by compensatory tibial external rotation in diplegic or by internal tibial rotation in a hemiplegic patient. Muscle tone imbalances also cause deformities of the hips, knees, ankles, and feet [10].

Charcot-Marie-Tooth Disease

Charcot-Marie-Tooth disease (CMT) is the most common of the seven types of hereditary motor and sensory neuropathies (HMSN). The two general types are demyelinating and axonal, but they are not distinguishable by clinical exam. As research has identified specific locations of different genetic abnormalities, the number of described subtypes has increased. Type IA CMT disease is the most common subtype and occurs via autosomal dominant transmission of a peripheral myelin protein 22 gene duplication on chromosome 17. The X-linked dominant form of disease is second most common and is attributed to a mutation of the connexin-32 gene. In general, demyelination of peripheral nerves and degeneration of the dorsal root ganglia and dorsal column of the spinal cord in the central nervous system occur, leading to progressive muscle atrophy, loss of proprioception, and loss of deep tendon reflexes. The most common deformity is worsening cavovarus foot posture with claw toes (Fig. 1.3) due to selective intrinsic and extrinsic (tibialis anterior

and peroneus brevis) muscle weakness and imbalance. The gastrocnemius-soleus complex is also weak and contracted, contributing to the cavovarus deformity and to the thin "stork leg." The hands can be affected and, less commonly, patients can develop scoliosis or hip dysplasia [11].

Arthrogryposis Multiplex Congenita

Arthrogryposis multiplex congenita (multiple congenital contractures) is actually a symptom complex of deformed, rigid, and sometimes dislocated joints that can occur as part of at least 300 different disorders. Amyoplasia is the classic and most common form of arthrogryposis that involves all four extremities. It is a nonprogressive syndrome characterized by extremities that appear cylindrical, fusiform, or cone shaped due to atrophied or absent muscles, decreased subcutaneous tissue, and diminished skin creases. In the lower limb, the foot deformities commonly seen include a rigid clubfoot or vertical talus. The knees may have either flexion or extension contractures. The hips are also very often involved, usually with flexion, abduction contractures, and hip dislocation noted in 15–30 % of patients with amyoplasia. These clinical findings result from decreased fetal movement. Fetal akinesia is due to failure of skeletal muscle development, often related to abnormal development of anterior horn cells in the spinal cord [12].

Skeletal Dysplasias

Children with various skeletal dysplasias present clinically with short stature and variable growth disturbances in the trunk and extremities. Specific deformities are typical for each type of dysplasia and are caused by specific structural abnormalities within the bones and cartilage. The underlying genetic mutation and mode of transmission, autosomal dominant for most but not all, are known for the majority of bone dysplasias. In the lower extremities, specific angular deformities and involved joints are characteristic for each diagnosis.

Benign and Malignant Tumors

Tumors, both benign and malignant, have the potential to cause limb deformities. Solitary lesions, even benign simple bone cysts, can be complicated by growth arrest due to pathologic fracture, juxtaposition to the physis, or injury during therapeutic curettage or injection [13]. The risk of growth disturbance is inherently higher with malignant tumors as they are much more aggressive. Treatment of malignancies may require a wide resection, with or without an amputation, often including the nearby physis that leaves the child with an increasing length discrepancy with growth.



Fig. 1.4 A 14-year-old male patient with juvenile rheumatoid arthritis (JRA). Note enlarged small joints of the hands. Image courtesy Dr. Hitesh Shah

Polyostotic diseases such as fibrous dysplasia and Ollier's disease are known to cause severe limb deformities. In fibrous dysplasia, expanding fibro-osseous tissue in the intramedullary canal causes repetitive microfractures, pain, and progressively bowed long bone deformities such as the classic shepherd's crook appearance of the femur [14]. Ollier's disease, a form of multiple enchondromatosis, produces islands of cartilage in the diaphyseal and metaphyseal regions that encroach upon the physis to cause inhibition of growth. Ollier's is often unilateral, so it is not only associated with angular deformities such as distal femoral varus, but also tends to cause severe lower limb length discrepancies [15].

Inflammatory Disorders

Children with inflammatory arthritis such as rheumatoid conditions have chronic joint inflammation that causes pain and muscle spasms, which eventually results in hip and knee flexion contractures, growth disturbances, and angular deformities. However, these patients will have typical features such as small joint involvements in upper extremity during earlier stage of the disease, which provides a clue to the diagnosis (Fig. 1.4). In children with hemophilic arthropathy, the repeated intra-articular bleeding causes synovitis that ultimately results in joint contractures secondary to cartilage degradation from both direct compression from the hypertrophic synovial pannus and proteolytic enzyme release [16]. Along with the intra-articular hemorrhage, there is hyperemia that often stimulates growth, resulting in angular deformity if there is asymmetric involvement of the adjacent physis or limb length discrepancy if symmetric involvement occurs [17].

Table 1.4 Common overgrowth and undergrowth syndromes

Common overgrowth syndromes		
Beckwith–Wiedemann syndrome	Macroglossia, macrosomia, midline abdominal wall defects, ear creases or ear pits, and neonatal hypoglycemia	Mutation of 11p15, leading to overactivity of the IGF-2 gene
Proteus syndrome	Progressively worsening macrodactyly, hemihypertrophy, thickening of the skin, lipomas, subcutaneous tumors, verrucae, epidermal nevus and macrocephaly	Somatic activating mutation in AKT1 kinase in a mosaic manner
Neurofibromatosis type 1	Café-au-lait spots, cutaneous neurofibroma, axillary freckling, optic glioma, Lisch nodules, vertebral scalloping, tibial dysplasia	Mutation of NF1 gene located on chromosome 17 leading to disruption of Ras signaling controlled by neurofibromin
Klippel-Trenaunay syndrome	Nevus flammeus (port-wine stain), venous and lymphatic malformations, and soft-tissue and bony hypertrophy of the affected limb	In some patients only: angiogenic factor VG5Q mutation. Translocation (5:11, 8:14)
Common undergrowth syndromes		
Russell-Silver syndrome	Short stature; small, triangular faces; disproportionately normal head circumference; clinodactyly	In 10 % patients: maternal uniparental disomy on chromosome 7
Cutis marmorata telangiectatica congenita	Generalized or localized reticulated cutaneous vascular mottling, macrocephaly, glaucoma, hemiatrophy, vascular anomalies	Unknown

Vascular Malformations

Various generalized and local vascular malformations are another group of disorders that can cause significant limb length inequality of up to 10 cm in addition to hypertrophy in girth and skin alterations [18].

Congenital

Hemihypertrophy and Hemiatrophy

There are no clear guidelines in literature as to what constitutes hemihypertrophy or hemiatrophy. It is also difficult to diagnose these conditions as there exists a normal variation between two sides of the body. However, when clinically significant differences are found, it is important to investigate further, as most of them would follow under one of the conditions discussed below (Table 1.4).

Idiopathic Hemihyperplasia

Also known as idiopathic hemihypertrophy (IH) or nonsyndromic hemihypertrophy, this is a condition of unknown origin (Fig. 1.5). Its importance lies in the fact that IH can be associated with embryonal tumors [19] such as Wilm's tumor, adrenal cell carcinoma, and hepatoblastoma in some patients.

In up to 80 % of patients with IH, no genetic abnormalities are found. When genetic abnormalities are found, these most commonly are uniparental paternal disomy of 11p15, loss of maternal methylation of *KCNQ1OT1*, or hypermethylation of H19. All these are mosaic abnormalities [20]. These abnormalities are very similar to genetic abnormalities found with Beckwith–Wiedemann syndrome. There is some evidence that the patients who have above-mentioned genetic abnormalities have a higher likelihood of developing embryonal tumors [21].



Fig. 1.5 A 9-year-old female with idiopathic hemihypertrophy of right lower extremity. The patient did not develop any neoplasm

Beckwith–Wiedemann Syndrome

Beckwith–Wiedemann syndrome [22, 23] is characterized by neonatal hypoglycemia, macroglossia, visceromegaly, and hemihypertrophy [24]. Most patients have a birth weight above the 90th percentile. Deletions and translocations in chromosome 11, near the locus of insulin-like growth factor

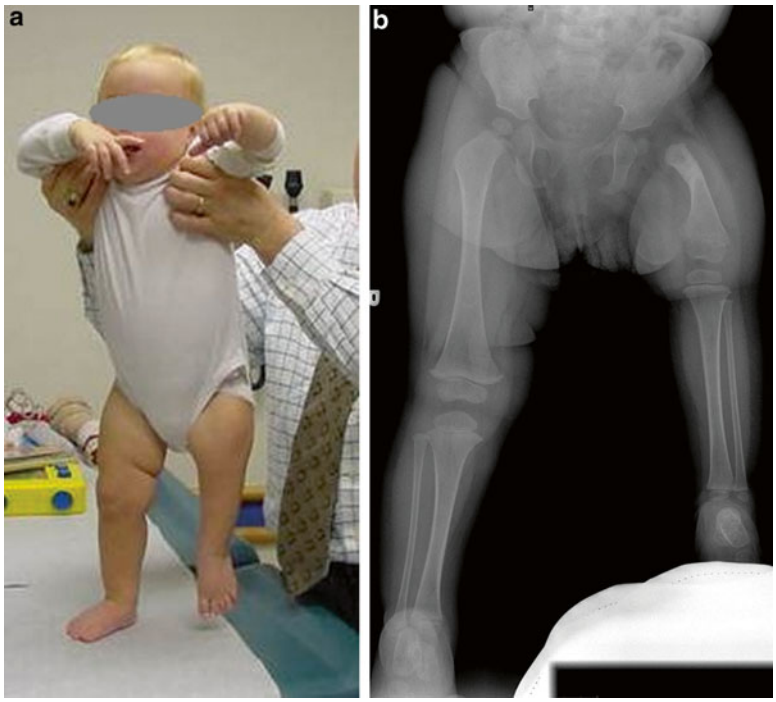


Fig. 1.6 (**a**, **b**) An 18-month-old male patient with focal femoral deficiency. Note short femur and apparent pseudarthrosis/absence of proximal femur on the left side

(IGF) II, can be identified in 70 % of the patients. As with IH, patients with Beckwith–Wiedemann syndrome are more prone to having embryonal tumors. The overall rate is 7–9 % in patients with Beckwith–Wiedemann syndrome, but those with the presence of hemihypertrophy are at substantially greater risk (24–27 %) [20, 24, 25].

As molecular techniques cannot always identify all the patients with tumor risk (especially in IH), serial abdominal ultrasound every 3 months up to age 7 and evaluation by a clinical geneticist is recommended [20] for both conditions.

Congenital Deficiency of the Long Bones

Longitudinal Tibial Deficiency

Commonly known as tibial hemimelia, this is a syndrome of partial to complete absence of the tibia at birth. In most cases the etiology is unknown. In a minority of cases, there is an autosomal dominant pattern. These specific entities include:

1. Warner syndrome: Tibial hemimelia–foot polydactyly–triphalangeal thumb syndrome
2. Tibial hemimelia-micromelia-trigonobrachycephaly syndrome
3. Tibial hemimelia diplopodia syndrome
4. Tibial hemimelia-split hand and foot syndrome

The heritable tibial hemimelia is usually bilateral and associated with anomalies of toes and hand. In a review by Schoenecker et al., 79 % of patients had associated congenital anomalies, with abnormalities of the hip, hand, or spine occurring alone or in various combinations [26].

Congenital Fibular Deficiency

Also known as fibular hemimelia, this condition includes partial or complete absence of fibula along with variable involvement of postaxial (lateral half) bones of the lower extremity. The exact etiology is not known and the cases are sporadic. Up to 15 % of patients have associated femoral anomalies [27].

When the fibula is partially absent/short, the spectrum of abnormalities include ipsilateral shortening of femur, hypoplastic lateral femoral condyle, cruciate ligament deficiency, ball and socket ankle, tarsal coalitions, and absent lateral rays. Children with hypoplastic or absent fibula often have associated tibial shortening with anteromedial bowing and marked equinovalgus deformity.

Congenital Femoral Deficiency

This is a condition of variable deficiency of the femur. In its mildest form the femur is short, while in severe case the proximal femur is absent and the distal femoral shaft creates a pseudoarticulation at the hip (Fig. 1.6). In some cases there is radiographic discontinuity noted between femoral neck and shaft. Similar to other long bone deficiencies, the etiology is unknown. However, there is an autosomal dominant inheritance noted in femoral hypoplasia–unusual facies syndrome [28].



Fig. 1.7 A newborn male with congenital dislocation of the right knee. The patient was initially treated by serial splinting and later with a quadriceps release

Congenital Dislocation of the Knee

The proper terminology for this condition is congenital knee hyperextension, with knee dislocation representing the most severe variety (Fig. 1.7). The incidence of congenital hyperextension is less than 1 per 1,000 [29]. Bilateral congenital dislocation of the knee (CDK) is usually associated with neuromuscular syndromes such as Larsen's or Beals' syndrome. Persistent hyperflexion of the hip is indicative of ipsilateral hip dislocation, which is commonly presented in neurologic conditions such as arthrogryposis and spinal dysraphism.

The most commonly accepted theory for etiology of CDK is abnormal fetal positioning. Subsequent akinesia (secondary to the neuromuscular conditions mentioned above) causes hyperextension leading to quadriceps atrophy and fibrosis.

Congenital Coxa Vara

Congenital coxa vara is a developmental abnormality characterized by a primary cartilaginous defect in the femoral neck with an abnormal decrease in the femoral neck–shaft angle, shortening of the femoral neck, relative overgrowth of the greater trochanter, and shortening of the affected lower limb. It is rare with an incidence estimated to be 1 in 25,000 live births. When present bilaterally, the condition is often associated with other autosomal dominant disorders such as cleidocranial dysostosis, spondylometaphyseal dysplasia, and metaphyseal dysostosis, Jansen type. Isolated congenital coxa vara is also believed to have a genetic etiology as it can occur in families and in twins. The coxa vara associated with congenital femoral deficiency is considered a separate entity as it does not share similar radiographic and clinical findings with congenital coxa vara.

Congenital Bowing of the Tibia

Anterolateral Bowing of the Tibia

"Congenital" Tibial Dysplasia

"Congenital" tibial dysplasia is associated with the development of pseudarthrosis of the tibia, which often develops subsequently and is usually not present at birth. As a result, the term dysplasia will be used instead of "congenital pseudarthrosis of the tibia." It is a very rare disorder affecting 1 in 140,000–190,000 [30–32].

Anterolateral tibial bowing and pseudarthrosis are associated with NF 1 in up to 55 % of the patients. Some authors have found an even higher association [32]. Fibrous dysplasia is present in up to 15 % of patients. The amniotic band syndrome (causing local constriction, eventually leading to pseudarthrosis) is also rarely associated with this type of tibial dysplasia [11].

Congenital Posteromedial Bowing of the Tibia

The deformity is present at birth and the tibial bowing is accompanied by a calcaneus foot and dorsiflexion contracture of the ankle. It is secondary to intrauterine malposition [33] (Fig. 1.8).

Anteromedial Bowing of the Tibia

This deformity of tibia is commonly associated with congenital fibular deficiency, as noted in the previous section.

Developmental Conditions

It has long been recognized that there are periods of physiologic genu varum and valgum in early childhood development. Newborns have genu varum which transitions to neutral alignment just before age 2, and then to maximum valgus around age 3 or 4. The valgus gradually decreases until the age of 7 or 8, when it becomes stable at the 5–7° of tibiofemoral valgus seen in adults [34].

Genu Varum

Physiologic Genu Varum

Physiologic genu varum is most common and has been defined radiographically as more than 10° of bilateral femoral-tibial varus after 18 months of age [35]. The entire lower limb appears bowed with both the distal femur and proximal tibia physes and epiphyses appearing normal and symmetric. These children often walk early, before 12 months of age, and a family history of bowlegs is common.



Fig. 1.8 (**a**, **b**) A 9-year-old male patient with congenital posteromedial tibial bowing. This patient does not have significant calcaneus deformity. Image courtesy Dr. Hitesh Shah

Tibia Vara/Blount's Disease

Tibia vara occurs when the apex of deformity occurs at the site of growth retardation at the posteromedial aspect of the proximal tibial physis and results in persistent or progressive bowing. There are two or three forms of the disease described by different authors based on age of onset, but all agree that the most common cause of pathologic genu varum is infantile tibia vara or Blount's disease (Fig. 1.9). In 1937, Blount characterized the deformity as an abrupt angulation just below the proximal tibial physis, an irregular physeal line, and a wedge-shaped epiphysis with a "beak" at the medial metaphysis. Apparent lateral subluxation of the proximal end of the tibia is often present [36].

It can sometimes be difficult to distinguish physiologic genu varum from early stages of infantile Blount's disease as they seem to occur along the same continuum of bilateral bowing. There may be a positive family history, but Blount's disease is considered a developmental disorder rather than congenital since the radiographic features are not seen in patients younger than 1 year [37]. Children with infantile Blount's are typically early walkers and often obese, placing enough force on the medial proximal tibial physis to impair normal longitudinal growth of the medial proximal tibial physis. Finite-element analysis of the knee has shown that a compressive force sufficient to retard physeal growth by the Heuter-Volkmann principle is produced on the medial tibial plateau of a 2-year-old in the 90th percentile for body weight and with a 20° varus deformity during single-limb stance [38].



Fig. 1.9 A 4-year-old male patient with Blount's disease, which is more common in patients of African American origin

A distinct form of tibia vara occurring later in childhood has been called late-onset or adolescent Blount's disease. Frequently, these patients are morbidly obese males in their early adolescence without any known insult to the proximal

Table 1.5 Common causes of genu varum

Physiologic
Blount's disease
Focal fibrocartilaginous dysplasia
Metabolic bone disease: rickets
Renal osteodystrophy
Posttraumatic
Post-infectious
Congenital deficiencies
Skeletal dysplasias: metaphyseal chondrodysplasia, spondyloepiphyseal dysplasia, multiple epiphyseal dysplasia, achondroplasia, pseudoachondroplasia
Neoplastic disease: fibrous dysplasia
Osteogenesis imperfecta

Table 1.6 Common causes of genu valgum

Unilateral	Posttraumatic physeal injury
	Proximal tibia metaphyseal fracture (Cozen)
	Neoplastic:
	Osteochondroma
	Focal fibrocartilaginous dysplasia
	Fibrous dysplasia
	Enchondromatosis (Ollier's)
Bilateral	Physiologic
	Metabolic: rickets
	Skeletal dysplasias:
	Chondroectodermal dysplasia (Ellis-van Creveld)
	Pseudoachondroplasia
	Morquio (MPS IV)
	Spondyloepiphyseal dysplasia
	Neoplastic: multiple hereditary exostoses

tibia to explain the medial physeal growth retardation around the knee. Similar to the infantile form, the etiology is not fully resolved, but the most popular theory is mechanical injury from excess weight, with or without preexisting mild varus deformity [39].

Proximal focal fibrocartilaginous dysplasia is a rare, idiopathic disorder with presentation similar to infantile Blount's. Clinically, the abrupt varus deformity is more distal than the knee joint line and corresponds to cortical sclerosis at the medial metaphyseal-diaphyseal junction of the proximal tibia at the insertion of the pes anserinus [40].

There are numerous other non-physiologic causes of genu varum (Table 1.5) that can be distinguished on radiographs.

Genu Valgum

Excessive physiologic genu valgum is frequently idiopathic. It is pathologic when the tibiofemoral angle increases after 7 years of age and walking becomes difficult as the knees rub together. Although it is less common than genu varum, there is still a wide range of causes [41] (Table 1.6, Fig. 1.10).



Fig. 1.10 A 6-year-old male status 1 year postproximal tibial metaphyseal fracture on the right who developed a genu valgum deformity. The patient was observed serially, and eventually the proximal tibia was remodeled completely

Acquired Sequelae and Complications

Residual Hip Deformities

Lower limb deformity or length discrepancy can occur as a result of several acquired conditions that are often evident in the child's history. A favorable outcome of pediatric hip conditions such as developmental dysplasia of the hip (DDH), SCFE, and Perthes disease is largely dependent on the congruency of the hip joint and avoidance of avascular necrosis (AVN). Residual hip deformity due to growth disturbances and AVN presents as coxa vara, coxa valgum, or short femoral neck depending on the location and extent of premature capital femoral epiphyseal closure as well as overgrowth of the greater trochanter that causes an abductor lurch.

Physeal Fractures

Fractures involving the physis are at risk for growth arrest due to direct injury to the chondrocytes and premature physeal closure. The Salter-Harris classification in combination with the mechanism of injury and degree of initial displacement can predict physeal disruption. In Salter-Harris type I and II injuries, the separation occurs primarily through the hypertrophic zone in the horizontal plane. Growth slows at

least temporarily, but the physis can appear histologically normal by 25 days [42]. In types III and IV, the fracture passes through all layers of the physis, creating a potential conduit between the bone of the metaphysis and epiphysis. Blood vessels invade this gap, allowing migration of osteoblasts to form a bony bar which tethers further longitudinal growth [42]. This can manifest as a leg length discrepancy or an angular deformity depending on the location and size of the physeal arrest or bony bar.

Non-physeal Fractures

Fractures do not necessarily have to directly involve the growth plate to result in a deformity. Long-bone fractures, especially femoral shaft fractures, may lead to deformity by malunion, overgrowth, or unrecognized ipsilateral physeal injury at the time of initial trauma. Valgus deformity can develop after a metaphyseal fracture of the proximal tibia without an associated fibular fracture (see Fig. 1.10). This was first described by Dr. Cozen, and may be due to soft-tissue interposition, stimulation of the medial physis, or tethering of the lateral physis by the intact fibula [43].

Musculoskeletal Infections

Sequelae of pediatric musculoskeletal infections, both osteomyelitis and septic arthritis, include limb length discrepancies and angular deformities from injury to the physis. Growth plate chondrocytes can be damaged by the infection itself or by the surgical debridement performed to treat it. Septic arthritis can also lead to joint contractures, articular cartilage destruction from proteolytic enzymes, and osteonecrosis from increased intra-articular pressure. The latter two complications may not cause deformity until several years after the acute septic episode.

Iatrogenic Causes

Iatrogenic injury to the growth plate can cause angular deformity, limb shortening, or overgrowth, depending on the extent of physeal damage. This can occur during closed treatment of a physeal fracture by repeated or overaggressive reduction attempts in the acute phase as well as manipulation after partial healing. During a closed manipulation of a postoperative joint contracture or casting for a congenital dislocation, such as the knee, excessive force can create a physeal separation. Any surgical procedure near or across the physis has a potential for permanent iatrogenic injury, including transphyseal hardware, improperly placed intramedullary nails, or instrumentation for guided growth. Drilling tunnels and securing grafts across growth plates for anterior cruciate ligament reconstructions in skeletally immature patients can also cause genu valgum, recurvatum, or overgrowth [44].

The purpose of this chapter was to provide an introduction to the vast number of etiologies of lower limb deformities and to organize them into a logical and clinically relevant framework. The remaining chapters in this book will delve further into the diagnosis, classification, and management of specific disorders.

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Clinical Evaluation Including Imaging

2

Joseph J. Gugenheim

Introduction

The majority of pediatric lower extremity deformities presenting for evaluation are normal physiologic angular and/or rotational conditions that resolve spontaneously without expensive and unnecessary treatment, diagnostic imaging, or follow-up. However, limb length discrepancy and asymmetric and/or severe angular deformities often do require imaging and follow-up. Thus, the clinician must distinguish between the physiologic angular and rotational conditions, which require only reassurance to the family, and deformity secondary to pathologic conditions, which requires follow-up and possible intervention. The Pediatric Orthopedic Society has defined normal as that which occurs within two standard deviations of the mean [1]. Angular or rotational conditions within this range are considered *variations*; those outside this range are considered *deformities* [2]. The common use of the term *deformity* to include both variations and deformities is used in this chapter. Accurate assessment of a child's lower extremities requires quantifying the deformity and knowing the mean value and standard deviations for the child's age. Fortunately, the history and physical examination and sometimes imaging are usually all that are needed to determine the diagnosis, prognosis, and recommendations for the family.

History

First, the clinician must determine why the child is brought in for evaluation. Is it because the referring physician wants to determine the diagnosis and need for treatment? If so, the family may not require much counseling and reassurance.

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Do the parents or grandparents want to "do something before it is too late," or do they want treatment, even if it is not indicated, because physicians prescribed treatment in the past to family members of previous generations [3]? These situations require a clear explanation with great patience why treatment is not necessary. The author, who has been practicing for almost 40 years, likes to humorously explain that when he began his practice, we physicians treated these conditions with special shoes, braces, exercises, shoe modifications, and inserts and had 100 % excellent results. We congratulated ourselves for being brilliant physicians for producing such excellent results until we learned that the children straightened up, not because of us, but in spite of us! By explaining that the evaluation of any treatment in the practice of medicine requires comparing treated to untreated patients and that such evaluations show that treatment in many conditions does not affect the outcome, accelerate the natural resolution, or prevent later disability, the physician can explain to the parents that treatment and further testing are not indicated [4].

Are the parents worried about cosmesis? Are they worried about future athletic performance? Are they worried about damage to the feet, knees, hips, or spine? Are they worried that their bowlegged toddler may grow up to resemble a bowlegged elderly relative with painful unicompartmental knee arthritis?

Is the child coming to the physician because of pain? If so, an etiology besides physiologic angulation/rotational variations should be sought.

When evaluating a lower extremity deformity or physiologic variant, the clinician should include questions regarding the relevant factors as described in Table 2.1.

The clinician should determine at what age the deformity was first noticed. Intoeing which does not appear until approximately age 2 years is usually due to resolution of the physiologic external rotation contracture of the infant hip, unmasking the persistent tibial torsion and/or proximal femoral antetorsion.

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Does it seem to be getting better, worse, or remaining the same? For example, bowing of the lower extremities is a normal consequence of the in utero position and will resolve spontaneously (Fig. 2.1); the gradual improvement in the bowing helps establish the diagnosis and rules out Blount disease or metabolic bone disease, two conditions that require a more extensive work-up and treatment.

Is there any history of fractures, lower extremity injuries, bone or soft-tissue infections, or previous surgery? The development of an angular deformity or leg length discrepancy following a fracture or infection may indicate a partial or complete injury to the growth plate.

Is there any family history of orthopedic, neuromuscular, renal, or endocrine disorders? Is there any family history of short stature? A family history of hypophosphatemic rickets, Charcot–Marie–Tooth disease, or kidney failure with renal rickets should alert the clinician to conditions that may be inherited.

Table 2.1 Relevant aspects of history (besides the onset, duration, progression, and prior treatment of the lower limb deformity)

Age
Gender
Number of siblings
Birth order
Length of pregnancy
Obstetric presentation
Presence or absence of difficulty with delivery
Birth weight
As active or less active than siblings in utero
Developmental motor milestones
Family history of orthopedic, neuromuscular, renal, or endocrine disorders

Physical Examination

Although many children who present for evaluation have an obvious diagnosis, one must always perform an examination that includes at least the vital signs (height, weight, body mass index (BMI)), upper extremities, spine and trunk, neurological, gait, and lower extremities. Although the gait and lower extremity exam will be the most detailed, the other components are necessary and can be performed quite rapidly.

Vital Signs

Height and weight should be measured at the office visit instead of asking the family to furnish this information because parents often are quite inaccurate with their child's stated height and weight [5]. The BMI is a proxy for adiposity and can be calculated from the formula

$$\mathrm{BMI} = \frac{\mathrm{Weight}\left(\mathrm{kilograms}\right)}{\mathrm{Height}^2\left(\mathrm{meters}\right)} = \frac{\mathrm{Weight}\left(\mathrm{pounds}\right)}{\mathrm{Height}^2\left(\mathrm{cm}\right)} \times 4536$$

The percentile for age-related Z score should be determined from charts available from the Centers for Disease Control (http://www.cdc.gov/growthcharts/clinical_charts.htm).

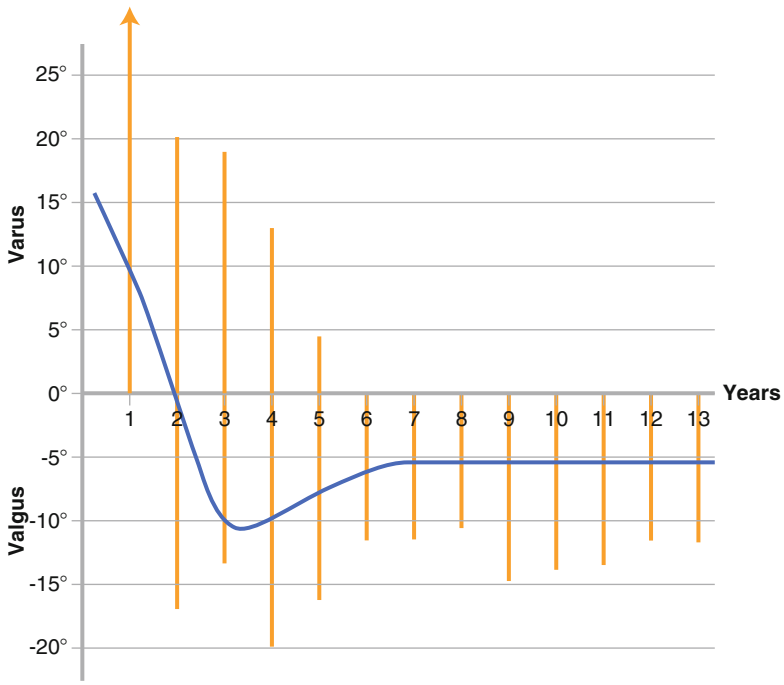


Fig. 2.1 The development of the tibiofemoral angle in children during growth; the standard deviation is ±8°. Adapted with permission from Zaleske DJ. Metabolic and endocrine disorders. In: Morrissy RT, Weinstein SL, editors. Lovell and Winter's Pediatric orthopedics, 4th edition. Philadelphia: Lippincott-Raven; 1996 [58]

Very large age-related *Z* scores may indicate a condition associated with obesity such as Blount disease or Prader–Willi syndrome. Very low *Z* scores may indicate malnutrition, malabsorption, chronic metabolic disease, or skeletal dysplasia.

Upper Extremity

An abnormal carrying angle of the elbow, especially if symmetric, may indicate a skeletal dysplasia. Joint laxity with excessive motion, such as hyperextension of the elbows or interphalangeal joints, may indicate a connective tissue disorder. Palpable metaphyseal enlargements may indicate a rachitic disorder or multiple hereditary exostoses.

Spine and Trunk

The rib cage should be palpated for a rachitic rosary: swellings at the costochondral junction are indicative of a rachitic disorder. Screening for scoliosis is performed by checking for thoracic and/or lumbar asymmetry with forward bending.

Neurologic

A quick but quite sensitive screening battery for the detection of neurological disease affecting the lower extremity can be done in children and adolescents of walking age by having them walk on their heels in dorsiflexion, on their toes (metatarsal heads) in equinus, and hop on one leg at a time. To perform these maneuvers, the child utilizes quadriceps (L2, L3, L4), hip extensors (L5, S1, S2), hip abductors (L5, S1), ankle plantar flexors (L5, S1, S2), and ankle dorsiflexors (L4, L5). If there is a high level of suspicion for the presence of a neurological abnormality, a more detailed exam must be done, including searching for pathologic primitive reflexes.

Gait

The examiner should watch the child walk. Sometimes children walk in a certain pattern as instructed by their parents when aware that the physician is watching. Thus, it is helpful to watch the child walk from the waiting area to the exam room when he or she is unaware of being examined. One should watch the child walk both with shoes and barefoot because shoes may hide dynamic conditions such as searching toes or dynamic forefoot adduction, two conditions attributed to contraction of the abductor hallucis creating a concave medial border of the foot during gait.

Foot Progression Angle

The foot progression angle is the angle between the axis of the foot and the line of progression, with intoeing expressed as negative values and out-toeing expressed as positive values [2, 6]. A study of gait in 160 children (up to 14 years of age) concluded that the normal angle of gait was approximately 10° external at all ages with a slight decrease after age 7 [7] (Fig. 2.2). The mean foot progression angle is a positive value (external rotation) throughout childhood and adulthood. However, negative values (internal rotation) are considered normal (within two standard deviations) throughout childhood and early teens [2]. Etiologies contributing to an internal rotation gait include excessive acetabular anteversion, excessive femoral anteversion, internal tibial torsion, metatarsus adductus (MTA), and searching toe. Etiologies contributing to an external rotation gait include acetabular retroversion, physiologic external rotation contracture of the hips, femoral retroversion, external tibial torsion, and positional calcaneal valgus. These conditions may be additive or may compensate for each other [8].

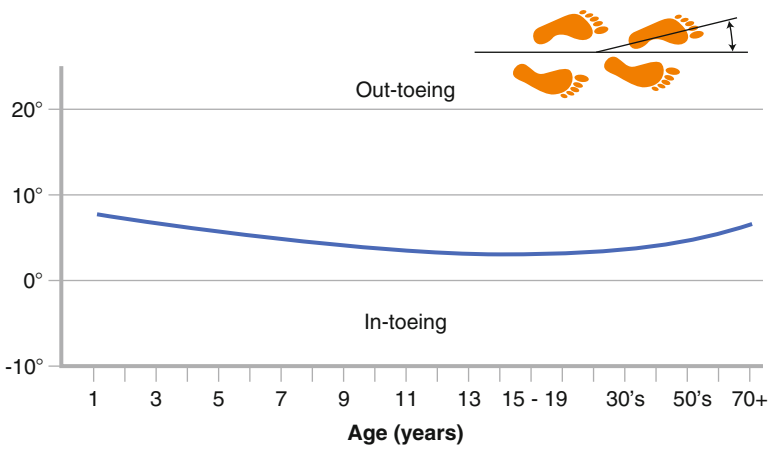


Fig. 2.2 The foot progression angle changes with age. The normal range is very broad. Adapted from [4]

Short Leg Gait

A child with a limb length inequality may walk with the foot and ankle in equinus or with an abductor lurch on the short side during stance phase of gait. Other compensatory mechanisms for a short lower extremity include flexing the contralateral knee and/or dropping the ipsilateral hemipelvis.

Equinus

Unilateral equinus is often seen in patients with hemiparetic cerebral palsy, some unilateral foot deformities, and limb length inequality. An equinus position in stance phase of gait is the most common compensating mechanism for a limb length inequality. Bilateral equinus may indicate idiopathic toe walking or neuromuscular disease, such as cerebral palsy or muscular dystrophy.

Trendelenburg Sign

The child should stand on one foot while the clinician observes for the pelvis dropping toward the opposite side. Normally, the child should not allow the pelvis to drop for at least 15 s. A positive sign with the pelvis dropping indicates abnormal ipsilateral femoral pelvis anatomy (such as acetabular dysplasia, dislocated or subluxated femoral head, proximal femoral deformity) or abductor weakness.

One must compare the gait appearance to the findings on the exam table. For example, forefoot adduction during gait but not in the nonweight-bearing position is frequently due to the normal phasic activity of the toe flexors and tibialis posterior muscles in stance phase; these muscles insert medial to the midline of the foot and may act as forefoot adductors during gait. The great toe may abduct during stance phase of gait. This condition, called wandering or atavistic toe, is attributed to the phasic activity of the abductor hallucis muscle [4, 8].

Lower Extremities

Supine Exam

A general assessment of the lower extremities in the supine position on the exam table includes evaluation of the limb lengths, as seen by the levels of the plantar aspects of both feet and the measurement of the distance from the anterior superior iliac spine to the medial malleoli. Any asymmetry of thigh circumference, leg circumference, or foot length should also be noted. If a discrepancy in limb lengths is noted, the patient should be examined with blocks underneath the short leg to determine when the pelvis is level. This method has been shown to be the most reliable clinical measurement for leg length discrepancy, although the studies were done in adults [9, 10].

Varus (bow legs) and valgus (knock knees) angulations at the knee are measured with a goniometer on the anterior aspect of the extremity with the patellas forward. Measuring the distance between the knees or medial malleoli to quantify varus or valgus is less reliable; there may be variability due to adiposity, size of the patient's legs or thighs, and limb torsion. Bowing of the lower extremities is normal until age 2 years. Persistent varus after age 2 years or excessive valgus may be a manifestation of a traumatic, congenital, developmental, or metabolic disease [11–16] (Table 2.2).

Although radiographs can distinguish between normal physiologic bowing of infancy and rare conditions requiring intervention, routine radiographic screening and referral are not cost effective and expose children to unnecessary radiation [11, 17]. The *cover-up test* is a useful screening to assess

Table 2.2 Etiologies of lower extremity deformity

Varus
Traumatic
Proximal tibial physeal injury (may also be valgus)
Congenital
Tibial hemimelia
Developmental
Achondroplasia
Campomelic dysplasia
Congenital pseudarthrosis of tibia
Hypochondroplasia
Metaphyseal chondrodysplasia (Schmid, Jansen, Spahr types)
Neurofibromatosis
Ollier disease (multiple enchondromatosis) (may also be valgus)
Osteogenesis imperfecta (may also be valgus)
Multiple epiphyseal dysplasia (may also be valgus)
Multiple hereditary exostosis (may also be valgus)
Spondyloepiphyseal dysplasia
Thanatophoric dysplasia
Tibia vara (Blount disease)
Metabolic
Vitamin D-deficient rickets
Hypophosphatemic rickets
Valgus
Traumatic
Proximal tibial physeal injury (may also be varus)
Proximal tibial metaphyseal fracture sequela (Cozen phenomenon)
Congenital
Congenital deficiency of femur
Posteromedial bowing of tibia
Fibular hemimelia
Developmental
Ellis-van Creveld dysplasia
Metatropic dysplasia
Morquio syndrome
Multiple hereditary exostosis
Multiple epiphyseal dysplasia (may also be varus)
Metabolic
Chronic renal failure

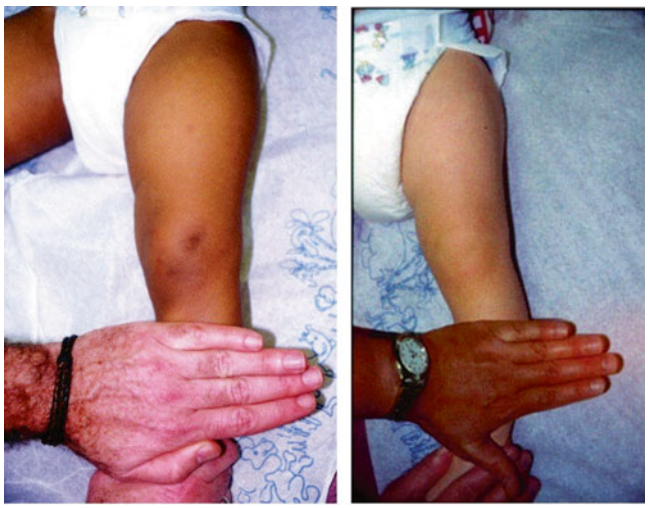


Fig. 2.3 Cover-up test of the proximal left tibia. (**a**) A varus or neutral alignment is a positive test. (**b**) A valgus alignment is a negative test. Courtesy of Jon R. Davids, M.D.

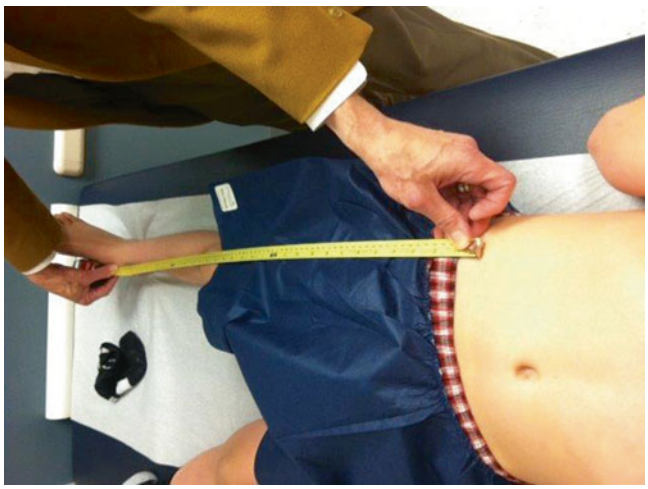


Fig. 2.4 The lower extremity length is measured from the anterior superior iliac spine to the medial malleolus with a metal tape measure

the alignment of the upper part of the lower leg in children presenting with bow legs between the ages of 1 and 3 years [17]. The lower extremity is positioned with the patella forward and the lower leg and foot are covered by the examiner's hand. A positive test (bowing in the upper tibia) or neutral test (a straight thigh–upper leg axis) is an indication for radiographic evaluation. A negative test (slight valgus at the upper tibia) indicates physiologic bowing. A positive cover-up test has a high sensitivity; a negative cover-up test has a high specificity and negative predictive value [17] (Fig. 2.3).

Limb length is measured from the anterior superior iliac spine to the medial malleolus with a tape measure (Fig. 2.4). If a limb length discrepancy is found, a cooperative child should stand with blocks underneath the short extremity to determine what size lift will level the pelvis. Older children and adolescents should be asked to report what amount of elevation for the short extremity makes them feel level and well corrected. Patients with long-standing limb length discrepancies usually report that they feel well corrected when the blocks under the foot undercompensate for the discrepancy because they have become partially accustomed to the discrepancy. Standing on blocks is more accurate (greater reliability) and also incorporates the foot and pelvis, unlike the supine tape measure technique [18].

The lower extremity may appear short if an ipsilateral hip or knee flexion contracture or an adduction contracture of the hip is present while standing; likewise the extremity may appear long if an ipsilateral hip abduction contracture is present. Any difference in the size of the foot should also be noted. Foot size discrepancies are seen in hemihypertrophy, neurological disease, and vascular disease.

To determine if a discrepancy is due to a femoral or tibial difference, the child should sit on the exam table with the hips and knees flexed 90°, the patellas forward, and feet unsupported. If there is a femoral discrepancy, the knees will appear to be at a different level when viewed from above; if a tibial discrepancy is present, the feet will be at different levels when viewed from the front. In many cases, there is a discrepancy in both segments.

Prone Exam

The prone exam (with measurement of hip rotation, thigh–foot angle, and foot alignment) is an excellent qualitative assessment of rotational conditions affecting the lower extremity.

Femoral Version and Antetorsion

Hip rotation in the prone position is the net result of soft-tissue rotation contracture, acetabular version, and femoral version (anteversion or retroversion). Axial rotation of the acetabulum and femur within two standard deviations of the age-related mean value is considered normal and is termed version; values outside two standard deviations are termed torsion [4].

Hip rotation should be evaluated with the child prone and the hip extended because the hip functions largely in extension during normal gait. Also, hip flexion in the supine position relaxes the anterior hip capsule, causing an inaccurate assessment of rotation [16]. The child should be positioned prone, with one hand lightly on the pelvic area to detect what degree of rotation rocks the pelvis off the table. The knee is flexed 90°. The leg serves as an indicator of rotation, measuring its angulation from vertical [6] (Fig. 2.5). Normal age-dependent values for internal rotation and external rotation have been published [2, 4, 6] (Fig. 2.6).

The normal hip arc of motion, the sum of internal and external rotations, is approximately 80°–100° [6, 19], with greater external rotation than internal rotation until the age of 3–5 years [4, 7, 20]. Due to the in utero position, infants

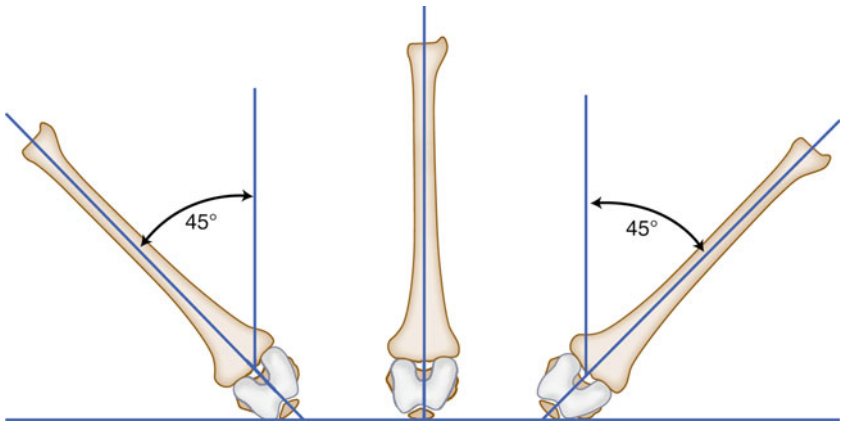


Fig. 2.5 Assessment of hip rotation with 45° internal and 45° external rotation. With kind permission from Springer Science+Business Media: Paley D. Principles of deformity correction. Berlin, Heidelberg; 2002 [23]

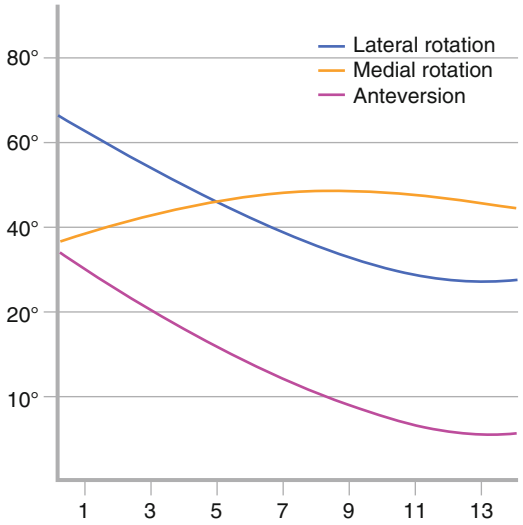


Fig. 2.6 Values for hip rotation and anteversion, according to age (*horizontal axis*). Adapted from [4]

often have an external rotation contracture of the soft tissues around the hip at birth that gradually resolves by age 18 months. This soft-tissue contracture can mask the underlying femoral antetorsion. This physiologic soft-tissue contracture is at its maximum value at birth and is the usual etiology for external rotation exceeding internal rotation until age 18–24 months [20]. Femoral antetorsion (abnormally high value for anteversion) manifests as an increase in hip internal rotation and a decrease in external rotation without affecting the total rotational arc. It is the most common cause of intoeing in early childhood, tends to occur more in girls, and is usually symmetric [8]. As the physiologic soft-tissue external rotation contracture resolves, internal rotation of the hip increases [8]. For this reason, parents often report that their child became more pigeon toed or walked with more intoeing at age 18–24 months.

Other etiologies of excessive external rotation include acetabular retroversion, as seen in Down syndrome [21], and femoral retroversion, as seen in slipped capital femoral epiphysis (SCFE), coxa vara, and proximal focal femoral deficiency (PFFD). Jacquemier et al. defined normal age-related values of acetabular version in a study of 143 children between ages 1 and 15 years [22]. Because the significance of abnormal acetabular version in infants and children is unknown at present, radiographic studies are rarely done to measure acetabular version; therefore, the true prevalence of acetabular retroversion in children is largely unknown. Coxa vara and PFFD usually exhibit a decreased arc of rotation (less than 90°) on the prone exam and are detectable on the supine exam as decreased hip abduction or, in unilateral cases, a limb length discrepancy.

A severe valgus deformity of the proximal tibia will cause an apparent increase in external rotation and decrease in internal rotation without affecting the total rotational arc, whereas a severe varus deformity will cause an apparent increase in internal rotation and decrease in external rotation (Fig. 2.7), but this is rarely significant in an evaluation of pediatric patients [23].

In conclusion, in the vast majority of cases of infants with more external rotation than internal rotation in the prone position with the gradual diminution of the discrepancy, the etiology is a physiologic external rotation contracture of infancy (PERCI) especially between birth and age 18 months. Increased internal rotation compared to external rotation is often indicative of femoral antetorsion.

In most cases, a qualitative clinical examination, based on comparing hip internal and external rotations, is sufficient without a quantitative imaging study. Imaging to quantify femoral version is indicated only if hip rotation is significantly asymmetrical, operative correction is planned, or version must be measured to determine the cause of failure of prior surgical procedure around the hip [4]. Most authors conclude that femoral anteversion is not harmful [8, 16, 24], although it may require surgical correction when associated with excessive external tibial torsion [4, 19], abnormal acetabular version [19, 25], neuromuscular disease [26], or developmental dysplasia of the hip (DDH) [25, 26].

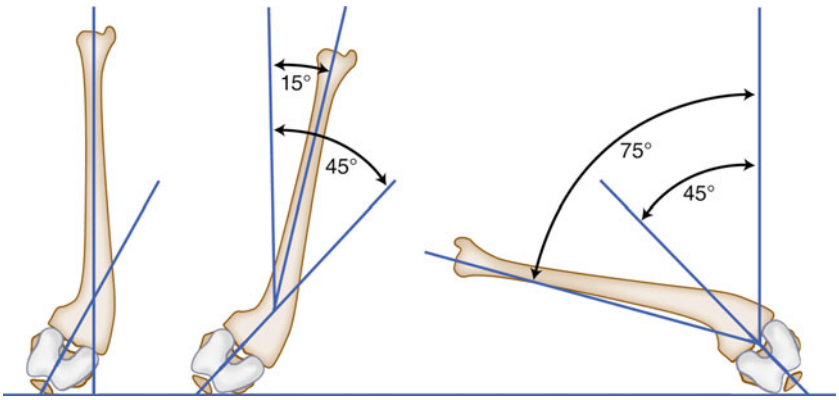


Fig. 2.7 A 30° valgus deformity of the proximal tibia results in an incorrect assessment of 75° external rotation (instead of 45°) and 15° internal rotation (instead of 45°). Adapted with permission from Lippincott Williams and Wilkins/Wolters Kluwer Health; Jacquemier M, Jouve JL, Bollini G, Panuel M, Migliani R. Acetabular anteversion in children. J Pediatr Orthop. 1992;12(3): 373–5 [22]

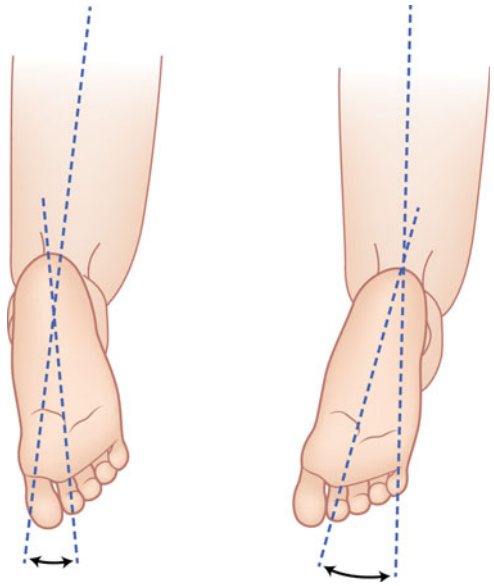


Fig. 2.8 The thigh–foot angle (TFA) assesses tibial torsion. (**a**) External tibial torsion. (**b**) Internal tibial torsion. Adapted with kind permission from Springer Science+Business Media: Paley D. Principles of deformity correction. Berlin, Heidelberg; 2002 [23]

Tibial Torsion

Tibial version is normal in children and, by definition, torsion is abnormal [2, 4]. However, the term *tibial torsion* will be used here to include both normal and abnormal values because of its common usage. The thigh–foot angle (TFA) is the angle formed by the longitudinal axis of the thigh with the longitudinal axis of the hindfoot. The TFA is measured in the prone position with the hip in neutral rotation, the knee in 90° flexion, and the ankle and foot in neutral dorsiflexion (Fig. 2.8). Although this assessment of the TFA is a qualitative diagnostic test for tibial torsion, a goniometer may be used for quantifying this angle and to compare on subsequent exams. External tibial torsion is diagnosed and quantified with a positive value if the foot is externally rotated on the leg; internal tibial torsion is diagnosed and quantified with a negative value if the foot is internally rotated on the leg.

The TFA averages 5° internal (range 30° to +20°) in infants and averages 10° external (range 5° to +30°) by the age of 8 years [8]. Katz reported internal tibial torsion on the left side more frequently than the right in a series of 54 females and 45 males [27]. Normal age-related values have been published [2, 4]. Staheli et al. assessed the normal range and interobserver error. They found that the normal range, defined as within two standard deviations, is quite large, with a standard deviation of 4.9°–8.9°. They concluded that values from 5° to 30° were within the range of normal during childhood [2].

Tibial torsion can also be diagnosed and quantified by determining the transmalleolar axis (TMA). With the child in the sitting position with the hip flexed and patella facing forward, the angle between the TMA and the coronal plane is assessed [28, 29]. Measurement can also be done in the prone position by locating the transmalleolar axis, drawing a line perpendicular to this axis, and then measuring the angle this line forms with the long axis of the thigh. The result obtained by the TMA method yields a value a few degrees greater than the TFA method, but the TFA is easier to measure [2]. While the TMA method measures only the tibial and fibular rotations, the TFA method is also affected by hindfoot abnormalities. More accurate quantification by diagnostic imaging is rarely indicated unless corrective osteotomies are being planned. Since internal tibial torsion is a normal condition in infants and children and normally resolves without treatment, corrective osteotomies are rarely indicated.

Axial Foot Deformities

The alignment of the foot as a possible cause of intoeing or out-toeing can also be assessed with the child in the prone position. The examiner should note whether the medial and lateral borders of the foot are straight (Fig. 2.9). The heel bisector line, a line through the midline axis of the hindfoot and forefoot, passes through the second web space in a normal foot. Originally described by taking a photocopy of the foot, a method of making a permanent paper document, the child is held in a weight-bearing position on the photocopy machine.

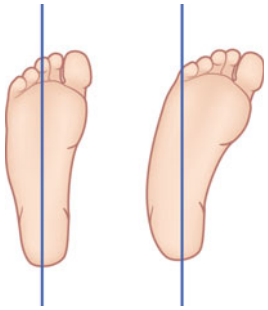


Fig. 2.9 The heel bisector line passes through the second web space in the normal foot (10A) and lateral to the second web space in metatarsus adductus. Adapted with permission from Smith JT, Bleck EE, Gamble JG, Rinsky LA, Pena T. Simple method of documenting metatarsus adductus. J Pediatr Orthop. 1991;11(5):679–80 [30]

The method can also be used while examining the child prone [30]. If the heel bisector line passes lateral to the second web space, the forefoot is adducted (MTA). If the line passes medial to the second web space, valgus is present. A concave medial border of the midfoot also indicates MTA. If MTA is present, the hindfoot can then be held with one hand while the examiner's other hand abducts the forefoot to determine whether the MTA is rigid or supple. A normal foot should be flexible and exhibit slight forefoot abduction with gentle manipulation.

Diagnostic Imaging

Whether to do imaging studies after a history and physical exam often presents a dilemma. The expense and radiation exposure with imaging should be avoided if the diagnosis is obvious from the history and physical exam and the imaging will not affect the decision making. Any limb length discrepancy, asymmetric angular deformity, or angulation outside the range of normal for age requires a full-length standing anterior–posterior image of both extremities to quantify the angular deformities and/or limb length discrepancy. Davids et al. noted that routine radiographic screening of children with bowed legs is not cost effective and exposes them to unnecessary radiation and recommended using the cover-up test to screen children between ages 1 and 3 years. Children with a negative cover-up test do not require radiography [17]. Guidelines for performing imaging studies for rotational conditions are less clear. Children with rotational components more than two standard deviations outside the mean, short stature (less than twenty-fifth percentile), asymmetric hip abduction or rotation, a positive Trendelenburg sign, marked limb asymmetry in length or alignment, or discomfort during gentle range of motion of the lower extremity should be considered for diagnostic imaging. A pelvic radiograph including a lateral view may be needed to rule out disorders such as hip dysplasia and SCFE in a child presenting with a gait abnormality and pertinent history and clinical exam [8].

Assessment of Limb Length

Several techniques have been developed for the measurement of limb length discrepancies, including orthoroentgenograms [31], slit scanography [32], standing teleroentgenograms [23, 33], scanograms [34], computerized tomography (CT) scanogram [35], and ultrasound [36]. The ideal method should be accurate, readily available in the physician's office, and technically easy. Its cost and radiation dosage should be low. Its measurements should exhibit good reproducibility (Table 2.3).

A single-exposure standing teleroentgenogram taken with the patella straight forward with a distance of 10 ft between the beam source and the film offers the best combination of benefits and fewest shortcomings (Fig. 2.10). It can easily be performed. Radiation exposure is greater than the CT scanogram technique [35] but less than the three-view scanogram [33]. With the advent of digital radiography and picture archiving and communication systems (PACS), the image can be manipulated to minimize radiation exposure and film storage is not a problem. By using a large distance between the beam source and the film, parallax and magnification (of up to 5 %) are introduced. Measurements can be made directly with a ruler or with computer software [33]. The image is made in the weight-bearing position. Angular deformities and bone pathology that may contribute to angular deformity and length discrepancy such as osteochondromas, enchondromas, or malunited fractures are detectable (see Table 2.2). The contribution of each segment (pelvis, femur, tibia, and foot) can also be quantified. Intraobserver and interobserver measurement reliability is excellent with a teleroentgenogram [34].

A standardized radiographic technique avoids distortion and magnification due to divergence of the X-ray beam and is reproducible so that serial radiographs can be compared. A cassette with a variable grid to include the hip, knee, and ankle is used. The cassette measures 132.5×38.5 cm for larger patients and 94.5×38.5 cm for smaller children. The beam is centered at the knee with the beam source at least 10 ft from the cassette. Correct patient positioning requires patellas straight forward for the anteroposterior (AP) view and 90° orthogonal to the AP position for the lateral view. The knees should be in full extension, with the body weight distributed equally on both feet. If there is a leg length discrepancy, a block is placed under the shorter extremity to level the pelvis and to keep the knee extended with the weight equally distributed on both feet. In severely obese children or in the presence of severe angular deformity,

Table 2.3 Comparison of methods for assessing leg length discrepancy[a]

Methods	Reliability[b]	Accuracy[b]	Magnification	Approximate radiation exposure (mrads)	Approximate charges (US dollars)	Radiographic deformity analysis	Incorporation of height of the foot and pelvis	Typical availability in the USA	Weight bearing
Clinical									
Supine tape measure—"Real" (ASAS to malleolus)	+	+	None	None	Office visit	Not applicable	No	Yes	No
Supine tape measure—"Apparent" (umbilicus to malleolus)	+	+	None	None	Office visit	Not applicable	Partial	Yes	No
Standing blocks	++	+	None	None	Office visit	Not applicable	Yes	Yes	Yes
Imaging									
Teleoroentgenogram	++++	+++	~5%	42	$95[c]	Yes	Yes	Varies	Yes
Orthoroentgenogram	+++	+++	Minimal	200	$110[c]	No	No	Varies	No
Scanogram	++++	+++	Minimal	200	$110[c]	No	No	Varies	No
Computed radiography	++++	+++	Varies with technique (scanogram vs. teleoroentgenogram)	Varies with technique, less exposure than standard radiography	$137[d]	Varies with technique (scanogram vs. teleoroentgenogram)	Varies with technique	Varies	Varies with technique (scanogram vs. teleoroentgenogram)
Microdose digital radiography	+++	++++	None	2	$75[c]	Yes	Yes	No	Yes
Ultrasound	+++	++	None	None	Not reported	No	No	No	Yes
CT scan (digital localization image)	++++	++++	Minimal	60	$60[c]	Minimal	None	Varies	No
MRI	++++	+++	Minimal	None	Not reported	Not reported	Not reported	No	No

[a]Adapted with kind permission from Springer Science+Business Media: Sabharwal S, Kumar A. Methods for assessing leg length discrepancy. Clin Orthop Relat Res. 2008;466 (12)2910–22
[b]Please see text for detailed analysis. For simplicity, the increasing number of "+" signs indicates greater degree of reliability/accuracy
[c]Machen MS, Stevens PM. Should full-length standing anteroposterior radiographs replace the scanogram for measurement of limb length discrepancy? J Pediatr Orthop B. 2005;14:30–37
[d]Sabharwal S, Zhao C, McKeon JJ, McClemens E, Edgar M, Behrens F. Computed radiographic measurement of limb-length discrepancy. Full-length standing anteroposterior radiograph compared with scanogram. J Bone Joint Surg Am. 2006;88:2243–2251 [33]

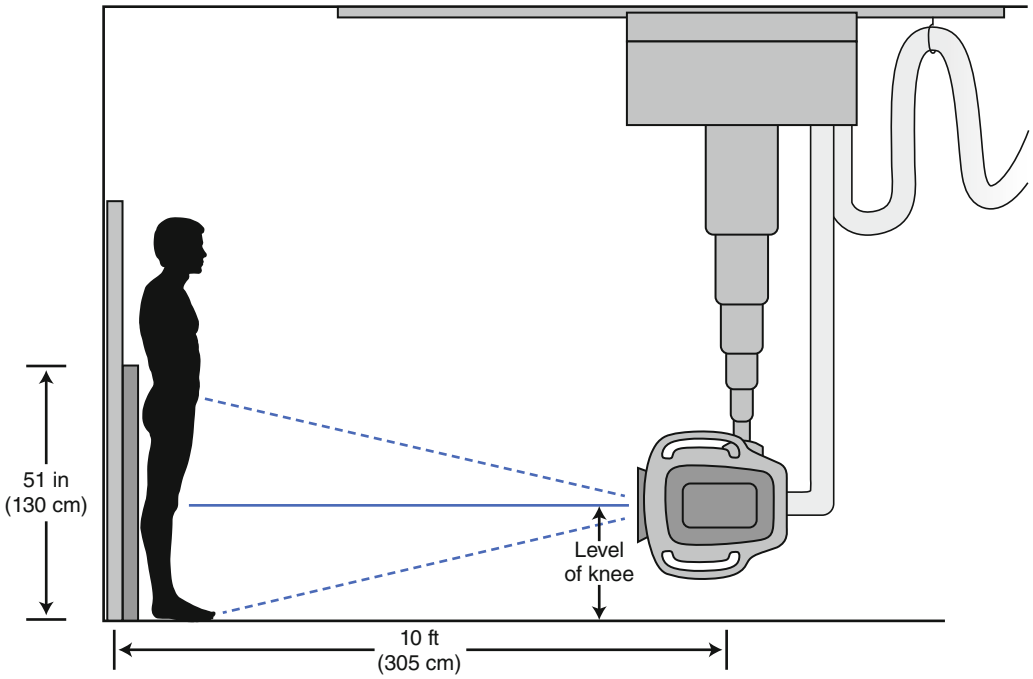


Fig. 2.10 A single-exposure standing full-length radiograph with patella forward is used to evaluate limb length and deformity. Adapted with kind permission from Springer Science+Business Media: Paley D. Principles of deformity correction. Berlin, Heidelberg, 2002 [23]

it may be necessary to perform separate AP radiographs of the two lower extremities. A magnification standard, such as a 30 mm ball bearing, can be affixed to the extremity at the main site of interest or at the knee when assessing leg length. The ball bearing should be positioned at the level of the bone, the same distance from the cassette as the bone. This technique quantifies magnification, which is usually about 5 % [33]. The ball bearing or another magnification marker must be affixed to the extremity at the level of the bone, not directly on the cassette (Fig. 2.11a, b). Placing a ruler with radiopaque graduations on the cassette may not accurately quantify magnification because the ruler is closer to the film than the bone.

Slit scanography and CT scanograms also exhibit reasonable accuracy and reliability [18], but they require special technology which may not be available in most clinicians' office setting. CT scanograms have the lowest radiation exposure compared to other radiographic methods and can be performed in the presence of flexion contractures. The CT scanogram can be performed very rapidly and magnification error is not an issue. However, since the scan is usually performed in a nonweight-bearing position, it is impossible to measure the contribution of the foot to the leg length discrepancy. Furthermore, it is difficult to manipulate the CT image for planning of deformity correction, although advances in digital imaging software may eliminate this problem in the future. Cost, availability of CT scanners, and need for scheduling continue to be obstacles to its use.

Slit scanography utilizes an X-ray beam that is tightly collimated to a thin transverse slit that exposes the film as the X-ray tube is moved from one end of the limb to the other [33]. Since the beam is always perpendicular to the film, theoretically magnification and parallax should be eliminated. Direct measurement using a tape measure is quite accurate and reliable [32]. Disadvantages include the facts that the patient is nonweight bearing and must remain still for the exposure and the equipment is not readily available in most clinicians' offices. Like the standard teleroentgenogram, it requires a special cassette (38.5×94.5 cm or 38.5×132.5 cm).

A three-view scanogram consists of three images with a ruler with radiopaque graduations placed vertically in the midline. Images of the hip, knee, and ankle are made with the beam perpendicular to the joint at each exposure to eliminate parallax and magnification. The technique is easily performed in most clinicians' offices, requiring only a ruler with radiopaque markings and standard radiography equipment. Its accuracy is comparable to the modified standing teleroentgenogram [33, 34]. Disadvantages include the slightly higher radiation dose [33], the necessity for the patient to remain still for all three images, the necessity to avoid any tilt of the ruler, and the necessity to avoid asymmetric positioning of the extremities in abduction or adduction. Deformity analysis cannot be performed, and midshaft pathology may be missed with a scanogram. Moreover, the contribution of the foot to the limb length discrepancy cannot be measured. Because of these shortcomings, scanograms

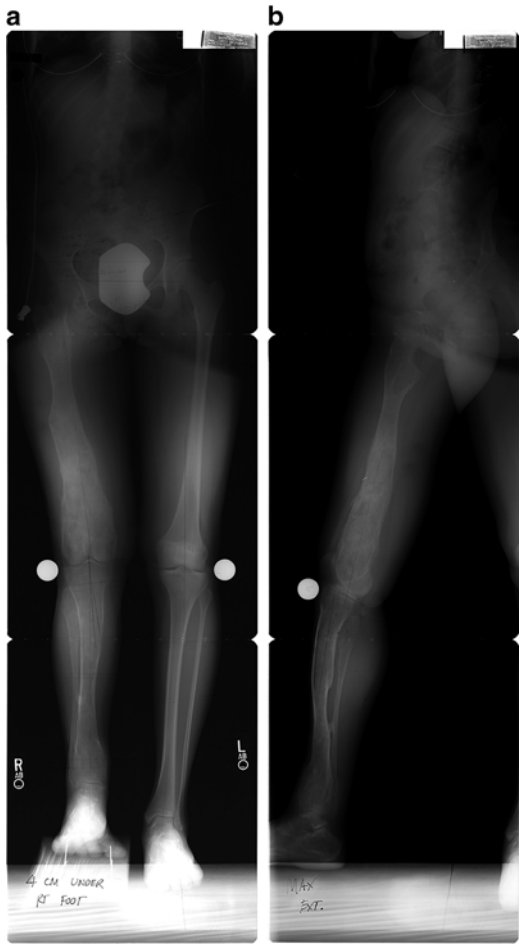


Fig. 2.11 (**a**) AP and lateral alignment radiographs of a 16-year-old girl with Ollier disease and a limb length discrepancy. She felt level with a 4-cm block under the right foot, but this block undercompensated for her discrepancy. (**b**) The lateral view is taken with the knee in maximum extension. Although the knee does not appear to extend completely, the lack of full extension is due to a juxtaarticular proximal tibial deformity, as quantified by proximal posterior tibial angle (PPTA). Note the 30-mm ball bearings at the levels of the knee to quantify magnification

are not recommended for the initial imaging technique, but offer convenience and accuracy for follow-up of femoral and/or tibial length discrepancies [33].

Orthoroentgenography is a radiographic technique of performing three exposures of the lower extremity on a long cassette. Similar to the scanogram, the beam for each exposure is centered over the hip, knee, and ankle to minimize magnification and parallax. By using rectangular collimation, the three exposures can be performed to obtain a full-length image from the hip to the ankle [31]. Direct measurement and deformity analysis on the image can be performed. However, the technique has many of the same disadvantages as the scanogram, including the inability to perform the study in a weight-bearing position, inability to measure the contribution of the foot to the length discrepancy, and need for the patient to avoid any movement while the three exposures are made. Higher radiation dose is also an issue.

In conclusion, the modified standing teleroentgenogram affords the most advantages for initial screening, since it will reveal the site of the discrepancy and any angular deformity and diaphyseal pathology. For follow-up, the standing teleroentgenogram, slit scanography, CT scanograms, or three-view scanograms may be performed, depending on the equipment and the absence of angular deformities or foot height discrepancy. If angular deformity and/or foot discrepancy is present, or deformity analysis is planned, the modified standing teleroentgenogram should be repeated.

Version Diagnostic Imaging

The presence of version (normal axial angulation) or torsion (abnormal version greater than two standard deviations from the mean value) is usually apparent from the physical exam without the need for diagnostic imaging. If the amount of version on physical exam is normal for the child's age, and the diagnosis is obvious and the imaging will not affect decision making, then no imaging studies are indicated. However, if the diagnosis is in doubt, if the suspected condition is outside the normal range for age, or if corrective surgery is being planned, imaging for diagnosis and quantification may be indicated. Since computerized tomography (CT) is usually necessary, one must weigh the advantages of information gained from the study against the disadvantages of radiation exposure and cost.

Acetabular Version

Computerized tomography measurements show that the normal acetabulum is anteverted approximately 13° in children from ages 1 to 15 years, with a large range of normal between ages 1 and 2 years [21, 22]. Visser et al. reported mean acetabular anteversion of 6.5° in a newborn and 16.5° in adults [37]. Excessive acetabular anteversion is often seen in children with DDH, and acetabular retroversion has been reported in children with Down syndrome [21]. McKibbin developed an "instability index" based on the sum of the acetabular version angle and the femoral anteversion angle. Values less than 20° or greater than 58° were associated with hip instability. He hypothesized that the version of the femur and acetabulum must develop in a coordinated manner [25]. Tönnis and Heinecke reported increasing pain and osteoarthritis in adult hips with a McKibbin instability index at the upper and lower limits of normal [19]. Currently there are no longitudinal studies of the development of acetabular version in normal and abnormal hips in children. Techniques for qualitative assessment using a single AP radiographic image of the pelvis have been described in adults but not in children [38, 39]. Until the relevance of quantifying acetabular version in

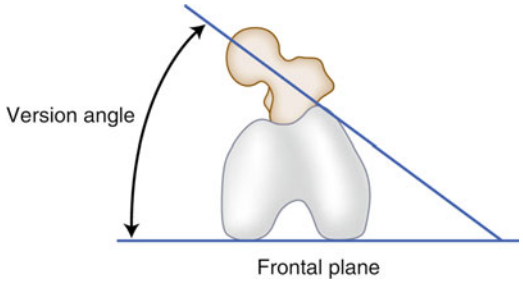


Fig. 2.12 Femoral version is the angle formed by the femoral neck and the plane of the femoral condyles. Adapted with kind permission from Springer Science+Business Media: Paley D. Principles of deformity correction. Berlin, Heidelberg; 2002 [23]

children is documented, computerized tomography and its high radiation exposure to the pelvic organs are probably not indicated with certain exceptions, such as preoperative planning for reconstruction of a hip dislocation associated with syndromes or following a failed initial hip surgery.

Femoral Version

Femoral version is the angle formed by the axis of the femoral neck with the tangent of the posterior aspect of the femoral condyles or transcondylar axis at the knee. The normal femur is anteverted, with the neck extending anteriorly from the coronal plane (Fig. 2.12). Normal values for infants and children have been published, with an average value of 30° to 40° at birth, declining to 8° in adult men and 14° in adult women [2, 4, 6, 19, 26].

Computerized tomography is currently the most accurate method of quantifying femoral version [40]. The equipment is readily available but exposes children to ionizing radiation. The morphologies of the femoral neck and greater trochanter sometimes make measurement difficult [41]. Various other radiographic techniques have been used, including biplanar radiography (RyderCrane [41] and Magilligan [42] techniques), fluoroscopy [43], CT scan, sinus wave biplanar conventional radiography, modified Heimann biplanar conventional radiography, and limited 3D volumetric tomography [40]. In order to minimize radiation exposure, ultrasound techniques have also been developed [44, 45]. One series showed that ultrasound measurements yielded lower values than the Magilligan technique, with larger discrepancies at greater degrees of torsion [44]. In contrast, another study in patients between 12 and 44 years of age found an anteversion value of 10° higher than the correct value and recommended ultrasound as a screening tool with radiographic methods if surgery is contemplated [45].

Different anteversion values arise due to the method of defining the distal reference line or plane to be compared to the proximal line or plane along the long axis of the femoral neck. Measuring the anteversion as a relationship between landmarks at the greater trochanter and long axis of the neck eliminates any contribution from the shaft. Even at the distal femur, the reference line or plane has been defined differently in various studies: as the flexionextension axis of the knee [43], as the plane bisecting the femoral condyles [42], and as a line tangent to the posterior aspect of the femoral condyles [19]. Because studies show that femoral antetorsion (excessive version) normally resolves and does not predispose to osteoarthritis, surgery is rarely indicated [8, 16, 24]. Antetorsion may not resolve in children with spastic cerebral palsy; in some such patients, corrective derotation osteotomies may be indicated [26].

When planning corrective femoral derotation osteotomies in children with cerebral palsy, accurate localization of the site of torsion proximal and distal to the greater trochanter is important. Kim et al. defined two components of femoral anteversion based on CT imaging: supratrochanteric torsion (STT) in the proximal femur and infratrochanteric torsion (ITT) in the distal femur. In normal children, the STT declines with age; the ITT showed a random distribution that did not decline with age for a net decrease in femoral antetorsion. In children with cerebral palsy, the ITT increased and the STT decreased, and the overall femoral antetorsion remained essentially unchanged. If derotation osteotomy is performed, it should be performed at the site of the torsion to avoid creating a new iatrogenic deformity and lever arm disease: muscle fatigue with or without a joint contracture [46]. Therefore, CT imaging may be indicated to localize the site of the torsion when planning derotation osteotomies.

Tibial Torsion

Similar to femoral version, tibial torsion rarely requires imaging for diagnosis or quantification. Computerized tomography has been described to measure tibial torsion as the angle formed by a line through the proximal tibial juxtaarticular area and the distal bimalleolar axis or distal articular surface [47]. Similarly ultrasonography measures the tibial torsion by the angle between the plane of the posterior aspect of the proximal tibia and the plane of the posterior aspect of the distal tibia [28]. These imaging techniques assess tibial anatomy, but because they use different bony landmarks, the values obtained differ slightly. Unlike clinical assessment of the thigh–foot angle or bimalleolar axis, CT or ultrasound negates any effect of foot pathology or mobility of the fibula. Ultrasound does not expose the child to radiation. Although experimentally produced tibial torsion increased pressure at the tibiotalar joint in cadavers [48], currently there is no known detrimental effect of physiologic tibial torsion in young children. Therefore, there is hardly any indication for performing such imaging studies in asymptomatic individuals.

Coronal Plane Angulation

Distinguishing physiologic bowing of the lower extremities in infants from infantile tibia vara (Blount disease) is a frequently encountered reason for orthopedic evaluation. Early and accurate diagnosis is important in order to avoid unnecessary treatment and radiation exposure in infants with physiologic bowing while preventing progression of the deformity in infants with tibia vara. The child with a negative cover-up test; normal height, weight, and BMI for age; negative family history for orthopedic disorders; and no other remarkable physical findings rarely needs a radiograph, but should be followed for resolution of the bowing [17].

Several studies have attempted to use radiographic measurements to distinguish normal physiologic bowing and tibia vara. The ideal radiographic imaging technique would have high sensitivity (such as not incorrectly missing a case of tibia vara and neglecting treatment) and high specificity (such as not incorrectly classifying a case of physiologic bowing as tibia vara and treating a child unnecessarily in infants before age 2 years). Langenskiöld and Riska classified six stages of infantile tibia vara, based on the radiographic appearance of the proximal tibia [49]. The classification system has several shortcomings. The radiographic changes are not seen before age 2 years; Langenskiöld stage is not related to prognosis and does not consistently guide treatment [50]. Interobserver agreement is good for the early and late stages but poor for the intermediate stages [50]. Salenius and Vankka described normal mean and standard deviations of the radiographic femoral-tibial angles for infants and children [14]. Although varus angles after age 2 years are abnormal, this method does not distinguish physiologic varus at the upper limit of normal from tibia vara before age 2 years. O'Neill and MacEwen measured the distal femoral metaphyseal-diaphyseal angle (FMDA) and the tibial metaphyseal-diaphyseal angle (TMDA). The distal femoral metaphyseal angle (FMDA) is formed by a line perpendicular to the line connecting the medial and lateral edges of the physis and a line drawn through the midpoint of the femoral shaft at points 2 and 3 cm proximal to the epiphysis. The proximal tibial metaphyseal angle is measured in a similar manner (Fig. 2.13). Children with a TMDA greater than the FMDA or fibula longer than the tibia were more likely to have a persistent varus deformity which did not resolve by age 30 months [51]. Levine and Drennan measured the metaphyseal-diaphyseal angle (MDA) at the proximal tibia, formed by the intersection of a transverse line through the proximal tibial metaphysis and a line perpendicular to the long axis of the tibial shaft in children between ages 11 months and 30 months (Fig. 2.14). Only 3 of 58 patients with an MDA less than 11° developed radiographic changes of tibia vara, whereas 29 out of 30 patients with an MDA of 11° or more developed radiographic signs of tibia vara [52]. Using Levine's and Drennan's radiographic technique, Feldman and Schoenecker developed guidelines with improved sensitivity and specificity by using 9° and less as the criterion for physiologic bowing and 16° or greater as the criterion for tibia vara. For patients with an MDA between 9° and 16°, the authors considered bracing only if there is instability with walking and recommended evaluating the patient at 4-month intervals until the bowing resolves or the characteristic signs of Blount disease are seen [53]. McCarthy et al. used the femoral-tibial ratio (FTR), defined as the femoral MDA divided by the tibial MDA, to differentiate between infantile tibia vara and physiologic bowing. If the tibial MDA is greater than the femoral MDA, the femoral-tibial ratio is less than one, indicating probable infantile tibia vara instead of physiologic bowing. If the FTR was less than one, indicating physiologic bowing, the authors reported a false-negative rate of 10 % (one in ten cases of infantile tibia vara would be missed) and a false-positive rate of 7 % (approximately 1 in 14 cases of bowing would be incorrectly diagnosed as infantile tibia vara). They found good inter- and intraobserver reliability, with little effect of rotation position for the radiograph [54]. Stricker and Faustgen emphasized the importance of neutral limb rotation when measuring the TMDA and the tibial-femoral angle [55].

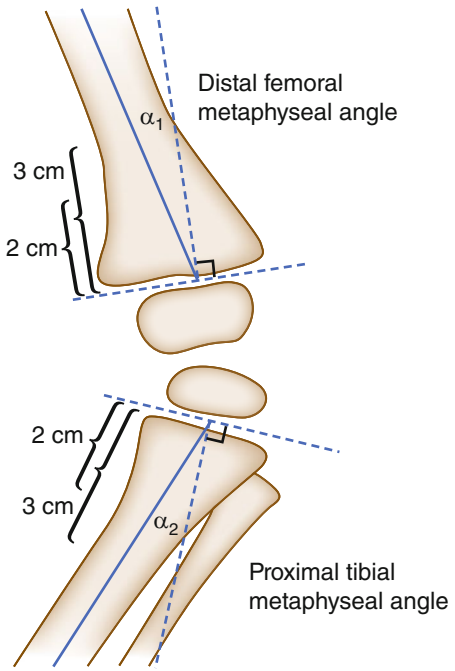


Fig. 2.13 The distal femoral metaphyseal-diaphyseal angle (FMDA) α1 and the proximal tibial metaphyseal-diaphyseal angle (TMDA) α2 of O'Neill and MacEwan. Adapted with permission from O'Neill DA, MacEwen GD. Early roentgenographic evaluation of bowlegged children. J Pediatr Orthop. 1982;2(5):547–53 [51]

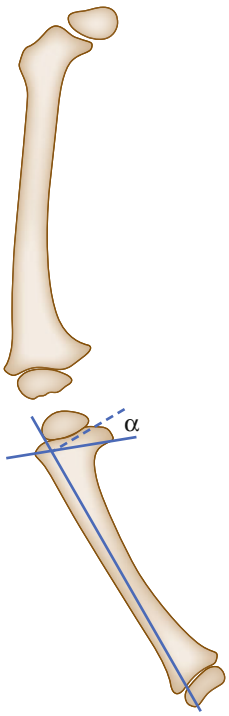


Fig. 2.14 The tibial metaphyseal-diaphyseal angle (MDA) of Levine and Drennan. Adapted from [52]

Because none of these methods guarantees perfect sensitivity and specificity, a combination of methods should be utilized, including measuring the FMDA and TMDA on an alignment radiograph in neutral rotation. An FTR less than one or an abnormally large TMDA warrants serious consideration of the diagnosis of early infantile tibia vara. However, there is no single value of an abnormally high TMDA that will differentiate between physiologic bowing and infantile tibia vara with perfect sensitivity and specificity.

Quantitative Deformity Analysis

The physician should use radiographic imaging to make a diagnosis, quantify the deformity, and plan correction of the deformity. A single-exposure weight-bearing teleroentgenogram or similar digital image from the hip joint to the floor is used to identify and quantify coronal (AP) and sagittal (lateral) plane deformities. This technique minimizes distortion and magnification due to divergence of the X-ray beam and is reproducible so that serial radiographs can be compared. Measuring traditional full-length radiographs was reported to be more reliable than measuring paper images or digital radiographs [56].

After adequate imaging, the clinician must analyze the deformity to quantify the components of the deformity. Accurate analysis is necessary to make a diagnosis and plan surgical correction if necessary.

Table 2.4 The six deformity components

The three angulation components are:	
Apex medial (valgus)	Angulation in the AP (*XY*) plane
Apex lateral (varus)	
Apex anterior (procurvatum)	Angulation in the sagittal (*YZ*) plane
Apex posterior (recurvatum)	
External rotation	Angulation in the *XZ* plane
Internal rotation	
The three translation components are:	
Translation along the *X* axis	Medial translation, lateral translation
Translation along the *Z* axis	Anterior translation, posterior translation
Translation along the *Y* axis	Lengthening, shortening

Skeletal deformity is a vector. Like all vectors, deformity has magnitude, direction, and location.

Magnitude of a skeletal deformity has six components: Three angulations and three translations (Table 2.4). Although these components of the deformity may coexist, they are independent of each other. The six components are measured in an XYZ three-dimensional coordinate system.

A deformity may have any or all of these components. A deformity with components in more than one plane is an oblique plane deformity, not a biplanar deformity. The magnitude of the deformity in the oblique plane (the true plane of the deformity) is greater than the magnitude of any of its orthogonal components.

In order to document the presence of a deformity on a radiograph, there must be at least one of the following: mechanical axis deviation (MAD), abnormal joint orientation angle(s), and limb length inequality. Conversely, to document accurate correction radiographically, it is both necessary and sufficient to correct the mechanical axis of the extremity, joint orientation angles, and limb length inequality. Abnormal axial angulation (malrotation) may be quantified by computed tomography if necessary, but it is usually assessed by physical examination.

Mechanical Axis and Anatomic Axis

The entire lower extremity, the tibia, and the femur have a *mechanical axis* in the coronal plane. The mechanical axis of the lower extremity is a straight line from the center of the femoral head to the center of the ankle. The mechanical axis is the weight-bearing line during quiet standing. In a normal lower extremity, this line passes 010 mm medial to the center of the knee [23]. The femur and tibia each has a mechanical axis. The femoral mechanical axis is a straight line from the center of the femoral head to the center of the knee; the tibial mechanical axis is a straight line from the center of the knee to the center of the ankle mortise. No matter how deformed a femur or tibia is, whether due to a malunion or a developmental deformity, the mechanical axis is, by definition,

Table 2.5 Hip–knee–ankle angle (degrees), tibial-femoral angle (degrees), and mechanical lateral distal femoral angle (mLDFA) (degrees)[a]

Age (years)	Hip–knee–ankle angle[b]	Tibial-femoral angle[b]	mLDFA[c]
	Mean and standard deviation	Mean and standard deviation	Mean and standard deviation
1<2	3.6±5.6	3.8±4.2	94.8±3.8
2<3	2.5±3.2	9.1±3.0	90.7±3.4
3<4	2.0±2.8	9.0±3.2	89.0±2.5
4<5	1.7±1.7	8.6±2.0	87.5±2.5
5<6	2.3±2.6	9.0±3.3	88.3±1.1
6<7	1.9±2.1	7.6±2.0	87.7±2.3
7<8	0.4±2.9	6.7±2.6	86.8±2.2
8<9	0.7±2.4	6.0±1.9	88.1±1.9
9<10	0.1±2.8	6.9±2.5	88.1±1.9
10<11	0.6±2.1	6.4±2.1	87.9±1.8
11<12	0.2±2.1	6.4±1.7	87.2±1.8
12<13	0.2±2.5	6.5±2.3	87.0±2.1
13<14	0.1±2.3	6.7±2.4	87.5±1.9
14<15	0.1±2.3	6.6±2.6	87.4±1.9
15<16	1.0±2.1	5.3±2.1	87.7±1.6
16<17	1.1±2.2	5.3±1.7	87.6±1.5
17<18	0.1±2.6	5.9±2.2	87.7±1.6

[a]Adapted from Sabharwal et al. [13, 57]
[b]Negative values indicate valgus angulation; positive values indicate varus angulation
[c]Angles less than 90° indicate valgus angulation and greater than 90° indicate varus

a straight line from the center of the joint at the proximal end to the center of the joint at the distal end.

The mechanical axes of the femur and tibia intersect at the center of the knee to form the hip–knee–ankle angle. In adults, the mechanical axes of the femur and tibia are almost collinear, forming a mean angle at their intersection at the knee of 1.2±2.3° varus [57]. Thus the mechanical axis of the lower extremity nearly coincides with the mechanical axis of the femur and mechanical axis of the tibia with very slight deviation medial to the center of the knee.

In children, however, the normal hip–knee–ankle angle ranges from 3.6±5.6° varus from age 1 to 2 years to 2.5±3.2° valgus from age 2 to 3 years, and then declines to 0.3° varus after age 7 years (Table 2.5). The changes in the lower extremity alignment, as quantified by the hip–knee–ankle angle, beginning as varus until age 2 years, changing to valgus at age 2–3 years, and stabilizing as very slight varus from age 7 years onward, correlate closely with similar changes in the alignment of the distal femur, and, to a lesser degree, of the proximal tibia [13].

The femur and tibia also have an *anatomic axis*. The anatomic axis is defined as a straight line determined by a series of mid-diaphyseal points. The anatomic axis is where an intramedullary nail is placed. In the femur, the AP anatomic axis differs from the AP mechanical axis. Unlike the mechanical axis, the anatomic axis is unaffected by abnormalities of the head or neck, such as coxa vara or avascular necrosis of the femoral head. By definition, the femoral mechanical axis intersects the distal femur at the center of the knee, midway between the femoral condyles. The femoral anatomic axis intersects the distal femur more medially, at approximately the junction of the concave intercondylar notch and convex medial femoral condyle (Fig. 2.15). The tibial anatomic axis is parallel and slightly medial to the tibial mechanical axis, but the difference is insignificant, so for most purposes the two axes of the tibia can be considered identical.

Although using the mechanical or anatomic axis to analyze deformity should yield similar results, the mechanical axis method is preferred for several reasons: The goal of correction is to restore the mechanical axis of the entire extremity. Also, in a severely curved femur or tibia, the deformed bone will not have a single anatomic axis. In the operating room, if a sufficiently large film cassette is not available to visualize the entire femur, the anatomic axis method may need to be used to verify adequate correction.

The femur and tibia have an anatomic axis in the sagittal plane as well. In the lateral radiographic view, the mechanical axes of the femur and tibia are almost never used; only the anatomic axis is used for deformity analysis. Similar to the AP anatomic axis, the lateral anatomic axes of the femur and tibia are straight lines determined by a series of mid-diaphyseal points. Since the femur has a slight bow in the sagittal plane, the line is determined by a series of mid-diaphyseal points closest to the section of interest.

Deformity analysis should be done in a stepwise manner (Fig. 2.16). The first step is to measure the lengths of the entire lower extremity, femur, and tibia bilaterally. The second step in determining the presence or absence of deformity is to draw the AP mechanical axis of the extremity and then answer the question: Does the anteroposterior (AP) mechanical axis pass through the center of the knee? The distance from the AP mechanical axis to the center of the knee in millimeters is the *MAD*. The mechanical axis normally passes through the center or slightly medial to the center of the knee (defined as medial MAD) for children and adults with the exception from ages 4 to 6 years, when it passes very slightly lateral to the center of the knee (lateral MAD) [13]. If there is AP MAD outside the normal range, there is deformity. Medial MAD indicates lower extremity varus; lateral MAD indicates valgus. The presence of MAD outside the normal range confirms the presence of deformity, but the absence of MAD does not confirm the absence of deformity (such as due to abnormal but compensatory joint orientation angles).

Joint Orientation Angles

The third step to discover deformity or to confirm correction of deformity is measurement of the joint orientation angles. Normal joint orientation angles and mechanical axis within

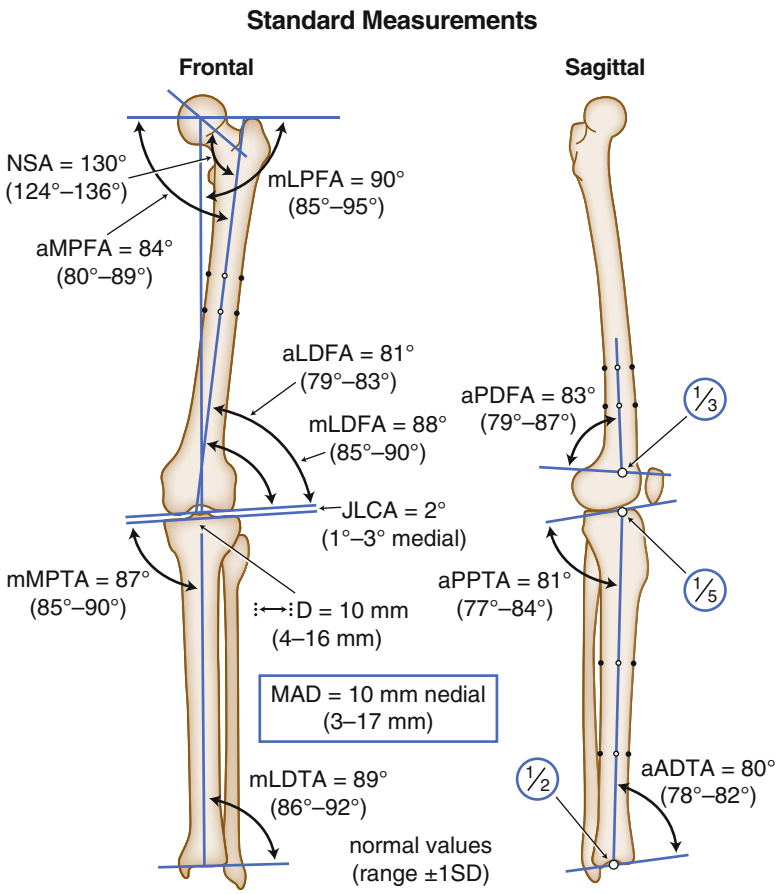


Fig. 2.15 Standard measurements for joint orientation angles and mechanical axis deviation. Adapted with kind permission from Springer Science+Business Media: Paley D. Principles of deformity correction. Berlin, Heidelberg; 2002 [23]

range of normal are both necessary and sufficient to rule out deformity or to confirm correction of deformity. If either is abnormal, there is deformity. The femur and tibia have a proximal joint orientation angle and a distal joint orientation angle on both the AP and lateral radiographs. The joint orientation angle is formed by the intersection of the joint orientation line with the axis, either mechanical or anatomic, of the respective bone. The joint orientation lines for the femur and tibia are described in Table 2.6.

The joint orientation angles are abbreviated by five letters. The first letter is a lower case "a" or "m," which designates anatomic or mechanical, respectively, depending on which axis is used to form the angle. Usually a (anatomic) or m (mechanical) is only used to differentiate between the two techniques of determining the joint orientation angles of the femur in the AP plane. In the lateral view of the femur, and AP and lateral views of the tibia, the joint orientation angle is formed by the anatomic axes. The second letter is either M (medial), L (lateral), A (anterior), or P (posterior), which designates the location of the angle with respect to the axis of the bone so that the normal value of the angle is 90° or less. This nomenclature is not used for the femoral neck–shaft angle because of the long-standing convention of measuring the relationship between the neck and the shaft as the larger of the two supplementary angles at this site. The third letter is F (femur) or T (tibia). The final letter is A, the abbreviation for angle.

Figure 2.15 shows the normal joint orientation angles for the lower extremity in adults. However, the age-specific pediatric values should be used for children less than 7 years of age [13].

A deformed femur or tibia has a deformity *magnitude*, deformity *direction*, and deformity *level*.

The *magnitude* of the deformity is the magnitude of the angle formed by the intersection of the proximal axis and distal axis of the bone. In a normal bone, the proximal and distal axes of the bone form an angle of zero; the two axes are collinear.

The proximal and distal axes of the bone can be drawn by using the long segment method or the short segment method (Figs. 2.17 and 2.18).

The *long segment method* is used if the deformity is near the middle of the bone (mid-diaphyseal). The anatomic axis (a series of mid-diaphyseal points) of each segment is drawn, and the angle formed by the intersection of these lines determines the magnitude and level of the deformity in the plane in which it is measured. The joint orientation angles at each end of the bone, formed by the intersection of

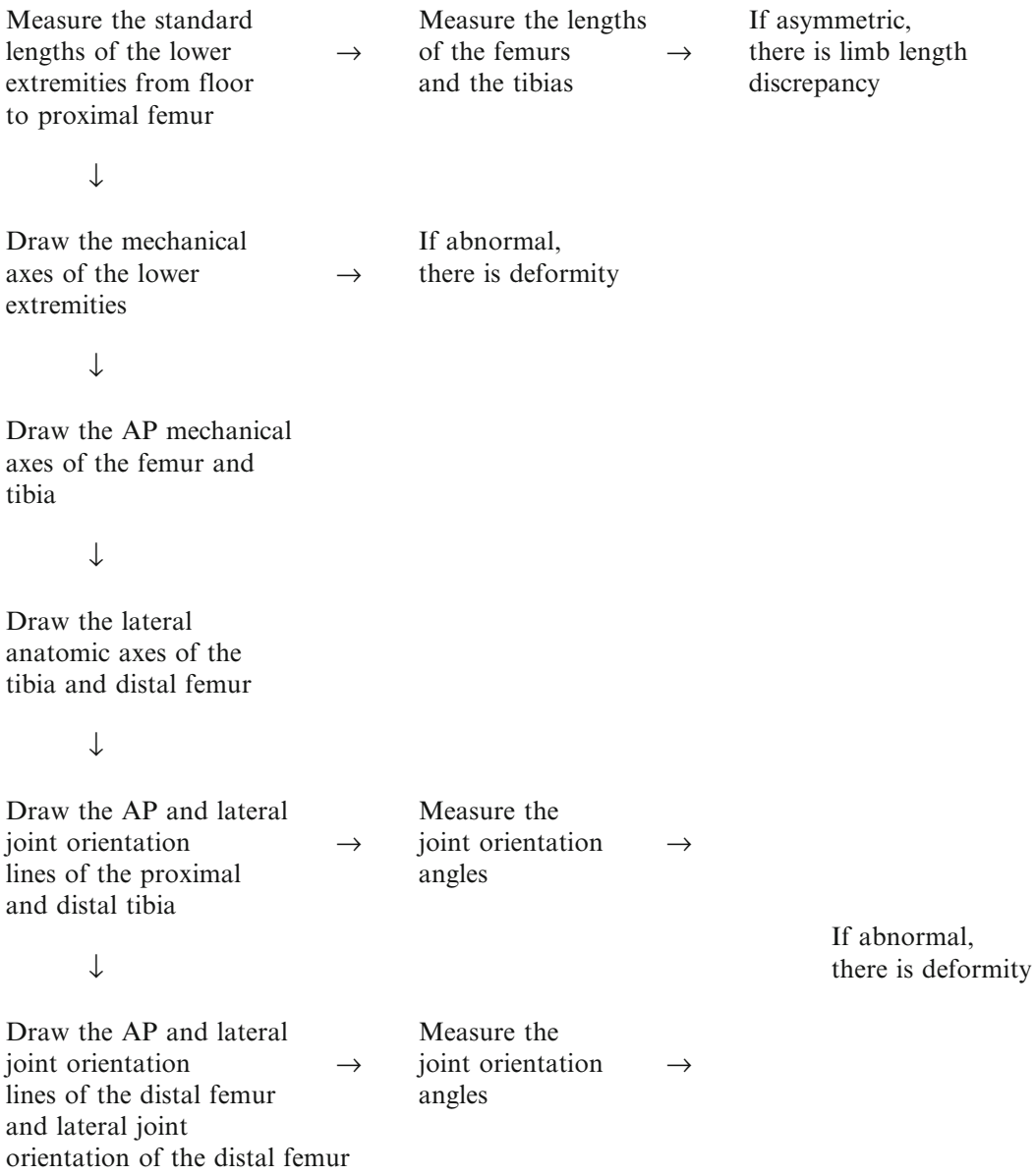


Fig. 2.16 Steps for detecting deformity on a radiograph of the lower extremities

Table 2.6 Joint orientation lines of the femur and tibia (see Fig. 2.17)

AP plane	
Proximal femur	Line from tip of the greater trochanter to the center of the femoral head (mechanical axis method). Longitudinal axis of the femoral neck (anatomic axis method)
Distal femur	A line tangential to the femoral condyles
Proximal tibia	A straight line from the lateral corner to the medial corner of the tibial plateau
Distal tibia	A line across the subchondral bone of the distal tibia at the ankle mortise
Lateral plane	
Proximal femur	Neck–shaft angle but rarely used in the lateral plane
Distal femur	A line connecting the anterior and posterior extent of the distal femoral physes or the site of the closed physis
Proximal tibia	A line across the subchondral bone of the proximal tibial plateau
Distal tibia	A line between the anterior and posterior corners of the distal tibia

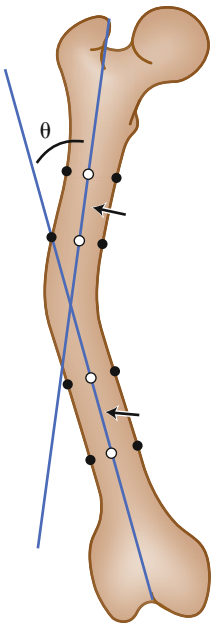


Fig. 2.17 The long segment method quantifies mid-diaphyseal deformities. It is θ, the angle formed by the intersection of the anatomic axes of the proximal and distal segments of the femur. Adapted with kind permission from Springer Science+Business Media: Paley D. Principles of deformity correction. Berlin, Heidelberg; 2002 [23]

the joint orientation line and the respective axis, must be checked. If abnormal, there is juxtaarticular deformity in addition to the mid-diaphyseal deformity.

If, however, one segment is too short (too close to the adjacent joint to draw a straight mid-diaphyseal line) or if the joint orientation angle of a long segment is abnormal (a juxtaarticular deformity), the *short segment method* is used. The magnitude of the deformity is the magnitude of the angle formed by the intersection of the long and short segments. The axis of the long segment of the bone being measured is determined by drawing the axis as a series of mid-diaphyseal points, and the short segment axis is drawn by utilizing the normal joint orientation angle, determined either by the contralateral limb joint orientation angle, if normal, or by the population normal value. The short segment axis must be drawn at its position on the joint orientation line in a normal skeleton.

This method of determining the long anatomic axis and short anatomic axis of the bone is valid for the femur and tibia in both the AP and lateral views. In certain situations in the femur, such as diaphyseal bowing or surgeon's desire to restore the *mechanical* axis to normal, the mechanical axis method is used by drawing the proximal mechanical axis and the distal short segment mechanical axis, as determined by the mechanical lateral distal femoral angle.

Because the AP mechanical and AP anatomic axes of the femur differ, the AP proximal femoral axis is drawn by first drawing the proximal joint orientation line (center of the femoral head to the tip of the greater trochanter) and then drawing a straight line distally from the center of the femoral head with a mechanical lateral proximal femoral angle (mLPFA) equal to the contralateral side if the contralateral femur is normal or the population normal value (90°) if the contralateral side is abnormal. One must not use the mechanical axis at one end of the femur and anatomic axis at the other end.

Oblique Plane Deformities

A deformity seen in both the AP and lateral views is an *oblique plane deformity. Biplanar deformity* is a misnomer because the deformity is only in a single plane (the plane of maximum deformity) between the AP and lateral planes (Fig. 2.19). Using the previously described techniques, the magnitudes of the AP (coronal) plane and lateral (sagittal) plane deformity are measured. The true magnitude of the deformity can be determined by either the graphic or the trigonometric method. The graphic method uses the Pythagorean theorem (Fig. 2.20):

$$\theta = \sqrt{\theta^2_{AP} + \theta^2_{lat}}$$

where θ = magnitude of the true or resultant angle of deformity
θAP = magnitude of the angular deformity in the AP plane
θlat = magnitude of the angular deformity in the lateral plane

Notice that the true angular deformity is greater than either orthogonal (AP and lateral) component for oblique plane deformities.

The graphic method, which utilizes the Pythagorean theorem, is a good approximation, but the exact magnitude of the resultant angle can be calculated using the trigonometric method from the equation

$$\theta = \tan^1 \sqrt{\tan^2 \theta_{AP} + \tan^2 \theta_{lat}}$$

where $\tan^1$ = arc tangent
$\tan \theta_{AP}$ = tangent of the angular deformity in the AP plane
$\tan \theta_{lat}$ = tangent of the angular deformity in the lateral plane

The graphic method is easier to use and yields an approximation within 0° to 2° for physiologic angular deformities.

The *direction* of the deformity is the orientation plane of the true (resultant) angular deformity. It is the plane of maximum deformity magnitude. The true orientation can be approximated by using the graphic method (see Fig. 2.20). Although it is only an approximation, it is quite accurate within the range of physiologic deformities. The orientation

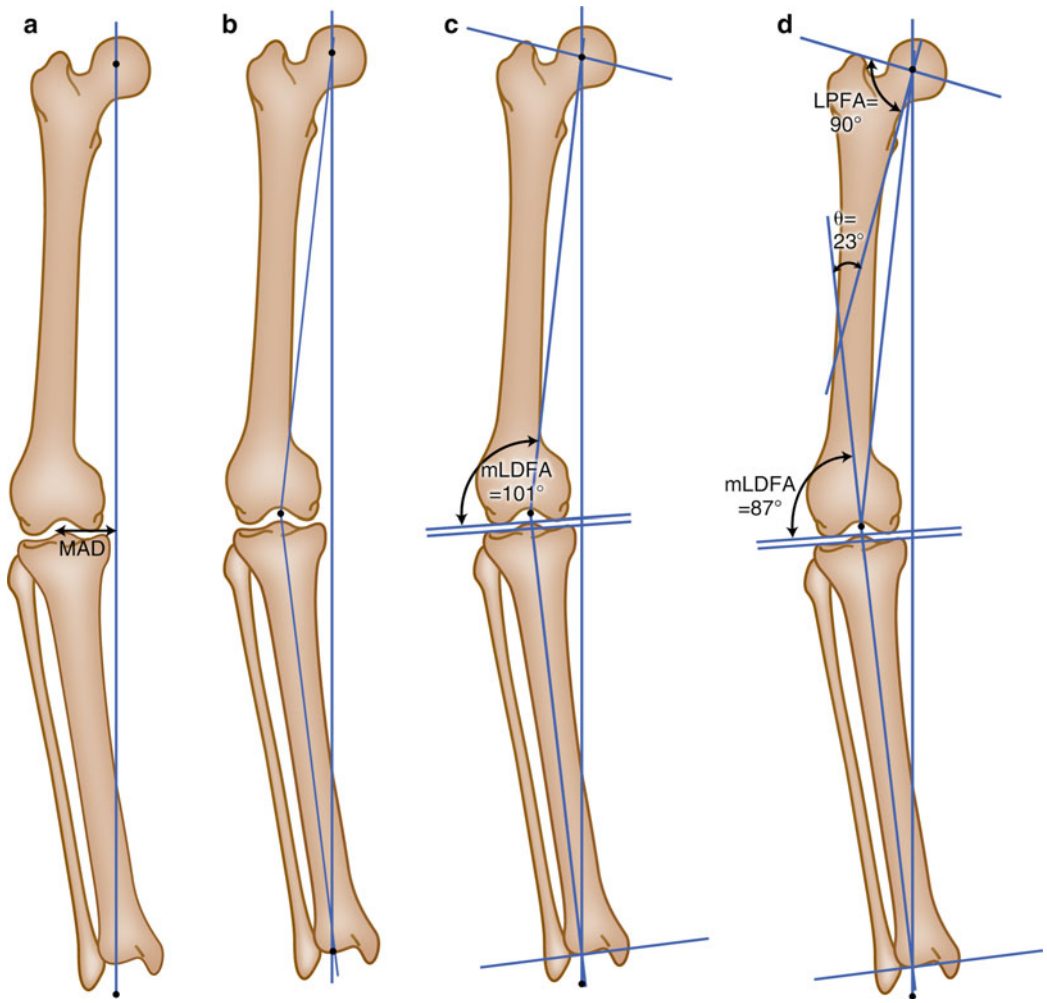


Fig. 2.18 The short segment method quantifies the deformity when one segment is too short to draw the anatomic axis. The axis of the short segment is drawn from the joint orientation lines, and the intersection of these two axes quantifies the deformity angle θ. (**a**) The mechanical axis of the lower extremities is drawn, confirming the presence of a deformity, as quantified by the mechanical axis deviation (MAD). (**b**) The mechanical axes of the femur and tibia are drawn. (**c**) The joint orientation lines of the femur and tibia are drawn. The mechanical lateral distal femoral angle (mLDFA) is 101°, which is abnormal. (**d**) The proximal and distal mechanical axes of the femur are drawn from the joint orientation lines. The intersection of the axes forms the angle θ, the magnitude of the deformity. Note that this method uses the mechanical axes of both segments of the femur. One must not use the *anatomic* (diaphyseal) proximal axis with the *mechanical* distal axis. Adapted with kind permission from Springer Science+Business Media: Paley D. Principles of deformity correction. Berlin, Heidelberg; 2002 [23]

angle α can be measured by drawing two perpendicular lines representing the magnitudes of the AP and lateral angulations. The angle between the resultant diagonal lines and the horizontal axis represents the orientation of the oblique plane deformity, the direction of the true deformity angle. This angle is the angle formed by the intersection of the plane of maximum deformity with the coronal plane.

The trigonometric method for measuring the orientation angle α yields the exact value and can be calculated from the formula

$$\alpha = \tan^{1} \frac{\tan \theta_{\text{lat}}}{\tan \theta_{\text{AP}}}$$

The *level* of the deformity is the location of the intersection of the proximal and distal axes. If the level differs on the AP and lateral radiographs, there is either translation or there is an additional angular deformity on one of the views.

The third component of angulation in the *XZ* plane is rotation around the *Y* axis, currently quantified with imaging studies. Since the magnitude is usually obtained by CT software, the measurement will not be discussed here.

Translation, if present, may occur in one, two, or three planes. The magnitude of translation is the length of the line drawn perpendicular from the axis (either mechanical or anatomic) of one segment to a point on the axis of the other segment. Developmental and congenital deformities do not

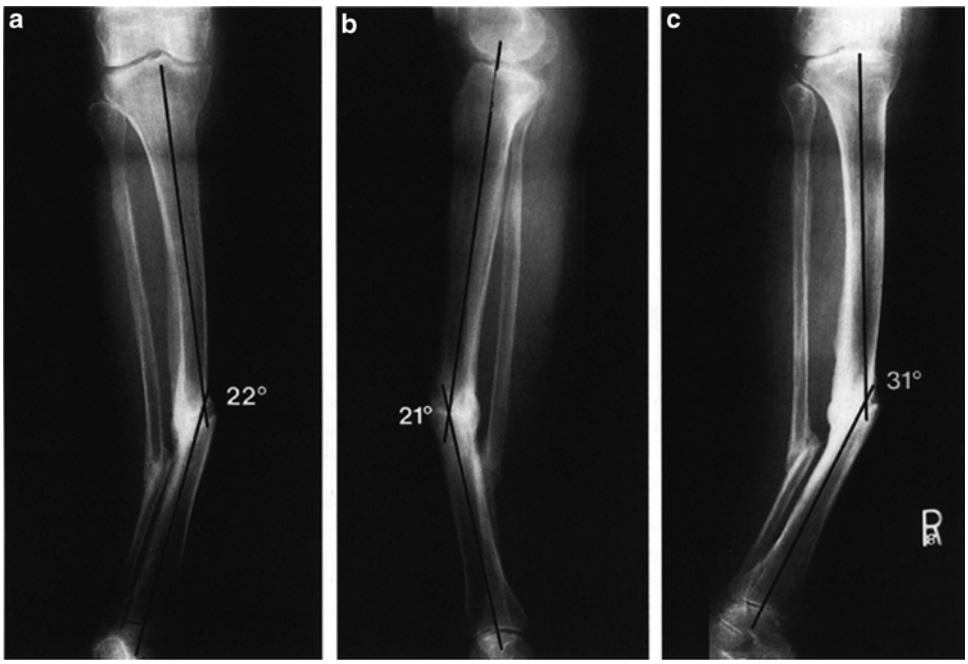


Fig. 2.19 (**a**, **b**) Oblique plane angular deformity of the tibia. Radiographs show angulation on the AP (22°) and LAT (21°) projections of magnitude. This is not a biplanar deformity but rather a single plane of angulation where the true plane of angulation lies between the frontal and sagittal planes. (**c**) Oblique radiograph obtained perpendicular to the plane of angulation shows the maximum angulation (31°). With kind permission from Springer Science+Business Media: Paley D. Principles of deformity correction. Berlin, Heidelberg; 2002 [23]

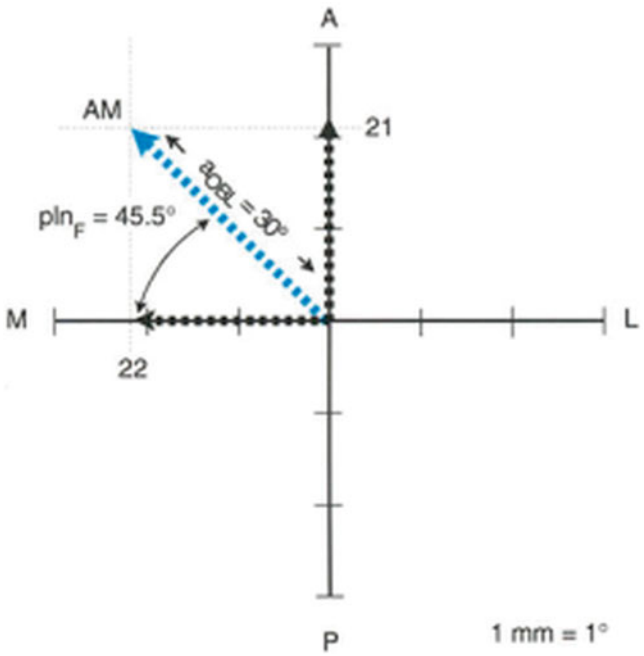


Fig. 2.20 The graphic method can be used to calculate the resultant oblique plane deformity and to determine the plane of the deformity. A vertical vector with a magnitude of 22° represents the apex anterior deformity. A horizontal vector of 21° represents the apex lateral (varus) deformity. The resultant angulation (30°) is represented by the diagonal of the rectangle formed by the AP and lateral angulations on the graph. The plane of the angular deformity is oriented 45.5° from the coronal plane. With kind permission from Springer Science+Business Media: Paley D. Principles of deformity correction. Berlin, Heidelberg; 2002 [23]

have a translation component. Translation with angulation is associated with high-energy fractures, where one fragment is angulated and translated with respect to the other by the energy of impact. Like angulation, the translation components in the orthogonal planes can be used to calculate the true magnitude of translation and its orientation. Unlike angulation, the graphic method using the Pythagorean theorem yields the exact magnitude and orientation instead of an approximation.

Box 2.1. Pitfalls

- For maximum accuracy, use metal tape measure, not cloth.
- During the prone exam, stabilize the pelvis on the exam table to avoid rocking the child side to side, which will give a false high estimation of hip rotation.
- Do not draw the AP mechanical axis at one end of the femur and the AP anatomic axis at the opposite end. The mechanical and anatomic axes are interchangeable elsewhere in the lower extremity, but not in the AP femur.

Laboratory Studies

Serum and urine chemistry studies are rarely needed to evaluate lower extremity deformities. However, if a rachitic disorder is present, serum and urine studies will be helpful in determining the etiology (Table 2.7).

In summary, many youngsters are brought to the physician's office by concerned parents, often very anxious to get

Table 2.7 Chemical findings in various forms of rickets [58]

Serum							Urine		Miscellaneous serum and urine values
			Alk		25(OH)	$1,25(OH)_2$			
Type of rickets	Ca^{2+}	P	Phos	PTH	Vit D	Vit D	%TRP	Ca^{2+}	
Vitamin D-deficient rickets	↓ or →	↓	↑	↑	↓	↓	↑	↓	
Dietary phosphate deficiency	→	↓	↑	→	→	→	→	→	
Gastrointestinal rickets	↓	↓	↑	↑	↓	↓ or →	↓	↓	↓ Absorption from gastrointestinal tract
Vitamin D-resistant rickets									Amino acids, sugar, water, base in urine
I. Phosphate diabetes	→	↓↓		→	→	→	↓↓	→	
II. Reduced $1,25(OH)_2$ vitamin D production	↓	↓	↑	↑	→	↓↓	↓	↓	
III. End-organ insensitivity	↓	↓	↑	↑	↑ or →	↑ or →	↓	↓	
IV. Renal tubular acidosis	↓	↓	↑	↑	↑ or →	↑ or →	↓	↑	Na ↓, K ↓, C1 ↑; acidosis alkaline urine
Renal osteodystrophy	↓	↑	↑	↑↑	↓↓	↓↓	?	↓↓	BUN ↑

↑ increased, ↓ decreased, → unchanged or normal, *Alk Phos* alkaline phosphatase, *BUN* blood urea nitrogen, *Ca* calcium, *Cl* chloride, *K* potassium, *Na* sodium, *P* phosphate, *PTH* parathyroid hormone, *TRP* tubular reabsorption of phosphate, *Vit D* vitamin D forms

treatment. When evaluating a child or adolescent with a suspected lower extremity deformity, the clinician must first determine if there is true deformity or simply normal pediatric limb development. The latter needs no treatment but can safely be observed for natural resolution while giving reassurance to the family. This determination can often be performed by a history and physical examination. If the components of the history and physical are consistent with the diagnosis of normal childhood development, imaging studies are not necessary. When imaging studies are deemed to be necessary, they must be performed using techniques which maximize diagnostic accuracy yet minimize expense and radiation exposure. These imaging studies can then be evaluated to make a diagnosis of deformity instead of normal development and to quantify the deformity to plan correction. Laboratory studies are rarely needed but may be useful to determine if there is a metabolic etiology.

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Decision Making in Lower Extremity Deformity Correction

3

Mark L. Miller and J. Eric Gordon

Introduction

Lower extremity deformities can vary widely with regard to their etiology, magnitude, integrity of the soft-tissue envelope, and natural history. Because of this variation, it is difficult to create a formulaic method of approaching the patient with a lower limb deformity. This difficulty is compounded by differences in the patient's environment, surgeon's skills, resources available to treat the patient, and ability of the patient or family to comply with a treatment program. Because of these differences, it is important to assess how each of these factors may influence the treatment method that is ultimately chosen and to follow a consistent approach in analyzing the deformity and planning the surgical correction. The nature and location of the deformity should guide the surgeon's choice of implant, not vice versa.

When approaching lower extremity deformities, it is important to develop a comprehensive problem list outlining the pathology associated with the involved limb. In some situations these lists may be quite short, outlining only a simple uniplanar angulatory deformity. In other situations, however, these lists can be quite complex, combining multiple deformities in different planes and locations, soft-tissue issues, joint instability, and psychological and family issues that can be extremely challenging.

It is important to remember that even with a consistent approach to decision making, it is not uncommon that multiple approaches to similar clinical deformities and symptoms may be appropriate and that there may not be a single "best" approach. Surgeon skills and patient/caretaker preferences can often influence the final decision making in these situations and it is important that the surgeon discuss the various options and potential complications with the patient and/or family prior to making a final decision.

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Clinical Evaluation

When evaluating a patient, there is a temptation for surgeons to look at radiographs of the patient and make decisions regarding the ultimate treatment without taking into account other factors. Like other areas of medicine, it is important for the surgeon to obtain a history of the problem, perform a careful physical examination, and obtain appropriate laboratory studies and imaging prior to making a diagnosis and developing a treatment plan.

When obtaining a history, it is important to elicit from the patient or family a chronologic account of the development of the deformity as well as current symptoms of pain, instability, and functional limitations. The surgeon should understand the limitations that the patients and their caretakers perceive as well as their expectations of treatment. The presence of joint instability, limitation of motion, pain, neurologic symptoms, signs of infection, and previous treatment are crucial to elicit from the patient or family. Past medical history, such as coagulopathy or bleeding issues, compromised immune system, and chronic medical conditions, can also influence the decision making.

Examination of the patient generally starts with observation of the individual's gait. Factors that contribute to the ultimate plan include general conditioning (for instance, is the patient obese?). The examination should also include an assessment of motion and stability of the major lower extremity joints. Limitation of joint motion or instability can have a direct impact on both the etiology and the treatment of deformities. For example, a hip adduction contracture can lead to the development of knee valgus in patients and attempts to correct the knee valgus without addressing the hip adduction contracture can lead to gait problems after correction of the knee deformity as

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well as postoperative recurrence. Children with congenital limb length discrepancies frequently have hip, knee, and/or ankle instability of the shorter extremity, which can have a substantial impact on subsequent limb lengthening. Additionally, rotational profile should be assessed by physical examination, documenting femoral anteversion [1] and tibial torsion clinically.

Imaging studies provide information about the static deformity in both the anteroposterior and lateral planes. It is crucial when evaluating the radiographs that the surgeon ascertains not only the presence of malalignment, but also the location of one or more deformities. This can be best accomplished by either mechanical axis planning or anatomic axis planning utilizing full-length radiographs of the entire lower extremity including the hip, knee, and ankle. This portion of the evaluation has been addressed in the previous chapter. Components of the deformity can include angulation, translation, length, and rotation. Of these components, angulation, translation, and length can be largely described from the radiographs. Rotation is often best ascertained by examining the patient, although specialized rotational studies such as CT scans and MRI can complement the physical examination in selected situations.

One must elucidate why the patient and/or family are seeking medical care for their deformity. Is the patient simply bothered by the appearance of the extremity or is the patient having pain? Often families seek care for growing children because of concerns not about the current state of the limb but because of concerns that the deformity will worsen or that pain or disability will result at a later time. It is important to engage the patient and caretaker regarding the natural history of the deformity and counsel them as to how the surgical correction may or may not address their concerns. Occasionally the patient or caretaker may be seeking care for a complaint that is only minimally or not related to the patient's deformity.

One must also evaluate the social situation of the patient as well as the psychological issues and family dynamics when planning correction of a deformity. In particular, the surgeon should take into account the ability of the patient and family to comply with a proposed treatment regimen. For example, gradual correction of deformities utilizing a circular external fixation system in a patient with autism might be ill advised. The ability of patients and families to comply with postoperative protocols should be evaluated prior to finalizing a surgical plan and prearrangements for the anticipated need for physical therapy and follow-up visits may be necessary in certain situations. The surgeon's skill set and available medical resources should also be taken into account when developing a plan. Relevant cultural norms should also be considered and may dictate pursuing reconstructive options that might otherwise be contraindicated and lead the surgeon and patient to accept perhaps less function in order to be more culturally acceptable. For example in some parts of the world there is a strong cultural bias against accepting amputation as part of what otherwise might be a preferred treatment.

Table 3.1 Important elements of a problem list to describe in planning a lower extremity deformity correction

Uniapical deformity versus multiapical deformity
Coronal, sagittal, and rotational parameters of each deformity
Soft-tissue envelope/vascularity of bone
Patient/caretaker perception of problem
Patient comorbidities
Prior treatment successes and challenges

The Problem List

The history, physical examination, imaging studies, patient and family perceptions of the problem, and dynamics of the situation should be brought together by the surgeon to form a problem list (Table 3.1). This list should summarize the deformities, pertinent pathology, and comorbidities as they impact the treatment of the deformity, patient perceptions of the problem, and potential obstacles to treatment whether they are related to the patient or the environment in which treatment is occurring. It is important to remember that the problem list is simply a problem list and not necessarily a surgical agenda.

After a problem list has been developed, the anatomy and biology of the underlying deformity must be considered. In particular the surgeon should decide whether the location of the deformity is more amenable to acute or gradual correction based on the anatomy of the neurovascular structures and postoperative or posttraumatic scarring. Are the physes open, leaving the option of growth modulation to correct the deformity? The presence of active or prior infection is another important factor to be considered, as are factors such as the presence of a pseudarthrosis.

When considering the problem list in conjunction with the anatomy and biology of the problem, the surgeon should be able to outline options for an appropriate treatment agenda for addressing the deformity comprehensively over time. Some of these plans may be relatively simple comprising an acute correction of a uniapical deformity. Other cases may require a much more complex plan, including a series of interventions over a number of years such as for an infant with proximal femoral focal deficiency.

Surgical Indications: General

The decision about whether or not to correct a given deformity can be quite individual for each patient and there are no absolute rules. Relative indications for surgical correction of a deformity include the presence of pain or a deformity

Table 3.2 Relative indications for surgical correction of lower extremity deformity

Persistent pain
Mechanical axis in zone 2 with symptoms
Mechanical axis in zone 3 or greater with or without symptoms
Uncompensated symptomatic hindfoot deformity
Sagittal plane deformities impeding gait and function

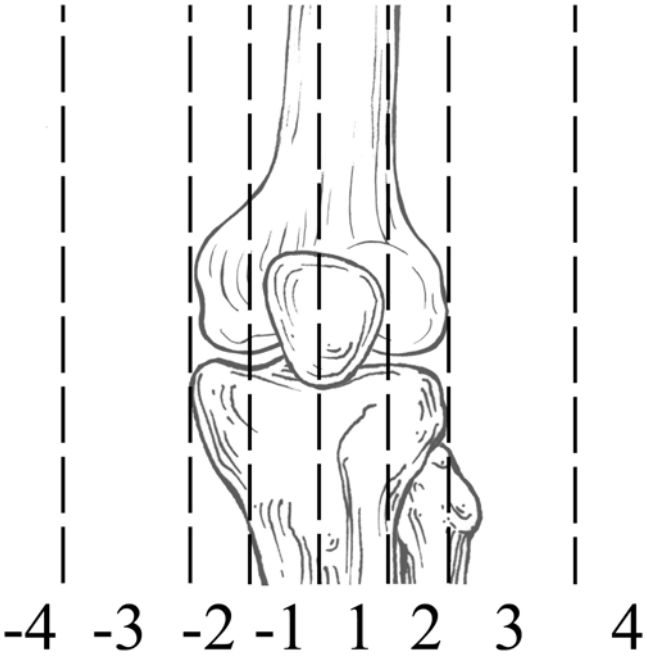


Fig. 3.1 Depiction of zone of mechanical axis deviation at the knee. Zone 1 is within the tibial spine. Zone 2 is within the tibial condyles. Zone 3 is within the knee joint width away from the center of the knee joint. Zone 4 is greater than one knee joint width from center of knee joint [22]

with a natural history of substantial progression (Table 3.2). In general, current or anticipated limb length discrepancies greater than 2 cm should be addressed.

Surgical Indications: The Knee

Angulatory deformities about the knee should generally be corrected if the mechanical axis falls within zone 2 and the patient is symptomatic and should be corrected if the mechanical axis is beyond zone 2 (Fig. 3.1) even if the patient is asymptomatic. When the deformity about the knee requires correction, the joint orientation angles such as the lateral distal femur angle (LDFA), the medial proximal tibial angle (MPTA), and the joint line congruency angle (JLCA) should be measured. If the JLCA is more than 5° in conjunction with a bony deformity, we prefer to address the ligamentous laxity as well. If the deformity about the knee involves only the femur or the tibia and the other bone is normal, correction should nearly always occur within the affected bone. If both bones are abnormal but the majority of the deformity is within one of the bones, there may be some consideration to correcting only the bone with the majority of the deformity if less than 5° of abnormality is present in either the LDFA or MPTA. If greater than 5° of abnormality is noted in both the LDFA and MPTA, we prefer to address the deformity at both sites.

For sagittal plane deformities about the knee, one must consider both the bony deformity within the distal femur and proximal tibia as well as the soft-tissue constraints about the knee joint. The goal of treatment of sagittal deformity about the knee is functional range of motion in both extension and flexion. My preference is to develop a surgical plan that achieves knee extension within 5° of full extension and without hyperextension of greater than 5° as well as knee flexion of at least 90°. For patients with limited extension, one must consider concomitant hamstring, iliotibial band, or posterior capsule releases while correcting the bony sagittal deformity. Likewise, one can consider quadriceps lengthening to augment a deformity correction in a patient with limited knee flexion.

Surgical Indications: The Ankle

The decision to correct ankle alignment must be made in conjunction with a careful examination of the hindfoot. Patients with substantial deformity of the ankle may be clinically well aligned with a compensatory hindfoot deformity (Fig. 3.2). Patients with such well-compensated deformity and with limited subtalar motion may not be well served by a decision to correct the ankle and thus "create" a more visible deformity by uncovering the fixed hindfoot abnormality.

Relative Contraindications

Contraindications to surgery are necessarily somewhat vague and include a number of patient-related factors including unrealistic expectations on the part of the patient and caretakers, their inability to follow through with the necessary outpatient components of a treatment program, as well as situations where the risks of surgical intervention outweigh the potential benefits to the patient (Table 3.3). For instance, patients with severe mental illness or with limited ability to comprehend the treatment plan may not have the ability to comply with more complex treatment protocols including gradual correction using external fixation. Although more complex treatment protocols may be contraindicated in these patients, simplified alternatives may represent better choices. For example, for some patients the

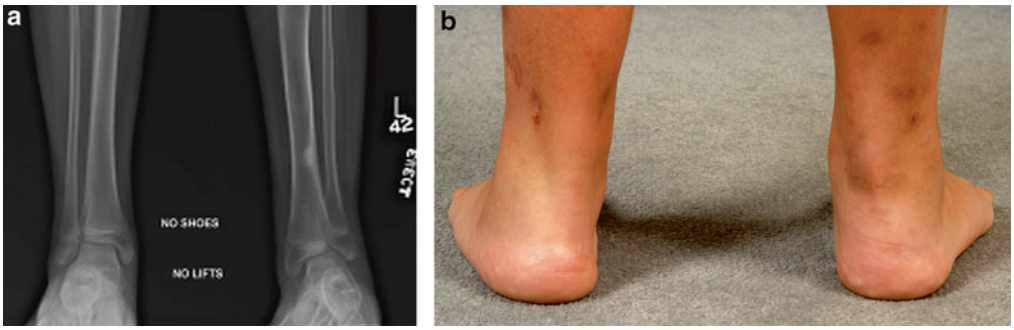


Fig. 3.2 Radiographs (**a**) and clinical images (**b**) of a patient with distal tibial valgus and compensatory hindfoot varus. Note the radiographic valgus of the left ankle and the apparent normal clinical alignment of the left hindfoot

Table 3.3 Relative surgical contraindications to performing lower extremity deformity correction

Unrealistic patient/family expectations
Patient/family unable to comply with postoperative protocol
Potential complications of treatment outweigh benefits to patient

difficulties involved in complying with instructions and a physical therapy program associated with a limb lengthening may be able to comply easily with the less strenuous instructions and therapy associated with a closed femoral shortening (Fig. 3.3).

Surgical Options

The plan for each deformity may comprise any of a number of different techniques that might be applicable in a given situation (Table 3.4). There is no compelling reason to avoid combining techniques when clinically applicable. General categories that should be considered include soft-tissue surgery, physeal bar resection, growth modulation, acute correction with internal or external fixation, or gradual correction with external fixation. A brief description of the techniques follows with a general description of indications and contraindications.

Soft-Tissue Modification

Soft-tissue surgery can be effective either as a solitary procedure or in conjunction with bony surgery. Modification of the soft tissues is not limited to procedures that directly approach the soft tissues themselves but can also be an intended consequence of bony procedures. In particular, tightening of the lateral collateral ligament at the knee can be performed by translating the fibular head distally either with a fibular osteotomy and gradual bone transport with an external fixator or in cases of adolescent tibia vara utilizing a circular external fixator. In these cases a tibial osteotomy can be performed while leaving the fibula intact and securing it distally to the tibia. The angulatory deformity is then corrected while lengthening gradually, thus transporting the fibula distally in relation to the proximal tibia resulting in tightening of the lateral collateral ligament (Fig. 3.4).

Physeal Modulation or Ablation

Physeal modulation can take many forms. Physeal bar resection following traumatic or developmental partial arrest can be performed successfully in situations where a partial arrest of the physis, as documented by progressive deformity, is present that involves less than 50 % of the physis with more than 2.5 cm of growth remaining at the involved physis and more than 2 years of growth expected [2, 3]. Advantages of physeal bar resection include the relative simplicity involved in postoperative care and relatively rapid patient recovery and return to activity (Fig. 3.5). Disadvantages of physeal bar resection include a high failure rate and frequent late closure of the physis necessitating close follow-up and further surgical treatment.

Complete epiphysiodesis by percutaneous drilling or utilizing either plates or staples can be performed in skeletally immature individuals who have adequate growth remaining to contribute to the correction of a limb length discrepancy [4]. This has been recommended in situations where the projected limb length discrepancy is between 2 and 5 cm. The technique however can be utilized in larger limb length discrepancies as part of a comprehensive treatment plan. Temporary hemiepiphysiodesis using medial and lateral staples or plates has been reported [5] but is not widely utilized due to concerns about permanent physeal closure and possibility of rebound growth following implant removal.

Hemiepiphysiodesis by plate, screw, drilling, or Blount's staples should be considered in patients who are skeletally immature and who have adequate growth available to reasonably expect correction of an angulatory deformity [6].



Fig. 3.3 Femoral shortening case study. (**a**) Preoperative radiographs of a 17-year-old female with a 7 cm left greater than right limb length discrepancy due to a right-sided posttraumatic femoral growth arrest. (**b**) Intramedullary saw completing distal cut within femur. (**c**) Intramedullary saw beginning proximal cut within femur 7 cm proximal to distal cut. (**d**) Completion of proximal intramedullary cut. (**e**) Intramedullary wedge splitting the medial cortex of the intercalary segment. (**f**) Completion of splitting intercalary segment. (**g**) Reduction of proximal and distal segment with solid intramedullary nail. (**h**) Postoperative radiographs demonstrating healed femur after closed femoral shortening

These deformities can be in the coronal, sagittal, or oblique planes (Fig. 3.6). Physeal modification can be utilized even in situations where the physis is abnormal [7] such as chronic renal failure and hypophosphatemic rickets. Deformities such as proximal tibial valgus following a Cozen's-type fracture which tend to recur following osteotomy correction are particularly amenable to treatment with plate hemiepiphysiodesis. Following correction of the deformity in a patient who has substantial growth remaining, the metaphyseal screw can be removed percutaneously and the plate and epiphyseal screw can be left in place allowing continued growth (Fig. 3.7). If the deformity recurs, the metaphyseal screw can be replaced percutaneously resulting in re-hemiepiphysiodesis and repeat correction.

Table 3.4 Available techniques to perform deformity correction

Soft-tissue modification
Physeal modulation or ablation
Osteotomy with acute correction
Osteotomy with gradual correction

Acute Correction with Osteotomy

Osteotomies with acute correction are particularly indicated in angulatory deformities in patients with minimal limb length discrepancies allowing correction of the deformity without placing undue stress on the neurovascular structures. Most often the acute correction is combined with rigid internal fixation although it can be utilized in conjunction with external fixation or Kirschner wire fixation and cast immobilization. The choice of implants is determined by the site of the osteotomy and can potentially include intramedullary fixation, screw fixation, locking plates, or non-locking plates.

Intramedullary fixation can be helpful when combined with percutaneous diaphyseal osteotomy techniques such as in patients with an isolated femoral rotational deformity [8, 9]. Intramedullary stabilization can also be helpful when angulatory deformities exist in the diaphyseal region after either traumatic malunion or in cases of hypophosphatemic rickets with residual femoral lateral bow (Fig. 3.8). In these cases often the percutaneous osteotomy can correct both the apparent genu varum deformity and the coxa vara that



Fig. 3.4 Case example of adolescent tibia vara treated with circular external fixator and tightening of the lateral collateral ligament. (**a**, **b**) AP and lateral preoperative radiographs of a 12-year-old male with tibia vara. (**c**, **d**) Immediate postoperative radiographs depicting a ring circular external fixator with fixation of the fibula distally into the tibia and no proximal transfibular fixation. (**e**) Postoperative radiographs after fixator removal showing distal transport of the fibula relative to the proximal tibia, thus tightening the lateral collateral ligament



Fig. 3.5 Case example of a 9-year-old female with a posttraumatic physeal bar formation in the right distal femur. (**a**) Standing AP demonstrating right distal femur physeal bar. (**b**, **c**) Coronal and sagittal slice of a CT scan showing lateral distal femoral bar with less than 20 % of physis involvement. (**d**) Fluoroscopic image showing curettage of physeal bar. (**e**) Postoperative AP after physeal bar resection. K-wires are placed in order to follow eventual growth; radiolucent cranioplast is placed on the epiphyseal side to limit reformation of bar. (**f**) Immediate postoperative standing AP. (**g**) Standing radiographs demonstrating growth within the distal femoral physis 2 years postoperatively. (**h**) Standing anteroposterior radiograph demonstrating continued growth with migration of the cranioplast proximal to the physis with resultant recurrence of the physeal bar associated with a limb length discrepancy and valgus deformity 3 years postoperatively

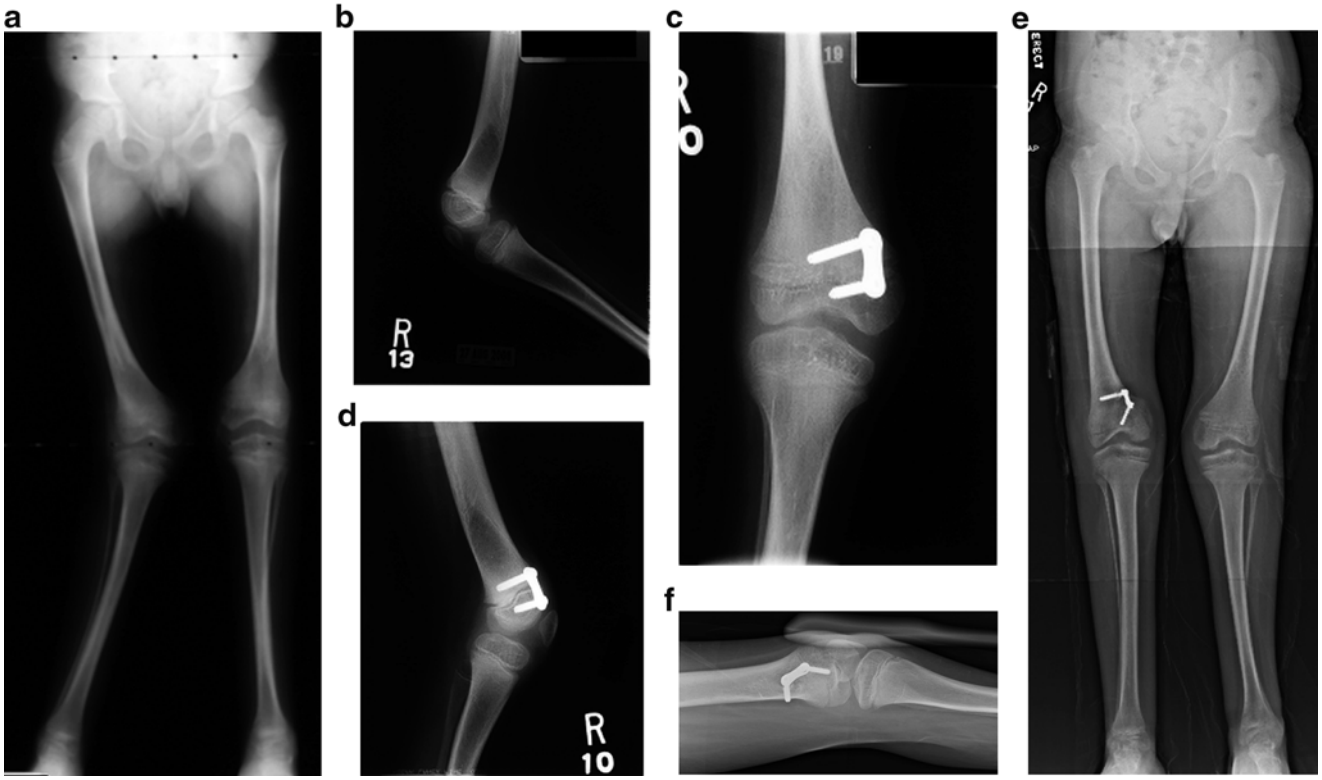


Fig. 3.6 Oblique plane deformity of the right distal femur in a 6-year-old male with arthrogryposis. (**a**) Standing AP demonstrating right distal femoral valgus. (**b**) Lateral of the right knee in less than maximal extension showing a knee flexion contracture of approximately 25°. (**c**, **d**) AP and lateral of the knee showing hemiepiphysiodesis of the anteromedial distal femur, promoting distal femoral varus and procurvatum. (**e**, **f**) Standing AP and maximal extension lateral views 18 months postoperatively with resultant correction of deformity. The patient had full knee extension

can produce a significant abductor lurch during gait. The disadvantage of intramedullary fixation is the limitation in the magnitude of the correction that can be obtained. Intramedullary stabilization results in a comparatively straight diaphyseal segment when combined with a diaphyseal osteotomy. Intramedullary stabilization can also be utilized with blocking screws in conjunction with acute correction of a supracondylar distal femoral or proximal tibial osteotomy but this use is technically difficult for the surgeon who is not familiar with the use of such screws. Proper mechanical alignment of the proximal and distal segments can also be obtained with temporary external fixator assistance prior to intramedullary reaming and fixation with an intramedullary nail. Other contraindications to intramedullary fixation include open physes with substantial remaining growth in the proximal tibia and the inability to pass an intramedullary nail thorough an obstructed intramedullary canal. In addition intramedullary nail should be generally avoided in the presence of active infection.

Screw fixation provides the surgeon with the ability to stabilize osteotomies with some inherent bony stability while achieving rigid correction of the deformity. Cannulated screws are particularly helpful when performing osteotomies in the epiphyseal region that require rigid control of the osteotomy. One specialized use of these screws is to stabilize intra-articular osteotomies in the proximal tibia. This can be performed in cases of medial tibial plateau depression due to delayed treatment of early-onset tibia vara with a resulting "pagoda" deformity [10]. These tibial plateau osteotomies can be effectively stabilized after an acute correction using cannulated screws with an incomplete osteotomy and allograft cortical bone as a structural graft (Fig. 3.9).

Plates provide a rigid method of stabilizing osteotomies following an acute correction. They are particularly useful in stabilizing metaphyseal osteotomies after correction of either angulatory or rotational deformities. Plates utilized can be conventional plates, locking plates, or blade plates [11]. In particular, blade plates and locking plates (Fig. 3.10) can be used to stabilize osteotomies that have little or no inherent stability and locking plates can be utilized with a submuscular technique that may minimize soft-tissue dissection. Advantages to their use include the ability to begin early motion at adjacent joints, rigid fixation, and immediate correction of the deformity (Fig. 3.11). Disadvantages to the use of conventional plates are the relatively larger amount of soft-tissue dissection that is necessary, the lack of ability to

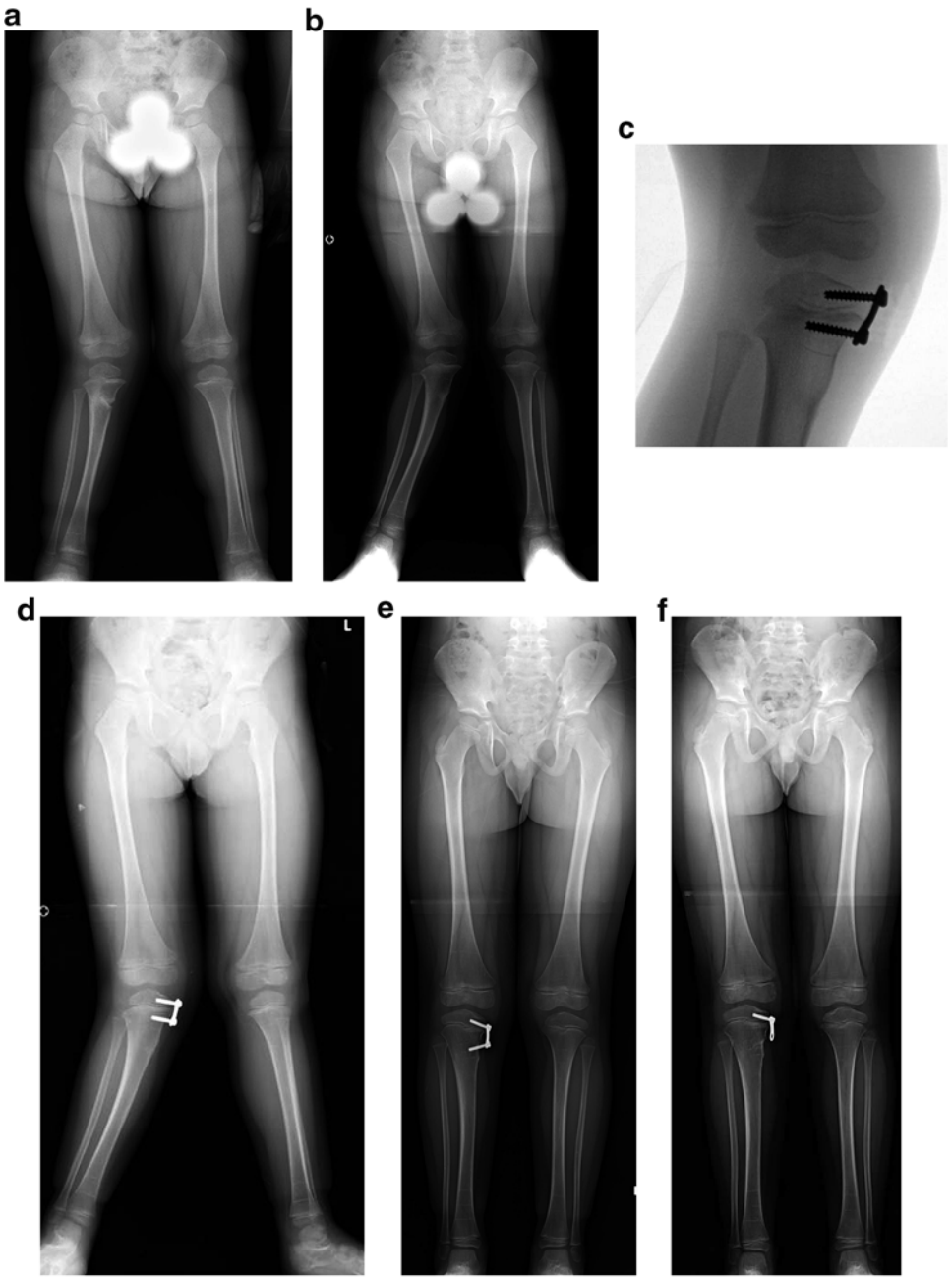


Fig. 3.7 Four-year-old male with a proximal tibial valgus deformity after proximal tibia fracture (Cozen's fracture). (**a**) Standing AP after cast treatment for right proximal tibia fracture. (**b**) 6-month postinjury film demonstration proximal tibial valgus deformity. (**c**) Fluoroscopic image showing placement of hemiepiphysiodesis plate medially. (**d**) Immediate postoperative standing AP. (**e**) 9-month postoperative radiographs showing normal mechanical axis. (**f**) Metaphyseal screw is removed after deformity correction in order to facilitate replacement of hemiepiphysiodesis if deformity recurs

adjust the correction postoperatively, and the difficulties associated with soft-tissue coverage in areas with little overlying soft tissue with resulting issues with infection.

Gradual Correction with Osteotomy

The workhorse of complex deformity correction is gradual correction. Gradual correction is effective in treating deformities with components of angulation, rotation, translation, and/or length. Gradual correction can be achieved with monolateral external fixation, circular external fixation utilizing the classic Ilizarov-type hinges and distractors, or computer-controlled six-strut circular external fixation. Most recently, self-lengthening nails have combined some of the advantages of gradual correction with internal fixation [12]. Self-lengthening nails however provide no ability to correct angulatory deformities gradually but do allow for acute correction of angulatory deformities with gradual lengthening. Advantages of gradual correction include minimal soft-tissue dissection and early weight bearing with the ability to deal with problems that arise during the postoperative period and

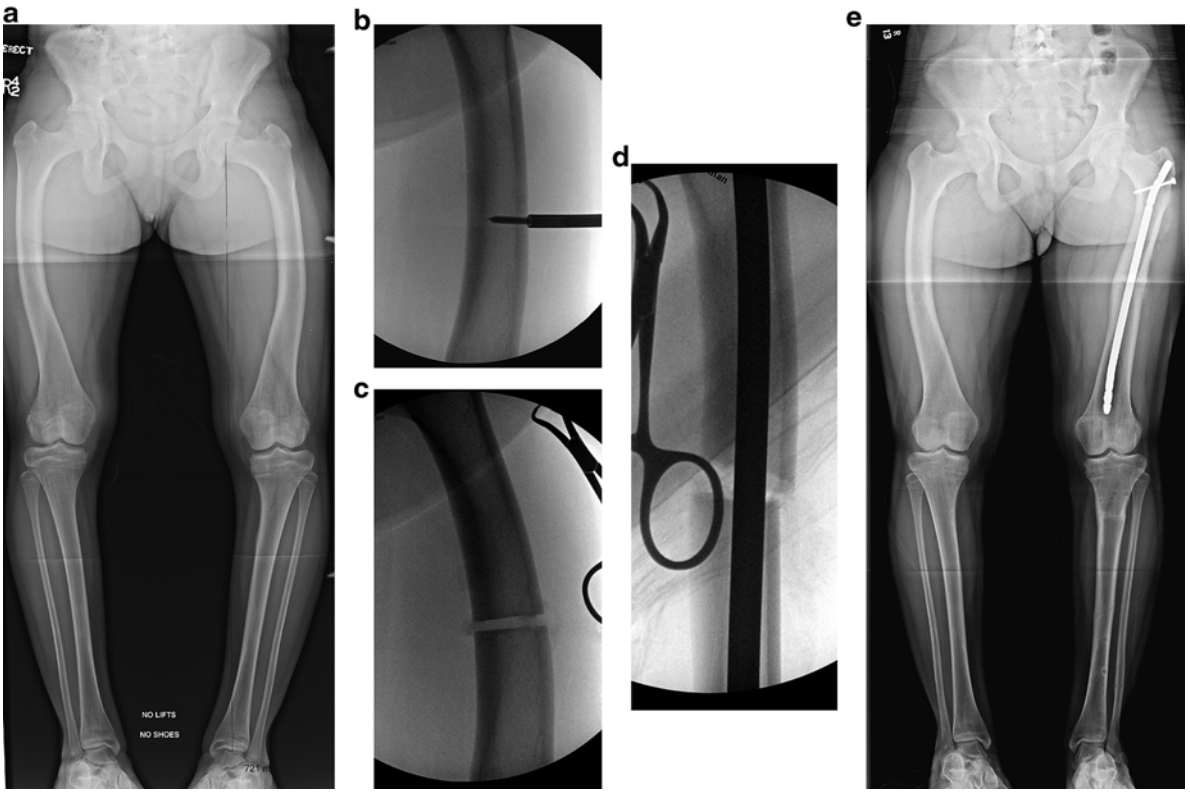


Fig. 3.8 Use of intramedullary nail for deformity correction. (**a**) Standing AP of 12-year-old female with X-linked hypophosphatemic rickets and midshaft bowing of both femurs at the isthmus. (**b**) Fluoroscopy demonstrating percutaneous diaphyseal femoral osteotomy using drill. (**c**) Completion of diaphyseal femoral osteotomy. (**d**) Insertion of pediatric femoral nail. As the osteotomy was performed at the isthmus and deformity, this results in an opening wedge osteotomy medially. (**e**) Postoperative standing AP demonstration correction of right femoral deformity. A proximal tibial osteotomy with gradual correction using a circular external fixator was used to correct the tibial deformity

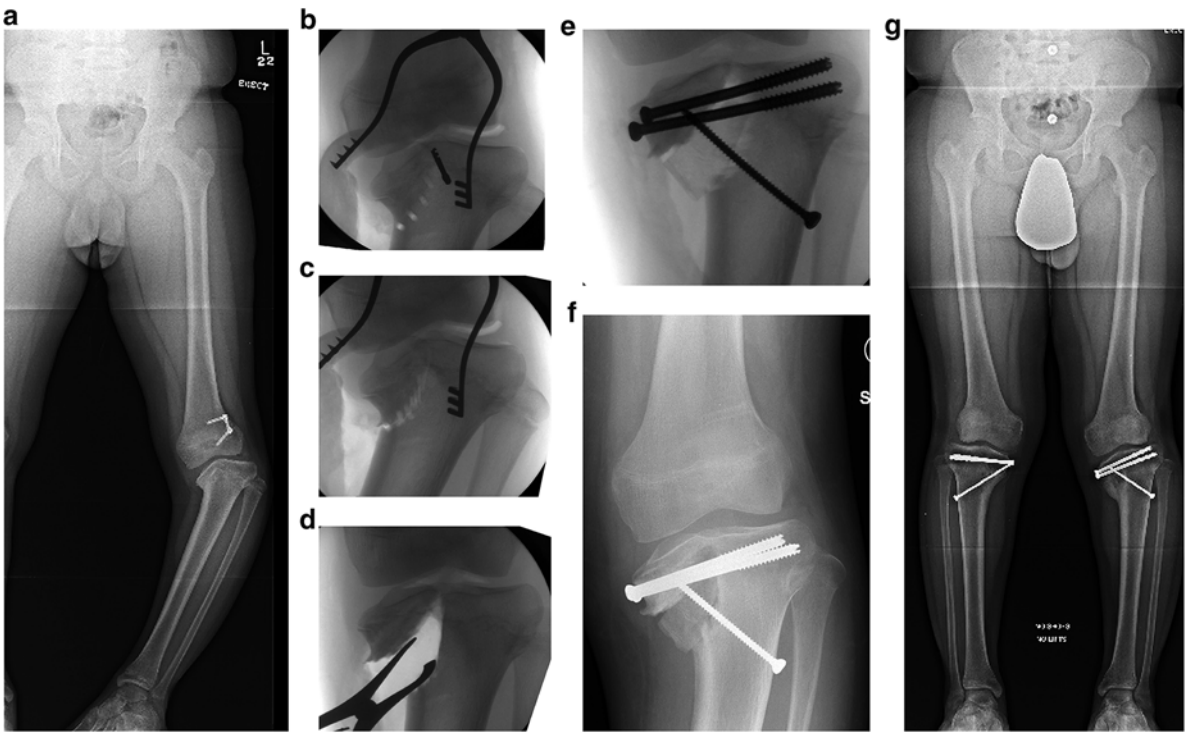


Fig. 3.9 Twelve-year-old male with Blount's disease and proximal tibia intra-articular deformity. (**a**) Standing AP demonstrating intra-articular proximal tibial deformity and medial growth arrest (Langenskiold type VI). (**b**, **c**) Fluoroscopy demonstrating drill holes along the path of intra-articular osteotomy for tibial plateau elevation, completion of osteotomy with an osteotome. (**d**) Laminar spreader simulating correction of intra-articular deformity. (**e**, **f**) Intraoperative fluoroscopy and postoperative radiographs demonstrating fixation with large fragment screws after structural bone graft placement. (**g**) Healed osteotomy sites with persistent proximal tibial varus. Tibial plateau elevation corrects intra-articular deformity but often does not correct overall varus deformity completely

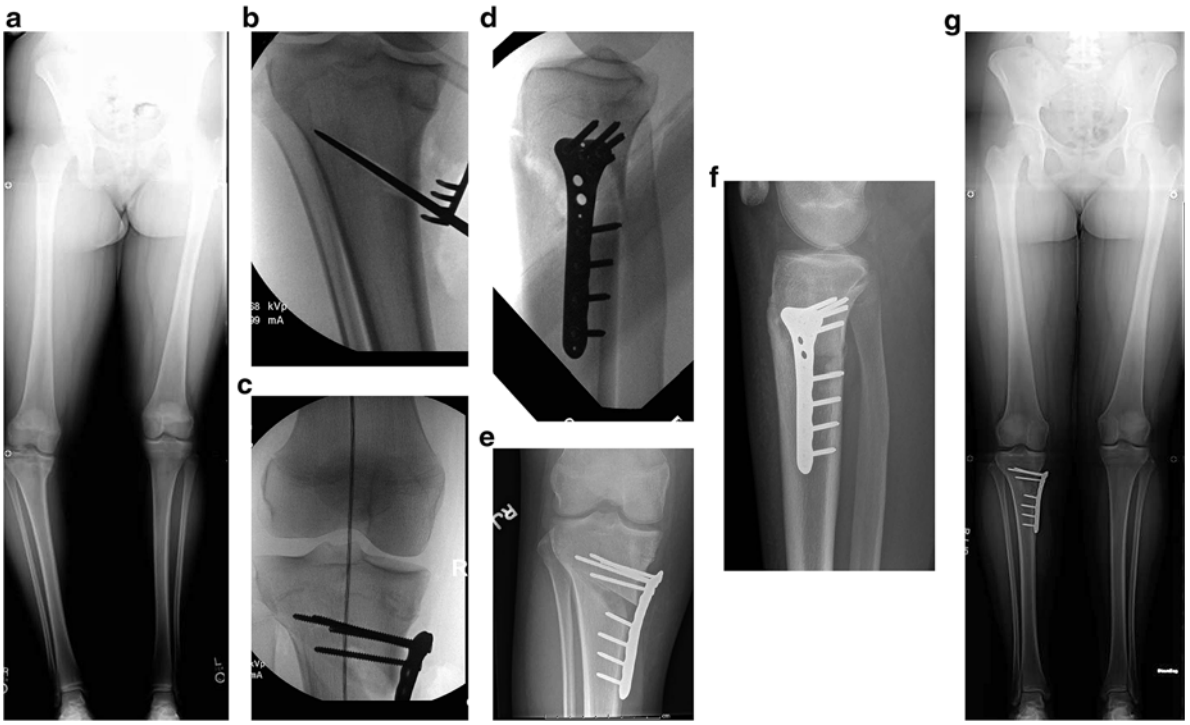


Fig. 3.10 Fourteen-year-old female with Blount's disease. (**a**) Standing AP demonstrating right proximal tibia varus deformity. (**b**) Oblique osteotomy heading from distal medially to proximal laterally with care to preserve lateral cortex. (**c**) Fluoroscopic image demonstrating restoration of mechanical axis. (**d**) Lateral fluoroscopic view. (**e**, **f**) Postoperative AP and lateral view showing correction of deformity. (**g**) 18-month postoperative standing AP demonstrating deformity correction

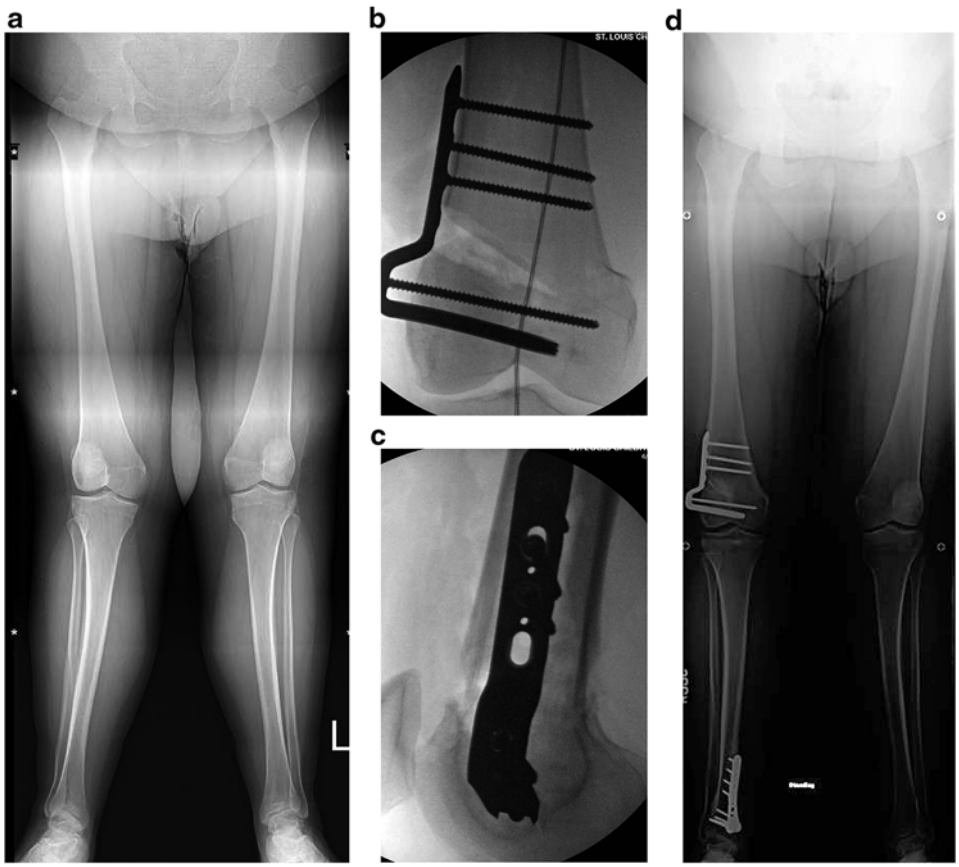


Fig. 3.11 Sixteen-year-old female with distal femoral valgus deformity. (**a**) Standing AP demonstrating distal femoral valgus deformity. (**b**, **c**) AP and lateral fluoroscopic images showing distal femoral osteotomy with blade plate correction of deformity. (**d**) Postoperative radiographs demonstrating restoration of mechanical axis. Of note, patient had a concomitant derotational distal tibial osteotomy for excessive external tibial torsion



Fig. 3.12 Thirteen-year-old female with tibial pseudarthrosis, status post-multiple attempts at correction and healing with Peter Williams rod. (**a**, **b**) AP and lateral of the right tibia demonstrating deformity and occurrence of a new distal tibial pseudarthrosis. (**c**, **d**) AP and lateral views of Ilizarov application with multilevel osteotomy, proximal osteotomy for tibial lengthening, and distal tibial osteotomy for deformity correction and bone transport into pseudarthrosis site. (**e**, **f**) AP and lateral views after frame removal and pseudarthrosis healing

the ability to "fine-tune" the correction to optimize final alignment [13–16]. Disadvantages of gradual correction include complex postoperative care requiring substantial compliance with a physical therapy protocol and rigid adherence to a follow-up schedule by the patient and family with the possibility of damaging adjacent joints by the injudicious use of distraction at an osteotomy site. In addition, external fixation systems tend to be disliked by patients and issues with pain and pin tract problems are often noted [17–21]. As the lower limb deformities become more complex, the degree of required patient compliance and the physician learning curve also increase substantially. The trade-off, however, is the increasing ability to deal with complex three-dimensional deformities and achieve accurate corrections (Figs. 3.12 and 3.13).

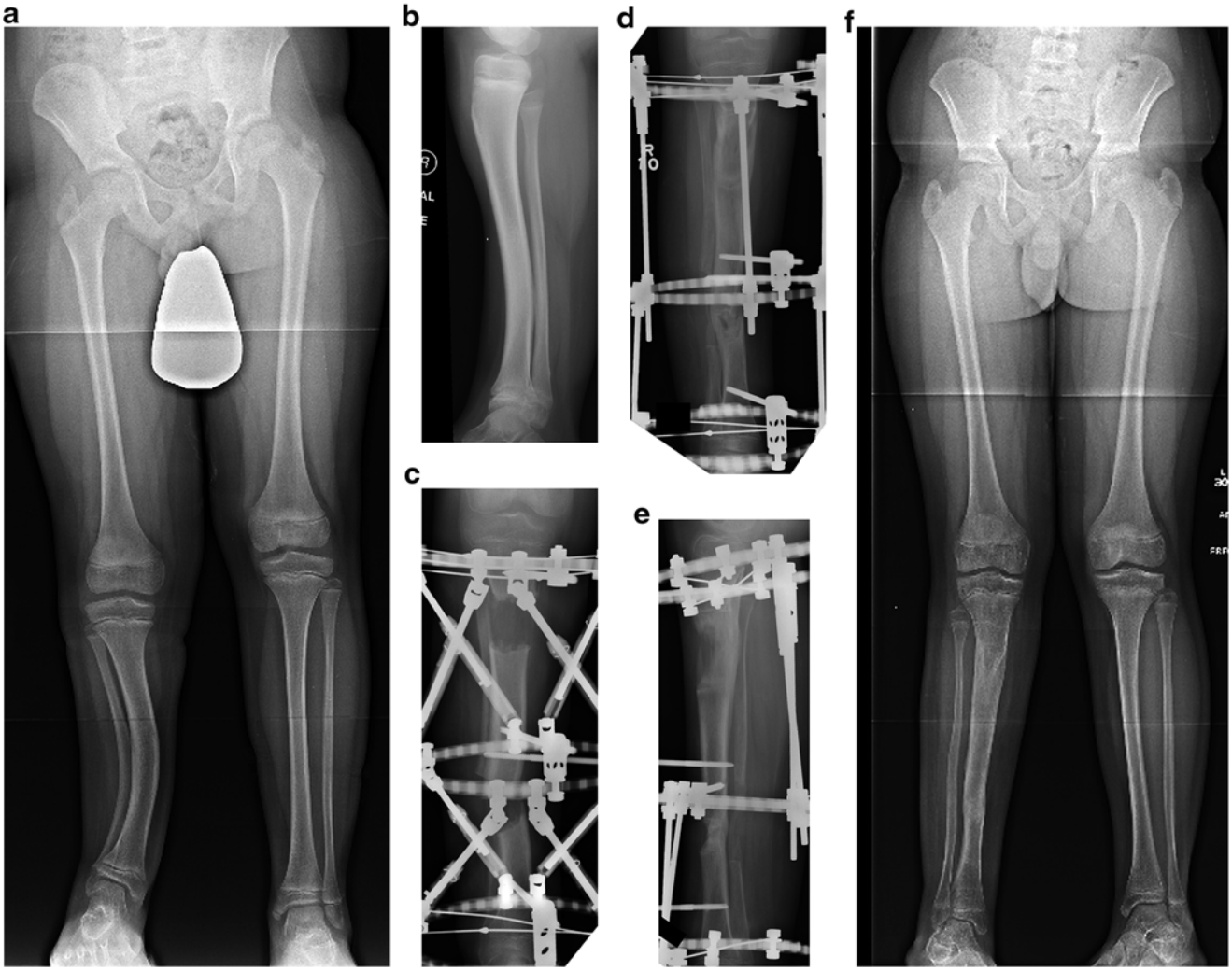


Fig. 3.13 Ten-year-old male with posteromedial bowing and limb length discrepancy. (**a**, **b**) Standing AP and lateral view demonstrating deformity and limb length discrepancy. (**c**) AP radiograph showing application of bi-level circular external fixator. A proximal tibial osteotomy was performed for lengthening and a midshaft osteotomy for deformity correction. (**d**, **e**) AP and lateral view demonstration of healing of regenerate and correction of deformity. (**f**) Standing AP showing restoration of mechanical axis and correction of leg length discrepancy

Summary

In summary, decision making in lower extremity deformity correction is a necessary but arduous task. One must develop a complete problem list which addresses the pertinent difficulties associated with the patient's deformity, perceived problem, and ability of the caretaker and patient to implement the plan of care. This problem list then should help guide the location of the correction and select which mode or combination of modes of deformity correction is optimal, whether it be soft-tissue modification, physeal modulation/ablation, or osteotomy though either an acute or gradual approach. The approach(es) selected should then guide the choice of implant. With advances in technology devices such as self-lengthening nails and computer-controlled six-strut circular external fixators have become more readily available and accessible, and the complexity of deformities that can safely be addressed has increased dramatically. It is still important to start with a comprehensive evaluation of each patient and his or her environment and apply the principles of deformity correction to develop a customized treatment plan.

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Growth Modulation for Angular and Length Correction

4

Peter M. Stevens

Introduction

As pediatric orthopedists, we are often consulted to evaluate and manage evolving deformities of the lower extremities that, left untreated, may have lifelong, adverse impacts upon gait and the spine. Generally speaking, the era of "corrective shoes," orthotics, and bracing has yielded to surgical management, because the natural history of pathological deformities is inexorable progression. In 1933, Phemister introduced the concept of open epiphysiodesis for the management of angular deformities and limb length inequality [1]. With the advent of fluoroscopy, this method evolved to the comparatively less invasive percutaneous growth arrest [2–4]. However, the acknowledged drawback of such ablative methods is that they are permanent and, hence, irreversible. Therefore, open or percutaneous epiphysiodesis must be delayed until adolescence and perfectly timed, in order to avoid over- or under-correction of a given deformity. The vagaries of determining skeletal maturity and of parental compliance and timely follow-up may make this goal optimistic [5]. The same drawbacks pertain to other experimental methods of chemical, electrical, or radiofrequency ablation, which are permanent and irreversible.

Given these limitations many still consider osteotomy to be the "gold standard" for progressive angular deformities such as Blount's disease, rickets, and idiopathic genu valgum. Some feel compelled to cut bone(s) in order to correct associated torsional deformity. However, the potential disadvantages of osteotomy include blood loss, neurovascular compromise, immobilization, over- or under-correction, and malunion or nonunion [6]. Comparative cost, cast immobilization, weight-bearing restrictions, unsightly scars, and difficult hardware retrieval must be considered as well.

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The appeal of temporary, reversible, epiphysiodesis is that, while being more cost effective, it circumvents the above list of iatrogenic problems. Haas was the first to document this, using a wire loop, initially in dogs and eventually in human subjects [7, 8]. Soon thereafter, in 1947, Blount introduced his staple for correction of angular deformities and limb length inequality and this technique was widely practiced for decades [9]. Because the physis is dynamic and the forces of growth so powerful, there was a propensity for staples to eventually (or precipitously) bend, break, or migrate (Fig. 4.1). The use of multiple staples failed to consistently address these problems. Furthermore, staple removal often proved challenging, sometimes resulting in local bone loss that could compromise the reversibility of this technique. This placed doubt upon the wisdom and necessity of using such rigid implants; their popularity has therefore declined.

Frustrated by some of the aforementioned problems with stapling, the concept of using an extra-periosteal, non-locking plate, serving as a tension band, was developed in 2002, and introduced in 2004 [10, 11]. Since then, similar implants have been introduced into the market, worldwide (Fig. 4.2). It is not critical which implant one selects; rather it is the tension band principle that counts. However, one should avoid locking plates because the rigid construct will predispose to implant failure.

In resource-challenged regions of the world, the 8-plate or facsimiles may not be available. In such an environment, an option is to take a 1/3 semi-tubular plate or a pelvic reconstruction plate and cut it into 2-hole segments [12–14]. The potential disadvantages relate to the relative bulkiness or stiffness (will not reverse bend) when compared to the real device. Some favor the use of quad plates (4-hole) in heavy children, particularly with Blount's disease. Inevitably, a variety of plate shapes have been proposed (step-off, hinge plates, H-plates) and the controversy is unresolved as to whether titanium or stainless steel implants are superior. Unfortunately, there is currently no literature to support or refute these ideas. Suffice it to say that any device that is flexible and permits differential growth can work.

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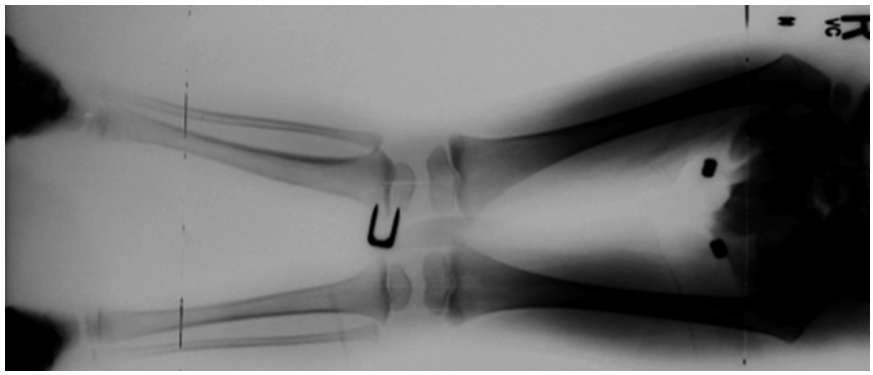


Fig. 4.1 A 4-year-old girl, with posttraumatic tibia valga (Cozen's phenomenon), treated by stapling. Rigid implants are subject to failure through bending, breakage, or migration, as a result of the inexorable forces of physeal growth. Note the extrusion of her single staple; occurring inexplicably, 9 months following implantation



Fig. 4.2 Since the introduction of the 8-plate in 2004, many facsimiles have been produced worldwide and used successfully for guided growth. The common denominators for success are low profile and non-locking

An alternative strategy, popularized by Metaizeau, involves the insertion of large-diameter (6.5 or 7.3 mm) cancellous screws across one or both sides of the physis (percutaneous epiphysiodesis using transphyseal screws, or PETS) [15]. While also minimally invasive, the theoretical objection is the necessity of violating the physis with a large threaded implant. Series reports of PETS are largely confined to the adolescent age group [16]. It has not been described in younger children or in sagittal or oblique plane deformities. Thus, PETS has comparatively narrow ranges of age and applications and its potential reversibility remains in question.

Deformities of the Lower Extremities

General

With the notable exception of physiologic varus and valgus, lower extremity deformities often progress with growth. While angular deformities at the knee are most obvious and perplexing, one must examine the patient for limb length discrepancy, rotational deformities, and ligamentous laxity that represent confounding factors. Also, examine the hip and ankle clinically and, when indicated, radiographically, for coexisting or compensatory angular deformities. If the deformity is unilateral, then the normal leg serves as a comparison. However, oftentimes deformities are bilateral and not necessarily symmetrical. One must be cognizant of remaining growth and the predicted natural history of a given problem. If a deformity is thought to be physiologic or borderline in severity, a follow-up after 6 months may clarify this and affect the timing of intervention. If there is concurrent deformity of the hip and/or ankle, simultaneous guided growth may be employed [17].

Angular: Frontal

Physiologic

It is well recognized that children under the age of two may present with symmetrical and painless bowing of the legs, associated with ligamentous laxity and intoeing. With observation alone, all components of this deformity will spontaneously correct, without treatment. While there is no need for intervention, parental education, reassurance, and periodic follow-up are tantamount to success. Likewise, physiologic genu valgum should be managed expectantly, without bracing, physical therapy, or shoe modification, up until the age of 6. These self-limiting conditions are a contraindication to surgical intervention of any kind [18–20].

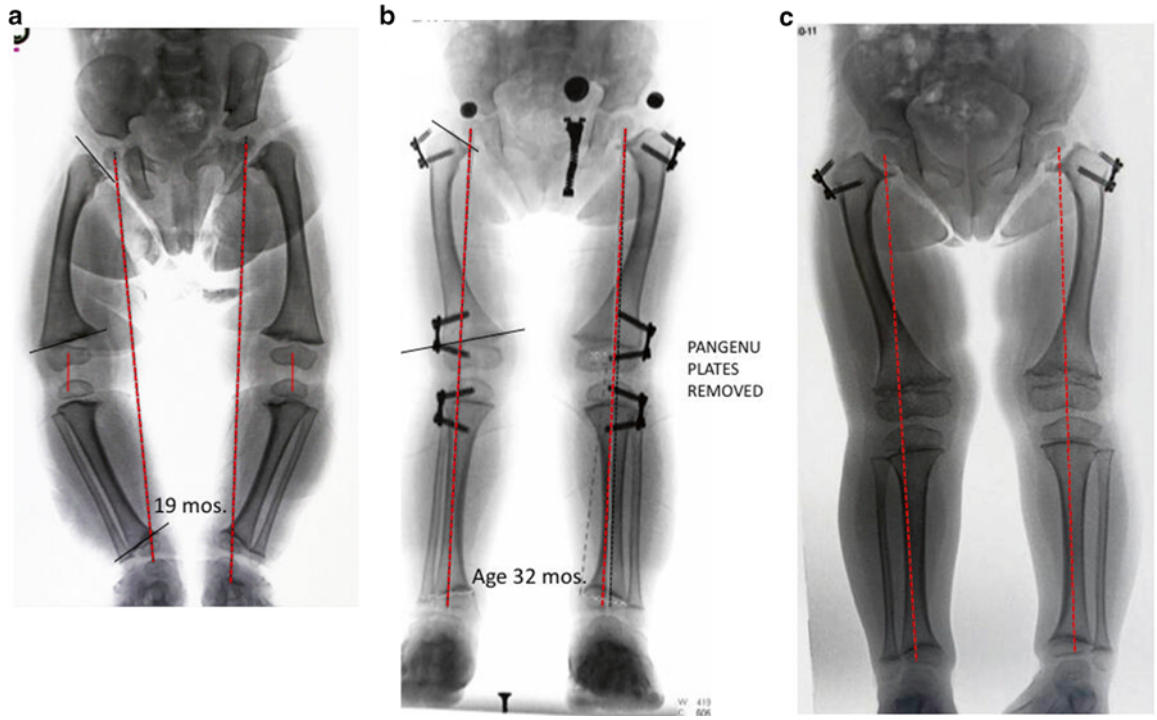


Fig. 4.3 (**a**) This 19-month-old girl with Schmid-type chondroepiphyseal dysplasia, presented with a waddling gait and hip pain (due to stress fractures of the femoral neck). (**b**) 13 months following bilateral, 3-level guided growth (outpatient/no cast), her mechanical axis neutral; her gait has become normal and hip pain abated. (**c**) At age 6, she remains asymptomatic, with neutral alignment. Note the indirect resolution of her ankle varus. Her femoral neck fractures remain healed; the plates will be left in situ, pending further growth. Proximal femoral osteotomies appear to be unlikely

Pathologic

In contradistinction to the above-noted problems, the pathologic deformities may be unilateral or bilateral and multilevel. The etiology may be idiopathic, developmental (Blount's), acquired (Cozen's), metabolic (rickets), genetic (neurofibromatosis, hereditary multiple exostoses, Schmid dysplasia), or dysplastic (fibrous dysplasia, Ollier's), to name a few. Deformities may occur in the frontal, sagittal, or oblique plane, compounded by torsion and ligamentous laxity. They are insidious in their progression, and refractory to attempts at orthotic management or physical therapy.

Regardless of the underlying etiology, individually and collectively these deformities constitute the indication for guided growth, as the treatment of choice, instead of osteotomy (salvage procedure). Clinically, genu valgum is often associated with anterior knee pain and a circumduction gait; patellar instability may ensue. It is documented by measuring the intermalleolar distance when standing and prone. Genu varum may also be seen with knee pain and a waddling, Trendelenburg-positive gait, often in association with intoeing, ligamentous laxity, and lateral thrust. Furthermore, when the deformity is unilateral, progressive, relative limb length discrepancy is common.

While knee deformities represent, by far, the most common indication for guided growth, there are specific deformities such as ankle valgus or coxa vara that may also lend themselves to the same principle and technique. Timely guided growth may prevent or forestall the need for distal tibial or proximal femoral osteotomies. In some patients, multilevel guided growth, to simultaneously address concomitant deformities, is efficient and well tolerated [17] (Fig. 4.3).

Treatment

Timing/Angular

When dealing with progressive angular deformities, the previously preferred method of epiphysiodesis—now largely of historic interest—involved permanent arrest by either open [1] or percutaneous techniques [21, 22]. These required serial scanograms and bone age determinations with calculations using various methods [23–25]. Guided growth with tension band plates is more forgiving in terms of timing and calculations. On a standing AP teleroentgenogram, the relative limb lengths and mechanical axis are documented. The latter normally will bisect the horizontal knee at an 87 degree angle. The mechanical axis should fall within the center two quadrants (zones +/− 1) of the knee (Fig. 4.4). When it is in medial or lateral 2 or 3, surgical intervention is generally warranted. If two levels of deformity, for example femur+tibia, are identified, then pan

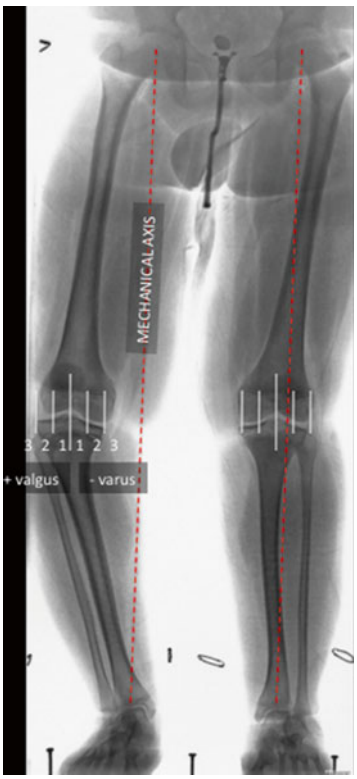


Fig. 4.4 The mechanical axis (*dotted red line*) comprises a vertical line connecting the center of the femoral head to the center of the ankle; normally, this bisects the knee. If you divide the knee into quadrants, medial zones (−) 1–3 reflect increasing varus and lateral zones (+) constitute valgus. While zone 1 may warrant observation, zones 2 or 3 call for treatment, in order to protect the physes and the knee

genu guided growth is indicated, in order to preserve a horizontal knee joint line.

In children older than 10, the anatomic angles, as codified by Paley and Sabharwal et al. (Table 4.1), can be useful to monitor during correction; these reference joint/shaft orientation for the femur and tibia [26, 27]. The joint congruence angle is typically 0° (parallel lines), indicating ligamentous balance/integrity. There may be concomitant deformity in both the femur and tibia. By envisioning the knee joint horizontally, one can readily detect multilevel deformities. For example, in patients with Blount's disease, there may be concomitant varus or valgus deformity of the distal femur that must be taken into account [26].

To benefit from guided growth, the age of the patient is not critical, provided that there is at least a year of predicted growth remaining. Guided growth has been safely and successfully utilized in patients ranging from 18 months to 18 years in chronologic age. The latter figure reflects the delayed/prolonged growth observed in some children with neurologic delays or metabolic syndromes. Commencing as young as age 4, patients with progressive or symptomatic deformity (such as Klippel-Trenaunay-Weber, Beckwith-Wiedemann, neurofibromatosis), it may be preferable to correct the problem, even if it requires repeated guided growth subsequently, rather than trying for perfect timing or resorting to osteotomy. The risks of delaying surgical realignment include compensatory deformity of the hip, knee, or ankle, ligamentous laxity, and patellar instability.

Table 4.1 Anatomic angles [26]

Abbreviation	Anatomic angles	Degrees
MPFA	Medial proximal femoral angle	90
LDFA	Lateral distal femoral angle	81
PMTA	Medial proximal tibial angle	87
LDTA	Lateral distal tibial angle	87
PPTA	Posterior proximal tibial angle	81
PDTA	Posterior distal tibial angle	90
JCA	Joint congruence angle	0–3 Apex medial

Technique/Angular

Under tourniquet control, the designated physis is approached, through a 3-cm incision. The dissection is deepened through fascial and submuscular planes, down to the bone, preserving the periosteum. A Keith needle is inserted into the physis, checking its position in the frontal and sagittal planes, with the fluoroscope. The plate is then inserted, placing the small center hole over the Keith needle to align it with the physis. It is desirable to position the plate midsagitally, unless an oblique plane deformity is being corrected. For example, a knee flexion/varus deformity may call for a single anterolateral tension band plate. One plate per physis is usually sufficient. The smooth 1.6-mm guide pins are inserted; they need not be parallel, but must avoid the physis and joint. Each hole is predrilled to a depth of 5 mm in order to facilitate advancement of the 4.5-mm cannulated screws, without undue torque.

While screw length is arbitrary, and sizes may be mixed, the longest practical screw is usually recommended. Each is inserted to be snug with the plate. It is then important to remove the guide pins, which may be binding on the screw, and further tighten each screw to countersink within the plate. This should be documented fluoroscopically. This critical maneuver will avoid 3-point bending stresses, reported in some patients with Blount's disease, which could produce stress fracture of the screw. This is a potential technical error that may compromise any screw, be it titanium or stainless steel, cannulated or solid. Once the hardware is secured, the wound is closed in layers and a soft dressing applied. Most patients are discharged from same-day surgery with (optional) crutches. Full weight-bearing and activities are encouraged as tolerated.

Follow-Up

Routine follow-up is recommended every 3 months (for angle) or every 6 months (for length) until correction is achieved (Fig. 4.5). For angular correction, most deformities

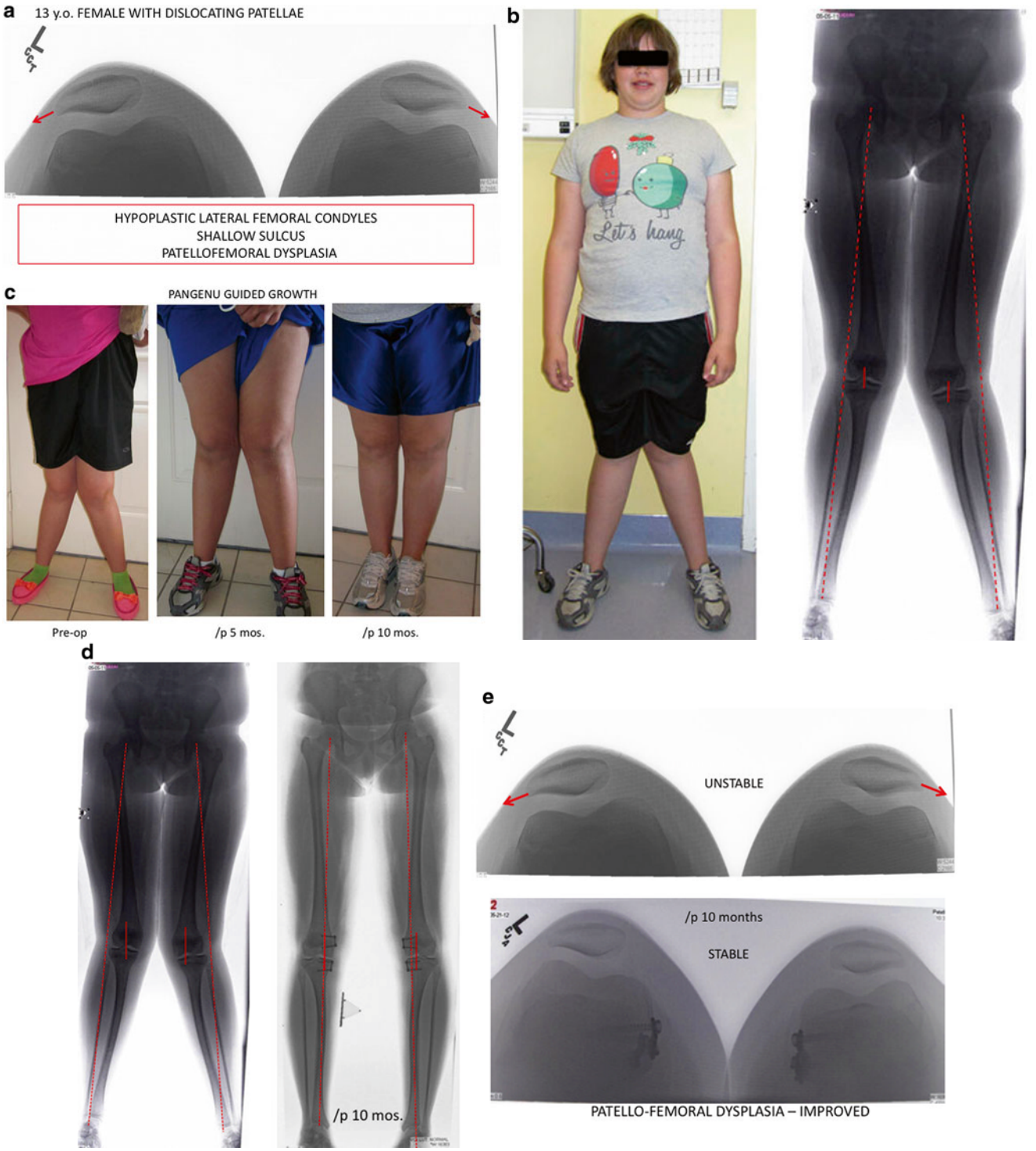


Fig. 4.5 (**a**) This 13-year-old girl presented with "patellofemoral dysplasia" manifest by bilateral patellar instability. (**b**) Clinically, her intermalleolar distance measured 20 cm. Her mechanical axis is lateral zone 3. It was apparent that idiopathic genu valgum (femur + tibia) was the proximate cause of her patellar dislocations and recognized that an MPFL reconstruction or patellar tendon transfer would not suffice to solve her problems. (**c**) Having hit her adolescent growth spurt, her deformities completely corrected within 10 months of pangenu plate application and her patellae stabilized, without direct intervention. (**d**) Radiographic comparison of before vs. 10 months after pangenu guided growth, whereupon the plates were removed. (**e**) "Sunrise" radiographs demonstrating improvement in the patellofemoral sulcus depth, presumably as a consequence of normalizing the mechanical axis and directing the patella in front of the femur

will be resolved within 12–18 months. Certain syndromes and bone dysplasias such as achondroplasia may require more time. There is no proven limit as to how long one can safely restrict one side of the physis. Since growth is unabated by the presence of a unilateral flexible tether, premature physeal closure is unlikely. Clinical correction is corroborated by full-length, weight-bearing teleroentgenogram, noting limb lengths and the mechanical axis. In children younger than 10, or with delay in skeletal maturity, it may be advisable to allow slight overcorrection of the mechanical axis into medial or lateral zone 1. In the event of rebound/recurrent deformity, the process may be repeated. Once the screws have reached maximum divergence, the narrow waist of the plate accommodates reverse bending of the plate, facilitating further correction. This characteristic may not be shared by the more rigid quad plates.

Angular: Sagittal/Oblique

Knee

Fixed knee flexion deformity (FKFD) may be associated with conditions including cerebral palsy, spina bifida, and arthrogryposis. It is important to differentiate between dynamic and fixed contracture, because the treatment is very different. Both conditions will limit the straight leg raise; with the former, the popliteal angle will change as the hip is flexed in the supine position. In addition to the typical supine straight leg raise, it is helpful to examine the patient prone, with the hip extended, relaxing the hamstrings. If the knee remains flexed and the foot does not reach the table, this indicates a fixed rather than dynamic knee deformity that will not respond to hamstring recession (Fig. 4.6).

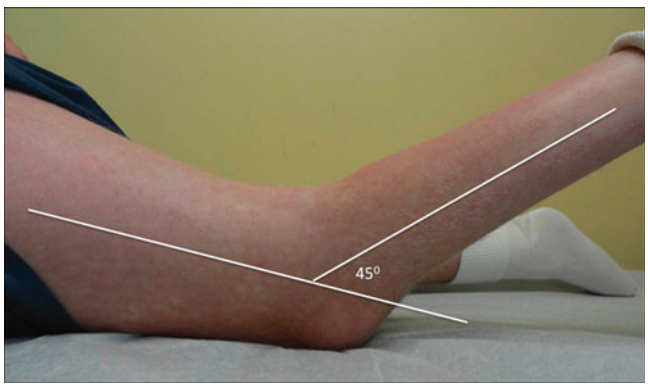


Fig. 4.6 Examination in the prone position places the hip in maximum extension, thereby eliminating the hamstrings as a source of deformity. With fixed knee flexion deformity (45° shown here) the ankle does not rest on the table. A hamstring recession will not resolve this deformity. An extension osteotomy may be averted with guided growth of the anterior distal femur

Often in children with lower limb spasticity bilateral, FKFD results in a progressive crouch gait that is refractory to bracing and physical therapy. The shortened stride length and anterior knee pain, often seen with radiographic evidence of patella alta +/− avulsion fractures, may preclude sustained ambulation. Supracondylar osteotomy of the femur +/− patellar advancement has enjoyed recent popularity. However, recurrent deformity is common and represents a major drawback.

An alternative approach is to employ guided growth of the distal anterior femur to gradually correct the sagittal or oblique plane deformities. It is important to place the plates medial and/or lateral to the sulcus (intracapsular, but nonarticular) in order to avoid irritation of the patella [28]. While the initial postoperative pain and crepitus may exceed the expectations of the parents and physical therapist, these symptoms abate as the legs gradually straighten. In contradistinction to osteotomies or external frames, knee flexion range is not compromised. Postoperative bracing [usually floor reaction] may still be helpful until the crouch deformity fully corrects. Typically, this takes 12–18 months following guided growth treatment. Overcorrection into recurvatum is rare, as is recurrence. In the authors' experience, simultaneous or subsequent advancement of the patellar tendon is usually not required because knee pain typically abates as the knee gains extension and patella alta is rendered unimportant. Furthermore, concomitant hamstring lengthening is also unnecessary. In fact, as the knee straightens, the hamstrings may actually pull upon the ischium, extending the pelvis and alleviating the hip flexion deformity, while preserving hip extensor power. Finally, apparent equinus is improved indirectly as the knee extends and the heel may reach the floor. Thus, by primarily addressing the FKFD with anterior tension band plates, "single-event, multilevel" correction is realized. In contradistinction to supracondylar osteotomy, posterior capsulotomy, or frames, knee flexion is preserved while extension is gained (Fig. 4.7).

Another application for sagittal plane correction of the proximal tibia is shown in Fig. 4.8. In this posttraumatic deformity, there was no defined physeal bar. Had one been identified, guided growth may be employed, at the time of epiphysiolysis, to expedite angular correction.

Ankle/Frontal

Progressive ankle valgus is common and insidious sequelae of a variety of conditions (Table 4.2). The detection is more challenging than knee deformities because of the relatively short height of the foot. It may be mistaken for an overcorrected club foot, planovalgus, or pronated foot. The critical radiographic study is a weight-bearing AP of the ankles. The normal lateral distal tibial angle (LDTA) should

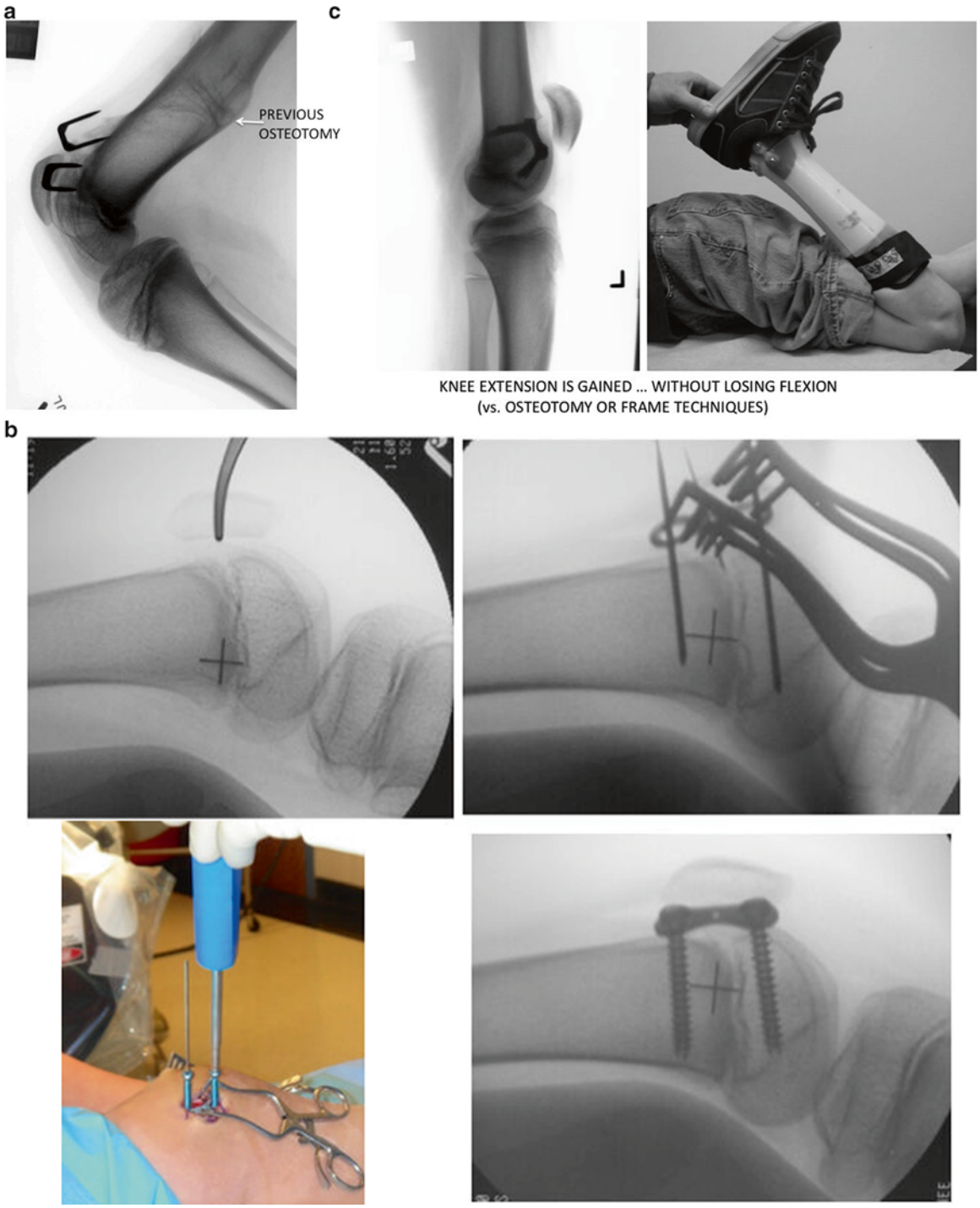


Fig. 4.7 (**a**) This 7-year-old patient with congenital fixed knee flexion deformity (FKFD) had a previous extension osteotomy (*arrow*) at age 3, with the inevitable recurrence of her deformity. Anterior femoral staples migrated and had to be removed. (**b**) Intraoperative images show the technique of sagittal plane correction by applying tension band plates to the anterior distal femur, one on each side of the sulcus. (**c**) In contradistinction to osteotomy, or frame application, knee extension is achieved, without losing correction. Regarding patella alta, if the patient is ambulatory and pain free, there is no requirement that the patellar tendon be advanced

be approximately 90°. As the deformity progresses, there is often lateral pain around the ankle and hindfoot due to impingement of the distal fibula upon the talus or calcaneus. The distal fibular epiphysis may enlarge in response to the increased weight-bearing load imposed as the ground reaction force shifts laterally. This situation does not lend itself to correction with orthoses.

A simple method of treatment is guided growth of the distal medial tibia. While a transphyseal screw can work, one is applying a rigid implant across the open and very dynamic

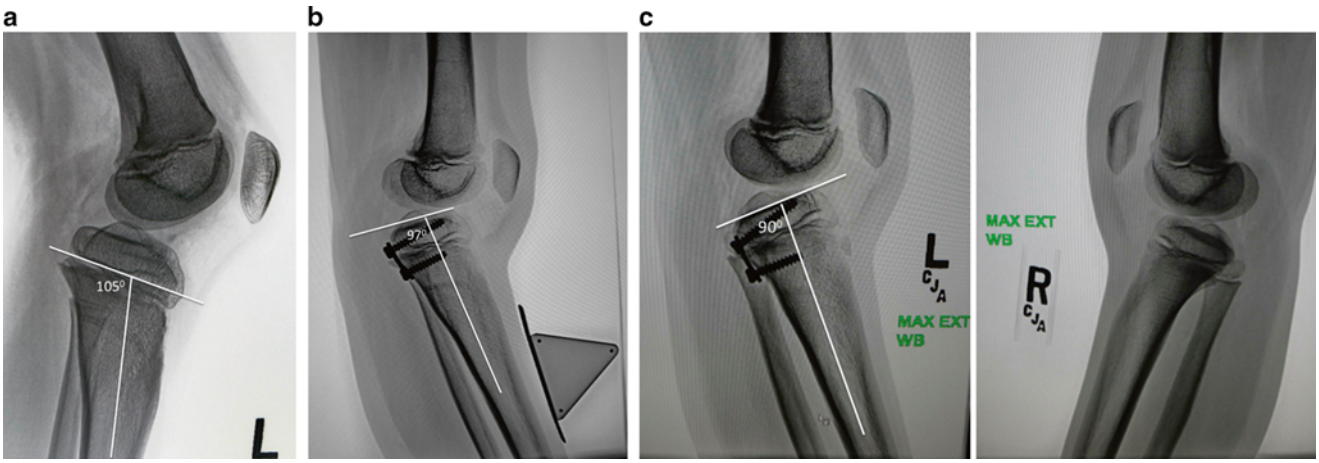


Fig. 4.8 (**a**) This 8-year-old girl suffered a proximal tibial transphyseal fracture on a trampoline. While the fracture healed uneventfully, she presented with a symptomatic, 20° recurvatum deformity. The preoperative lateral view demonstrates posterior proximal tibial angle of 102°, with the normal value being 81°. (**b**) 6 months following posterior guided growth, to effect correction in the sagittal plane, note the diverging screws. (**c**) 12 months postoperatively, the screws continue to diverge, as a result of redirected growth. The opposite knee is shown for comparison. She has become asymptomatic. When the recurvatum is fully corrected, the metaphyseal screw will be removed, percutaneously. Her growth will continue to be monitored on a regular basis and the screw may be reinserted p.r.n.

Table 4.2 Etiology of ankle valgus

Dysplastic	Neuromuscular	Genetic	Posttraumatic
Club foot	Cerebral palsy	Rickets (or metabolic)	Fracture
Postaxial hypoplasia	Spina bifida	Down syndrome	Osteotomy
Tibial pseudarthrosis	Neurofibromatosis	MHE	

physis. This may lead to implant failure or difficult removal. It is preferable to utilize a flexible, extra-periosteal tension band that will produce more rapid correction and is simple to remove. In comparison to the knee, the time to correction of the ankle is slower, often taking 18–24 months. It is recommended to overcorrect into slight varus (LDTA of up to 95°), in anticipation of rebound valgus, which is relatively common. The concerns expressed regarding hardware prominence have noit borne out in practice. However, if the child is wearing an AFO, it is desirable to bend or pad the brace p.r.n. to avoid rubbing.

Ankle/Sagittal

At the distal tibia, one may insert an anterior plate for residual fixed equinus or a posterior plate for calcaneus deformity, rather than resort to supramalleolar osteotomy. In a similar manner, a posterior plate may address calcaneus deformity of the ankle. If the deformity is greater than 15°, however, an osteotomy may be required.

Complications of Guided Growth

Perioperative complications pertaining to tension band plate insertion are decidedly rare, provided that the routine precautions are exercised. These include protecting and preserving the periosteum, and avoidance of violating the physis or joint with the screws. With respect to the latter, it may be helpful to perform an arthrogram in young children, in order to outline the entire chondroepiphysis (Fig. 4.9).

There have been reports of *broken screws*. Most commonly this occurs in Blount's disease and involves the metaphyseal screw, not at the head/shank junction, but where the exposed screw shank enters the cortex. This may occur

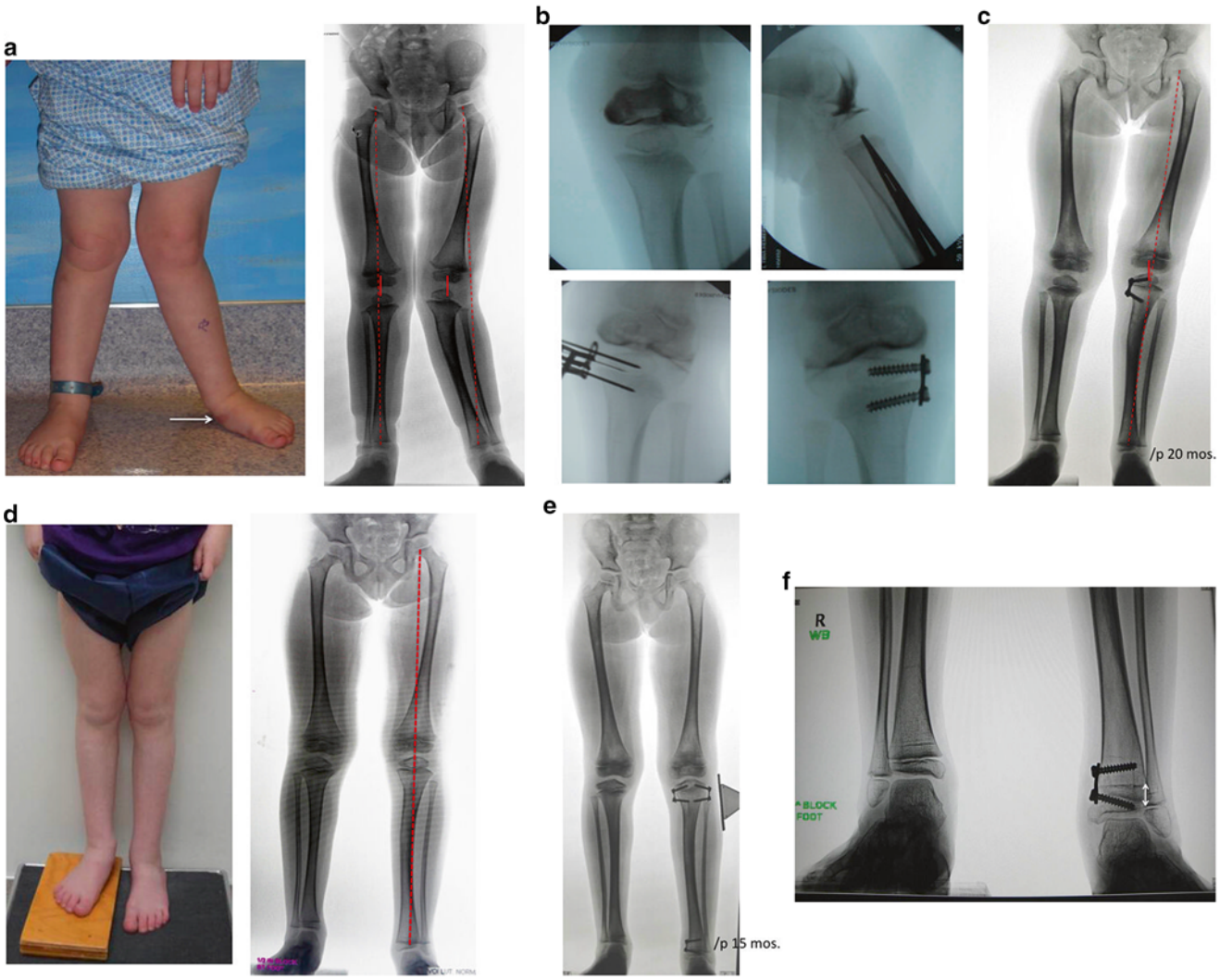


Fig. 4.9 (**a**) A 3-year-old girl with Cozen's posttraumatic genu valgum, presenting 18 months following proximal tibial fracture. Note the small hemangioma on the instep (*arrow*). (**b**) An arthrogram was employed to better delineate the chondro-epiphyses and aid in placing the guide wires and cannulated screws. (**c**) Full correction of the angular deformity took 20 months, whereupon the implant was removed. Follow-up continued to monitor for recurrent valgus and LLD. (**d**) At age 7, she is noted to have a 2.5 cm discrepancy and symptomatic ankle valgus. These were attributed to her congenital hemangioma causing growth imbalance. This was managed via bifocal guided growth—proximal tibia for length inhibition and distal medial tibia for valgus. (**e**) 15 months later, her length discrepancy measures 1.2 cm and the ankle valgus has resolved. (**f**) Note the asymmetrical growth line in the left distal tibia, with correction of the ankle valgus to neutral. The strategy is to wait for 9 months, allowing slight overcorrection into ankle varus, and then remove the metaphyseal screws from the distal and proximal plates. Following 6 months of reprieve from guided growth, allowing growth to resume, the metaphyseal screws may be reinserted. Follow-up will be continued until maturity, repeating this process of screw removal/reinsertion as indicated

regardless of the material (stainless vs. titanium), design (cannulated vs. non-cannulated), or size of the screw (Fig. 4.10). This situation is best avoided by anticipation and awareness. Steps to avoid this situation include the following: (1) remove guide pins and alternately tighten the screws to bend the plate in situ, or (2) remove plate and increase its convex bend to better contour to the lateral cortex of the tibia. When encountered, broken screws may be percutaneously exchanged, as needed, rather than resorting to osteotomy. Other options to consider are the use of dual plates or quad plates, and the choice of solid (non-cannulated) screws. Without corroborative evidence, these choices reflect surgeon preference.

The issue of *rebound growth* imbalance and recurrent deformity, which is more common under the age of 10, is not a true complication. Rather, it is an educational challenge, requiring the surgeon to establish parental expectations, maintain vigilance, and pursue meticulous follow-up. The rebound deformity phenomenon can generally be detected within 12 months of implant removal. Properly informed, most parents (and patients) would prefer repeat guided growth to any osteotomy.

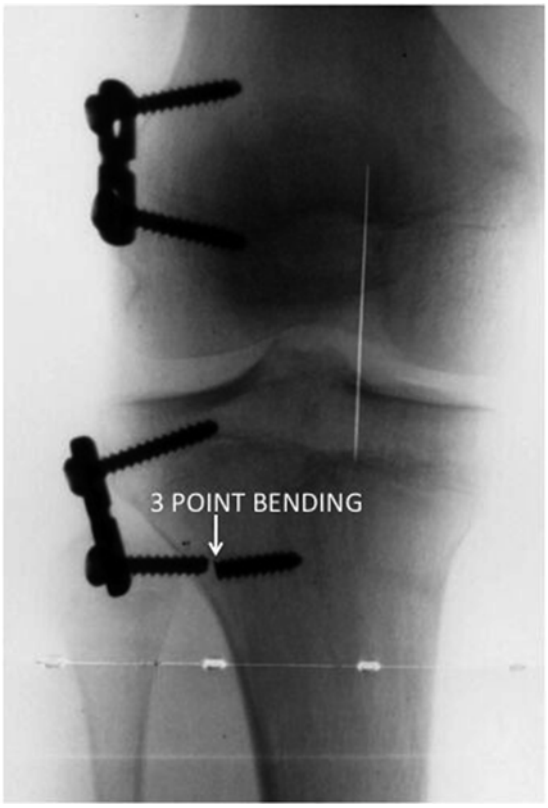


Fig. 4.10 If there is no contact between the plate and tibial metaphysis, the screw be subject to 3-point bending, regardless of the screw size and material. This 4.5-mm solid stainless screw demonstrates such a technical error

Management

Weighing the potential for rebound growth, causing recurrent deformity (more common under the age of 10), may be desirable to permit slight overcorrection (of varus into medial zone 1 valgus—or vice versa). This will extend the "mileage" of a given procedure. If rebound is deemed likely, one option is to remove just the metaphyseal screw (Fig. 4.11) and continue observation, reinserting the screw if and when rebound is documented. In other words, the modular nature of reversible guided growth lends itself to a myriad of conditions and situations. Frequent minimally invasive, outpatient surgeries may be preferable to major, more invasive techniques.

Length: Anisomelia

Physiologic

It is widely accepted that limb length discrepancy of up to 1.2 cm is well tolerated by most adults, requiring no treatment. When lesser discrepancies are identified, it is appropriate to see the growing child once or twice a year to document any progression and intercede accordingly. While shoe lifts may be prescribed, they are compensatory and few children will consistently wear them.

Pathologic

Whether the etiology is congenital, developmental, or acquired, if the predicted discrepancy will exceed 1.2 cm at maturity, then surgical treatment may be warranted. It is important to see the growing child at least annually, in order to identify the trend (static vs. progressive), educate the parents, and anticipate the predicted discrepancy at maturity.

The most practical annual radiograph to obtain is the teleroentgenogram, which is a full-length, weight-bearing AP of the legs [29]. It is helpful to level the pelvis with an appropriate height wooden block under the shorter leg. If one uses a calibration marker, then relative limb lengths may accurately be measured, along with the mechanical axis [30]. Traditionally, shoe lifts were employed until adolescence when open or percutaneous epiphysiodesis was undertaken. Some authors still favor permanent epiphysiodesis over reversible instrumented techniques [31]. However, problems inherent in determining skeletal maturity have frequently led to over- or under-correction, especially when using permanent and irreversible techniques. With the advent of reversible techniques—formerly using staples and more recently, extraperiosteal tension band plates—the timing is much more forgiving and the philosophy has changed accordingly.

Treatment: Length

Timing: Over vs. Under the Chronologic Age of 10

The typical application for guided growth inhibition is during adolescence, hoping to achieve limb equalization at skeletal maturity. Regardless of the implants chosen (staples, tension band plates, Metaizeau PETS), there is a latency effect before longitudinal growth is actually inhibited. This should be taken into account and epiphysiodesis undertaken 6–12 months in advance of what was formerly deemed "ideal." Some authors prefer drilling the physis, in order to avoid implants and the potential need for removal [31]. However, by using reversible technology, there is leeway because the implants may be removed if correction occurs before skeletal maturity is reached.

Comparatively young children with "gigantism" due to specific conditions such as Trevor's Disease, Klippel-Trenaunay-Weber, hemangioma, Beckwith-Wiedemann

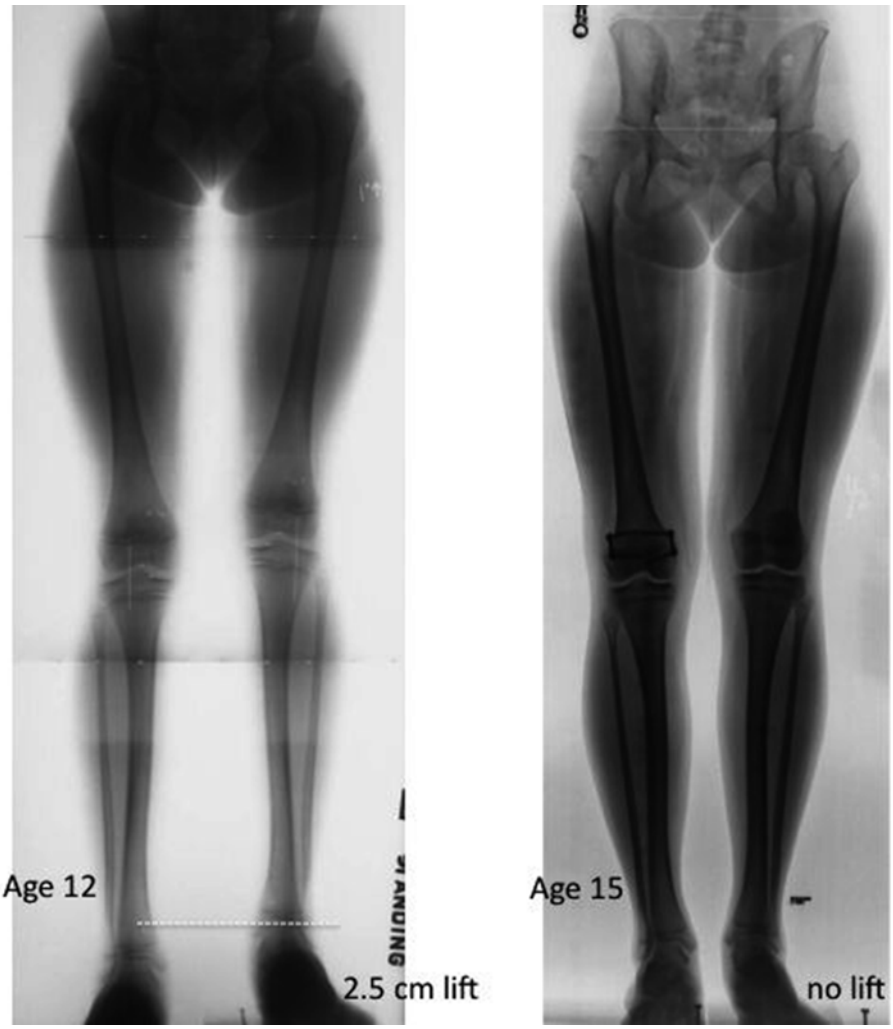


Fig. 4.11 As a sequela of DDH, this 12-year-old girl became symptomatic with a 2.5-cm limb length discrepancy. Three years following guided growth, her limb lengths are equal

syndrome, and neurofibromatosis may be candidates (starting as young as age 4) for intermittent longitudinal restraint of the overgrown limb, via guided growth (Fig. 4.12). Similarly, children with limb dysplasia, such as postaxial hypoplasia, may be considered for serial/intermittent guided growth of the normal, longer limb. The goal is to forestall, minimize, or perhaps obviate limb lengthening, with all of its associated risks.

In patients undergoing major long bone resection/limb salvage for oncologic disease, intermittent guided growth of the contralateral limb may mitigate some of the problems encountered with expanding implants. When employing this strategy, it is important to respect the 2-year window of opportunity, because, if a given physis is restrained indefinitely, the physis could permanently shut down. Therefore, one strategy is to remove the metaphyseal screws, wait for 6 months, and reinsert them (see Figs. 4.9 and 4.12). This may serve as an adjunct to contralateral limb lengthening, or circumvent the need for it altogether.

Treatment/Length

Despite utilizing the same implants and surgical approaches, there are subtle differences in technique. Specifically, the screws are placed in a divergent orientation within the plate (Fig. 4.13). This will mitigate against screws bending and may reduce the aforementioned latency period.

Complications/Length

Length correction is less dramatic than angular. Consequently, follow-up every 6 months is sufficient for monitoring progress. The proximal lateral tibia may pose problems and lead to iatrogenic genu varum. This problem may be encountered whether one uses staples or tension band plates. If such an angular drift is noted, one should remove the medial tibial plate [or metaphyseal screw], allowing gradual normalization of the mechanical axis, and reimplant when the alignment

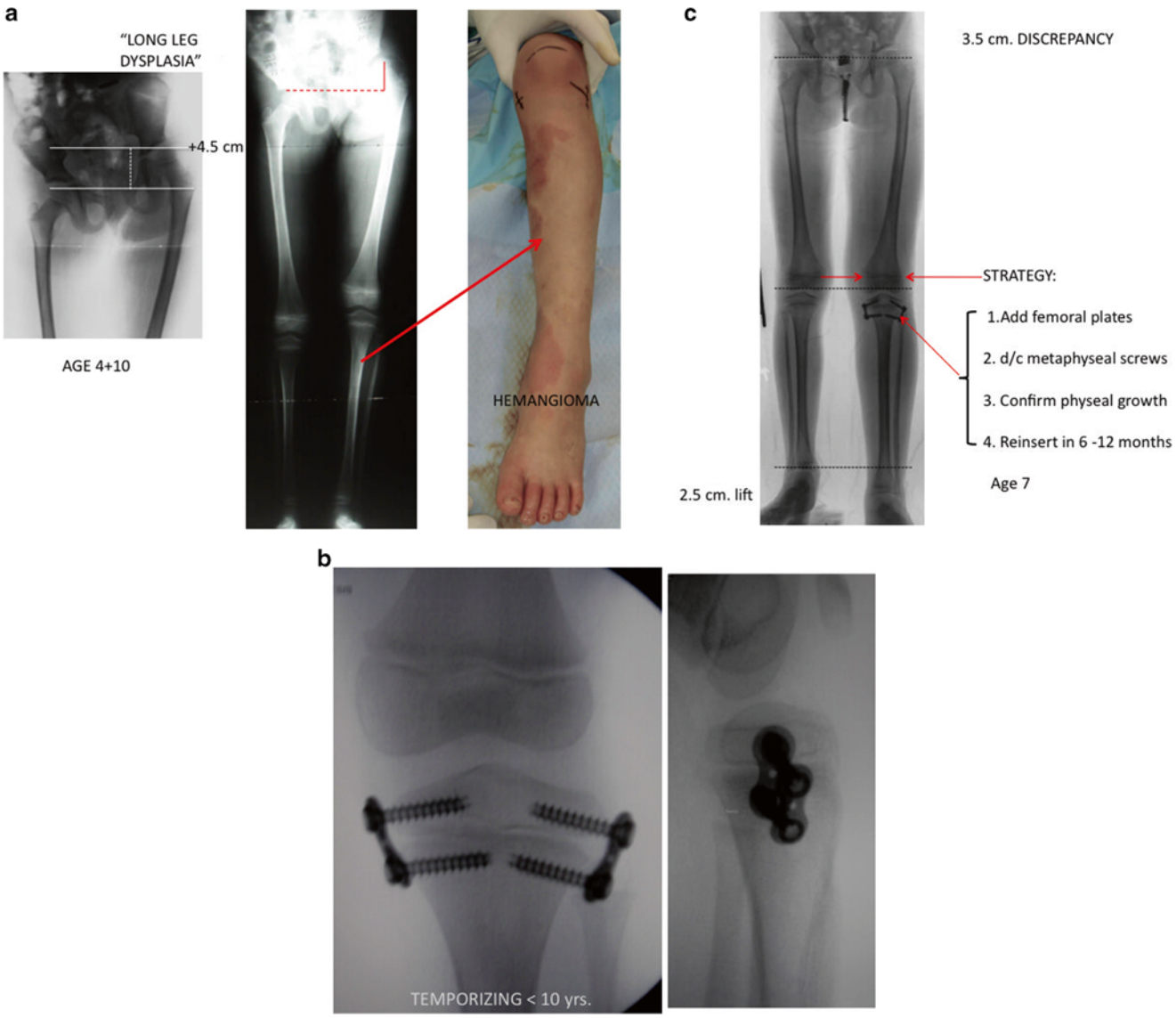


Fig. 4.12 (**a**) This girl presented with hemangioma causing a progressive LLD (4.5 cm), left hip and back pain, and projected 6- to 7-cm discrepancy (if untreated) at maturity. (**b**) She started at 4 + 10 years with guided growth of the proximal tibia. The fibula was not prominent, and thus was not included. (**c**) 2 years later, her discrepancy measured 3.5 cm. The empirical tolerance of physeal restraint is 2 years. (**d**) Therefore, the tibial metaphyseal screws were removed and femoral plates added. (**e**) Asymptomatic at age 9, with a 2.5 cm discrepancy (would have been 5–6 cm by now). (**f**) At age 10, with a 2-cm discrepancy and varus drift of the mechanical axis into zone 2. This was managed by reinserting the lateral metaphyseal screw in the tibia and percutaneously drilling the proximal fibular physis. When the axis returned to neutral over the ensuing 8 months, medial screws were added. (**g**) A summary of guided growth procedures as of age 11 + 4, when her limb lengths became equal for the first time in her life. The financial savings and reduced risk, as compared to osteotomy/shortening of the involved leg, or frame/lengthening of the normal right leg, are obvious

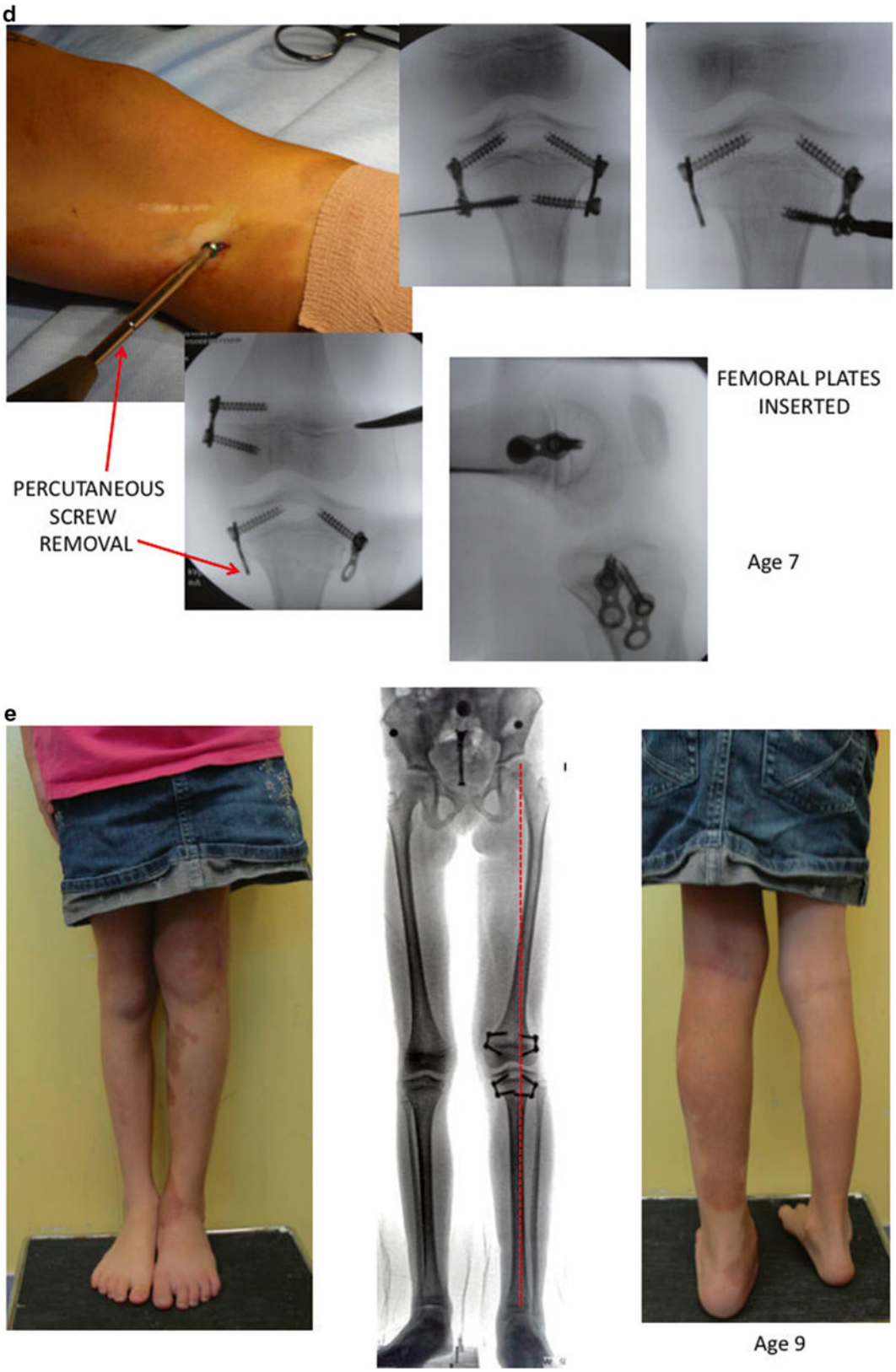


Fig. 4.12 (continued)

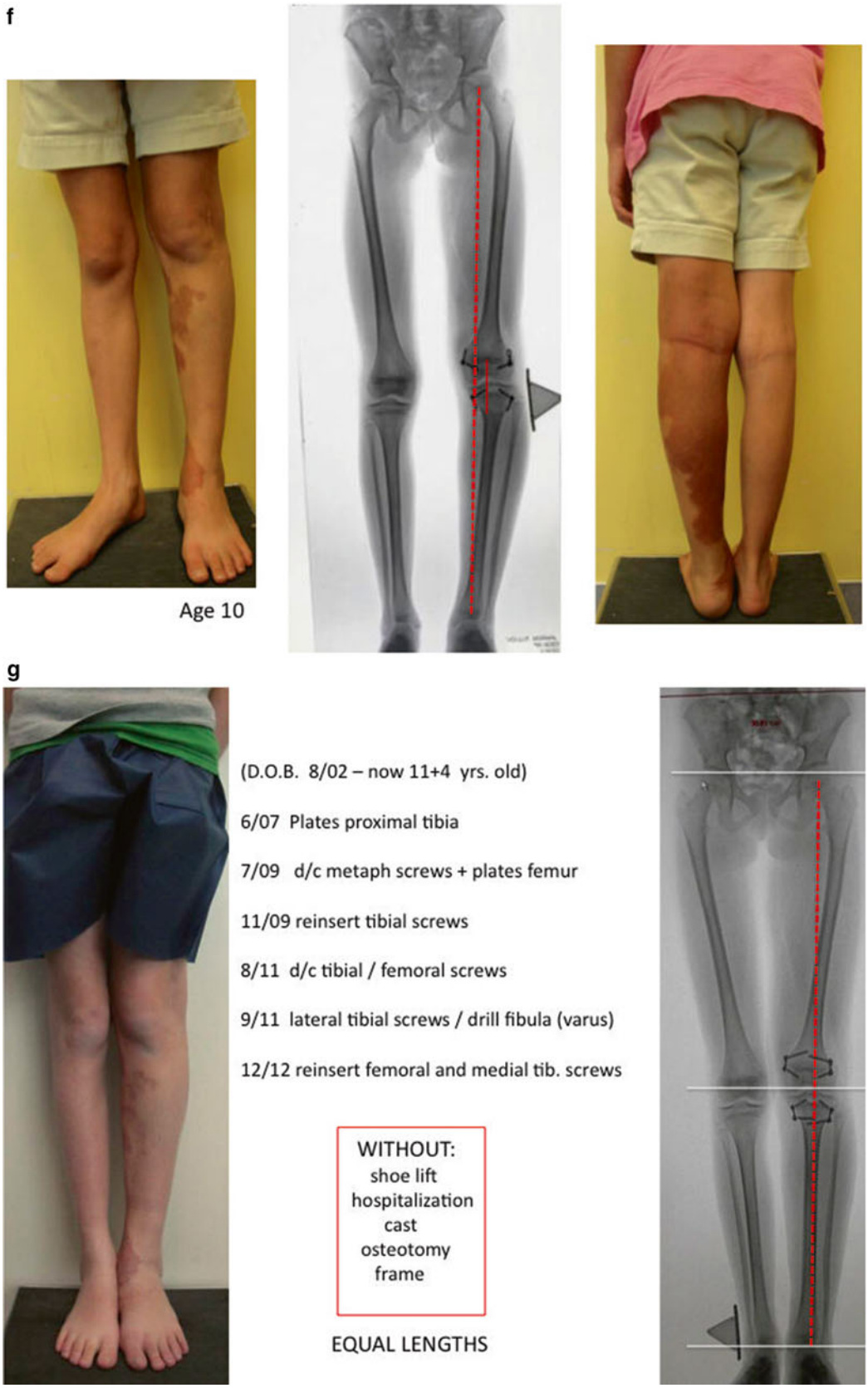


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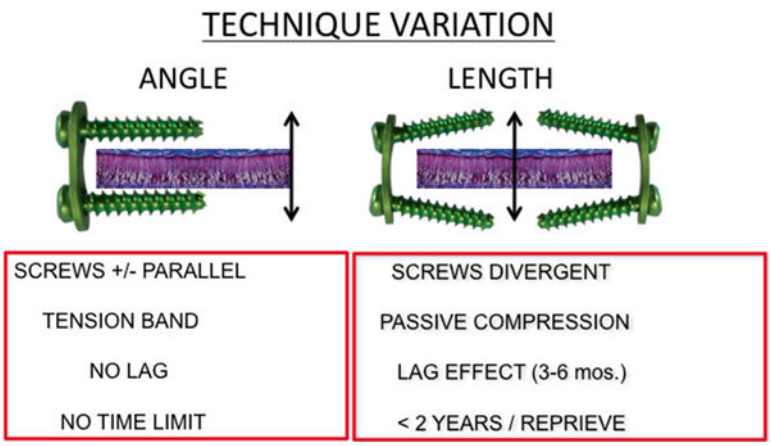


Fig. 4.13 When effecting angular correction, the screws are placed more or less parallel to each other and there is no time limit. Most angular deformities correct within 12–18 months. When placed for length inhibition, the screws are placed divergent to each other. This may reduce the lag time and avoid screw bending. After 2 years of growth inhibition, one may remove the implant, or just the metaphyseal screw(s), wait for 6 months, and repeat the process p.r.n.

is neutral (see Fig. 4.12). In some cases, unopposed fibular growth may contribute to the deformity. If this is suspected, or if the fibular head is relatively prominent, it may be desirable to percutaneously drill the proximal fibular physis.

Summary

The classic method of epiphysiodesis, described eight decades ago, has evolved to a more versatile, less invasive, and reversible method, such as employing a low-profile tension band plate. This technique lends itself to the correction of both angular deformities and limb length discrepancy and may safely be applied in very young children, without the risk of permanent physeal closure [provided the periosteum is protected]. Bilateral and multilevel deformities may be addressed, on an outpatient basis. The complication rate and relative cost are very low, compared to osteotomy. This method should be considered the treatment of choice for a wide variety of conditions, reserving osteotomy for salvage situations.

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Physeal Bar Excision

5

Karl E. Rathjen and Anthony I. Riccio

Introduction

As with any clinical dilemma in the growing child, management of a physeal growth disturbance begins with careful consideration of the natural history of the pathologic process. Appropriate treatment of a physeal arrest starts with understanding the patient and disease characteristics that may act in concert to create limb deformity. Knowledge of the nature and characteristics of the arrest, an understanding of normal physeal behavior, and an appreciation of factors influencing future growth allow the clinician to assess the potential for deformity if a physeal arrest was left untreated. Appreciation of these factors is also essential for generating realistic expectations for restoration of normal growth following surgical intervention. To ensure optimal outcome, the treatment choice must be tailored to the individual patient and, in many instances, the individual physis.

Etiology and Prognosis

Physeal injury can occur via both direct and indirect mechanisms. While physeal fractures account for the majority of physeal arrests and premature physeal closures, growth disturbance is also reported following osteomyelitis, vascular insult, radiation exposure, as well as neoplastic and tumor-like conditions [1–12]. Infantile Blount's disease is yet another cause of physeal growth disturbance. Unlike other etiologies, physeal arrest in infantile Blount's disease is likely the result of sustained altered physiologic function within the physis itself [3, 13, 14].

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Growth disturbance can occur from physical loss of the physis, altered physeal physiology without loss of physeal structure, or disruption of normal physeal architecture. Disruption of normal physeal architecture may produce an osseous bridge across the cartilaginous physis, which is termed a physeal bar (Fig. 5.1). However, it is important to realize that physeal growth disturbance may produce deformity without physeal arrest (Fig. 5.2). The etiology of a physeal growth disturbance appears to have some bearing in the successful restoration of growth following resection of a physeal bar. In the authors' experience, growth arrests associated with fractures and infantile Blount's disease carry a better prognosis for resumption of growth following epiphysiolysis than arrests resulting from infection, radiation, or intraosseous space-occupying lesions [11]. As the treatment strategies can differ based upon the etiology of a growth disturbance, an understanding of the nature of the physeal disturbance is essential.

Trauma

It is well documented that the physis' cartilaginous nature provides a path of minimal resistance to forces traversing an extremity. The majority of physeal fractures course through the hypertrophic zone of the growth plate, which is mechanically the weakest zone of the physis and therefore the most susceptible to disruption. Physeal fractures that progress from the hypertrophic zone into the adjacent metaphysis (Salter–Harris II fractures) course away from the germinal and proliferative zones and therefore away the regions of maximal growth and metabolic activity. As a result, these fractures generally have a lower rate of physeal arrests than those that course from the hypertrophic zone, through the germinal and proliferative zone and towards the epiphysis [15] (Salter–Harris III and IV fractures) [16, 17] (Fig. 5.3).

In addition to the Salter–Harris classification, fracture location can be prognostic for the development of a growth

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Fig. 5.1 AP radiograph of bilateral ankles. Note the "Harris growth arrest" lines (*arrowheads*) on both sides. On the right ankle the Harris line is asymmetrical and terminates medially at the area of physeal arrest or "bony bar" (*arrow*)

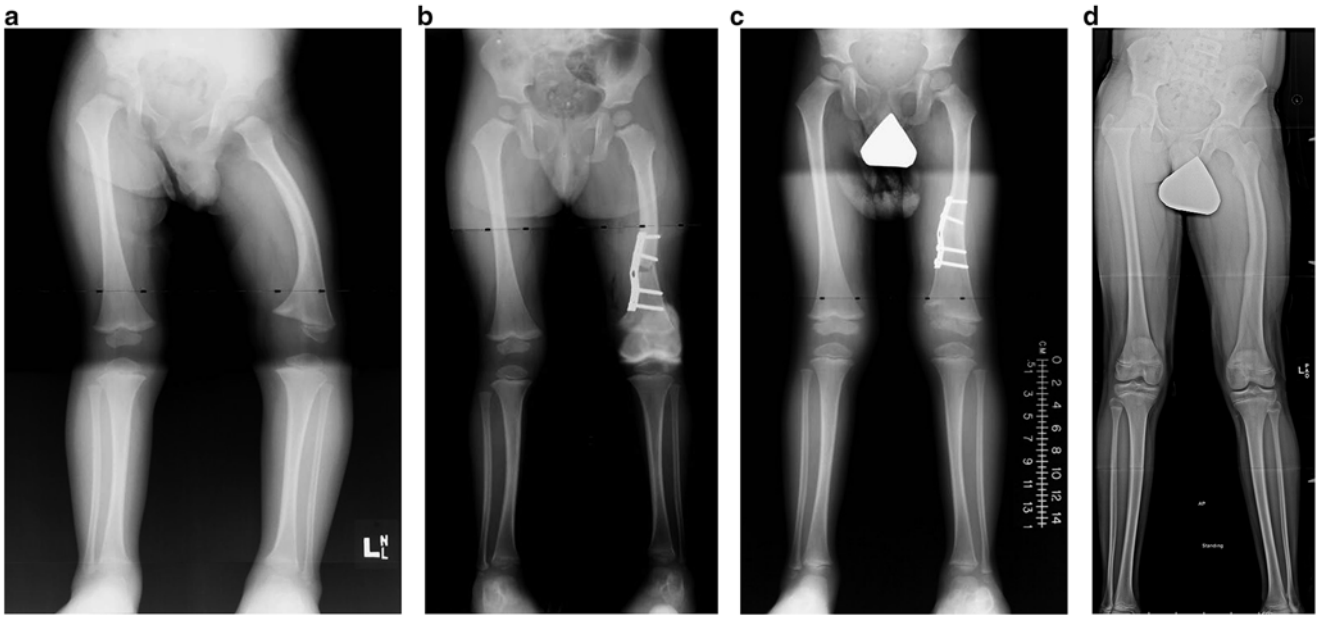


Fig. 5.2 (**a**) A 2-year-old with a history of neonatal sepsis. MRI at this age showed no physeal bar and a relatively normal cartilaginous epiphysis. Despite the significant diaphyseal deformity and appearance of the secondary ossification center, the patient was felt to have mild (albeit long-standing) growth disturbance without physeal arrest. (**b**) Intraoperative radiograph with opening wedge osteotomy and arthrogram which demonstrates normal cartilaginous anatomy of the distal femur. (**c**) With restoration of normal limb alignment and mechanical forces, the secondary ossification of the distal femur begins to ossify. This is an excellent example of the Heuter-Volkmann principle. (**d**) 9 years postoperatively, there has been relatively normal growth. Note the absence of angular deformity, the pelvic obliquity associated with limb length inequality, and the premature closure of the ipsilateral distal femoral growth plate

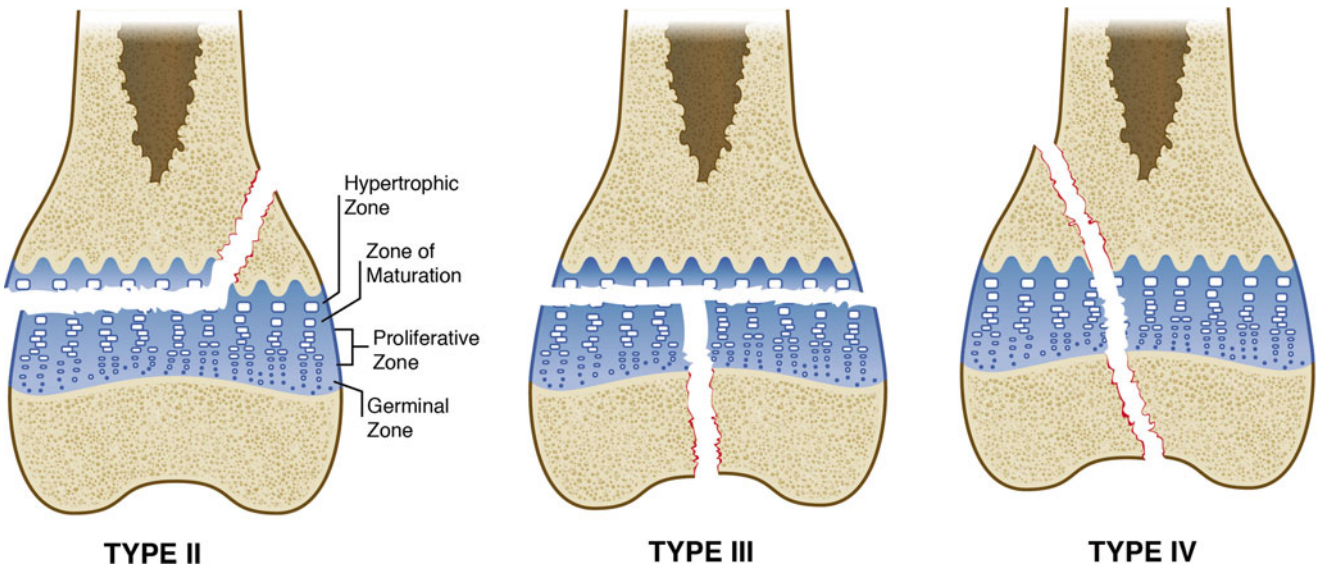


Fig. 5.3 Salter–Harris type II, type III, and type IV injuries. Note that the germinal zone of the physis is disrupted in the type III and IV injuries

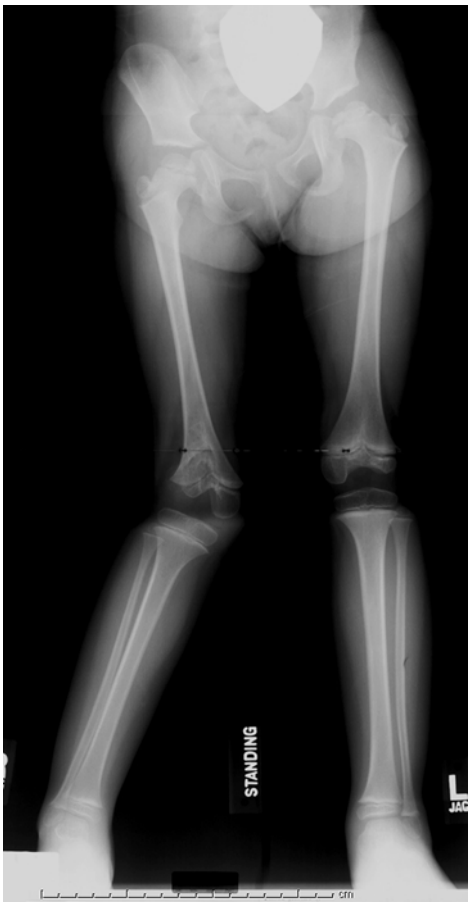


Fig. 5.4 AP bilateral lower extremities of a 4-year-old with a history of neonatal sepsis. Note the shortening and angular deformity of the right leg. Note also that the central growth arrest has created intra-articular abnormality of the right knee. Note the irregularities in the left proximal and distal femoral epiphyses

arrest. Fractures involving the distal femoral physis and, to a lesser extent, those involving the medial malleolus are notorious for relatively high rates of growth arrest [11, 17].

Infection

Osteomyelitis and septic arthritis can lead to both physeal growth disturbance and growth arrest. As with trauma, physeal destruction from musculoskeletal infection can result in both complete and partial growth disturbance resulting in angular deformities, altered joint morphology, and limb length differences [7]. Widespread systemic neonatal sepsis can result in multifocal physeal arrests and severe deformity (Fig. 5.4).

Neoplasm and Tumorlike Conditions

Growth disturbance from physeal injury has been reported in the presence of both malignant and benign neoplasms [18, 19].

Physical encroachment of the growth plate by these lesions is the usual mechanism of physeal damage, although additional injury can occur during treatment (surgical or radiation). Growth disturbance resulting from space-occupying lesions can be challenging to treat as it is often difficult to predict the degree of altered growth resulting from irradiation and the behavior of physeal tissue preserved following tumor resection or a limb salvage operation. Small physeal bars are only rarely encountered, usually obviating epiphysiolysis as a surgical option. Completion epiphysiodesis and staged or concurrent deformity correction or limb lengthening procedures are more commonly required.

Vascular Insult

Limb hypoperfusion due to either severe systemic hypotension or local limb ischemia resulting from vascular injury, altered venous outflow, or compartment syndrome can result in altered physeal physiology and growth disturbance [20–22].

This relatively rare cause of abnormal growth plate function has been associated with both partial and complete physeal arrests. A small proportion of growth arrests without an obvious attributable cause may in fact have resulted from unrecognized transient vascular injury insufficiently following local trauma without fracture or injury remote to the area of arrest in the ipsilateral limb.

Other Causes of Physeal Injury

Rare etiologies of growth plate disturbance include burns, frostbite, electrical injury, radiation exposure, and repetitive stress injuries. Growth disturbance due to repetitive stress is commonly encountered in the young athlete. Altered proximal humeral physeal growth is well described in children who engage in extensive overhead throwing activities [23, 24]. Fortunately, cessation of the offending activity usually results in restoration of normal physeal function.

Assessment of the Abnormal Physis

Timely identification of a growth disturbance, differentiation between partial and complete physeal arrests, determination of the location and extent of a physeal bar, and an accurate assessment of remaining growth of the involved physis are all necessary for complete assessment of the abnormal physis and selection of appropriate treatment.

Timely Identification

Early identification of a physeal growth disturbance usually affords the clinician a wider array of management options and can simplify management by allowing for treatment of the growth arrest without concomitant management of an associated deformity. Careful serial radiographic follow-up of physeal injuries is recommended until the clinician is confident regarding the resumption of normal growth. Even when normal growth ensues, radiographic surveillance until skeletal maturity is advised as physes with a history of injury are known to close prematurely [25]. If identification of a growth disturbance is delayed and a significant acquired deformity has developed, both the deformity and the growth disturbance usually require treatment.

Partial Versus Complete Physeal Arrest

The effect of a bar (bony, cartilaginous, or fibrous) on the adjacent physeal growth varies with location and size of the arrest. Large bars and those bars associated with extensive injury to surrounding physeal cartilage usually retard growth of the entire physis causing a complete growth arrest (Fig. 5.5). Such bars usually result in a limb length discrepancy without associated intra-articular or angular deformity. Smaller central bars and those that are more peripherally located usually result in a partial physeal growth disturbance. As the bar causes local restriction of growth in the presence of normal physeal function remote to the bar, angular deformity, limb length difference, tenting of the physis, and intra-articular deformity can ensue (see Fig. 5.4).

Resumption of growth following resection of a physeal bar relies upon the integrity and health of the normal physis surrounding the physeal bar. There is no role for physeal bar resection in the presence of a complete growth arrest of the entire physis; therefore, differentiating between a partial and complete disturbance is the first step in developing a management strategy.



Fig. 5.5 AP bilateral lower extremities of a 9-year-old who sustained a distal femoral physeal fracture 2 years earlier. Note the premature complete closure of the left distal femoral physis and the pelvic obliquity associated with mild limb length inequality

Location of Physeal Arrest

Partial growth arrests are classified based upon their location within the physis. Determination of location of a physeal bar is important, as it is predictive of the potential deformity that might ensue if left untreated. Furthermore, the choice of surgical approach for a bar resection is largely dependent upon its location.

Physeal bars are generally classified as central or peripheral [10]. Peripheral bars, as the name suggests, involve an outer edge of the growth plate. If left untreated, peripheral bars are likely to create marked angular deformity in a child with considerable growth remaining. We typically classify peripheral bars as Type A arrests (Fig. 5.6). Central bars are sub-classified based upon the amount of healthy physis surrounding the arrest. Type B arrests are surrounded circumferentially by normal physeal cartilage. As the normal physis continues to grow around a Type B bar, tenting of growth

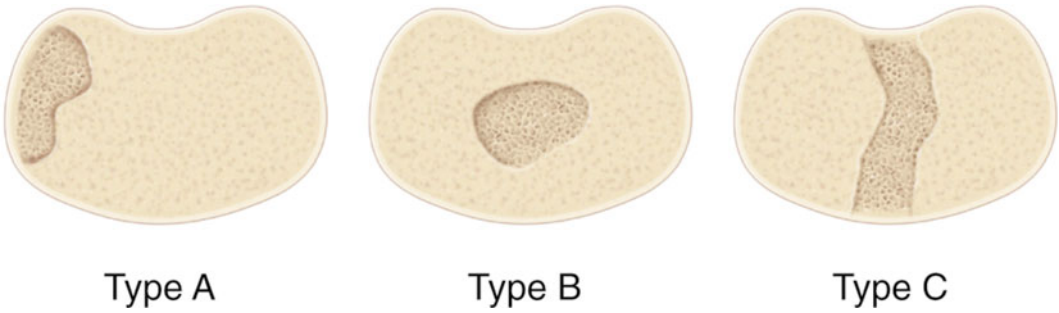


Fig. 5.6 Anatomic classification of physeal arrest. Type A: Peripheral. Type B: Central surrounded by normal physis. Type C: Central traversing the physis completely. Reprinted with permission from Herring JA (ed). Tachdjian's Pediatric Orthopedics: From the Texas Scottish Rite Hospital for Children. 5th ed. Philadelphia: Elsevier; 2013

plate, and eventually the epiphysis, can develop. This may ultimately result in distortion of the underlying articular cartilage. A Type C arrest is a variation of a central bar in which the area of arrest traverses the entire growth plate in one direction with healthy physis present on the remaining two sides. This type of arrest is often seen following Salter–Harris III and IV fractures of the medial malleolus in which physeal bars frequently extend from anterior to posterior with normal physis found medially and laterally (an example is shown in Fig. 5.10).

Resection of a bar while minimizing concomitant damage to the surrounding normal physis is of paramount importance regardless of the type of arrest encountered. The importance of classification lies in the fact that accomplishing this goal in the presence of central bars (Types B and C) may be more challenging than in those located peripherally.

Extent of Physeal Arrest

Determination of the extent of physeal involvement in a partial growth plate arrest is the next step in developing an appropriate treatment strategy. Size of a physeal bar, typically documented as a percentage of the entire physis, has been shown to correlate with successful outcomes following resection surgery [4, 11, 26, 27]. Successful restoration of growth has been reported to be more likely in bars that comprise less than 25 % of the involved growth plate [27]. While it is important to note that no firm cutoff exists as to a percentage of physeal area beyond which bar resection should not be attempted, restoration of meaningful growth following excision of arrests that involve greater than 50 % of the growth plate is less likely. Although three-dimensional imaging is now nearly universally used to assess the extent of a physeal arrest, we believe that it is important to understand the process of "mapping" a physeal arrest in two planes and that this exercise can ensure that the surgeon has a comprehensive understanding of the three-dimensional anatomy of the physis [3, 25, 28–31]. This nuanced knowledge will likely aid the surgeon who attempts resection of a physeal arrest.

Plain Radiographs

Standard orthogonal radiographs are an appropriate initial diagnostic study for any patient with a suspected physeal arrest. Though advanced imaging modalities allow for more accurate determination of the presence, extent, and exact location of a physeal arrest, much information can be garnered from plain radiography. Physeal narrowing and areas of bridging can often be seen using plain radiographs prior to the development of deformity (as demonstrated in Figs. 5.1 and 5.5). More readily determined, however, are the presence and orientation of growth arrest lines, which often are the earliest heralds of altered growth [4]. These thin sclerotic lines develop at the growth plate during most physeal insults. If normal growth resumes, this line becomes progressively distant from the physis. If normal symmetric physeal growth resumes, the arrest line will remain parallel to the physis. In the presence of a growth arrest, the line (if the arrest is peripheral) or lines (if the arrest is central) will converge towards the area of abnormal growth. Identification of such convergence should alert the clinician to a likely arrest and will usually prompt additional imaging to assess the extent of the bar (see Fig. 5.1).

Computed Tomography

Computed tomography (CT) is often the imaging modality of choice for assessment of a physeal bar. A CT scan can generally be obtained quickly and without sedation, which is often necessary for magnetic resonance imaging (MRI). Modern image reconstruction and manipulation software usually allow for excellent visualization of both normal and

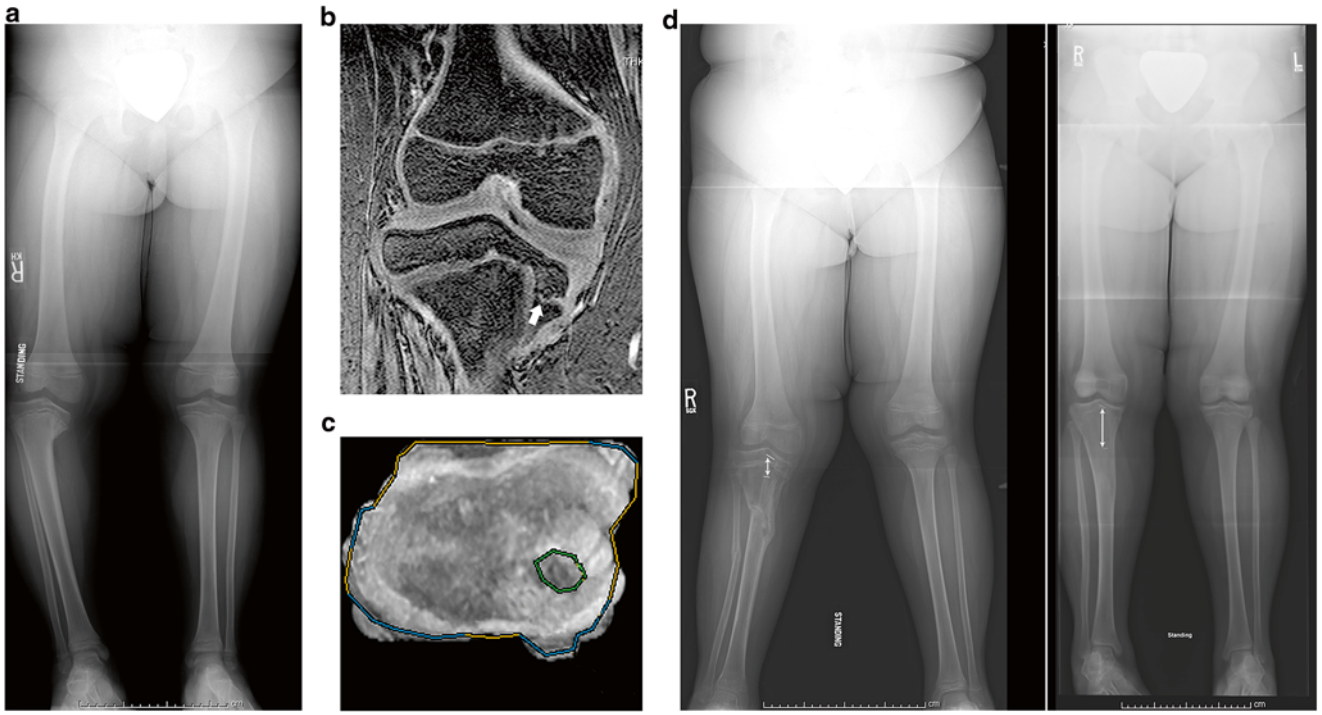


Fig. 5.7 (**a**) AP bilateral lower extremities of an 8-year-old with infantile Blount's. (**b**) Coronal MRI demonstrating a small physeal bar in a proximal tibia (*arrow*). (**c**) Modern software can facilitate construction of a "map" which defines the area of physeal closure. (**d**) Early postoperative and final standing radiographs. Note the resumption of growth as demonstrated between the markers (*arrows*)

diseased physis, thereby allowing for very accurate determination of the extent and location of a physeal bar (see also Fig. 5.10a) [30].

Magnetic Resonance Imaging

MRI is a highly sensitive test for the detection and delineation of areas of physeal arrest. This study allows for the localization of both normal physeal cartilage and regions of bony bars. Furthermore, the use of three-dimensional MRI utilizing fat-suppressed spoiled gradient-recalled echo sequences allows for manual reconstruction of the growth plate, thereby allowing the clinician to accurately map an area of growth arrest which can be invaluable in developing a surgical plan for epiphysiolysis (see also Fig. 5.7c) [3, 29, 31, 32]. Accurate mapping of a physeal bar should include localization in relation to known and identifiable anatomic markers which can be useful during surgical resection [33, 34] (Fig. 5.7).

Growth Remaining

The greater the growth potential in a physis, the more likely it will resume growth following resection of a physeal bar, and the greater the benefit from the restoration of growth. The amount of growth remaining from a particular physis is determined by which physis is involved and the age of the patient. A number of studies have assessed longitudinal limb growth and allow general estimates of yearly growth from each long bone physis [35–37]. As physiologic or skeletal age may differ significantly from chronological age; the former is important to consider when assessing growth potential. Traditionally, this is most commonly achieved using standards for skeletal age published in atlas format using radiographs of the left hand and wrist [38]. Skeletal age can also be determined using AP and lateral radiographs of the elbow [39, 40]. This information, in conjunction with the knowledge that extremities typically grow until a bone age of 14 in females and 16 in males, can provide estimations of the amount of growth remaining in a particular physis.

Surgical Indications

Traditional indications for physeal bar excision include a growth plate arrest occupying less than 50 % of the involved physis in a child with more than 2 years or 2 cm of growth remaining at the involved physis. Interestingly, there is surprisingly little scientific evidence to support this "dogma" [4, 12, 27]. Because of the modest results of physeal bar resection in most modern reports, we believe that physeal bar resection is ideally performed in patients in the first decade

of life, and patients closer to skeletal maturity (i.e., only 3 or 4 years of limb growth remaining) may be better treated with completion epiphysiodesis, with or without a contralateral epiphysiodesis to mitigate any limb length difference. Consideration can be given to resection of larger bars in very young children as success might spare the child from multiple subsequent procedures. Lastly, excision of a physeal arrest is ideally performed prior to the development of angular deformity. Once deformity is present, consideration must be given to concomitant corrective osteotomy.

Surgical Technique and Pitfalls

Once the determination has been made to resect a physeal bar, meticulous preoperative planning should address the following considerations: need for concomitant deformity correction, approach for bar resection, interposition material following resection, and radiographic markers in the epiphysis and metaphysis to aid in assessment of future growth.

Role of Osteotomy

The decision to surgically correct any concomitant angular deformity should be the first consideration when planning a physeal bar resection. In our experience, it is unlikely that a successful bar resection will "overgrow" to correct angular deformity. Furthermore, we believe that it would be unwise to plan for "guided growth" via physeal tethering in an injured physis. Thus, osteotomy and acute correction of deformity are often the most attractive option when a physeal bar is associated with clinically unacceptable angular deformity. Acute correction of the angular deformity to restore normal alignment has the added theoretical benefit of eliminating any asymmetrical forces across the physis which may be contributing to abnormal physeal growth via the Heuter-Volkmann principal. If acute angular correction is performed, careful consideration and planning must be given to determine appropriate fixation of the osteotomy. Fortunately, many of these patients are young and can be managed with relatively simple constructs including variations of "pins and plaster" (Fig. 5.11). Obviously, more extensive internal or external fixation is another option.

Surgical Approaches (Tips and Tricks)

Once the need for osteotomy to correct angular deformity has been assessed, the determination can be made regarding the best approach to resect the physeal bar. Peripheral bars are simplest and are approached directly (Fig. 5.8). Central bars can be more challenging and can be addressed in a variety of ways, including through a metaphyseal window or tunnel (perhaps most common) (Fig. 5.9), percutaneously (ideal for Type C bars as illustrated in Fig. 5.10), and directly through an osteotomy (most commonly an option when performing concomitant angular deformity correction). Although distraction through the physis prior to bar resection has been reported [41], we have no experience with this technique and have concerns that distraction across the physis may further damage the germinal layer and lead to premature closure. The surgeon should select and approach that will give him or her the confidence that he or she can reach the entire extent of the bar. Most commonly the bony bar is resected with a high-speed burr. It is paramount that the surgeon visualizes normal physis separating the epiphysis and metaphysis. Again this is relatively straightforward for peripheral bars but can be more challenging for central bars. Fiber-optic lighted suction, dental mirrors, and arthroscopes may aid in confirming a normal physis "360°" around the resection of a central bar. Additionally, radio-opaque dye can be placed in the resection bed to allow fluoroscopic assessment of the extent of the resection (Figs. 5.9 and 5.11).

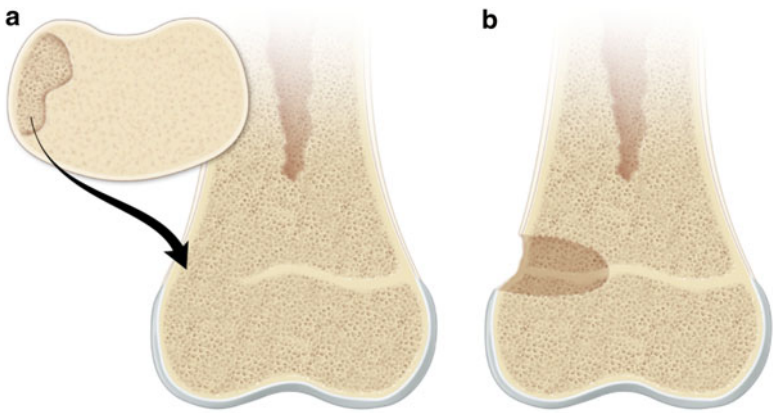


Fig. 5.8 Illustration of peripheral physeal bar resection

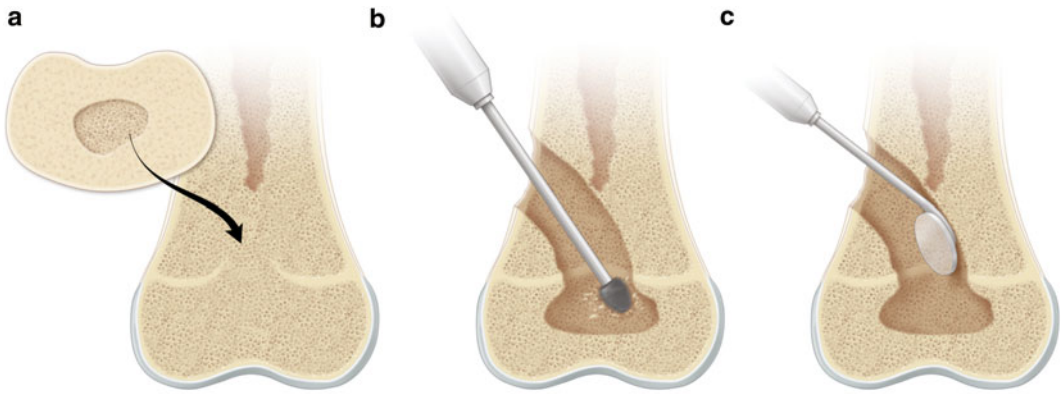


Fig. 5.9 Illustration of central physeal bar resection. Reprinted with permission from Herring JA (ed). Tachdjian's Pediatric Orthopedics: From the Texas Scottish Rite Hospital for Children. 5th ed. Philadelphia: Elsevier; 2013

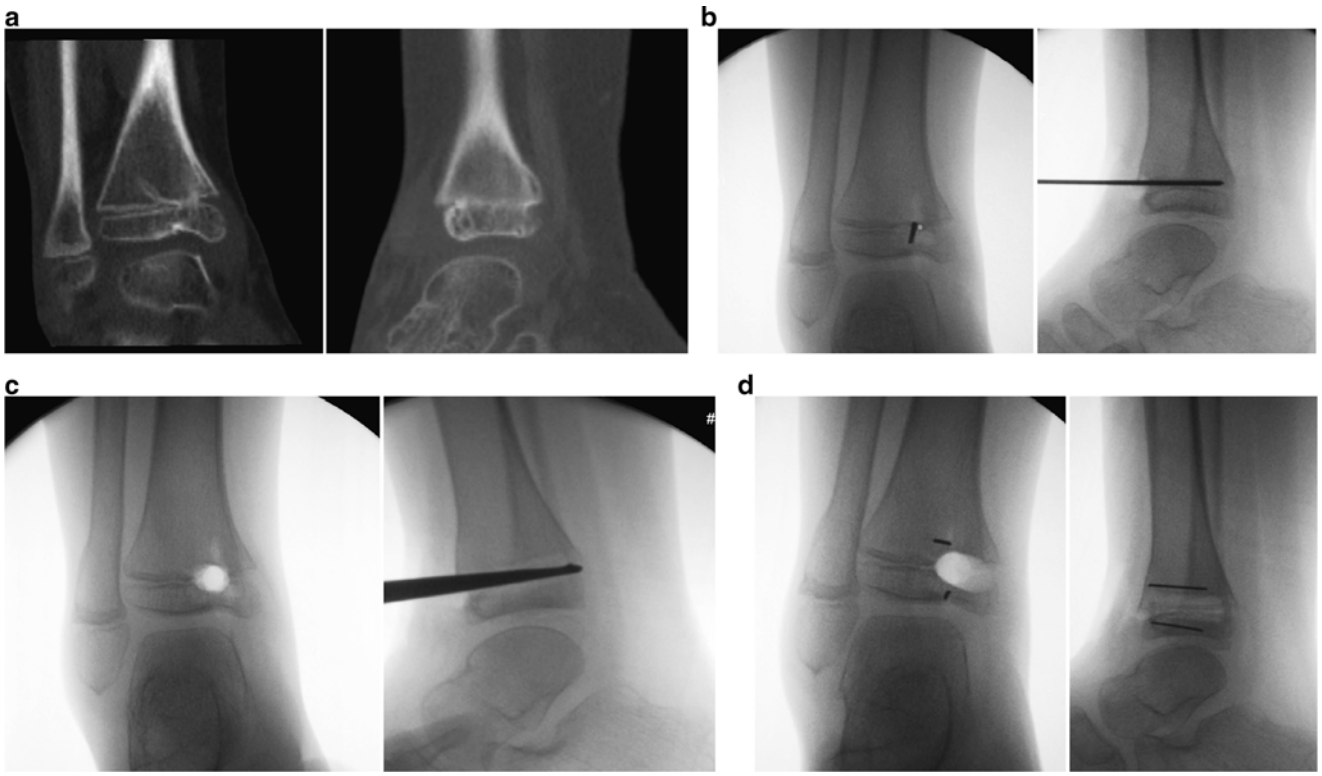


Fig. 5.10 (**a**) Coronal and sagittal CT scan demonstrating Type C central bar following a Salter–Harris IV fracture of the medial malleolus. (**b**) Intraoperative fluoroscopy is used to place a guide pin through the area of physeal closure. (**c**) A cannulated reamer is placed over the guide to resect the bony bar. Resection is confirmed with a curette. (**d**) Final appearance after radiographic markers are placed to facilitate postoperative assessment

Interposition Material

Once the surgeon is satisfied that the bar has been completely excised, interposition material should be placed in an effort to limit recurrence. Interposition materials have included bone wax, fat, silicone, and cranioplast [4, 6, 9, 27]. (Silicone is no longer clinically available.) While a few recent investigations have assessed the possibility of cartilaginous cells as an interposition material, they are, to our knowledge, not yet clinically viable [42–44]. Subsequently, for peripheral bars we ensure that the adjacent periosteum has been widely and sharply excised from the resection area and apply bone wax

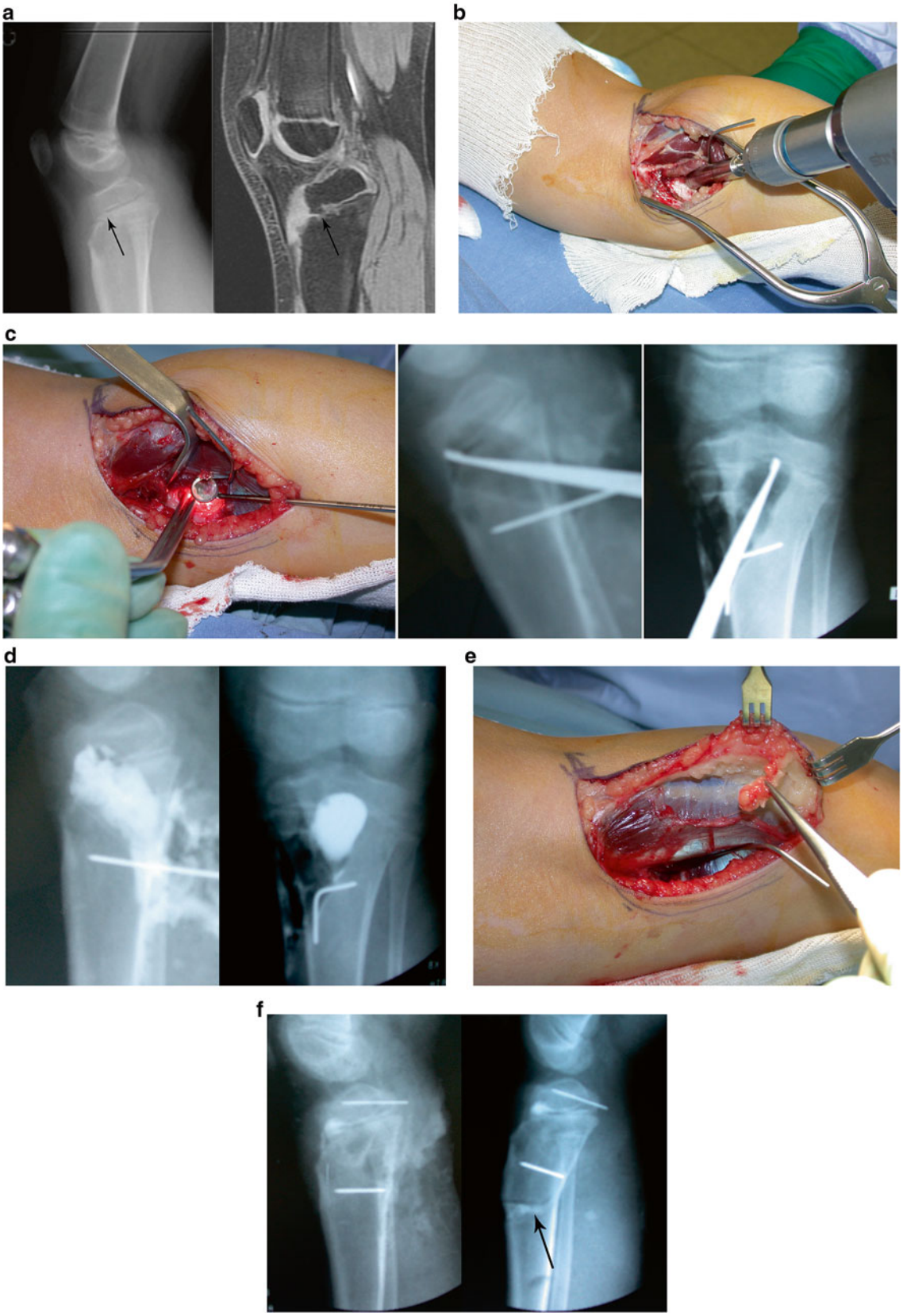


Fig. 5.11 (**a**) Lateral X-ray and MRI showing a recurvatum deformity and an anterior physeal bar (*arrow*) following a proximal tibial fracture. (**b**) Cannulated ACL reamer is used to create a tunnel through the posterior tibial metaphysis after placing a guide wire in the physeal bar. (**c**) Fiber-optic lighted suction, dental mirrors, and fluoroscopy are used to confirm adequate resection of the bar. (**d**) Radio-opaque dye can be used to document the extent of the resection. (**e**) Local fat is harvested to use as an interposition material. (**f**) Lateral radiograph before and after opening wedge osteotomy with allograft (*arrow*) to correct angular deformity. Note the radio-opaque markers in the epiphysis and metaphysis to help with postoperative assessment of subsequent growth

to the exposed metaphyseal and epiphyseal bone. For central bars we prefer local fat interposition. Although cranioplast has the purported advantage of completely and permanently filling the resection void, we believe that it has several disadvantages when further reconstructive procedures are required. Namely that it is difficult to remove and leaves a bony deficit post-removal, often exactly at the level of deformity, significantly complicating subsequent procedures.

Radiographic Markers

The surgeon's final intraoperative task is to place small radiographic markers in the epiphysis and metaphysis to serve as markers to make comparison measurements on radiographs over time. We usually use small k-wires, although vessel clips are also effective. It is imperative that the surgeon communicate to the family the need to follow the child to skeletal maturity. Close follow-up can allow early detection of recurrence and allow completion epiphysiodesis prior to the development of angular deformity. Even with excellent resumption of growth the surgeon should expect the involved physis to close earlier than the contralateral limb as the patient nears skeletal maturity [27]. In our experience nearly 75 % of patients undergoing a physeal bar resection will require a subsequent procedure [12].

Clinical Outcome

The reported results of physeal bar resection are variable. Perhaps the most cited report is Peterson's 1984 report where some growth was noted in 70 of 71 cases. They noted a wide variation in the growth of the operated bone relative to the normal bone (0–200 %) with a mean of 84 % in patients followed to maturity. Perhaps not completely appreciated, they also noted that in some cases the "normal" bone was treated with an epiphysiodesis which "favorably influences the result expressed as a percentage" [10]. Williamson and Staheli reported "near-normal" longitudinal growth (>93 % of the contralateral limb segment) and no progression of angular deformity in 11 of 29 patients [27]. More recently, Hassler and Foster reported results more in line with our experience, noting fair or poor results in 14 of 22 patients [33].

Summary

Physeal bar excision is an important tool in the armamentarium in treating children with acquired limb deformity. The surgeon considering physeal bar resection should be familiar with the anatomy and physiology of the normal physis and the etiology and anatomy of the physeal growth arrest and have an understanding of skeletal maturation. Because of the complexities of the factors involved, the outcome of attempted bar resection can be unpredictable. As such, the surgeon should educate patients and families of the importance of close clinical follow-up as well as the likelihood of needing subsequent procedures. Despite the unpredictable nature of physeal bar resection, it can result in meaningful and gratifying resumption of growth in carefully selected patients.

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Acute Deformity Correction Using an Osteotomy

6

Vrisha Madhuri and Sangeet Gangadharan

In this chapter, we discuss the indications, principles, specific considerations, choice of hardware and complications of acute correction of lower limb deformities using an osteotomy.

Principles of Acute Deformity Correction

General Considerations

It is generally preferable to perform an acute correction when the magnitude of angular correction required is small and the deformity is in a single plane. In children, the bone gap created by an opening wedge osteotomy heals well due to better healing potential and a thick, biologically active periosteum. The skin overlying the affected area should be healthy as acute correction can cause skin necrosis because of stretch or impaired vascularity. Long-standing deformities are associated with soft-tissue contractures on the concave side of the deformity and this must be taken into account when planning.

Age Considerations

The outcome of treatment of certain deformities depends on the age at which the corrective osteotomy was performed. This is due to correction or worsening of the deformity or the underlying bony pathology with increasing age. In some conditions there is a high prevalence of recurrence when correction is done at an earlier age. For instance, in congenital posteromedial bowing of the tibia, it is wise to defer surgery till at least after 3–4 years of age, since spontaneous correction of the deformity is noted to a large extent, and the complication rate and chance of recurrence are higher if done at a younger age 1. Soft tissue as well as bony correction of deformities in cerebral palsy also tend to fail if performed before 8–10 years of age due to worsening spasticity with growth and persisting muscle imbalance [2].

The magnitude of correction should take into account the possibility of progressive deformity with future growth. Physeal injuries and conditions such as Blount's disease where only a portion of the growth plate is affected may require fusion of the healthy (lateral) portion of the open physis or overcorrection of the deformity (Fig. 6.1). In some situations, deformity correction may be combined with treatment of the pathology such as excision of a physeal bar (Fig. 6.2) [3, 4].

Principles Pertaining to Correction of Lower Limb Alignment

Minor changes in lower limb alignment following corrective osteotomy often remodel in children. Bone remodelling following correction occurs in both sagittal and coronal planes, although such remodelling is greater in the plane of joint movement and when the osteotomy is close to the growing end of the bone. This remodelling occurs at the site of osteotomy in accordance with Wolff's law [5]. This law infers that remodelling will occur on the compression side by laying new bone. Pauwels postulated that malalignment of bone produces a differential growth at the physis which tends to realign the shaft perpendicular to the major joint reaction forces [6]. This observation is similar to the epiphyseal pressure rule of Heuter-Volkmann (growth is retarded by increased mechanical compression] [7]. Any residual deformity would tend to decrease by remodelling in 4–5 years following surgery [8]. Seventy-five percent of this remodelling occurs at the physis and 25 % by bone drift or translation [9, 10].

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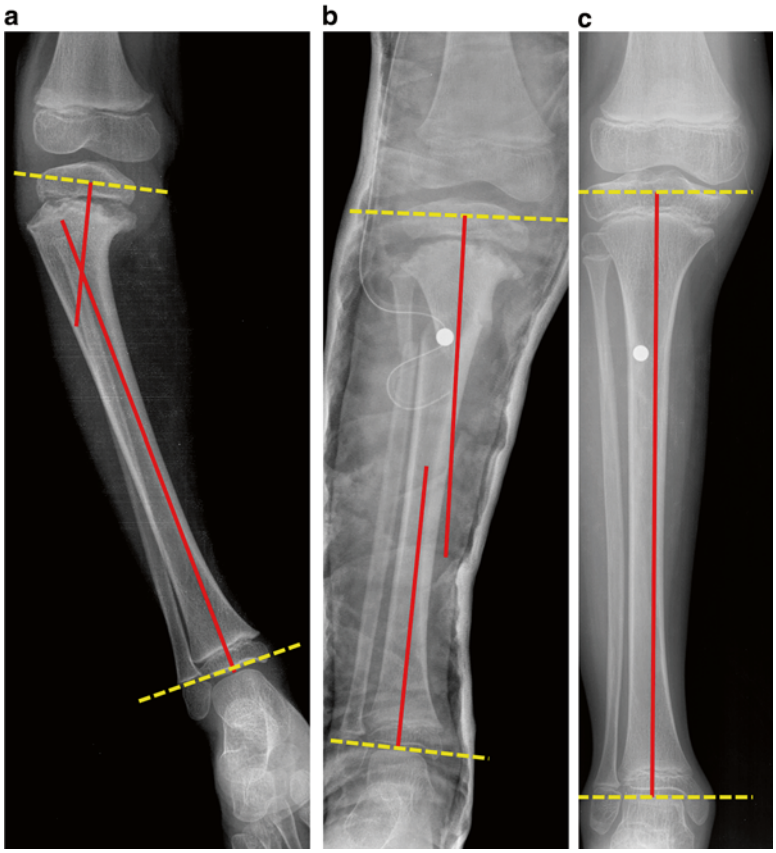


Fig. 6.1 (**a**) Radiograph of a 7-year-old girl with Blount's disease. Apex of the deformity is seen at the intersection of the two lines (*red lines* indicate the proximal and distal tibial mechanical axes). (**b**) Overcorrection by Rab's osteotomy. (**c**) Radiograph at 3-year follow-up shows good alignment of the mechanical axis of the tibia

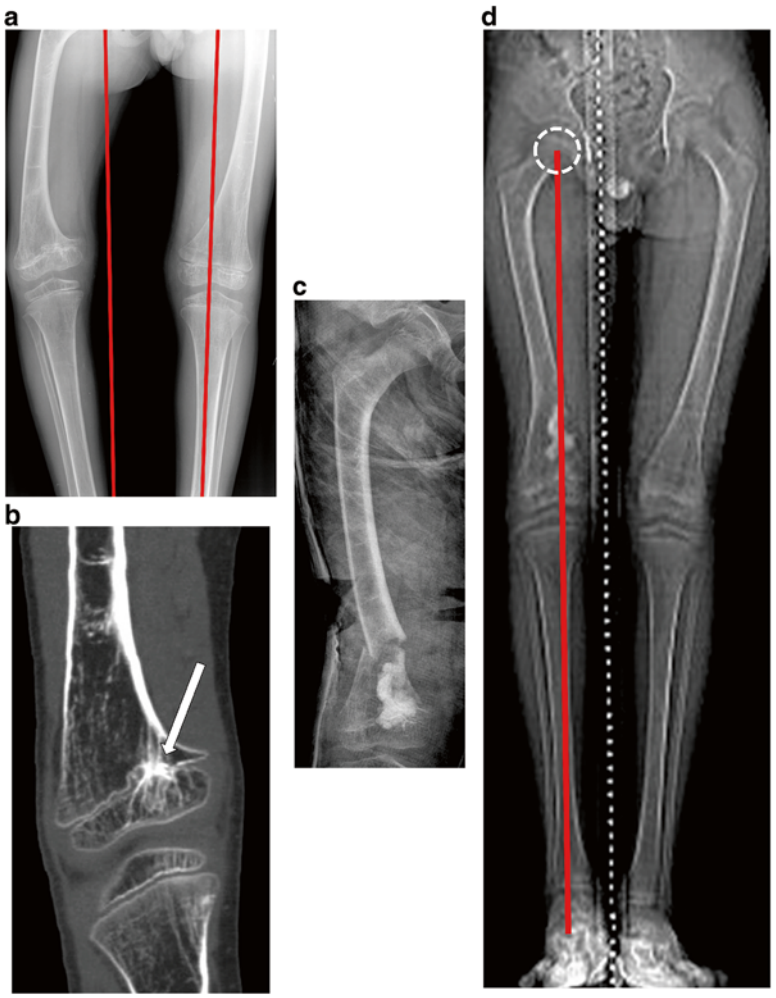


Fig. 6.2 (**a**) Right-sided genu varum in a 7-year-old boy with hemi-epiphyseal arrest of the right distal femur (*red lines* represent the mechanical axes of both lower limbs). (**b**) CT scan shows a peripheral physeal bar (*white arrow*). (**c**) Bar resection and cement interposition were performed in addition to corrective osteotomy of distal femur. (**d**) CT scanogram (supine) shows restoration of the mechanical axis and proximal migration of cement indicating restoration of growth at 18-month follow-up. A supine CT scanogram was taken in this case; however, a standing full-length radiograph is a more accurate measure of alignment and should be preferred

Thus a perfect anatomical alignment may not be necessary; however long-term follow-up is required in children with growth remaining.

Influence of Level of Osteotomy

In children, often the osteotomies are at a site distant from the apex or center of rotation of angulation (CORA). When planning an osteotomy, one should consider various factors such as the proximity of the CORA to the adjacent physis and/or joint, presence of pathological bone and neurovascular structures in the vicinity. Overall, metaphyseal osteotomies have a better healing potential than diaphyseal ones. The ease of rotational correction is the same at both levels in case of femur, but the proximal shaft is preferable as large corrections at the distal third would be theoretically at the expense of an undesirable alteration in the patella-femoral dynamics [11]. Any corrective osteotomy or lengthening should not be performed at the site of pathological bone to avoid healing problems [12, 13]. The exceptions are where the entire bone is pathologic like in osteogenesis imperfecta or fibrous dysplasia.

Adjuvant Fibular Osteotomy: When and Where?

The fibula acts as a strut to prevent large angular or rotational corrections of the tibia. Thus, with opening wedge correction of the tibia, an adjuvant fibular osteotomy is often performed along with tibial osteotomy.

- Fibular osteotomy should be avoided in its proximal third to safeguard the common peroneal nerve [14]. In the middle third there is a risk of damage to peroneal vessels [15–17].
- Classic recommendation of doing a distal tibial de-rotation without a fibula osteotomy is for rotational corrections of up to 35° [18]. However, de-rotational osteotomies of up to 55° internal and 45° external have been reported without fibular osteotomy and without neurovascular compromise [19]. We however prefer a fibular osteotomy while doing a rotational correction of greater than 30°.
- We tend to perform osteotomy at the proximal and middle third junction in case of proximal tibial angular osteotomy and close to distal and middle third junction in distal tibial/rotational osteotomies.
- A segment of the fibula should be resected when the desired angular correction in tibia is considerable [20]. Fibula osteotomy, if performed at a site far away from CORA, leads to wide displacement of the fibula and in such situation may require resection of the projecting portion of fibula. A distal tibio-fibular fusion should be performed to avoid ankle instability when a fibular pseudarthrosis is anticipated following such resections [21].
- At least 10 % of the total fibular length should be preserved distal to the osteotomy to avoid ankle instability [22]. Our experience shows that proximal fibular migration occurs if the fibula does not unite, irrespective of the level of osteotomy.

Principles Pertaining to Specific Osteotomies

Opening Wedge Osteotomy

The axis of deformity correction lies on the convex cortex. The overall length of the limb increases following deformity correction. The final length achieved will be equal to the sum of the lengths of the convex border of the deformity (Fig. 6.3a). If the combined total correction exceeds 2.5–3 cm of lengthening, shortening may be required as a cautionary measure since significant lengthening could compromise the neurovascular status. The nerve stretch in certain circumstances may be further influenced by the direction of rotation such as the risk of iatrogenic injury being somewhat less in external than internal rotation when performing a proximal tibial osteotomy because of tethering of the peroneal nerve at the proximal fibula [23]. When the axis of correction lies away from the osteotomy site further lengthening is achieved; for instance in calcaneal lengthening osteotomy the axis of correction lies away from the calcaneum, i.e. at the talar head [24] (Fig. 6.4).

Closing Wedge Osteotomy

The axis of deformity correction lies on the concave side of the deformity. The overall length of the limb increases following deformity correction and is equal to the sum of the concave borders of the deformity as shown in Fig. 6.3b. Generally, a closed wedge osteotomy in a straight bone reduces the limb length whereas the bone length increases or remains the same with an open wedge osteotomy, for instance with compensatory osteotomies for the proximal femur in Perthes disease and non-union of the neck of femur (Fig. 6.3c).

Dome Osteotomy

Dome osteotomy is ideal for deformities with CORA close to the joint line such as in physeal or metaphyseal injuries near the knee or ankle. The displacement of the osteotomy is directly proportional to the radius of correction. Dome osteotomy causes minimal length alteration as the CORA is usually at or near the site of deformity in the metaphysis. The advantage of such an osteotomy is good bony contact and intrinsic stability. However this technique is limited by its inability to correct rotational malalignment.

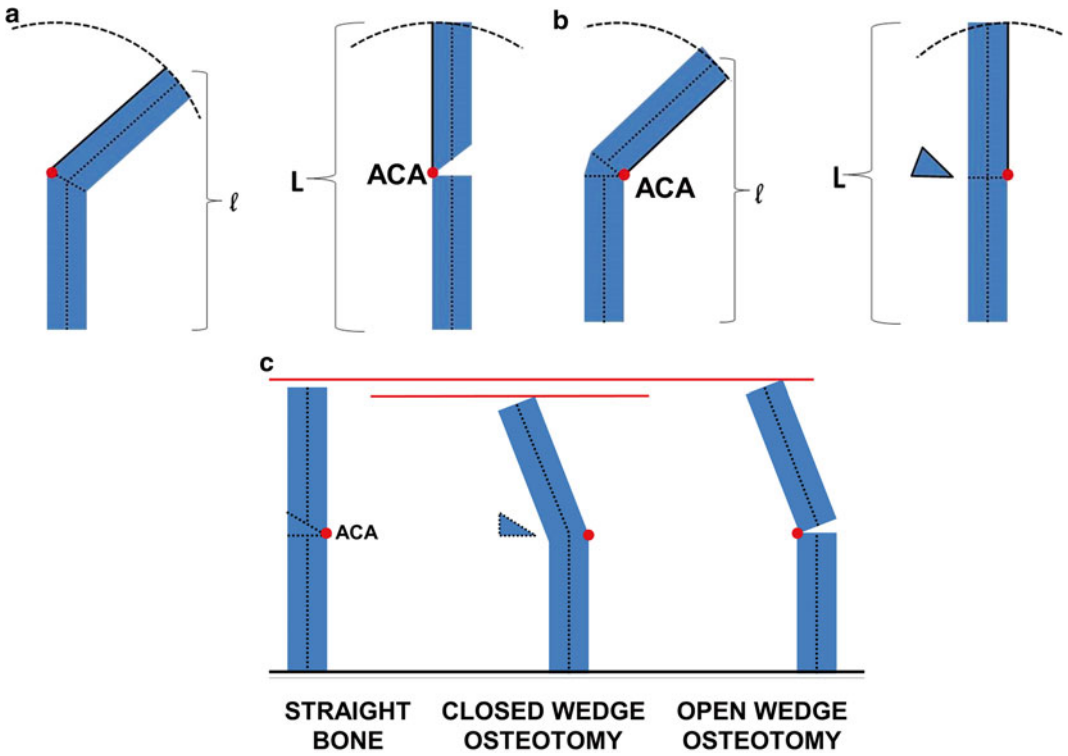


Fig. 6.3 (**a**) Open wedge osteotomy in a deformed bone of length "l" showing increase in length of the bone. The final length "L" is the sum of convex borders of the deformed bone. (**b**) Closed wedge osteotomy in an angular deformed bone showing increase in length of the bone. The final length "L" is the sum of concave borders of the deformed bone (*ACA* axis of correction of angulation). (**c**) In a straight bone an open wedge angular osteotomy causes less shortening than a closed wedge as shown by the *red lines*. Hence, the former is preferred when minimal lengthening is desired along with corrective osteotomy

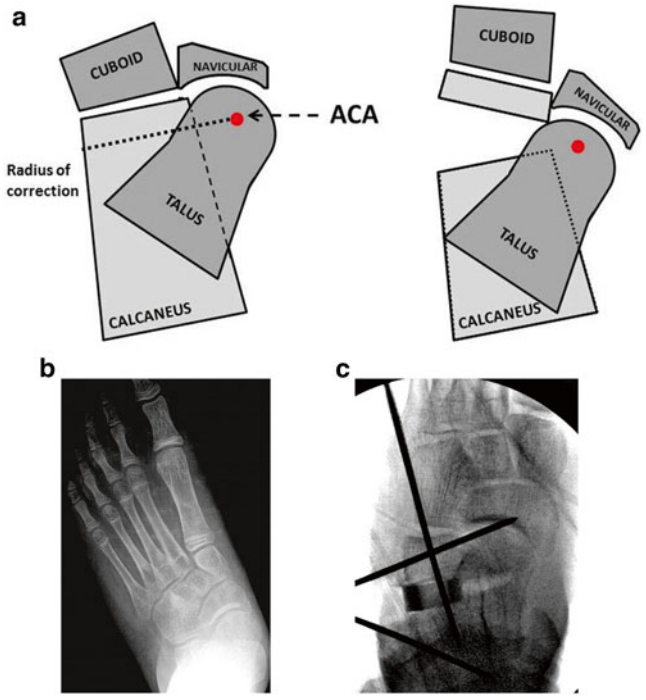


Fig. 6.4 (**a**) Axis of correction of angulation (ACA) is located away from the calcaneum, i.e. at the talar head. The effective lengthening along with open wedge osteotomy can be seen. (**b**) Preoperative and (**c**) intraoperative radiographic views of left calcaneal lengthening osteotomy and the increase in talar head coverage brought about by it

Angular Correction and Translation

An angular osteotomy involves shifting of mechanical axis in the direction of correction of the deformity; that is, the mechanical axis shifts medially in varus and laterally in valgus osteotomy. Hence translation of the distal fragment has been advised in order to restore the mechanical axis [23]. A valgus proximal femoral osteotomy requires lateralisation of the distal fragment. However when doing an angular correction below the knee the direction of translation is opposite, i.e. lateral translation for varus and medial translation for a valgus osteotomy. Glard et al. showed that proximal femoral varus osteotomy with medial translation in Perthes disease did not affect the mechanical axis deviation at the knee [25]. There are a number of studies which show remodelling of the femoral neck-shaft angle following varus osteotomy with or without translation [25–30]. In a child, do we need to translate the distal fragment when performing angular correction to optimise the mechanical axis? Sanghavi et al. found that after varus de-rotation osteotomy, the neck-shaft angle tends to revert to normal depending on the age at which the operation was undertaken [26]. Tercier et al. showed that following proximal femoral osteotomy in Perthes, the resulting mechanical axis deviation is usually subtle and clinically insignificant in children [27]. Following varus osteotomy of the proximal femur in cerebral palsy

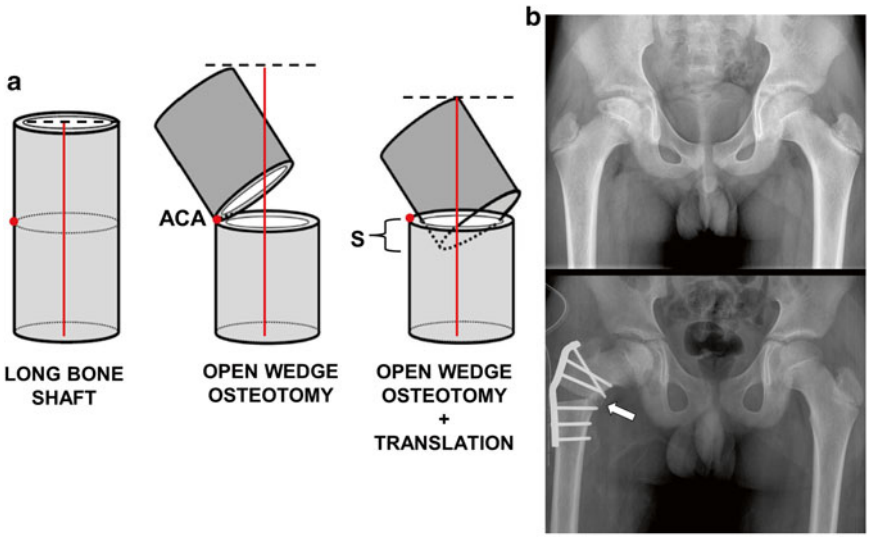


Fig. 6.5 (**a**) Effect of angular correction on length of a straight bone. *Red solid lines* indicate length of the straight bone. Increase in length is seen following opening wedge osteotomy without translation. Translation has relatively less effect on the final limb length. "S" is the amount of shortening effected following translation. *ACA* axis of correction of angulation. (**b**) *Top*: Radiograph of a child with Perthes disease of the right hip. *Bottom*: A varus de-rotation osteotomy was performed. The medial intact periosteal hinge is shown (*white arrow*)

60 % of the correction is lost after 8 years by remodelling [28]. In pelvic support osteotomy it has been shown that up to 30° of valgus angulation at the osteotomy site does not cause any significant change in mechanical axis [31]. Therefore it is our assumption that translation of the distal fragment during angular correction of proximal femoral deformities is not mandatory in younger children unless the magnitude of correction is outsized, such as greater than 30°. This observation is also supported by the acceptability of overriding/bayoneting fracture reduction in younger children where after a few years complete restoration of the anatomy occurs [32]. The advantages of not translating are that the periosteal hinge medial or lateral stays intact and the final lengthening achieved is greater than with translation (Fig. 6.5).

Oblique Osteotomy

An oblique osteotomy corrects multi-planar deformities at the same time. A meticulous preoperative planning is essential as it involves correction of the deformity in various planes through a single osteotomy. Lesser the obliquity of the plane, lesser is the bony contact after angular correction but easier and greater is the rotational correction such as in Rab's osteotomy (Fig. 6.6).

Lengthening/Shortening Osteotomy

Lengthening osteotomy in a single stage is best achieved either by a step cut/long oblique osteotomy to maintain adequate contact while lengthening at the same time such as in distal fibula, metatarsals or proximal ulna. Acute lengthening of 1.7 cm and 3.2 cm for the tibia and femur, respectively, has been performed with minimal complications in adults [33]. Opening wedge angular correction can some time offset the shortening caused by varus angulation and gain in length to some extent, for instance in Perthes disease (see Fig. 6.4a). It is important to keep in mind the change in length ratio as compared to the companion bones in case of osteotomies of leg and foot [34]. Acute shortening of up to 3 cm can be performed safely in young adults with open tibial fractures with bone loss [35]. Further shortening should be gauged based on the clinical assessment of limb vascularity. Fibular shortening osteotomy can be combined with proximal tibia corrective osteotomy in achondroplasia to correct the lateral collateral laxity and fibular overriding [36]. The major disadvantage of lengthening/shortening osteotomy

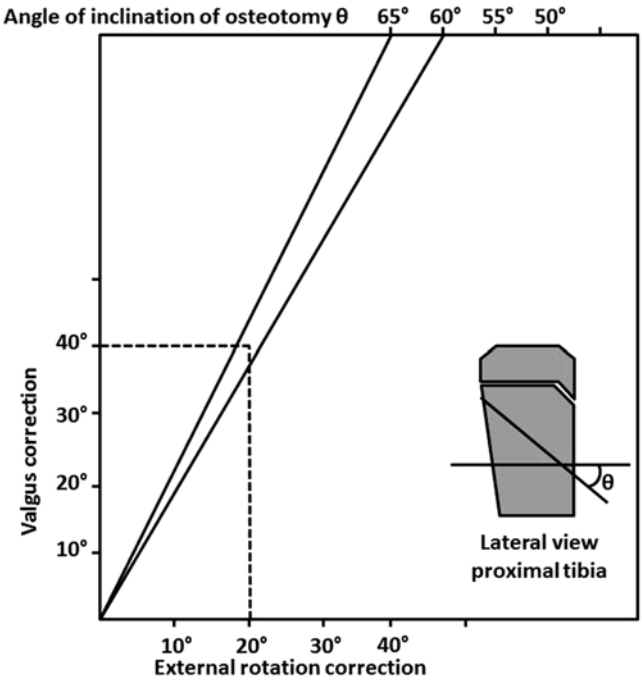


Fig. 6.6 In this example the amount of desired external rotational (20°) and valgus angular correction (40°) (indicated by *dotted lines*) is plotted on the graph. θ, the angle of inclination of proximal tibial osteotomy with the horizontal lies between 60 and 65°. Adapted from [74]

with acute correction is that the amount of correction safely gained is limited. If the magnitude of angular deformity correction is considerable, either a shortening osteotomy or distraction osteogenesis is preferred to avoid significant risk to the neurovascular structures and soft-tissue tightness (Fig. 6.7).

Special Considerations

Deformity Secondary to Physeal Injury

Angular deformity secondary to physeal insult is usually progressive until skeletal maturity. The following strategies can be adopted for such corrections:

- Overcorrection of the deformity to compensate for future progression of deformity.
- Combination of acute corrective osteotomy and excision of physeal bar and interposition.
- Acute corrective osteotomy combined with a growth modulation technique: This would however invoke a shortening effect due to arrest of either side of the physis.
- An opening wedge osteotomy would simultaneously address the existing limb length discrepancy.

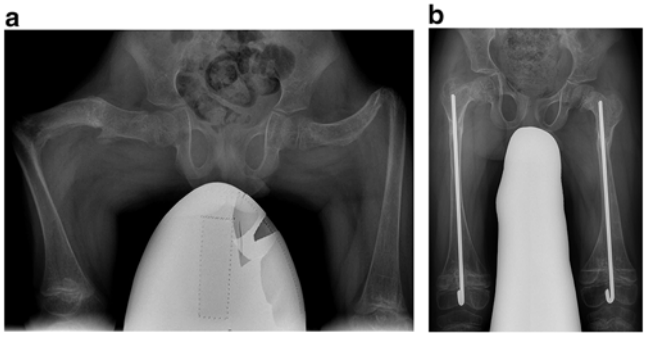


Fig. 6.7 (**a**) Radiograph of a 10-year-old girl with osteogenesis imperfecta and bowing deformity of both femora. Restoring the length of the femur is limited by the stretch on neurovascular structures and soft tissues. (**b**) Hence, corrective osteotomies are combined with shortening

Mango Slice Effect

In the Indian subcontinent, raw mango slice, a savoury seasonal snack for children, is split into multiple radial cuts for better seasoning. This makes a good illustration for osteotomy with multiple open wedges for diaphyseal deformity as seen following rickets and other metabolic disorders. The concept and execution are explained in the following illustration. Small corrections at two or more CORAs allow lengthening and deformity correction spread over the length of bone (Fig. 6.8).

The Deformity Memory Effect

Paediatric congenital and developmental conditions such as developmental coxa vara, fibular hemimelia and posteromedial bowing of tibia have a memory because of associated soft-tissue contractures. Following an osteotomy, either the deformity can recur by bending the callus or slow relapse occurs due to abnormal forces on the growing physis [Heuter-Volkmann]. In these situations the osteotomies should be combined with soft-tissue lengthening or shortening of the bone (Fig. 6.9).

Cozen's Phenomenon

Certain fractures of the proximal tibia in a growing child are associated with a proximal tibial valgus deformity. These deformities may correct spontaneously over a few years, and thus an acute corrective surgery for the tibial valgus should be deferred for at least 3–4 years following the fracture [37, 38] (Fig. 6.10).

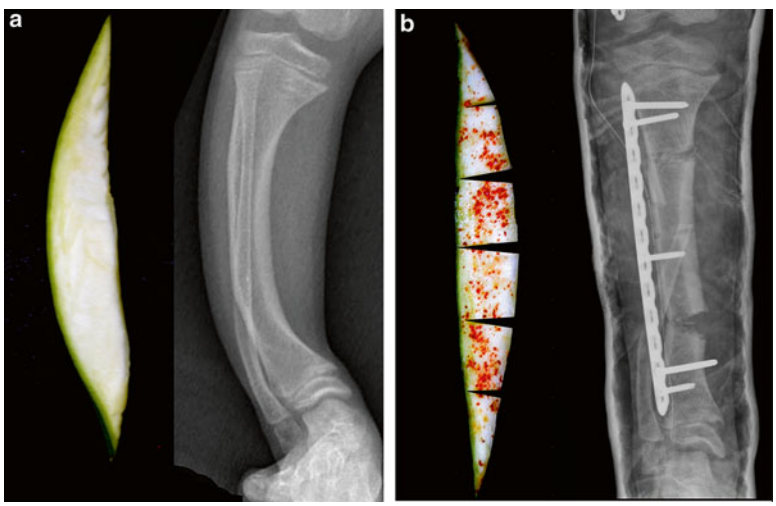


Fig. 6.8 (**a**) An intact mango slice and (**b**) a slit, seasoned one can be compared with multiple opening wedge osteotomies in a bowed tibia

Box 6.1

- Acute deformity is preferred in children and when the magnitude of deformity or length discrepancy is relatively small.
- Soft-tissue constraints should be addressed simultaneously where needed, to avoid recurrence.
- Knowledge of the natural history of the underlying pathology is crucial before executing an acute osteotomy.
- Anatomical alignment of mechanical axis is not mandatory in younger children because of remodelling potential.
- A dome osteotomy can be performed when the CORA is juxta-articular.
- Fibular osteotomy may be necessary for tibial osteotomies for large correction.



Fig. 6.10 Cozen's phenomenon. (**a**) Three-year-old boy with proximal tibia fracture. (**b**) Radiograph after 4 years shows tibial valgus deformity

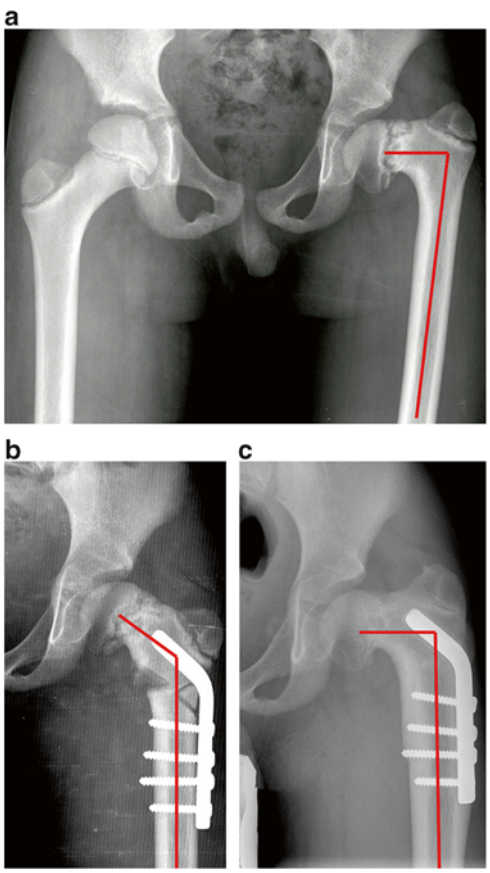


Fig. 6.9 The deformity memory effect. (**a**) Developmental coxa vara in a 15-year-old boy. (**b**) Corrective osteotomy and blade plate fixation. (**c**) Radiograph at 16-month post-op follow-up shows reduction of neck-shaft angle (*red angles*) in spite of the implant position remaining unchanged

Indications

The indications for acute correction with an osteotomy in children are varied and include correction of certain uniplanar or multi-planar skeletal deformities. The indications include angular and rotational correction as well as treatment of mild length discrepancies.

Site-Specific Osteotomies

Metaphysis

Frontal plane angular deformities around the knee [genu varum or genu valgum] related to conditions such as Blount's disease, metabolic bone disease, physeal arrests and coxa vara are often treated with acute correction via a metaphyseal osteotomy (Fig. 6.11).

Epiphyseal

Osteotomies may also be performed at epiphyseal level to accomplish joint elevation such as medial hemi-plateau elevation for Blount's disease [39–41].

Juxta-Apophyseal

Re-directional osteotomies of greater trochanter, calcaneal and tibial tuberosity improve muscle function by optimising the lever arm dysfunction [42–45] (Fig. 6.12).

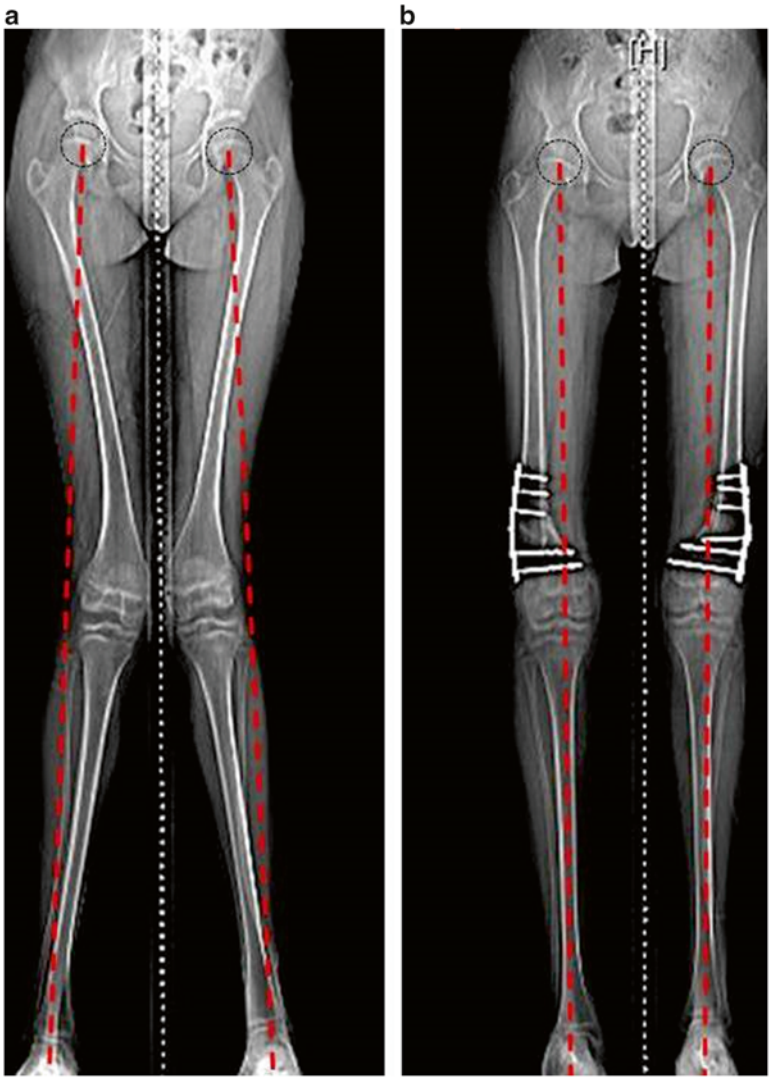


Fig. 6.11 CT scanograms (supine) comparing (**a**) pre-op and (**b**) post-op mechanical axis alignment (*dotted red lines*) in a 14-year-old girl with bilateral genu valgus. She underwent bilateral varus osteotomies of distal femora and fixation with locked compression plates. Supine CT scanogram was taken in this case. However, a standing full-length radiograph is a more accurate measure of alignment and should be preferred

Diaphysis

The common indications for diaphyseal osteotomies include metabolic bone disorders such as rickets, osteogenesis imperfecta, renal osteodystrophy and deformities secondary to traumatic or pathological fractures due to osteomyelitis and cystic lesions of bone. Posteromedial bowing of tibia and congenital limb deficiency form a special subset of diaphyseal osteotomies due to their propensity for complications due to associated soft-tissue inelasticity requiring special measures to avoid complications, for instance, postponing the deformity correction till the child with posteromedial tibial bowing is older and using a locked plate to prevent fractures or bowing of the regenerate that may cause recurrence of the deformity. Congenital pseudarthrosis of tibia is a rare disorder where the primary aim of treatment is to achieve union along with deformity correction and limb lengthening (Fig. 6.13).

Metabolic bone disorders often exaggerate the natural curves of shafts of long bones, often leading to anterolateral bowing in the femur and tibia. The associated diaphyseal deformities are typically a generalised bow and multi-apical in nature, requiring multilevel osteotomies for correction. Severe forms of osteogenesis imperfecta often cause grotesque diaphyseal deformities. The poor quality of underlying bone warrants an intramedullary device to splint the bone after correction (see Fig. 6.7).

Certain Considerations Based on Location of the Deformity

Proximal Femur

In order to avoid damage to the capital physis, most of the proximal femoral osteotomies in children are carried out distant from the site (apex) of deformity. Such acute correction reorients the femoral head in valgus or varus, extension or flexion and rotation and is also used for shortening the

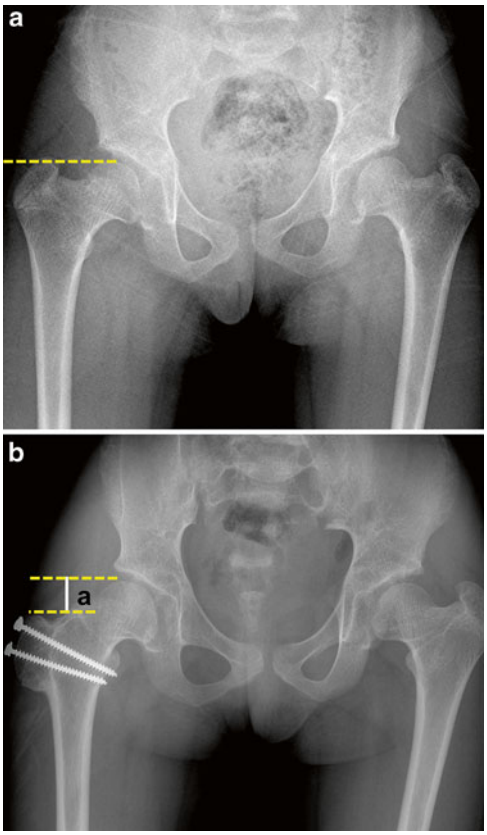


Fig. 6.12 (**a**) A 10-year-old girl with skeletal dysplasia with shortened abductor lever arm and abnormal articulo-trochanteric distance bilaterally. (**b**) Distal and lateral transfer of right greater trochanter was performed. The increased length of abductor lever arm following trochanteric transfer improves abductor power and gait and eliminates trochanteric impingement. a, restored articulo-trochanteric distance

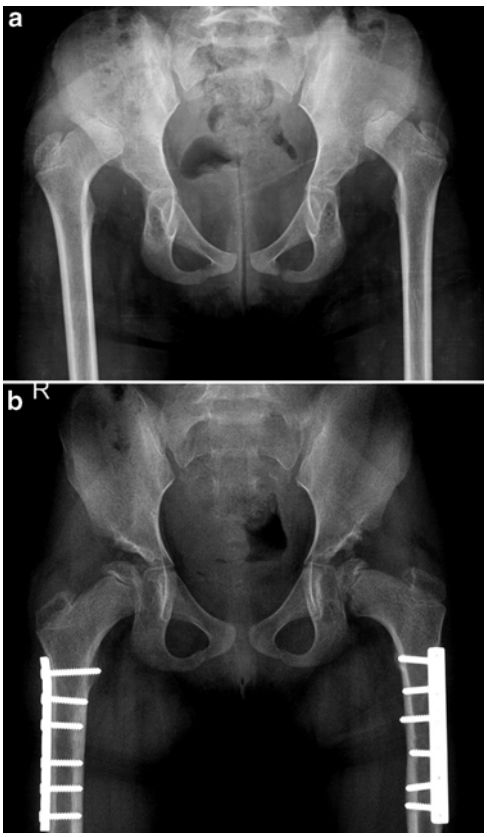


Fig. 6.14 (**a**) Radiograph of a 6-year-old girl with bilateral dysplastic hips. (**b**) Open reduction was performed in conjunction with femoral shortening osteotomy to avoid avascular necrosis and de-rotation to correct anteversion and reduce the hip

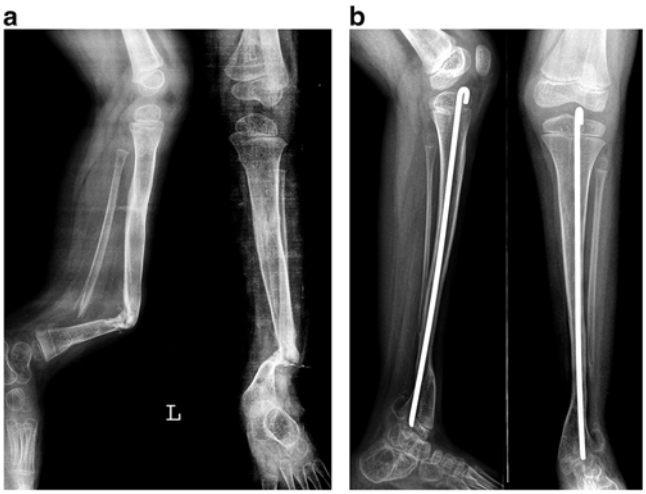


Fig. 6.13 (**a**) Anteroposterior and lateral views showing congenital pseudarthrosis of tibia in a 6-year-old boy. (**b**) Six years following corrective shortening osteotomy, resection of hamartomatous periosteum, bone grafting and revision intramedullary rodding

femoral segment in certain cases of long-standing dislocated dysplastic hips.

Most of these osteotomies cannot be performed at the CORA due to the presence of pathological bone at the site of deformity as in proximal focal femoral deficiency or because of proximity to the proximal femoral physis and the tenuous blood supply to the femoral head [12, 13]. The common example is Perthes disease where proximal femoral varus or valgus osteotomies realign the articulation to either increase femoral head coverage or avoid impingement by the deformed head (Fig. 6.14). A combination of angulation, shortening and rotation is sometimes necessary to achieve optimal reorientation, for instance a proximal femoral osteotomy in a child with a congenital and neurogenic hip dislocation. In developmental coxa vara realignment of the physis to a more horizontal position prevents relapse.

Acute osteotomy of the intertrochanteric and subtrochanteric regions of femur orients the proximal femoral physis more horizontally, thus converting shear forces to compressive forces.

Sub-capital realignment osteotomies are an example of correction at the level of deformity (CORA) in slipped

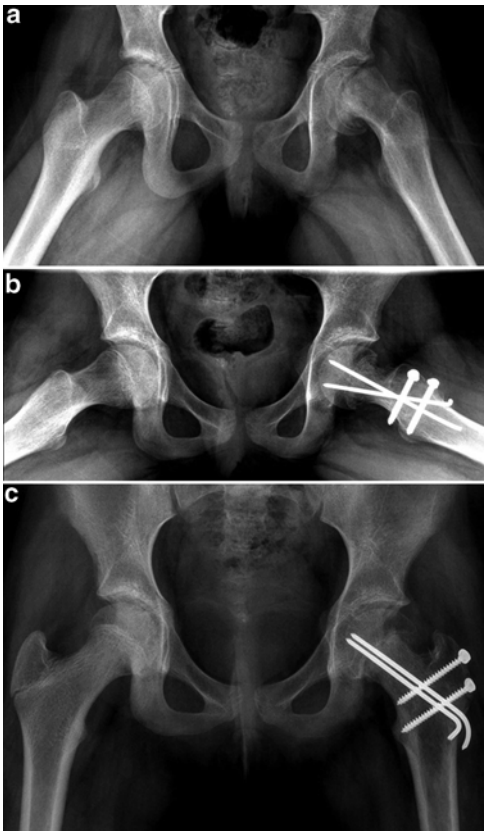


Fig. 6.15 (**a**) Acute on chronic SCFE left hip in a 12-year-old girl. (**b**, **c**) A modified Dunn's osteotomy was done to realign the head and avoid femoro-acetabular impingement

capital femoral epiphysis but are rarely done because of the risk of vascular compromise to the femoral head blood supply (Fig. 6.15).

Osteotomies Around the Knee

Flexion deformity of the knee associated with intractable joint contractures and muscle imbalance such as myelodysplasia, arthrogryposis, cerebral palsy and poliomyelitis can be corrected by supracondylar extension osteotomy of the femur. Such osteotomies reorient the arc of knee motion allowing full apparent extension and improve the child's gait and function. A distal femur-shortening osteotomy can counter the long-standing flexion contracture without significantly affecting the hip extensor power. Correction of coronal plane angular deformities by closed or open wedge is the commonest indication around the knee. While this is now being rapidly replaced by the growth modulation technique, correction via an osteotomy has a role when the deformity is severe and growth potential is limited or where rapid correction is desirable. Lower limb deformities in children with certain skeletal dysplasias where growth modulation techniques play a limited role can also be treated by

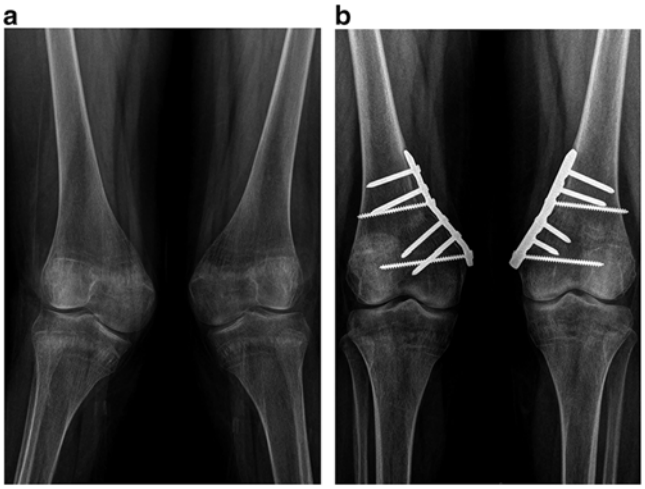


Fig. 6.16 (**a**) Bilateral genu valgus in a skeletally mature girl. (**b**) Bilateral closed wedge osteotomies and metaphyseal plate fixation were performed. Closing rather than open wedge osteotomies were performed to avoid common peroneal nerve injury. In femoral valgus deformity corrections greater than 30° we prefer to decompress the nerve

acute corrective osteotomy. The correction of deformity around the knee requires due consideration to neurological and vascular structures due to their proximity to the osteotomy site (Fig. 6.16).

Acute Correction of Length Discrepancy

Correction of minor shortening can be obtained by acute lengthening. Brachymetatarsia correction by acute lengthening of up to 15 mm can be performed safely without any neurovascular compromise [34]. *Trans*-iliac osteotomy is a rarely done procedure which achieves lengthening when angular correction is not sought. In relative femoral neck lengthening, displacement osteotomy of the trochanter is combined with reshaping of the head, neck and trochanter to eliminate femoro-acetabular impingement and lengthen the lever arm (Fig. 6.17). Acute lengthening of the distal fibula by a step-cut or long oblique osteotomy is another example [43].

Reorientation Osteotomy of Acetabulum

Ganz, triple, Dega's and Salter's osteotomy in dysplastic acetabulum augments femoral head coverage and redistributes hip joint force [45, 46].

Osteotomy with Adjuvant Procedures

Sometimes acute correction of the deformity can be combined with another procedure. This can be unifocal such as physeal bar excision with corrective osteotomy in hemi-physeal

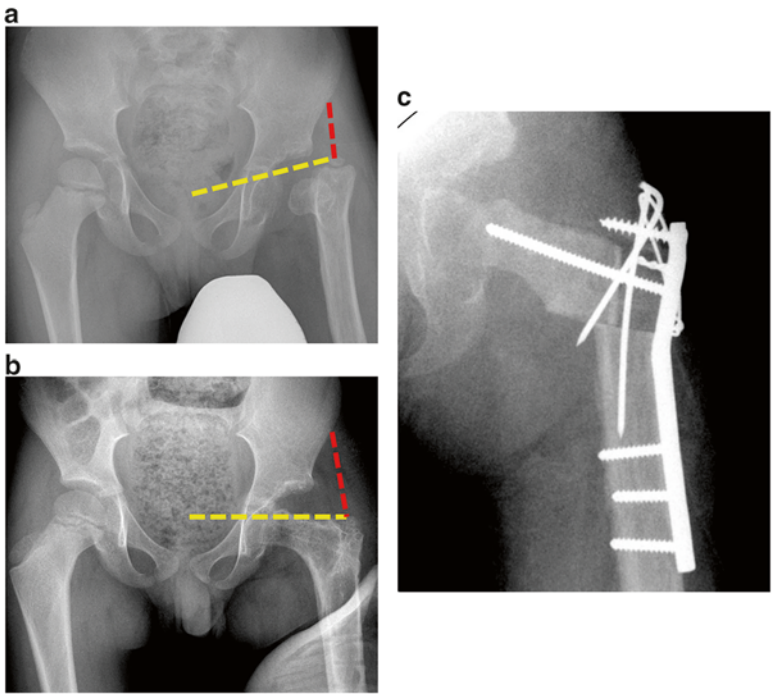


Fig. 6.17 (**a**) Radiograph of a 6-year-old boy with Choi's type IVa [44] post-septic sequel left hip. (**b**) A neck-lengthening osteotomy with distal transfer of the greater trochanter shows optimisation of the hip abductor lever arm function. *Red- and yellow-dotted lines* indicate abductor lever arm and moment arm, respectively. (**c**) Immediate post-operative radiograph

arrests around the knee (see Fig. 6.2). Alternatively it could be bifocal, where it is not optimal to do the angular correction at the same site as that of lengthening, for instance pelvic support osteotomy with distal femoral lengthening [47, 48]. Guided growth procedures can also be performed along with acute osteotomy, e.g. lateral proximal tibial guided growth along with ipsilateral Rab's osteotomy for a child with Blount's disease [49]. To counter the muscle imbalance brought about by cerebral palsy or meningomyelocele and other neuromuscular disorders, corrective osteotomy is combined with a muscle-balancing procedure, thereby avoiding relapse. For example, patellar tendon reefing is done following supracondylar extension osteotomy for fixed flexion deformity of the knee in cerebral palsy, and tibialis anterior transfer to the heel is combined with a calcaneal osteotomy in a child with excessive ankle dorsiflexion and weak plantarflexors.

Disease-Specific Indications

In certain conditions like osteogenesis imperfecta, when the child is non-ambulatory, the indication for a corrective osteotomy is to aid seating on a wheelchair, better hygiene and prevent further deformities rather than aligning mechanical axis. A long-standing dislocated or destroyed painful hip in a child can be managed by a pelvic support osteotomy combined with excision of the femoral head.

Box 6.2

- Apophyseal reorientation osteotomies (calcaneal, greater trochanter and tibial tuberosity) lengthen the lever arm for optimal joint function.
- Metabolic bone disorders usually cause multiapical deformities which often require acute multilevel corrective osteotomies.
- Acute shortening osteotomy may be performed primarily as in proximal femur to avoid avascular necrosis or in conjunction with deformity correction to avoid stretch on neurovascular structures.
- Various realignment osteotomies of the proximal femur are described to increase joint coverage, reduce impingement, realign capital physis and increase the arc of motion.

Planning

Rules Governing Osteotomy Planning

Growth considerations have to be kept in mind when planning deformity correction. Hence osteotomy planning in children is somewhat different from that of adults where accurate line diagrams and metaphyseal angles are drawn with a view to achieve precise alignment of the knee mechanical axis.

While restoring the mechanical axis is important in children, the deformity correction involves aligning the proximal and distal physes parallel to each other and perpendicular to the ground rather than merely achieving exact anatomical alignment. However in older children nearing skeletal maturity, the principles of restoring the mechanical axis used for adults are more applicable. While there are several methods for preoperative planning, we will illustrate our preferred planning methods for acute correction of the tibial and femoral deformities in children.

Planning for Tibial Deformity Correction

Step 1: Finding the CORA

The aim of surgical correction via an osteotomy is to achieve near-normal mechanical alignment and joint orientation of the lower extremity. After obtaining standing radiographs in the orthogonal planes, the joint lines at the knee and ankle are drawn (yellow-dotted lines) as shown in Fig. 6.18b. Red lines form the anticipated mechanical alignment angles of the tibia with the yellow-dotted lines tangential to the knee joint and talar dome. We consider the talar dome as the reference line for distal mechanical axis since the epiphysis and physis of the distal tibia are often misshapen in angular deformity in growing children. The mechanical axes intersect each other at the CORA and the angle [α] thus subtended is the magnitude of correction of the deformity (Fig. 6.18b). When the CORA is located in the bone near the obvious apex of deformity (area where the deformity visually appears to be maximum), correction can be obtained by a single osteotomy of magnitude "α" at this level.

Step 2: Finding the Axis of Correction for Two CORAs

In bowing deformities, the CORA could be located away from the obvious apex of deformity, suggesting the presence of two or more CORAs [23]. In such situations, two points

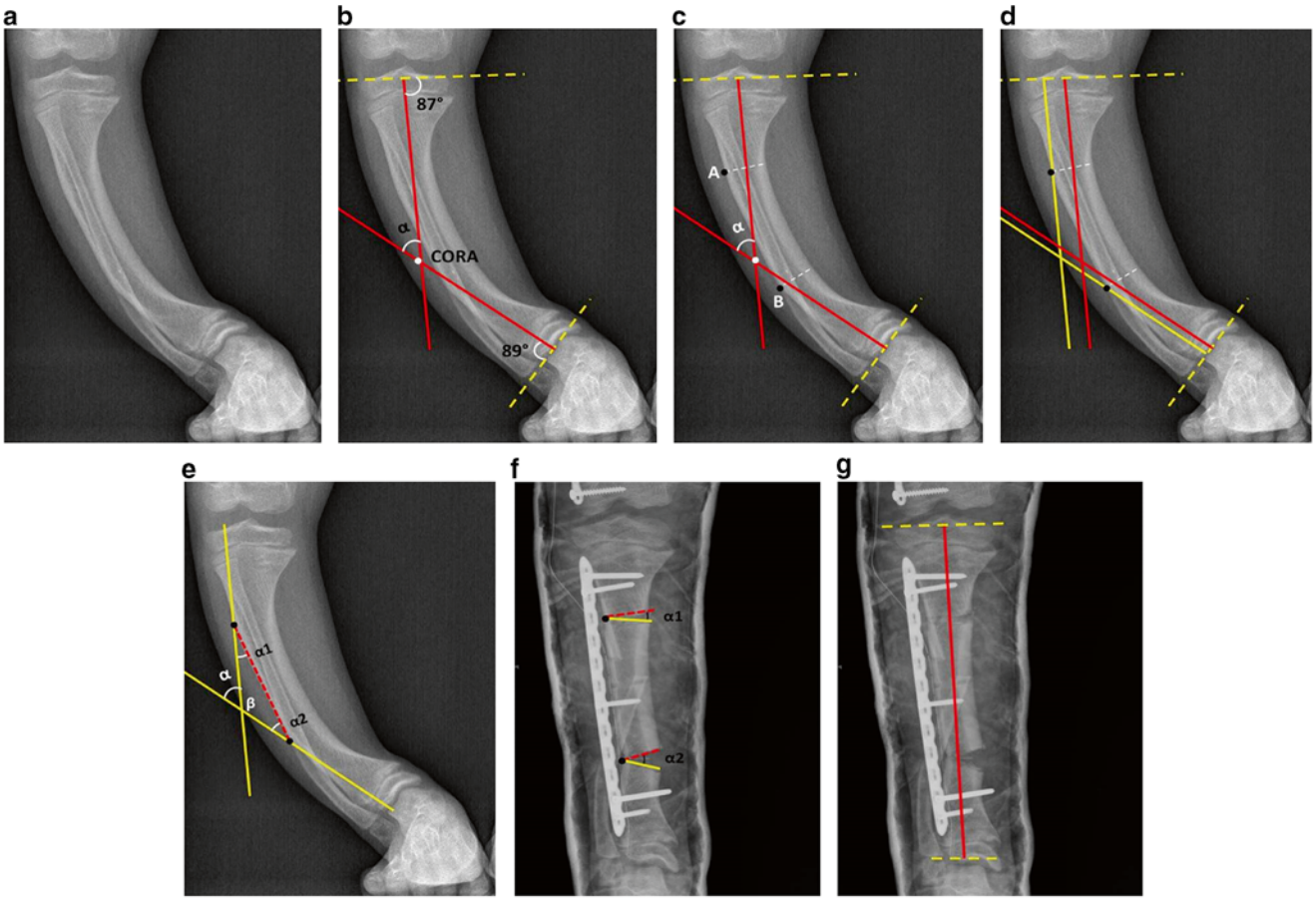


Fig. 6.18 Steps in planning correction of tibial deformity. (**a**) Bowing deformity of the tibia due to metabolic bone disease. (**b**) *Yellow-dotted lines* drawn tangential to the tibial plateau and talar dome. Mechanical axes [*red dotted*] of proximal and distal tibia are drawn from the centre of the former landmarks. The point of intersection of these lines is the CORA of the deformity and the angle "α" subtended is the magnitude of correction. (**c**) Two suitable sites for osteotomy "A" and "B" are plotted on the convex or concave tibial border based on execution of an open or closed wedge osteotomy, respectively. (**d**) Lines (*yellow solid*) are drawn parallel to the proximal and distal mechanical axes through these points. (**e**, **f**) The two points are connected by a line (*red dotted*), thus forming a triangle with α1, α2, and β angles. An opening wedge osteotomy of "α1" magnitude proximally and "α2" distally is planned. (**g**) Final radiographs after executing the osteotomies showing the corrected mechanical axis

(A and B) are plotted at the desired osteotomy sites on the cortex of the bone (Fig. 6.18c). These points form the centre of the arc of correction of the deformity. Lines (yellow) parallel to the mechanical axis of the individual fragments [red] are drawn through the proximal and distal points, respectively, as shown (Fig. 6.18d). These points are now connected by a single line (dotted red) (Fig. 6.18e, f). The sum of the two acute angles, α1 and α2, subtended in the triangle formed is equal to the total magnitude of correction "α".

Explanation:

The obtuse angle "β" in the triangle = 180° − [α1 + α2] (sum of all angles in a triangle = 180°)

Also, β = 180° − α (angles on a straight line add to 180°)

Hence, α = α1 + α2

The equation proves that the magnitude of multiple wedge osteotomies at the two desired points should sum up to the total magnitude of deformity correction.

The osteotomies can be thus planned at the marked points. It is prudent to confirm the mechanical axis alignment intraoperatively with the Bovie cord method [50]. The cord is stretched between the centre of the hip and talar dome following correction. The correction is then fixed using appropriate fixation device. If desired, the CORAs could be identified on the concave border and closing wedge osteotomies carried out.

Contrary to the popular practice, we do not draw the mid-diaphyseal line as this would produce two fixed apices at sites which may be unacceptable to undergo osteotomy. The advantage of the described technique is that the osteotomy sites can be chosen by the surgeon in consideration to adjacent structures like neurovascular bundle, physis, pathological bone or unhealthy skin.

Planning for Femoral Deformity Correction Other than Coxa Vara

The mechanical and anatomical axes for the femur are different. There are no paediatric standards for proximal lateral femoral angle and the greater trochanter may be incompletely ossified or unossified in young children. This requires an alternative planning for femoral deformity correction.

Step 1: Plotting Mechanical Axis for Distal Femur

Standing orthogonal films are obtained in both anteroposterior and lateral view. The mechanical lateral distal femoral angle (mLDFA = 88°) is drawn (see Chap. 2). This gives the distal mechanical axis [red line] as shown in Fig. 6.19b.

Step 2: Plotting Proximal Femoral Mechanical Axis

As there are no standards for mLPFA for different age groups in children we consider plotting the mechanical axis as follows. The mid-diaphyseal line is drawn as shown in Fig. 6.19c. A line parallel to this is drawn from the centre of capital femoral ossific nucleus [black dotted]. The proximal femoral mechanical axis is a line angled 7° lateral to this line (Fig. 6.19d). The point of intersection of these lines is the CORA and the angle subtended gives the magnitude of correction. The axis of correction should be located in the axial plane of the CORA depending upon whether an open, closed or neutral wedge osteotomy is desired. In case of bowing deformity with two or more CORAs **step 2** shown in Fig. 6.18e,f for tibial correction can be carried out. In case of coxa vara, the principle of deformity correction lies in aligning the capital femoral physis as horizontal as possible.

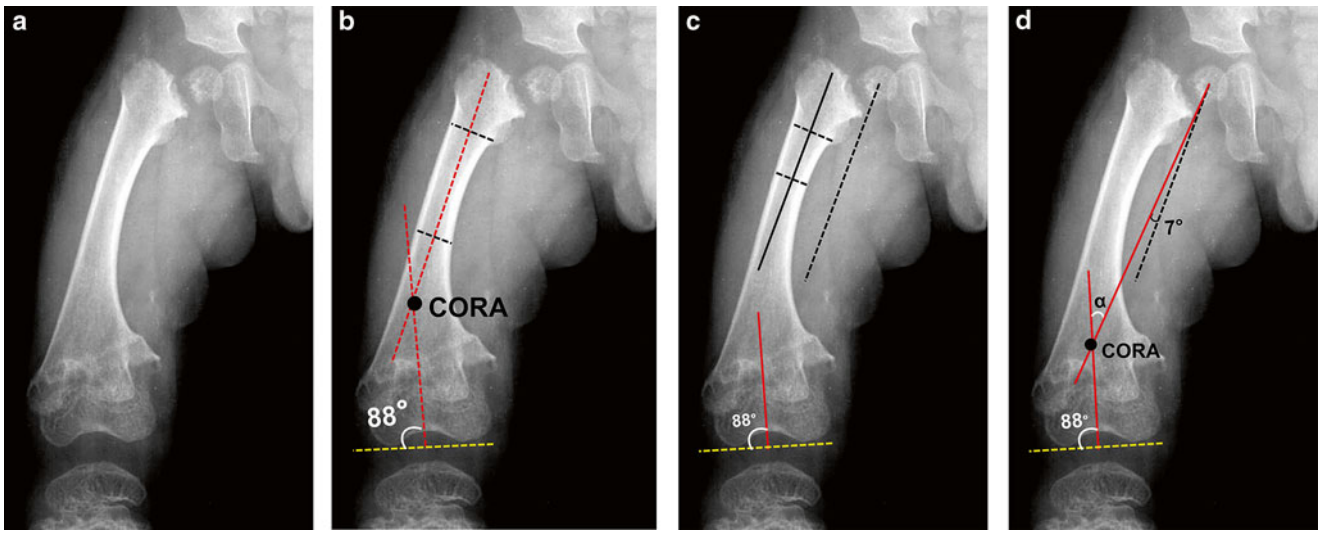


Fig. 6.19 (**a**) A femoral bowing deformity in a 4-year-old boy with unossified greater trochanteric apophysis. (**b**) mLDFA and mid-diaphyseal line for the proximal femur are plotted (*red solid line*, mechanical axis; *black solid line*, anatomical axis). (**c**) *Black-dotted line* is drawn parallel to the anatomical axis from the centre of capital femoral ossific nucleus. (**d**) The mechanical axis (*red solid line*) at the proximal femur is drawn from the same point at 7° to the *black-dotted line*. Intersection of the two axes is the CORA and "α" is the magnitude of deformity correction

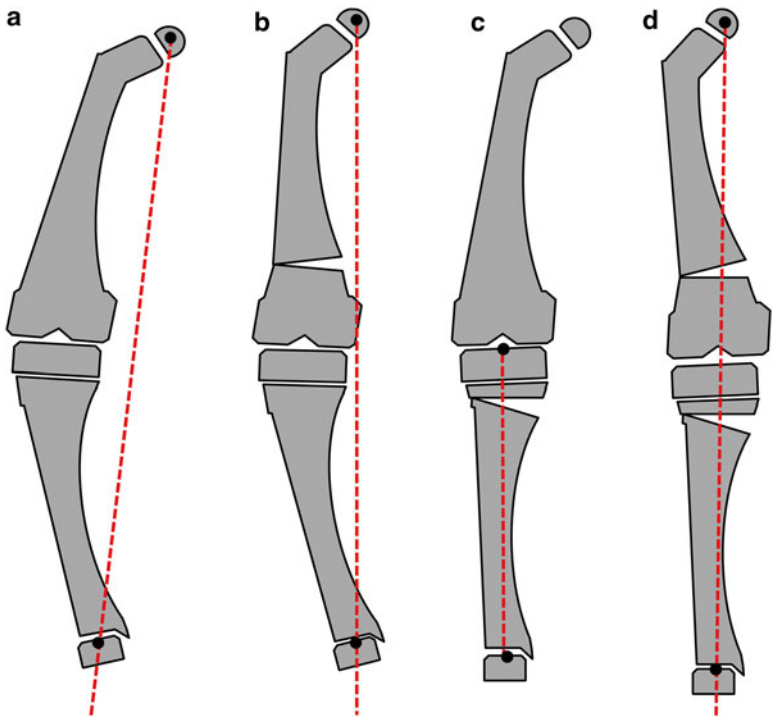


Fig. 6.20 (**a**) Illustration showing mechanical axis deviation (MAD) in a limb with tibial and femoral deformities. (**b**) Distal femoral corrective osteotomy without addressing the tibia deformity. Cross-checking MAD with Bovie cord between hip and talar dome as reference points is not possible as tibial axis is abnormal. (**c**) Cross-checking correction of tibial deformity is possible with Bovie cord using the centres of tibial plateau and talar dome as reference points. (**d**) The corrected tibial mechanical axis allows the mechanical axis of the limb to pass through the centre of the knee joint when the femoral head and the talar dome are the reference points

In all other cases we wait for the neck-shaft angle to remodel once the mechanical axis is restored. In adolescents where the trochanter is ossified, mLPFA and mLDFA are plotted as described in Chap. 2.

Planning for Multiple Bony Deformities in the Same Limb

Children with deformities secondary to metabolic disorders or osteogenesis imperfecta often present with multiple bony deformities in the same limb. When planning such sequential corrections, single or multi-staged, we perform the tibial correction first which has essentially the same mechanical axis and anatomical axis. For checking the femoral alignment, if the tibial alignment is normal, the Bovie cord is stretched between hip and ankle. Following correction of the femoral deformity, the cord should pass through the centre of knee if the mechanical axis of the lower extremity is restored. However, when the tibia is also deformed, in spite of restoring the femoral mechanical axis, the cord may lie on the medial or lateral side of the knee if the tibia has residual varus or valgus deformity, respectively (Fig. 6.20b). There is no such problem when checking the tibia as the Bovie cord can be stretched between the centre of the knee and ankle, and if it lies along the axis of tibia, the tibial deformity correction is confirmed independent of the hip reference point (Fig. 6.20c).

If the femoral deformity is corrected first, the cord has only one reference point, i.e. the centre of femoral head, the distal reference point being outside the corrected mechanical axis, i.e. the centre of talar dome. On the other hand, the tibial deformity can be corrected without using limb mechanical axis for reference as the anatomical and mechanical axes are the same in case of tibia. Hence when ipsilateral femoral and tibial deformities are being addressed with acute osteotomy correction, either at the same sitting or in a staged fashion, we recommend performing the tibial correction first (Fig. 6.21).

Box 6.3

- Judicious preoperative planning with standing radiographs or CT scanogram is essential before executing acute corrective osteotomy.
- Consider talar dome as a reference line for drawing the mechanical axis of distal tibia as the distal tibial epiphysis is often misshapen in deformities of the tibia.
- Intraoperative confirmation of mechanical axis alignment with a Bovie cord is prudent for accurate correction of deformities.
- When dealing with concomitant deformities of ipsilateral tibia and femur, it is wise to correct the tibial deformity first to avoid malalignment.

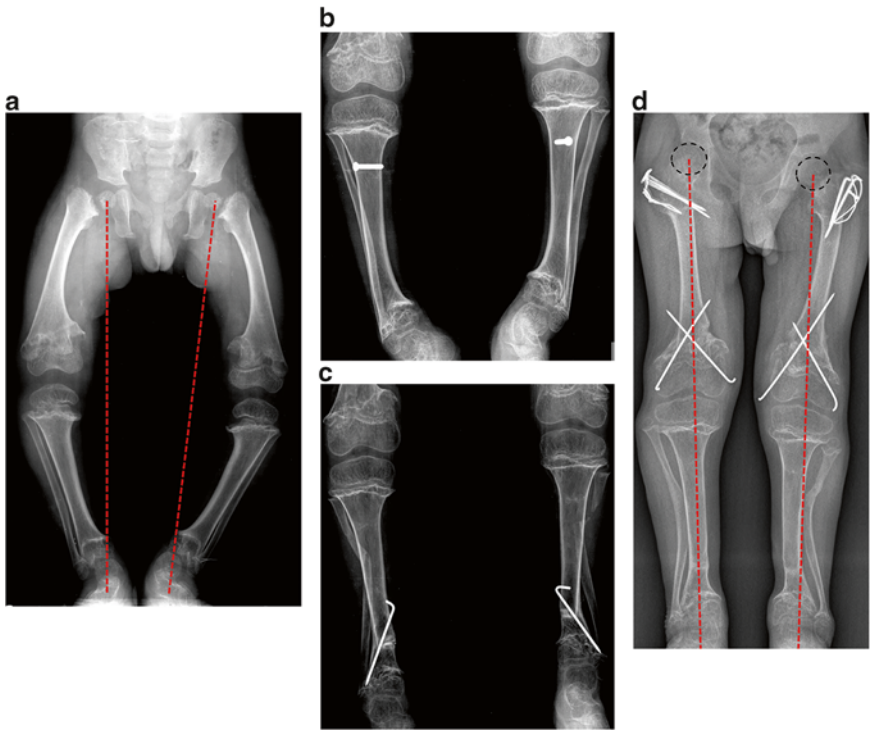


Fig. 6.21 (**a**) Preoperative standing radiograph of a child with spondylometaphyseal dysplasia. He underwent serial corrections of lower limb deformities. (**b**, **c**) The tibial deformity was addressed initially. (**d**) This was followed by correction of femoral deformity

Hardware Considerations

Casts are often used following acute deformity correction in young children. They are often used to maintain deformity correction after no fixation or minimal fixation with screws or wires [50]. The casts need to be well moulded to avoid loss of correction in cast. In the operating room, correction of the deformity in the cast is checked by the cautery cord method. A well-done cast wedging allows for fine-tuning of deformity correction (see Fig. 6.22). However, casts have to be used with caution in acute correction as post-operative swelling may lead to compartment syndrome.

Kirschner Wires

Smooth Kirschner (K) wires are safe and easy to use especially in infants and toddlers where plaster casts are often needed. The smooth wires are less traumatic to the physis and allow for fine-tuning of the correction in the post-operative period by wedging of the casts. They are also used when corrective osteotomies are combined with other procedures such as physeal surgeries in the vicinity and where the anticipated healing times are short and the forces across the osteotomy site are not excessive. Examples of K wire use are Bowen's technique for tibia vara correction, supramalleolar osteotomy, calcaneal lengthening osteotomy, Salter's innominate osteotomy and metatarsal osteotomies for hallux valgus correction [18, 51, 52].

Screws

Screws are useful for small fragments and it is possible to do post-operative adjustment in one plane only when a single screw is used, e.g. in Rab's osteotomy (see Fig. 6.1) [53]. They are also used for fixation in trochanteric (see Fig. 6.12) and tibial tuberosity displacements. They can cross the growth plate near or after physeal closure, as in osteotomy involving mal-united condyles and malleoli.

Dynamic Compression Plate

A contoured dynamic compression plate (DCP) in children can hold the angular correction required for small deformities. With such internal fixation devices a predetermined correction can be achieved by translating the bone fragments to the plate. Rotational correction can be achieved by inserting K wires on either side of the planned osteotomy at the desired angle and make them parallel at the time of fixation. This technique is easy for the beginners as the proximal fragment can be predrilled and tapped prior to osteotomy and is under control during osteotomy and fixation [54].

Tension Band Wiring

Even large fragments such as proximal femur fragments during coxa vara correction can be fixed in children with tension

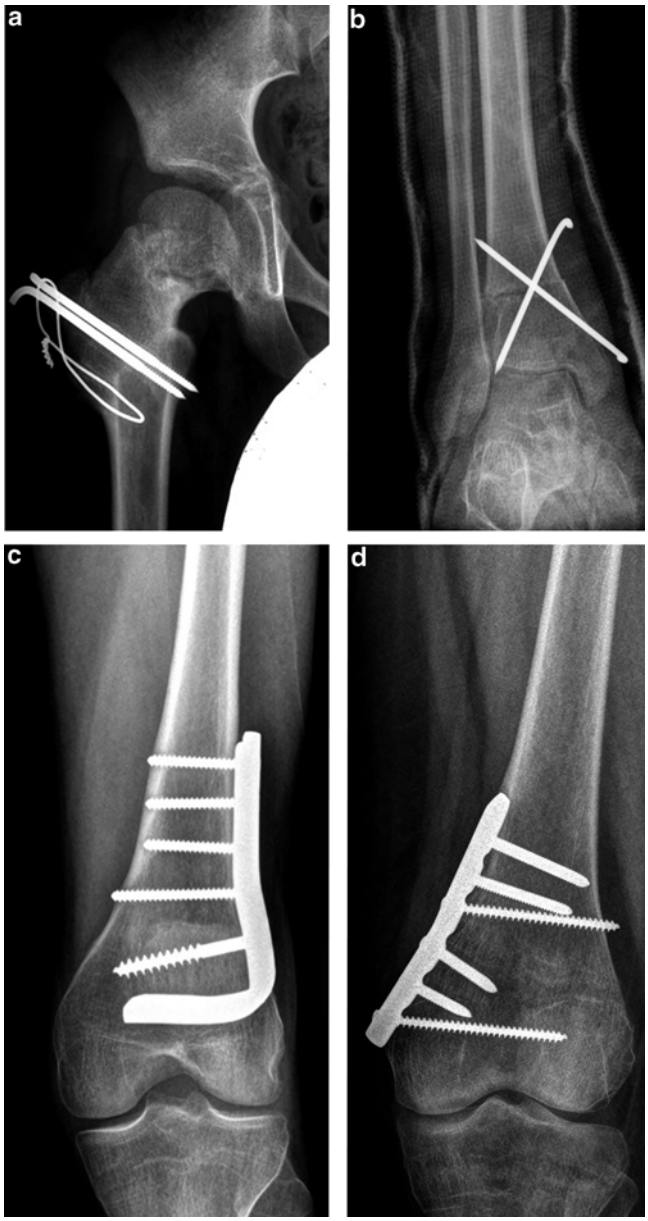


Fig. 6.22 Shows use of various implants at different sites. (**a**) Distal transfer of the greater trochanter fixed with tension band wiring. (**b**) Use of Kirschner wires and plaster cast for fixation of supramalleolar corrective osteotomy. (**c**) Lateral closed wedge osteotomy of distal femur and fixation with blade plate for genu varus deformity. (**d**) Use of metaphyseal plate for juxta-articular fixation following medial closed wedge osteotomy for correction of genu valgus deformity

band wiring (Fig. 6.22). It is a useful technique in closing wedge osteotomy. Another example is distal transfer of greater trochanter [55]. There is lesser degree of physeal damage when correction is desired in younger children wherein screws or plates cannot be used.

Blade Plate

Specific osteotomy plates such as condylar blade plates with preset angles can be used for proximal tibial as well as proximal and distal femoral metaphyseal corrections (see Fig. 6.22). Minor adjustment in angular correction is possible by varying the angle of insertions/entry points in condylar plates. Rotational correction can also be achieved by directing the blade in appropriate anteversion. However, these devices are less forgiving and require a thorough understanding of the local anatomy and the constraints of the implant design.

Locked Plates (LCP)

These are ideal for holding correction in osteoporotic bones and in multiple osteotomies within a single bone. They permit early weight bearing and promote improved bone healing due to reduced plate bone contact. Metaphyseal locked plate allows better hold in a short fragment near the joint (see Fig. 6.22). Paediatric varus and valgus hip LCP and distal femoral LCP for osteotomies around hip and lower femur allow multiple locked screw fixations, thus achieving not only rigid fixation but also avoiding early or late joint penetration as can be seen with some of the more conventional plating systems.

Intramedullary Devices (Such as Rush Rods and Fassier-Duval Telescoping Rods)

These devices are preferred when the bone quality is poor and cannot be corrected completely by medical management such as in osteogenesis imperfecta, metabolic disease and resistant forms of rickets and renal osteodystrophy. The rate of reported complications with telescoping rods has been high [56, 57]. Rush rods can also be used for corrective valgus osteotomy in the proximal femur in proximal femoral focal deficiency (PFFD). Overlapping rush rods can also be used to accommodate future growth in metabolic bone disorders and osteogenesis imperfecta (Fig. 6.23).

External Fixation and Limb Reconstruction Systems

Acute correction with external fixation offers potential advantages of allowing dynamic compression, distraction following acute correction, permitting minor realignments

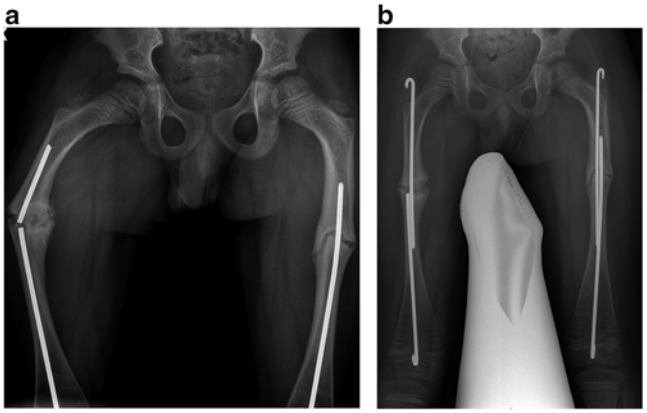


Fig. 6.23 (**a**) An 8-year-old boy with osteogenesis imperfecta presented a month after a trivial fall. Radiograph shows healing fractures of both femora and broken Rush rod in right femur. (**b**) Bilateral double Rush rodding was done to accommodate future growth of bone and fracture beyond the bone was protected by the implant

post-operatively and having a lower incidence of peroneal nerve palsy and compartment syndrome following a proximal tibial osteotomy in obese or adolescent patients [58]. The stability offered by circular external fixators often allows patients to weight bear as tolerated. The fixator can span the physis till healing of the osteotomy and then be removed, thereby not significantly affecting growth.

Bone Void Fillers

Autografts, allografts and varied combinations of commercially available synthetic bone substitutes can be used as fillers or scaffolds following an opening wedge osteotomy as promoters of bone healing. Such fillers may also provide some structural support in maintaining alignment following an acute correction. Ceramic-based bone graft substitutes include hydroxyapatite, calcium phosphate, tricalcium phosphate or bioglass. Hydroxyapatite-based scaffolds are the most preferred substitutes for bone reconstruction as natural bone has similar stoichiometry [59]. However, with the exception of autografts, these substitutes do not have an osteogenic potential. Synthetic bone graft substitutes are often brittle leading to loss of correction and cannot be used to fill large defects in long bones [60]. Resorption of hydroxyapatite-based ceramics is faster in young children [61]. In a recent study Balakumar et al. showed that hydroxyapatite tricalcium phosphate calcium silicate scaffold (HASi) incorporates faster and can be used safely for metaphyseal defects in children and adolescents [62] (Fig. 6.24).



Fig. 6.24 (**a**) Use of hydroxyapatite-tricalcium phosphate-calcium silicate (HASi) scaffold as a wedge to maintain correction in calcaneal lengthening osteotomy. (**b**) Non-union and resorption of neck of right femur following fracture in an 11-year-old girl treated by neck reconstruction by autograft (*arrow*) and valgus subtrochanteric osteotomy. Follow-up radiograph shows evidence of union

Adapted Fixation Techniques

Fixator-Assisted Plating

Maintaining accurate alignment can be difficult when using plate fixation following acute deformity correction. The fixator-assisted plating (FAP) technique, as described by Bar-On et al., uses a temporary external fixator to acutely correct the deformity and stabilise it before application of the plate [63, 64]. Fixator-assisted manipulation of osteotomy fragments using either a tubular, rail or ring fixator allows easy placement of LCP or DCP by minimally invasive technique, thus maintaining the biology and also decreasing the need for assistance during surgery. Indications for this technique include angular or rotational correction of metaphyseal or diaphyseal, multi-planar and multi-apical deformities.

Technique

The technique described by Bar-On et al. involves use of a monoplanar external fixator and locked plates [63]. Metaphyseal plates are used for juxta-articular deformities

and straight narrow plates for diaphyseal deformities. External fixator pins are introduced beyond the proximal and distal ends of the selected locked plate. When this is not possible, the half-pins can be closer together; however, their placement should avoid the anticipated site of plate fixation. The osteotomy is placed as close to the CORA as possible. Bar-On et al. recommend two locked screws per segment. Rozbruch described a similar technique in adults employing a designated deformity correction frame such as Taylor spatial frame or multiaxial correction frame [64]. The fixator placement must not impede subsequent internal fixation. The deformity is then incrementally corrected intraoperatively with this fixator.

We use a slight modification of the above techniques by placing the proximal and distal pins at right angles to the mechanical axis and fixed to the clamps of a monolateral fixator or a two-ring circular fixator. For tibial deformities, one or two half-pins are passed parallel to the proximal and distal joint lines, each under image intensifier in the plane of maximum deformity and preferably on the concave side of the deformity as distraction holds the alignment better. For the femur, the distal pin is inserted parallel to the joint line and proximal pin is inserted in the proximal shaft angled 7° caudad to a perpendicular (valgus with respect to the anatomical axis in the anterolateral plane) to the femoral shaft (Fig. 6.25a). When rotational correction is required, the pins are aligned at the desired magnitude of rotational alignment in each fragment. In a large epiphysis like that of the femur, the pins can be passed through the distal femoral epiphysis if necessary. A two-ring Ilizarov frame with two wires in each fragment may be used in corrections close to the ankle (Fig. 6.26).

A hinged monolateral or ring fixator permits fine-tuning of correction before plate insertion. Acute correction by osteotomy is carried out at or near the CORA and alignment is confirmed in both planes. In case of multilevel deformity correction or femoral corrective osteotomy, we use the Bovie cord technique to confirm final alignment. The locked plate is introduced by making longitudinal incisions proximally, distally or through the osteotomy site [63]. One end is fixed with a K wire to achieve plate alignment in close proximity to the shaft of the bone following correction and fixed with distal and proximal locking screws. At this stage, there is the opportunity for minor translation of the underlying bone fragment by adjusting the screw lengths or use of a non-locking cortical screw in the distal or proximal fragment of the plate to achieve desired displacement of the fragment. Prominence of plate may be avoided by pre-contouring the plate. Pre-contouring the plate with locked screws engaged in the plate is recommended to avoid distortion of the screw holes. Alternatively a contoured DCP can also be used. Post-operatively toe touch ambulation is usually permitted.

Fixator-assisted deformity correction offers fine control of alignment, eases correction and requires minimal assistance during application of internal fixation. The main advantage over conventional techniques of correction is that there is less risk of losing correction. We often use a similar technique when plating at the end of gradual lengthening with a fixator to shorten time in external fixation.

The technique, however, is difficult to use in smaller children. A difficult exposure or plate introduction due to presence of fixator can be avoided by judicious preoperative planning and outlining plate position with a skin marker [63]. Another problem is that a large pin tract may act as a stress riser. This could be prevented by extending the plate beyond the fixator or providing cast protection for a short period. The technique is also unsuitable for corrections of large magnitude involving bone shortening as actual contact between the two fragments is lost. We have had no pin tack infection thus far when doing locked plate following acute or gradual deformity correction using external fixation but there is a theoretical risk of deep infection.

Supracutaneous Locked Plating

Supracutaneous plate is an attractive option for acute deformity correction or shortening in the presence of poor skin condition or infection [65–67]. The indications are similar to those for using external fixation except that it allows fixation of small metaphyseal fragment when a metaphyseal locked plate is used. The additional advantages are its lightweight, low profile and easy concealment under regular clothing especially in a small child. We extensively use this technique for open fractures as well. Supracutaneous locked plate can serve as a lightweight external fixator in the presence of or prior to infection. The usual indications are deformity correction secondary to osteomyelitis sequel, physeal arrest or pathological fractures. The technique involves initial deformity correction at the non-union/mal-union site. In the presence of a gap non-union shortening is affected and the fragments are held with a conventional or fixator-assisted technique (Fig. 6.27). One centimetre thick folded linen is inserted between the plate and skin to aid screw length measurement. A supracutaneous locked plate holds this position with a minimum of four cortices (two locked screws) on either side. Following this the fixator is removed. We do not hesitate to put one screw in the epiphysis when the fragment is very small as the plate is removed in a matter of few weeks to months and the effect on growth is negligible. Pin care protocols are followed and removal is done when all four cortices at the osteotomy site are noted on radiographs.

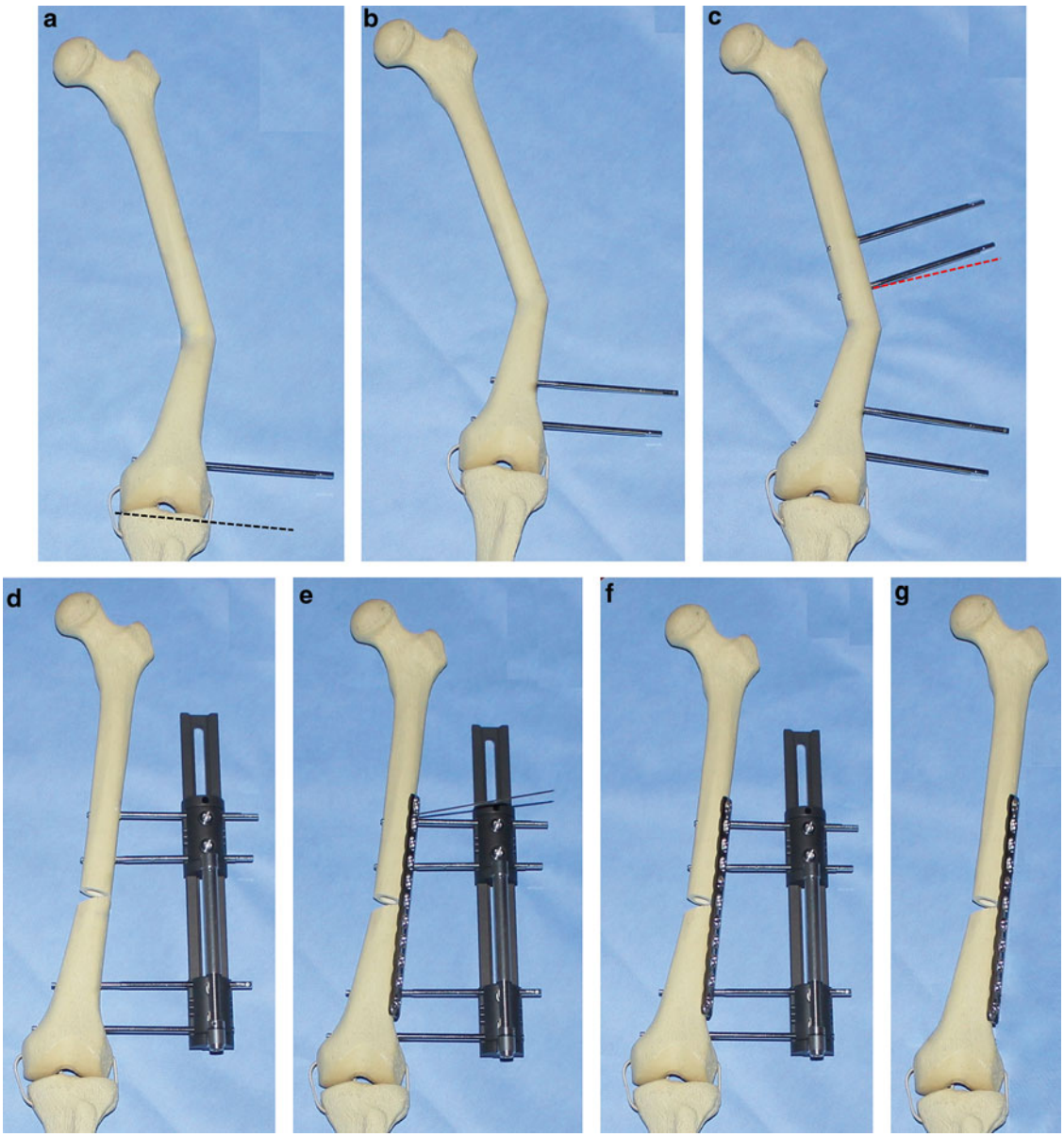


Fig. 6.25 The saw bone model illustrates the technique of deformity correction with limb reconstruction system (LRS) and subsequent plating. (**a**) Distal pin is placed freehand parallel to the knee joint line (*dotted line*). (**b**) Second pin is placed using the LRS pin template. (**c**) Second set of screws are angled 7° caudad to a perpendicular to the underlying diaphysis (*red-dotted line*). This restores the mechanical axis of femur which lies at 7° to the anatomical axis of the proximal fragment. (**d**) Osteotomy is carried out at or near the CORA and deformity corrected using the monolateral fixator. (**e**) A locked compression plate is aligned parallel to the shaft anterior to the fixator and temporarily stabilised with a K wire proximally and locked screw distally. (**f**) Remaining two screws proximally and one distally are inserted through the locking holes and K wire removed. (**g**) LRS removed

Box 6.4. Fixation Techniques and Implants

- Internal fixation with K wire and/or plaster cast allows minor post-operative realignment during moulding or by cast wedging.
- Metaphyseal locking plates offer better hold of short metaphyseal fragments following corrective osteotomy.
- Deformity correction in poor bone quality can be stabilised with intramedullary devices which permit growth and provide internal splinting.
- An external fixator following corrective osteotomy offers added advantage of compression, distraction and minor post-operative realignments.
- Bone void fillers provide structural support and act as promoter of bone healing as well.
- Fixator-assisted plating technique allows deformity correction and stabilisation of fragments until plate fixation is done.
- A locked compression plate can be used as a low-profile and lightweight external fixator following acute osteotomy.

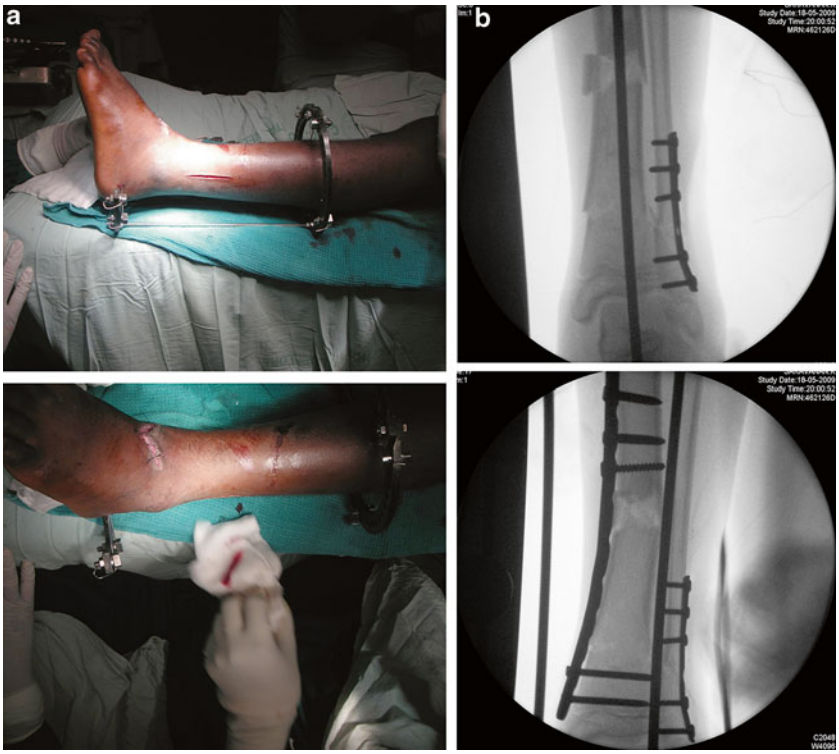


Fig. 6.26 (**a**) Photographs show use of Ilizarov frame in the fixator-assisted technique for reduction and maintaining alignment of fracture fragments in a 17-year-old boy with an open fracture tibia. (**b**) Intraoperative radiographs show attainment of alignment and the connecting Ilizarov rods, which are seen overlying the skin

Complications

The complications in acute deformity correction include complications of osteotomies, fixation failure and lengthening associated with acute deformity correction. The risk of complications may vary with age, the site of deformity, underlying pathology, proximity to physis and adjacent neurovascular structures. There is no consensus in literature on the amount of acute deformity correction and frequency of complications. Angular deformity correction has been noted to have more complications than rotational corrections [68].

Acute Complications

Neurological

The rate of neurological complications following acute deformity correction is varied. There is evidence that acute correction of a proximal tibial varus deformity is associated with a greater risk of peroneal nerve palsy than with gradual correction [69]. Some studies have shown no complications whereas others have reported a variable rate of such injuries ranging from 5 to 20 % [68]. The neurological deficit could be due to direct intraoperative injury, ischemia, acute stretch or fibular osteotomy [68]. Acute correction of a deformity via an osteotomy can potentially stretch the adjacent nerve(s) when the osteotomy and correction result in lengthening of the limb, especially if neurovascular bundle lies on the concave side of the deformity. This iatrogenic risk is less with a closing wedge than an opening wedge osteotomy. The degree of nerve stretch also depends on the mobility of the neurovascular bundle in the surrounding soft tissues and is related to the magnitude of angular correction and its distance from the centre of correction (Fig. 6.28). The neurologic risk could further increase when rotational correction is attempted; for instance, acute internal rotational correction in the proximal tibia can often lead to common peroneal nerve palsy. Another cause of peroneal nerve palsy is a fibular osteotomy, especially in the proximal third. Ideal position for fibular osteotomy is closer to the lower and middle third junction as the peroneal nerve is relatively less mobile in the upper third [15]. Judicious use of intraoperative SSEP monitoring and prophylactic nerve decompression in certain high-risk instances could minimise the risk of iatrogenic nerve injuries [68].

Compartment Syndrome

Concomitant acute lengthening with deformity correction, pressure on vessels or soft-tissue oedema following surgery can lead to a compartment syndrome. When performing a proximal tibial osteotomy with acute correction, it is advisable not to close the deep fascia after deformity correction. Prophylactic anterior compartment fasciotomy and insertion of a drain have also shown to decrease the chance of

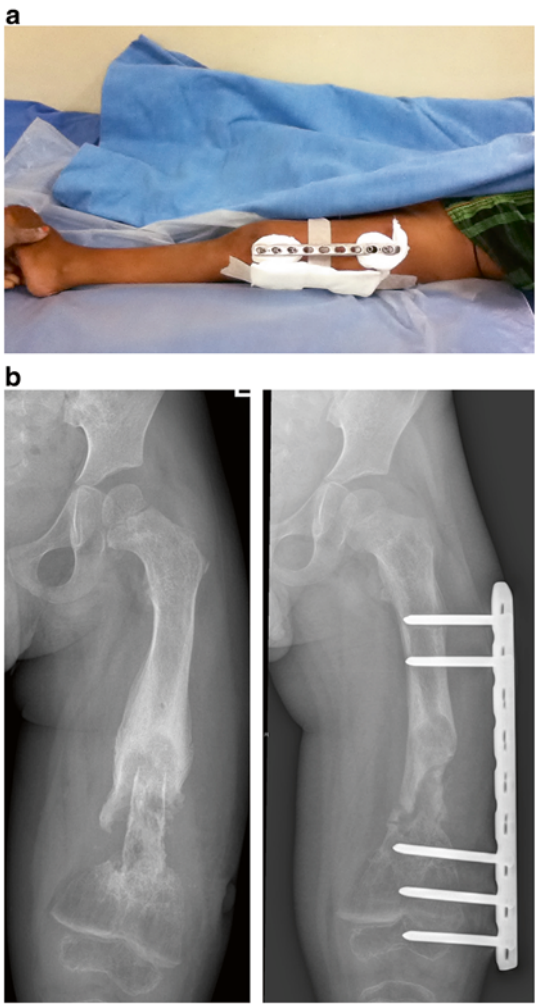


Fig. 6.27 (**a**) Picture of a 6-year-old boy with chronic osteomyelitis left femur with a supracutaneous locked plate fixation. (**b**) Pre- and post-operative radiographs show corrective angulation and shortening osteotomy following sequestrectomy

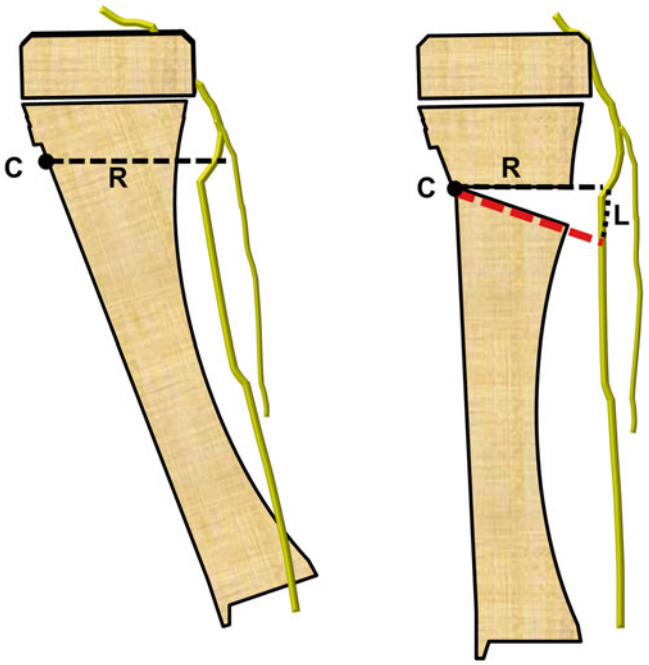


Fig. 6.28 Illustration of valgus deformity of tibia. Effect of nerve stretch brought about by an opening wedge osteotomy is shown. The amount of stretch effected "L" on the nerve is directly proportional to the magnitude of angular correction "θ" and this can be calculated by the formula $L = 2\pi R \frac{\theta}{360^\circ}$ where "R" is the distance of the relatively immobile nerve segment at the level of osteotomy from "C", the centre of rotational correction. With kind permission from Springer Science + Business Media: Paley D. Principles of deformity correction. Berlin, Heidelberg; 2002

compartment syndrome in acute correction for Blount's disease [70]. Opening wedge osteotomy, large displacements and multiple osteotomies in the same or different segments of the same limb predispose to the development of a post-operative compartment syndrome.

Vascular

Vascular injury may occur secondary to direct injury, instrumentation or deformity correction. Such iatrogenic injuries may be due to drill/osteotome/oscillating saw/inappropriate retractor placement or implant placements such as screws, external fixator pins or excessive stretch following open wedge osteotomy. The popliteal artery is at specific risk for osteotomies around the knee joint, e.g. Rab's osteotomy. The peroneal vessels are at risk when doing an adjuvant fibular osteotomy at its middle third [16].

Skin and Wound Problems

When the CORA is away from the site of correction large displacements of the bone occur and these could result in risk to the skin and soft tissue and wound dehiscence. Other causes for skin necrosis are poor skin conditions prior to surgery, subcutaneous bones such as tibia and hardware prominence especially in subcutaneous areas such as femoral condyles.

Iatrogenic Fractures

Intraoperative fractures can occur, as in osteogenesis imperfecta, or a longitudinal split, or while trying to stabilise fragments in the presence of excessive soft-tissue tension.

Physeal Injury

Physeal damage can occur when correcting close to the joint or due to multiple penetrations by the fixation devices such as nails or wide Kirschner wires. Use of intraoperative image intensifier is hence advised when dealing with juxta-physeal deformity corrections.

Late Complications

Joint Stiffness

Soft-tissue adhesions following acute osteotomy correction are often the cause of joint stiffness more so around the knee and the ankle. Tethering of the ilio-tibial band and the quadriceps may also occur when external fixators are applied to the thigh. Excessive intra-articular pressure following a juxta-articular acute opening wedge osteotomy correction could lead to sequel such as chondrolysis stiffness along with chondrolysis and avascular necrosis can be associated with distal transfer of the greater trochanter.

Vascular

There are reports of delayed presentation of pseudoaneurysms caused by vascular injury related to instruments such as an osteotome, or the penetration of vessels by fixator pins, screws or wires [71]. Once recognised the treatment options include ligating the involved vessel if insignificant for the vascularity of the limb, embolisation and vascular repair.

Recurrent Deformity

Recurrence of the deformity following an osteotomy is likely in the presence of metabolic bone disease, especially if the underlying metabolic disorder is not corrected or controlled. Other causes are residual soft-tissue contractures requiring soft-tissue lengthening or muscle balancing such as in neuromuscular foot deformity (Fig. 6.29). In coxa vara, the presence of an open physis and failure to appropriately realign the capital physis increase the risk of recurrence. In some of the congenital limb deficiency and posteromedial bowing there is a deformity memory and this causes a recurrence by bowing of an immature callus. Lack of planning for further disturbed growth in the affected physis, poor bone quality and ongoing disease process such as cystic lesions and fibrous dysplasia are other causes. Recurrence may also be related to the severity of soft-tissue deformity or magnitude of initial deformity, poor choice of implant or premature weight bearing. Correction or optimising the underlying metabolic disorder prior to surgical correction is essential, and supplemental external or internal splinting following osteotomy correction is required in those with impaired metabolic state [50]. However, the level of osteotomy, whether metaphyseal or diaphyseal, may not be related to the risk of recurrence [50, 72]. In children with cerebral palsy, rotational deformities tend to recur if de-rotation is performed in children less than 10 years of age [2].

Non-union and Delayed Union

Non-union and delayed union are rare in children but could occur in the presence of excessive motion at the osteotomy site, decreased bone contact and bone gap associated with large angular corrections and thermal necrosis due to use of power instruments such as an oscillating saw. This is also more likely when combining acute correction with gradual lengthening. Some of this can be avoided by adding translation and collapse which increases stability and bone contact and decreases length gained at the osteotomy site. Possibility of poor healing is high when doing acute corrections of greater than 30° with associated lengthening [73].

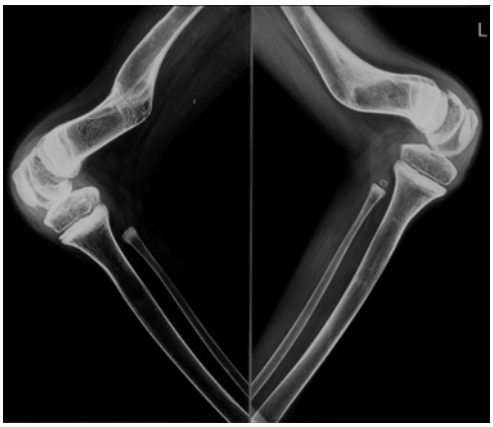


Fig. 6.29 Radiograph of 7-year-old boy referred for recurrence of flexion deformity of both knees 2 years following corrective femoral osteotomy for the same

Hardware-Related Complications

In children, the peri-implant complications are high because of the greater difference in the stiffness of the implant and bone. As the underlying bone is smaller and the overlying soft-tissue envelope less bulky, implant prominence is greater. Intramedullary implants being less elastic, if not perfectly aligned with the longitudinal mechanical axis, tends to cut out or bone bends at the implant bone junction. The suggested periods for implant removal will vary from 9 months to one and a half years, based on the underlying cause. In some conditions such as osteogenesis imperfecta, these implants are not removed, but revised with further growth.

Even after the healing of osteotomy in deformity correction in osteoporotic bone, fractures or bowing can occur at the implant bone interphase and cause the hardware to fail. Wrong choice of hardware, especially in those with osteopenia, is another cause of implant failure or recurrence of deformity (Fig. 6.30).

Cast-Related Complications

Patients with anaesthetic limb, impaired cognition and spasticity have a predisposition to develop skin and soft-tissue sores related to overlying casts. To minimise the risk of such cast-related problems, bony prominences should be

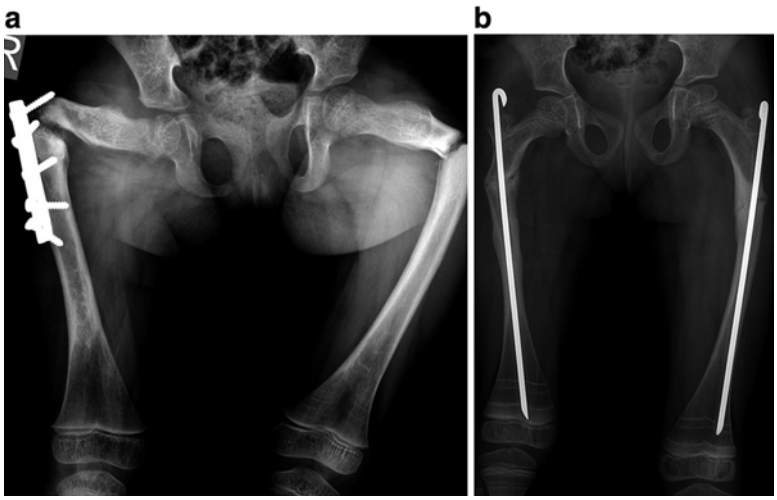


Fig. 6.30 (**a**) Radiograph of a 10-year-old girl with osteogenesis imperfecta (OI) shows fixation failure due to poor choice of implant and soft bones. (**b**) The ideal implant for OI would be an intramedullary device

adequately padded and attempts at gaining further correction by wedging the cast must be avoided, especially in these at-risk children. If cast wedging is done, it should include removal of cast wedge on the collapsing side to avoid excessive pressure on the underlying skin and soft tissues. Tight plaster casts in the acute post-operative phase should be avoided to avoid compartment syndrome.

Muscle Weakness

When performing a closing wedge osteotomy and substantial shortening, there is a relative lengthening of the overlying muscle-tendon unit. For instance, a closing wedge supracondylar extension osteotomy can be associated with quadriceps insufficiency and weak knee extension. In some cases where the weakness persists despite physical therapy, one may require surgical measures such as patellar tendon reefing.

Box 6.5. Staying Out of Trouble When Doing Acute Deformity Correction

- Leave the deep fascia open following acute deformity correction to minimise the risk of compartment syndrome.
- Judicious use of intraoperative SSEP monitoring and nerve decompression in high-risk cases is recommended to avoid iatrogenic nerve palsy.
- Physeal injury can be avoided by use of intraoperative imaging and avoiding multiple penetrations of the physes by nails and wires for juxtaphyseal osteotomies.
- The bony pathology and associated soft-tissue contracture or muscle imbalance should be addressed during deformity correction in order to minimise recurrence.

Acute Versus Gradual Correction

The complication rates with acute correction are generally higher than those of gradual correction in the tibia [58, 69]. Gradual correction of tibia vara has shown to correct angular deformity more accurately than acute correction [58, 69]. The possible cause could be the flexibility to correct minimal malalignments with the gradual correction techniques, especially with use of computer-assisted technology. However, the acute correction requires less supervision. Before choosing acute or gradual correction for managing a child with a lower limb deformity, the treating surgeon should consider factors such as the patient's growth remaining, magnitude of current and anticipated deformity and limb length discrepancy, availability of resources, familiarity with instrumentation and psychosocial issues.

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Gradual Deformity Correction

7

David S. Feldman, Adam M. Kurland,
and Abdel Majid Sheikh Taha

Introduction

Deformity correction for traumatic, congenital and developmental conditions in the pediatric population may be quite complex and the deformities can be quite severe. In children, the deformities may have their apex (or apices) at the level of an adjacent joint or growth plate which then requires an osteotomy away from the site of the deformity and requires translation at the osteotomy site to restore a normal mechanical axis of the affected limb segment. Additionally, deformities may be multi-apical and are almost always in the oblique plane. The patient may also have a limb with compromised overlying soft tissues.

The severity of the deformity and these other confounding factors often make it advantageous to perform a correction gradually. The ability to correct a deformity gradually gives the surrounding soft tissues sufficient time to accommodate. Gradual correction may also allow for a more accurate correction and a diminished risk of compartment syndrome, nerve injury, and non-unions [1, 2].

Gradual correction provides the opportunity to make changes after the procedure, in an office setting, resulting in a more accurate overall correction with less morbidity than an acute correction. This chapter reviews the history, biology, clinical indications and considerations, specific uses, complications, and how to avoid complications during the gradual correction of the lower limb.

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Box 7.1

- Gradual correction is often safer than acute correction
- Gradual correction allows for adjustments postoperatively

History

The modern day history of gradual correction takes it origins from the work of Codivilla in 1905 [3] and based on our research was first utilized clinically in 1934 at the Hospital for Joint Diseases in New York. De Bastiani and Ilizarov expanded the use of this work and explained and elucidated the biology of distraction osteogenesis. While this method is utilized for limb lengthening, a gradual correction to correct a limb deformity employs the same principles. Current technology continues to utilize these concepts with both, computer-assisted management with external fixation and more recently with intramedullary devices. Distraction osteogenesis is the biology required for gradual correction through an osteotomy site.

Box 7.2

- There are many devices that allow for gradual correction, internal and external.
- Historically external fixation has been utilized for gradual correction.

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Biology

In distraction osteogenesis, whether utilizing internal devices or more traditionally external fixation devices, the intact periosteum is likely responsible for bone formation [4]. Following an osteotomy, the patient or caretaker is typically instructed to begin distracting the bone after a period of 5–10 days to allow the intraosseous blood supply to recover. After this latency period with an ideal rate [5–7] (usually 1 mm/day, 1.5 mm/day with an intramedullary rod in some cases) and an ideal rhythm (traditionally 4×/day but continuous "auto distraction" is likely even better), new bone will form in the distraction gap. While leaving the intramedullary blood supply undisturbed at the time of the osteotomy was thought to be crucial for this regeneration, this has been questioned because the blood supply likely regenerates 1 week after cutting the bone. It is known that the intact periosteum is the most important structure in distraction osteogenesis. The gradual tension generated with distraction allows for filling in of the bone as it is angled and/or lengthened.

Understanding how the bone is formed during the distraction has been debated in the literature and it is likely that both intramembranous and enchondral ossification occurs within the regenerate bone. Distraction osteogenesis is similar to fracture healing, in which an initial hematoma transforms into fibrocartilaginous tissue and fibrous tissue. Within 2–3 weeks this tissue matures to a striated appearance of the regenerate [6, 8–10].

Many modalities have been described to enhance bone formation including electrical stimulation and low-intensity pulsed ultrasound [11–13]. Combining these modalities with the gradual allowance of weight bearing may accelerate the process of remodeling of the regenerate and add to its strength by increasing its bone density (refer to Chap. 28 for more details).

Box 7.3
- Distraction osteogenesis is the biologic principle utilized in gradual correction.
- The regenerate can be enhanced with different types of modalities.

Clinical Uses

Almost any degree of deformity can be corrected with gradual correction (Fig. 7.1). Gradual correction, as the name implies, allows for the slow and precise correction of deformity. The deformity may be angular, rotational, translational, or any combination of these (Figs. 7.1, 7.2, 7.3, and 7.4) [14]. Limb length inequality can also be addressed using this method (Fig. 7.5). This technique is typically associated with less disruption of the soft tissue envelope surrounding the deformity than with acute correction [1]. The soft tissue envelope includes many vital structures, such as blood vessels,

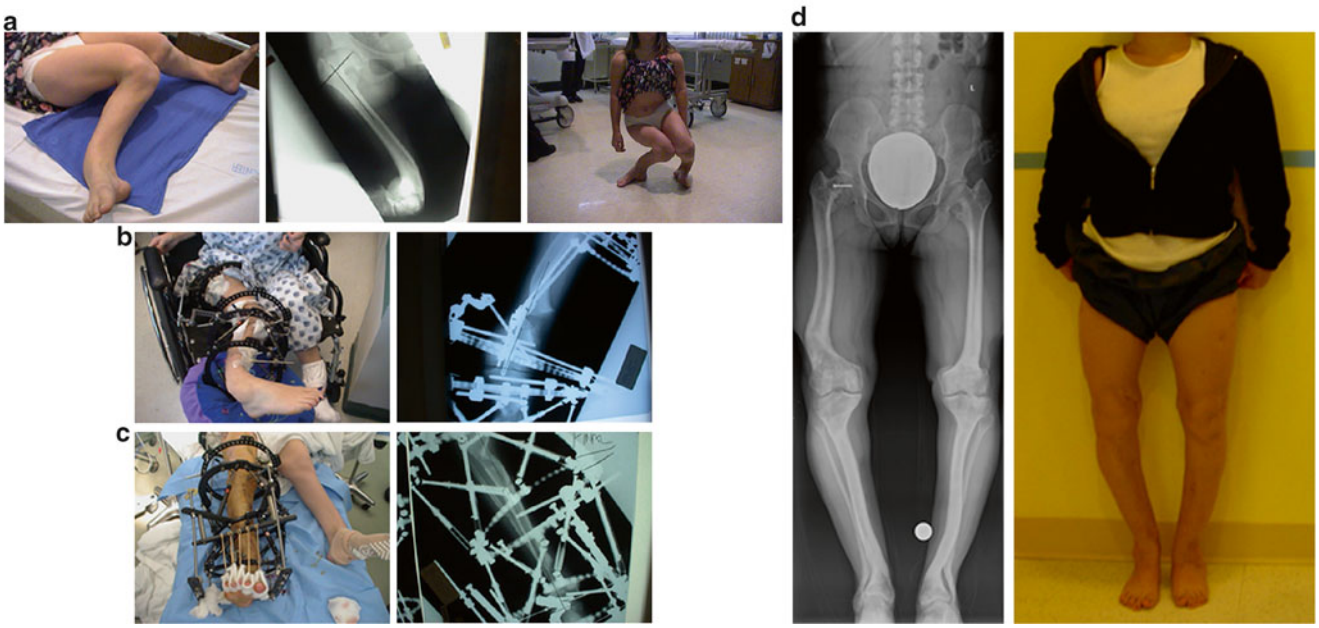


Fig. 7.1 Severe deformity. A patient with pseudopseudohypoparathyroidism (pseudoPHP) with complex, multiplanar and multiapical deformities of both lower extremities using the Ilizarov technique and the Taylor Spatial Frame™. This case demonstrates the possibility through gradual correction to correct very severe deformity. (**a**) Preoperative clinical and radiographic view. (**b**) During correction. (**c**) After one side is completed. (**d**) Final result

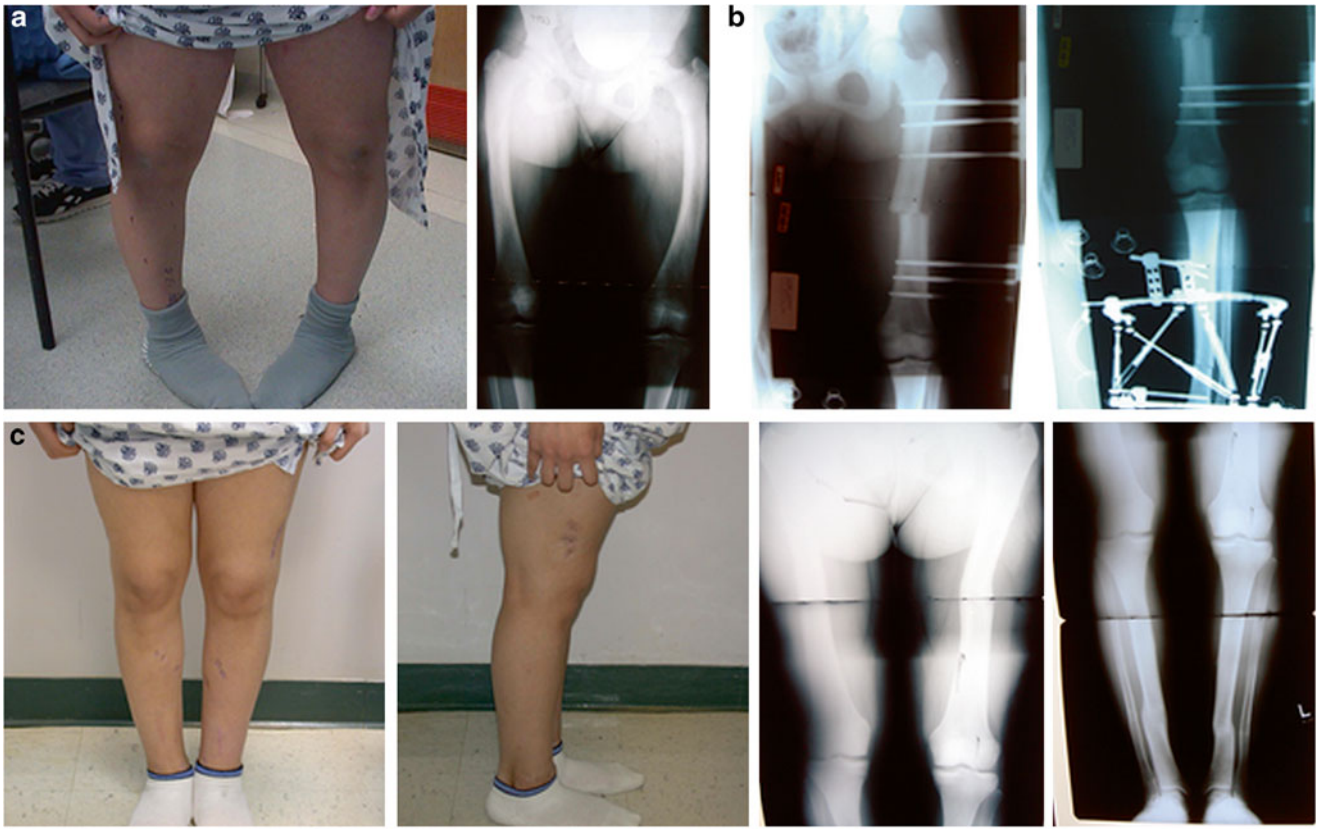
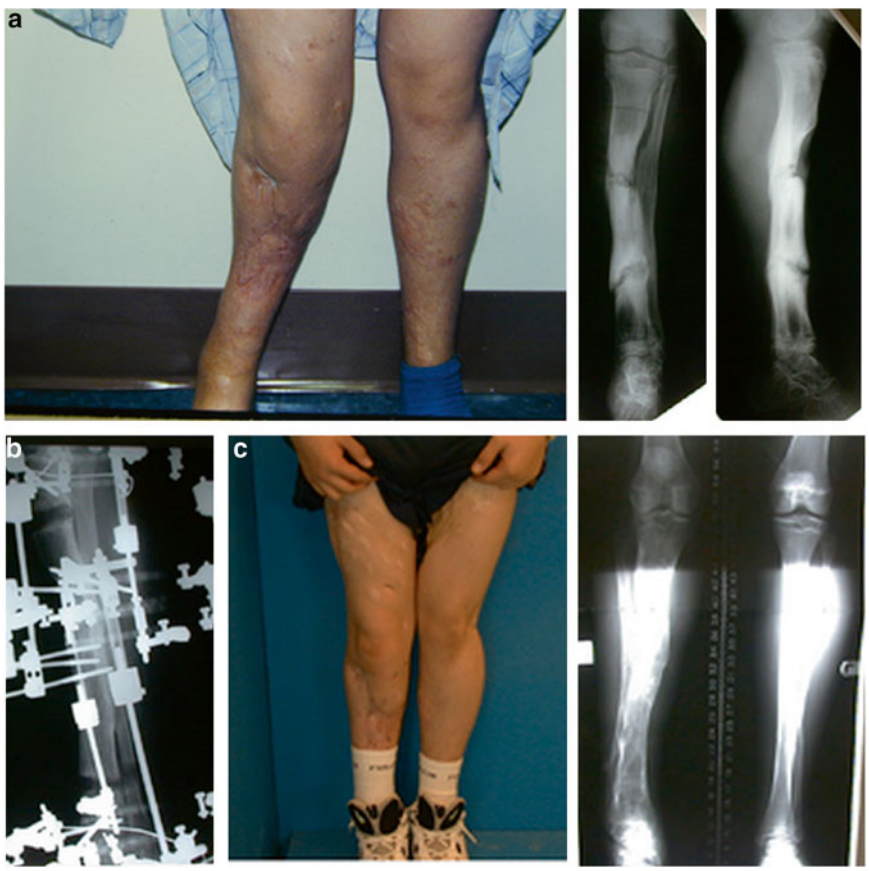


Fig. 7.2 In the case of hypophosphatemic rickets with a multiapical deformity and rotational deformities of the tibia and fibula, a femoral deformity resolved to one location and was corrected acutely while the tibial and fibular rotational deformities were gradually corrected with the use of a circular external fixation device. (**a**) Preoperative clinical and radiographic view. (**b**) During correction. (**c**) Final result

Fig. 7.3 Translation and angular deformity—This deformity of infected non-unions demonstrates a case in which the translational deformity cannot be corrected at the site of deformity due to poor soft tissue and a single vessel leg. This translational deformity is corrected by two angular osteotomies away from the site of the deformity. (**a**) Preoperative clinical and radiographic view. (**b**) During correction. (**c**) Final result



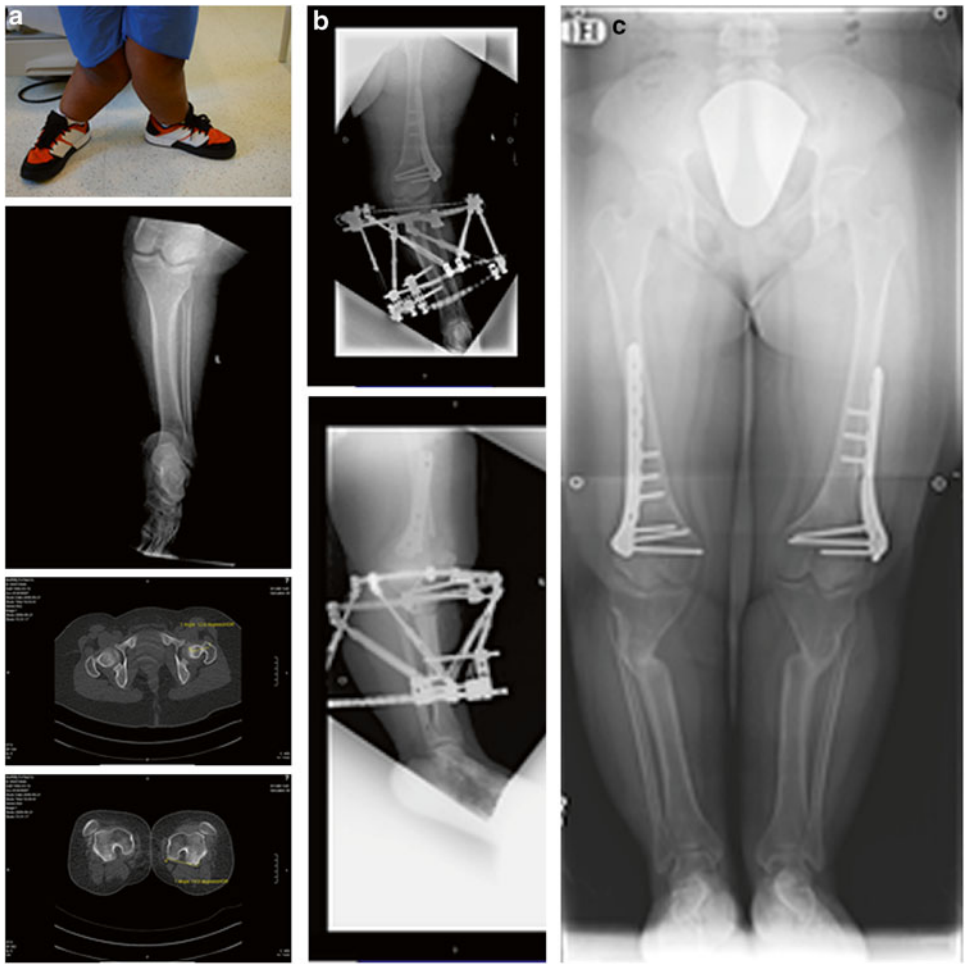


Fig. 7.4 In a patient with Ellis–van Creveld disease with femoral retroversion and tibial angular deformity, deformity correction was performed via an extra-articular osteotomy of the femur with derotation and gradual correction of the tibial angular deformity with external fixation. Note the initial patellar dislocation was reduced once the rotational deformity was corrected. (**a**) Preoperative clinical and radiographic view. (**b**) During correction. (**c**) Final result

nerves, muscles, and ligaments, which have different physical properties from bone and thus may not tolerate the forces of acute correction, especially in the setting of a large deformity. For instance in the case of a severe valgus deformity around the knee joint, performing an acute correction can stretch the common peroneal nerve, possibly resulting in a postoperative nerve palsy and subsequent foot drop [14]. Gradual correction allows for monitoring of the nerve function or dysfunction and a release of the nerve may be performed during the correction process, should symptoms develop.

Blount disease, a condition ideally suited for gradual correction, is an oblique plane (anterolateral) deformity of the proximal tibia that presents in young children and adolescents (see Fig. 7.6). The deformity is often at the level of or close to the proximal tibial physis and may be associated with internal tibial torsion [15, 16]. Historically, the operative correction of Blount disease was associated with malunions and delayed unions as well as compartment syndrome and nerve injury (most commonly the nerve to the extensor hallucis longus). Many of these complications can be minimized with gradual correction [17, 22]. Compartment syndrome occurs either due to swelling in the compartment obstructing venous return or compression or injury to the anterior tibial artery as it transverses the interosseous membrane. Both of these etiologies, i.e., swelling and injury to the artery, are much less common with gradual correction. Rarely are prophylactic fasciotomies required in gradual correction.

Box 7.4

- There are many clinical uses for gradual correction.
- Protection of nerve function and the soft tissue envelop are key components to gradual correction.

Analysis of Deformity

Planning for deformity correction follows the same principles whether performing a gradual or an acute correction. One must analyze the axes in the coronal, transverse and in the sagittal planes [18, 19], identify the apex of each deformity, plan the level of the osteotomy (or osteotomies), and decide



Fig. 7.5 Deformity correction with limb length inequality. This case demonstrates tibial varus and femoral rotation concurrent with length inequality of both bones. An acute derotational osteotomy of the femur and bifocal osteotomy of the tibia were performed. Deformity correction and lengthening of the tibia were achieved using a hexapod external fixation device. (**a**) Preoperative radiographic view. (**b**) Clinical and radiographic view during correction. (**c**) Final result

on the surgical technique and hardware to be used. In addition to analyzing the axes and determining the degree and location of the deformity, the patient's age and remaining growth are also important factors. Patients that are still growing can typically benefit from growth modulation procedures particularly when the deformity is close to the joint (refer to Chap. 4 for more details) [20]. To ensure accurate correction of a deformity of the lower limb, both the individual features of the bone being osteotomized and the limb as a whole must be understood. The overall alignment of the extremity, the alignment of the tibia and femur independently, the joint alignment (laxity), and rotational deformity require evaluation (refer to Chaps. 2 and 3 for more details).

Of the many factors that affect bone healing, the quality of the underlying bone is perhaps the most important [21] and should be the first step in assessing the deformity. The potential to heal dictates the success of the procedure, the extent of fixation needed, and the possibility of hardware failure. The ability of a bone to heal differs substantially between various conditions (Figs. 7.7 and 7.8). For instance, in multiple enchondromatosis, the bone has a higher neoplastic potential when compared to normal bone and may heal at a slower rate (see Fig. 7.7) [22]. Osteogenesis imperfecta is an example in which the bone density and architecture is weak [23], making it more difficult to maintain alignment without supplemental measures such as preoperative bisphosphonates and the use of an intramedullary device instead of an external fixator.

The deformity must be quantified and should be analyzed on the anteroposterior (AP) and lateral radiographs. Varus or valgus angulation and the medial or lateral translation are measured on the AP view. Procurvatum or recurvatum and anterior or posterior translation are measured on the lateral radiographs. Rotational alignment is measured clinically using the thigh-foot axis with the patient in prone and if necessary may be supplemented with a CT scan or MRI. Length is measured using a standing full-length lower extremity radiograph. To quantify the deformity, mechanical axis deviation (MAD) and joint laxity, a standing patella forward full-length AP and lateral radiograph of both lower limbs are used [2]. Femoral and tibial deformities are further quantified on patella forward AP and lateral views of the



Fig. 7.6 Angular deformity. This is a case of untreated Blount disease in an adult. To elevate and restore the joint line, an intra-articular osteotomy was performed. The deformity was then gradually corrected using a hexapod external fixation device. (**a**) Preoperative clinical and radiographic view. (**b**) During correction. (**c**) Final result

individual long bones. In the case of the very young child with unossified epiphyses, an intraoperative knee arthrogram may be performed to outline the femoral condyles and tibial plateau to more accurately assess the deformity present in the tibia and/or femur (refer to Chap. 2 for more details).

Box 7.5

- Thorough analysis of the deformity including an assessment of the quality of the bone is essential prior to gradual correction.
- A thorough clinical exam and standardization of imaging studies allows for accurate deformity analysis.

Planning the Osteotomy

Determining the level of the osteotomy is an integral part of the presurgical plan. Ideally the osteotomy should be located at the apex of the deformity. At times, the osteotomy cannot be performed at the apex of the deformity due to the proximity of the joint, the physis, or compromised surrounding soft tissues. These issues necessitate moving the osteotomy away from the apex. For example in Blount disease, the apex is usually in proximity to the knee joint or the proximal tibial physis, making it impractical both to osteotomize in these areas and to safely obtain satisfactory fixation at the osteotomy site. Additionally, one must avoid performing the osteotomy proximal to the tibial tubercle since this may lead to patella

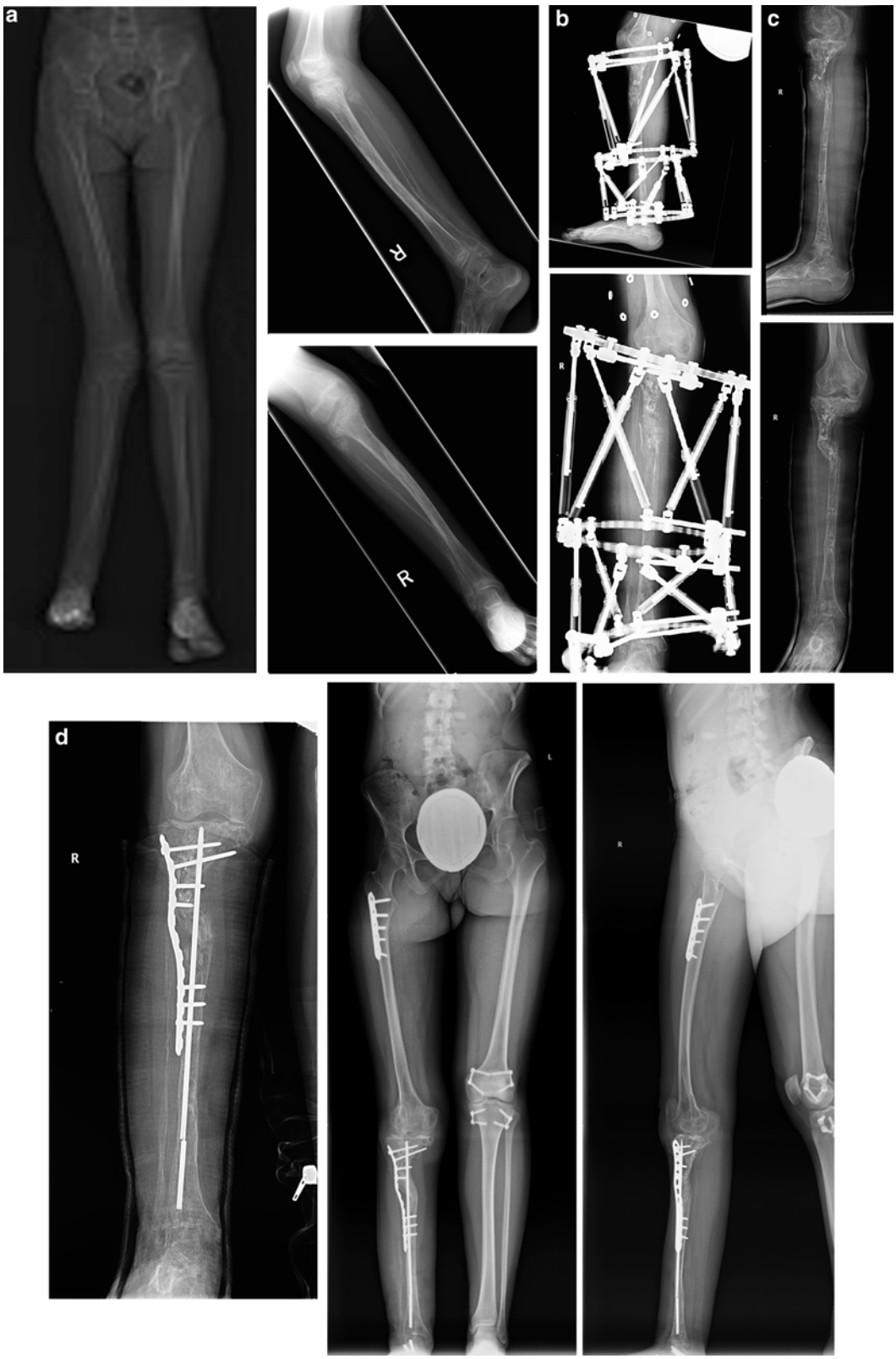


Fig. 7.7 Poor regenerate secondary to an underlying condition. An 11-year-old patient with hemangiomatosis and tibial bowing underwent deformity correction with hexapod external fixation. Hemangiomatosis prevented proper healing of the osteotomized bone, which necessitated stabilization of the bone internally with a rush rod and locking plate. (**a**) Preoperative radiographic view. (**b**) During correction. (**c**) Delayed union. (**d**) Final result

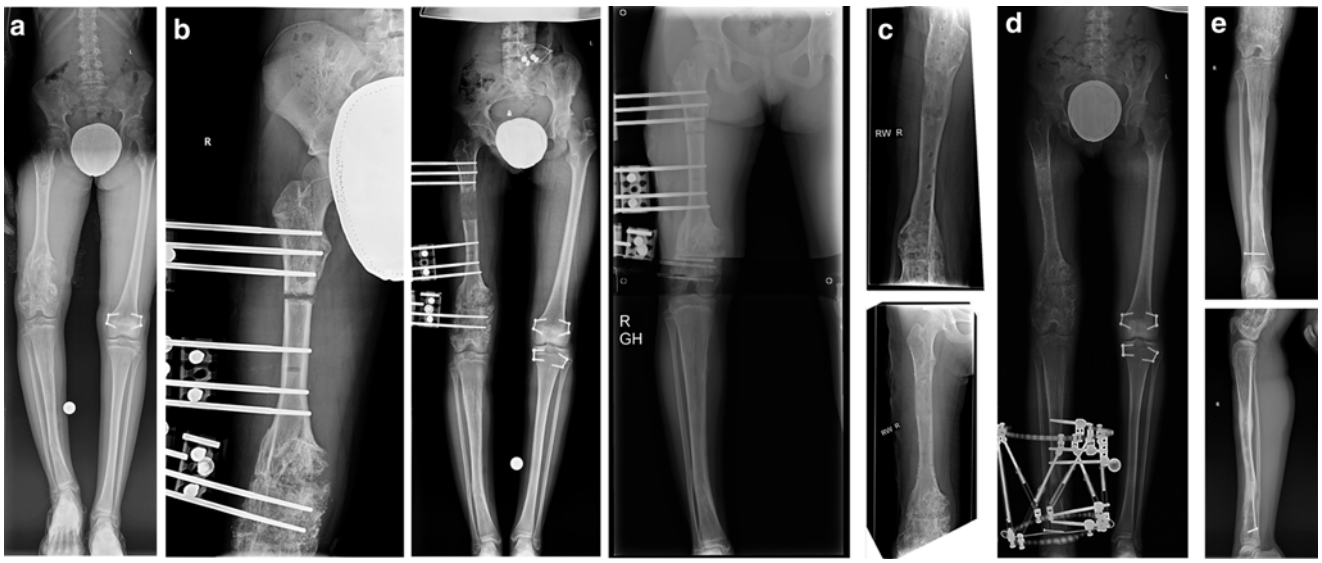


Fig. 7.8 Delayed healing secondary to underlying condition. This case demonstrates deformity correction and lengthening in the setting of Ollier's disease. The patient had limb length inequality and varus deformity and underwent lengthening of the femur proximally with a monolateral fixator and deformity correction of the distal femur with the same monolateral frame. The tibial deformity was treated with a hexapod fixation device. The patient underwent simultaneous growth modulation on the contralateral side. (**a**) Preoperative radiographic view. (**b**) During lengthening. (**c**) Femur after femoral lengthening and deformity correction. (**d**) During tibial deformity correction. (**e**) Final result, tibia

tracking problems and injury to the tibial physis [24]. However, performing the osteotomy away from the apex has its consequences. Translation of the bone is necessary in such cases in order to restore the normal mechanical axis and allow the joint lines to remain parallel to the floor. The further the osteotomy is from the apex of the deformity, the larger the magnitude of the required translation. Additionally, it is necessary to keep in mind that gradual translational correction always begins with distraction. As long as the surgeon accounts for these factors preoperatively, they should not prove problematic.

The anticipated outcome of deformity correction is a normal mechanical axis without oblique joint lines (Fig. 7.9). In most cases, the need to have a straight bone is less important than having a corrected mechanical axis (see Fig. 7.1). Exceptions to this are the cases of metabolic bone diseases, such as hypophosphatemic rickets (see Fig. 7.2) or osteogenesis imperfecta in which a straight bone is often necessary for proper healing and the avoidance of later stress fractures.

Box 7.6
- Factors affecting the location of the osteotomy include the etiology and the location of the deformity.
- Bone quality may affect the osteotomy location and the type of fixation utilized.

Devices

Many techniques and various types of fixation devices can be utilized once the preoperative planning phase has been completed. External fixation remains the gold standard for gradual deformity correction as it allows easy and accessible postoperative manipulation of the deformity. Both circular and monolateral options exist. Many circular systems utilize the principle of the hexapod fixator in which there are two rings: a fixed reference ring and a mobile ring (see Figs. 7.1, 7.2, 7.3, 7.4, 7.5, 7.6, 7.10, 7.11, and 7.12). This configuration allows for a three-dimensional movement of the mobile ring relative to the reference ring. The final configuration is the result of a combination of many uniplanar movements namely anterior–posterior translation, medial–lateral translation, and internal–external rotation. It is simply many vectors adding up over time to give the final vector correction, which is the desired orientation of the distal fragment [2]. Many hexapod systems are available including the Eisenberg fixator™, the Ortho-SUV™, the Smart Correction™, the Spider Frame™, the TL-Hex™ and the Taylor Spatial Frame™. These devices can be utilized to correct deformity not only of the tibia [2] or femur [25] but of the foot [26, 27] and upper extremity as well.

Monolateral devices, which are more typically used in acute correction, may also be utilized in gradual deformity correction (Fig. 7.13). The multi-axial correcting (MAC™)

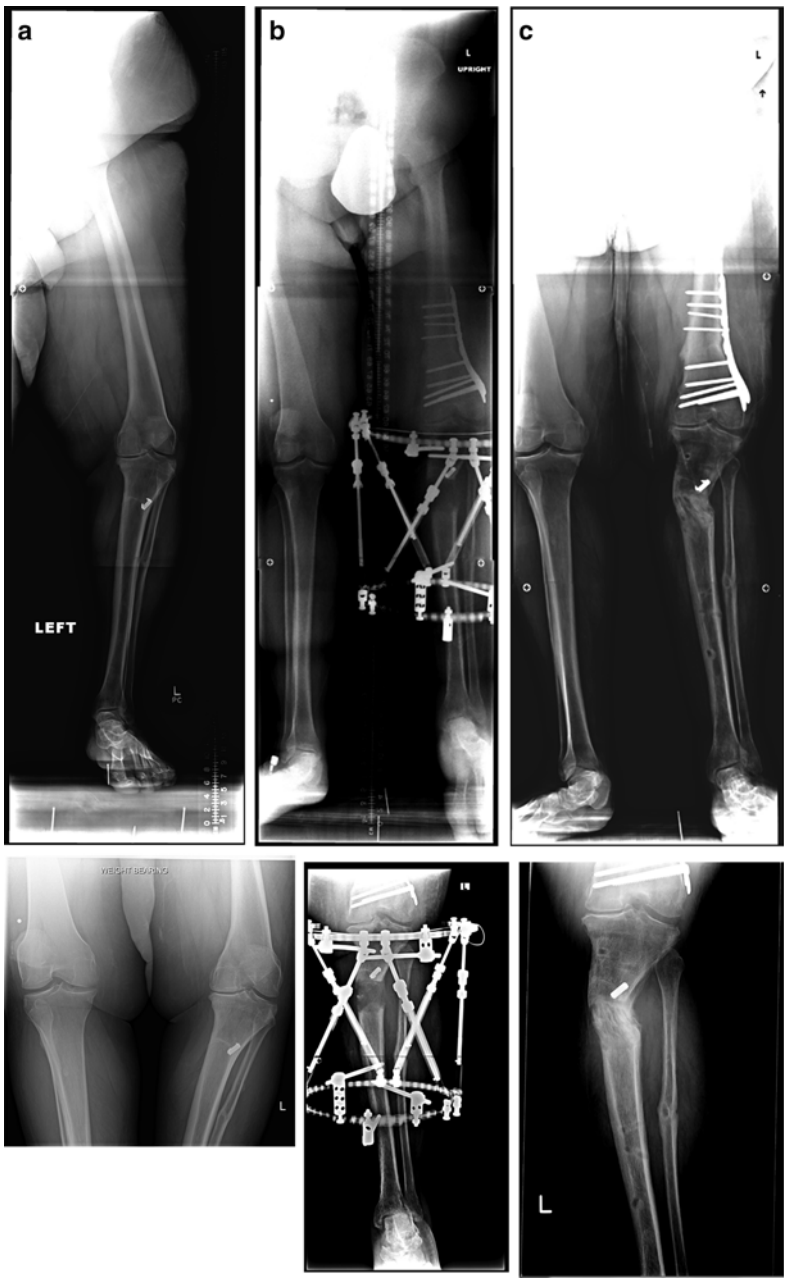


Fig. 7.9 Surgical correction at the location of the deformity, i.e., tibia or femur, is essential. This patient initially presented with 20° of femoral varus and a near normal tibia. The patient's varus was surgically corrected by 10° via a distal femoral osteotomy, leaving 10° of varus unaddressed. The patient then underwent a tibial osteotomy with external hexapod fixation in an attempt to address the remaining varus deformity. This resulted in a corrected mechanical axis but a valgus deformity in the proximal tibia and created oblique joints of the knee and ankle. The patient's resultant MPTA was 103° (15° of valgus) and aLDFA 94° (13° of varus). This oblique joint line creates sheer stress in the joints involved. The patient now has pain in his ankle and knee that will require a revision osteotomy. (**a**) Preoperative radiographic view. (**b**) During deformity correction. (**c**) Final result with oblique knee and ankle joints with pain. This osteotomy will require a revision

fixator system is an example of a monolateral fixation that allows for gradual correction in all planes. The MAC™ system allows for two planes of angulation, two planes of translation, compression, distraction and rotation. The adjustments are usually done in the outpatient setting. This system does not employ the use of computer software to plan the correction [28–30].

A locking plate, used in concert with external fixation (lengthening over a plate), mimics internal fixation creating a stable fixed angle construct within the limb. It is inserted percutaneously, allowing minimal iatrogenic soft tissue injury, and provides stability when locked after distraction osteogenesis. The locking plate allows for early removal of the frame without concern of loss of correction through bending of the regenerate [31] (refer to Chap. 8 for more details).

With the advent of internal lengthening devices, the use of internal fixation to achieve correction is gaining popularity. One such method is the reverse planning technique which achieves both length and deformity correction. As the name implies, this technique involves planning the correction in reverse, i.e., starting with the desired deformity correction and length and determining the appropriate position of the self-lengthening intramedullary nail (IM) at the time of the osteotomy. Blocking screws are used to obtain optimal nail position and to prevent rod migration and loss of correction (Fig. 7.14) [32] (refer to Chap. 9 for more details).



Fig. 7.10 Recurvatum. This 14-year-old patient with recurvatum and limb length inequality of the tibia secondary to posttraumatic physeal arrest underwent proximal osteotomy, deformity correction, and tibial lengthening with external hexapod fixation. (**a**) Preoperative clinical and radiographic view. (**b**) During correction. (**c**) Final result

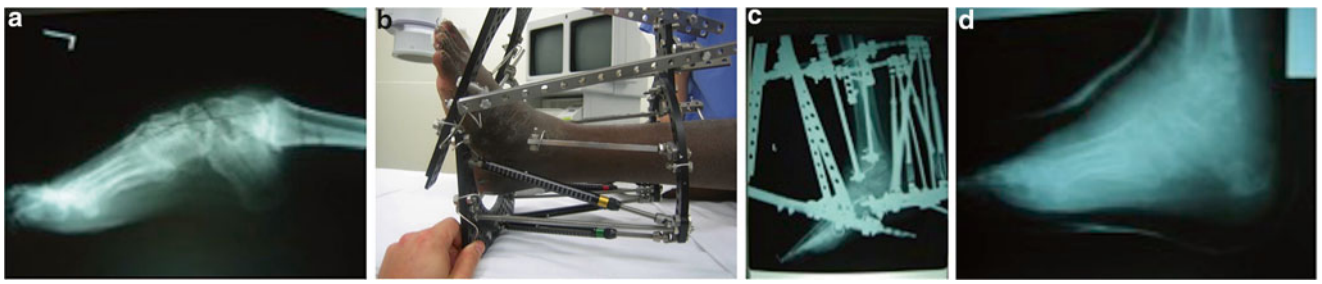


Fig. 7.11 Gradual deformity correction of untreated clubfoot in a young adult with external fixation. (**a**) Preoperative radiographic view. (**b**) Intraoperative clinical view. (**c**) During correction. (**d**) Final result

Box 7.7

- Circular fixators, monolateral fixators, intramedullary devices and hybrid techniques can be utilized for gradual correction.
- Blocking screws are often needed when intramedullary devices are used.

Postoperative Care

The use of gradual deformity correction demands a compliant patient (and caretaker) and a devoted surgeon. Applying the fixation device is only the start of the journey. The surgeon must be committed and able to accommodate seeing the patient on a frequent basis. The patient and caretaker must follow instructions and adhere to the postoperative regimen, which includes the correction schedule, physical therapy, and weekly office visits. Regular and close follow-ups minimize complications and allow for early detection when untoward events occur. Some complications, such as joint subluxations and dislocations, if unattended, may be deleterious and life altering for the patient.

Patients can typically be discharged within 2–3 days postoperatively, but must be instructed on the proper postoperative program prior to their discharge. Protected weight bearing with the aid of crutches or a walker is typically permitted immediately following surgery. While an inpatient, physical therapy should commence to teach the patient exercises to maintain and restore range of motion of local joints. Physical therapy should be continued several times per week as an outpatient, with range of motion exercises and progressing to strengthening exercises (refer to Chap. 12 for more details).

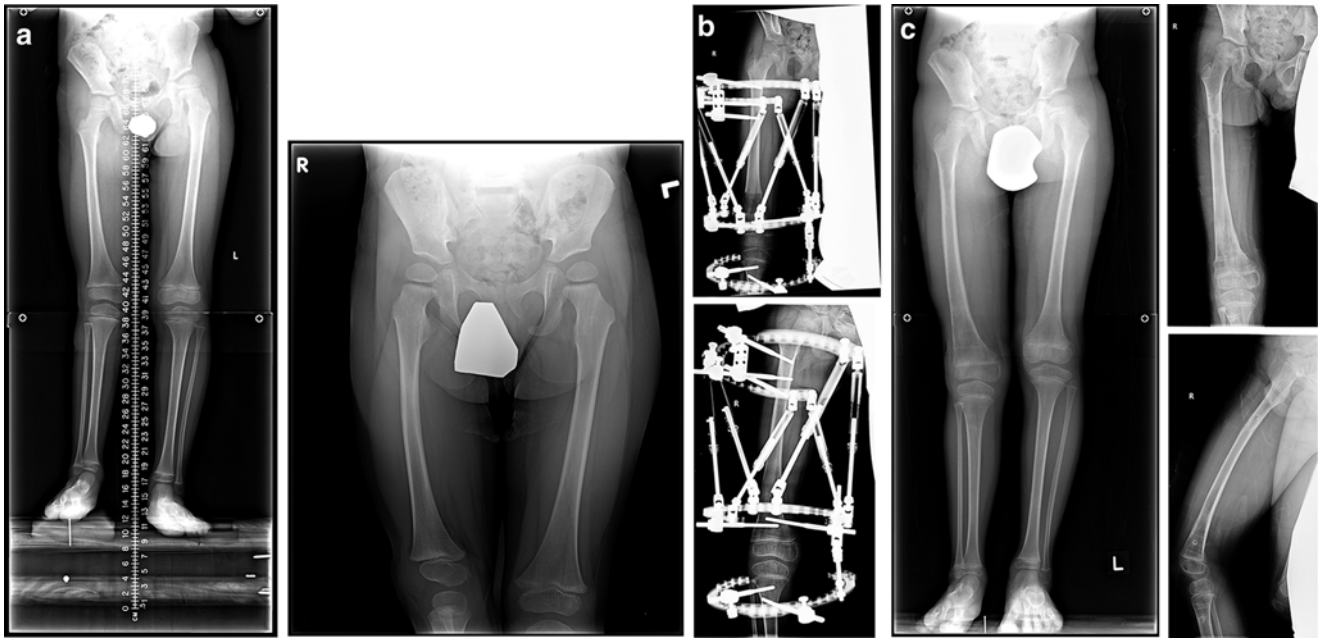


Fig. 7.12 Protecting against joint subluxation and dislocation. This case demonstrates distal femoral deformity and limb length inequality secondary to a congenital femoral deficiency and associated knee joint laxity. The patient underwent deformity correction and femoral lengthening with a knee spanning hexapod fixator to prevent knee subluxation and dislocation. (**a**) Preoperative radiographic view. (**b**) Knee stabilized by external fixator during correction. (**c**) Final result

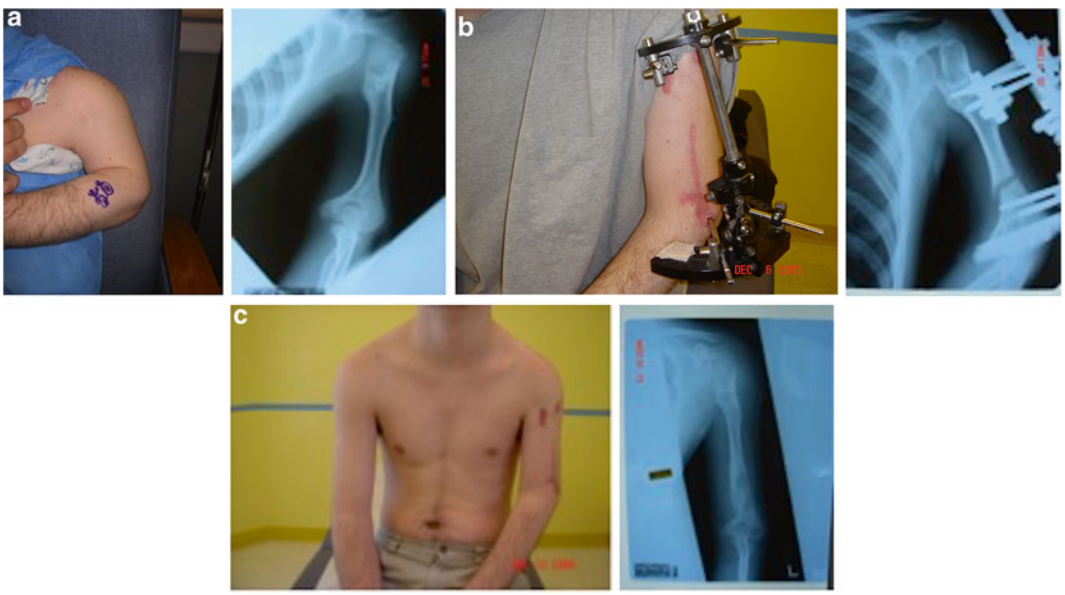


Fig. 7.13 Deformity correction with monolateral fixation device. A 15-year-old patient with humeral deformity and limb length inequality secondary to neonatal sepsis-related physeal arrest. Deformity correction and limb lengthening was performed using a monolateral fixation device. (**a**) Preoperative clinical and radiographic view. (**b**) During correction. (**c**) Final result

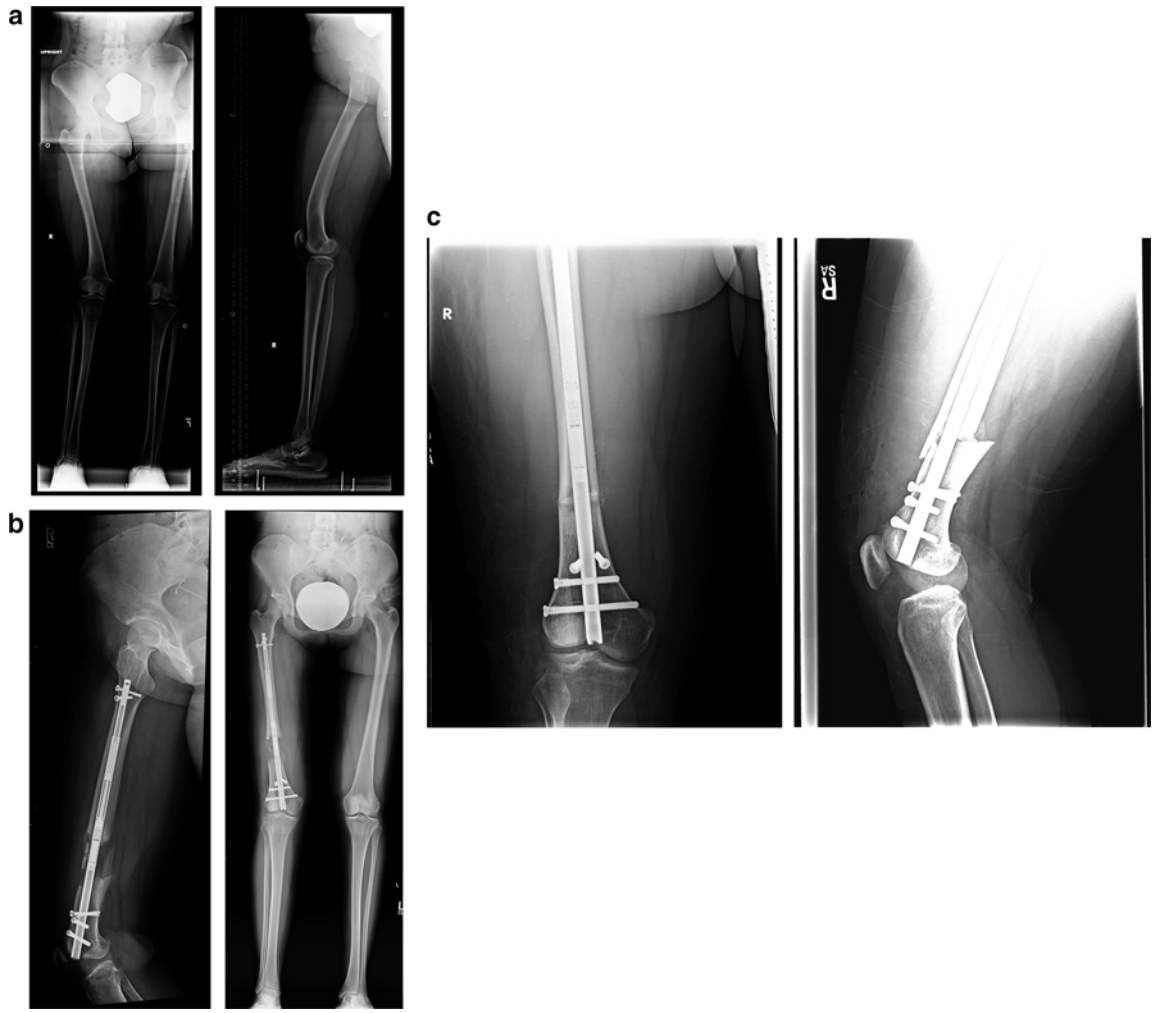


Fig. 7.14 Acute deformity correction with gradual lengthening: intramedullary rod with blocking screws. (**a**) Preoperative radiographic view. (**b**) Immediate postoperative radiographic view. (**c**) During correction

Follow-up visits should typically be scheduled weekly for the first month and then every 2 weeks until correction of the deformity has been completed. When tricortical consolidation is observed on AP and lateral radiographs, the frame is often dynamized for 2 weeks prior to removal. Upon removal, the patient is allowed to foot-flat weight-bear with the use of assistive devices, followed by gradual increase in weight bearing until the return of full function [33].

Box 7.8

- A dedicated patient and surgeon with close follow-up are necessary for successful deformity correction.
- Physical therapy and a home exercise program are necessary to protect the adjacent joints.

Potential Complications

The potential complications of performing gradual correction can primarily be divided into those that are related to surgery more generally and those that are specific to this type of procedure. General complications include those related to anesthesia, infection whether at the surgical site or at a pin site [34] and delayed union or non-union, which is a potential complication of any surgery that involves an osteotomy.

Procedure specific complications include joint stiffness, joint instability, subluxation, and potential dislocation. The surrounding joints remain at risk during the correction phase and lengthening process. The ankle, knee, and hip all may be subjected to significant strain during gradual correction due to incidental limb lengthening. The hip is susceptible to dislocation both during and after femoral lengthening, especially with a dysplastic hip. If the patient presents with acetabular dysplasia, this should be treated prior to femoral lengthening and deformity correction. Pelvic pins can also be utilized to stabilize the hip if it is inherently unstable (Fig. 7.15). Furthermore, an adductor or psoas release may be required to reduce stresses on the hip.



Fig. 7.15 External fixator spanning the hip joint utilizing pelvic pins to stabilize the pelvis during deformity correction. Radiographic view, pelvic pins

The knee is particularly at risk when treating congenital femoral deficiencies and must be observed postoperatively for subluxation. To adequately protect the knee joint and ensure maintenance of full extension, a hamstring and/or iliotibial band release including the lateral intermuscular septum may be necessary. The knee and hip are further at risk for dislocation if femoral rotation is not properly corrected (Fig. 7.16).

During tibial lengthening the ankle is also at risk for a variety of complications. If the ankle becomes tight in equinus, treatment initially includes physical therapy, but should tightness persist a percutaneous tendoachilles lengthening or gastrocnemius recession may be required. Fibular migration may occur proximally, especially when performing deformity correction expected to result in an increase of length greater than 2.5 cm. To preemptively protect against this, a syndesmotic screw is inserted through the distal fibula into the distal tibia (Fig. 7.17). Performing a gradual deformity correction, as opposed to an acute correction, lessens the incidence of nerve damage; however, it remains a concern. In cases of genu valgum greater than 15–20°, a peroneal nerve release may be required to prevent motor dysfunction, dysesthesias, and pain on the dorsum of the foot.

Box 7.9

- Joints can sublux or dislocate around the area of correction and must be protected.
- Spanning fixators can be used to protect the joint during deformity correction.
- Soft tissue releases may be utilized to prevent and/or treat contractures.

Summary

Gradual deformity correction has grown in popularity from the early 1900s through Ilizarov and De Bastiani to the present day. Instrumentation has evolved allowing for a more facile gradual correction of complex deformities utilizing external fixation and intramedullary lengthening devices. The future entails ever-improving devices and a better understanding of the use of biologics to enhance the regenerate bone.



Fig. 7.16 Hip and knee complications: adolescent with a congenitally short femur that had previously undergone a successful 4-cm lengthening. The patient now had a limb length inequality of 5 cm and varus. Bifocal deformity correction and lengthening was performed with a monolateral external fixator that was placed on the femur and did not span the hip or knee joint. Postoperatively the patient developed a subluxed hip and a rotary subluxation of the knee followed by a dislocation of the hip, which was subsequently relocated. Additionally the patient developed a femur fracture through the regenerate following removal of the fixator that was plated. The patient's femur was fixed in 70° of anteversion and developed osteomyelitis. (**a**) Preoperative radiographic view of the dysplastic hip and varus femur. (**b**) Radiographic view of bifocal femoral deformity correction and lengthening with monolateral fixation. (**c**) Femur fracture. (**d**) Plated femur fracture. (**e**) Osteomyelitis in the femur, fixed in 70° of anteversion. (**f–h**) Progression of the destructive arthropathy and depression of the lateral plateau as well as the rotary subluxation of the knee joint. (**i**) Subluxation of the hip. (**j**) Dislocated hip with a triangular femoral head. (**k**) Intraoperative supine clinical view of the lower extremities prior to surgical relocation of the hip and treatment of anteversion. (**l**) Radiographic view of the relocated hip and knee. The view of the knee demonstrates the extent of the arthropathy. (**m**) Radiographic view of the total hip and knee replacements

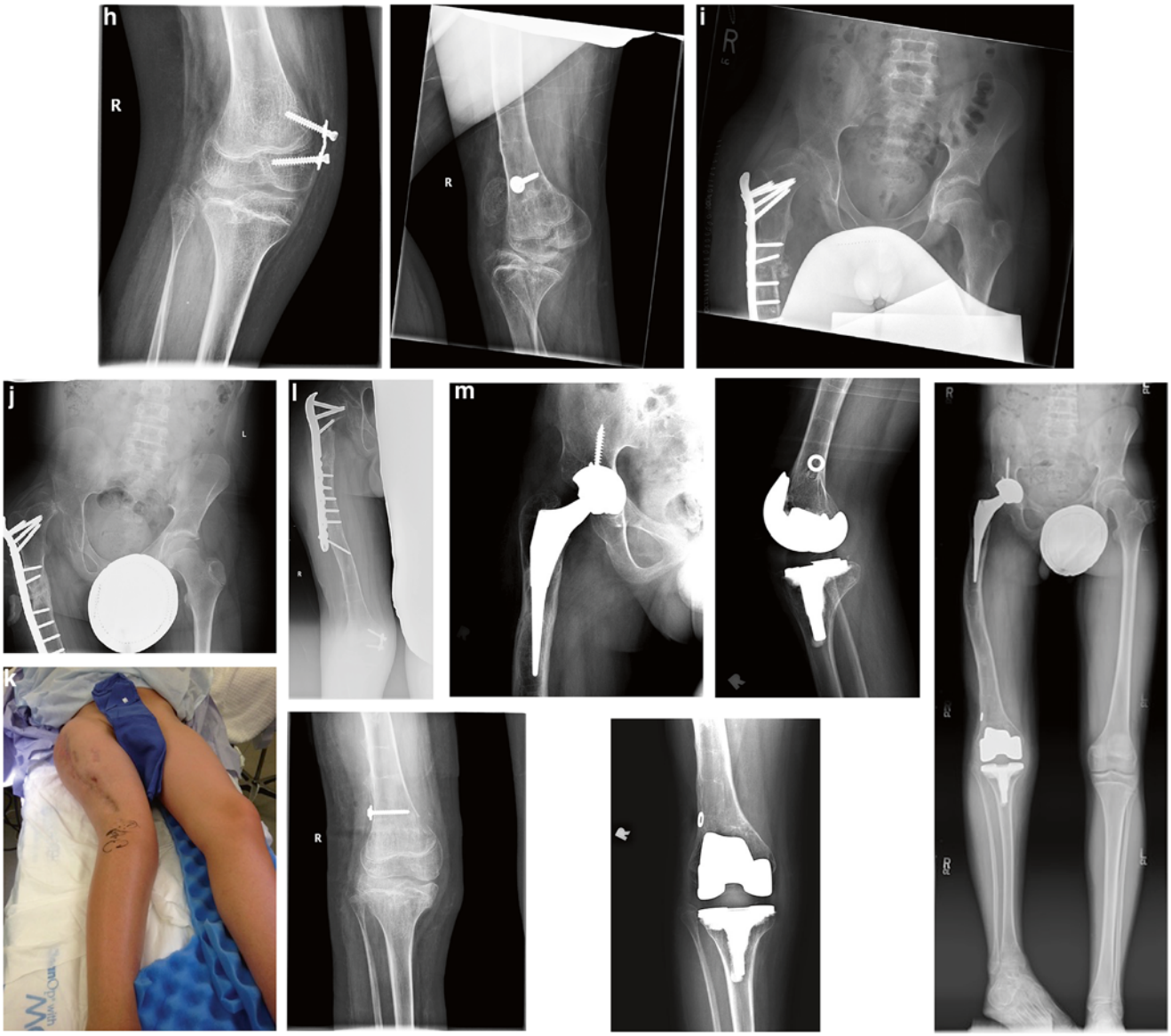


Fig. 7.16 (continued)

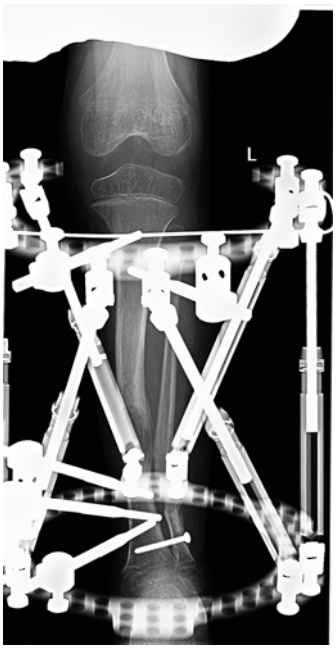


Fig. 7.17 Syndesmotic screw through distal tibia and fibula to prevent fibular migration during deformity correction. Radiographic view, syndesmotic screw

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Hybrid Techniques for Limb Length and Deformity Correction

8

Mark T. Dahl and Chang-Wug Oh

Introduction

The primary indications for bone lengthening and deformity correction include limb length discrepancy resulting from congenital and posttraumatic conditions such as fibular hemimelia, congenital short femur, fracture malunion, and premature growth arrest. Suitable patients include selected children, adolescents, and adults. Distraction osteogenesis (DO) has been successfully used over the last half-century to lengthen bone [1]. Critical to successful DO is an optimal rate and rhythm of distraction for which external fixation has been a reliable tool [2, 3]. The disadvantages of external fixation frames are well known to include pin tract infections, pain, soft-tissue tethering, and joint stiffness. Limb lengthening complications such as joint contracture, stiffness, subluxation, fracture, residual deformity, and chronic pain are well described [4]. The prolonged treatment of bone lengthening with external fixation can be shortened by substituting internal fixation for the external fixator during the consolidation phase of lengthening, thereby saving the patient prolonged external fixation.

Despite these extraordinary gains in the field of limb lengthening and deformity reconstruction, patients and their families must endure months of pin site maintenance and ever-present external fixation. The resulting soft tissue scarring can lead to further contracture and adjacent joint difficulty. Additionally, Ilizarov's methods take years and scores of cases for surgeons to master the techniques [4]. Even after years of experience with bone regeneration, surgeons commonly confront yet another new complication arising from the treatment.

Lengthening Over Nail (LON)

The technique, first proposed by Bost and Larsen, used an intramedullary pin to prevent development of deformity during lengthening in addition to the external fixator used to gain length [5]. Paley, Herzenberg, and Bhave introduced the modern method of lengthening over nail (LON) in 1997 (Figs. 8.1 and 8.2) [6, 7]. These authors described 29 patients treated with an intramedullary rod inserted and an external fixator applied concomitantly at the time of the osteotomy for lengthening. They compared this group to 31 previously operated Ilizarov lengthenings. Lengthening over an intramedullary nail reduced the average duration of external fixation by nearly one-half. Knee range of motion returned to normal 2.2 times faster in the group that had lengthening over a nail. There were six fractures of the distraction bone in the Ilizarov group. In the group that had had lengthening over an intramedullary nail, one nail and one proximal locking screw failed. Complications were reduced by 25 % in the nailing group. The authors described a pattern of over-reaming by 2 mm, essentially under-sizing the nail preventing entrapment of the rod. They described placing pins intra-cortically, leaving them out of the marrow space to avoid deep infection. The advantages of lengthening over an intramedullary nail thus include a decrease in the duration of external fixation, protection against lengthening site fracture, and earlier rehabilitation.

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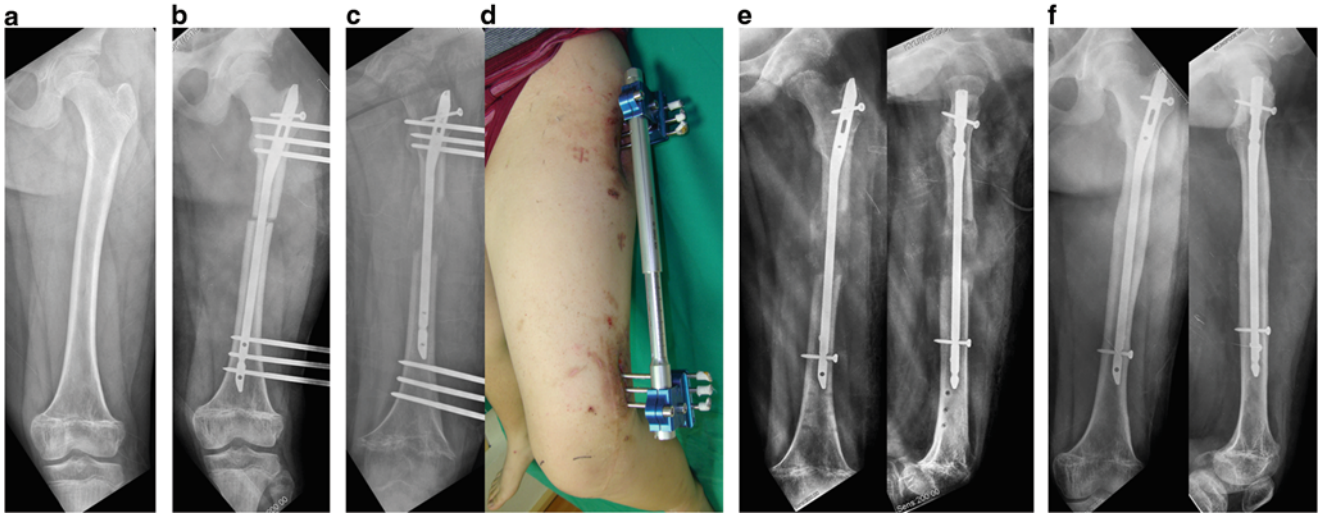


Fig. 8.1 (**a**) A 12-year-old boy had a femoral shortening of about 4.5 cm due to the growth arrest of distal femur. (**b**) Lengthening over an IM nail was performed using a trochanteric entry tibial nail and a monolateral external fixator. (**c**, **d**) The target length was achieved. (**e**) Two interlocking screws were fixed at the distal segment and the external fixator was removed. (**f**) 1 year later, the distraction callus had healed



Fig. 8.2 This soldier was struck by an IED (improvised explosive device), causing open brain trauma and open fractures of the left femur and right forearm. (**a**–**c**) A 10-cm leg length discrepancy (LLD) with angulation, rotation, and translation of the femur developed. (**d**–**f**) Acute correction of the angulation, translation, and rotation was followed by lengthening over nail of 10 cm. (**g**) Functional gait was restored

Gordon et al. described their initial experience using LON in preadolescents in a retrospective series [8]. Nine children (average age, 9 years and 10 months) with femoral length discrepancy were treated with femoral lengthening over a humeral intramedullary nail. After nail insertion, a monolateral external fixator was placed and lengthening was performed through a proximal osteotomy. No avascular necrosis or proximal femoral valgus was noted at a minimum of 2 years follow-up. The authors concluded that the "technique is effective but has a high rate of complications, including osteomyelitis, which developed in two of the nine patients." The technical challenge of pin placement remote from the intramedullary device requires practice, but is necessary to prevent the risk of bacterial contamination of the canal from the external fixator pins.

Box 8.1

- When positioning the external fixator pins or wires, contact with the IM nail should be avoided.
- Over-reaming of the medullary canal by 1.5–2 mm may be needed to under-size the diameter of nail, and thus prevent binding of the nail during lengthening.

Plate-Assisted Lengthening (PAL)

Iobst and Dahl first introduced this technique in 2007 [9]. This study is a description of the surgical technique and a retrospective examination of six patients treated using a technique designed for children with open growth plates as a means to remove the fixator sooner than with a fixator alone. A percutaneous locking plate is inserted at the beginning of the lengthening, along with a circular external fixator. Axis deviation was a problem in two cases, requiring adjustment during the locking of the plate. The plate is percutaneously locked at the completion of lengthening, at which time the fixator is removed. The procedure is tedious because of fixator hardware interfering with access for locking screws. The goal of shortening fixator time was accomplished with a mean external fixation index of 0.42 months/cm. Oh et al. also described a technique similar to limb lengthening with a submuscular locking plate [10, 11]. Submuscular plating has the advantage of providing angular stability and preserving the periosteal and endosteal blood supply, which is beneficial for distraction osteogenesis. The main indication of this technique is when an intramedullary nail is not a viable option, as in patients with a narrow medullary canal, bone deformity, open physes, or joint contractures (Figs. 8.3, 8.4, and 8.5).

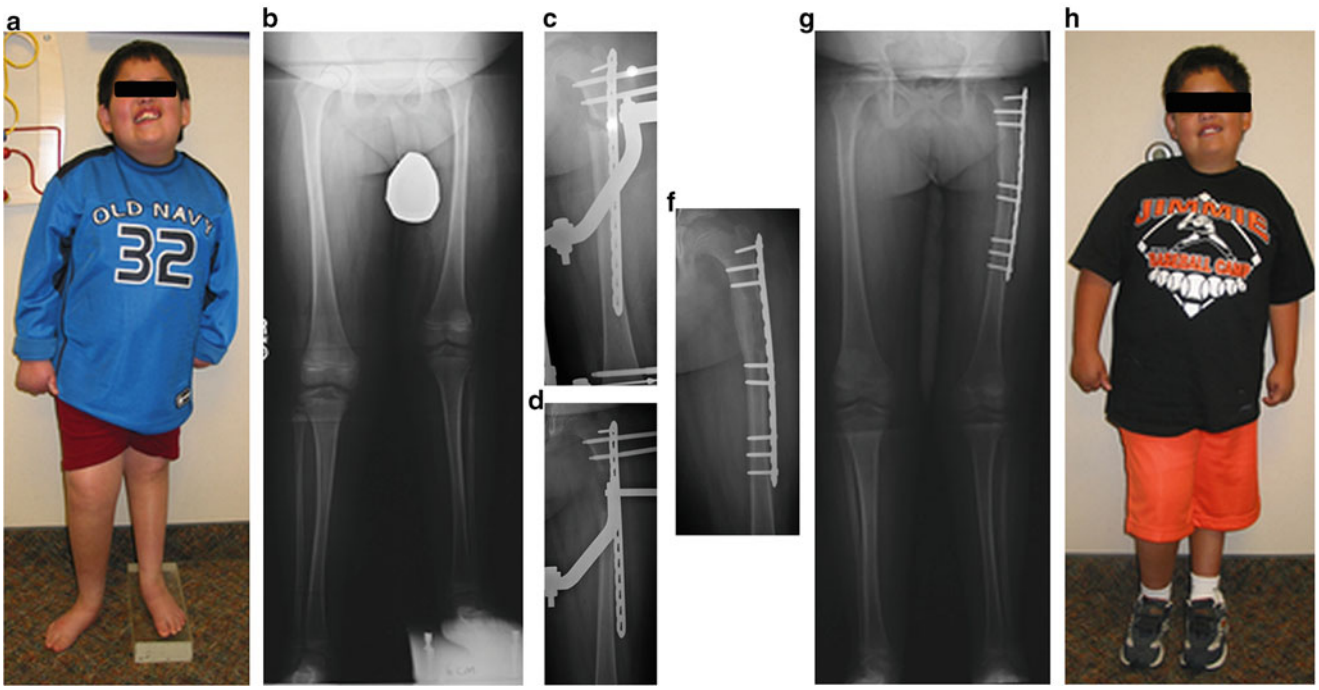


Fig. 8.3 (**a**, **b**) Clinical photo and AP X-ray of 6-year-old boy with 5.5-cm LLD secondary to congenital femoral hypoplasia. (**c**) AP X-ray of femur at 1 week of distraction. (**d**) AP X-ray of the femur at 3 weeks distraction. (**e**) AP X-ray of the femur at 7 weeks, after plate locking and frame removal. (**f**, **g**) X-ray and clinical photo at completion of treatment

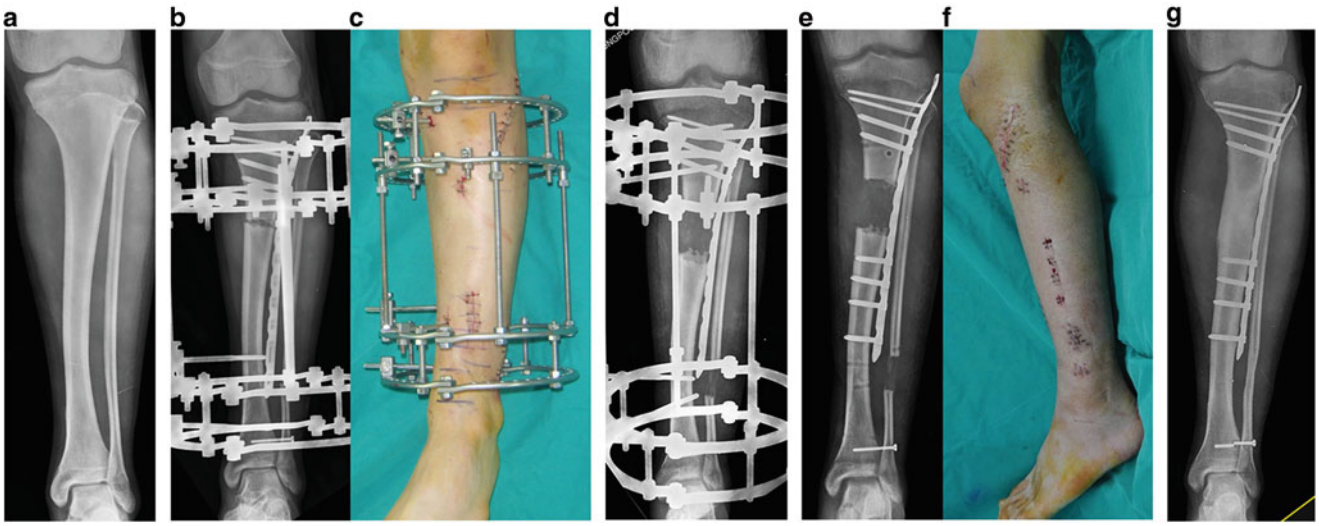


Fig. 8.4 (**a**) A 15-year-old boy had a tibial shortening of about 4.5 cm due to congenital postero-medial bowing. (**b**, **c**) A submuscular locking plate was placed laterally without involving the physeal plate, and a ring external fixator was fixed. After achieving the target length (**d**), four screws were percutaneously placed in the distal segment (**e**) and the external fixator was removed (**f**). Seven months later, the distraction callus had healed without complications (**g**)

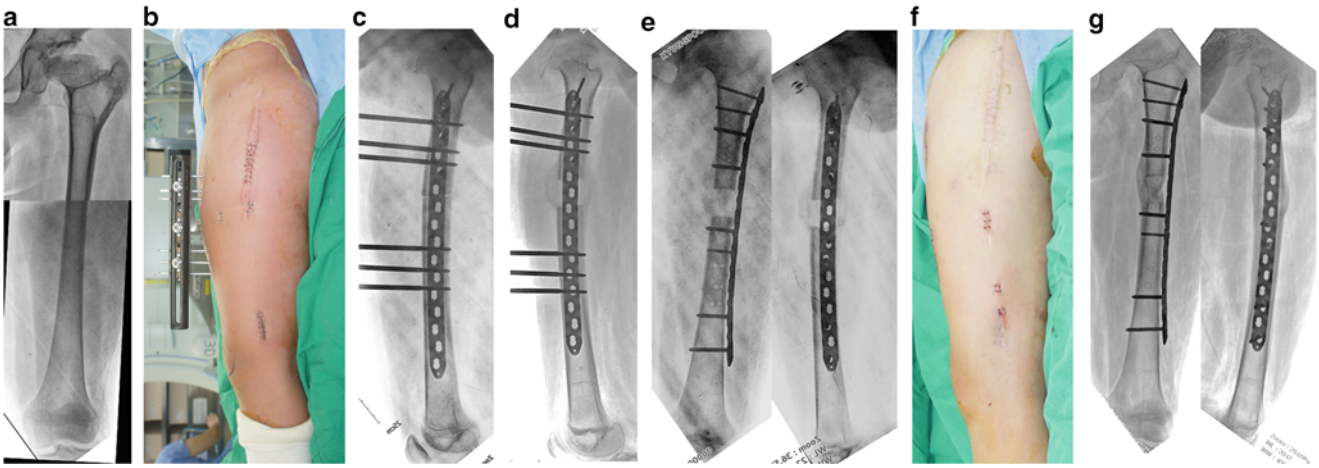


Fig. 8.5 (**a**) A 14-year-old boy had a femoral shortening of about 3.5 cm due to the Perthe's disease. (**b**, **c**) Submuscular locking plate was placed laterally, and a monolateral external fixator was fixed anteriorly to lengthen the femur. After achieving the target length (**d**), four screws were percutaneously placed in the distal segment (**e**) and the external fixator was removed (**f**). (**g**) 10 months later, the distraction callus had healed uneventfully

Box 8.2

- When placing the external fixator, half-pins or wires should not contact plate or screws, to avoid cross-contamination, which may evolve into deep infection.
- To give sufficient stability, a longer plate should be chosen unless an anatomical contraint is present. Also, enough screws are needed to stabilize each segment.
- Protection from full weight bearing and the use of a protective device (such as a patellar tendon bearing brace in tibia lengthening) is recommended until sufficient consolidation of the distraction callus is achieved.
- When fixing a screw in skeletally immature patients, the adjacent physis should not be violated.
- Insertion of screws in the distal segment at the end of distraction period should be performed prior to removal of the external fixator. This will prevent loss of correction that was gained at the lengthening site.

Lengthening and Then Plating (LAP)

This hybrid technique employs an external fixator lengthening, with the external fixator replaced by a percutaneously inserted plate at the completion of the bone lengthening (Figs. 8.6 and 8.7) [12]. Rozbruch and Fragomen published a retrospective case-matched comparison between LAP and a more typical Ilizarov technique with 27 extremities in each group [13]. The time wearing the frame and external fixation index were lower in the LAP group (4.5 vs. 6.2 months and 1.5 vs. 2 months/cm). Axis deviation was observed in seven patients in the LAP group and six in the classic group. Varus malalignment with plate breakage occurred in two patients in the LAP group. Pin-tract infection was more common in the classic group (12 vs. 2). No deep infections occurred in the LAP group.

The advantage to plating after lengthening is that the circular fixator allows for a precise correction of alignment before the plating. Care must be taken to leave open access for the plate insertion, so that contaminated pin sites do not later cause infection of the plating site. Oh et al. noted that this technique is beneficial, especially when an IM nail is not appropriate for children [14]. Rozbruch described that residual deformity may lead to plate breakage [13].

Box 8.3

- LAP technique is beneficial in children who have an open physeal plate or a small intramedullary canal, as the IM nailing may be difficult to perform.
- Because this technique does not require exposing the distraction area and soft tissue dissection is kept to the minimum, the risk of deep infection can be minimized.
- Caution is still needed not to involve pin tracks of the external fixator, which can be a source of delayed infection.

Bone Transport with a Plate

For the segmental bone defect, trifocal and tetrafocal methods of bone transport with multiple osteotomies and two or three levels of bone regeneration can shorten treatment times. However, the prolonged duration of external fixation required to enable consolidation of distraction callus is a disadvantage and is frequently associated with complications,

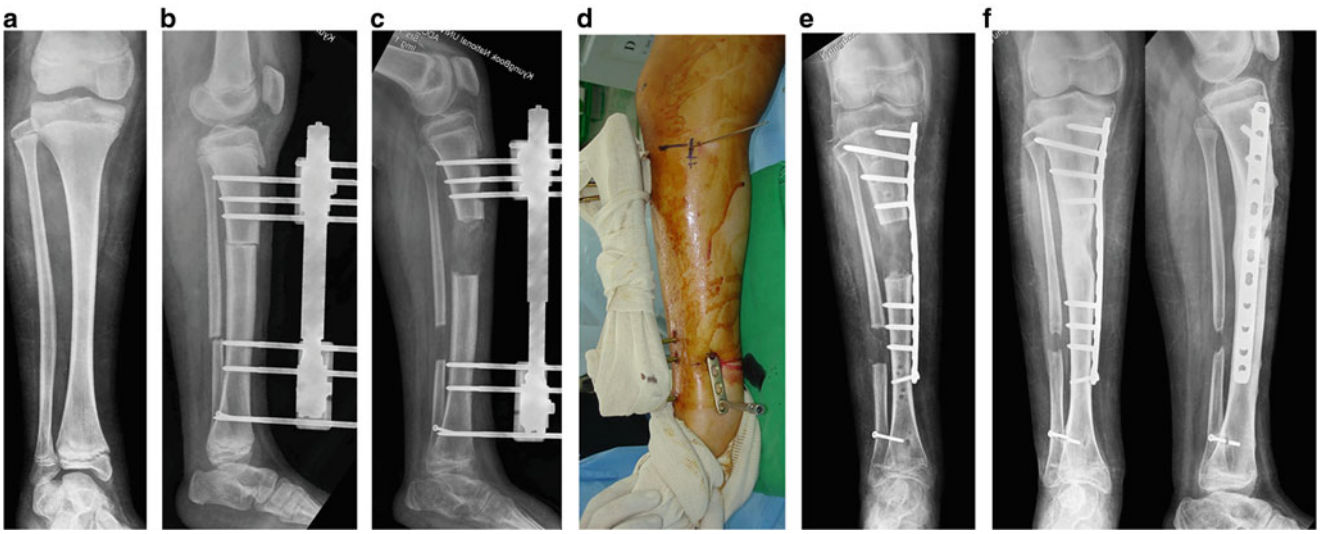


Fig. 8.6 (**a**) An 11-year-old boy with a fibula hemimelia had a tibial shortening of about 4 cm. (**b**) A monolateral external fixator was fixed anteriorly to lengthen the tibia. (**c**) The target length was achieved. (**d**) Then the percutaneous plating was performed on the medial aspect of the leg, and (**e**) the external fixator was removed. (**f**) The distraction callus had healed uneventfully

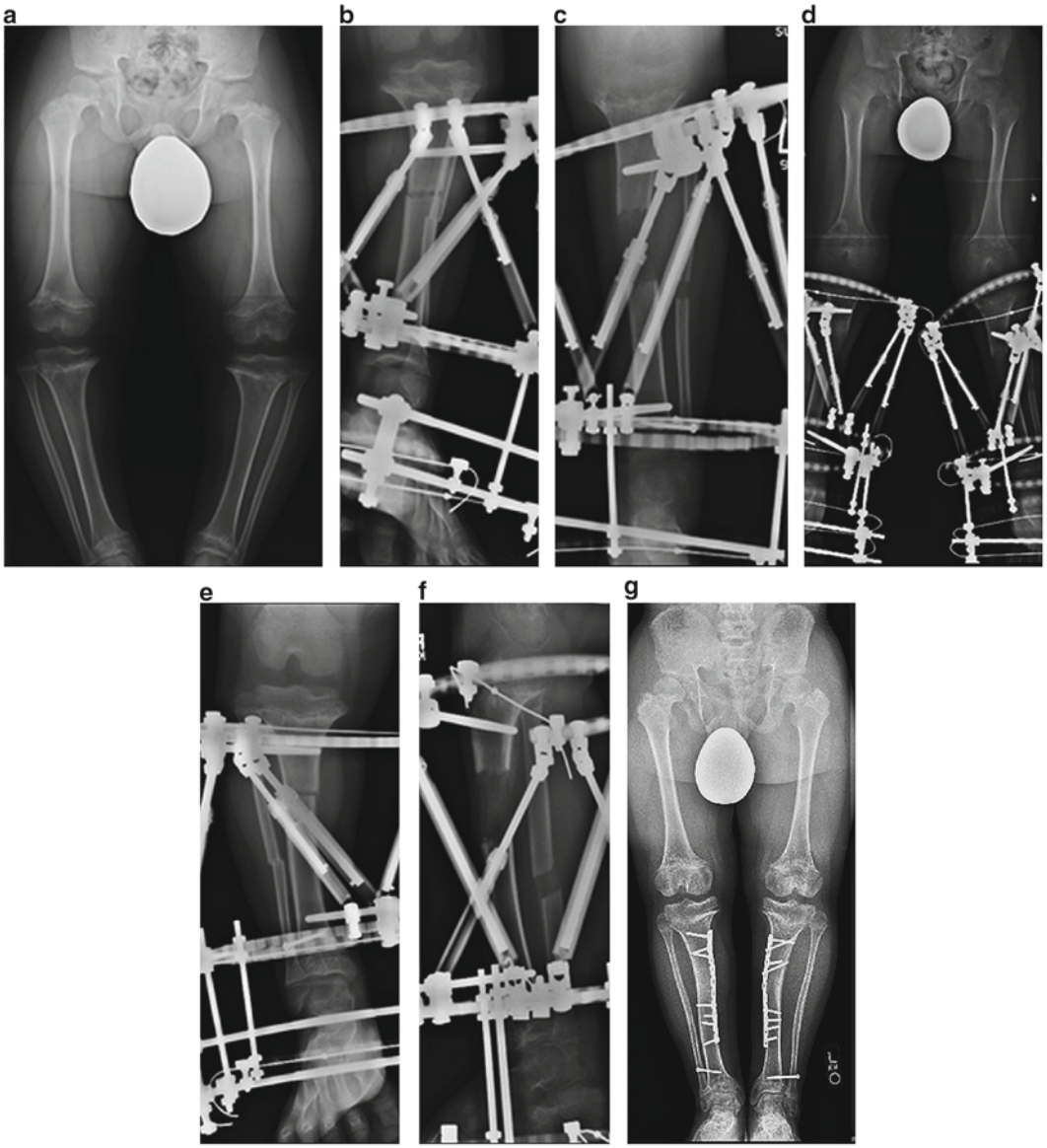


Fig. 8.7 (**a**) Full-length standing AP X-ray of a 12-year-old boy with hypochondroplasia and bilateral symptomatic genu varus. They desired a knee correction and lengthening to improve stature. (**b–f**) X-rays showing the initial frame application, followed by simultaneous angulation and length correction. Part (**g**) shows healing after percutaneous contoured plating of the lengthening sites with simultaneous frame removal

such as pin tract infection, joint stiffness, discomfort, and refracture. Bone transport over an IM nail has been conducted with the aim of reducing the duration of external fixation, but the proximity of the pin to the nail introduces the risk of deep infection and failure of fixation [15]. Oh et al. introduced "bone transport with a plate," which requires a shorter period of external fixation (Fig. 8.8) [16]. Because pre-inserted plate and additional screw fixation at the transported segment add stability to the construct, this technique can eliminate the fixator time required for union at the distraction area as well as the docking site. The risk of spreading infection can be minimized by placing the fixator half-pins sufficiently distant from the plate or screws. This technique can be used in segmental bone defects following trauma or infection, in both adults and children.

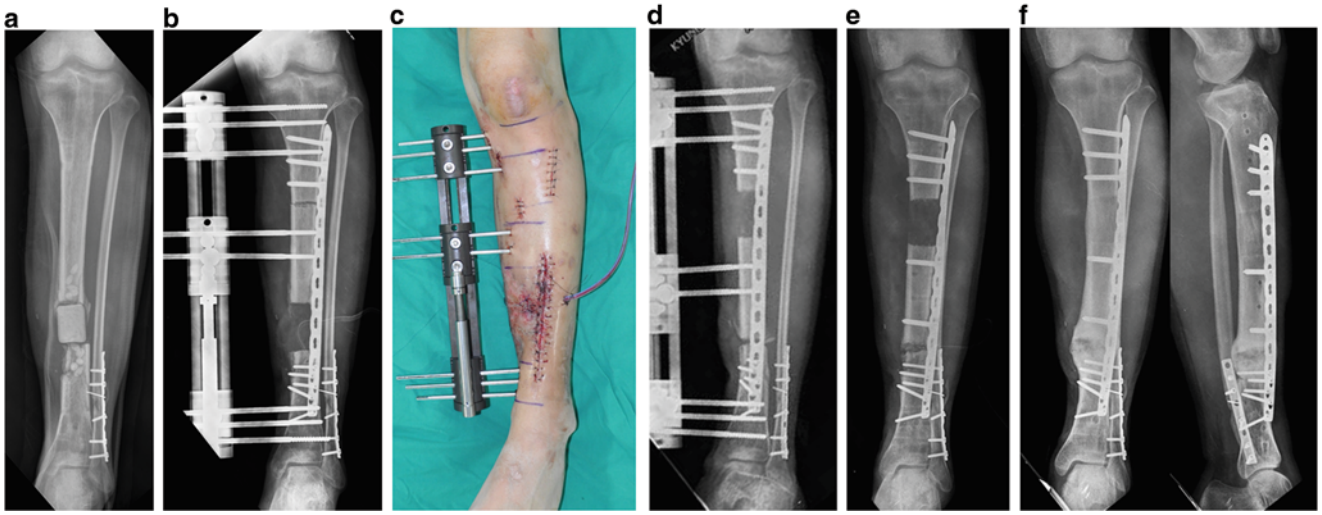


Fig. 8.8 (**a**) An 18-year-old male patient had a segmental tibial defect filled with an antibiotic-impregnated cement block. (**b**, **c**) A laterally based locking plate and a medially based monolateral external fixator were applied for bone transport. (**d**) With a gradual distraction osteogenesis, docking of the transport segment was achieved. (**e**) Then two percutaneous screws were fixed at the transported segment and autogenous bone grafting performed at the docking site. (**f**) Radiographs after 10 months show satisfactory union achieved at the distraction callus and docking site

Box 8.4

- Compared to bone transport over a nail, this technique may further reduce the external fixation time because the screw fixation at the transported segment adds stability and can eliminate the time required for union at the docking site.
- In order to minimize the risk of secondary infection, the external fixation pins are inserted sufficiently remotely from the plate or screws.
- If the remaining segment is too short to insert an IM nail, such as in the case of juxta-articular bone defects, the use of transport with a plate can be very useful.

Lengthening and Then Nailing (LATN)

This technique, described by Rozbruch in 2008 [13], employs a conventional deformity correction and external fixator lengthening, with the external fixator replaced by a percutaneously inserted locked nail at the completion of the bone lengthening. Its greatest indication is in those cases where acute intraoperative deformity correction is most difficult. The circular fixator gradually corrects deformities, at the completion of which a locked nail is inserted (Fig. 8.9).

Lengthening and Then Screw Fixation

Ilizarov taught us that the most extreme foot deformities must be corrected gradually to stretch the soft tissues and neurovascular structures into the desired position. Conditions such as severely deformed clubfeet can be corrected in this way with or without osteotomy. Prolonged subsequent external fixation protects the regenerate bone and corrected position, but often results in foot stiffness. Inserting percutaneous screws, pins, or staples at the time of completion of deformity correction allows for earlier removal of the fixator. In unpublished experienced, one of the authors (MD) has used this technique a few times in recent years (Fig. 8.10). Several weeks of external fixation time can be saved. The percutaneous, fluoroscopic-guided insertion of cannulated screws thus protects the osteotomy sites along with casts or braces.

Summary

A variety of hybrid techniques have been established to improve the patient experience during limb length and deformity correction, by reducing external fixation time to the latency and distraction phases. Each of these techniques shares the requirements of careful preoperative planning, prevention of cross-contamination of external fixator pins, and skill with internal fixation devices.

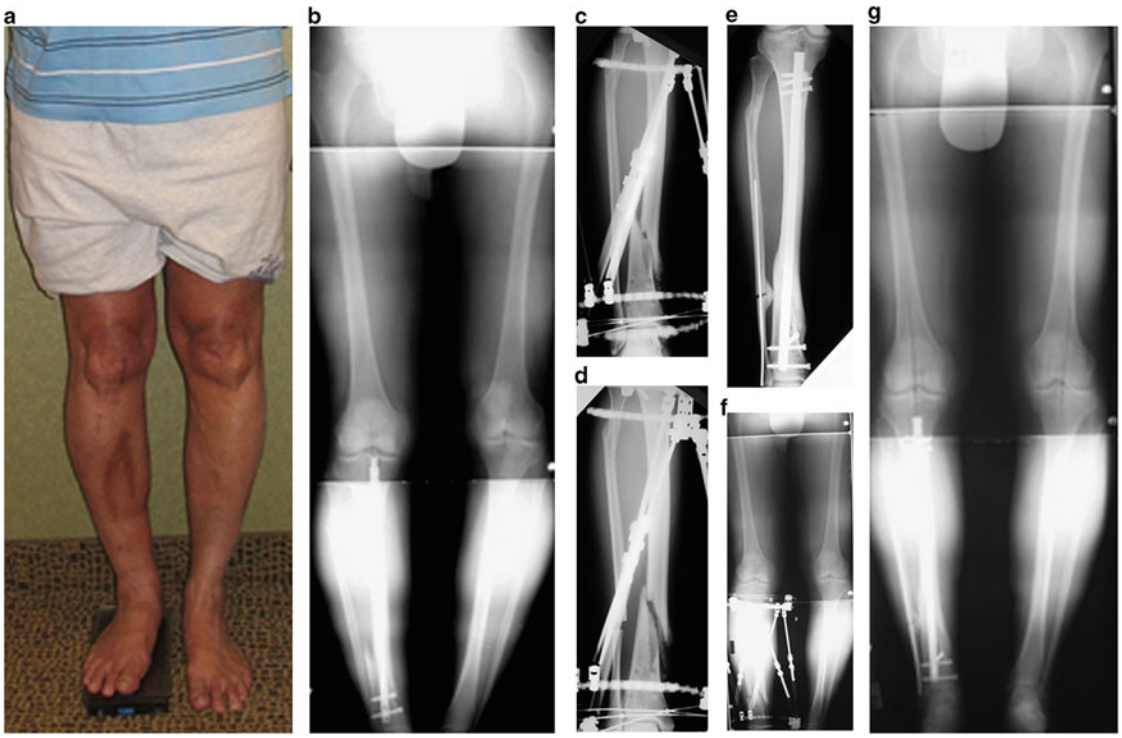


Fig. 8.9 (**a**, **b**) This young man's tibia fracture was treated with an IM nail, resulting in a nonunion, malunion with shortening of 2.5 cm, varus angulation and lateral translation. (**c**, **d**) The existing nail was removed, fibula osteotomized, 6-axis circular fixator applied with gradual length and deformity correction. (**e**) When length was restored, a locked IM nail was placed in the tibia and an IM pin placed in the fibula. (**f**, **g**) Completed lengthening and healing with equal leg lengths was achieved

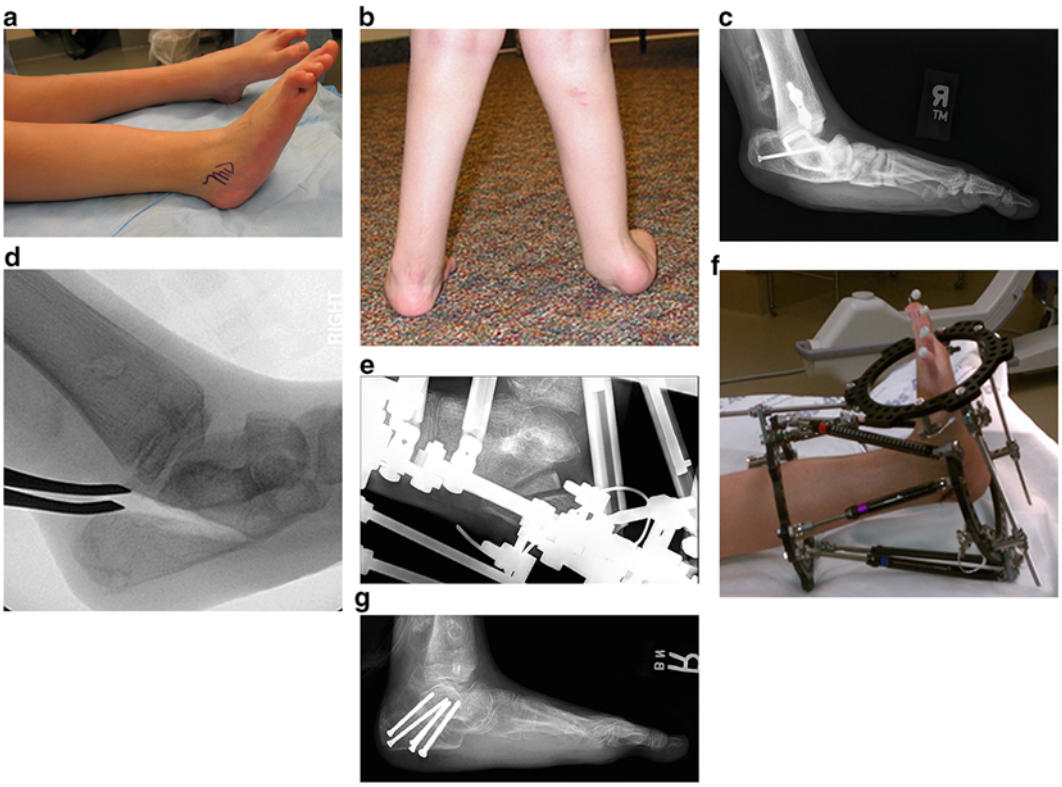


Fig. 8.10 (**a**, **b**) This 11-year-old girl has bilateral fibular hemimelia, with the right foot in extreme equinovalgus. (**c**) Prior surgery failed to create a plantigrade foot. (**d**) A posterior calcaneal osteotomy is partially opened acutely, limited by soft tissue tolerance. (**e**) Interpositional grafting maintains separation of the osteotomy. (**f**) Gradual correction with a circular external fixator is ideal for this deformity. (**g**) Maintaining external fixations until healing is unnecessary, as multiple internal fixation screws provide the support needed and spare the patient an additional 2–3 months in the frame

Box 8.5

- Lengthening over nail reduces external fixation time and facilitates early rehabilitation.
- Plate-assisted lengthening in children reduces fixator time.
- Lengthening and then nailing allows for complex deformity correction followed by nailing.
- Complex foot correction can be done gradually, but secured with screws to hasten fixator removal.

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9 Motorized Intramedullary Lengthening, an Emerging Technology for Limb Length and Deformity Correction

Mark T. Dahl

Introduction

The first description of limb lengthening is credited to Codivilla in 1905 [1]. Limb lengthening has evolved from an open step-cut osteotomy, distracted by bed-confined traction, to Ilizarov's methodology of distraction osteogenesis whose principals include a blood preserving corticotomy, a latency period, regular rate and rhythm of distraction, stable circular external fixation, and functional use of the extremity during treatment. Three decades ago, a small cadre of surgeons learned Ilizarov's methods directly from him, and they are now passing this information on to the next generation of limb reconstruction surgeons. With this "passing of the torch," we now have the option of a technical refinement to the process, that is, remotely controlled, motorized, telescopic, intramedullary lengthening nails. The discomfort associated with the prolonged use of external fixation of any kind is often poorly described in the literature, but is well known to patients and surgeons. Although the Ilizarov methodology included many innovative biological and mechanical concepts that were previously unknown, external fixation is not without its problems, which include length of time required for the treatment, pin site infection, soft tissue tethering, and scarring at the pin sites, with patients often expressing dissatisfaction during the process. This chapter describes the latest methods of delivering Ilizarov's principles to patients by fully implanted, motorized, telescopic, intramedullary lengthening nails.

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Indications

The primary indications for bone lengthening and deformity correction with motorized intramedullary lengtheners include limb length discrepancy resulting from congenital and post-traumatic conditions such as fibular hemimelia, congenital short femur, fracture malunion, and premature growth arrest. Suitable candidates must have completed growth at the adjacent physis, have adequate canal diameter and bone length to accept the available nails, and have a canal suitable for intramedullary nailing. These patients include selected children, adolescents, and adults. Distraction osteogenesis (DO) has been successfully used over the last half-century to lengthen bone. An optimal rate and rhythm of distraction is critical to successful DO, for which external fixation has been a reliable tool [2, 3]. The disadvantages of external fixation frames are well known and include pin tract infections, pain, soft-tissue tethering, and joint stiffness [4]. Limb lengthening complications such as joint contracture, stiffness, subluxation, fracture, residual deformity, and chronic pain are well described [5]. Bone lengthening with a fully implantable device is desirable in order to avoid external fixation, mechanical integrity and accurate control of distraction are mandatory. As with any intramedullary rod, lengthening must occur along the anatomic axis of the bone, which will result in a translation deformity, the effect of which is determined by the original anatomy and the magnitude of the lengthening [6]. The prediction and prevention of these iatrogenic deformities using a novel preoperative planning technique known as "reverse planning" is essential in preventing abnormal mechanical loads in the future [7]. Remote-controlled, motorized, internal lengthening devices (MILD) have recently become available for use in the femur and tibia [8–10].

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History

Ilizarov's remarkable discovery of bone regeneration, commonly referred to as the Ilizarov method, has opened windows into the treatment of many conditions previously considered as hopeless maladies.

It is now possible to combine Ilizarov's principles with those of intramedullary fixation for limb lengthening by using preoperative planning, less invasive surgery, temporary intraoperative external fixation, specific reaming techniques, and intramedullary fixation. Telescopic nails with miniature motors are remotely controlled by either radio frequency or magnetic energy [8, 11]. These techniques require precise planning and execution because only axial length can be adjusted after the surgery without an additional operation.

Distraction osteogenesis (DO), largely introduced by Ilizarov, now enables us to lengthen bones in children with conditions such as fibular hemimelia or a congenitally short femur without the additional need for bone grafting [2, 3]. We spare these children additional surgeries by tapping the body's capacity to form regenerate bone, but the new bone requires the prolonged use of external fixators throughout consolidation. Fractures, stiffness, and residual deformity are often a result of months in external fixation. With DO, we are also able to achieve union in congenital pseudarthrosis of the tibia more predictably, a formerly hopeless condition which often resulted in amputation after multiple attempts at union with bone graft, pins, and casting [12]. The correction of severely deformed limbs from developmental, infectious, and traumatic conditions such as Blount's disease by using simultaneous, gradual angular, rotational, and length corrections can now be more precisely performed with web-based computer analysis than with the Ilizarov device with six-axis correction [13]. Massive bone defects resulting from trauma, infection, and tumor can be filled by spontaneously regenerating the patient's own bone by compressing the defect site while growing new bone at a remote site [14].

Despite these extraordinary gains in the field of limb lengthening and deformity reconstruction, patients and their families must endure months of pin site maintenance and the ever-present external fixation. The resulting soft tissue scarring can lead to further contracture and difficulty with mobilizing the adjacent joint(s). Additionally, it takes years and scores of cases for surgeons to learn and then become experts in Ilizarov's methods. Even after years of experience with bone regeneration, surgeons commonly confront yet another new complication arising from the treatment [5].

Intramedullary fixation to supplement external fixation during limb lengthening has lead to a new generation of limb lengthening technology, described in a separate chapter as "hybrid lengthening". These techniques include lengthening over nail and plate assisted lengthening, each of which reduces the time spent in external fixation [15–17].

The "Holy Grail" of limb lengthening could be considered an intramedullary nail capable of bone fragment stabilization, gradual deformity correction, and lengthening, obviating the need for external fixation altogether. Such a device is not currently available since intramedullary nails are only capable of bone stabilization and gradual lengthening or shortening. The correction of angulation, translation, and rotation must be done acutely by the surgeon at the initial surgery, limiting its applicability due to potential neurovascular compromise and damage to potential bone regeneration.

A Brief History of Internal Limb Lengtheners

Bliskunov described an internal lengthening device that consisted of an intramedullary femoral rod bolted to the iliac wing and lengthened by a ratchet mechanism [18]. Cole introduced a device, the intramedullary skeletal kinetic distractor (ISKD), that is actuated by the inherent rotational characteristics of gait [19]. The Albizzia nail, introduced in France by Guichet, also has mechanical actuation [20]. The Albizzia and ISKD are still in use, but require the patient to perform intermittent axial rotation of the limb to effect distraction. These modalities perform well, albeit rate control difficulty is reported with each [21, 22] (Figs. 9.1 and 9.2).

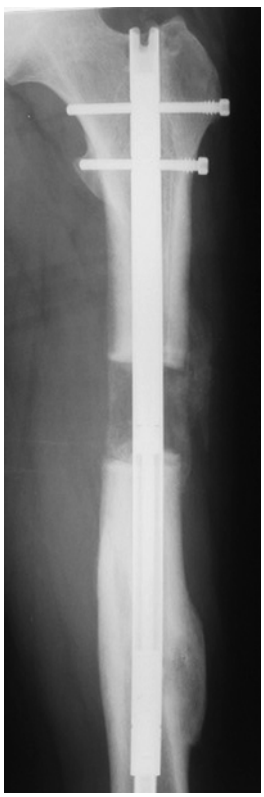


Fig. 9.1 The ISKD was found to be most useful for posttraumatic femoral length discrepancies

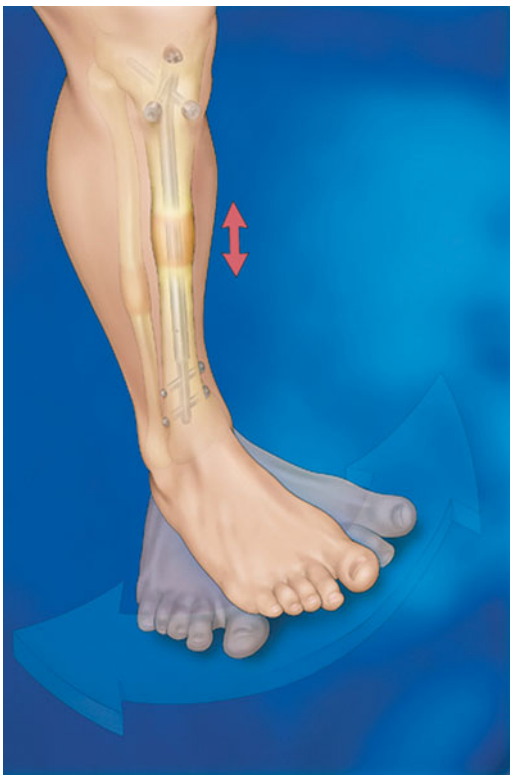


Fig. 9.2 The ISKD and Albizzia nails are activated by manual limb rotation



Fig. 9.3 The FITONE nail has an attached cable and antenna. The transformer box shown converts electric current to radiofrequency waves, transmitting energy transcutaneously

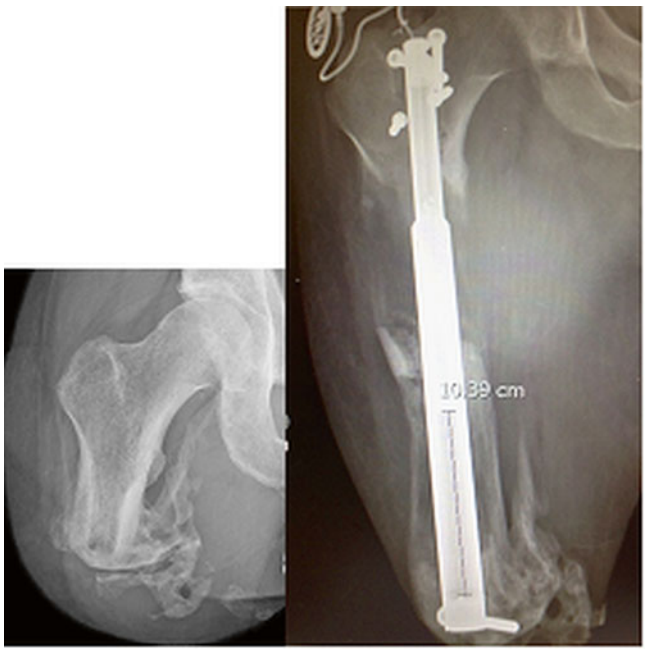


Fig. 9.4 (**a**, **b**) The FITBONE stump lengthener was used to lengthen a traumatic above knee amputation for a total of 18 cm, allowing the patient to be fitted with prosthesis and walk without assistive devices

A new era has begun with the introduction of intramedullary lengthening devices whose distraction is controlled by an external power source which causes an internal actuator to effect the desired amount of distraction. Baumgart and Betz first introduced such a device in 1992 [8]. Baumgart further developed specific insertion tools for the FITBONE nail (Wittenstein Intens, Igersheim, Germany), and published a novel planning method in 2009 [7]. The FITBONE device employs an electric motor imbedded in the telescopic rod, which is activated by intermittent transcutaneous transmission of radiofrequency waves to a subcutaneous receiver that converts these waves into an electrical impulse discharged via a connecting cable (Fig. 9.3). Distraction only occurs when the transducer is placed directly over the receiver, allowing precise control of distraction rate and rhythm. This device has since undergone multiple improvements, and the current generation has been used in over 2,000 cases worldwide. Models suitable for antegrade, trochanteric, and retrograde femoral use are available. A tibial model is in use as well. A bone transport device and a model for short amputation stump lengthening are also available (Fig. 9.4a, b). The FITBONE device requires specific training under the direction of its inventor, Professor Rainer Baumgart or a designee. The FITBONE device is currently available in the USA only under an FDA-approved compassionate use exemption.

The PRECICE nail (Ellipse Technologies, Irvine, CA) was FDA-approved and introduced in 2012. The telescopic nail has a magnetic drive mechanism that is activated by a handheld external electromagnetic controller. Similar to FITBONE, the surgeon instructs the patient in its use, and the device is specifically programmed by the surgeon to prevent application of an improper rate. The patient or family member then performs the distraction by applying the controller directly over the internal magnet in the limb three

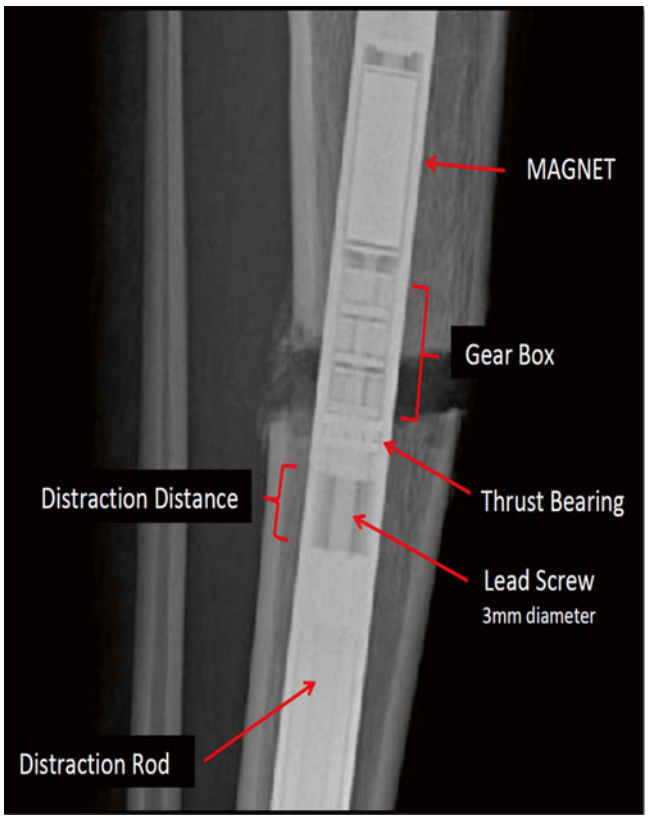


Fig. 9.5 The PRECICE mechanism has a spindled magnet attached to gear boxes which rotate against the distraction rod

to four times daily, with the rate and rhythm being adjusted according to weekly clinic visits and radiographs. This device can apply either compression or distraction, does not require a cable/antenna-receiver, and is fully FDA-approved. The manufacturer recommends specific advanced instruction for its users. Either device requires removal after complete corticalization has developed, typically 1–2 years after placement. The PRECICE device has now been used in over 800 cases worldwide (Bart Balkman, Ellipse Technologies, personal communication) (Fig. 9.5).

General Guidelines for Motorized Intramedullary Limb Lengthening

As with all limb lengthening procedures, appropriate surgeon education is essential. The properly educated surgeon then organizes a dedicated team of nurses, therapists, and orthotists to ensure the comprehensive care of these patients who often have complex medical and psychosocial issues.

Preoperative studies include a standing film of both lower extremities with block leveling of the pelvis. Foot height differences are considered to be part of the tibial discrepancy. A standing lateral film of the femur or tibia is necessary, inspecting for obvious angulation as well as the more subtle variation in the canal thickness often observed in longstanding deformities. All X-rays are made with a magnification marker so that the segment length and canal diameter can be accurately measured.

The Reverse Planning Method

The Reverse Planning Method described by Baumgart begins with defining the final ideal correction accounting for length, angulation, and translation [7]. This method can be used with any approach to the femur or tibia, but is particularly necessary for femoral lengthening, as intramedullary lengthening along the anatomic axis of the femur will cause medial translation of the knee and lateralization of the mechanical load to the knee. This coronal plane translation has been estimated at 1 mm per centimeter of lengthening, but depends on the magnitude of the preoperative deformity, the location of the corticotomy, the neck-shaft angle, and the length to be achieved [23]. Deformity correction with an intramedullary nail must then occur acutely intraoperatively, anticipating the future position of the knee. Intramedullary lengthening of the tibia does not typically lead to similar translation problems since the tibial anatomic axis normally approximates its mechanical axis.

It is important to execute the following steps with precision in the preoperative planning process (Fig. 9.6a–g).

Step 0

Make a full-size tracing of the radiograph of the entire lower limb, marking reference points, angles, and lines. Once the bone to be corrected and lengthened is determined by the malalignment test [24], consider its curvature in both the coronal and sagittal planes. If a straight track for the nail cannot be achieved, an alternate corticotomy site or a second osteotomy should be considered. If the medial proximal tibial angle (MPTA) is normal (85–90°), continue the tibial mechanical axis line to a point well above the current femoral head location (see Fig. 9.6a).

Step 1

Draw and extend the mechanical axis of the tibia proximally with sufficient length to account for the lengthening goal (see Fig. 9.6b).

Step 2

Draw the location of the femoral head in its final position after lengthening along the mechanical axis has occurred (see

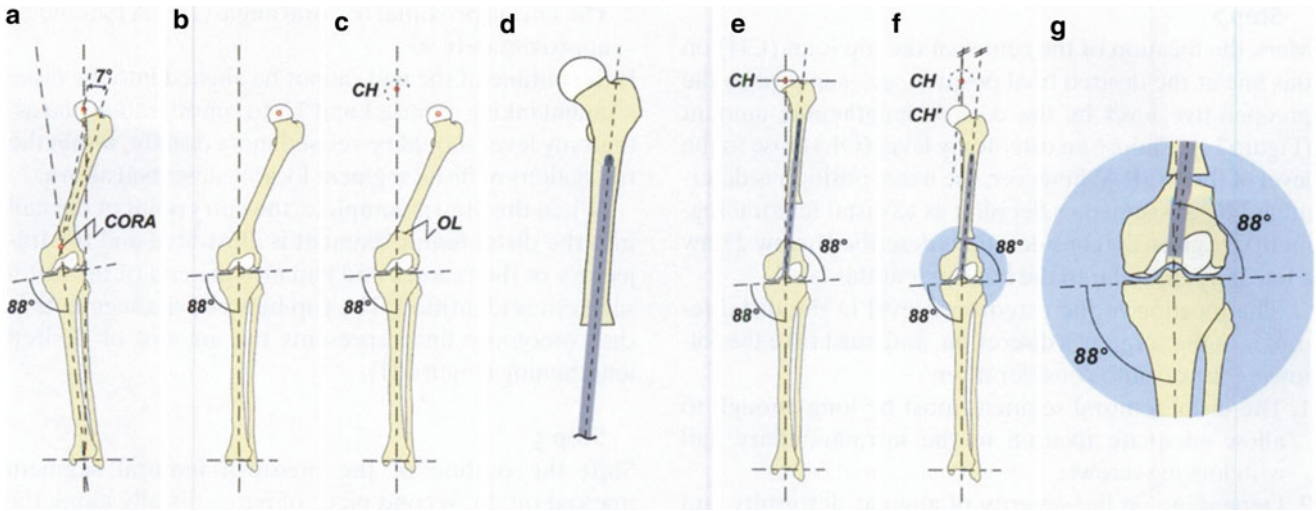


Fig. 9.6 (**a**–**g**) Reverse Planning Method for retrograde nailing of the femur

Fig. 9.6c). Choose a corticotomy level near the deformity apex, but of sufficient distance from the knee joint to allow for both locking and blocking screw insertion (generally 7–10 cm from the knee joint line). Depending on the severity of the deformity and the magnitude of the planned lengthening, selection of a corticotomy site too distant from the knee may result in excess cortical reaming and undesirable conditions for healing.

Step 3

A second tracing is made outlining the proximal femur to the level of the corticotomy (see Fig. 9.6d). The anatomic axis is then defined and drawn, and the nail is outlined to scale. In this step, the surgeon must consider the effect of reaming in both planes, being certain that cortical integrity is maintained to accommodate the size of implant and characteristics of the bone.

Step 4

After overlapping the second tracing on the first, and placing the femoral head in its future location, the papers are then swiveled until the anatomic axis lines are matched. The nail tracing should enter the distal fragment in a position of anatomic harmony, and the mechanical lateral distal femoral angle (LDFAm) should approximate 90° (see Fig. 9.6e).

Step 5

Shift the outline of the second drawing distally along the anatomic axis line to the point where contact is made between the corticotomy ends (see Fig. 9.6f). This image of the bone segment and nail position, including angulation and translation, now becomes the intraoperative goal (see Fig. 9.6g). It will replicate the desired intraoperative position with regard to sagittal translation and angulation, and should allow the exact nail position to be duplicated. If coronal deformity exists, the corticomy can be manipulated directly with an osteotome and/or indirectly with Shanz pins and sterile bumps.

Visual Aids

Creating visual aids for surgery can be helpful in many ways. The preoperative plan, printed full size, is secured to the surgical suite wall, next to the patient radiographs. Skin markings are then created. The fluoroscope is brought into the field in the AP projection. The hip, knee, and ankle reference points and lines are marked on the overlying skin with a sterile marking pen. The anatomic and mechanical axes are marked on the bone to be treated. The patella and infrapatellar ligament are marked since they are used to maintain consistent AP positioning. The proposed corticotomy site is marked (Fig. 9.7).

The selected nail is opened and the proximal, distal, and telescopic portions are indicated with skin clips to identify the desired reaming depth. Anatomic hazards to be avoided, such as growth plates, are also marked. These marks are then used for visually referencing Shanz pin insertion and reaming paths, and for assisting the surgeon in reducing X-ray exposure (Fig. 9.8).

Venting the Canal

Reaming a closed long bone causes elevation of intramedullary pressure and intravasation of marrow contents, with the potential for creation of fat embolism. This complication is



Fig. 9.7 Sterile markings are made on the surface of the skin to aid canal preparation

best avoided by venting the canal to decrease the marrow pressure [25]. The planned corticotomy site is routinely selected as the venting site. This site is drilled (vented) in percutaneous or open fashion using a subperiosteal approach prior to instrumenting the canal. A series of 4–5 mm drill holes allow intramedullary reamings to spill at the corticotomy site, also adding biological support to the lengthening site (Fig. 9.9a, b).

The actual corticotomy will be performed at a later point in time after the canal preparation.

Shanz Pins

Shanz pins are inserted with c-arm guidance in the lateral position. The femoral condyles are rotated so as to overlap each other on a lateral view, and the pin is inserted from lateral to medial, just inside the posterior cortex at the junction of the metaphysis and epiphysis, and out of the way of the future passage of the nail (Fig. 9.10).



Fig. 9.8 Reaming depth can be guided by applying skin clips at the appropriate levels

Entry Points

Ideal entry points are required for proper intramedullary nailing and are defined by several authors [26, 27] (Fig. 9.11a, b).

The correct starting point for trochanteric entry is actually on the further-most point medial on the tip of the trochanter without entering the piriformis fossa.

Reaming Techniques

As the telescopic nails are straight and the canal is not, one must either over-ream a curved canal to accommodate the straight nail, or 'fit' rigid reaming to match the straight nail. After the canal has been vented and the Shanz pins placed, reaming can proceed with attention to appropriate entry points suitable to the approach chosen [26, 27].

Conventional Reaming

The canal is first vented. A bulb tipped guide wire is placed. Flexible reamers prepare a cylindrical path but curve at multiple apices, thus requiring over-reaming of 2 mm to accommodate a straight nail. Corticotomy is generally performed after this "over-reaming" with flexible reamers.

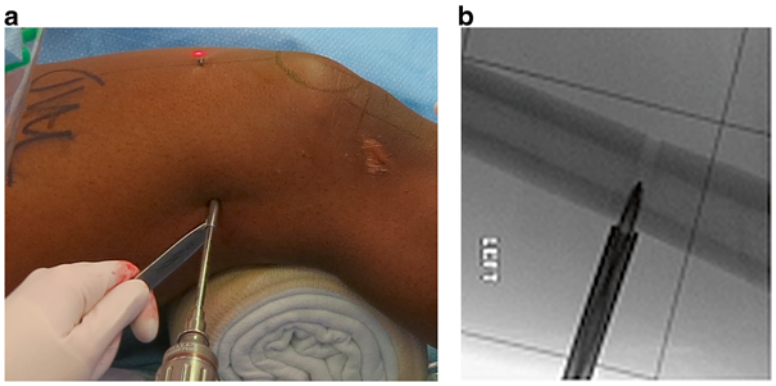


Fig. 9.9 (**a**) Venting the canal at the level of the future corticotomy is done with a series of drill holes, thus creating a subperiosteal pocket into which reamings will spill. (**b**) A 5-mm drill bit with sleeve serves this purpose



Fig. 9.10 The knee joint skin mark assists parallel Shanz pin insertion without switching c-arm position. The proximal pin is placed rotationally parallel to the distal pin, at the level of the lesser trochanter, perpendicular to the shaft, and posterior to the future nail path

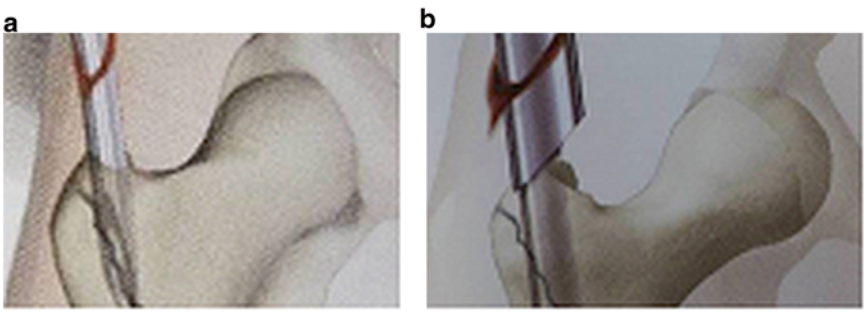


Fig. 9.11 (**a**) Trochanteric entry is at the medial-most tip of the trochanter. (**b**) Reaming sleeves protect soft tissue

Rigid Reaming

The canal is first vented. Canal preparation with rigid reamers creates a well-defined pathway for the straight nail with a future line-to-line fit. Blunt or sharp rigid reamers can shape curved canals with asymmetric wall thickness to accept the nail (Fig. 9.12a, b).

Reaming sleeves passed over insertion dilators protect soft tissues from the reamer blades and prevent spillage of marrow contents into the joint or soft tissues.

Rigid reamers are inserted within the tubes along the planned pathway, and are increased in 0.5 mm increments to 0.5 mm greater than the nail diameter. After canal preparation on the "near side" of the corticotomy, the corticotomy is completed with an osteotome and the deformity is corrected to match the plan in step 5. With the plan achieved, reaming then proceeds on the "far side" of the corticotomy to the 0.5 mm greater than the nail diameter.

Regardless of reaming technique, the nail is inserted with simple hand pressure, reaming slightly more as necessary to

avoid mallet use, and potential damage to the micromotor. Alignment is checked, the corticotomy is compressed, and locking screws are inserted.

Blocking Screws

Blocking screws (Poller) are known to be useful in trauma and reconstruction applications of intramedullary fixation for correcting and preventing deformity [28, 29]. They function to narrow a canal in epiphyseal and metaphyseal areas, and thus guide nails in juxtaarticular applications (Fig. 9.13 a–c).

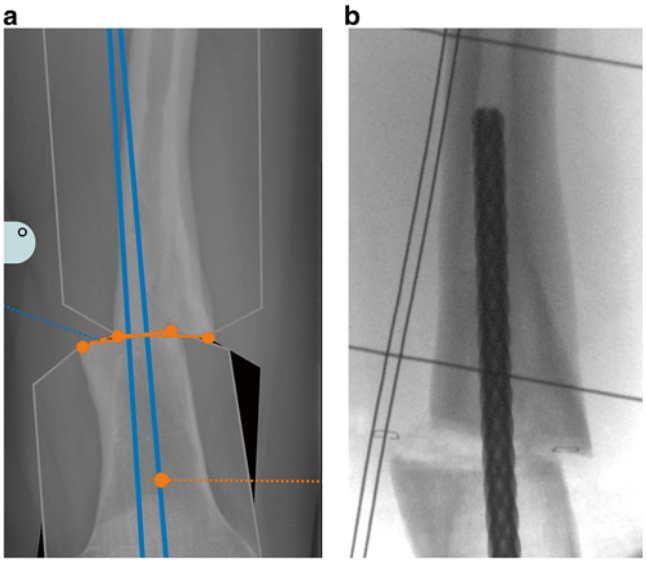


Fig. 9.12 (**a**) Carefully inspecting the canal preoperatively determines if the canal will accept an intramedullary nail. (**b**) In specific instances rigid reamers may be used to "shape" the inner canal

We have learned from our bone lengthening experiences that deformities developing during lengthening are a common event. Asymmetric muscle forces imposed by unequal muscle mass, muscle anatomy, and two-joint muscles create the predictable deformities of varus-procurvatum during femoral lengthening and valgus-procurvatum during tibial lengthening. The development of such deformities can be corrected by adjustments of the fixator in the outpatient clinic when lengthening with circular external fixation. When lengthening with intramedullary techniques, these deformities must be anticipated and prevented with the appropriate use of blocking screws, as later correction requires additional surgery.

Antegrade Femoral Lengthening

Patient Indications

Antegrade femoral lengthening is indicated for patients with leg length discrepancy originating from the femur. Angular and rotational deformity of the proximal to mid femur can be corrected acutely, with the latency period suitably delayed to account for the correction magnitude. A piriformis entry can be used for adults. A trochanteric entry has been shown to be safe for preadolescents as young as age 12 years [30] (Fig. 9.14a, b).

Lengthening along the anatomic femoral axis will result in 1 mm of knee medialization for every 10 mm lengthened, potentially resulting in an undesirable transfer of load laterally [18]. Assessment of the weight-bearing line on preoperative full-length AP X-rays will be helpful in determining whether this amount of medial translation will be harmful to the knee in the future. Acute deformity correction can be assisted by temporary intraoperative placement of Shanz

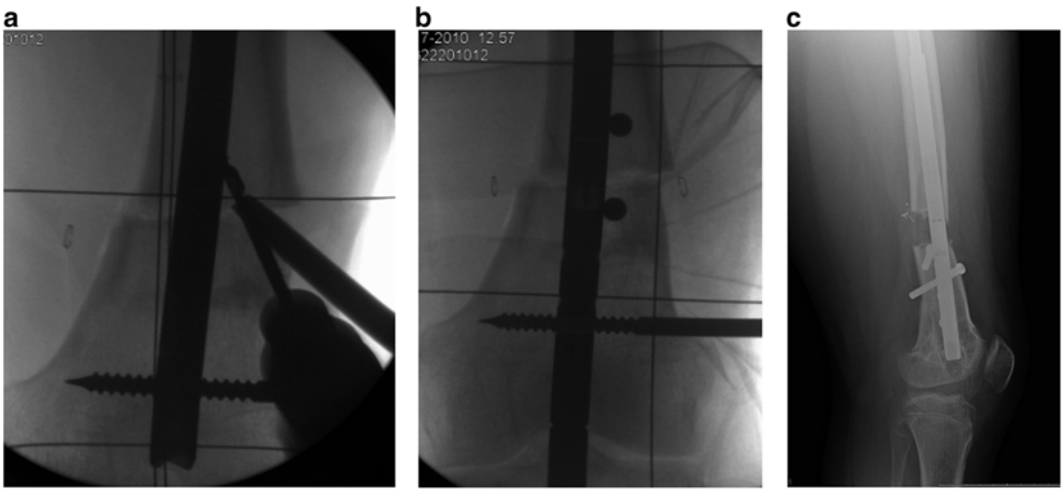


Fig. 9.13 (**a**) Blocking screws are placed against the nail. (**b**) Blocking screw location is specific to each case, generally on the "concave" side of potential deformity. (**c**) A single posterior screw maintained this extension osteotomy

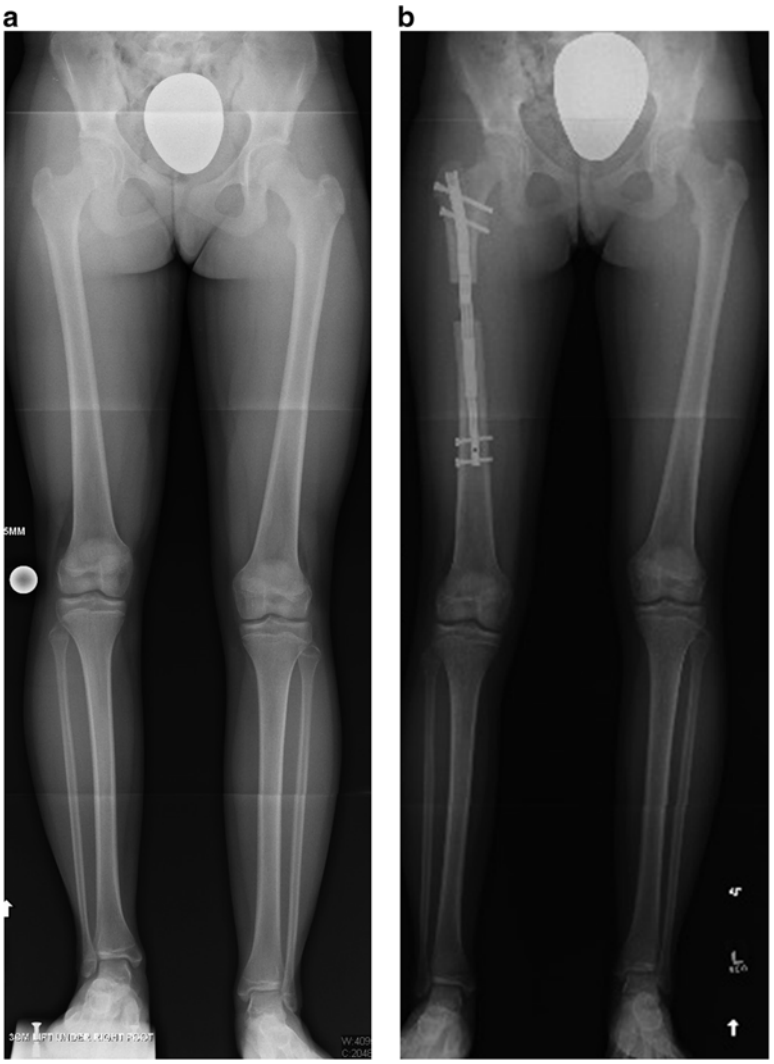


Fig. 9.14 (**a**) A 16-year-old girl with a 3.5-cm idiopathic right femoral shortening with preoperative weight bearing line 2 mm medial to the medial spine, suggests an antegrade lengthening will result in lateralization of 3.5 mm. (**b**) A trochanteric entry best avoids the hip vasculature, and resulted in the predicted and acceptable weight bearing line 1 mm lateral

pins, and progressive deformity during lengthening can be prevented by blocking screws (Fig. 9.15a–c).

Planning for Antegrade Femoral Lengthening

A graphical method of planning has been described for length and deformity correction [31]. The corticotomy should be made at the apex of the deformity in the coronal and sagittal planes, when possible.

Antegrade Femoral Technique

A radiolucent table and a bump under the ipsilateral buttock are used. Sterile draping must provide adequate exposure of the proximal buttock for nail entry. The ipsilateral arm is supported across the chest and protected during reaming. At the planned corticotomy level, a 1-cm incision is made and multiple holes are drilled in a transverse fashion, venting the canal. Shanz pins are placed in the proximal and distal segments away from the nail tract to mark rotation and control the bone segments.

Careful preoperative evaluation of individual hip anatomy allows determination of the ideal piriformis or trochanteric entry. For example, a proximal femoral flexion deformity requires a more posterior entry point. Acquired hip deformities such as anterior displacement of the greater trochanter can malign the entry point as well. The hip is adducted and a wire is inserted into the intramedullary canal through the piriformis fossa or medial tip of the greater trochanter. A 2-cm incision is made over the wire, and a soft-tissue protector is inserted. The intramedullary canal is opened with a cannulated drill, and then reaming is performed by either of the previously described methods. The assembled nail is inserted upto the corticotomy site. An osteotome is used to finish the corticotomy, with completeness confirmed by fluoroscopy. Deformity correction is completed directly by osteotome

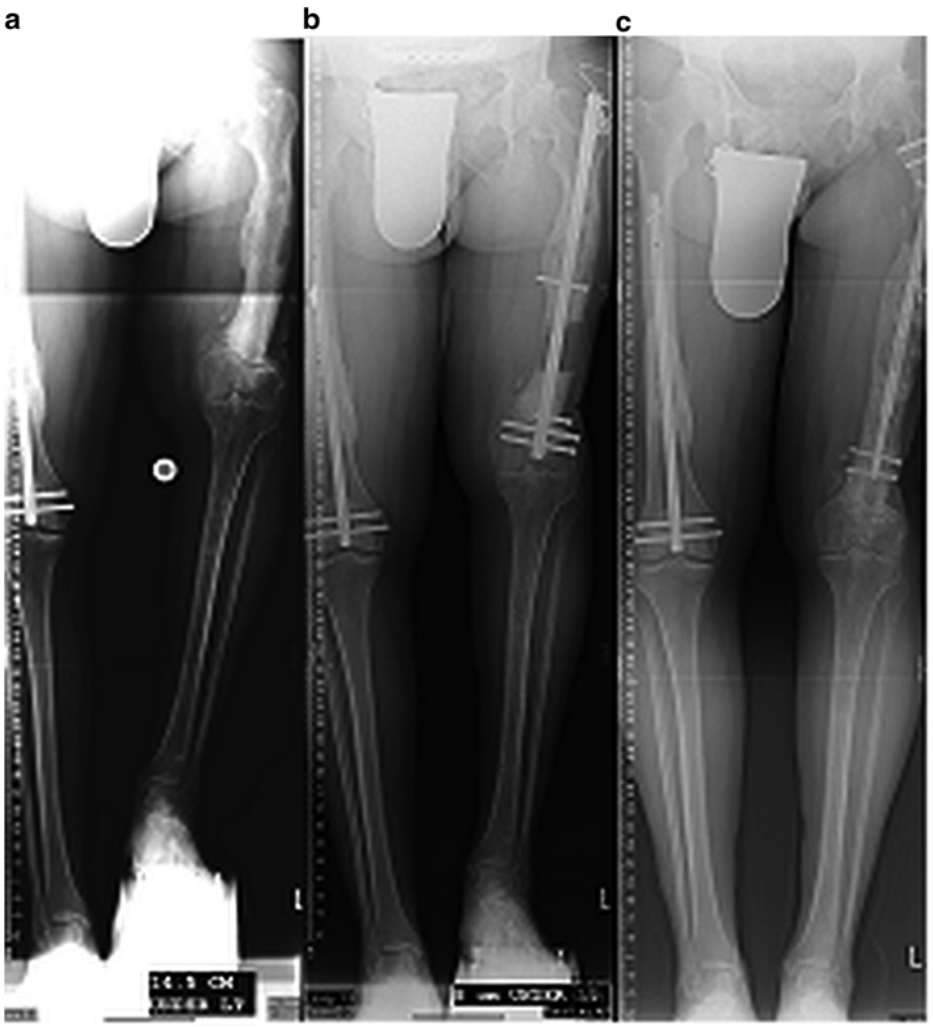


Fig. 9.15 (**a**) Post traumatic knee ankylosis makes an antegrade approach to this femoral lengthening necessary. (**b**) Childhood Perthes altered proximal femoral anatomy; making piriformis entry more suitable for the first femoral lengthening of 8 cm. (**c**) The second lengthening was similarly approached through the piriformis fossa

Box 9.1. General Guidelines Summary

1. Blocking screws should be employed to prevent deformity at insertion or during lengthening. They are less likely needed if the deformity and corticotomy apex is at the isthmus, and/or the lengthening is small, less than 3 cm.
2. Blocking screws are most commonly needed in the distal femur and proximal tibia, as these locations often have large canal-to-nail size ratios.
3. Postoperative monitoring is done weekly during distraction and monthly during consolidation.
4. Preoperatively plan each case with life-size drawings or computer graphic simulation. The Reverse Planning Method is most useful in retrograde femoral applications.
5. Nail length is planned to allow 6–7 cm of the wider portion of the nail in the distracted segment at the completion of distraction.
6. The canal must be vented before reaming.
7. Isthmic bone diameter must be able to accommodate reaming 2 mm greater than the nail diameter for flexible reaming, and 0.5 mm for rigid reaming.
8. Shanz pins control segments and prevent rotational deformity, particularly in femoral lengthenings where the corticotomy is not well visualized through the wound.
9. Periosteum-sparring, non-heat-generating corticotomy should be positioned at or near apex of the deformity.
10. Comminution of the osteotomy should be avoided, as this creates nail instability, may prevent acute correction, encourages lengthening deformity, and may lead to poor bone regenerate.
11. The nail should be advanced by hand pressure only in order to avoid damage of the distraction motor and gear boxes.

manipulation of the osteotomy site or indirectly by bumps and a surgical assistant. The corrected position of the osteotomy may be secured with a femoral distractor or sterile bumps. The nail is passed across the osteotomy site correcting any residual deformity. Rotation is optimally set by rotating the Shanz pins into their pre-osteotomy position, and the proximal interlocking screws are inserted via the targeting jig. The corticotomy is manually compressed. Proper alignment is checked and adjusted using an alignment grid, long alignment rod, or Bovie cord. Distal locking screws are inserted using the "perfect-circle" freehand technique. Iliotibial band release is performed for lengthening of greater than 3 cm and in cases of congenital origin.

Retrograde Femoral Lengthening

Patient Indications

Retrograde femoral lengthening is indicated for skeletally mature patients with the any of the following:

1. Where antegrade femoral lengthening will result in excessive axis deviation or translation
2. Hip arthrodesis or deformity of the proximal femur preventing antegrade femoral nailing
3. Distal femoral deformity suitable for acute correction and retrograde nailing (Fig. 9.16a, b).

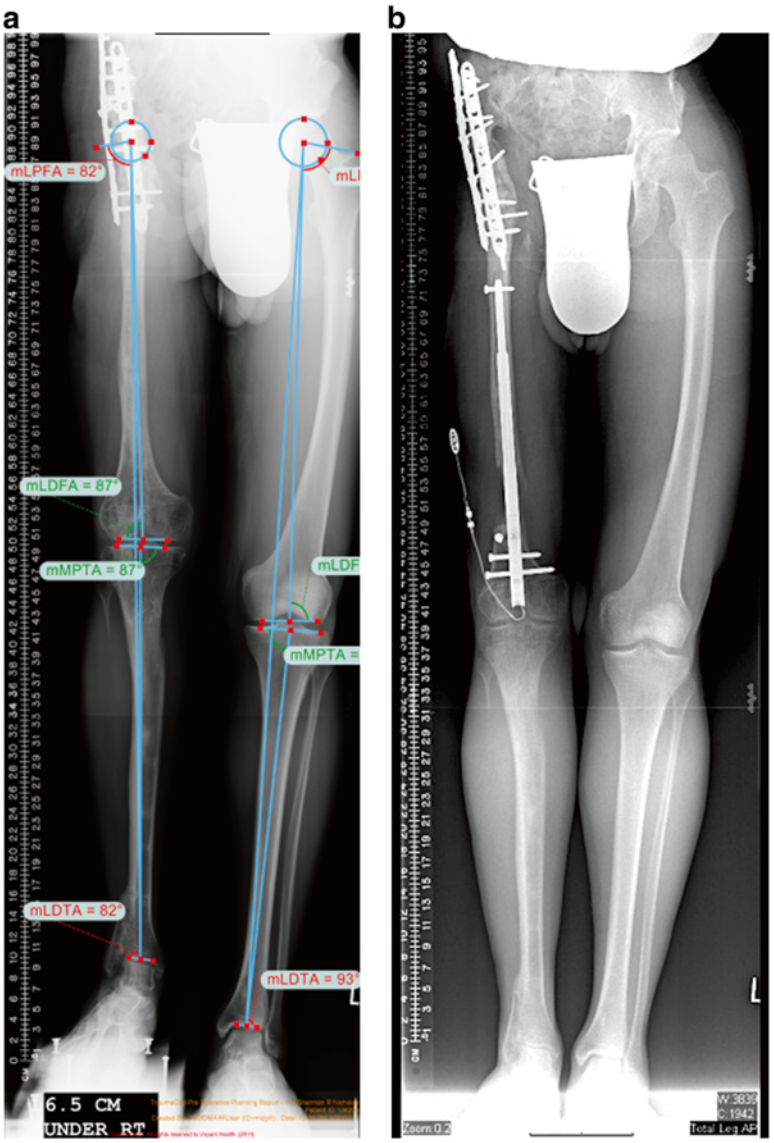


Fig. 9.16 (**a**) Hip arthrodesis was performed after proximal femoral osteosarcoma resection. (**b**) A retrograde femoral lengthening was the necessary approach for the 6.5-cm limb length discrepancy

A retrograde femoral approach is also indicated in skeletally immature patients with premature arrest of the distal femoral physis, provided canal size and bone length is sufficient for straight nail insertion. This technique requires entry through and protection of the knee joint as described by Watson and others [26, 27].

Planning

Retrograde femoral lengthening is routinely planned with the Reverse Planning Method as described by Baumgart [7]. This method of planning can be performed with X-ray tracings of the preoperative full-length standing radiographs, or with commercially available software systems. In either case, it is recommended that the surgeon reproduce the actual size of the entire limb using preoperative, intraoperative, and completed correction X-rays for intraoperative use.



Fig. 9.17 The skin incision for retrograde femoral lengthening is transverse or longitudinal, with a longitudinal incision through the ligament

Retrograde Femoral Surgical Technique

A radiographic grid is placed under the patient, and the hip position is centered over the grid marker. The orientation of the distal joint line, patella, and the anatomic and mechanical femoral axes are marked with sterile skin markers. The corticotomy level, future nail position, and junction of the telescopic portion of the nail are marked with skin clips to guide reaming depth and direction.

The femoral canal is vented through the future corticotomy site by drilling multiple holes through a 10-mm lateral incision. Two Shanz pins are placed, one distal and another proximal to the corticotomy site, in a lateral to medial direction and rotationally parallel.

The infrapatellar ligament is split longitudinally through a 10 mm infrapatellar incision, and a 3 mm guide wire is inserted to a point in the intercondylar notch anterior to the PCL origin, and directed along the path that will allow for reaming of the distal bone segment as planned (Figs. 9.17 and 9.18a–d).

Biplane fluoroscopy guidance for determining the exact entry point avoids hyaline cartilage injury. The articular surfaces are protected from reamers and reamings with a protective sleeve so that insertion, removal and exchange of reamers never touch the articular surfaces, and never actually enter the joint.

Reaming just to the corticotomy level in 0.5-mm increments is performed. The corticotomy is then completed with an osteotome at the venting site. Drilling six to eight 5-mm holes should be adequate to avoid bone spikes at the corticotomy since they may prevent acute deformity correction. The osteotomy site is then angulated, translated, and/or rotated to the preoperatively planned position and held there by a femoral distractor or an assistant. Reaming of the diaphysis is performed to 0.5 mm greater than the diameter of the nail. The nail is inserted with hand pressure, and alignment is checked with the grid. Additional reaming is performed if

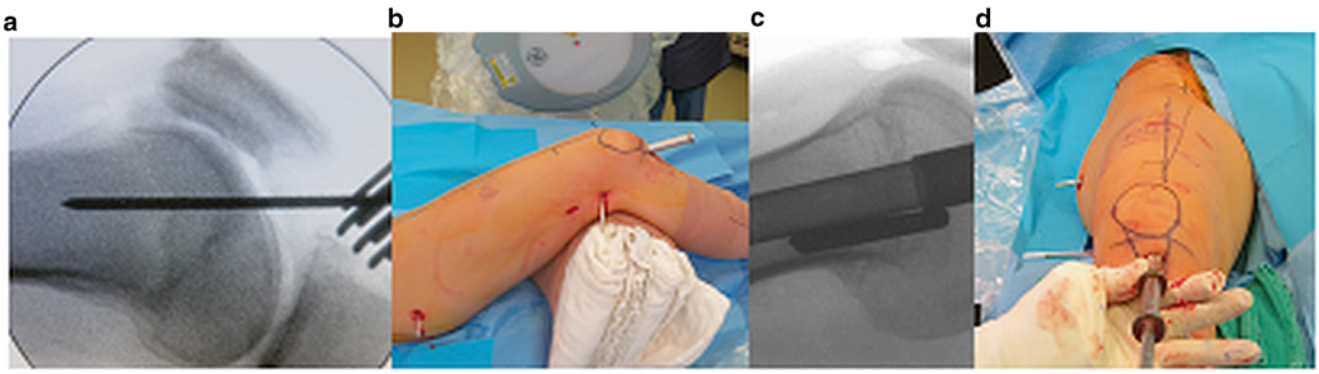


Fig. 9.18 (**a**) A 3-mm guide wire is inserted to the center of the notch, just anterior to the PCL origin and posterior to the articular surface. (**b–d**) Reaming sleeves are inserted, protecting the joint and ligament from reamings and reamer damage

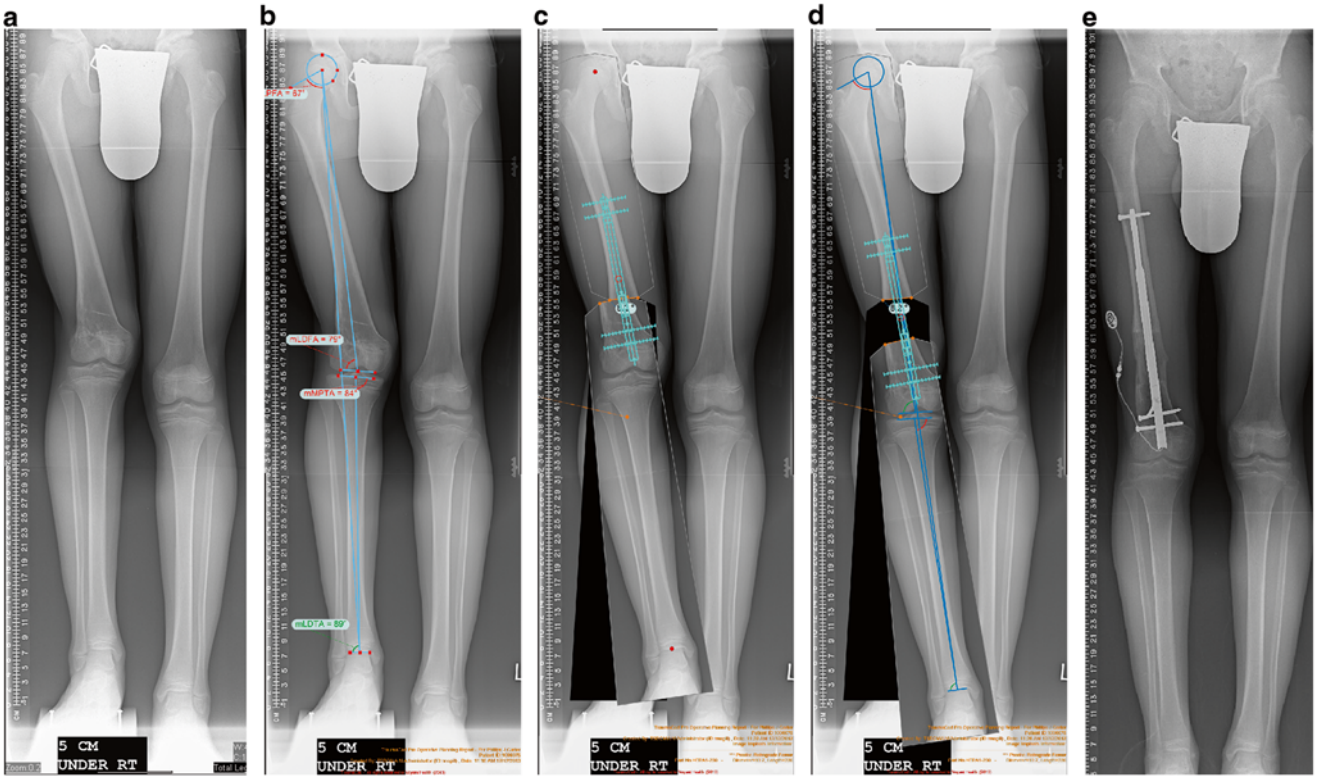


Fig. 9.19 (**a**) Distal femoral physeal closure after fracture resulted in valgus and shortening. (**b**) Properative computer-assisted planning defines, weight bearing line lateral 12 mm, valgus mechanical distal lateral femoral angle 79°, and axial discrepancy of 5 cm. (**c**) Retrograde nail size, position, and translational correction determined by Baumgart's Reverse Planning Method. (**d**) Mechanical axis and weight bearing line and length predicted to be restored to normal after length achieved. (**e**) Full length radiograph demonstrates ideal correction and achievement of goals

the nail does not pass easily. Adjustments in alignment are made if needed and locking and blocking screws are added.

This 14-year-old boy had premature asymmetric distal femoral physeal arrest of the right side, resulting in 9° of valgus and 5 cm of shortening. Preoperative full-length standing X-ray with a lift under the short limb (b) computer-assisted planning (c) corticotomy with intraoperative correction (d) end of lengthening with neutral mechanical axis (e) end result with ideal alignment and matching length (Fig. 9.19a–e).

Tibial Lengthening

Patient Indications

Candidates for tibial lengthening with a telescopic nail are skeletally mature patients with a tibial length discrepancy of 2–6 cm, modest deformity thought to be safely correctable acutely, and an open medullary canal with adequate diameter and length to accommodate the nail.

Planning

Tibial anatomic and mechanical axes are collinear in the undeformed tibia so that planning for translation is unnecessary. A modest tibial deformity can be acutely corrected to allow passage of a telescopic nail, provided that the apex of the deformity (level of the correction) is amenable to stable fixation, and the surrounding neurovascular structures can safely tolerate the acute correction. For example, proximal tibial valgus deformities are at greater risk for peroneal nerve palsy than are proximal tibial varus deformities. Blocking screws are inserted during canal preparation for corrections in the proximal third of the tibia since valgus and procurvatum may develop during lengthening. Corrections in the lower half of the tibia where the bone is wide may not have adequate distal purchase at the end of lengthening, and may also require enhanced fixation with blocking screws. These features limit acute corrections to the region of the junction of the proximal and middle thirds of the tibia, and sometimes to the diaphysis.

Tibial Surgical Technique

Position the patient supine on a radiolucent table with the image intensifier on the opposite side. Sterilely prepare the entire limb, including the foot, leg, thigh, and hip. Perform a fibular osteotomy at the junction of the middle and distal thirds through the internervous plane between peroneal and soleus muscles. Insert a 4.5-mm fully threaded cortical screw distally from the fibula to the tibia with the foot in dorsiflexion. Proximal fibular attachment to the tibia can be accomplished in selected cases using the Rancho technique [32].

A prophylactic anterior compartment fasciotomy is performed in patients at risk for developing a compartment syndrome, such as those with substantial deformity corrections, previous neurovascular compromise, or those anticipating greater lengthening goals (greater than 3 cm). Perform a gastrocsoleus recession if there is existing equinus contracture, or for lengthenings more than a few centimeters or with congenital etiology. Further protection against equinus can be achieved by static or dynamic dorsiflexion bracing or extraarticular pinning as described by Herzenberg [33].

Use an infrapatellar incision, and longitudinally split the tendon or approach the tibia medial to the tendon. Obtain a starting point in the center of the intraarticular area under biplanar image intensifier control.

Choose the tibial corticotomy level at the deformity apex, at least 8 cm from the proximal joint line, thereby allowing room for both locking and blocking screws. Plan the nail length such that there will be 6–7 cm of the wider portion of the nail in the distal segment after distraction is complete [33]. The corticotomy is performed through a 2–3 cm vertical incision, with a tiny elevator lifting the medial and lateral periosteum. Multiple 5 mm drill holes will vent the canal.

Shanz pins are placed rotationally parallel to each other, yet parallel to the adjacent joints in the coronal plane. The tourniquet, if used, must be deflated before entering the canal and reaming. Ream in 0.5-mm increments with either rigid or flexible reamers, and do so with soft tissue protection and a flexed knee supported on a padded triangle. Posterior and medial blocking screws are placed at this time, guiding the reamers along a path to prevent a valgus and procurvatum deformity.

The corticotomy is completed with an osteotome, the knee is flexed, and the nail inserted by hand pressure. The proximal interlock jig will require removal to confirm alignment, as the knee will not fully straighten with the jig in place. When satisfied with alignment, insert the proximal interlocking bolts first. Manually compress the corticotomy. "Perfect circle" free-hand insertion of the distal interlocks is now completed.

Deformity correction can be achieved with tibial Shanz pins placed posterior proximally and distally, with each pin oriented parallel to the adjacent joint. Fixator assisted nailing techniques can be used [23]. Blocking screws should be liberally used to prevent intraoperative deformity and prevent deformity from developing during lengthening (Fig. 9.20a–e).

Intraoperative Nail Testing for All Limb Lengthening Techniques

All nails are tested intraoperatively, before closure, to ensure mechanical function and that subsequent distraction at the osteotomy site will occur postoperatively.

Post-operative Management

Encourage isometric exercises and active range of motion of adjacent joints throughout the lengthening process. Use a cam walker boot to support the foot and ankle in neutral position in tibial lengthenings. Use a knee immobilizer to support full knee extension during femoral lengthenings. Restrict weight bearing to 30 lb pressure maximum until three cortices are evident on X-rays (generally 3–4 months). Oral opiates should be adequate for pain control. Avoid nonsteroidal anti-inflammatory drugs (NSAIDs) beyond the first few days postoperatively, as these agents can slow regenerate bone formation. Consider 2–6 weeks of thromboembolism prophylaxis appropriate to the patient's risk factors. Begin physical therapy, stressing ankle dorsiflexion and knee extension for tibial lengthenings, and knee and hip extension for femoral lengthening. Weekly postoperative physical therapy visits are advised to emphasize the need for continued therapy in preventing contractures in all patients, but most importantly in those with congenital etiologies (Fig. 9.21).

Recommend calcium and vitamin D supplements, and a diet high in protein and carbohydrates, and low in fat. The importance of the latency period must be considered [34]. Latency is commensurate with the patient's age, health, bone being treated, bone quality, location of the corticotomy, and magnitude of the acute correction performed. Generally, the latency period is slightly longer in intraoperatively performed corrections than with gradual corrections, 5–7 days in a child's femur, and 7–10 days in a child's tibia. Adults require latency of 7–10 days in the femur and 10 days in the tibia. Acute rotational and translational corrections require an additional 2 days of latency.

The rate and rhythm of distraction should begin at 0.25 mm every 6 h, and then adjusted based on weekly assessments of the X-ray appearance of the bone regenerate density, volume, contour, and consistency.

Physical therapy sessions and principles previously outlined must be followed [35]. The weekly follow-up visits

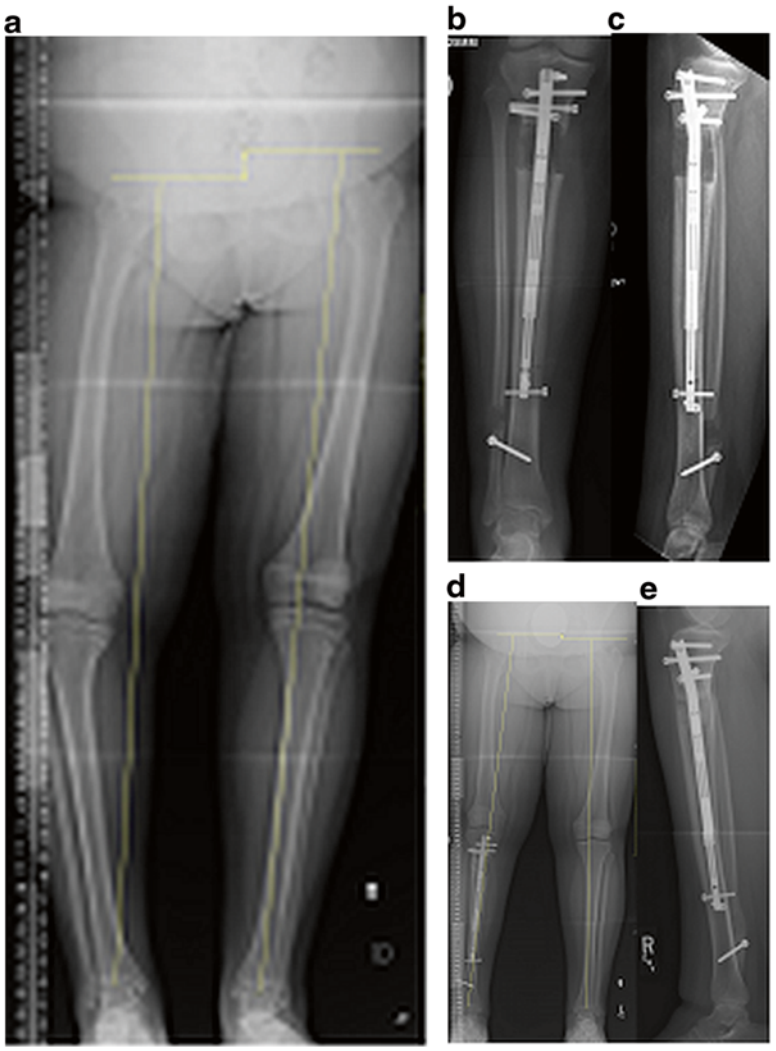


Fig. 9.20 (**a**) Adolescent Blount's with 15° varus, 10° procurvatum, 10° internal rotation, and 3 cm of shortening. (**b**, **c**) Acute interoperative varus, rotation and procurvatum correction with fixator assisted nailing, then gradual lengthening. (**d**, **e**) Deformity correction maintained throughout lengthening by proximal posterior and lateral blocking screws

during the distraction phase can be extended to monthly visits during the consolidation phase. Patients must be informed to prevent contracture, improve motion, and not to exceed the 30 lb weight-bearing restriction during this phase. Full weight-bearing is not allowed until corticalization is radiographically evident on three of four cortices. Physical therapy can be modified to strength and endurance training once three cortices are evident. Removal of the implant is performed 1 year after surgery, provided the regenerate bone is circumferentially corticallized.

Complications of Motorized Intramedullary Lengthening

Little has been written on the subject of complications of motorized limb lengthening, as the technology is so new. Green and Dahl presented a poster exhibit on the subject as it relates to all methods of lengthening at the AAOS annual meeting in 2013 [36]. The authors point out that certain complications of limb lengthening are intrinsic to any bone lengthening, and can occur regardless of the technique or device used.

Limb lengthening complications include:

1. Failures of preoperative assessment
2. Intraoperative complications
3. Incomplete corticotomy
4. Iatrogenic deformity during insertion
5. Premature consolidation
6. Failures of bone formation
7. Device failure
8. Joint contracture
9. Joint subluxation
10. Deformity during lengthening
11. Neurological consequences
12. Muscular consequences
13. Psychological aspects
14. Social/familial aspects



Fig. 9.21 Proper therapy compliance allowed the patient illustrated in Fig. 9.20 to maintain nearly full knee range during treatment

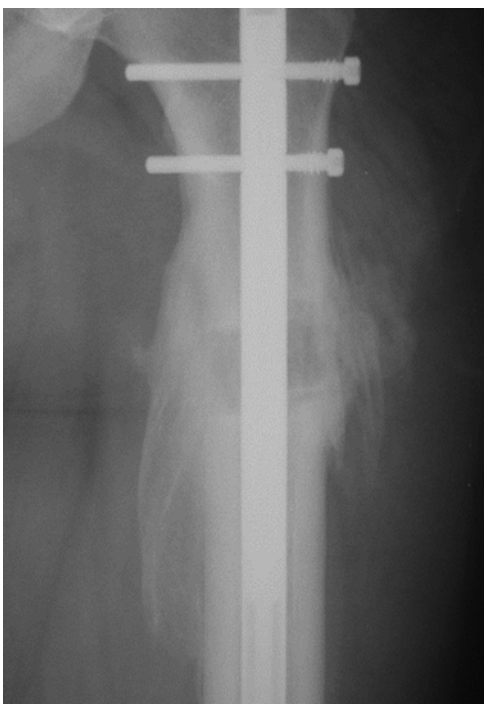


Fig. 9.22 Excessive bone formation in a patient with remote head trauma prevented distraction beyond 2 cm despite increasing the lengthening rate to 1.5 mm/day

15. Fracture after lengthening
16. Deformity after lengthening
17. Stiffness following lengthening
18. Surgical site infection

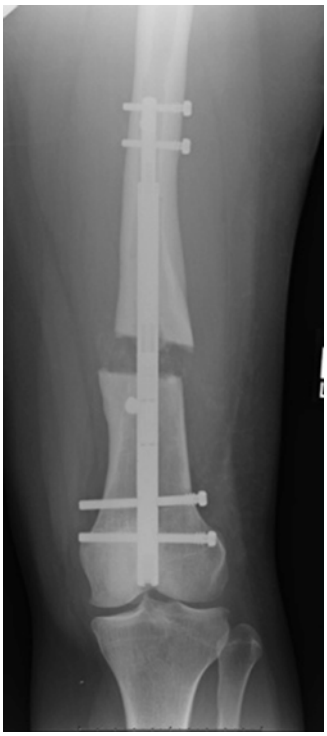


Fig. 9.23 Atrophic regenerate in a multiply operated femur requires a delay and/or slower lengthening rate until the regenerate improves

Pin and wire site infection has been eliminated altogether.

Premature consolidation is most likely to occur in the young patient with femoral lengthening without acute deformity correction. A clue to this occurrence in fixator lengthenings, extreme wire or pin site deflection on X-ray, is not evident in intramedullary lengthenings. A failure of the corticotomy to continue separation at the rate expected signals the clinician to increase the rate of distraction (Fig. 9.22).

Poor new bone formation seems to occur more commonly with intramedullary lengthenings than with fixator lengthenings, as reaming damages the marrow source of regenerate bone, and acute corrections damage the periosteal source of new bone. Atrophic bone formation should be corrected by temporarily stopping the lengthening for 3–5 days, and then resuming the lengthening at a slower rate until the regenerate improves. Delayed healing, primarily in adults, can be expected in 4 % and can be minimized by ideal corticotomy technique, a longer latency period, weekly monitoring of X-rays, and slowing the rate or temporarily stopping the lengthening in the face of poor regenerate. If the regenerate does not improve with these maneuvers, a reversal of the lengthening can be done with PRECICE device. The penalty for continued lengthening in the face of poor regenerate bone is a nonunion, which may require nail exchange and/or bone grafting (Fig. 9.23).

Fractures occurring through regenerate bone should no longer occur, as the nail protects the regenerate bone. Femur

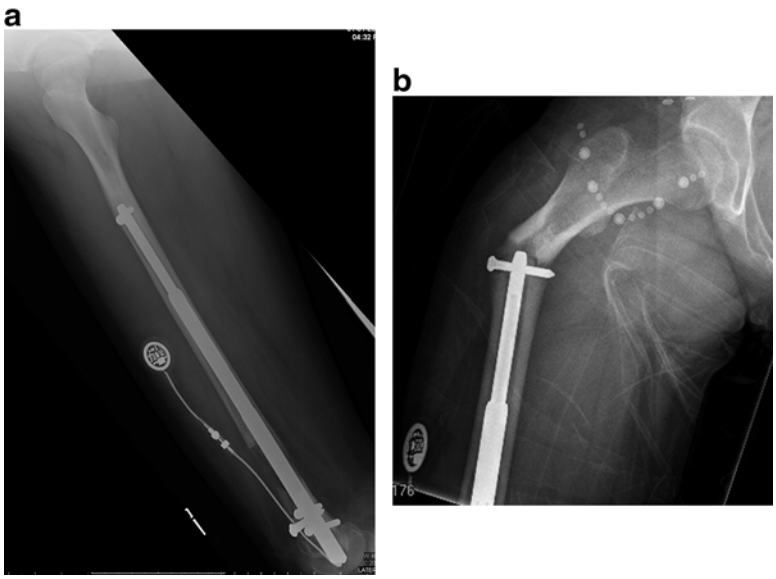


Fig. 9.24 (**a**) This 14-year-old female with congenital short femur required additional reaming, with resulting anterior cortical thinning. (**b**) After completing the lengthening a fracture developed through the proximal locking bolt at the area of maximal cortical thinning on the tension side of the femur

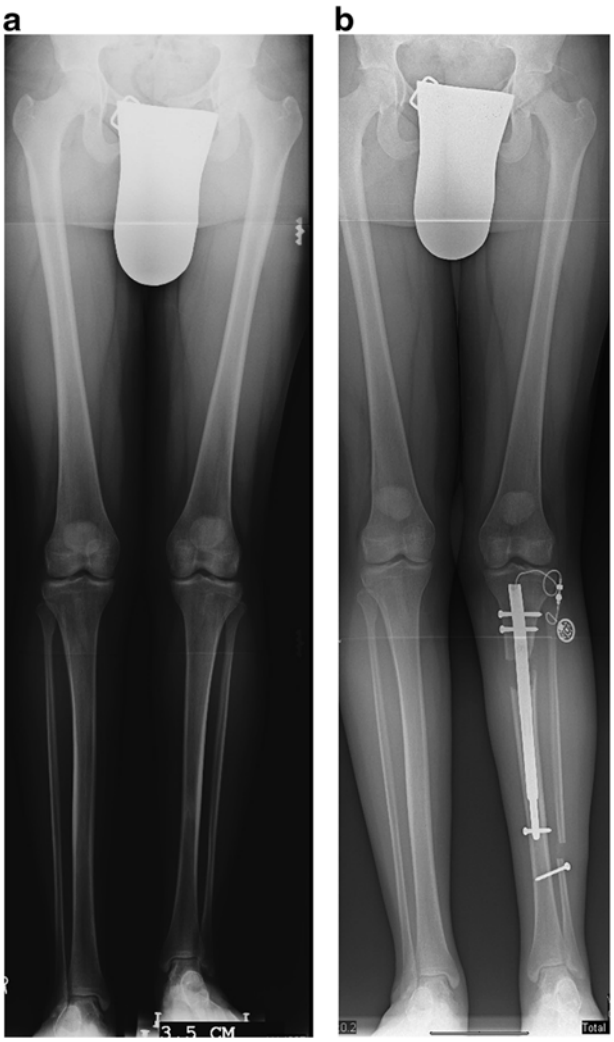


Fig. 9.25 (**a**) Congenital short tibia of 3.5 cm with normal axial alignment. (**b**) Valgus deformity developed during lengthening. Note the large space in the proximal metaphysis allowed the nail to migrate laterally during lengthening

fracture at the tip of a lengthening nail can occur when the bone is too small for the device (children), and excessive or eccentric reaming has occurred to accommodate the nail (Fig. 9.24a, b).

Deformity during distraction, a known complication of fixator lengthening, is at risk to occur in intramedullary lengthenings too. This occurs most commonly in the distal femur or the proximal tibia where the wider metaphyseal bone may allow angulation around an intramedullary nail despite adjacent locking screws. Blocking screws should be placed on the concave side of the anticipated deformity of varus/procurvatum in the femur and valgus/procurvatum in the tibia. Surgical site infection is rare, but deep infection has been reported in a patient previously having undergone prolonged external fixation in the past (Figs. 9.25a, b and 9.26).

Contracture or subluxation of the hip, knee, or ankle in congenital cases can occur, just as in external fixator lengthenings. While doing intramedullary lengthenings in congenital etiology, protection of the knee joint by spanning is not possible, requiring more strict maintenance of the ability to fully extend the knee. The joint dislocation cascade of knee flexion contracture, followed by knee translation, progressing to posterior subluxation, and then to frank dislocation must be prevented at the beginning. Prevention methods require awareness, vigilance, and modest lengthening goals, as these soft tissue complications are intrinsically related to the underlying diagnosis and host condition. Physical therapy directed at stretching, and joint specific static and dynamic bracing are of value as in any lengthening [35]. The ankle joint is protected from equinus deformity by static splinting, stretching, and in certain congenital cases, an extraarticular screw, removed after completion of the lengthening.

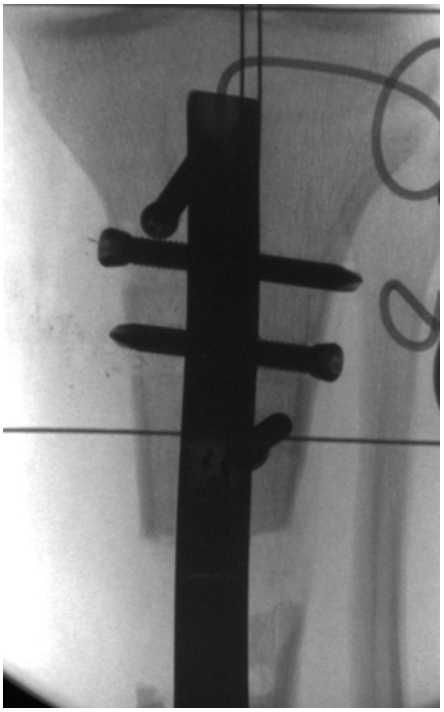


Fig. 9.26 Blocking screws serve to narrow a capacious canal, in this example, correcting proximal tibial valgus

Outcomes

Early clinical case series of the FITBONE and PRECICE devices report favorable outcomes and patient satisfaction, with relatively few implant problems [10, 11, 37]. High accuracy of distraction, maintenance of joint motion, favorable alignment, and fewer complications have been noted, compared with external fixation lengthenings [9].

Patients report significantly lower levels of pain with motorized intramedullary lengtheners, compared with external fixators [38]. Reports of mechanical device failure such as nail breakage of the FITBONE are rare (4 in 2,000 cases) (Roman Stauch, Intens, personal communication). Nail breakage of the PRECICE is reported to be 11/600 and is more common in bilateral applications, as patients with these applications have a greater problem with restricted weight-bearing compliance. Failure to distract is reported in 12/600 with the PRECICE device. Transmitter, receiver, cable, or motor failure of FITBONE is reported to be 16/2,000 (Personal communication, Bart Balkman 2014).

Just as in Ilizarov methodology, proper surgeon education and assembly of a lengthening team of nurses, therapists, and orthotists at centers of excellence cannot be overemphasized.

Summary

Critical to the success of a motorized internal lengthening nail are combining proper surgical training, accurate preoperative planning, minimally invasive surgery, mechanical integrity of the construct, and ideal control of the rate and rhythm of distraction. Early designs that were mechanically actuated had problems with rate control resulting in bone formation complications. The two current designs use either magnetic or electrical control, and have reliable use while eliminating pin and wire complications and fixator-associated pain. While internal lengthening has obvious advantages, there are specific patient indications. Children with open growth plates or small bones are not suitable candidates. External fixation and associated hybrid techniques such as lengthening over nail and plate-assisted lengthening continue to be necessary for certain cases. The correction of severe foot and joint deformities will still require external fixation for the foreseeable future. Improvements in instrumentation and expansion to other conditions will broaden the safety and efficacy of these devices. The motorized intramedullary lengthening nail is an important new tool for the limb length and deformity correction surgeon.

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Part II

Related Concepts and Management Options

Biomechanically Based Clinical Decision Making in Pediatric Foot and Ankle Surgery

10

Jon R. Davids

Introduction

The understanding and classification of functional deviations and deficits at the ankle and foot are based upon an appreciation of normal function during the gait cycle [1–3]. The interaction between the ankle, foot and the floor is a critical element of normal gait. Function of the ankle and foot is determined by a complex interaction of anatomy, physiology, and physics. Proper ankle and foot alignment is required for optimal function of the knee and hip during gait. Disruption of normal function of the ankle and foot may disrupt knee and hip function, compromising the energy efficiency of gait and in extreme cases precluding the ability to ambulate.

Clinical decision making for the management of ankle and foot deformities in children can be standardized by the use of a diagnostic matrix (Table 10.1) [4]. This paradigm is based upon the collection and integration of data from five sources: the clinical history, physical examination, plain radiographs, observational gait analysis, and in complex cases associated with certain disease processes (e.g., cerebral palsy, myelodysplasia, and hereditary sensorimotor neuropathies), quantitative gait analysis (which may include kinematic/kinetic analyses, dynamic electromyography (EMG), and dynamic pedobarography).

This chapter will begin with an overview of normal ankle and foot function during the gait cycle. This will provide a framework for the identification of common (or coupled), and uncommon (or uncoupled), segmental malalignments of the ankle and foot. This will be followed by an overview (principles and indications) of the most common interventions (i.e., soft tissue surgery, guided growth, osteotomies, and arthrodeses) utilized to correct these segmental malalignments. Finally, a standardized approach for the preoperative, intraoperative, postoperative, and surveillance of ankle and foot alignment and function will be presented.

Ankle and Foot Function During Normal Gait

The understanding of ankle and foot function during normal gait is facilitated by considering the lower leg to consist of four segments: the tibial or shank segment, the hindfoot (talus and calcaneus), the midfoot (navicular, cuneiforms, and cuboid), and the forefoot (metatarsals and phalanges) [1–3, 5, 6] (Fig. 10.1a, b). It is also helpful to consider the foot to consist of two columns: the medial column (talus, navicular, cuneiforms, great toe metatarsal, and phalanges), and the lateral column (calcaneus, cuboid, lesser toe metatarsals, and phalanges) [7] (Fig. 10.2a–c). Standardized, consistent terminology is required to describe the alignment of the separate segments of the ankle and foot [8]. Movement of the plantar aspect of the segment in question during the gait cycle is described as *inversion* (towards the midline) or *eversion* (away from the midline). Movement of the distal aspect of the segment in question during the gait cycle is described as *adduction* (towards the midline) or *abduction* (away from the midline). *Supination* is a combination of inversion and adduction. *Pronation* is a combination of eversion and abduction. Rotation of the segment about its longitudinal axis towards the midline is described as *internal rotation*. Rotation of the segment about its longitudinal axis away from the midline is described as *external rotation*.

The gait cycle is a period of time beginning with the initial contact of the reference foot with the ground, continuing through ipsilateral stance and swing phases until the subsequent ipsilateral initial contact. Stance phase occurs when the reference limb is in contact with the ground. Swing phase occurs when the reference limb is not in contact with the ground. The interaction of the ankle and foot with the ground during the stance phase of the gait cycle is described by the

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Table 10.1 The diagnostic matrix for the assessment of the ankle and foot deformities

Source	Information
Clinical history	Pain
	Tripping
	In-/Out-toeing
Physical examination	Gross foot shape weight-bearing/non weight-bearing
	Flexible/rigid
	Plantar callous pattern
Radiographic examination	Segmental alignment weight-bearing, AP and LAT views
Observational gait analysis	Foot contact with floor (3 rockers)
	Foot progression angle
	Foot clearance in swing phase
Quantitative gait analysis	Kinematics
	Kinetics
	Dynamic EMG
	Pedobarography

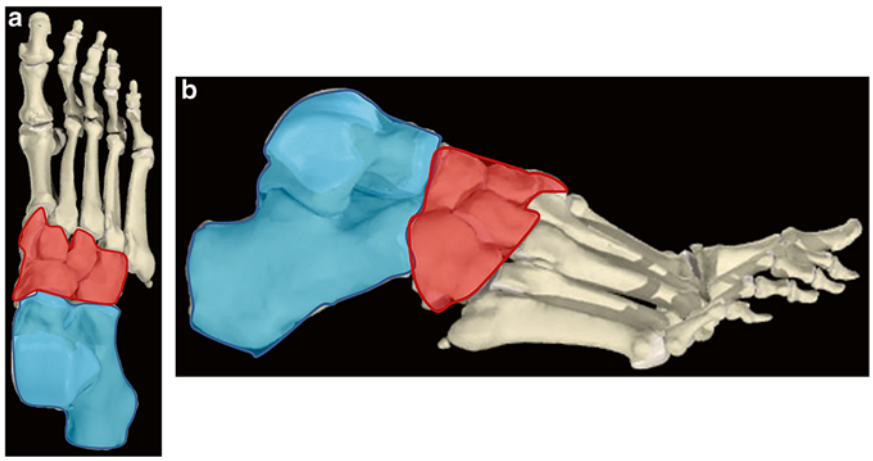


Fig. 10.1 Three segments of the foot. (**a**) Diagram of the anteroposterior view of the right foot. The hindfoot segment is *blue*, the midfoot segment is *red*, and the forefoot segment is *white*. (**b**) Diagram of the lateral view of the right foot. The hindfoot segment is *blue*, the midfoot segment is *red*, and the forefoot segment is *white*

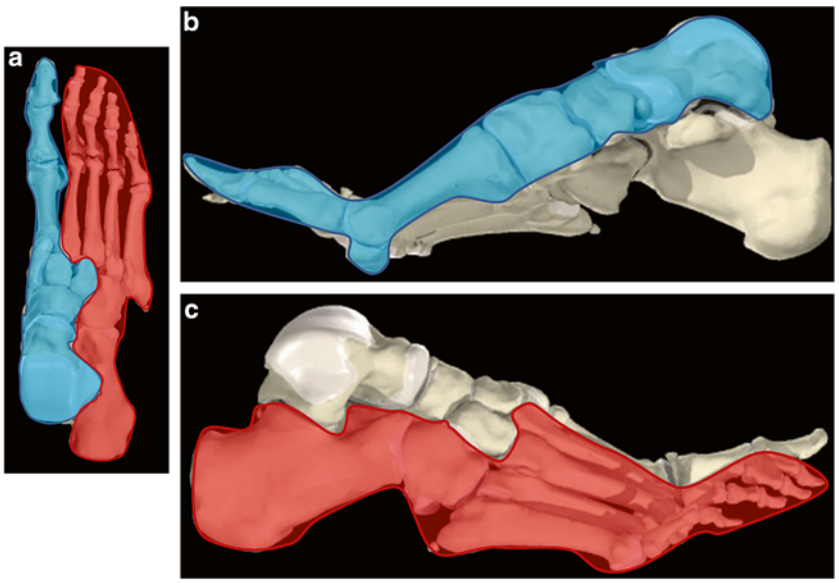


Fig. 10.2 Two columns of the foot. (**a**) Diagram of the anteroposterior view of the right foot. The medial column is *blue*, and the lateral column is *red*. (**b**) Diagram of the lateral view (medial side) of the right foot. The medial column is *blue*. (**c**) Diagram of the lateral view (lateral side) of the right foot. The lateral column is *red*

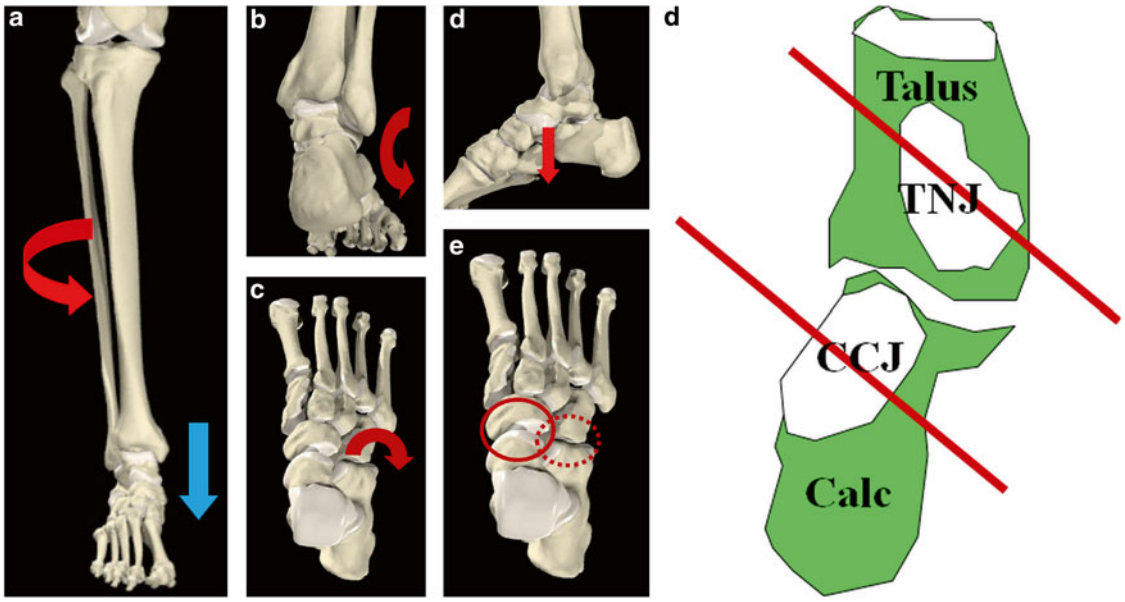


Fig. 10.3 Skeletal alignment of the lower extremity during the first rocker of stance phase. (**a**) The tibia is rotating internally (*red arrow*) and the ankle is plantarflexing (*blue arrow*). (**b**) Hindfoot alignment during the first rocker of stance phase consists of calcaneal eversion (*red arrow*). (**c**) Hindfoot alignment during the first rocker of stance phase consists of calcaneal abduction (*red arrow*). (**d**) Hindfoot pronation (eversion and abduction) forces the talus to plantarflex (*red arrow*) during the first rocker of stance phase. (**e**) This "unlocks" the primary joints of the midfoot (talo-navicular in *solid circle*, calcaneocuboid in *dashed circle*). (**f**) Coronal view of the articulation between the hindfoot and midfoot in first rocker of stance phase. Lateral is to the left, medial is to the right. *TNJ* talo-navicular joint, *CCJ* calcaneocuboid joint, *Calc* calcaneus. The main axes of the TNJ and CCJ are parallel (*red lines*), which allows motion and "unlocks" the midfoot

concept of three rockers [2, 3]. In normal gait, the heel is the first part of the foot to contact the ground at initial contact. The ankle subsequently plantar flexes until the foot is flat on the floor. This motion is controlled by the eccentric activity of the ankle dorsiflexor muscle group. The *first, or heel rocker*, occurs from heel strike to foot flat during the loading response subphase of stance. As the body progresses forward, the tibia advances forward over the foot, which is achieved by ankle dorsiflexion. This motion is controlled by eccentric activity of the ankle plantar flexor muscle group. The *second, or ankle rocker*, occurs as the tibia advances over the foot during the midstance subphase of stance. Immediately prior to the initial contact of the opposite foot, the heel of the reference foot rises off the ground and dorsiflexion occurs through the metatarsophalangeal joints of the forefoot. This motion is controlled by concentric activity of the ankle plantar flexor muscle group. The *third, or forefoot rocker*, occurs as the ankle begins to plantar flex during the terminal stance subphase of stance. This is an essential event during normal gait, as the largest moment generated by any single muscle group during the gait cycle is the internal plantar flexion moment generated by the ankle plantar flexor muscle group during third rocker in terminal stance [2, 3, 9].

In the stance phase of the normal gait cycle, the ankle and foot provide shock absorption during loading response (first or heel rocker), stability during midstance (second or ankle rocker), and a rigid lever during terminal stance (third or forefoot rocker) [2, 3]. During loading response, the tibial or shank segment rotates internally, and the ankle is plantarflexing (Fig. 10.3a). This results in eversion and abduction of the hindfoot, primarily through the subtalar joint (see Fig. 10.3b, c). Pronation of the hindfoot forces the talus to plantarflex, which "unlocks" the joints of the midfoot, which follows into pronation (see Fig. 10.3d, e) This coupled movement of the hindfoot and midfoot results in maximum flexibility of the foot, which allows the joints to contribute to shock absorption. During midstance, the tibial or shank segment is rotating externally, and the ankle is dorsiflexing (Fig. 10.4a). This results in inversion and adduction of the hindfoot, primarily through the subtalar joint (see Fig. 10.4b, c). Supination of the hindfoot forces the talus to dorsiflex, which "locks" the joints of the midfoot, which follows into supination (see Fig. 10.4d, e) This coupled movement of the hindfoot and the midfoot results in restoration of the longitudinal arch of the foot and maximum rigidity of the foot, which enhances stability. During terminal stance, the tibial or shank segment continues to rotate externally, and the ankle continues to dorsiflex. As the body progresses forward, the center of pressure beneath the foot advances distally into the forefoot. Because the segments of the foot are aligned to promote maximum rigidity, the forefoot is stable as it is loaded. The rigidity of the foot segments provides an optimal

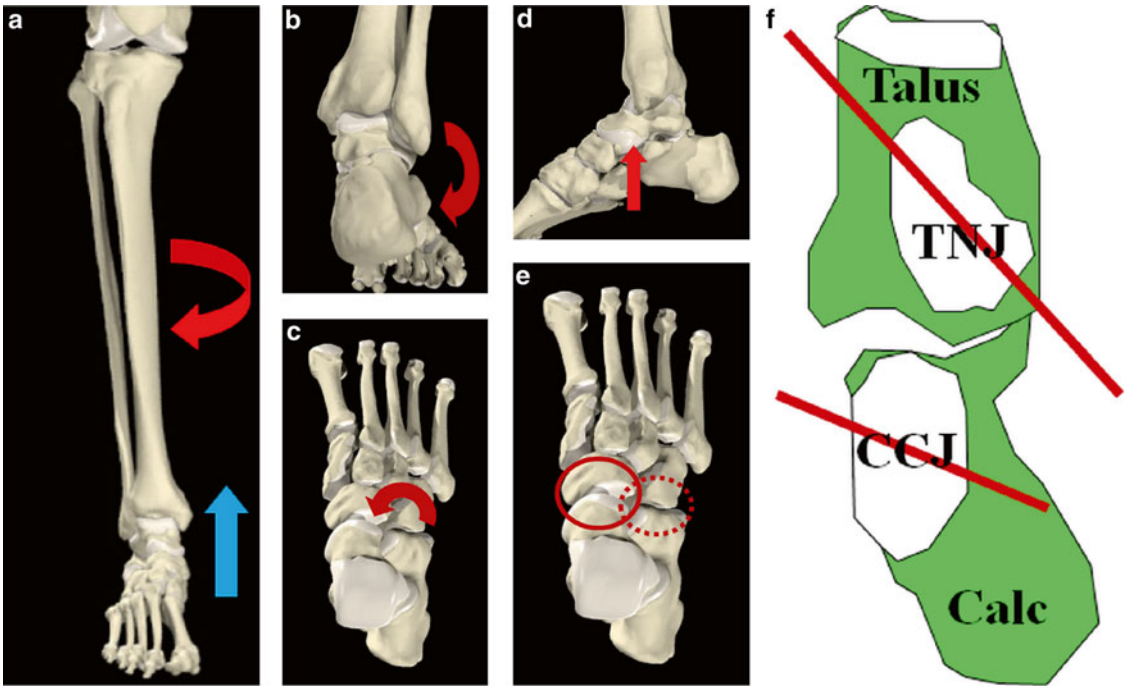


Fig. 10.4 Skeletal alignment of the lower extremity during the second rocker of stance phase. (**a**) The tibia is rotating externally (*red arrow*) and the ankle is dorsiflexing (*blue arrow*). (**b**) Hindfoot alignment during the second rocker of stance phase consists of calcaneal inversion (*red arrow*). (**c**) Hindfoot alignment during the second rocker of stance phase consists of calcaneal adduction (*red arrow*). (**d**) Hindfoot supination (inversion and adduction) forces the talus to dorsiflex (*red arrow*) during the second rocker of stance phase. (**e**) This "locks" the primary joints of the midfoot (talo-navicular in *solid circle*, calcaneo-cuboid in *dashed circle*). (**f**) Coronal view of the articulation between the hindfoot and midfoot in second rocker of stance phase. Lateral is to the left, medial is to the right. *TNJ* talo-navicular joint, *CCJ* calcaneocuboid joint, *Calc* calcaneus. The main axes of the TNJ and CCJ are no longer parallel (*red lines*), which restricts motion and "locks" the midfoot

lever arm to the ankle plantar flexor muscles during terminal stance. Normal, expected segmental alignment patterns of the ankle and foot during the stance phase of the gait cycle, as described above, are the consequence of coupled movements between the anatomical segments.

In the swing phase of the normal gait cycle, the foot and ankle contribute to clearance and pre-positioning for the subsequent stance phase [2, 3, 6]. During pre- and initial swing the tibia or shank segment is rotating externally and the ankle is plantar flexing. The segments of the foot are "unlocked" as the limb is unloaded. During mid swing the tibia or shank segment is rotating internally and the ankle is dorsiflexing. These coupled motions serve to functionally shorten the limb and promote clearance. During terminal swing these coupled motions continue and the foot is maintained in a plantigrade alignment, perpendicular to the anatomical axis of the tibia or shank segment. This pre-positioning of the foot during terminal swing will result in a heel strike at the initial contact, which is the optimal alignment for the ankle and foot as the extremity enters the subsequent stance phase in loading response.

Box 10.1

- The lower leg consists of four segments: the tibial or shank segment, the hindfoot (talus and calcaneus), the midfoot (navicular, cuneiforms and cuboid), and the forefoot (metatarsals and phalanges).
- The interaction of the ankle and foot with the ground during the stance phase of the gait cycle is described by the concept of three rockers; first or heel rocker, second or ankle rocker, and third or forefoot rocker.
- In the stance phase of the normal gait cycle, the ankle and foot provide shock absorption during loading response (first or heel rocker), stability during midstance (second or ankle rocker), and a rigid lever during terminal stance (third or forefoot rocker).
- In the swing phase of the normal gait cycle, the foot and ankle contribute to clearance and pre-positioning for the subsequent stance phase.

Segmental Malalignment Patterns of the Ankle and Foot

Segmental malalignments of the ankle and foot may be categorized as coupled or uncoupled. *Coupled* segmental malalignments represent exaggerations of normal segmental alignments that occur during the gait cycle (as described above). The three most common coupled segmental malalignments are equinus, equinoplanovalgus, and equinocavovarus. Equinus is characterized by excessive plantar flexion of the hindfoot relative to the ankle, with normal midfoot and forefoot alignment (Fig. 10.5a, b) Equinoplanovalgus is characterized by equinus deformity of the hindfoot, coupled with pronation deformities of the midfoot and forefoot (Fig. 10.6a, b). The lateral column of the foot is functionally and/or structurally shorter than the medial column. Ankle valgus and hallux valgus deformities are frequently seen in association with equinoplanovalgus foot segmental malalignment (see Fig. 10.6c, d) Equinocavovarus is characterized by equinus deformity of the hindfoot, coupled with supination deformity of the midfoot and variable malalignment of the forefoot (Fig. 10.7a, b) The lateral column is functionally and/or structurally longer than the medial column. Compensatory ankle valgus deformity may be seen in association with equinocavovarus foot segmental malalignment.

In all three coupled segmental malalignment patterns, heel strike at initial contact does not occur, disrupting the first or hindfoot rocker and shock absorption function in loading response. Equinus and equinocavovarus malalignment patterns disrupt the second or ankle rocker by blocking ankle dorsiflexion, compromising stability function in midstance. Equinoplanovalgus malalignment maintains the mid- and forefoot segments in an "unlocked" alignment, compromising stability function in midstance, which may result in excessive loading of the plantar, medial portion of the midfoot. All three coupled segmental malalignments may compromise the ability of the ankle plantar flexor muscles to generate an adequate internal plantar flexion moment during third or forefoot rocker. The hindfoot malalignment associated with equinus and equinocavovarus malalignment patterns shortens the length of the plantar flexor muscles, compromising their ability to generate tension, as described by the length-tension curve for skeletal muscle [10, 11]. With equinoplanovalgus, the moment generating capacity of the ankle plantarflexor muscles is further compromised by the malalignment of mid-and forefoot segments, which effectively shortens the lever arm available to this muscle group during the third or forefoot rocker. All three segmental malalignment patterns of the ankle and foot may inhibit ankle dorsiflexion in swing phase, compromising clearance in midswing and proper positioning of the foot and ankle in terminal swing.

Uncoupled segmental malalignments are alignment patterns between the hind-, mid-, and forefoot that never occur during the gait cycle. Equinocavovalgus is an example of an uncoupled segmental malalignment pattern (Fig. 10.8a, b). Uncoupled segmental malalignments of the ankle and foot are relatively uncommon, and are frequently the consequence of deformity following previous surgery.

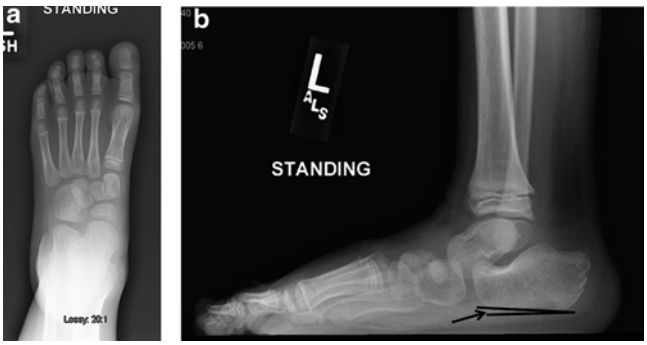


Fig. 10.5 Plain radiographs of the foot in a child with equinus deformity. (**a**) Anteroposterior view shows normal segmental alignment. (**b**) Lateral view shows hindfoot plantarflexion (diminished calcaneal pitch, indicated by *solid arrow* towards angle formed by *solid lines*, normal is approximately 20°), with otherwise normal segmental alignment

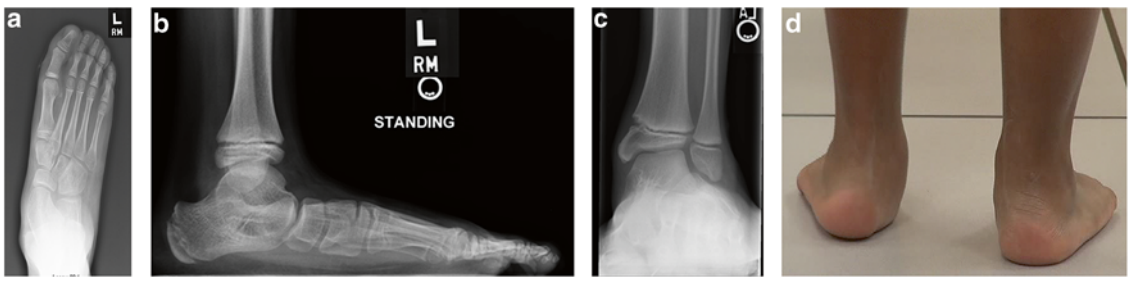


Fig. 10.6 Plain radiographs of the foot in a child with equinoplanovalgus deformity. (**a**) Anteroposterior view shows hindfoot pronation, talonavicular uncoverage, forefoot abduction and hallux valgus. (**b**) Lateral view shows hindfoot plantarflextion, midfoot pronation (excessive naviculocuboid overlap), and forefoot pronation (excessive overlap of the metatarsals). (**c**) Anteroposterior view of the ankle shows ankle valgus deformity (increased tibiotalar angle, lateral wedging of the distal tibial epiphysis, and a high fibular station). (**d**) Clinical photograph of the left hindfoot in weight-bearing for this child. There is significant hindfoot valgus deformity, which the radiographs show to be a consequence of both tibiotalar (ankle) and talo-calcaneal (subtalar) valgus malalignments

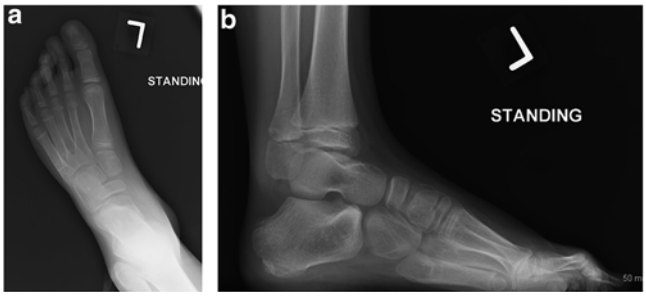


Fig. 10.7 Plain radiographs of the foot in a child with equinocavovarus deformity. (**a**) Anteroposterior view shows hindfoot supination (excessive overlap of the talus and calcaneus), and forefoot adduction (medial deviation of the great toe metatarsal relative to the talus). (**b**) Lateral view shows hindfoot varus (parallelism of the talus and calcaneus), midfoot supination (diminished naviculocuboid overlap), and forefoot supination (excessive stacking of the metatarsals)

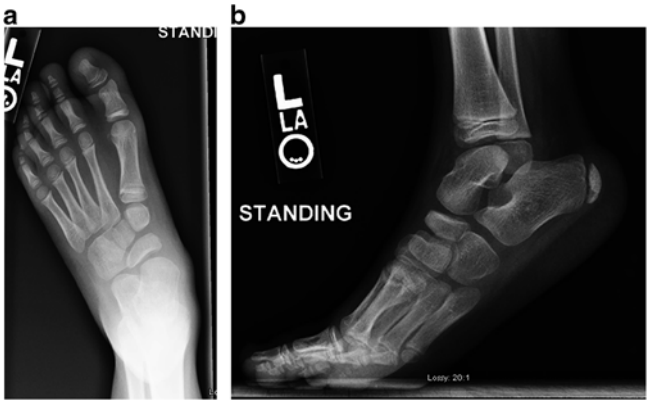


Fig. 10.8 Plain radiographs of the foot in a child with equinocavovalgus deformity, which is an example of an uncoupled malalignment. (**a**) Anteroposterior view shows hindfoot pronation, talonavicular uncoverage, forefoot abduction and hallux valgus interphalangeus. (**b**) Lateral view shows hindfoot varus (parallelism of the talus and calcaneus), midfoot supination (diminished naviculocuboid overlap), and forefoot supination (excessive stacking of the metatarsals). The normal coupling between the three segments of the foot is disrupted, as shown by the dissonance between anteroposterior and lateral radiographic views

Box 10.2

- Segmental malalignments of the ankle and foot may be categorized as coupled or uncoupled. *Coupled* segmental malalignments represent exaggerations of normal segmental alignments that occur during the gait cycle. *Uncoupled* segmental malalignments are alignment patterns between the three segments of the foot that never occur during the gait cycle.
- The three most common coupled segmental malalignments are equinus, equinoplanovalgus, and equinocavovarus.

Surgical Interventions

Interventions to correct foot deformities in children may be selected to improve function and/or cosmesis. Both of these goals may be achieved by surgeries designed to improve foot shape. It is presumed that improved foot shape following soft tissue and skeletal surgery can restore both the stability function of the foot during the second or ankle rocker in midstance and the skeletal lever arm function of the foot during the third or forefoot rocker in terminal stance [12–15]. However, it is important to recognize that increased foot stiffness associated with many skeletal surgical procedures (e.g., arthrodesis) utilized to improve foot shape may compromise shock absorption function of the foot during the first or ankle rocker in loading response [16]. Cosmetic improvements following foot surgery are related to improved visual assessment of static standing foot alignment (particularly restoration of the medial longitudinal arch and toe alignment) and improved foot progression angle during stance phase.

Soft tissue surgeries include release, lengthening, or shortening of muscles, tendons, ligaments, and joint capsules; or transfer of the muscle tendon unit. Release of soft tissue structures is generally reserved for fixed ankle and foot deformities associated with progressive disease processes in subjects who have significant impairment and whose goals are to improve static alignment in order to promote brace wear, shoe wear, or foot position in a wheelchair, and to facilitate transfer level motor activities. Lengthening of soft tissue structures is appropriate for fixed ankle and foot deformities associated with static or stable disease processes in subjects whose goals are to improve alignment to facilitate dynamic functional motor activities. It is important to recognize that in most cases there is preexisting weakness of the muscle tendon unit that is being lengthened, and that all lengthening surgical procedures result in additional weakening. When operating on muscles and tendons, selective surgical lengthening techniques that minimize the subsequent weakness of the muscle tendon unit are therefore favored [10, 11]. Surgical procedures that partially (also called "split") or completely transfer the muscle tendon unit are reserved for completely dynamic ankle and foot deformities associated with static or stable disease processes in subjects who have relatively lower levels of motor impairment [17–19]. Partial and complete transfers are performed to address a dynamic muscle imbalance. Achieving perfect dynamic balance with all types of tendon transfer can be challenging. Over correction may occur with either partial or complete transfers, and under correction may be seen following partial transfers. Proper patient selection is essential.

Skeletal surgeries include guided growth, osteotomy, and arthrodesis. Guided growth can be utilized to correct ankle valgus deformity and metatarsus/phalangeal deformity

Table 10.2 Levels of ankle and foot deformity and treatment options

	Treatment options		
Levels of deformity	Pharmacologic/neurosurgery	Muscle tendon surgeries	Skeletal surgeries
I: Dynamic soft tissue imbalance, no skeletal deformities	Neuromuscular junction blockade	Partial or complete tendon transfers	Usually not necessary
	Selective dorsal rhizotomy		
	Intrathecal baclofen		
II: Fixed soft tissue imbalance, no fixed skeletal deformities	Not appropriate as isolated intervention	Sequential lengthening	Usually not necessary
		(Myotendinous Junction Recession, Tendon Lengthening)	
III: Fixed soft tissue imbalance, with structural skeletal deformities	Not appropriate as isolated intervention	Sequential lengthening	Osteotomy or arthrodesis
		Appropriate in conjunction with skeletal surgery	(Lengthening, shortening, angular, rotational)

associated with juvenile hallux valgus [20–22]. Typically two or more years of growth remaining is required to achieve correction by guided growth strategies. Osteotomy and arthrodesis techniques may correct deformity by addition (i.e., lengthening), subtraction (i.e., shortening), angulation, or rotation. Acute skeletal lengthening techniques are preferred as they utilize coupled segmental relations between the segments of the foot to achieve correction [12, 23, 24]. These procedures require a bone graft, and in most cases internal fixation. The use of allograft is favored over autograft, though late allograft collapse during the re-ossification phase of graft incorporation has been reported [25]. Acute skeletal shortening procedures are used for the correction of the most rigid foot deformities, which are usually associated with congenital conditions (e.g., arthrogryposis) or peripheral neuropathies and myopathies (e.g., dystrophin deficient muscular dystrophies).

Clinical decision-making for surgery is guided by the classification of foot deformities into three levels (Table 10.2). Level I deformities are characterized by dynamic soft tissue imbalance. Skeletal anatomy is normal. Level II deformities are characterized by fixed or myostatic soft tissue imbalance. However, the underlying skeletal segmental malalignments are flexible and correctable on manipulation. Level III deformities are characterized by structural skeletal deformities that are usually associated with fixed or myostatic soft tissue imbalance. For foot deformities associated central nervous system conditions (e.g., cerebral palsy) it is not always possible to determine preoperatively if the deformity is level II or III. In such cases, sequential soft tissue lengthening is performed first, followed by intraoperative assessment of segmental foot alignment with stress radiographs under fluoroscopy (Fig. 10.9). If correction of alignment is determined to be insufficient, then sequential skeletal surgery, focused on the segment(s) that remain malaligned is performed.



Fig. 10.9 Intraoperative clinical photograph of the use of a "foot pusher" to achieve simulated weight bearing lateral radiographs

Box 10.3

- *Soft tissue surgeries* include release, lengthening, or shortening of muscles, tendons, ligaments, and joint capsules; or transfer of the muscle tendon unit.
- *Skeletal surgeries* include guided growth, osteotomy, and arthrodesis.
- *Clinical decision-making for surgery* is guided by the classification of foot deformities into three levels

Assessment Tools and Indications

Clinical decision making for the management of ankle and foot deformities in children integrates a range of data by the use of a diagnostic matrix (see Table 10.1) [4]. The integration of data from multiple sources results in a degree of redundancy that improves decision making and quality of outcomes. When the data is consistent across the fields, and the problem is common, the confidence in decision making should be high. When the data is apparently inconsistent across the fields, or the problem is unusual, then the confidence in decision making should be lowered. Surgical treatment paradigms for coupled segmental malalignments of the ankle and foot are more advanced and generally more effective than those for uncoupled segmental malalignments. The latter deformities are individually unique, and therefore require careful, case-by-case surgical planning and treatment.

Different types of ankle and foot deformities are best evaluated with different combinations of assessment tools at different points in the course of management (Table 10.3). Plain radiographic views at all points include standing anteroposterior and lateral views of the foot, and a standing anteroposterior view of the ankle. Additional views of the hindfoot (Cobey view) and subtalar joint (Harris heel view), while not part of the routine radiographic assessment paradigm, may be used to further assess for overall hindfoot alignment and the presence of a talocalcaneal coalition. In the case of the latter, computed tomography scan (CT) is required to confirm the diagnosis and for adequate planning prior to surgical management. True or simulated weight bearing is essential; foot segmental alignment may be dramatically different in loaded versus unloaded conditions, and non-weight bearing views are of little value. Qualitative and quantitative assessment of plain radiographs should be done is a systematic fashion, referring to normative data to objectively describe the ankle and foot segmental alignment (Table 10.4) [7, 26]. Accurate assessment of the causes of hindfoot alignment in the coronal plane (i.e., determining the relative contributions of deformity at the tibiotalar and talocalcaneal joints) requires analysis of the anteroposterior radiograph of the ankle in addition to the views of the foot [27] (see Fig. 10.6) For the *preoperative assessment* of the valgus and varus foot, kinematics is limited by the reliance on a single segment foot model that can notaccount for abnormal midfoot alignments [28, 29]. Dynamic electromyography (EMG) is only necessary for feet with dynamic varus deviations, to sort out the relative contributions of the gastrocsoleus complex, the tibialis posterior, and tibialis anterior muscles [30, 31]. Pedobarography is indicated for valgus and varus malalignments [32]. The relation between static standing foot alignment (as indicated by plain radiographs) and dynamic foot loading (as indicated by pedobarography) is complex [33, 34]. The former is not always a good predictor of the latter, and when there is apparent discrepancy between the two modalities, priority should be given to the pedobarograph data as it is a closer measure of actual function [32, 34]. *Intraoperative assessment* relies

Table 10.3 Assessment tools for the ankle/foot

	Assessment tools						
	History	Physical examination (ROM, OGA)	Standing/stress radiographs	Kinematics	Kinetics	EMG	Pedobarography
Deformity	Preoperative						
	Preoperative						
Equinus	X	X	X	X			
Valgus	X	X	X	Limited	X		X
Varus	X	X	X	Limited	X	X	X
	Intraoperative						
Equinus			X				
Valgus			X				
Varus			X				
	Postoperative						
Equinus	X	X	X				
Valgus	X	X	X		X		X
Varus	X	X	X		X		X
	Surveillance						
Equinus	X	X					
Valgus	X	X					
Varus	X	X					

ROM range of motion, *OGA* observational gait analysis, *EMG* electromyography, *X* appropriate tool to use

Table 10.4 Normal radiographic angle measurements [7, 26]

Ankle and foot segments and columns*	Normal mean ±1 SD*	Increased value (>Mean + 1 SD)	Decreased value (<Mean − 1 SD)
Hindfoot			
Tibiotalar Angle (anteroposterior view, degrees)	1.1 ± 3.75	Eversion	Inversion
Calcaneal Pitch (lateral view, degrees)	17 ± 6.0	Calcaneus	Equinus
Tibiocalcaneal Angle (lateral view, degrees)	69 ± 8.4	Equinus	Calcaneus
Talocalcaneal Angle (lateral view, degrees)	49 ± 6.9	Eversion	Inversion
Midfoot			
Naviculocuboid Overlap (anteroposterior view, percentage)	47 ± 13.8	Pronation	Supination
Talonavicular Coverage Angle (anteroposterior view, degrees)	20 ± 9.8	Abduction	Adduction
Lateral Talo-First Metatarsal Angle (lateral view, degrees)	13 ± 7.5	Pronation	Supination
Forefoot			
Anteroposterior Talo-First Metatarsal Angle (anteroposterior view, degrees)	10 ± 7.0	Abduction	Adduction
Metatarsal Stacking Angle (lateral view, degrees)	8.2 ± 9	Supination	Pronation
Columns			
Medial-Lateral Column Ratio (lateral view)	0.9 ± 0.1	Abduction	Adduction

For direction on how to measure each angle or distance ratio on plain radiographs, see reference [7]
* See figure 10.2

primarily on stress radiographs, which requires the use of a "foot pusher" device to obtain simulated weight bearing views of the foot (see Fig. 10.9). *Postoperative assessment*, once recovery, healing, and rehabilitation following surgery have been completed, should be as quantitative as possible, mirroring the preoperative assessment to allow objective assessment of outcome in multiple domains. Finally, *surveillance* is guided by the history and physical examination, with additional assessment tools utilized only if a problem has been identified.

Box 10.4

- Clinical decision making for the management of ankle and foot deformities in children integrates a range of data by the use of a diagnostic matrix, which considers and integrates data from five domains.
- Coupled segmental malalignments are easier to treat surgically than uncoupled segmental malalignments.
- Different types of ankle and foot deformities are best evaluated with different combinations of assessment tools at different points in the course of management

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Pediatric Joint Contractures

11

Christopher Iobst

Introduction

Joint contractures can be a debilitating cause of lower extremity deformity. They can occur in isolation or in combination with bony deformity. Mild joint contractures can often be managed nonoperatively, while severe contractures may require surgical interventions. All contractures can be frustrating to deal with, both for the patient and the physician, as they often recur in the growing child despite best efforts. It is important to learn how to identify, evaluate, and manage joint contractures in pediatric patients. This chapter reviews the current methods for evaluating and treating equinus contractures, knee flexion contractures, and knee extension contractures in children.

Equinus Contracture

Equinus contracture is described as the inability to fully dorsiflex the ankle through the tibiotalar joint. Equinus contracture interferes with gait by decreasing stability in stance phase, by removing the smooth transition of the body over the foot, and by causing inadequate clearance of the foot in the swing phase [1]. Although the term equinus is frequently used, a working definition remains lacking. Digiovanni et al. have offered the following standardization of terms to describe the inability to dorsiflex through the tibiotalar joint: gastrocnemius equinus represents maximal ankle dorsiflexion of less than or equal to 5° with the knee in full extension, and Achilles tightness represents maximal ankle dorsiflexion of less than or equal to 10° with the knee in 90° of flexion [2].

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Pathoanatomy

Equinus contracture can be an unrecognized underlying factor in many of the pathologies associated with the foot and ankle. In a normal gait pattern, the center of pressure on the foot is approximately the second metatarsal-cuneiform joint [3]. The pull of the ankle plantarflexors, the gastrocnemius and soleus muscles, counteracts the body's anteriorly displaced center of pressure and prevents the body from falling forward in stance. In an equinus deformity, however, the foot's center of pressure moves about 3 cm distally and 3 mm laterally to its normal location [3]. This abnormal loading of the forefoot and midfoot in stance can lead to chronic mechanical overload and structural breakdown of the foot and ankle. It has been demonstrated that patients with metatarsalgia or related forefoot/midfoot symptoms had less average maximal ankle dorsiflexion with the knee in extension than a control population without foot symptoms [2]. Equinus also interferes with gait by preventing the tibia's forward progression over the foot during stance phase, which shortens the stride length. Another abnormal gait finding may be hyperextension of the knees, which develop as a compensatory mechanism to allow an equinus ankle to maintain a plantigrade foot position.

Equinus positioning of the foot has frequently been called an Achilles contracture. However, this is a misnomer because the majority of the perceived stiffness or stretch occurs within the muscle bellies themselves, not in the tendon [2]. The tendon is responsible for only about 3–5 % of this change in position [2]. Therefore, although physicians often describe "Achilles" tightness and address the Achilles tendon directly to treat this disorder, the origin of the problem is often at the level of the gastrocnemius and soleus muscles [2].

Equinus deformity can be divided into two main types, muscular and osseous. Muscular equinus can be further subdivided by identifying the primary cause, gastrocnemius equinus or gastrocsoleus equinus. It is important not to overlook osseous deformity that can mimic soft tissue contractures. For example, distal tibial procurvatum, forefoot

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equinus, and flattop talus can each mimic an ankle equinus contracture [4]. It is also possible to have a combination of muscular and osseous equinus.

Clinical Evaluation

While obtaining the history, the physician should try to ascertain the underlying cause of the contracture (neuromuscular disease, iatrogenic, congenital, etc.) as this can influence the natural history and help the surgeon make appropriate treatment recommendations. It is also important to find out the extent to which the contracture affects the child's function and whether or not any previous treatment has been attempted.

Proper examination of the foot and ankle for equinus requires a thorough understanding of the relevant anatomy. Equinus deformity may be caused by contracture of the gastrocnemius alone or a contracture of both the gastrocnemius and the soleus. Because the gastrocnemius originates above the knee joint and the soleus does not, it is possible to differentiate the contribution of each muscle to the equinus contracture by physical examination. The Silfverskiold test evaluates maximal ankle dorsiflexion with the knee in full extension and then again with the knee in 90° of flexion which relaxes the gastrocnemius muscle (Fig. 11.1) [5]. It is critical for both components of this test, that the examiner inverts the subtalar joint and supinates the forefoot, prior to dorsiflexing the ankle. This locks the talonavicular joint and neutralizes the hindfoot, preventing any motion through the midfoot, which could be misconstrued as ankle dorsiflexion. If the ankle joint dorsiflexes past neutral equally well with both the knee extended and flexed, there is usually no equinus contracture present. However, if the ankle joint dorsiflexes with the knee flexed but does not reach the same dorsiflexion with the knee extended, then there is gastrocnemius equinus. On the other hand, if the ankle dorsiflexion is less than neutral with both the knee flexed and extended, then it can either be gastroc-soleus equinus or osseous equinus. Osseous equinus can be determined by identifying bony deformity such as distal tibial procurvatum or intra-articular exostoses on a lateral radiograph of the ankle. Some foot deformities, such as a cavus foot with forefoot plantarflexion, can also be mistaken for an equinus deformity.

Imaging

Radiographically, equinus can be evaluated on a weight-bearing lateral radiograph in which the patient stands with the knee straight. The magnitude of the equinus can then be determined from the standing lateral radiograph by measuring the tibial-sole angle [6]. This is calculated by drawing a line along the sole of the foot from the plantar aspect of head of first metatarsal to plantar aspect of calcaneus and joining it with a line drawn along the long axis of the tibia (Fig. 11.2). The degree of equinus deformity can be categorized as mild (less than 20° plantarflexion), moderate (20°–40°) and severe (greater than 40°) [7].

Management Strategy and Decision Making

Nonoperative

Most mild and some moderate equinus will respond to physical therapy with stretching, casting and bracing. Because early age at first surgery in children with cerebral palsy is a major risk factor for recurrent equinus deformity, temporizing measures to postpone the first surgery should be considered in younger children [8, 9]. Options include the

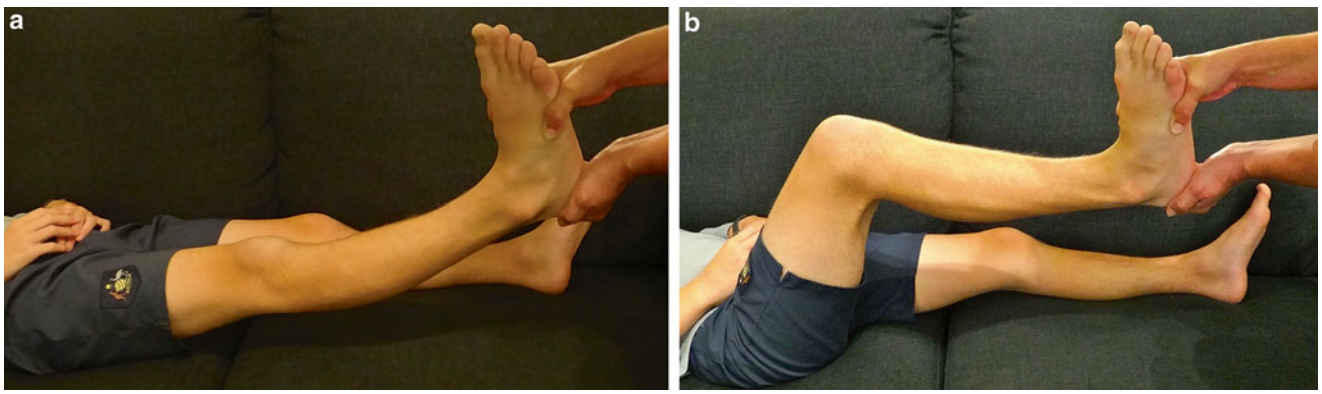


Fig. 11.1 The Silfverskiold test is used to determine the contribution of the gastrocnemius to the overall equinus contracture. The foot is dorsiflexed with the knee extended and then again with the knee flexed to 90°. If there is more dorsiflexion of the ankle possible with the knee flexed than with the knee extended, a gastrocnemius contracture is present. The test should be performed with the forefoot supinated and the sub-talar joint inverted to lock the midfoot joints

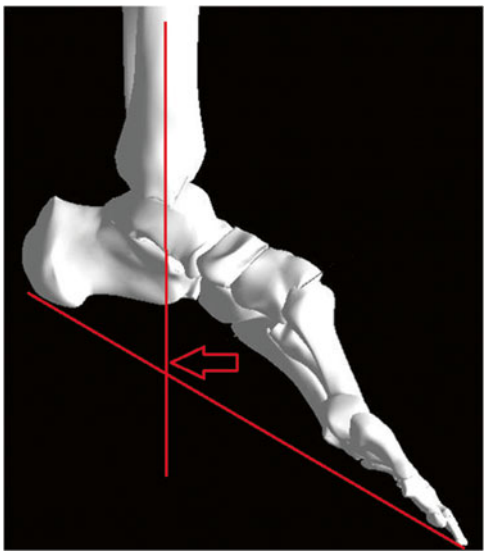


Fig. 11.2 The magnitude of the equinus can be measured on a lateral radiograph of the ankle by measuring the tibial-sole angle. Equinus deformity will have a measurement of greater than 90°

use of botulinum toxin A, casting and orthoses. Serial casting should be performed with the patient in a prone position to obtain the most effective correction [10]. Botulinum toxin A has had successful short-term outcomes and may allow surgery to be delayed until after the age of 6 years in children with cerebral palsy [9]. Planning of surgical procedures can be based on the results of selective botulinum toxin A injections. Potential problems include the development of antibodies to botulinum toxin A and the cost of the treatment. Botulinum toxin A should only be used in muscles that produce dynamic deformities in the absence of fixed contracture.

Operative

Lengthening of Muscles and Tendons

If the patient demonstrates an equinus deformity with gait but the ankle has sufficient dorsiflexion past neutral on passive range of motion examination, the equinus is dynamic, and surgical treatment may not be needed. However, if the child has failed conservative management or demonstrates tripping, falling, or forefoot pain with functional impairment, then surgery should be considered [11]. The presence of a fixed equinus contracture at the ankle, with dorsiflexion to less than neutral is also an indication to operate. An important principle governing surgery for equinus is to use the most conservative gastrocnemius lengthening procedure possible to achieve approximately 5° of dorsiflexion with the knee extended [11]. The amount of lengthening required for a specific degree of fixed equinus is often much less than surgeons estimate. The appropriate surgical treatment for equinus deformity must not only achieve adequate length of the gastrocnemius–soleus complex to allow dorsiflexion in

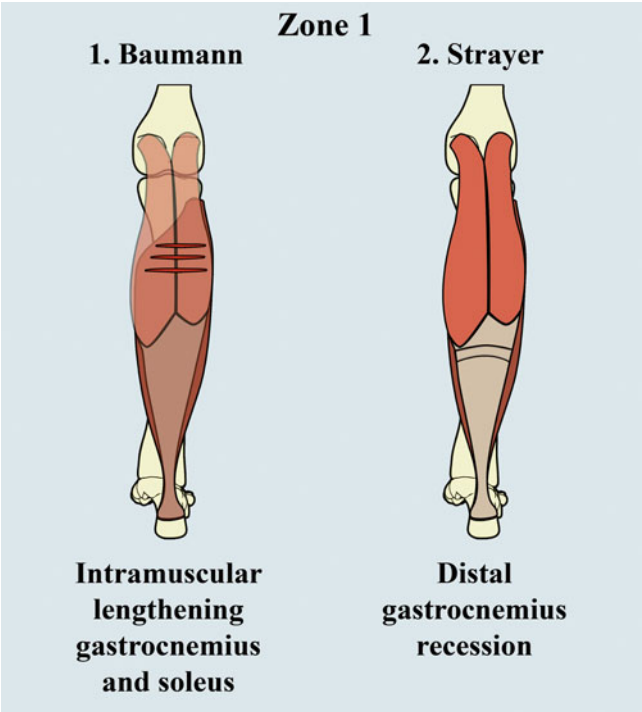


Fig. 11.3 Zone 1 level surgeries include the Baumann procedure and the Strayer procedure. Illustration created by and reprinted with permission of Daniel Biggers

stance phase, but also preserve adequate strength and moment generation to control advancement of the tibia over the foot [11]. The most devastating complication of surgery for equinus deformity is excessive weakness of plantarflexion with a calcaneus gait. Surgery for recurrent equinus is possible but there are currently no reliable methods with which to shorten an over-lengthened gastrocnemius–soleus complex [11]. Therefore it is better to err on the side of undercorrection especially in patients with cerebral palsy and follow the philosophy "a little equinus is better than calcaneus" [11]. Overcorrection may result in a severe crouch gait and the loss of walking ability [11].

Operations designed to lengthen the triceps surae can be assigned to one of three discrete anatomical zones. Zone 1 starts at the gastrocnemius origin and ends at the most distal fibers of the medial belly of the gastrocnemius [9]. The operative procedures in Zone 1 include the Baumann and the Strayer distal gastrocnemius recession (Fig. 11.3). The procedure described by Baumann involves intramuscular lengthening of the gastrocnemius and soleus performed in the interval between the gastrocnemius and soleus fascia [13]. The gastrocnemius may be lengthened selectively, or differential lengthening of the gastrocnemius and soleus may be performed by varying the number of cuts in the fascia overlying the two muscles. Herzenberg et al. found it more effective to make a second more distal incision in the gastrocnemius muscle rather than adding an incision in the soleus [14].

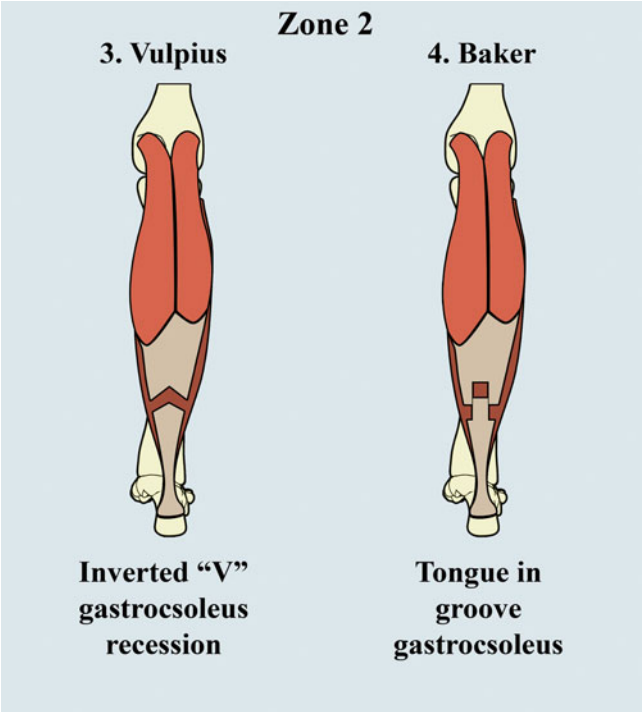


Fig. 11.4 Zone 2 level surgeries include the Vulpius and the Baker procedures. Illustration created by and reprinted with permission of Daniel Biggers

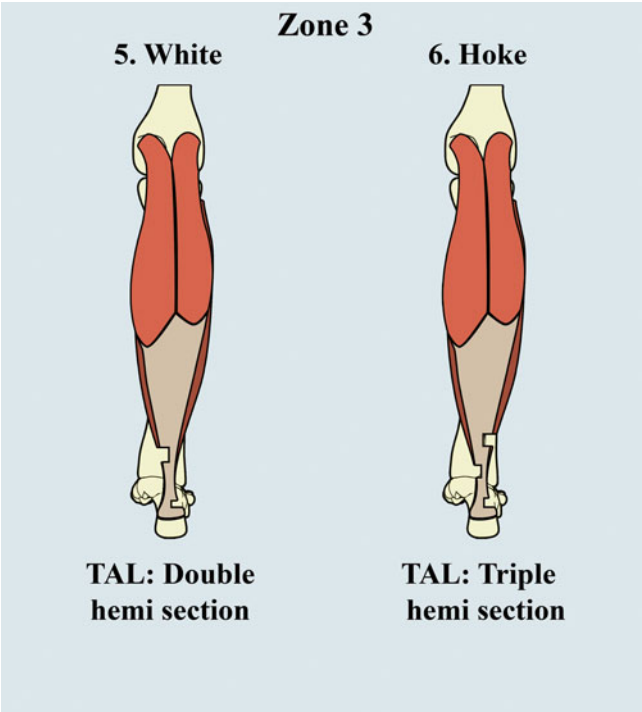


Fig. 11.5 Zone 3 level surgeries include the White and Hoke procedures. Illustration created by and reprinted with permission of Daniel Biggers

At the junction of Zones 1 and 2 it is possible to perform selective lengthening of the gastrocnemius alone, as described by Strayer [15]. It is also possible to perform differential lengthening of the gastrocnemius and soleus by separating the gastrocnemius fascia from the soleus fascia and dividing the gastrocnemius aponeurosis and soleus fascia separately in a technique known as the modified Strayer procedure [11].

Zone 2 extends from the distal end of the medial belly of the gastrocnemius muscle to the end of the soleus muscle [9]. This zone is characterized by condensation of the gastrocnemius fascia into a broad aponeurosis, which lies superficial to the soleus fascia. The layers are most accurately referred to as the conjoined gastrocnemius aponeurosis–soleus fascia [12]. The two Zone 2 layers can very rarely be separated and then only partially and by sharp rather than blunt dissection [12]. Zone 2 procedures include those of Vulpius and Baker and involve performing a gastrocnemius–soleus complex recession with the use of a single transverse division of both the gastrocnemius aponeurosis and the soleus fascia (Fig. 11.4). Neither selective nor differential lengthening of the two layers that comprise the conjoined tendon is possible in Zone 2 with these procedures [12]. The Zone 2 procedures are neither anatomically nor biomechanically equivalent to the Zone 1 procedures [12]. The Vulpius procedure is often grouped with the Strayer procedure and referred to as a gastrocnemius recession, when it is in fact a gastrocnemius–soleus complex recession [12].

Zone 3 extends from the end of the soleus muscle to the insertion of the Achilles tendon on the calcaneus [9]. The gastrocnemius aponeurosis and the soleus fascia provide two groups of fibers that are distinct at first, then rotate, and finally blend together within the substance of the Achilles tendon [12]. Zone 3 procedures include all forms of lengthening of the Achilles tendon, as well as heel cord advancement (Fig. 11.5). Zone 3 lengthening of the Achilles tendon should be used when there is a contracture of both the gastrocnemius and the soleus of greater than 30° and no improvement in ankle dorsiflexion is seen with knee flexion [11].

Achilles tendon lengthening may be performed by open or percutaneous techniques. In the open technique, a longitudinal incision is made just lateral to the Achilles tendon. The tendon is lengthened in Z-fashion and repaired with heavy non-absorbable suture. Percutaneous lengthening can be performed by either two or three transverse partial tenotomies. The first tenotomy is made proximal to the calcaneal insertion and a little more than half of the tendon is divided by inserting a scalpel longitudinally into the center of the tendon, turning the blade out toward the edge of the tendon, and pushing the tendon down onto the blade. The opposite half of the tendon is then divided by using a similar maneuver 3–4 cm more proximally. In a patient with a valgus hindfoot, the distal cut is through the lateral half of the tendon. In a varus hindfoot, the distal incision is through the medial half. After completing the incisions, the ankle should be dorsiflexed

gradually with the heel inverted. The foot should be dorsiflexed to neutral. It is important to verify the integrity of the tendon after a percutaneous lengthening is performed. The surgeon should squeeze the calf and simultaneously watch the ankle. If the ankle plantarflexes when the calf is squeezed, then the tendon can be presumed to be intact within the tendon sheath. If there is no plantar flexion, the tendon may have been ruptured by the lengthening procedure and an open repair will be needed.

The stability of the musculotendinous unit after a particular lengthening procedure as well as the amount of lengthening achieved are directly related to the residual cross-sectional area of the soft tissues remaining intact at the site of the lengthening [12]. For example, after a Baumann intramuscular tenotomy, all of the soleus and gastrocnemius muscle fibers, plus the intact fascia on the posterior aspect of the gastrocnemius and the anterior aspect of the soleus remain intact. This confers excellent stability and resists further lengthening of the musculotendinous unit [12]. The Baumann procedure appears to yield good results in children with spastic diplegia and mild equinus deformities with no reported overcorrections [16]. Zone 2 procedures achieve greater lengthening than Zone 1 procedures [12]. This may be related to the decreasing cross-sectional area of muscle at the site of the fascial division. In Zone 3, the stability of the musculotendinous unit after a percutaneous triple hemisection depends entirely on side-to-side links between the largely parallel collagen fibers of the tendon, which have been divided at three contiguous levels [12]. This may contribute to overlengthening and poor outcomes, especially in ambulatory children with spastic diplegia [12].

Therefore, there is no single surgical procedure that should be considered a panacea for all patients with equinus deformity. Rather, the history and physical examination will help guide the surgeon to decide which procedure may be more appropriate for each clinical situation. For example, in children with spastic hemiplegia and equinus deformity, the Silfverskiold test usually reveals a substantial fixed contracture of both the gastrocnemius and the soleus [12]. A Zone 2 lengthening (Vulpius) or a Zone 3 procedure (percutaneous hemi-section) might be selected, depending on the severity [12]. In contrast, a Zone 3 procedure in patients with spastic diplegia is associated with a very high prevalence of calcaneus deformity and crouch gait at long-term follow-up [12]. In these patients, the Silfverskiold test usually reveals either an isolated contracture of the gastrocnemius or a contracture of the gastrocnemius that is greater than that of the soleus [12]. Therefore, a Zone 1 procedure such as the Baumann or Strayer would be recommended. The type of cerebral palsy should be carefully considered when deciding on the type of surgical procedure and overall management of an equinus deformity [9].

The stability of the gastrocnemius–soleus musculotendinous unit after surgical lengthening may also be relevant to the decisions about postoperative care and rehabilitation protocols [12]. The intrinsic stability after Zone 1 or 2 lengthening may permit very early weight-bearing and relatively short periods of cast immobilization. The relative instability after Zone 3 lengthening and the documented tendency for continued lengthening under small loads may require longer immobilization and delayed weight-bearing [12].

A typical postoperative regimen after equinus surgery consists of a below-the-knee cast with the foot in a neutral dorsiflexion for 3 weeks. Unrestricted weight bearing beginning the first postoperative day is prescribed if a Zone 1 or Zone 2 procedure was performed. Casts are removed after 3 weeks to check healing and to cast molds for an ankle–foot orthosis. The standard postoperative ankle–foot orthosis is solid with hinges inserted but not activated [11]. Casts are reapplied and removed at 6 weeks after surgery where they are replaced by the ankle–foot orthoses.

The greatest risk after equinus surgery is recurrence and it correlates strongly with the age of the patient at surgery. Children with cerebral palsy who undergo heel cord lengthening at 7 years or younger are particularly at risk for recurrence [8]. Recurrence of equinus is also possible in cases of congenital disease, such as arthrogryposis, or with the loss of function of the anterior compartment muscle(s), as in severe burn cases, poliomyelitis, or nerve injury [6]. Recurrence in posttraumatic equinus is rare as long as the anterior tibial muscles are functional. Recurrent equinus can be corrected with a Vulpius gastrocnemius–soleus complex recession in Zone 2 [12]. In some cases of repeated lengthening the Achilles tendon becomes scarred and adherent, so the subsequent lengthenings must be done using the open technique.

When performing a lengthening the surgeon must be careful not to position the foot in excessive dorsiflexion. Overstretching of the tendon can lead to excessive length and calcaneus gait. Calcaneus can also occur after lengthening if there is an unrecognized flexion contracture of the knee. If the knee remains crouched and the Achilles tendon becomes longer, the ankle will drop into excessive dorsiflexion during stance phase and create an inefficient gait pattern.

External Fixation

While mild to moderately severe equinus contracture can be addressed with soft tissue lengthening, severe equinus deformities often cannot be corrected with a single open surgical procedure because the skin and neurovascular structures may not tolerate such an acute stretch. The Ilizarov method of gradual correction can be used in these situations. The risk of a neurovascular stretch injury during treatment can be further reduced by decompressing the posterior tibial neurovascular bundle at the tarsal tunnel during frame application,

especially in the presence of any associated varus deformity [4]. If any neurologic compromise occurs during the gradual distraction, the rate of the distraction can be slowed or temporarily stopped to allow for neurological recovery. Nerve decompression should be considered when nerve function does not improve or if any new motor deficit is noted [4].

In addition to providing a method for correction of severe contractures, external fixation has several other advantages. During external fixation treatment there will be access to the soft tissues for wound care, especially in the presence of abnormal soft tissues such as scars, burns, and compromised skin [4]. External fixation also allows for adjusting the rate of deformity correction during the postoperative period and can allow for joint range of motion and early weight bearing [4].

All ankle external fixator constructs need to allow for distraction of the ankle joint space prior to beginning the deformity correction. This should be performed at the time of frame placement. In a simple equinus correction, the ankle should be distracted 2–5 mm compared with preoperative radiographs [6]. This limits cartilage compression of the ankle joint and any rockerbottom deformity that may occur as the ankle is brought into dorsiflexion [6]. The distraction of the hindfoot, however, must be done in a posterior-inclined direction. If distraction is performed in a purely axial direction, parallel to the tibia, the talus will tend to sublux anteriorly [6]. Fixation should be applied to both the talus and the calcaneus to prevent overdistraction of the subtalar joint during the correction of the tibiotalar joint contracture [4].

Types of External Fixation Constructs

There are three different types of external fixator constructs that may be used to correct equinus contractures depending on the hinge system used: constrained, unconstrained, and virtual. The constrained fixator has a single hinge centered on the axis of rotation of the ankle (Fig. 11.6). The normal trans-malleolar axis is 80° in the coronal plane and 84° in the sagittal plane [6]. Therefore, the axis created by the hinges is positioned on a slightly oblique plane with respect to the horizontal from the tip of the lateral malleolus to just distal to the tip of the medial malleolus [6]. Anterior subluxation of the talus can be avoided by carefully positioning the hinges slightly inferior to the axis of the ankle by 4–5 mm [6]. This produces a counter-push on the foot during correction and directly opposes anterior subluxation of the talus [6]. The constrained fixator allows for range of motion of the joint during contracture correction.

An unconstrained or flexible frame can be used for simple, unidirectional deformities with no associated bony deformities [6] (Fig. 11.7). Frames for the correction of a simple equinus contracture require much less rigidity than those for bony instability. Consequently, this construct consists of only a tibial ring block, a calcaneal half ring, and a metatarsal half ring [6]. The use of a footplate or connecting bars between the calcaneal and metatarsal half rings is usually not needed for simple equinus correction. A single olive wire is placed in the calcaneus and in the metatarsals and attached to the respective half ring. The calcaneal wire should be placed relatively

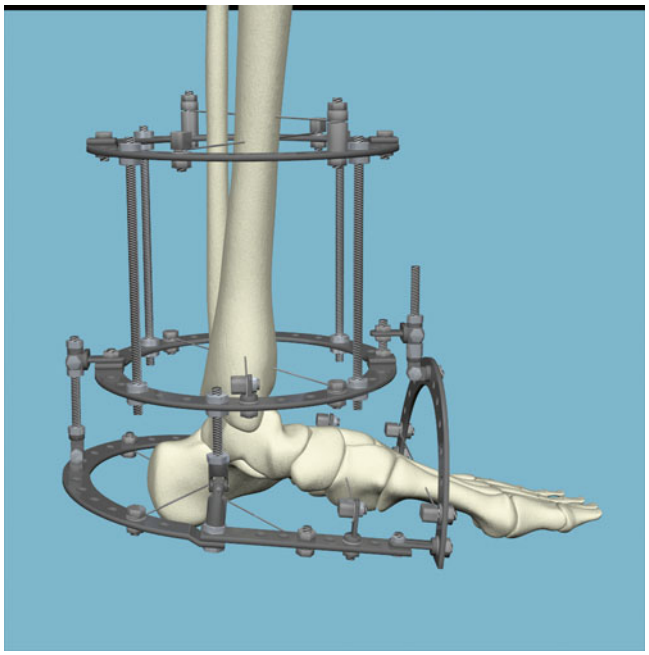


Fig. 11.6 An example of a constrained external fixator with hinges placed at the center of rotation of the ankle. Illustration created by and reprinted with permission of Daniel Biggers

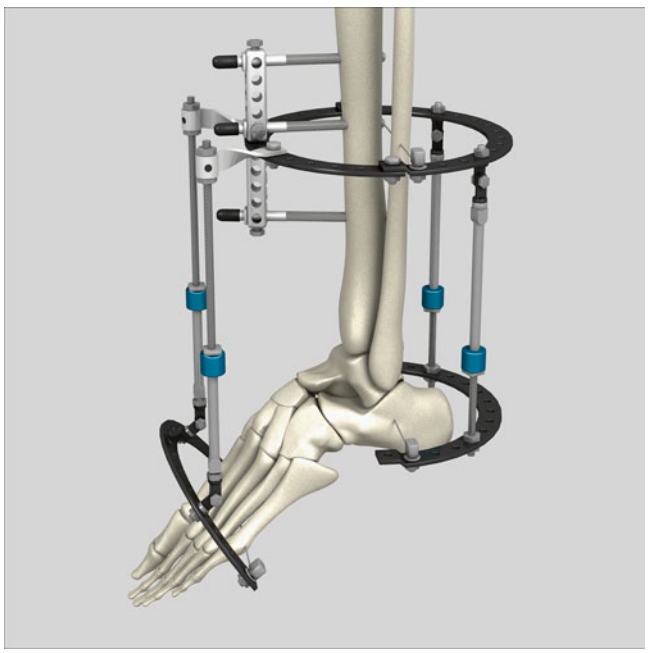


Fig. 11.7 An example of an unconstrained external fixator for correction of an equinus deformity



Fig. 11.8 An example of a Taylor Spatial Frame™ used for correction of an equinus deformity. The lateral process of the talus is chosen as the origin. Illustration created by and reprinted with permission of Daniel Biggers

proximally and posteriorly to prevent wire cutout and to increase its biomechanical advantage [6]. Instead of building a hinge at the center of rotation like the constrained frame, the unconstrained frame allows rotation to occur naturally at the tibiotalar joint as the correction progresses.

The Taylor Spatial Frame™ (Memphis, TN) allows the surgeon to correct the equinus deformity around a virtual hinge (Fig. 11.8). The lateral process of the talus is generally chosen as the "origin." In severe contractures, the tibial ring can be mounted obliquely to create a larger space posteriorly for the struts to fit. The tibiotalar joint can be distracted acutely with the frame in the operating room or the distraction can be built in to the deformity program. It is also possible to prevent anterior subluxation of the talus by accounting for it during data input in the deformity software program. A typical comprehensive spatial frame deformity program will define the deformity as plantar flexed (amount of equinus to correct), short 5–10 mm (to distract the tibiotalar joint), anteriorly translated (to prevent anterior talar subluxation), and slightly internally rotated (the ankle naturally externally rotates as it dorsiflexes).

Postoperatively, deformity correction with the external fixator is started as soon as the patient can tolerate it comfortably. Weekly or biweekly postoperative radiographs should be taken to follow the deformity correction and to ensure that the ankle remains distracted 2–5 mm without any subluxation [6]. The equinus deformity should be overcorrected by 10°–15° if possible and held with the fixator for a minimum of 4–6 weeks further to allow for soft tissue adaptation [6]. The corrected position should be maintained with a cast for approximately 4 weeks after frame removal and with functional bracing thereafter. However, once the Ilizarov fixator is removed the deformity may recur, especially as the child grows [17]. Increasing the duration of time in the fixator does not seem to prevent recurrence [17]. Any motor imbalance should be identified and addressed with tendon transfer or bracing to try to prevent recurrence. The parents and patients should be made aware that additional procedures are most likely going to be necessary to maintain the foot in a plantigrade position as the child gets older.

Box 11.1. Equinus Deformity

- Recurrence is common after equinus surgery especially in children under age 7 years.
- In an external fixation construct for equinus correction, distraction must be performed first to prevent impaction of the tibia and talus.
- Watch for anterior subluxation of the talus during correction of equinus with an external fixator.
- The surgeon should choose the appropriate zone for soft tissue lengthening based on the physical examination, the underlying diagnosis, and the amount of equinus contracture present.

Knee Flexion Contracture

Etiology

Knee flexion contractures commonly develop in children with neuromuscular disease such as cerebral palsy. They are also associated with congenital deformities of the lower extremities such as congenital femoral deficiency, tibial hemimelia, congenital dislocation of the patella, arthrogryposis, popliteal pterygium syndrome, sacral agenesis, and certain skeletal dysplasias [18]. Knee flexion contractures are considered to be more disabling than knee extension contractures [19]. As little as 5° of extension loss can create a noticeable limp, strain the quadriceps muscle and lead to patellofemoral pain [20]. Murray and Fixsen found that only 46 % of their patients with arthrogryposis and knee flexion contractures became community ambulators whereas 89 % of the children with extension contractures did [21]. Unlike hip and ankle contractures, the other lower extremity joints or the lumbar spine cannot compensate for knee flexion contractures [22]. Hoffer et al. found that a knee flexion contracture of greater than 20° was incompatible with functional ambulation [23].

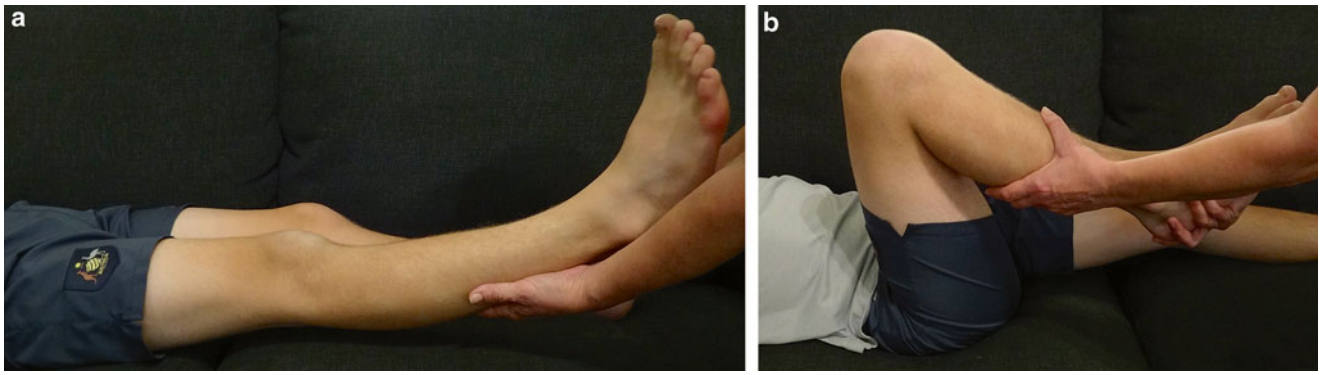


Fig. 11.9 Examination of the knee should evaluate maximum knee extension and maximum knee flexion

Biomechanics

In children the hinge point of the knee is defined as the intersection of the posterior femoral cortex and the physis or physeal scar [24]. Normally, the mechanical axis of the limb will pass slightly anterior to the hinge point of the knee allowing the knee to lock when the lower limb is fully extended [24]. This permits the quadriceps muscle to rest during prolonged standing [24]. In a knee flexion contracture, however, the center of gravity falls posterior to the knee. This arrangement places increasing demand on the quadriceps to resist the progressive crouch and energy expenditure increases during a crouched gait. This constant activation of the quadriceps muscle will result in fatigue and increases the oxygen cost during such pathologic gait [25]. Knee flexion contracture also functionally shortens the limb creating a gait disturbance. Consequently, a fixed knee flexion deformity exceeding 10° can lead to anterior knee pain, decreased endurance, patella alta, and progressive crouch gait in ambulatory patients [25]. In wheelchair users, knee flexion contracture may impair standing, transfers, and activities of daily living.

Clinical Evaluation

The physical examination for knee flexion contracture should focus on observation of the gait pattern from both the frontal and sagittal plane. A typical knee flexion contracture gait pattern will include greater than normal knee flexion during midstance and an inability to extend the knee fully at the end of swing phase continuing into initial contact [18]. Walking will also exhibit decreasing cadence and step length. A focused examination of the knee should include an assessment of the maximal knee extension, maximal knee flexion and the total arc of motion present (Fig. 11.9). As the range of motion is evaluated, it is important to determine if there is firm or soft endpoint to maximal knee extension. A flexion contracture of the knee with a soft endpoint will likely have an improvement from soft tissue procedures [22]. The stability of the knee should be tested and the function of the quadriceps evaluated. One relative contraindication to surgical treatment is the absence of active quadriceps function because of the high likelihood of recurrence of the flexion deformity [18, 26]. The flexibility of the hamstring muscles must be assessed and can be measured via the popliteal angle. This angle is created first by positioning the patient supine on the examining table, flexing the hip to 90° and then maximally extending the ipsilateral flexed knee. The acute angle between the vertical and maximal extension of the tibia is the popliteal angle. There is variability in measured popliteal angles with a mean value of 26° in normal children 4 years and older [18]. The examining physician must carefully assess all of the lower extremity joints for strength, spasticity and contracture. While tight hamstrings are generally the cause of a crouch knee gait, quadriceps and gastrocsoleus weakness may also lead to excessive knee flexion in stance phase [18]. A bent-knee gait may also be a compensation for equinus and toe-walking if the popliteal angle is normal [18]. The hip must also be examined because correction of hamstring contractures without addressing concomitant hip flexion contractures will lead to increased hip flexion and forward lean during gait [18]. Furthermore, distal femoral or proximal tibial deformities may create or exaggerate a knee flexion deformity. In patients with skeletal dysplasias, for example, the knee flexion deformity may be the result of bony or articular cartilage abnormalities rather than any soft tissue tightness [18]. Patients with congenital knee flexion contractures that have never reached full extension positioning may develop compensatory deformities of the distal femur and proximal tibia [19]. Syndromic and congenital contractures such as arthrogryposis are also occasionally associated with lateral displacement of the extensor mechanism and the mechanical axis [26]. Each particular problem needs to be recognized and addressed at the time of surgical correction in order to prevent recurrence.

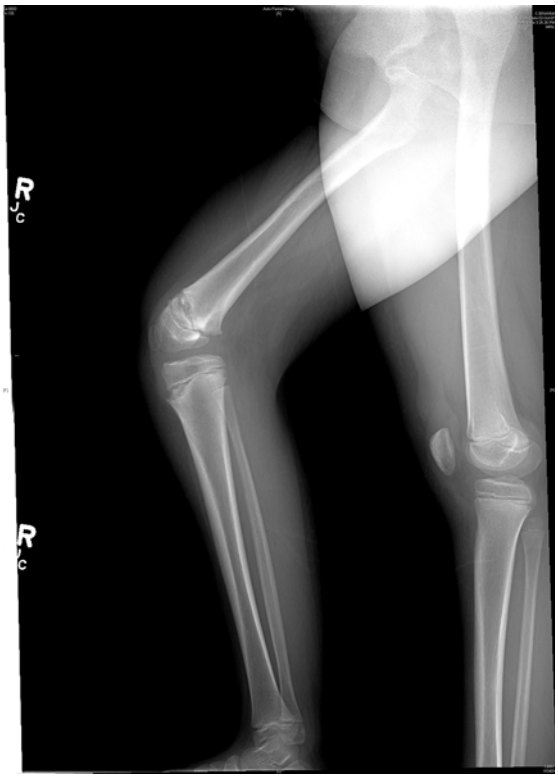


Fig. 11.10 Lateral radiograph of the lower extremity demonstrating a knee flexion contracture. The image was taken with the knee in maximum extension

Imaging

The radiographic evaluation of a knee flexion contracture requires modification of the usual anteroposterior radiographs of the femur, tibia, or knee. Because of the flexed position of the knee these views can be difficult to obtain. Similarly, long leg anteroposterior films are rendered meaningless by the flexed knee [22]. Obtaining individual tibia and femur posteroanterior radiographs with the child in the prone position is another method to overcome such obstacles in positioning the extremity for radiography [22]. Lateral radiographs with the knee in maximum flexion and extension may help define the knee's range of motion and demonstrate any tendency towards tibial hinging or subluxation with changes in the arc of motion [22]. In long-standing knee flexion contractures with reduced pressure and motion at the joint, the lateral radiograph will help identify any compensatory changes of the proximal tibia. The resulting dome shaped epiphysis can cause posterior tibial subluxation during deformity correction [19].

The most meaningful way to evaluate the lower extremity sagittal plane alignment is with a full-length standing lateral view radiograph of the lower limb with the knee joint maximally extended [24] (Fig. 11.10). This view allows evaluation of the mechanical axis of the lower extremity, which is defined as a line connecting the center of the femoral head to the center of the ankle. In a normal situation the mechanical axis should pass slightly anterior to the hinge point of the knee. If the lower extremity has fixed flexion deformity at the knee the mechanical axis will pass posterior to the hinge point of the knee. The anterior cortical lines of the distal femur and proximal tibia should also be measured [24] (Fig. 11.11). These lines represent the overall position of the knee joint and will determine if the knee joint is in a neutral position or a fixed flexion position. In a normal limb the anterior cortical lines of the femur and tibia should be collinear [24]. If these lines are not collinear, then the angulation between the anterior cortical lines of the distal femur and proximal tibia should be measured to quantify the total amount of flexion deformity. On the lateral radiographs, the distal femoral and proximal tibial joint orientation lines as measured by the posterior distal femoral and posterior proximal tibial angles should be drawn [24]. This analysis helps determine if a deformity is entirely the result of a contracture or if it has a bony component. The comparison of the amount of deformity determined by the joint analysis and the amount of deformity denoted by the anterior cortical line measurement allows the soft tissue component of the deformity to be determined. If the degree of deformity measured by the anterior cortical lines is not equal to the degree of deformity determined by the joint analysis, then a soft tissue component is present [24]. Multiple combinations of bone deformity and soft tissue contribution can result in the same clinical deformity.

Decision Making and Management Strategy

Decision Making

Knee flexion contractures can occur in combination with other lower extremity deformities. In limbs with a knee contracture and a coronal plane deformity of the femur or tibia it can be difficult to differentiate the contribution of each to the total limb malalignment [22]. As noted earlier, flexion contracture of the knee prevents proper positioning of the limb for radiographic analysis making it challenging to accurately measure the magnitude and location of the bone deformity. In these cases, it is easier to correct the knee flexion contracture first and then, once the knee is straight, corrective osteotomies can be accurately planned [22]. Hip deformities and contractures create additional obstacles to correcting knee contractures, maintaining correction, sitting, and ultimately ambulating after the correction. Hip flexion contractures greater than 30°, extension contractures, and rotational abnormalities that prohibit positioning the knee directly anteriorly need to be addressed prior to knee contracture correction [22]. If the knee is treated first, it will be difficult to maintain the knee correction while treating the hip.

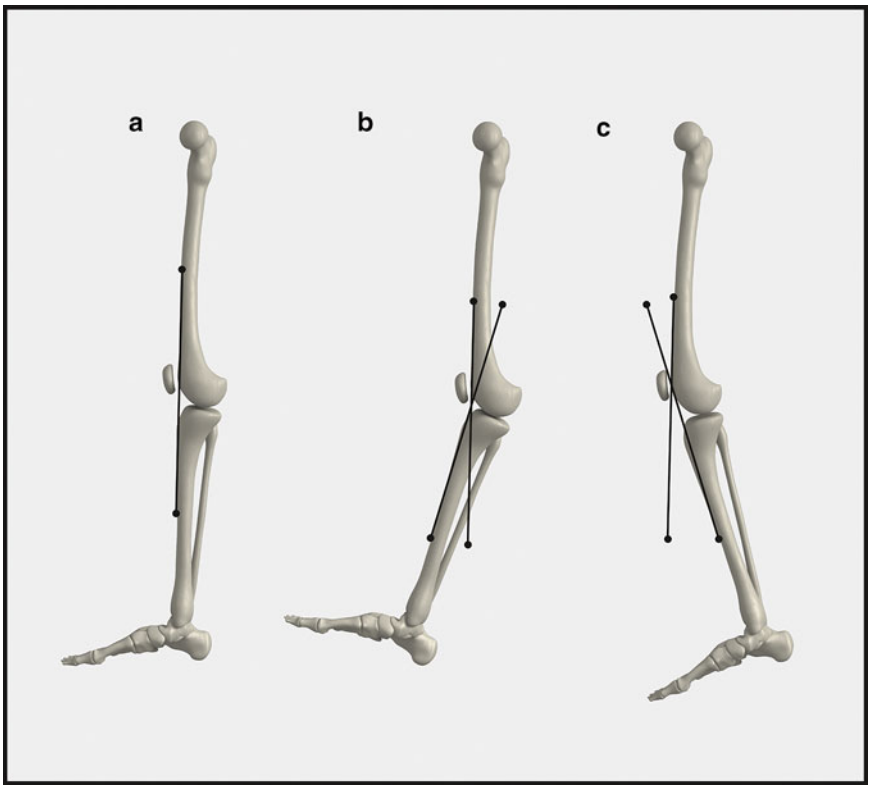


Fig. 11.11 In normal alignment, the anterior cortical lines of the distal femur and the proximal tibia should be collinear. If the lines are not collinear, then the angle created represents the magnitude of the extension or flexion contracture present. Illustration created by and reprinted with permission of Daniel Biggers

Nonoperative

In most cases of knee flexion contracture, an initial attempt at nonoperative management should be considered. In general, patients with relatively normal hip and ankle function can tolerate a knee flexion contracture of less than 20° [22]. The quadriceps' function is bolstered when the body's center of gravity is brought anterior to the knee by leaning forward with the hip locked in extension and using the plantar flexors to stabilize the ankle. Nonoperative methods include stretching by physical therapy, serial casting, hinged cast bracing, adjustable splinting, skin traction, dynamic spring-loaded splinting, botulinum toxin A, and bracing. These methods are generally successful in patients with mild flexion contractures, or are used to complement posterior soft-tissue knee release to gradually finish the correction. Bracing with knee–ankle–foot orthoses or floor reaction ankle–foot orthoses may improve gait efficiency if there is quadriceps or ankle weakness [19]. The posterior push on the knee from the brace in stance phase can improve mild dynamic knee flexion contractures [18]. Mild contractures can also be managed by serial casting. A serial casting protocol of weekly sequential wedging demonstrated that patients who were less than 12 years old and had an initial flexion contracture of less than 15° were likely to maintain correction 1 year postoperatively [27]. Dynamic splinting, which delivers low torque, prolonged duration stretching was found to improve active range of motion by an average of 23° in a review of eight studies in the literature [28]. The major disadvantage with these methods, however, is the amount of corrective force that may be applied with casts, splints, and braces is limited by the low tolerance of the skin to direct pressure [29]. Additionally, the knees must be carefully monitored for the development of posterior tibial subluxation and recurrence rates are high. Two recent reviews of the literature reached similar conclusions that stretching does not have clinically important effects in joint mobility, pain, spasticity, or activity limitation in people with, or at risk of, contractures [30, 31].

Operative

Few patients with clinically significant knee flexion contracture can be managed definitively with nonoperative techniques. If conservative treatment does not prevent the progression of fixed knee flexion deformity, then surgical options should be considered. The type of surgical intervention chosen should be based on the cause of the contracture, its magnitude, its duration, the presence of tibiofemoral subluxation, neurologic involvement, and the patient's ability to participate in treatment [29]. The most common surgical options include soft tissue releases, anterior distal femoral epiphysiodesis, extension osteotomy of the distal femur, and arthrodiastasis using an external fixator. Prerequisites for any knee flexion contracture surgery include the absence of a concomitant hip flexion deformity of more than 15° and the presence of adequate quadriceps and gross neurologic function of the affected limb.

Soft Tissue Releases

Soft tissue release procedures described in the literature may take different forms: distal hamstring tenotomy or lengthening, proximal hamstring release, or hamstring tendon lengthening combined with posterior knee capsulotomy. Hamstring lengthenings are indicated when the popliteal angle is greater than the flexion contracture [22]. In children, knee flexion contractures of less than 20° are often responsive to hamstring lengthening alone [22]. However, these soft tissue releases may not be sufficient to gain full correction in a more severely contracted knee. In this situation, the long-standing altered resting length of the popliteal vessels and sciatic nerve may prevent full correction safely [29]. Neurovascular complications are more common if full extension is performed immediately at surgery in order to immobilize the patient in a straight cast. The surgeon should be careful not to aggressively stretch the posterior structures of the knee or a nerve palsy of all or part of the sciatic nerve can occur. The sciatic nerve does not appear to tolerate elongations, especially when abruptly applied, of more than 5–12 % of its original length without injury [32]. Hip flexion with the knees extended in casts can further stretch the sciatic nerve [18]. If a nerve palsy is recognized, the cast should be immediately removed and the knee allowed to flex. Postoperative serial casting requires vigilance. The surgeon must also be wary of any developing skin necrosis, joint cartilage compression, or joint subluxation that may occur while serial casting [29].

For children with flexion contractures of the knee, proximal hamstring release is usually not recommended as the primary soft tissue procedure as it can cause anterior pelvic tilt and hyperlordosis of the lumbar spine [18]. Most surgeons prefer to perform distal hamstring releases instead. A typical release pattern includes an intramuscular aponeurotic lengthening of the semimembranosus, a Z-lengthening of the semitendinosus, and either a tenotomy or a Z-lengthening of the gracilis. When lateral hamstrings are included in the procedure, an intramuscular aponeurotic lengthening of the biceps femoris is recommended. Usually two separate transverse cuts are needed in the fascia of the semimembranosus and biceps femoris to obtain adequate lengthening [18]. Medial hamstring lengthening alone is usually sufficient in patients with mild crouch and moderately increased popliteal angles. The addition of lateral hamstring lengthening will improve maximum knee extension during stance phase. This is potentially helpful in patients with more severe crouch but it also increases the risk of developing anterior pelvic tilt [18]. If there is a hard end point to the range of motion in extension then the addition of a posterior capsulotomy may be indicated.

Correction of knee flexion contractures greater than 60° usually involves additional complete posterior capsular release with or without femoral shortening [18]. The femur must be shortened to decompress the posterior neurovascular structures. In most cases, the femur can be safely shortened acutely by 3 or 4 cm. In these large, long-standing contractures, the surgeon should be aware that there might also be a fibrous adhesion in the anterior part of the knee that blocks full extension [18].

After hamstring lengthening, a knee immobilizer can provide sufficient immobilization for 3 or 4 weeks. If there is any residual contracture, a long-leg cast is applied. Early weight bearing and ambulation are encouraged. The greatest improvements in the knee flexion contracture are seen within the first year after surgery [18]. However, in a growing child recurrence over time is common, with the return of the flexion deformity estimated at around 3° per year [32].

Bony Correction Options

An alternative concept to the soft tissue releases for knee flexion contracture is to alter the alignment of the distal femur to create knee extension. Two methods for achieving this correction have been described: distal femoral extension osteotomy and guided growth with anterior distal femoral hemiepiphysiodesis. Extension osteotomies of the distal femur are indicated for children with contractures no greater than 45° [22] (Fig. 11.12). Although larger contractures have been treated with extension osteotomies, there is increasing risk of a neurovascular stretch injury. While shortening of the femur helps to protect the neurovascular structures, it exacerbates the limb shortening already caused by the angular osteotomy. While the extension osteotomy can produce a straight knee there are several concerns with this technique. Distal femoral extension osteotomies does create a secondary deformity (angulation and shortening) instead of correcting the primary soft tissue deformity and may lead to abnormal joint-loading forces on the posterior femoral condyles in the ambulatory patient [22, 29, 32]. Because of rapid bone remodeling in the younger child, extension osteotomies have a high recurrence rate and should probably be delayed until after 12 years of age [19].

Guided growth treatment using an anterior distal femoral hemiepiphysiodesis is another option for fixed knee flexion deformity exceeding 10°. Younger patients (<12 years old) have demonstrated the best results with this technique and correction of fixed knee flexion deformity up to 40° has been accomplished [25]. Guided growth using tension-band plates appears to have a faster correction rate and fewer complications than staples [25, 33]. Contraindications for guided growth treatment include less than 12 months of predicted growth remaining and flexion deformities that are caused by a dynamic etiology [25]. This technique does not seem to impair knee flexion or knee stability, which is a possible advantage over the soft tissue releases. However, because the hemiepiphysiodesis correction is gradual, it may take a few years to obtain a meaningful correction in large contractures, which may not be a suitable time frame for some patients.

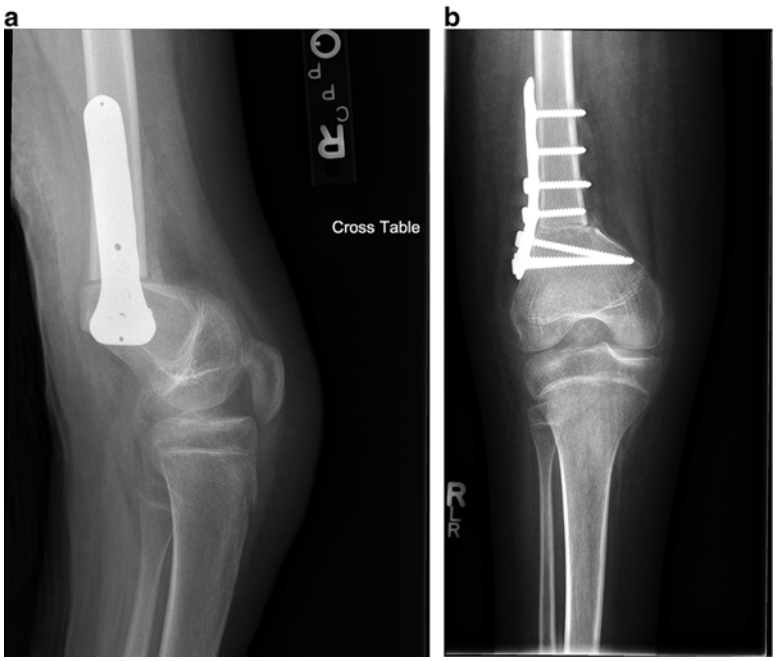


Fig. 11.12 Distal femoral extension osteotomy stabilized with a locking plate to correct a knee flexion contracture (case provided by Steven Frick, MD)

External Fixation with Gradual Soft Tissue Correction

Gradual extension of knee flexion contracture with circular or monolateral external fixation can be used on essentially any magnitude of contracture severity. This option has many potential advantages especially in managing patients with severe flexion contractures. Acute correction of long-standing severe contractures risks stretching the sciatic nerve and its branches and can create large skin defects posteriorly. Hypertension caused by the sudden stretching of the sympathetic nerves located around the blood vessels can also occur [32]. In these situations, the sciatic nerve may be more tolerant of gradual correction and skin problems may be more easily managed with external fixation. With external fixation, tibial subluxation can be addressed during contracture correction and no femoral shortening is required, even with large deformities [22]. In flexion deformities secondary to popliteal pterygium, the contracted skin and fascia can be gradually stretched. The rate of contracture distraction can be decreased as necessary to protect soft tissue and neurovascular viability.

In external fixation of knee flexion contractures, the fixator should be applied with its hinge carefully positioned on the knee joint center. The true center of knee rotation can be approximated running through the origins of the medial and lateral collateral ligaments [22]. Under image intensification in the operating room, the knee is positioned with the posterior and distal femoral condyles overlapping in the lateral projection. When the knee is too immature to visualize the femoral condyles well, an arthrogram may help better visualize the femoral condyles [19]. The knee joint center of rotation can be identified at the intersection of a line along the posterior femoral cortex and the physis or physeal scar [24] (Fig. 11.13).

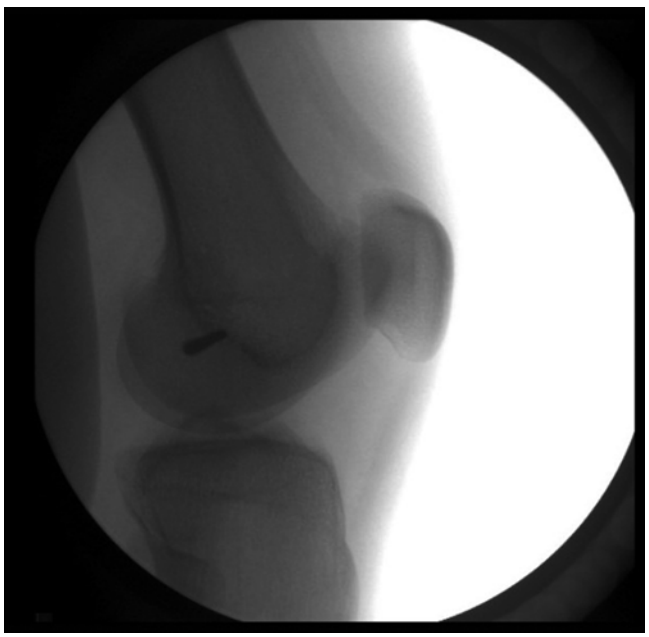


Fig. 11.13 The center of rotation of the knee can be identified at the intersection of the posterior femoral cortex line and the physis (or physeal scar)

When using a Taylor Spatial Frame™, this point can be chosen as the "origin" (Fig. 11.14). It is important to avoid placing the hinge too posterior or too proximal to the center of rotation [22]. Posterior placement will aggravate the tendency for the tibia to sublux posteriorly and proximal placement will lead to anterior tibiofemoral impaction in full extension.

Patients with known preoperative subluxation of the tibia on the femur should first have reduction of the subluxation

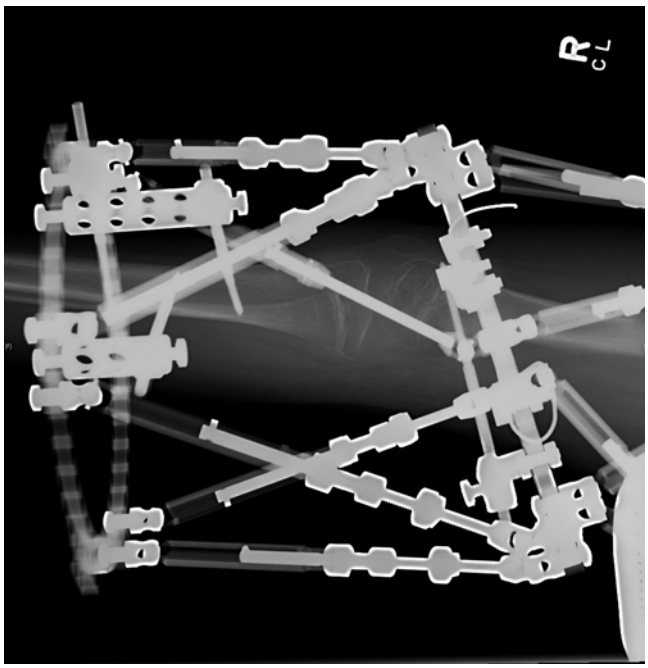


Fig. 11.14 External fixation used to gradually correct a knee flexion contracture (same patient as in Fig. 11.10)

with the external fixator followed by distraction and gradual extension of the knee [22]. To help prevent tibial subluxation from occurring during the correction process, an arch wire technique can be used in the proximal tibial ring so that as the wire is tensioned it pulls the tibia slightly anterior on the femur [22]. In children with severe flexion contractures, it may not be possible to use full rings on the femur and the tibia, as they will impinge on one another posteriorly. A half or two-thirds ring can be used as a drop ring either in addition to or as substitution for the periarticular full rings [22].

There is some debate as to whether soft tissue release is necessary before attempting correction with an external fixator but most surgeons agree that distraction of the knee joint with the external fixator is mandatory prior to correcting the deformity. This initial distraction of 5–10 mm is important to avoid impingement and articular cartilage damage during correction [18, 19, 22, 26, 32].

The usual rate of correction with the external fixator is 1°–2° per day although this can be adjusted based on the patient's tolerance [22, 32]. Stiff knees require a slower rate of correction to reduce the chances of a periarticular fracture [22]. The external fixator construct should have a removable distractor to allow the patient to move the knee during correction treatment although such mobilization of the knee is not always well tolerated by the patients [19, 29]. Once the correction is completed, it is recommended that the frame be maintained at full extension for an additional few weeks to allow the soft tissues to adapt to the new position. The frame is removed in the operating room and the leg is measured for a knee–ankle–foot orthosis with locking knee hinges [22]. The extremity is then placed in a long leg cast with the knee in full extension, with a mold applied to prevent posterior tibial subluxation. The cast is removed in approximately 2 weeks and full-time knee–ankle–foot orthosis use is begun immediately. Physical therapy to regain knee motion is generally started at this time. After gradual correction with an external fixator, there is usually a loss of knee range of motion compared to the preoperative total arc of motion [29]. This range, however, has been redirected to a more useful and functional arc compared to preoperatively. Fractures are a potential complication when using external fixation, especially in the proximal femur. The surgeon should be aware of any limitation of hip range of motion that could lead to excessive stress on the femur with positioning. Fracture risk can be minimized by using half pins no larger than 33 % of the diameter of the bone, by avoiding unicortical pins, and by avoiding tight clusters of metaphyseal pins [19, 22].

Recurrence of deformity is still a frequent clinical problem after any surgical treatment of knee flexion contractures in children. Such procedures are best at redirecting the arc of motion but rarely increase the total arc of motion [18, 29]. Weak or absent quadriceps power has been identified as an important factor leading to recurrence [26]. Pediatric contractures can recur due to growth or from remodeling of extension osteotomies [22, 32]. In very stiff contractures, damage to the epiphysis can occur from excessive pressure between the femur and tibia when the knee is extended [18]. In some cases of recurrent severe knee flexion contracture despite multiple surgeries, amputation with prosthetic fitting may be appropriate. If there is severe limb length inequality, knee arthrodesis and a Symes amputation may provide a more satisfactory result than multiple failed attempts at correction [18].

Box 11.2. Knee Flexion Contracture

- The other lower extremity joints and the lumbar spine cannot compensate for a knee flexion contracture.
- Prone radiographs of the femur and tibia allow evaluation of the coronal plane in patients with knee flexion deformity.
- The standing full-length lateral view radiograph is critical to evaluate the sagittal plane alignment of a patient with a knee flexion contracture.
- Over-aggressive stretching of the knee into extension can cause a neurovascular injury to the limb after soft tissue releases.
- Correction of the knee flexion contracture with an external fixator should begin with distraction of the knee joint.

Knee Extension Contractures

While some joints such as the wrist or ankle can tolerate significant loss of motion and still be compatible with reasonable function, the knee is much less forgiving [34]. Knee stiffness can become so disabling that it limits the patient's ability to perform activities of daily living. Knee extension contractures are far less common than flexion contractures. During gait, extension contractures of the knee cause swing-through dysfunction as well as dampen the small flexion moment in mid-stance and thus make it difficult to walk efficiently [22]. Knee extension contractures also create problems with sitting comfortably.

Etiology

Common causes for knee extension deformity include scarring or fibrosis of all or part of the quadriceps after trauma, adhesions from intra-articular knee surgery and quadriceps tightness after femoral lengthening. The anatomic conditions that cause a loss of knee flexion can be both intra-articular (fibrosis and shortening of the medial and lateral para-patellar retinaculum, adhesions from the deep surface of the patella to the femoral condyles) and extra-articular (fibrosis of the vastus intermedius with adherence to the rectus femoris muscle and to the front of the femur, shortening of the rectus femoris, adherence of the quadriceps to fracture callus or regenerate bone, adherence of the skin to the underlying muscle, and pin site tethering on the lateral side of the femur) [34, 35].

Clinical Evaluation

The physical examination is similar to that already presented for knee flexion contractures. The physician needs to evaluate the arc of motion present and the stability of the knee, including the patella.

Imaging

Radiographs of the knee are recommended to search for any distal femoral or proximal tibial deformities that may create or exaggerate the knee extension deformity. The physician should search for any underlying lateral femoral condylar hypoplasia and genu valgum in patients with congenital femoral deficiency [36]. This finding can put the patient at risk for developing an obligate patellar dislocation with knee flexion.

Decision Making and Management Strategy

Nonoperative

Knee stiffness is expected to occur to a certain degree during femoral lengthening. Physical therapy focused on maintaining motion during the distraction and the consolidation phase is standard practice. However, certain patients do not respond to physical therapy [36]. The minimum acceptable amount of knee flexion for walking is 70° but this is not sufficient for normal daily activities and a minimum of 110° is preferable in young patients [34]. Patients may be considered candidates for surgery when they reach a plateau in gaining flexion during their physical therapy and when flexion remains less than 90° [34]. Patients who fail to improve knee range after at least one manipulation under general anesthesia should also be candidates for surgery [34].

Manipulation under anesthesia has been shown to improve the range of motion in patients with knee stiffness [37–39]. In patients with less than 90° of knee flexion, this technique can be used. It is designed to forcefully overcome the adhesions in the knee with the patient under general anesthesia and adequate muscle relaxation. The physician should flex the patient's ipsilateral hip to 90° and apply progressive force to the knee joint while holding the leg close to the knee. This should be done gradually and without excessive force. Audible and palpable lysis of the adhesions is often noted and the knee should develop an appreciable increase in motion. As more adhesive tissue accumulates in the knee, the chance of a successful manipulation decreases over time. Therefore, most authors recommend performing the manipulation within 3–4 months from the index surgery. Complications from the manipulation are related to the force needed to manipulate the knee and include fracture, hematoma, rupture of the extensor mechanism and wound dehiscence. The physician should be cognizant that skeletally immature patients are especially at risk for distal femoral physeal fractures while undergoing manipulation of the knee [38].

Operative

Quadricepsplasty is the term applied to describe surgical procedures to the quadriceps muscle designed to improve knee flexion [34]. Numerous techniques have been developed to treat knee extension contractures including the Thompson quadricepsplasty, the Judet quadricepsplasty, and their modifications [34–36, 40–45]. The literature suggests that the range of motion achieved intra-operatively may be the most important determining factor for the final gain of knee flexion [35]. Despite the improvement in knee range of motion that these procedures achieve, quadricepsplasty can create new problems such extensor lag, infection and iatrogenic injuries. Careful patient selection is paramount because these procedures require intensive postoperative therapy [34].

Patient and family compliance and motivation are essential for a satisfactory outcome.

Many extension contractures of the knee have both intra- and extra-capsular components that need to be addressed. Para-patellar incisions provide access to the scarred areas, patella, and distal quadriceps mechanism. Intra-capsular releases involve incising the lateral and medial retinaculum, the supra-patellar pouch, and any intra-articular adhesions. Extra-capsular contracture releases require extra-periosteal release of the quadriceps off the femur and the lateral intermuscular septum. Tethering between the skin and quadriceps muscle should also be released.

The Thompson technique, described in 1944, is based on isolating the rectus femoris completely from the vasti [40]. This involves stripping the rectus femoris from the other vastus muscles and releasing the vastus lateralis and vastus medialis from either side of the patella through a longitudinal midline incision. If the rectus femoris remains tight and is limiting flexion, then lengthening of the rectus femoris is done.

The combination of releases performed in the Thompson quadricepsplasty weakens the extensor mechanism and can cause an extensor lag. Up to 80 % of patients can develop an extensor lag after rectus femoris lengthening using the Thompson quadricepsplasty [34]. Extensor lag, if permanent, may be significant enough to affect the stability of the knee and require some patients to use orthotics long term [34]. The anterior midline incision used in this technique is another potential concern. The anterior location of the incision can create ischemia of the skin over the patella and delayed wound healing when the knee is flexed [35]. Finally, extensor mechanism rupture could occur intra-operatively if forceful manipulation of the knee is attempted as a way to circumvent lengthening of the rectus femoris [35].

A modification of Thompson's technique has shown more optimistic results [35]. In this technique, the connection of the vastus medialis to the superomedial corner of the patella is preserved and anterolateral or lateral incisions are used to avoid the problems with anterior incisions. These modifications also allowed early and intensive postoperative rehabilitation.

In 1956, Judet described his quadricepsplasty technique using a controlled staged correction of the knee stiffness [41]. It permits sequential release of the intrinsic and extrinsic components limiting knee flexion and affords the opportunity to stop as soon as adequate flexion is obtained, thereby minimizing disturbance to the quadriceps muscle [34]. The procedure is done through two incisions. The first is a short medial para-patellar incision to release intra-articular adhesions. The second is a long lateral incision allowing release of extra-articular adhesions. The final step, which is necessary in only 20 % of patients, is a release of the vastus lateralis origin from the greater trochanter [34]. The literature reflects that the Judet quadricepsplasty seems to have a much lower risk of extensor lag than the Thompson quadricepsplasty and avoids the skin issues of the midline anterior incision [34, 44]. Another promising modification of quadricepsplasty, called extra-articular mini-invasive quadricepsplasty, combines open release of the extra-articular contracture with arthroscopic release of the intra-articular contractures [45].

Several quadricepsplasty techniques have been developed specifically for patients with knee extension contracture after femoral lengthening. These patients usually do not have the intra-articular contractures often seen in posttraumatic or iatrogenic knee extension contractures [42]. Hosalkar et al. describe a technique using a lateral incision that combines a primarily extra-articular release with a V-Y lengthening of the quadriceps tendon [43]. They recommend repairing the quadriceps tendon with the knee in 60° of flexion. A second technique, the distal quadricepsplasty, is advocated by Martin et al. [36]. The authors recommend this procedure for patients with stable knees and symptomatic restriction of flexion after at least 3 months of unrestricted activities and intensive physical therapy, or those who develop obligate lateral patellar instability with knee flexion. In patients with patellar instability, residual external rotation or distal femoral valgus is corrected before or as part of the surgical procedure. Complete transection of the quadriceps tendon provides more potential length than the classic V-Y lengthening and allows for transposition to treat obligate patellar dislocation. They describe making a long oblique incision of the quadriceps tendon starting on the medial side as far distally as possible, and extending proximally and laterally as vertically as possible to exit in the lateral patellar capsule incision. In cases of patellar instability the distal or patellar portion in transposed medially relative to the proximal or quadriceps portion. To determine the amount of lengthening required, they recommend establishing a firm repair of the tendon at 80°–90° of flexion.

Khakharia et al. describe a more limited quadricepsplasty for femoral lengthening patients who are still in the external fixator [42]. At an average of 35 days from the end of distraction, patients with less than 60° of knee flexion were treated by a transverse fasciotomy to release the iliotibial band and the anterior fascia of the thigh. The anterior fascia of the vastus intermedius was also released with care not to incise any of the muscle tissue. Of the 16 patients treated with this method, one patient had a 10° extensor lag and one had clinical weakness. The authors do not recommend the limited quadricepsplasty for chronic knee extension contracture from non-lengthening related etiologies.

The postoperative management is an important component to the overall outcome of all quadricepsplasties. Most authors use a continuous passive motion machine that is applied in the recovery room and set to a 0°–60° arc of

motion and then advanced as tolerated. Weight bearing is allowed with crutches or a walker and a knee immobilizer to protect against inadvertent excessive knee flexion. Active-assisted knee flexion, passive flexion, active and passive extension, and isometric quadriceps exercises are encouraged postoperatively.

Box 11.3. Knee Extension Contractures

- Knee extension contractures are less common than knee flexion contractures.
- Patients should be considered for surgery if the knee flexion remains less than 90°.
- Because intensive postoperative therapy is required after quadricepsplasty, patient selection is critical.

Summary

A lower extremity joint contracture can be a frustrating condition to deal with for the patient, the family and the surgeon. Many treatment options can be effective initially but slowly recur over time as the child continues to grow. Equinus contractures do not always require an Achilles tendon lengthening. A careful physical examination and a thorough knowledge of the gastroc-soleus anatomy will help the physician choose the correct site for addressing an equinus contracture that will provide the optimal clinical outcome. Knee flexion contractures have multiple options for treatment that can be divided into two basic categories: acute correction or gradual correction. The options for acute correction include soft tissue releases and extension osteotomies. Gradual correction can be done with nonoperative techniques (physical therapy, bracing, botulinum toxin) or operative techniques (guided growth, external fixation). Knee extension contractures are less common than flexion contractures. Most treatment regimens recommend progressing from physical therapy, to manipulation under anesthesia to quadricepsplasty as necessary to achieve correction. New techniques for quadricepsplasty have been developed that appear promising.

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Physical Therapy During Limb Lengthening and Deformity Correction: Principles and Techniques

12

Anil Bhave, Erin Baker, and Mary Campbell

Introduction

Treating children who have lower limb deformities including shortening can present challenges for the physical therapist. Whether the deformity is related to a congenital, developmental, or acquired etiology, these patients often present with biomechanical compensations in their movement strategies in order to be as independent as possible while performing daily activities. When planning treatment of their lower limb deformities, besides assessing the child's psychosocial support system one must critically evaluate the mobility, strength, and stability across the weight-bearing joints to understand how mobility may be impacted during surgical correction. The goal of treatment for these children is primarily to improve their function and gait. In addition to physical abnormalities, these patients do suffer from body image issues [1]. Many patients report improved body image after limb lengthening [2].

In this chapter we discuss the salient principles and techniques employed by the physical therapist while treating children undergoing limb lengthening and deformity correction with conventional techniques using external fixation as well as more recently introduced intramedullary lengthening nails. We also briefly discuss specific rehabilitation issues amongst patients with congenital shortening of the lower limb as well as certain other clinical conditions such as achondroplasia and Perthes disease.

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Rehabilitation During Distraction Osteogenesis

Rehabilitation for limb lengthening and reconstruction surgery can be divided into four distinct phases: (1) inpatient phase (postoperative days 1–3), (2) lengthening or correction phase, (3) consolidation phase, and (4) post frame removal phase [3].

Inpatient Phase

Physical therapy begins on the first day following surgery. During this phase the therapist teaches the patient and family correct positioning in bed. Resting splints are made to maintain neutral positioning of the adjacent joints (Fig. 12.1). The patient and caretakers are instructed regarding range of motion and supine exercises for a home program. The physical therapist is responsible for teaching the patient how to ambulate with assistive devices, to maintain weight-bearing precautions, and maneuver stairs. Equipment such as walker, commode, wheelchair, and crutches are also ordered in preparation for discharge from the hospital. For patients undergoing limb lengthening, it is preferable to order wheelchairs with reclining back rests in order to avoid sciatic nerve stretch by not having patients sit upright with their hips flexed up to 90°. In addition, elevated leg rests are used to afford appropriate leg elevation and removable armrests are recommended for easy transfers to and from the wheelchair.

Lengthening or Correction Phase

Based on variables such as the underlying diagnosis, need for multi-segment lengthening, amount of deformity and social considerations, there are three different rehabilitation pathways that most patients take after surgery: outpatient physical therapy with one to two sessions per day for 3–5

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days per week, a day program with two to three sessions of therapy a day for 3–5 days a week, and inpatient rehabilitation with extensive physical therapy in a rehabilitation hospital [4]. The goals of physical therapy during the lengthening/correction phase are to maximize joint range of motion and maintain muscle tone and flexibility. Early joint mobilization is imperative to prevent contractures. Barker et al. [5] found that greatest loss of motion across joints occurred between surgery and commencement of distraction indicating pain as the primary cause. Aquatic therapy is a good adjunct to land therapy for patients with multi-segment lengthening and those undergoing bilateral lower extremity lengthening when full weight bearing is not allowed (Fig. 12.2).



Fig. 12.1 Orthoplast custom foot splints are fabricated prior to discharge from the hospital. In case of monolateral fixators a ring is attached to the fixator for easy application of the foot splint using Velcro straps

Consolidation Phase

In this phase, the goal of therapy is to maximize range of motion and continue to improve strength. Weight bearing and exercises that promote axial loading of the bone are encouraged. If patients have lost substantial knee range of motion, such as development of flexion contracture after tibial or femoral lengthening, then aggressive splinting across the knee joint with customized bracing or casting is done (Fig. 12.3). It is best to treat these contractures before the removal of external fixator since treatment of contracture after removal of fixator may lead to insufficiency fractures due to excessive stress and unresolved soft tissue tension.

Post Frame Removal

Gradual recovery of range of motion and functional strength through progressive exercises is the focus of physical therapy after removal of the fixator [4]. At this stage, there is a concern for fracture through the pin sites and at the newly formed bone of the lengthening regenerate [6]. It is recommended to be cautious in the first 4–6 weeks after fixator removal to allow for bone healing at these areas and therapy may be delayed for a month [7]. Casts and braces may be utilized during this phase for providing additional stability to the lengthened bone [4]. Once physical therapy is resumed, it is recommended that closed chain exercises be started before open chain exercises. Closed chain exercises are weight-bearing exercises, providing axial loading to further strengthen the lengthened bone segment. Open chain exercises are exercises where the distal limb segment moves around the proximal axis, this leads to increase torque and stress on the weaker newly lengthened bone segment.



Fig. 12.2 Chlorinated, heated, hydrotherapy pool for promoting increased range of motion and muscle strength

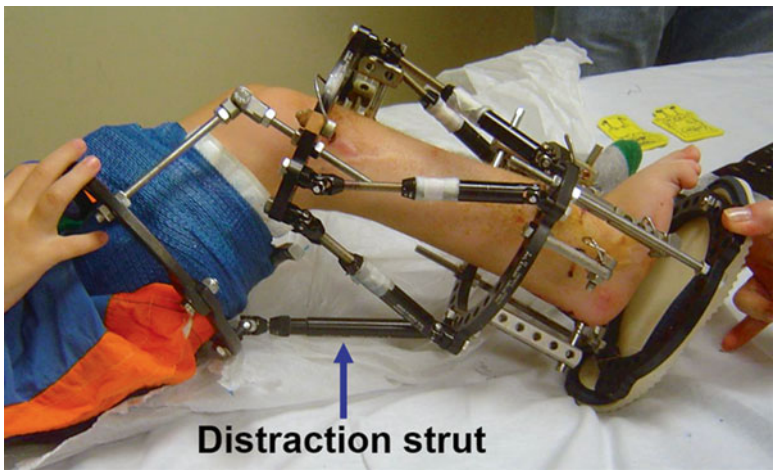


Fig. 12.3 Use of a Taylor Spatial Frame™ strut to apply an extension force in a patient who developed severe knee flexion contracture during tibial lengthening for fibular hemimelia limb deficiency. Thigh cuff using fiberglass casting is linked to the external fixator with hinges



Fig. 12.4 Careful stabilization of the femur and tibia are critical during therapy after fixator removal. Patient receiving hamstring stretch with posteroanterior mobilization. Therapist hands are placed as close to the joint line as possible

Careful stabilization of the bone segment as close to the joint line as possible is necessary to avoid bending stress on the bone during mobilization with physical therapy (Fig. 12.4). Manual mobilization with adequate stabilization of the lengthened bone is a safe and effective way to regain motion. Gait training and return to normative function are other additional goals during this phase. Recovery is slowest immediately following fixator removal and speeds up between 6 months to 1 year postoperatively. No further recovery is typically seen after 2 years following fixator removal [2]. During this period, home exercise program and participation in activities play a big part in the final journey of return to full functionality.

Box 12.1. Complications During Limb Lengthening

- Muscle contractures
- Muscle weakness
- Joint stiffness
- Nerve injury
- Joint subluxation

Role of Physical Therapy in Management of Complications

After the index surgery, patients can experience bone and/or soft tissue complications, which can compromise functional outcomes. Temporary soft tissue dysfunction occurs in the majority of patients who undergo limb lengthening and deformity correction procedures. The severity of these complications and obstacles to lengthening are dependent upon the diagnosis, amount of lengthening or correction, soft tissue quality, level of osteotomy, technique and device used for surgical correction, and nerve complications [1]. In addition to physical causes of complications there are social reasons such as poor pain tolerance and inadequate family support that can lead to suboptimal results.

Examples of soft tissue dysfunctions include muscle and tendon contractures, muscle weakness, joint stiffness, nerve injury, and joint subluxation. Bone complications include poor bone formation, angulations of the regenerate bone, and

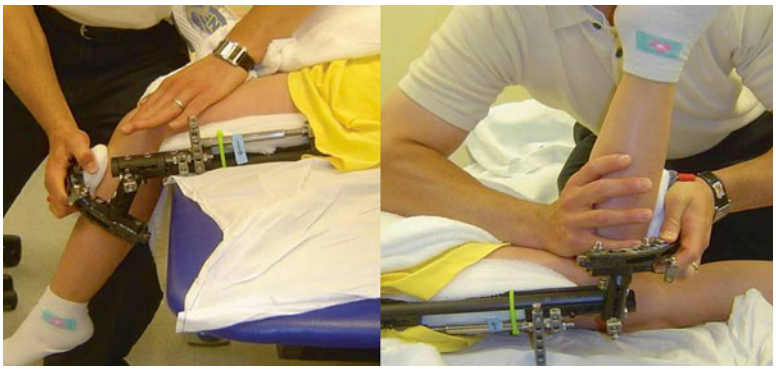


Fig. 12.5 Hand placement is critical for joint mobilization. Interior mobilization of the patella during knee flexion mobilization and correct posterior stabilization of the proximal tibia are important

insufficiency fractures. Therapeutic intervention is crucial to avoid permanent dysfunctions. The physical therapist needs to have adequate handling techniques and the skill to apply appropriate forces and stabilization in order to focus treatment targeted at the appropriate tissues (Fig. 12.5). It is a fine balancing act since excessive forces can cause bone to bend or fracture. The therapist must keep in mind that the stress strain curve for tissues in the human body alters with age [8]. Pediatric patients deform their soft tissue with much less force than adults. Bone and soft tissue complications can occur during lengthening and consolidation phases, as well as after the removal of the external fixator.

Muscle Contractures

Contractures occur when elastic tissues and contractile elements are unable to accommodate change in their length following limb lengthening and deformity correction. Myofascial tissues resist such elongation more than most other tissues and are therefore more prone to contractures. A relative shortening of diarthrodial or biarticular muscles (crossing two joints) and the fascial component is often the cause of such contractures [1]. For example, the gastrocnemius develops passive tension much more rapidly in response to bony lengthening than does the soleus.

When muscles are kept in a shortened position, often due to postoperative pain early in the lengthening process the muscle adaptively shortens by absorbing sarcomeres to maintain efficiency. Prolonged stretch through a muscle has been proven to add sarcomeres. The therapist must devise a way to provide prolonged stretch of such muscles across adjacent joints. For such stretch to be effective it must have four critical components: (1) adequate intensity, (2) long duration, (3) adequate frequency with multiple repetitions, and (4) an active stretch component [8].

Problematic muscles should be identified early in order to treat contractures effectively. In tibial lengthening, for example, problem muscles are the gastrocnemius and toe flexors. As a result, patients are prone to develop knee flexion, ankle plantarflexion, and toe flexion contractures. Likewise, during femoral lengthening, the quadriceps (especially the rectus femoris) and hamstring muscles are predisposed to develop contractures resulting in deficits of flexion and potential for fixed flexion deformity across the knee joint, respectively.

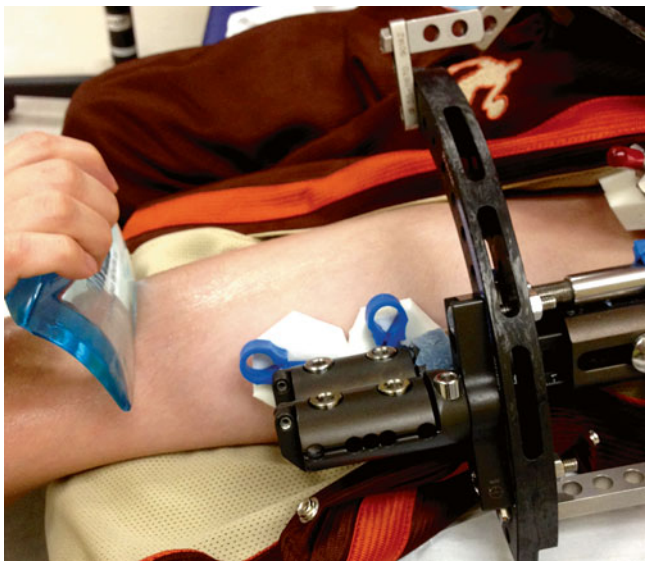


Fig. 12.6 ASTYM™ treatment for improving muscle flexibility and to promote remodeling of the fibrous tissue

Soft tissue restriction can be further complicated by the presence of scars from previous surgery, as is often seen in patients with long-standing limb deformities. Scar tissue lacks the elasticity of the native tissue and scar fibers are often laid down in a much more disorganized, rigid pattern, especially when the tissue is immobilized due to pain or lack of motion. Utilizing soft tissue mobilization with implementation, such as ASTYM™ (Fig. 12.6) and Graston™, can break down deficient, mature scar tissue and establish a more organized, pliable scar [8].

Managing muscle contractures effectively is vital for a satisfactory clinical outcome following limb lengthening and

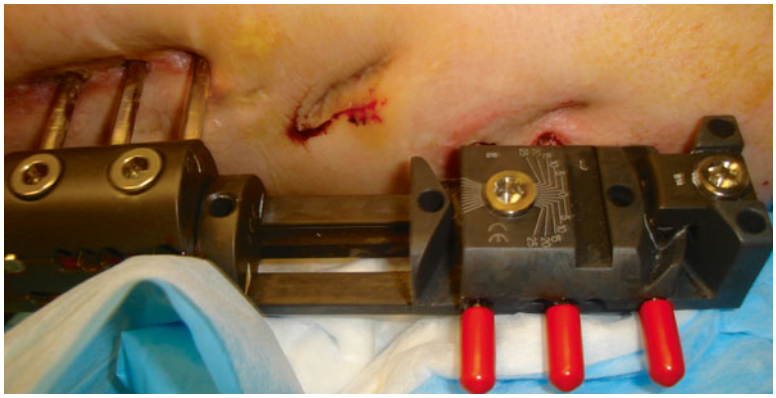


Fig. 12.7 Pin site around the hip joint can be painful due to movement of excess tissue during therapy. Massage around the pin site and use of tight wraps help reduce pain during therapy

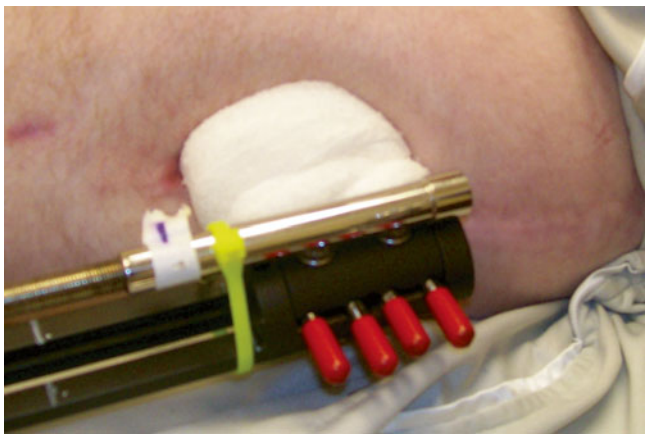


Fig. 12.8 A compression wrap around proximal femoral pins to avoid tissue motion during therapy. This technique reduces pain as well as risk of infection

deformity correction [1]. Passive stretching is an effective way to manage soft tissue contractures. Per our protocol, moist heat is applied prior to stretching to aid in collagen breakdown and improve stretch of connective tissue [8]. Analgesic medications administered 30 min prior to therapy help patients tolerate the stretch better. Reciprocal inhibition, activating the antagonist muscle before stretching the agonist, relaxes the muscle being stretched. Pin sites near the fascial and muscle planes are susceptible to inflammation. Thus gentle soft tissue mobilization and massage helps to manage tightness caused by adherent soft tissues around the pin. Another strategy to reduce pain around the pins during joint mobilization and stretching is to use tight pin wraps especially at sites near the knee or hip joint (Fig. 12.7). The tight pin wraps reduce skin motion during therapy and significantly reduce pain (Fig. 12.8). Stretching of biarticular muscles 10 times per session and uniarticular muscles five times a session with 20–30-s holds for each stretch is recommended. When stretching a biarticular muscle, stabilization at one joint is necessary during mobilization at the other joint to obtain maximum stretch. Ideally, a patient should have such treatments twice a day.

Positioning and splinting strategies should also be used to manage soft tissue contractures. Optimal positions vary by affected body part. For example, patients who have undergone tibial lengthening should be positioned in knee extension and ankle dorsiflexion (Fig. 12.9). Knee extension, along with hip abduction, is a desirable position for patients undergoing femoral lengthening. Commercially made static splints (Fig. 12.10) augment therapeutic passive stretching by counteracting the connective tissues' elastic response to passive stretching. Using a splint to place the muscle under tension for as many hours as possible, helps prevent contractures by obtaining plastic response in the connective tissue. Dynamic splints produce optimal plastic elongation (permanent elongation) of the connective tissue through a low load prolonged duration stretch (Fig. 12.11). Such a stretch also causes physiological remodeling of connective tissue through biochemical response. Dynamic splints work most effectively in treating knee contractures as they are a hinge joint. Be aware that splints work only in optimal positions and tension should always be increased gradually.

Commercially made dynamic splints do not always provide the customization needed for an effective stretch. Therefore, the authors developed and utilize a custom knee device (CKD) for contracture management during or after external fixation (Fig. 12.12). This is a semirigid device made of a polyester and fiberglass cast of the thigh and lower leg with an aluminum hinge across the knee to allow for progressive stretching into the restricted range of motion. Elastic bands are utilized to provide the needed tension and the rigidity of the casting materials allows for minimal loss of the stretch energy to the desired tissues. Knee joint subluxation can also be managed utilizing the CKD with added straps at the knee joint to help approximate the joint and reduce the subluxation. The more the patient wears this

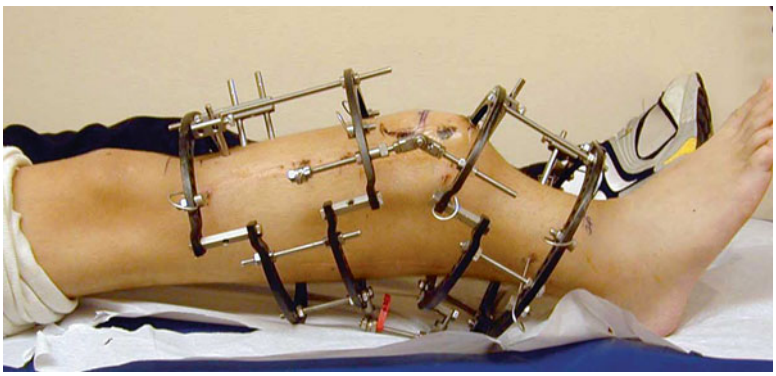


Fig. 12.9 Patient undergoing tibial lengthening and procurvatum deformity correction. Gastrocnemius and soleus contracture with lack of ankle dorsiflexion occurs commonly in these patients

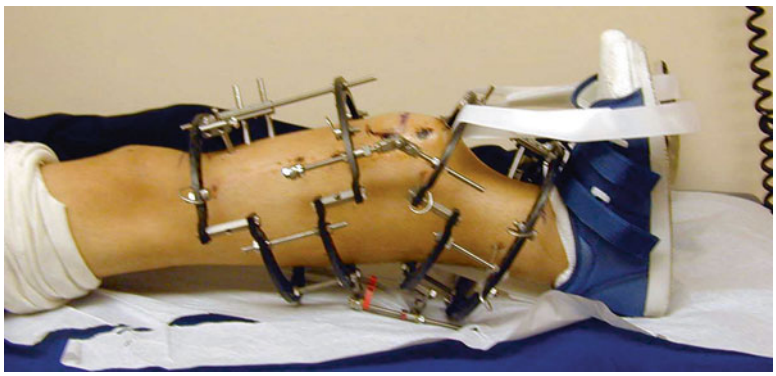


Fig. 12.10 Commercially available walking shoe modification with Velcro straps to keep ankle in neutral dorsiflexion position

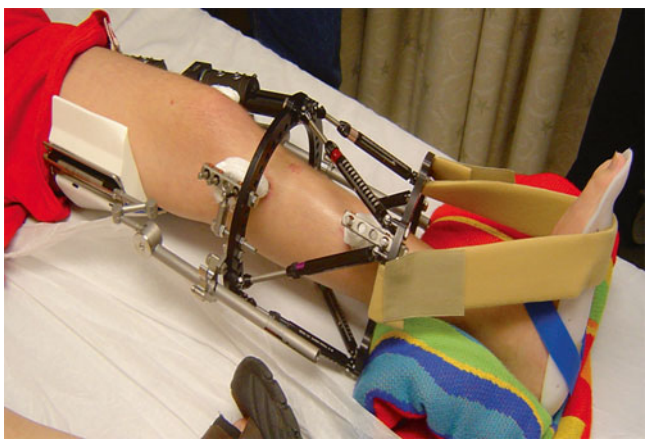


Fig. 12.11 Use of commercially available dynamic brace (Dynasplint™) attached to the proximal tibial ring to improve knee extension

device, the sooner the muscle truly lengthens and range of motion is restored. This concept can be utilized with tibial frames, as a thigh cast can be made with a half ring casted in and then attached to the tibial frame with a hinged joint. A telescoping strut can be added and the knee can be progressively straightened if a flexion contracture develops (Fig. 12.13). It is recommended to use these splints all night or at least 3–4 h per day.

Joint Stiffness

Muscle contractures, if not treated aggressively, lead to capsular and intra-articular adhesions. Contracture of biarticular muscles also introduces compressive force on the articular cartilage, which leads to a stiffness across adjacent joints [9]. Characteristics of a stiff joint include loss of motion and ease of movement within the joint's available excursion. Joint stiffness is best prevented with active and passive motion. Manual traction with joint mobilization is beneficial and recommended when feasible. However, such mobilization can only be performed on joints that are not included in the external fixation construct.

Muscle Weakness

Patients may experience muscle weakness due to disuse atrophy and neurogenic inhibition with associated pain. Electrical stimulation and hydrotherapy are two modalities that can be used to manage muscle weakness. Electrical stimulation is used as an adjunct to a strengthening program and to augment voluntary muscle contraction. The most common area for electrical stimulation is the quadriceps muscle (Fig. 12.14). In our experience, as long as there is no direct contact of electrodes to the half-pins, it has been safely



Fig. 12.12 Use of custom knee device (CKD) in a child with congenital femoral deficiency (CFD) undergoing femoral lengthening using monolateral fixator. Elastic bands offer continuous dynamic stretch in to extension. Patients are encouraged to use the device 8–14 h per 24-h period daily

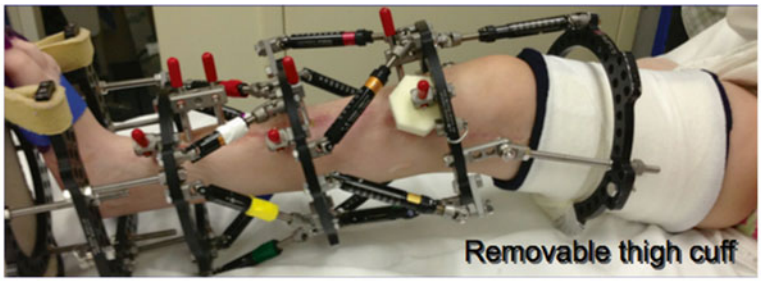




Fig. 12.13 Use of removable thigh cuff attached to tibial frame with hinges in a patient undergoing tibial lengthening. Taylor Spatial strut between ring incorporated in thigh cuff and proximal tibia ring allows for applying an extension stretch

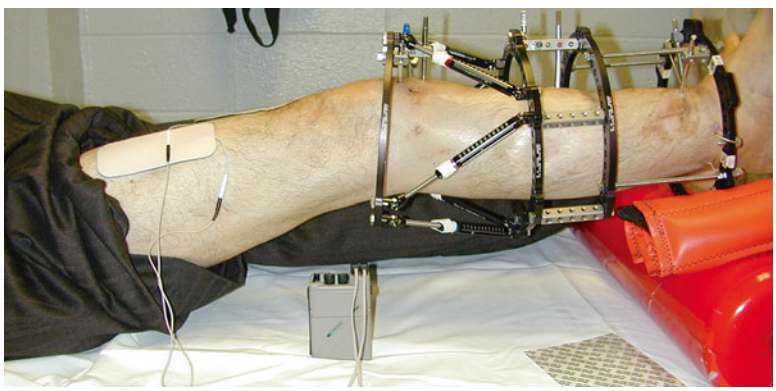


Fig. 12.14 Neuromuscular electrical stimulation of the quadriceps muscle with heel propped position. This position allows for active stretch as well as augmented strengthening of the quadriceps

tolerated by children undergoing conventional distraction osteogenesis treatment with external fixation.

At our center, typical electrical stimulation parameters are a biphasic pulse at a frequency of 35–70 Hz and pulse duration of 300–400 μs. In addition, an on-to-off ratio of 1:3 is recommended. Stimulation intensity should be adequate to cause strong contraction of the muscle. Patients tolerate newer electrical stimulation devices better using advanced frequency modulation software. Children tolerate higher intensity settings with these devices as compared to older devices. Family education for setup is recommended for incorporation of electrical stimulation into a home program.

Hydrotherapy helps avoid significant muscle weakness, especially in patients with bilateral external fixators or unilateral femur plus tibial fixators. Hydrotherapy also promotes increased active ROM (see Fig. 12.2). Buoyancy offsets fixator weight and facilitates muscle strengthening. Limb lengthening patients experience aquatic therapy in a

heated, clinically chlorinated pool. Hydrotherapy allows for weight bearing early in the lengthening process, even if there are weight-bearing restrictions on land. With the water up to the nipple line, patients can utilize the buoyancy of the water to provide their bones with protection from gravity's compressive forces to walk for continued cardiovascular health and muscle memory. The heated water brings muscle relaxation and freedom of motion, which allows for exercise and standing mobility. The chlorine levels at our center are kept at level 5 since patients get in with open pin sites. Purulent drainage from a pin site is a contraindication for getting into the pool. Daily assessment of pin sites are done by the treating land therapist as well as the aquatics therapist.

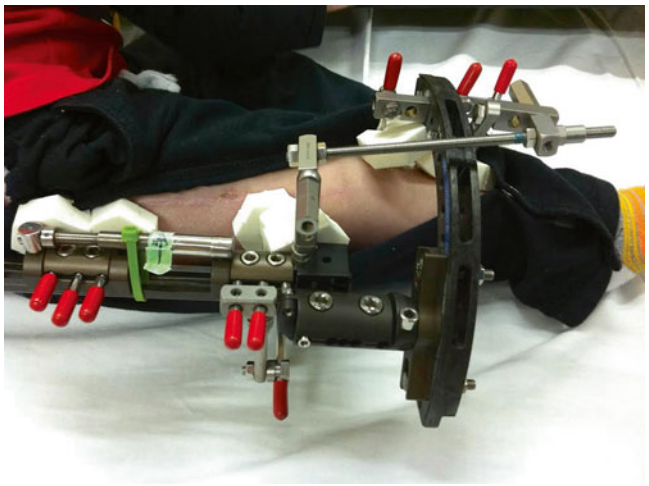


Fig. 12.15 Use of knee extension rod in a patient undergoing femoral lengthening for congenital femoral deficiency. Patients are recommended to use the extension rod all night long and 2 h on and 2 h off during day

Joint Subluxation

Joint subluxation is a dreaded complication of limb lengthening surgery. Subluxations are caused by lack of opposition of the joint surfaces, muscle and tendon contracture, and ligamentous laxity. If a subluxation is not recognized and treated early, it can result in permanent sequelae. This can lead to significant reduction in range of motion, pain, and early onset of arthritis due to articular cartilage erosion. Knee subluxation in patients who have undergone femur lengthening for congenital femoral deficiency usually involves posterolateral translation of the proximal tibia. Such patients may have cruciate deficient knee joints, hamstring contracture, quadriceps inhibition, tight iliotibial bands, and flexed knee posturing. The combined effect of these conditions leads to unopposed posterolateral pull on the proximal tibia and in some cases also lateral subluxation of the patella [1]. Knee joint subluxation can occur during femoral lengthening if the joint is not protected in the frame or during lengthening with an intramedullary lengthening device. It can also occur due to unresolved residual tension in the soft tissues post fixator removal. Due to the high frequency of subluxations in the congenital femoral lengthening most surgeons prefer to extend the frame to the tibia with a hinge at the knee joint (Fig. 12.15) [1].

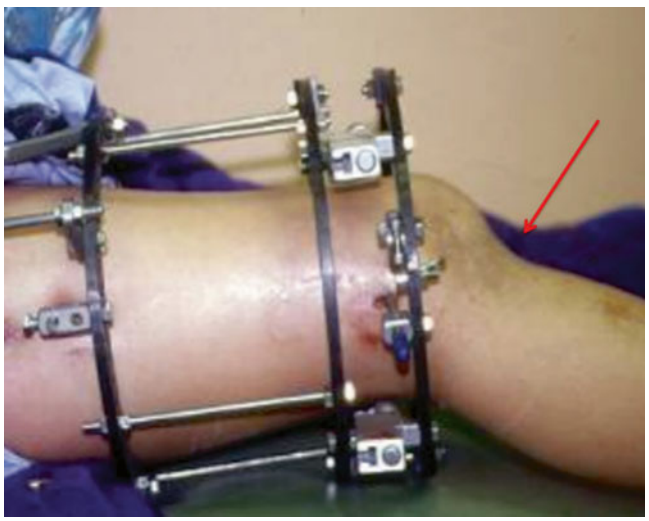


Fig. 12.16 Posterior subluxation of the knee in a patient undergoing femoral lengthening. The posterior subluxation of the tibia is demonstrated by the *arrow*. Due to prevalence of this complication, most surgeons use tibial frame linked to the femoral frame with hinges to prevent knee subluxation

Treatment for knee subluxation starts with early detection. Usual signs include posterior slope of the tibia (commonly called the ski slope sign) (Fig. 12.16), anterior pain on knee extension, and overall sudden increase in stiffness and pain. Physical therapy treatment consists of vigorous knee extension with proper proximal tibia anterior tracking via slings and manual mobilization techniques. Soft tissue mobilization techniques and augmented tissue stimulation to the lower kinetic chain using ASTYM™ is particularly useful. If the patient has an external fixator with no extension to the tibia, then manual therapy with joint distraction can also be employed. At night, the patient may also use tibial traction and dynamic splints. In the author's experience, mild to moderate subluxations respond to conservative regimens, while more severe ones may need surgery that includes distal iliotibial band and biceps femoris releases.

Hip subluxation is a rare complication of femoral lengthening. It presents as posterolateral migration of the femoral head and usually occurs in patients with acetabular dysplasia in the face of an adductor contracture during proximal femur lengthening. Adductor stretching and an abduction pillow can help prevent hip subluxation in proximal femoral lengthening. When hip subluxation occurs, patients usually need additional surgery including adductor release, followed by tibial skin traction and intensive therapy.

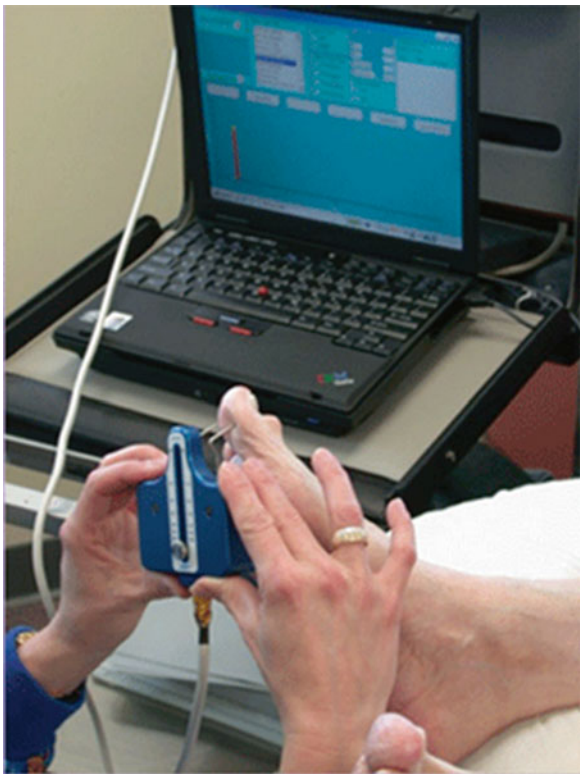


Fig. 12.17 Pressure specified sensory device (PSSD) (Sensory Management, Baltimore, MD). This device allows for on invasive monitoring of sensory function during lengthening

Nerve Injury

During the lengthening process, as the bony segments slowly distract, the soft tissue often becomes taut and restricted. It was thought that bony distraction over-stretches the nerve, but Nogueira et al. [10] found this is not always the case as no correlation was noted between amount lengthened and the onset of nerve signs. Patients often present to therapy with numbness and tingling peripherally with burning pain along nerve distributions. The most commonly compromised nerve is the peroneal nerve, more prevalent in tibial lengthenings and corrections. Classic signs are numbness and burning pain on the dorsum of the foot, especially between the first and second toes. In more severe cases, the extensor hallucis longus (EHL) becomes weak or worse, the tibialis anterior muscle weakness leads to a foot drop. The sciatic nerve can also be over-stretched, especially in large femoral lengthenings or simultaneous femoral and tibial lengthenings. In foot corrections, as in clubfoot, the medial plantar nerve is at greatest risk for compromise, with burning and tingling on the plantar surface of the hallux. If nerve compromise is suspected, the authors utilize the pressure specified sensory device, or PSSD (Fig. 12.17). This device can reliably test the light touch sensation of various nerve distributions with 100 % sensitivity and 87 % specificity [10]. The device also has the capability to store data for later comparison. One drawback of such a device or any sensation testing is that the patient needs to be of an age or maturity to participate reliably in the test. Usually, this is reserved for patient's aged 6 years and older. When sensory signs and symptoms of nerve compromise are present, the medical team will either slow down the lengthening or prescribe medications like Gabapentin (Neurontin™) or Lyrica™ to manage the pain associated with nerve problems. In some cases, especially those with motor weakness, urgent surgical release of the offending nerve is required. Considerations need to be made when providing therapy to patients who have nerve compromise. Bi-articular stretches, such as straight leg raises, should be avoided in favor of mobilization at each individual joint of the lower leg. This allows for soft tissues to be lax on one end while the opposite end is treated.

Weight-Bearing Considerations

Functional mobility training will depend greatly on the level of weight bearing allowed. During the lengthening and consolidation phases, weight bearing may be permitted, dependent on the type of fixator or device used and the bone being lengthened [4]. A ring fixator (such as the Ilizarov and Taylor-Spatial™) generally provides the underlying bone with substantial support to allow for 50–100 % weight bearing. In the classic Ilizarov frame where each segment has four rings and four wires are placed on each ring with specific tensile strength, all of the force that normally would go through the bone is transferred to the metal frame, protecting the osteotomy site and allowing for full weight bearing. The more recent versions of the circular frame utilize rings similar to that of the Ilizarov but favors half-pins instead of transosseous wires as bony anchors. These pins are generally better tolerated by the patients as they offer less tearing of the skin. If the foot is incorporated into the frame then a walking ring is added, fitted with a rubber tread to allow for safe ambulation (Fig. 12.18).

Monolateral rail fixators handle weight-bearing forces very differently. Typically, majority of the forces on the bone are lateral for the femurs and medial for the tibia. Without any counter to distribute forces in an even manner, weight bearing is restricted in such patients during lengthening to prevent loosening of the pins or worse, fracture at the pin site. Many monolateral fixators for femoral lengthening will have a hinge at the knee and an extension to the tibia, with a partial ring attached to pins in the tibia (see Fig. 12.15). This offers a great deal of additional stability at the osteotomy site, allowing for weight-bearing as tolerated during the lengthening process.

Even in cases where monolateral rails are used without a counter as in stature lengthening, weight bearing with a walker



Fig. 12.18 Application of walking ring to the tibial frame allows for pain free weight bearing and also one can adjust the length of the walking ring to accommodate leg length difference

is often allowed for transfers only. Repetitive forces through these devices weaken the bony interface, as would happen with unrestricted weigh-bearing. Restricted, controlled weight bearing for very short durations allows for stretching of the gastrocnemius muscles and the work induced is good for cardiovascular health.

These considerations around weight bearing will shape the plan of care in physical therapy. Ambulation during lengthening helps to maintain muscular and bony strength and may shorten the rehabilitation process after fixator removal [11].

Specific Musculoskeletal Disorders

Legg-Calve Perthes Disease

Legg-Calve-Perthes disease (commonly referred to as Perthes disease) is usually seen in children ages 4–10 years and is more common in boys [12]. Perthes disease has been treated using the principle of "containment" by long periods of immobilization, historically using hip spica casting to hold the femoral head in a most congruous position with the acetabulum to allow the best reshaping of the femoral head. Hip spica casts often bring difficulty with mobility of the child as well as hygiene issues. Utilizing a hip distractor external fixator alleviates these issues as well as assures a congruous position of the hip (Fig. 12.19). They hold the hip in 15°–20° of abduction, 10° of flexion and 10° of external

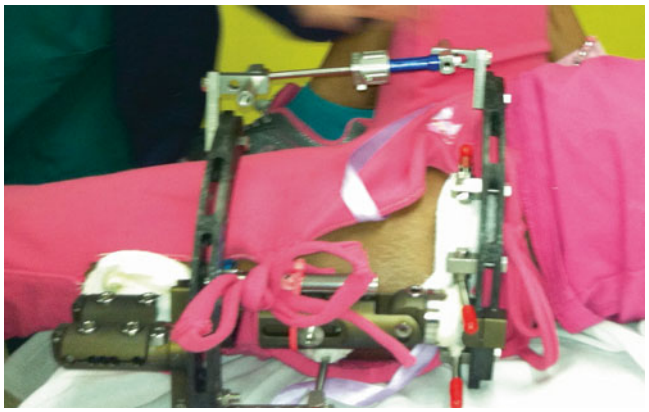


Fig. 12.19 Use of hip extension rod to maintain maximal hip extension in patient undergoing hip distraction for treatment of Perthes Diseases. Patients wear the rod all night long

rotation. A partial ring over the pelvis provides stabilization to a hinged joint at the axis of the sagittal plane for hip motion and a distractor bar is attached to the monolateral rail. The femur is gently distracted over a period of 10–14 days and held in position for approximately 4 months to allow for re-ossification and reshaping of the femoral epiphyses. Utilizing this frame can lead to improved joint mobility and decreased pain [13].

The focus of rehabilitation immediately after surgery is on standing mobility training, family education, and establishing a home program for the patient. Since the legs are being held in abduction and weight bearing through the effected limb is limited, a wide rolling walker is often needed initially to aid in ambulation. Patients can progress to axillary crutches eventually to allow for ambulation on stairs, but selection of crutches is a very individual choice, based on the patient's safety awareness and balance control. For long distances, these children still need a wheelchair, with consideration of the size of the patient and the extra width added by the frame laterally.

Only sagittal plane motion is allowed by the hip distractor. Therefore, only hip flexion and extension mobilization can be done while in the frame. The family needs to be educated in a home stretching program to maintain hip motion and prevent contractures from immobility and pain. A removable anterior telescoping bar is added to the frame to provide a progressive, sustained stretch across the hip into extension (see Fig. 12.19). After 4 months the frame is removed and the role of physical therapy increases exponentially. For the first 6 weeks post removal, the patient is wearing a hip abduction brace nearly at all times, giving the patient a very wide stance in gait. Weight bearing is allowed at this point and gait training is needed to learn to ambulate without a device.

Without frontal and transverse planes of motion across the hip during external fixation, abduction/adduction and

internal/external rotation motions are very limited after frame removal. Therapy focuses on regaining the hip motion, both passively through joint mobilization and actively through exercise and mobility training. Control of active hip motion is challenging in this population since the lumbar spine so readily provides foot placement in gait and movement in exercise—a compensatory strategy developed during the 4 months of non-use in the frame and potentially even longer, depending on how long the disease progressed prior to treatment. With an abduction contracture on the effected limb, the leg feels longer to the patient, and a compensatory trunk lean to the contralateral side is apparent. Furthermore, due to the young patient population, it becomes imperative that the family is involved in treatment, learning to cue the patient to aid in retraining and carry over the exercise program at home. Normalization of gait can take several months. In our experience, integrating yoga and other treatment modalities that provide postural awareness help in the recovery of hip motion and strength, and ultimately allow a return to independent activity.

Congenital Femoral Deficiency

Lengthening the femur of a child with congenital femoral deficiency (CFD) can be one of the most difficult lengthenings to manage. Up to 95 % of these children have deficient or absent cruciate ligaments in the knee and such instability can cause problems during lengthening [14]. As the femur gets longer, the pull of the muscles increases, especially that of the hamstrings. With underdeveloped quadriceps and pain limiting motion, the pull of the hamstrings can cause a posterior subluxation of the tibia on the femur. Having a soft tissue reconstruction to address the cruciate deficiency prior to femoral lengthening in a patient with CFD associated with ligamentous laxity lowers the risk of postoperative subluxation [15]. If subluxation occurs, lengthening is slowed or stopped and knee extension mobilization needs to be done with posterior support to the proximal tibia.

The half pins used to anchor the fixator across the knee joint can be problematic with knee flexion mobilization. Therapists' hand placement is important for the safety of the patient. The knee cannot be mobilized into flexion from the distal tibia because that can cause loosening of the tibial pins. Anterior to posterior tibial mobilization must be done proximally, with a posterior counter force to prevent pin loosening and fracture of the bone (Fig. 12.20). The loss of leverage and inefficient grip can make this mobilization difficult for the therapist. Knee flexion is often where the greatest loss of motion occurs during CFD lengthening. If the flexion measures 40° or less, it is often recommended that lengthening be stopped, at least temporarily, till further joint mobility is gained [9].



Fig. 12.20 Careful hand placement for knee flexion mobilization in a patient with femoral lengthening for CFD. The therapist's left hand is stabilizing the hinge and right hand applies flexion force as well as stabilizes posterior tibia

After removal of the fixator, therapy can begin to focus on mobility training as well as regaining motion to the preoperative level. Often children with CFD have acetabular dysplasia and weakness around the hip, especially involving the abductors. Gait training and exercise that focuses on closed chain control of the hip and trunk are needed, but often difficult for the patient. Gaining normal strength of the abductors may not be a realistic goal, given the number of prior surgeries including placement of intramedullary devices that may have been placed and removed from the femur in their lifetimes causing damage to the abductors if a trochanteric approach was used as well as the presence of hip dysplasia.

Treating a child or adolescent with an internal femoral lengthening device presents a similar set of potential problems, although the external fixation can provide stability to the knee and prevent subluxation. Often CKDs are made with the use of subluxation straps early in the process to encourage gentle knee extension and prevent posterior subluxation of the knee during such lengthening (Fig. 12.21).

Fibular Hemimelia

Fibular hemimelia is the most common lower extremity deficiency affecting males slightly more than females [16]. Tibial lengthening and deformity correction for the treatment

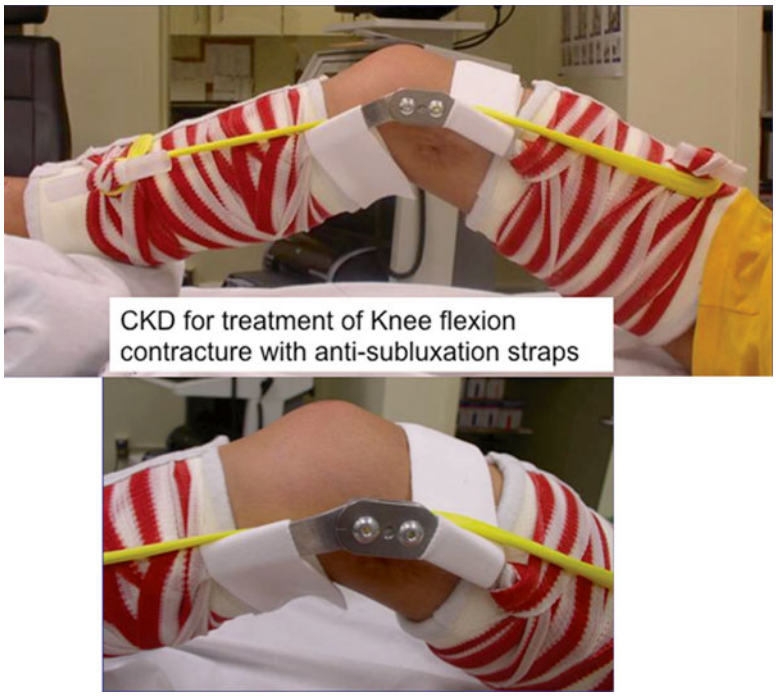


Fig. 12.21 Use of CKD bracing with special straps. The straps allow for an inferior mobilization of the distal femur and superior mobilization of the proximal tibia. The combination of CKD with elastic band and special straps allow for simultaneous correction of knee flexion contracture and subluxation

of fibular hemimelia is commonly done with a circular fixator. Knee extension and ankle dorsiflexion are the motions most compromised. The foot can be in or out of the frame. In our experience, the patient is more apt to lose range of motion at the knee and ankle if the foot is not included in the frame.

The peroneal nerve is also at risk of being compressed or overstretched during tibial lengthening [1]. Neurogenic symptoms can worsen with knee extension. Patients with the foot included in the frame are at a higher risk of peroneal nerve palsy with tibial lengthening as they are unable to plantarflex the ankle to provide relief to the nerve at the fibular head. Surgeons will often prophylactically release the peroneal nerve in such cases.

Patients with fibular hemimelia often have altered mechanics of the ankle joint. The mobility of the ankle is generally limited in these patients with some compensation at the midfoot and forefoot due to some compensatory hyper flexibility at these distal joints. It is important for the physical therapist to consider these compensatory mechanisms as gait training advances following removal of the fixator. Rocker bottom shoes may help the patient achieve a more normal gait pattern despite lack of ankle mobility.

Achondroplasia

When approaching a patient undergoing limb deformity correction due to underlying achondroplasia, it is important to take into consideration the unique morphologic features of this patient population. Skeletal characteristics in achondroplasia include but are not limited to increased lumbar lordosis with excessive anterior pelvic tilt, a more prominent sacrum, and a smaller spinal canal. These skeletal characteristics put the patient at an increase risk of symptomatic spinal stenosis during bilateral lower extremity lengthening. Besides neurologic deficits in the lower extremity, loss of bowel and bladder function can also occur due to tethering of the spinal cord and nerve roots. During treatment, the physical therapist should position the patient to compensate for these skeletal abnormalities while performing lower extremity mobilizations. This can be done by utilizing pillows and wedges to allow for hips to be in a flexed position and the spine to have decreased lordosis (Fig. 12.22). A prominent sacrum also puts patients at a higher risk of sacral wounds. It is important for patients to have time set during the day that they are lying prone to allow for decreased pressure on the sacrum to decrease risk of pressure wounds. Wheelchair cushions are also used to alleviate pressure on the sacrum in sitting.

Individuals with achondroplasia also present with unique soft tissue characteristics including ligamentous and joint laxity, and relatively redundant muscle tissue [17]. This relative laxity of the muscles decreases strength and leads to a waddle type gait pattern. Often, there is a noticeable trunk sway for compensation as well as a larger base of support during the gait cycle. These gait abnormalities provide the individual with increased stability without the use of core control and proper hip abductor mechanism function. Redundant muscle tissue along with ligamentous and joint laxity allows for increased flexibility early in the limb lengthening process, limiting the amount of motion lost in the early phase of lengthening.



Fig. 12.22 Prone positioning in a patient with achondroplasia undergoing four-segment lengthening. Use of a wedge allows for reduction of the hyperlordosis in patients

While undergoing bilateral lower limb deformity correction/lengthening, patients are allowed weight bearing for transfers only. In addition to muscular and cardiovascular benefits mentioned earlier, transfer training gives the patient and family increased independence at home. Patients can use push up blocks to help provide upper extremity leverage during lateral and anterior/posterior transfers from wheelchair to bed. Assistance should be given to the lower extremities, lifting from the full tibia rings if available. Core stabilization exercises can help the patient control the spine for transfers, particularly important in head and neck control. Aquatics become an important physical therapy modality for strengthening and gait training while the patients are non-weight bearing.

Physical therapy is performed on a daily basis throughout lengthening, with the primary focus to limit loss of motion through the use of soft tissue and joint mobilization. If lengthening can be carried out simultaneously in all four segments (bilateral femur and tibia), these patients are at a high risk for joint contractures, despite their soft tissue laxity. Loss of motion often occurs in ankle dorsiflexion, hindfoot eversion, knee flexion/extension, hip flexion/extension, and hip abduction. Patients with monolateral rail lengthening of the femur may experience increased tension in the distal iliotibial band due to the tethering at the pin sites [17]. Mobilizations can be performed to the distal iliotibial band by performing a gentle varus mobilization of the knee. If circular fixators are being used for the tibia, then foot splints can be attached to the circular rings to help limit loss of dorsiflexion.

Four-segment lengthening also contributes to increased risk of nerve injury. The physical therapist must continue to monitor for early signs and symptoms of neuropraxia during the lengthening process. Knee extension stretching should be performed with the hip in less than 60° of flexion. In severe cases the hip can be dropped over the side of the table into slight hip extension to avoid stressing the nerve pathway during knee extension mobilization (Fig. 12.23). Ankle dorsiflexion mobilization can be modified by being performed with the knee in slight flexion.

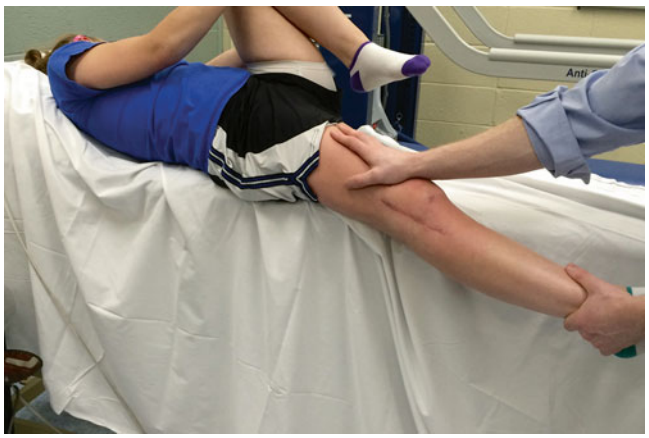


Fig. 12.23 Knee extension mobilization with hip in extension. This position allows for adequate knee extension stretch without compromising or stretching sciatic nerve

After lengthening is complete, patients are in external fixators until proper bone healing has occurred. During this consolidation phase, weight bearing is gradually increased and strengthening can be progressed. Loss of hip extension and ankle dorsiflexion during lengthening leads to an increase in lumbar lordosis while standing with a significantly flexed posture of the knees and hips. Standing stretches of the gastroc-soleus complex with use of tilt boards or wedges helps to improve erect posture and allow for the stretch-recoil necessary for proper push off during gait. Regaining hip extension is important to decrease the stress on the lumbar spine and spinal cord. Increasing hip extension also restores the stretch-recoil of the hip flexor mechanism to allow for appropriate propulsion of the lower extremity.

Immediately after external fixators are removed, patients are non-weight bearing for approximately 4 weeks. During this non-weight-bearing phase, the patient's family performs gentle range of motion exercises. When sufficient bone healing has occurred and weight bearing is allowed, physical therapy resumes with an emphasis on gait training. Patients habitually have a wider base of support after fixator removal, both from

compensatory gait prior to surgery and having walked with external fixators. Treatment strategies should focus on normalizing the base of support, increasing stride length, and improving timing of hip musculature activation. Global lower extremity and core strengthening facilitates the patient's overall independence with their functional activities and gait.

Internal Lengthening Devices

There has been an increasing interest at some centers in performing "internal lengthening" of the lower extremity using intramedullary nails. In our anecdotal experience, patients and families seem to prefer this method over using external fixators. Currently, the use of intramedullary lengthening devices in femur or tibia is generally limited to patients who have reached skeletal maturity, have minimal or no angular deformity and have adequate diameter of the intramedullary canal of the lengthened bone. Weight bearing is usually 30–50 lb. during lengthening. It is also important to remember that it is not the magnitude of load but the frequency of loading that can have deleterious effects on the stability of the intramedullary lengthening implant During the distraction phase weight bearing is typically limited to activities of daily living only, whereas in the consolidation phase weight bearing can be increased as maturation of the lengthening regenerate occurs.

Complications of internal lengthening are similar to that of the external fixator excluding pin site problems. The rate of complications or obstacles to lengthening using the ISKD™ device has been reported as high as 65 % [18]. These rates are similar to rates reported in the limb lengthening performed with external fixator.

In addition, due to lack of external fixation stability, the risk of joint subluxation is increased with the lengthening nails. Knee flexion contracture with subluxation is a common complication of internal lengthening especially in patients with congenital etiology (Fig. 12.24). Prophylactic bracing and adequate physical therapy are critical to prevent joint subluxation and development of permanent contractures. The magnitude of complications is also dependent upon the ability to control the rate of lengthening. If the internal lengthening device goes faster than 1–1.5 mm a day, the severity of complications is greater. The authors have successfully treated this problem of excessively rapid lengthening utilizing proper handling techniques as well as custom dynamic bracing (see Fig. 12.21). In a retrospective review of this approach in 20 patients (with 22 contractures, 9 subluxations), successful resolution of knee flexion contractures occurred in 19/20 patients (95 % success) and 8/9 subluxations (88 %) (Fig. 12.25) [19].

Knee flexion deficits are also a common obstacle to internal femoral lengthening. Restoration of motion is best achieved with daily therapy including patella and soft tissue mobilization.



Fig. 12.24 Knee subluxation in a patient undergoing internal femoral lengthening for congenital femoral deficiency



Fig. 12.25 Complete resolution and normal knee alignment of the patient in Fig. 12.24 after 6 weeks of customized therapy including joint and soft tissue mobilization, quadriceps electrical stimulation and use of CKD with special straps for 16 h per day

The authors prefer ASTYM™ to treat the entire kinetic chain for patient undergoing internal lengthening since there are no pins to interfere with the treatment. For femoral internal lengthening, special attention must be given to the tensor fascia lata and iliotibial band as well as the muscles that attach to the tensor fascia lata. Modified Ober positioning (Fig. 12.26)



Fig. 12.26 Iliotibial band stretching in a modified Ober position

with gentle varus mobilization of the tibia as well as prone knee flexion with external rotation of the hip and simultaneous internal rotation of the tibia over femur are useful techniques to target these tissues. Patellar mobilization along with regaining knee joint mobility is paramount. Special emphasis should be given to inferior patella mobilization with knee flexion mobilization and superior patella mobilization with knee extension mobilization. Patellar taping techniques are also a useful adjunct to therapy (Fig. 12.27). The goal of patellar taping is to neutralize lateral subluxation force due to tight lateral structures and encourage inferior movement of the patella.

In tibial lengthening with an internal device, common joint problems include knee flexion contracture and ankle equinus contracture. In cases in which the osteotomy is at the distal tibia, patients may also develop equino-varus contracture due to over pull of the tibialis posterior, in addition to the tightness of gastrocnemius and soleus. Dynamic splints for prophylactic treatment are recommended. CKD bracing with ankle dorsiflexion is useful to manage this. Knee extension mobilization and simultaneous ankle dorsflexion mobilization in addition to subtalar and foot mobilization is instituted. Due to the foot being a small lever, the therapist has to use good foot stabilization technique to obtain an effective stretch. As with all patients undergoing limb lengthening, it is important to continue to work on strengthening the quadriceps and hip abductors since these muscles are often atrophied due to prior immobilization and limited weight bearing.

Gait Considerations

Children with leg length discrepancy (LLD) may also have skeletal deformities, muscle contractures, joint subluxations and muscle weakness. In order to equalize their leg lengths,



Fig. 12.27 Patients with femoral lengthening have a tendency for lateral subluxation of the patella and patella alta. Patellar taping is a useful adjunct to help mobilize patella medially and inferiorly

children with LLD intuitively use compensatory mechanisms such as (1) equinus posturing of the ankle on the short side, (2) pelvic tilt with lowering of the pelvis on the short side, and (3) knee flexion on the long side (Fig. 12.28). Short leg gait has the following characteristics (1) stance time reduced on the short side, (2) stride length reduced on the short side, (3) cadence (steps/minute) increased, and (4) walking velocity marginally reduced. Limb equalization reduces these compensatory strategies and gait becomes more symmetrical [20].

When working with a child with a congenital cause of limb length discrepancy, there are also angular deformities of the bones to consider when evaluating and treating gait abnormalities. At the hip, there may be coxa vara or coxa valga which can impact hip biomechanics and function of the hip abductors. Considering the length-tension relationship of muscles, the abductors rely on the normal femoral neck-shaft angle to be 120°–135°. When this angle is changed the muscles are unable to provide the trunk with a stable base of support at the pelvis, leading to a compensatory Trendelenberg gait pattern.

If there is a fixed flexion deformity of the femur and/or an anterior pelvic tilt, then the hip extension needed for proper propulsion at the end of swing is not available. The child will instead utilize their lumbar spine to allow their center of gravity to move over the stance limb. Even with compensation,



Fig. 12.28 Pelvic obliquity compensation for leg length difference. This results in hip abduction of the short limb and hip adduction of the longer limb

the step length will be shorter on the effected side. An equinus contracture further compromises the child's propulsion.

During normal gait, knee range of motion provides shock absorption and stability in stance and toe clearance in swing. When one has a shortened limb, often the knee is quite stiff in gait, as there is limited need to flex it to clear the ipsilateral foot. Femoral deformities and small patellae can limit quadriceps muscles ability to fully activate. Couple this with cruciate ligaments deficiencies, and the affected child will generally lock his or her knee to prevent buckling and potential injury. Often lack of range of motion at the knee does not affect these patients until their leg length has been equalized and they need to start functioning more normally.

Gastrocnemius tightness is certainly the most common problem at the ankle, leading to difficulty in toe clearance once the limb lengths are equalized and decreased propulsion in stance, since it is not able to stretch and recoil normally. Congenital problems of the lower leg may present with procurvatum or recurvatum of the distal tibia. If there is procurvatum of the distal tibia, then the patient will be in excessive dorsiflexion throughout the gait cycle and propulsion will be difficult. With a recurvatum of the tibia, the ankle is in a plantigrade position, thus preventing the knee

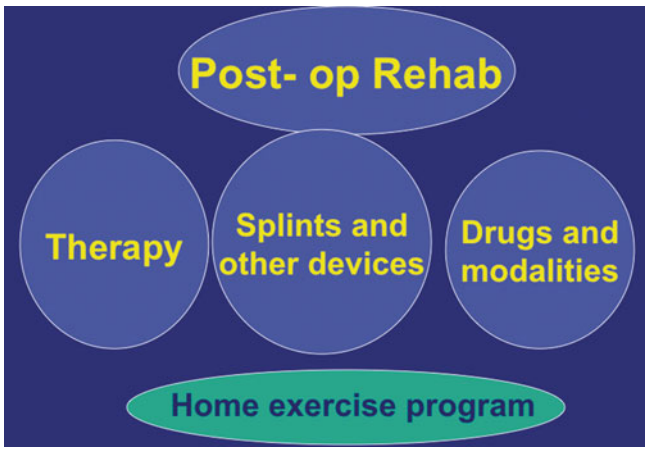


Fig. 12.29 Successful outcome for children undergoing limb lengthening and deformity correction can only be obtained by a carefully designed protocol for the individual patient that involves daily therapy, static or dynamic splints and strict adherence to home exercise program with parent or caregiver participation

from functioning normally in gait and prevents the body's weight from advancing over the foot. The knee will lock out early in gait and will decrease the stride length of the unaffected limb.

Rotational and angular deformities of the lower leg also present problems for all of the joints of the lower extremity as the ground reaction forces will become unevenly distributed at each joint causing abnormal wear and tear and inefficient gait. Normal foot progression is difficult and the possibilities of running and sporting activities are greatly limited the more severe the deformity is.

Summary

Treating patients with limb length discrepancies before, during, and after limb lengthening can be incredibly challenging. It takes understanding of the anatomy of the human body and how is interacts in mobility to effectively determine a proper plan of care for these patients. Customized therapy (Fig. 12.29) is the greatest tool to overcome the common pitfalls in lower limb lengthening and deformity correction to ensure the best clinical outcomes.

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13 Amputation and Prosthetic Management: Amputation as a Reconstructive Option

John A. Herring

When a child presents with a major lower limb deformity, whether congenital or acquired, many treatment options must be considered. Modern limb lengthening techniques with external frames and intramedullary lengthening devices have greatly expanded the treatment. In many cases these technological advances have enabled surgeons to effectively manage deformities which were untreatable in earlier times.

In a similar way, amputation and prosthetic strategies have also advanced dramatically in recent years. Over the last several decades a remarkable increase in social acceptance of visible disabilities has come about. Many factors have contributed to this progress, including the Americans with Disabilities Act, inclusion of Paralympic sports in concert with the Winter and Summer Olympic Games, and an appreciation for the remarkable achievements of prominent athletes with limb deficiencies (Figs. 13.1 and 13.2). Children, who in the past sat quietly on the sidelines because of their "disabilities," are now competing as varsity athletes, cheerleaders, and team captains, using highly functional state-of-the-art prostheses.

Initial enthusiasm for limb lengthening reconstruction for congenital deformities was very high. As the difficulties and complications of such interventions came to light, especially those encountered with "heroic" lengthenings, a reconsideration of the role of amputation has been appropriate [1–7]. Let us consider the example of a child born with fibular hemimelia, or congenital absence of the fibula, and we will compare Syme amputation to correction with the Ilizarov method. A child with a fibular hemimelia with a well-formed, four ray foot and a 10 % limb length discrepancy can expect an excellent result with one tibial lengthening and a contralateral epiphysiodesis and is not usually a candidate for amputation (Fig. 13.3). At the other end of the spectrum, a child with a very short tibia, marked anterolateral bowing, and a two ray foot can expect to achieve his or her full athletic potential with a single surgery and appropriate prosthetic management following amputation done at 11 months of age. (Fig. 13.4a, b) An Ilizarov approach would require three or more periods of frame management, with considerable loss of childhood experiences, with an end result of compromised function and cosmesis. The decision for management of the child with a deformity in between these two extremes is more difficult. The decisions are complex and emotionally laden and require extensive knowledge of all aspects of proposed treatment, and consideration of family and social dynamics [8, 9].

Treatment Concepts

Congenital deficiencies involving the lower extremities present with varying degrees of severity, some of which are best treated with amputation. Amputation in this context is often one part of a complex strategy involving removal of some bony and soft tissue elements while reconstructing other anatomic components. When applied appropriately, amputation becomes a very positive step in achieving the best functional and cosmetic outcome for the child (Box 13.1). Conditions in which amputation is often useful include congenital femoral deficiency, fibular hemimelia, and tibial hemimelia. Amputation for congenital pseudarthrosis of the tibia may be appropriate after failure of other methods to obtain a stable union with a functional extremity, and occasionally amputation is chosen as the primary management for this condition.

Deformity and dysfunction following severe trauma may also be an indication for amputation, either primarily or following attempts at reconstruction. Primary amputation may be indicated in young children when major epiphyses are injured in such a way that future growth is severely limited.

Many methods of limb reconstruction are appropriately used maintain lower extremity function after tumor excision. Allograft and internal prosthetic reconstruction have made a

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Fig. 13.1 (**a**) A young woman with proximal femoral deficiency. Her management was with Symes amputation and knee fusion. She is celebrating her first Paralympic gold medal in three-track skiing at Salt Lake City. (**b**) The same woman as she achieves another Paralympic medal, this time in track cycling. Overall she has now medaled in four Paralympic games

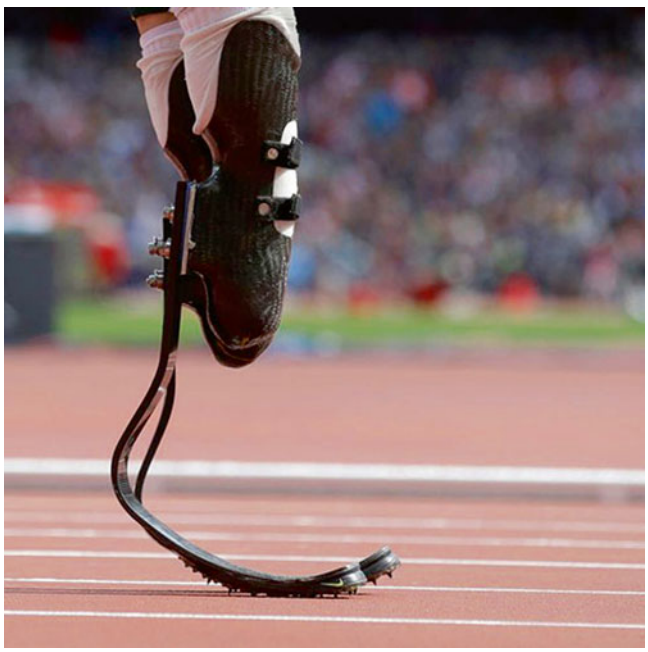


Fig. 13.2 Cheetah type carbon blades. These enable highly competitive performance in a number of sports including track and field, basketball, and football



Fig. 13.3 A boy's foot with fibular hemimelia. This four-rayed foot was functional and had good mobility. Note the Symes prosthesis on the other side, which had a more severe fibular deficiency

Box 13.1.
Amputation as a positive event:
- PFFD
- Fibular hemimelia
- Tibial hemimelia
- Trauma
- Others

positive impact on the management of malignant and aggressive benign tumors in children. Amputation at times becomes a solution to intractable complications such as infection and implant failure after these initial efforts. Amputation remains the best primary treatment option when the size and location of the tumor exceeds the limits of reconstructive options [10–18]. In addition, amputation may be appropriate when the patient presents with wide-spread disease, and amputation can provide pain relief and the ability to ambulate over a shortened life span.

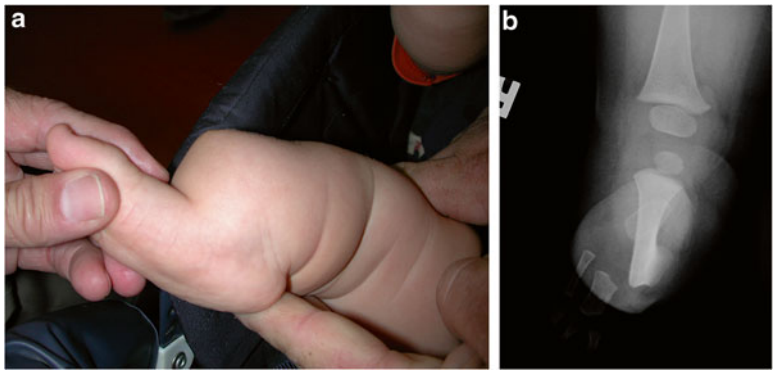


Fig. 13.4 (**a**) A more severe fibular hemimelia with marked shortening and angulation of the lower limb. The two-rayed foot was subsequently converted to a Symes amputation. (**b**) A radiograph of the same leg showing marked shortening and angulation of the tibia

Patient and Family Management

Management options for these abnormalities also vary greatly depending not only on the severity of the condition, but also on the available medical expertise and prosthetic support. Early on, the parents should be introduced to and educated about treatment methods which involve amputation as well as those which involve other methods of complex reconstruction. This education is markedly enhanced when such parents trying to make a decision for their child meet other parents, some of whose children are being managed by amputation and others by limb lengthening reconstruction. These encounters allow parents to ask other parents questions which they would not ask their doctors, and in fact, which the doctors are not really very good at answering.

Historically, amputation was a last resort, dreaded by the surgeon and feared by the patient and caretakers. As prosthetic devices have become more functional and cosmetic, and sports prostheses have allowed people with amputations to compete in sports at very high levels, the concept of amputation as a positive strategy has taken hold in society. Parents and children need to be informed about modern amputation techniques and prosthetic fitting which provide high function with a minimum number of surgical encounters and morbidity (Box 13.2). They will need the knowledge with which to compare amputation to limb lengthening reconstruction, typically requiring multiple procedures over time. They should be provided with honest information about complications and outcomes of all methods, frequency and duration of hospitalizations, and the availability and quality of prosthetic care.

Box 13.2
Education is essential:
- By physician
- By other parents (who have had a child with a similar diagnosis)
- With psychologic support

The comparative costs of prosthetic management and complex reconstruction are difficult to assess. Several studies have shown conflicting conclusions and consideration of all relevant factors is difficult. A most important cost to consider is the lost childhood or adolescence which occurs with multiple episodes of frame application and hospitalizations to manage complications [19–21].

Specific Conditions

Congenital Femoral Deficiency

There is great variation of femoral anatomy among children with congenital femoral deficiency (CFD), and several classifications are useful. The Gillespie classification [22–24] is based on length of the femur at presentation (Fig. 13.5). In type A the femur is at least 50 % as long as the normal femur, and in type B it is less than 50 % of the normal length. In type C there is almost no development of the proximal femur. The classification relates in general to treatment recommendations. The Hamanishi classification [25] illustrates the wide variety of anatomic presentations, but is not specifically tied to treatment (Fig. 13.6). The Paley classification [26] adds consideration of the mobility of the femoral head within the acetabulum, and is useful for some reconstruction methods (Fig. 13.7).

Treatment in Gillespie Type A

These patients, with at least 50 % of normal femoral length can usually be managed with lengthening and reconstructive methods. The outcome is best when the hip is stable and well formed, and when the upper femoral deformity or deficiency can be corrected. The greater the shortening of the femur, the greater is the number of corrective and lengthening procedures that will be required. The lengthening process in this condition is difficult and a number of authors report a high frequency of complications and failures because of shortening and contracture of all the tissues in the thigh [27–29].

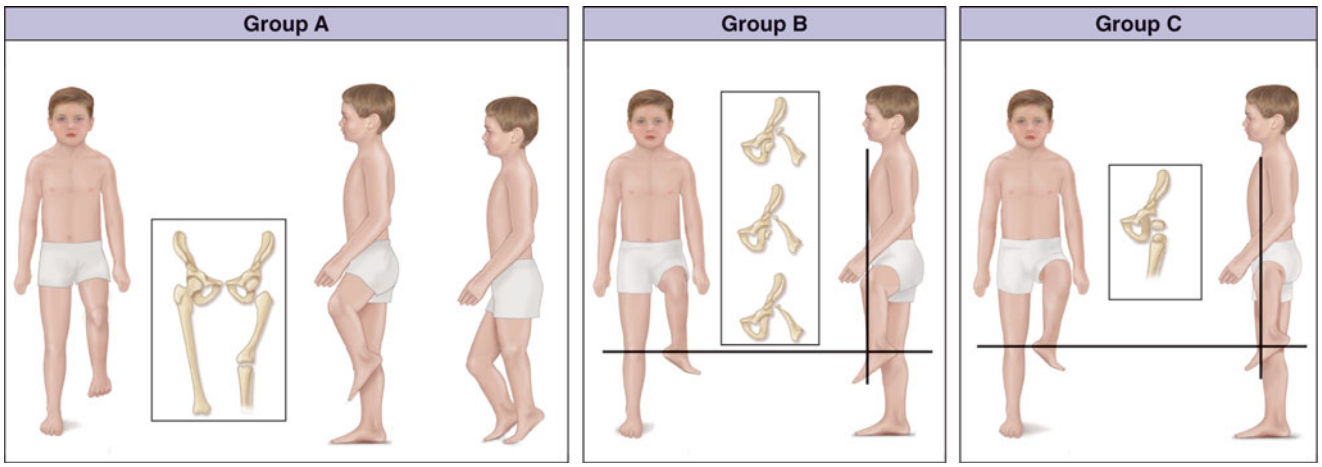


Fig. 13.5 The Gillespie classification of femoral deficiency. In type A the femoral length is more than 50 % of the normal side. In type B, the length of the femur is 50 % or less than the other femur. In type C, there is almost no femoral length. This image was published in Tachdjian's Pediatric Orthopedics, Herring JA, Limb deficiencies, 965–974, Copyright Elsevier 2014

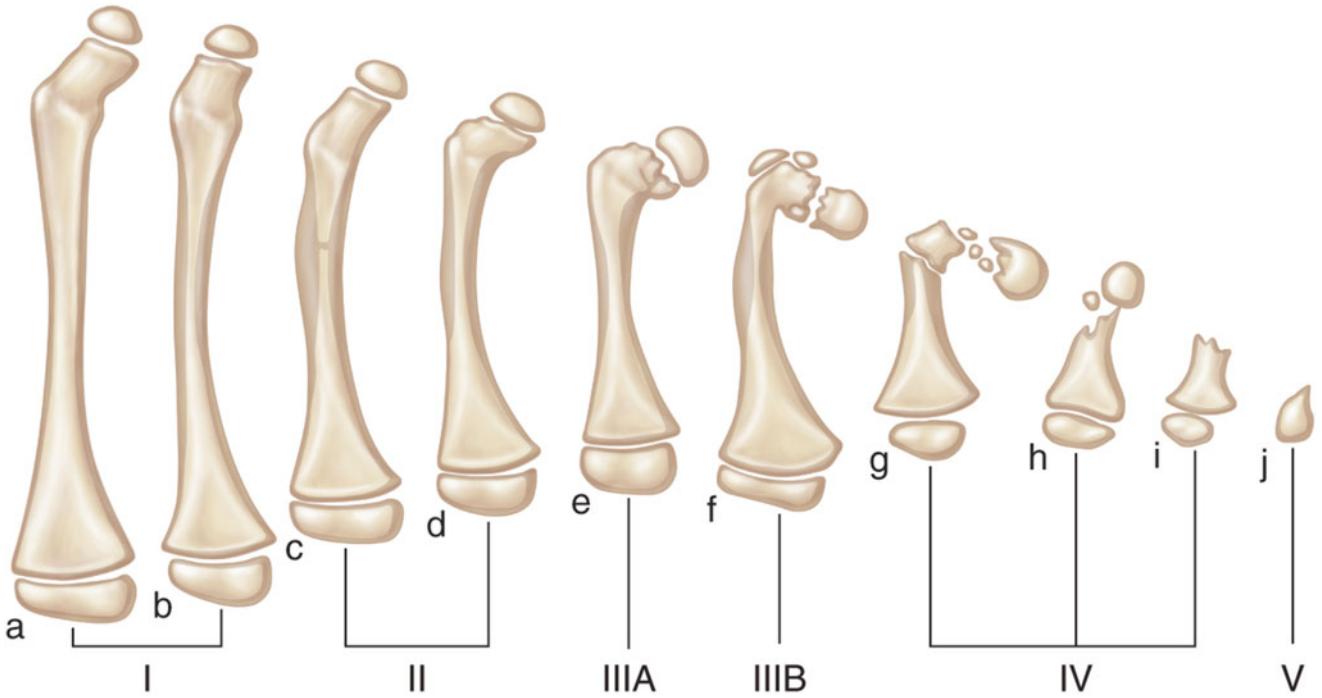


Fig. 13.6 The Hamanishi classification of femoral deficiency. This classification notes the multiple variations of the deformity. This image was published in Tachdjian's Pediatric Orthopedics, Herring JA, Limb deficiencies, 965–974, Copyright Elsevier 2014

Gillespie Types B and C

In these patients the femur is less than 50 % of the normal length, and lengthening and reconstruction will generally require three or more episodes of lengthening. Each of these will consume months of time, and complications often increase with successive lengthening. Likewise, the ultimate function likely decreases as the underlying anatomy is more deficient in cases with substantial femoral shortening. Thus, in this situation some variation of amputation is usually appropriate (Box 13.3). Patients with bilateral femoral deficiency are almost never candidates for amputation, nor are those with major upper limb anomalies requiring the feet for manual activities (Fig. 13.8).

The treatment options in CFD can best be understood relative to the presenting anatomy. Factors to consider include (1) the anatomy of the hip and its musculature, (2) relative shortening of the femur, (3) the range of motion of the knee, (4) the anatomy of the foot. The anatomy of the hip

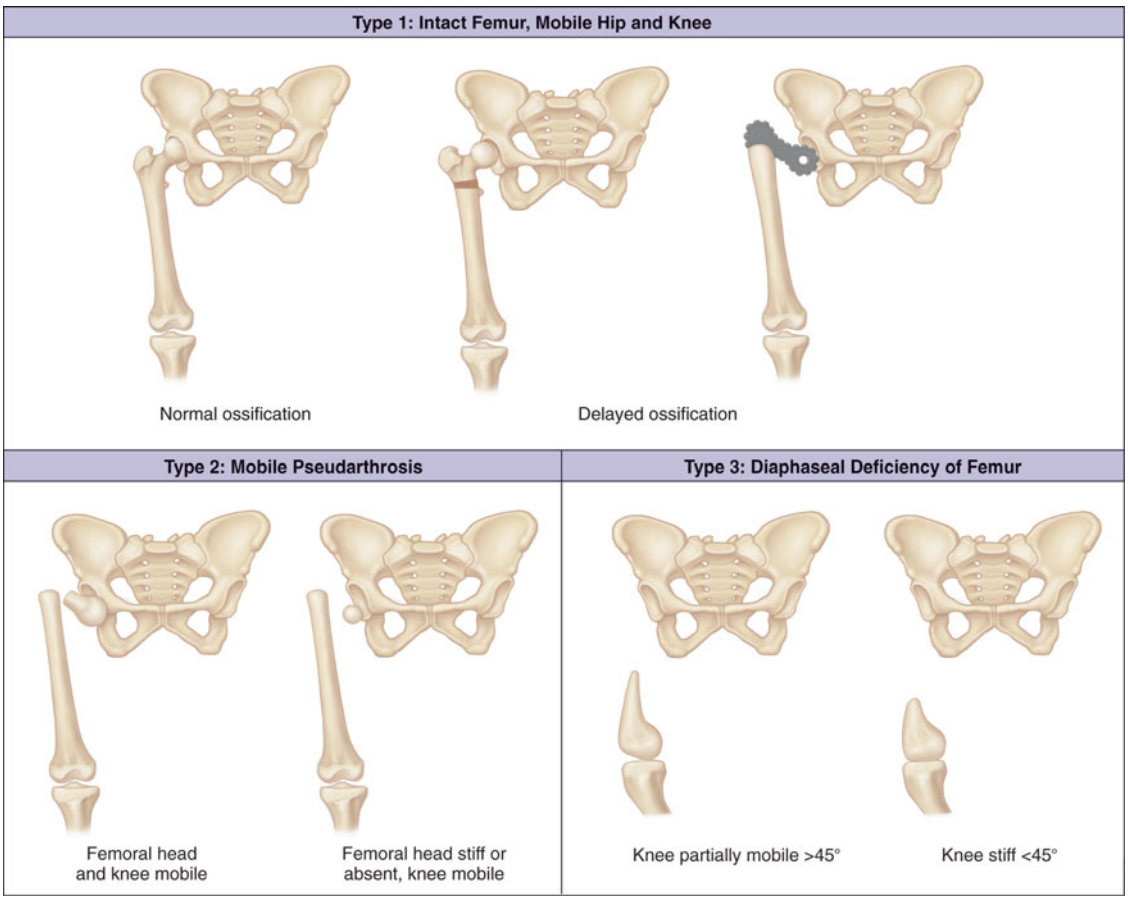


Fig. 13.7 The Paley classification of femoral deficiency. This classification takes note of hip and knee mobility. This image was published in Tachdjian's Pediatric Orthopedics, Herring JA, Limb deficiencies, 965–974, Copyright Elsevier 2014



Fig. 13.8 A radiograph of severe bilateral femoral deficiency, Gillespie type C, in which there is no femoral formation and the tibias articulate with the pelvis. Treatment alternatives are minimal in these children. This image was published in Tachdjian's Pediatric Orthopedics, Herring JA, Limb deficiencies, 965–974, Copyright Elsevier 2014

varies from relatively normal to severe deformity or sometimes absence of the femoral head and acetabulum. When possible, the anatomy of the upper femur should be reconstructed to restore bony continuity of the femur and restore the femoral neck shaft angle. Also, any acetabular dysplasia should also be corrected (Fig. 13.9). When the femoral head has not developed, or is immobile in the acetabulum, hip reconstruction is generally not feasible.

Knee arthrodesis with Symes amputation has been used for many years and provides good function. The child can be fitted with a prosthesis at walking age incorporating the foot in the prosthetic socket, or a Symes amputation can be done at walking age to facilitate prosthetic fitting. Arthrodesis of the knee is usually done at an older age, often age 3 or 4 years, when there is significant ossification of the knee structures. The arthrodesis improves the gait by reducing the flexed position of the thigh segment and aligning the mechanical axis of the limb under the weight line of the body. To have appropriate length of the thigh segment after arthrodesis, the upper tibial epiphysis is usually fused to the

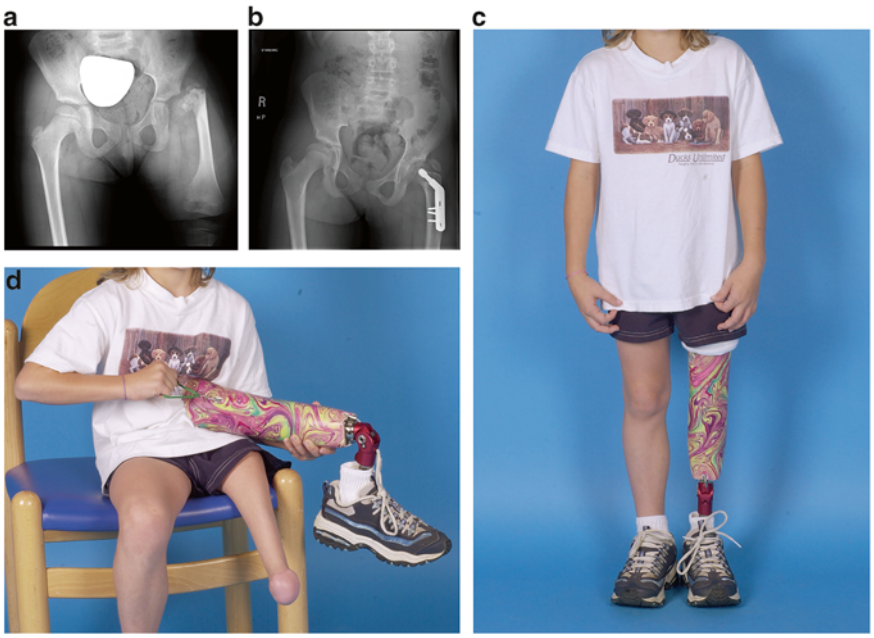


Fig. 13.9 (**a**) A Gillespie type B deficiency with less than 50 % of femoral length. There is a short femoral neck with a marked varus deformity. (**b**) A radiograph of the pelvis after proximal femoral valgus osteotomy and acetabuloplasty. (**c**) The same patient with her prosthesis. Her anatomic knee is intact and flexes just at the top of the prosthesis. She is an active varsity cheerleader with no functional limitations. (**d**) Her shortened thigh segment is evident as she sits without her prosthesis

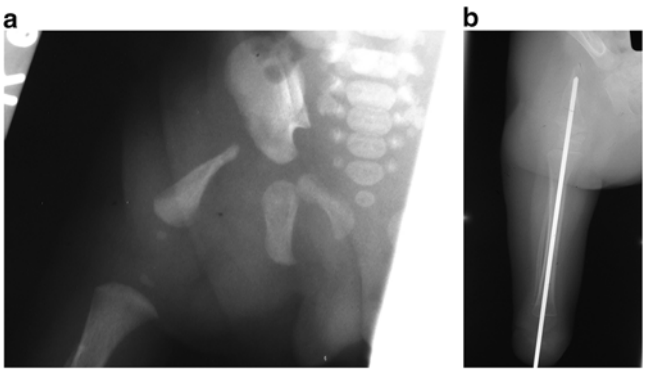


Fig. 13.10 (**a**) A radiograph of a type B femoral deficiency. Note the absence of femoral head or acetabular development. (**b**) The same patient after arthrodesis of the knee and Symes amputation



Fig. 13.11 A 10-year-old girl with a Van Nes type rotationplasty after tumor resection

distal femoral metaphysis with removal of the distal femoral epiphysis (Fig. 13.10). Ideally the end of the thigh segment should be at least 10 cm shorter than the contralateral knee to allow room for the prosthetic knee components. The relative growth of this segment is less than the normal femur by the amount of growth of the contralateral distal femoral epiphysis, since the homolateral epiphysis was removed at the time of knee fusion. As a practical matter, if the child is reaching adolescence and the knees are at the same level with a shorter prosthetic tibial segment, the surgeon should consider performing a proximal tibial epiphysis on the side of the limb deficiency.

As an alternative, rotation of the distal limb to achieve a Van Nes effect may be done at the time of knee arthrodesis [30, 31]. When there is no hip stability, the distal segment has a tendency to derotate with growth, and may have to be repeated.

When the proximal hip anatomy is not amenable to reconstruction, several procedures have been developed in an attempt to reduce the abductor limp which is usually quite noticeable. The most successful has been that developed by Ken Brown in which the femur is rotated 180° and fused to the pelvis in a vertical position [30, 31] (Fig. 13.11). In this position flexion of the rotated knee becomes hip flexion. The fusion of the femur to the pelvis, with full rotation turns the foot backwards with the toes facing posteriorly. The foot is placed in equinus into a prosthetic socket so that the ankle controls the prosthetic knee. When the ankle dorsiflexes the prosthetic knee flexes and when the foot plantar flexes the

knee extends. The anatomic ankle also provides proprioception of "knee" position which is an important advantage for the patient, for example when descending stairs.

Box 13.3.
Options with amputation:
Femoral length <50 %
Hip reconstruction
Knee fusion
Femoral-pelvic fusion
Symes amputation
Rotationplasty

Congenital Fibular Deficiency

While several classifications of congenital fibular deficiency (or fibular hemimelia) are used, we prefer the Birch classification [32], which is based largely on the severity of foot deformity and the relative limb length (Table 13.1). In general, a patient whose foot can be made plantigrade and which has three or more rays may be a candidate for lengthening. When the overall limb length inequality is 10 % or less, lengthening is preferred. With discrepancies between 10 and 30 %, either method may be appropriate. With discrepancies greater than 30 %, often requiring more than two limb lengthening procedures, we usually recommend an amputation (Box 13.4) (see Figs. 13.4 and 13.12).

Our preferred amputation technique for fibular hemimelia is a variation of the Symes procedure in which the ankle is disarticulated without removing either malleolus [33]. The heel pad remains as an excellent weight bearing structure,

Table 13.1 Proposed classification of congenital fibular deficiency based on clinical deformity and treatment based on classification[a]

Type	Characteristic	Treatment anticipated
Type 1 (foot preservable)		
1A	<6 % inequality	No treatment or orthosis or epiphysiodesis
1B	6–10 % inequality	Epiphysiodesis ± lengthening
1C	11–30 % inequality	1 or 2 lengthenings ± epiphysiodesis or extension orthosis
1D	>30 % inequality	>2 lengthenings or amputation or extension orthosis
Type 2 (foot nonpreservable)		
2A	Functional upper extremity	Early amputation
2B	Nonfunctional upper extremity	Consider salvage

[a]Shortening is calculated as a percentage relative to the contralateral limb. In bilateral cases, the longer limb is classified as Type 1A, and the shorter limb is classified as is done for unilateral cases

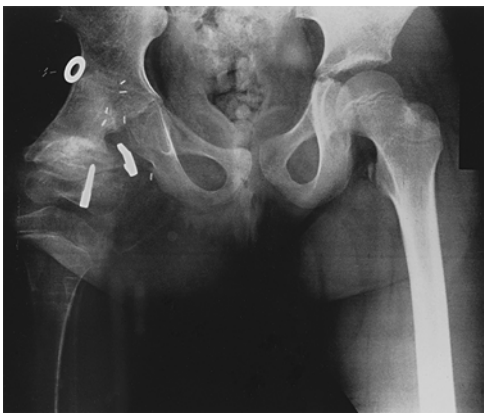


Fig. 13.12 A radiograph of a patient who had a Brown procedure. The proximal femur with a tumor was removed and the distal femur was rotated 180° and fused to the pelvis in an extended position. The staples are placed to stop the growth of the distal femoral epiphysis



Fig. 13.13 A 2-year-old girl treated with Symes amputation for fibular hemimelia

and the incision is anterior away from the weight bearing surface (Figs. 13.13 and 13.14). Some surgeons prefer the Boyd modification with fusion of the calcaneus to the distal tibial articular surface.

The limb length discrepancy can easily be equalized by the varying the length of the prosthetic limb. A moderately shorter limb is ideal so that there is room for prosthetic foot components. Most patients can ambulate well without the prosthesis and compensate for more severe length discrepancy by walking with the other knee flexed.



Fig. 13.14 Appearance of a Symes amputation for fibular hemimelia

Box 13.4.
Amputation considered for:
- Three-rayed foot or less
- Limb length discrepancy >30 %
- Optional for LLD 10–30 %
  *Never with missing upper extremity function

We prefer to perform amputation when the child reaches the developmental stage of cruising, meaning taking steps while holding on. The prosthesis can be available just as the child reaches walking age, and very little training is then necessary. Within a few hours the child comprehends, almost magically, what this addition to his or her anatomy is there for, and away the child goes.

Congenital Tibial Deficiency

The Jones classification [34] is very useful in the management of congenital tibial deficiency (or tibial hemimelia) (Fig. 13.15). In the type 1a, in which there is complete absence of the upper tibia, we consider amputation through the knee (knee disarticulation) to be the best management (Figs. 13.16 and 13.17) (Box 13.5). The older Brown procedure, in which the fibula is centered beneath a bulbar distal femur, was widely abandoned after studies showed almost universal long-term failure. Loder and Herring found that these created knees became stiff and non-functional over several years [35]. The femoral length should be monitored as the child grows, and a distal femoral epiphyseal arrest is often necessary so that the femur is 6 or 8 cm shorter than the contralateral femur at skeletal maturity. This allows space for the prosthetic knee component so that the prosthetic and anatomic knees are at the same level.

When the proximal tibia is present and the quadriceps actively extends the tibial segment, as in the Jones type 1b and 2, a very functional reconstruction is available. The fibula in this instance should be fused to the upper tibia, and the foot amputated at the distal fibular level, maintaining the heel pad. In the young child the upper tibial portion may be mostly formed of cartilage. Fusion of the fibula to the tibial segment requires enough bone formation in the upper tibia to accomplish fixation and fusion. In this situation we often will primarily amputate the foot to enable the child to ambulate in a prosthesis with support above the knee. As the bone formation increases, we perform a fusion of the fibula to the upper tibia with either pin or screw fixation. Once the fusion matures, the child will wear a Symes type prosthesis and usually function at the same level as the child with fibular hemimelia (Fig. 13.18).

Box 13.5.
Amputation appropriate for:
- Jones 1a (knee disarticulation)
- Jones 1b, 2 (Symes amputation, synostosis of tibia to fibula)
- Jones 3, 4-variable indications

The Jones 4 type abnormality has a distal diastasis between the tibia and fibula. Some of these deformities are best managed with Syme amputation and others with reconstruction (Fig. 13.19). These abnormalities vary considerably, and treatment must be individualized for the given anatomy. Elements which may favor amputation include degree of limb length discrepancy, lack of a distal tibial articular surface, relative size and stiffness of the foot. As with other conditions, the choice of treatment is between a complex and multistage reconstruction with an outcome which is hard to predict and a relatively simple amputation surgery which requires prosthetic wear for life.

Congenital Pseudarthrosis of the Tibia

In our experience, children with congenital pseudarthrosis of the tibia are candidates for amputation in two scenarios. The more common need for amputation arises following several surgical interventions with either failure to achieve union, or when refracture occurs. Often various types of surgery have been performed, and the likelihood of a subsequent procedure being successful is low (Fig. 13.20).

Over recent years a small number of families have been referred to us who desire primary amputation for their young child. They have reasoned that the child with an amputation

Type	Radiologic Description	No. of limbs
1a	 Tibia not seen Hypoplastic lower femoral epiphysis	6
1b	 Tibia not seen Normal lower femoral epiphysis	12
2	 Distal tibia not seen	5
3	 Proximal tibia not seen	2
4	 Diastasis	4

Fig. 13.15 Jones classification of tibial deficiency. Redrawn with permission from Herring JA, Cummings, DR. The Limb Deficient Child. In: Morrissy RT, Weinstein SL, editors. Lovell and Winter's Pediatric Orthopedics, vol 2, ed 4. Philadelphia: Lippincott-Raven; 1996

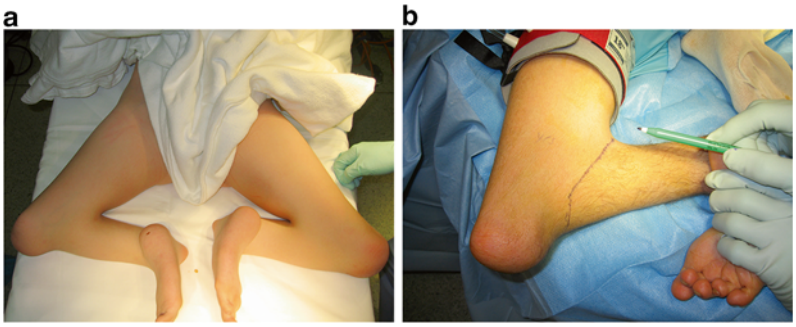


Fig. 13.16 (**a**) A 15-year-old boy with untreated complete tibial hemimelias, Jones type 1a. (**b**) Incision lines have been drawn for knee disarticulation surgery. The suture line does not impair the distal end bearing skin



Fig. 13.17 A Jones type 2 tibial hemimelia. There is a proximal tibial segment which is powered by the quadriceps and hamstrings. The subsequent treatment was a fusion of the tibia to the fibula and a Symes amputation

about the age of first walking will be able to function with prosthetic management and minimal future surgery or no need for protection from vigorous life and sporting activities. We have performed such amputations after careful evaluation of the child and parents' understanding. In addition to meeting other children with similar amputations, our psychologists also evaluate and educate the family.

We perform the amputation in this disorder at the level of the junction of the proximal and middle third of the tibia. Some have recommended Syme amputation with the suggestion that the pseudarthrosis may heal [36]. In earlier years we had no such healing after this approach, and some patients had pain at the pseudarthrosis site requiring reoperation. In addition, current trends for sports prostheses require significant available space for blades and other components that cannot be used with a longer residual limb.

A modification we prefer for this amputation, which also we use for traumatic amputations, is to remove a short segment of cartilage covered bone from the foot to block overgrowth of the tip of the residual limb. We will take a metatarsal head with a short segment of diaphysis and use this as an intramedullary osteochondral plug into the distal tibia. To block the fibular overgrowth we usually use a phalanx from a toe for the same purpose. I should note that we have no firm evidence at this time of the efficacy of this maneuver.

Tumor Reconstruction

While a discussion of the available amputation options for patients requiring amputation for malignancy is beyond the scope of this chapter, certain procedures have proven to be very functional. For proximal femur resections, the procedure described by Ken Brown with rotation plasty gives excellent functional results [30, 31]. The distal femur is rotated 180° and fused to the pelvis in a vertical orientation. The anatomic knee, which is placed at the level of the pelvis, functions as a hip with hinge flexion and extension; the ankle and foot power the prosthetic knee. The resultant function is very good (Fig. 13.21) [37–40].

When a femoral lesion can be excised leaving the proximal femur and hip intact, fusion of the tibia to the proximal femur in a 180° rotated position also provides excellent function, again using the foot to power the prosthetic knee (Fig. 13.22).

With tumors distal to the knee, function of the knee joint should be preserved, if at all possible. In unusual circumstances the distal tibia can be "flipped" to be fused to a remaining proximal tibial segment (Fig. 13.23).

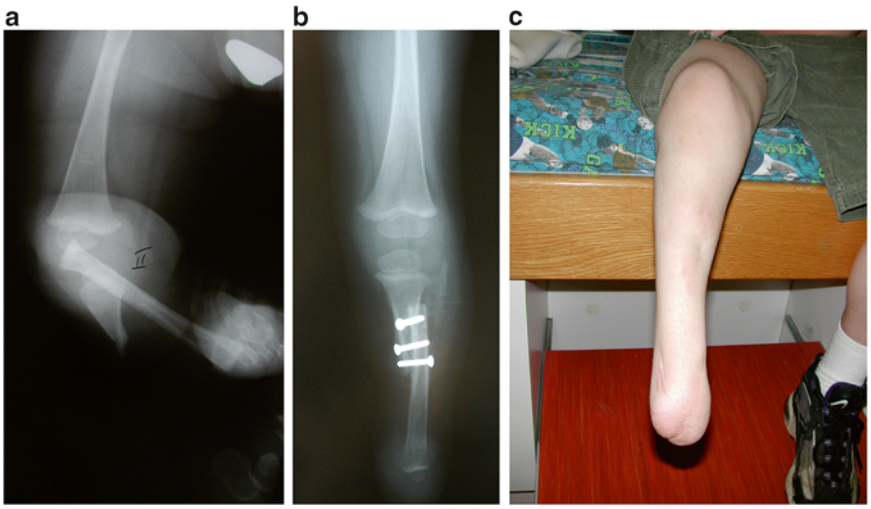


Fig. 13.18 (**a**) A Jones type 2 tibial hemimelia before treatment. (**b**) Radiograph of the limb after fusion of the fibula to the proximal tibial segment. (**c**) Appearance of the limb after fusion is complete. The patient had excellent athletic function



Fig. 13.19 A radiograph of a Jones type 4 tibial hemimelia with a distal diastasis between the tibia and fibula. In this instance there was no distal tibial articular surface and a diminutive foot, and a Symes amputation was performed

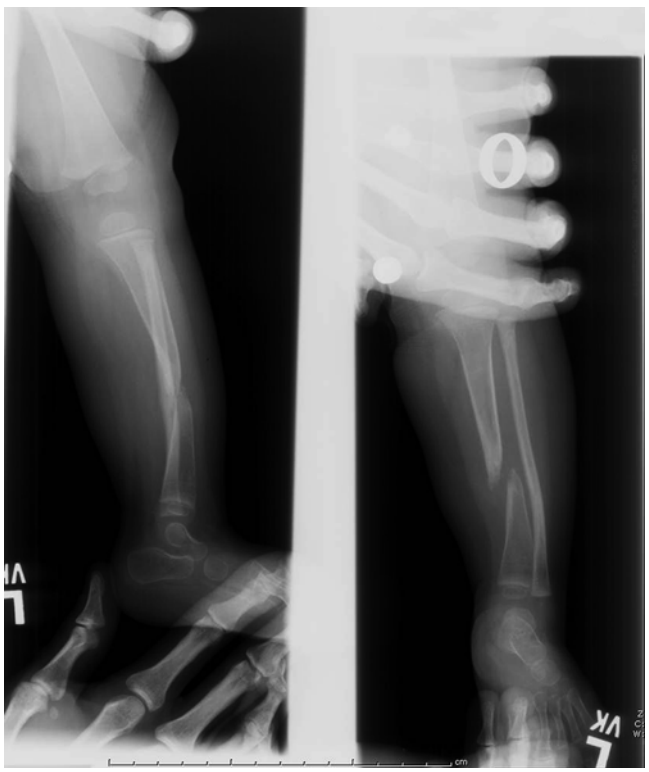


Fig. 13.20 A radiograph of a congenital tibial pseudarthrosis in a girl with neurofibromatosis. After a number of procedures she developed a late recurrent pseudarthrosis and underwent trans-tibial amputation

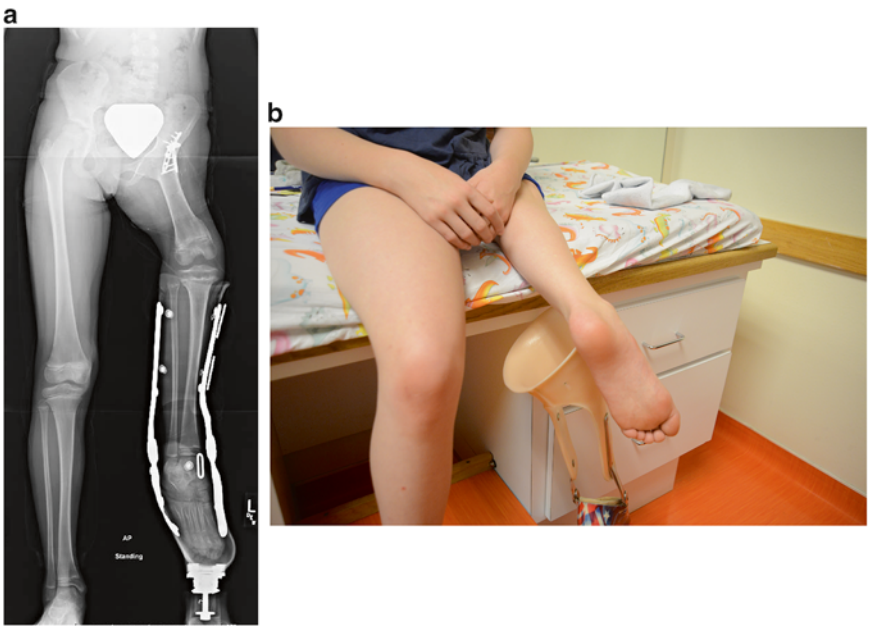


Fig. 13.21 (**a**) A radiograph of a patient treated for a proximal femoral tumor. Her remaining femur was rotated 180° and fused to the pelvis. A revision will likely be necessary to shorten the femur and reduce the abducted position. (**b**) A photograph showing the rotated and very functional foot



Fig. 13.22 A radiograph of a patient who had a distal femoral tumor excised and a fusion of the remaining proximal femur to the tibia, which was rotated 180°

Volume Changes with Oncology

Patients undergoing chemotherapy or multiple other procedures may experience significant volume changes of their residual limb from either weight loss or, conversely, edema. These children may benefit from prostheses with sockets which can be expanded and contracted as necessary.

Amputation After Trauma

Whenever possible the knee joint should be preserved at the time of amputation. Exposed, granulating areas on the upper tibia can be maintained with skin grafting. While some areas may break down, children have better healing properties than adults, and there is great functional benefit to maintaining the native knee joint (Fig. 13.24).

Prosthetic Considerations

First Prosthesis

We prefer to perform indicated amputations at the time when the child is pulling up or beginning to take steps. We then fit the child with a prosthesis at the time when he or she would naturally begin walking. When the amputation is at the knee level or above, the initial prosthesis is constructed without a knee joint. We usually add the knee to the next prosthesis as the child is 18 months or older. With bilateral absences we may wait longer to add an articulated knee to either or both prostheses depending on the child's walking abilities and the length of the residual limb.

Prosthetic Replacement for Children

Children in their growing years will require new prostheses about every 12–18 months. Varying the thickness of socks and liners can sometimes prolong the time of use of a prosthesis. Children with end bearing residual limbs, such as knee disarticulations and Symes amputations, can often wear



Fig. 13.23 (**a**) A radiograph of a patient who presented with a painful valgus right knee deformity. The density in the proximal tibia was an osteosarcoma. (**b**) An AP radiograph showing a complex allograft reconstruction of the tibia. (**c**) A lateral radiograph showing a complex allograft reconstruction of the tibia. (**d**) A radiograph showing the allograft after removal of the fixation devices. Subsequent infection and non-union required consideration for amputation. (**e**) The proximal tibia was retained and the allograft removed. The distal tibial segment was reversed allowing fusion of the more healthy distal tibia to the proximal segment with pin fixation



Fig. 13.24 A clinical photo of a knee and residual tibia following severe trauma. The patient had excellent function in spite of the compromised soft tissue coverage

their prostheses well after they have grown a fair amount (Fig. 13.25). It is not uncommon for a child to prefer the "old" prosthesis to the "new" one, especially if changes in design have been made. Gradually, and often after some modifications, the child will prefer the new limb. When possible the old limb can be modified to be used for swimming, water skiing, or other "rough" activities.

Length of the Residual Limb

A major advantage of the Syme or Boyd amputation is the ability of the patient to weight bear on the residual limb without a prosthesis. This provides the patient with options for activities without the prosthesis such as swimming, walking indoors, and mobility when the prosthesis is not available. The recent availability of prostheses with terminal devices such as the spring devices have changed the desires of many



Fig. 13.25 A young girl standing beside all of her previous prostheses. In a growing child a new prosthesis may be needed as often as every 12 months



Fig. 13.26 A clinical photograph of a transtibial amputation with marked overgrowth of the fibula. It is remarkable that in spite of bone penetration of the skin, infection is very rare



Fig. 13.27 A clinical photograph of a transfemoral amputation. In an effort to keep extra length, a segment of bone was covered with split skin graft. The extra femoral length was not needed and the wound healed after the poorly covered segment was removed

patients. These powerful blades provide the ability to run, jump, and participate in sports up to and including the Olympic level, and require 10 or more cm of space below the end of the residual limb. Some patients with well-functioning Symes amputations have returned requesting more proximal amputation in order to use such terminal devices. Before doing such an operation we make sure that the patient understands the both the functional losses as well as the gains, especially relative to losing the ability to walk without the prosthesis.

Residual Limb Overgrowth

Overgrowth at the distal end of trans-tibial amputations, as well as trans-femoral amputations and others, produce annoying problems for prosthetic wear. Usually a slender spike of bone grows from the distal periosteum and becomes prominent or protrudes from the end of the limb. There is usually a bursa around this bony spicule, which is filled with fluid and bone and cartilage fragments. These are exquisitely painful and often prevent wear of the prosthesis (Figs. 13.26 and 13.27).

The first line of treatment is to resect the overgrowth and shorten the residual limb so that there is ample distal soft tissue coverage. Many efforts have been proposed to reduce the frequency of overgrowth. Placement of a bone and cartilage plug into the canal of the transected bone is sometimes effective. Iliac crest bone and cartilage graft has been used for this purpose, as well as biologic elements such as metatarsal heads which may be available at primary amputation. Excision of distal periosteum at the time of revision has also been advised. Blockage of the distal canal with cement, bone wax, polytetrafluoroethylene felt, and other substances has also been advised [41–49].

End Weight-Bearing

Amputation levels which allow end weight-bearing have a number of advantages, especially in growing children. We often see children who are functioning well in a prosthesis that they have outgrown. These are usually children with either knee disarticulation or Symes-level amputations. While they have grown up out of the prosthetic socket, the ability of the limb to tolerate full weight-bearing has allowed these children to function normally.

The Future of Prosthetics

Future developments will likely add much to our ability to help children with missing limbs. We can expect stronger and lighter materials for prosthetic manufacture. New prosthetic knee mechanisms which provide powered knee extension for stair climbing have just become available and will likely become useful. Running legs continue to be improved to the extent that super-normal performance may soon be achieved. While these powerful blades may make it difficult to determine fairness for sports competitions, the amputee athlete will enjoy greater success on the playing field. Hopefully we will see the development of prostheses which will have permeable components which allow passage of air and moisture to free the wearer from problems of odor and skin rash. Transmission of sensation from prostheses to the person is on the horizon for upper extremity devices and could be useful in lower limb prosthetics as well. Finally, better durability and water proofing would be much appreciated.

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Working in Resource-Challenged Environments

14

Scott C. Nelson and Hugh G. Watts

Introduction

Due to the considerable responsibility that we as pediatric limb deformity surgeons have to our patients and their families, we should never compromise our surgical principles based on the circumstances or location in which we perform operations. The techniques outlined in the chapters of this book have applicability to every part of the world and thus will not be rewritten in this chapter. The limited resource environment presents some unique challenges which often create the need for improvisation and innovation, but this must not be allowed to compromise our results. Indications and surgical techniques may vary based on the type and severity of limb deformities being treated, but surgical principles should remain the same independent of geographic location.

Never before in history have both patient needs and available treatment options been so great. As globalization, communication, and technology increase so do our opportunities to develop and maintain international relationships with other health care providers. This presents possibilities for education and enhances patient care. The potential reasons for the high prevalence of congenital, post traumatic and post infectious limb deformities in resource limited locations includes the following: Lack of environmental regulation (teratogenic pollutants in the air, soil and water), lack of education, urbanization, transportation-related injuries, natural disaster, lack of prenatal care, consanguineous relationships, and limited access to health care. The world population is growing fastest in resource limited environments. The population on this planet is now more than seven billion persons—which is double what it was in the 1970s and quadruple what it was in the 1930s. Income disparity between wealthy and poor continues to increase as well (Table 14.1) [1, 2].

The healthcare resource disparity is even higher at more than 88–1 [3]. Meaning that in places like North America we spend more than 88 times the amount of resources on our own healthcare than what is available for many people in the world. In the USA and in other developed countries, the primary concern is toward the medical care of the aging population. But as Fig. 14.1 [4] shows, children make up a much larger portion of the population in Africa than in the USA. These two population samples exemplify the general trend in population proportion between developed and developing countries.

As pediatric limb deformity surgeons, there is an increasing urgency to engage in service where needs are increasing and resources are relatively decreasing (Box 14.1).

International relationships and mutual understanding between health care providers around the world are necessary to build sustainable programs that will best be able to serve the needs of children with limb deformities. This chapter is written both for surgeons wishing to provide humanitarian service in foreign countries as well as practicing surgeons who are citizens of countries with limited resources.

Box 14.1. Factors affecting prevalence of limb deformities in developing countries

- Lack of environmental regulation (teratogenic pollutants)
- Lack of education
- Urbanization
- Transportation-related injuries
- Natural disaster
- Lack of prenatal care
- Consanguineous relationships
- Limited access to health care

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International Collaboration

Volunteering and Hosting

National and international collaboration between surgeons can be a rewarding experience, both for the host and visiting surgeons and in the end, patient care is improved. Operating together with colleagues and other surgeons is a unique benefit of doing humanitarian work. Every opportunity to share knowledge and techniques with others should be sought, as it provides an opportunity for learning and expanding our ideas.

Good communication is essential to developing a working relationship between surgeons. This starts long before any planned time together and continues on long afterward. In the era of electronic communication it is now easy to do remote preoperative planning, and provide consultation in the follow-up period. Personal preparation as well as preparation of material and supply needs can be optimized by detailed communication.

Table 14.1 Relative wealth ratio between nations in the upper and lower percentiles of economic prosperity showing a severely progressive disparity between the richest and poorest countries in the world

Year	Wealth disparity
1820	4:1
1913	11:1
1950	35:1
2002	75:1

Commit to a Program

As limb deformity surgeons, we will be much more effective if we make a long-term commitment to a program in a resource limited environment rather than making first time visits to a wide variety of different locations. With repeat visits to a specific location expectations are established, relationships exist, communication is simplified and work can be done more effectively. Also, the value of long-term follow-up should be emphasized, both for the patient and for the surgeon. The quest for exploration and adventure is always intriguing and may be appropriate in certain situations. However, this should not be the motivating factor as such notions may dilute the quality and quantity of patient care.

Common Mistakes Made by Volunteers

1. Assume that you are going to teach someone your operative techniques. It is important to go with a humble attitude to first learn and then teach. Often times it is surprising how this attitude benefits everyone.

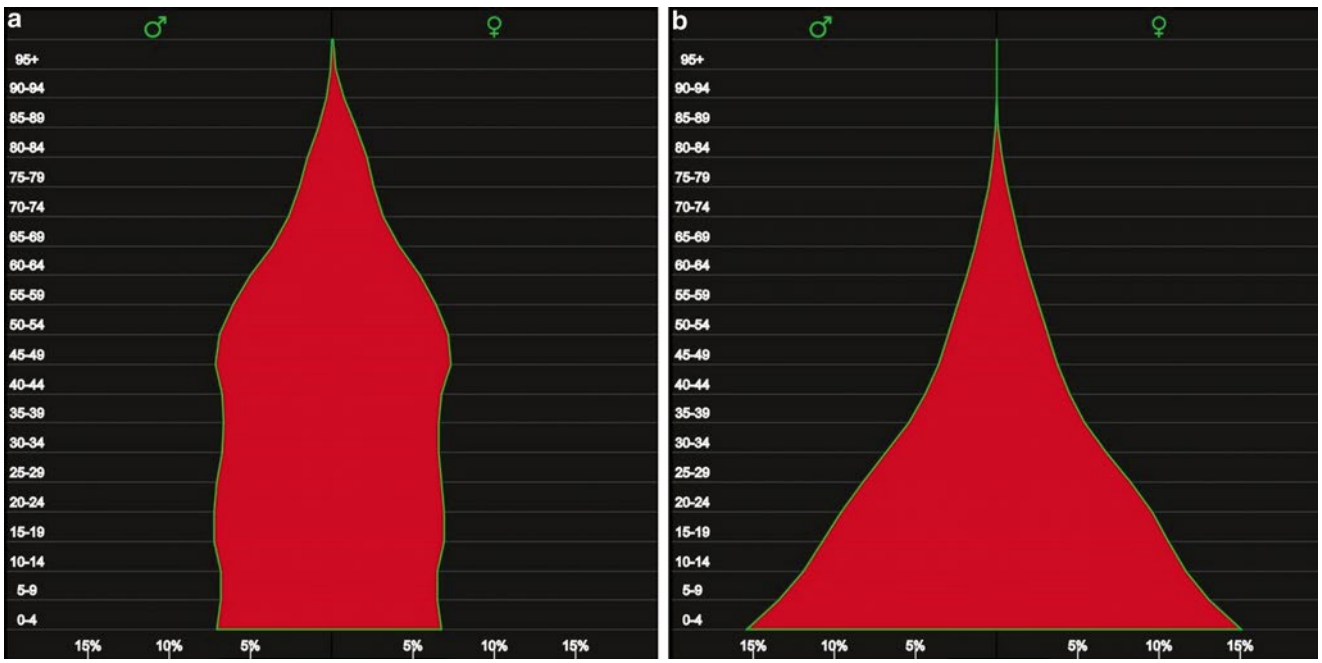


Fig. 14.1 US and African population pyramids. Population pyramids showing the distribution of the population by age in the USA (**a**) and Africa (**b**). 2010 statistics. http://www.worldlifeexpectancy.com/world-population-pyramid

2. Operate late every night and get as many cases done as possible. Situations vary and this may not be a mistake in all situations. However, relationships should be ultimately prioritized over pure production. Consideration must be given to local staff that often repeatedly host visitors and realize that your relationship with them is what will benefit everyone in the long term. As much as it pains the volunteer surgeon to leave behind a child with a deformity that could be corrected, it is sometimes best deferred to a more favorable time for the team and local staff.
3. Use materials and supplies as if you own the place. Opening up multiple packs of sutures, extra gloves, and other materials that may not be essential to the operation is wasteful and may be offensive to the local staff. Many of these items that we take for granted in our own countries are in short supply and are used very sparingly. Even if they are materials brought by yourself, it is best to respect the use of these with the local standard of discrimination since these items can be saved and used for future cases.
4. Make your gifts of materials and supplies the center of attention. Bringing equipment and gifts of supplies is really an obligation to have the privilege of operating in resource depleted hospitals. It must be remembered that volunteers coming to do charity work can be a financial burden on the hosting institution. Patient volume is often increased as are overhead costs for the hospital. It is wonderful if the hosting institution can afford to provide transportation, food and lodging, but in many situations they simply do not have the resources to do this and the mature volunteer should realize that it is often necessary to help with these expenses.

Common Mistakes Made by Hosts

1. Not being responsive to pre- and post-trip communications. Even in the era of modern communication our lives are busy and sometimes important emails and phone messages go unanswered.
2. Not having patients prepared and available for surgery. Hosts must understand that a visiting surgical team often times sacrifices thousands of dollars of lost income to come and do charity work. It is disrespectful if controllable factors have not been appropriately managed to make the most of the trip.
3. Planning operations that are not appropriate for the skills of the volunteers. This is avoided by adequate pre-trip communication and/or long-term relationships. In any case it is essential to know the capabilities of the visiting group.
4. Appropriate equipment exists but it is not available or missing. It is important to be organized and if equipment and materials exist then there is no excuse to not know where they are at the moment they are needed. Many times stock is lacking and this can be communicated ahead of time to the visiting teams who often have access to the needed items at their home institution. Asking for items not needed or that already exist in large quantities is often frustrating for volunteers who sometimes go to great efforts to acquire items only to find a large stock of them in the storeroom.

Understanding Repercussions of Good Work

It may be difficult to understand why, in spite of great need, you are not wanted. This is a common scenario and must be diplomatically dealt with as human needs sometimes may call for you to help in a situation where you are needed but not wanted. It must be understood that our acts of charity can have secondary repercussions. For example, how does the local pharmacist feel when we bring in a big box of medication and now his income is undermined? How do the surgeons across town feel when some Americans come and advertise for free surgery? How does the hosting hospital recuperate costs from increasing the hospital census by 500 % with non-paying patients? Although licensing regulations for physicians and import taxes on gifts in kind are often annoying and have been subject to corrupt government processes, it should be understood that the basis behind this is to protect the economic infrastructure of the medical community. There are no easy answers to these questions as the needs of the people are often times dire, and the barriers can sometimes seem insurmountable. An important concept to keep in mind when navigating these issues is to go with the idea that you are helping more than just individual patients. Go with the idea that you are going to help a doctor, an institution, a medical community, and a country as well as the individual patient (Box 14.2).

Keep Your Motives Pure

There are many reasons to do charity work. Travel, adventure, learning, publicity, and feeling good about oneself are some reasons that may motivate surgeons to volunteer, but should be put secondary to the sincere desire to provide a needed service in the most effective way. Is it OK to have ulterior motives as long as we are doing good work and people are getting helped? This is similar to having a conflict of interest with a particular implant company or letting personal economics affect your surgical decision making. Yes, patients can still be helped in a good way, but this can lead to poor judgment and places the priority on benefiting yourself more than the patient—a dangerous place to be.

Credentialing and Liability

Credentialing and medicolegal concerns are becoming more important issues even in resource limited environments and there are a few basic precautions and diplomatic principles

that should be understood. Although it can be liberating to practice in environments where bureaucracy and liability issues do not take precedence over patient care, it must be remembered that, this freedom should not encourage renegade behavior. Doing what is right for the patient should always be your guiding principle and you should make efforts to assure that those working around you realize that. Never should a surgeon work outside his expertise and/or perform procedures that he is not licensed to do in his home country. As a guest surgeon respecting the credentialing rules of a host institution and country is important. Many resource limited countries have licensing provisions for surgeons coming for short periods of time to do charity work. The general motive of the licensing entity in these countries is to protect the economic viability of local surgeons and make sure that there is a minimum standard of quality amongst foreign health care workers. Often times the credentialing process is rather simple or does not exist at all, but no matter what the case, having a gracious attitude and building relationships with the local staff and administration is an important part of the process.

Medicolegal issues are bigger concerns in North America than most other parts of the world. Nonetheless a few precautions are warranted. The issues vary greatly depending on the location, type of patients being treated, and organizational affiliations. Most legitimate organizations can provide some information on the subject and have provisions in place to protect volunteer surgeons. Many times in these environments it is the hospital or international organization rather than the individual surgeon that carries the burden of malpractice. However, one unique and potentially serious issue is the situation of a US surgeon operating on a US citizen in another country. There have been reports of such cases being litigated in the US court system.

Aside from the subject of formal litigation, disgruntled patients often access news media and internet venues that provide an easy and cost free way to damage the reputation of doctors and institutions. Whether the allegations are perceived or real, these types of patients can do significant damage. It goes to emphasize that in any part of the world maintaining good communication with patients can help to maintain a strong reputation and prevent misunderstandings and possible litigation.

Box 14.2. Keys to Successful International Collaboration

- Good communication
- Develop relationships through long-term commitments
- Be humble—first learn and then teach
- Do not overburden the system
- Respect the local infrastructure
- Understand the possible negative repercussions of good work
- Keep your motives pure—do not do the right thing for the wrong reasons
- Do not perform procedures that you are not properly qualified to do

Operative Challenges: Improvise Don't Compromise

Indications

In general, operative indications should follow the same basic concepts in any part of the world. This involves evaluating whether or not the benefits outweigh the risks. As circumstances vary, so does the relative relationship between benefits and risks. Efforts should not be spared in trying to obtain the necessary equipment and expertise to do an operation with the very best quality possible. We cannot let inconveniences and lack of proper equipment dictate what we do. Following the principle of doing it right or not doing it at all is important no matter where the location (Box 14.3).

Economics have to be considered but should not be the most important factor. They can have paradoxical implications in environments where the reuse of external fixation is commonplace. For example, treating a Blount disease with an expensive multiplanar fixator that can be reused many times may be cheaper than using internal fixation. Whereas in North America, those same sophisticated external fixators may be cost prohibitive for correcting a simple uniplanar deformity. The reuse of external fixator equipment can enable the surgeon to operate without regard to the cost of each additional component that is added for strength and/or cosmesis of the external fixator. Return trips to the operating room for adjustments to fixators can be simple and inexpensive in the developing world without the need for third party payers to approve the process.

The indications for limb salvage versus amputation deserve mention here as these are complex issues even in our own country where we understand the economics and many of the cultural issues. The relative importance of functional outcome, cultural views, and cost of reconstructive procedures versus cost of lifetime prosthetic maintenance take a paradigm shift and differ greatly from country to country depending on the availability of prosthetic services. Often times the expectation of performance is different in less developed environments. In economically privileged countries the debate centers on what option is best for high performance in sports and long distance running, whereas in economically challenged situations most people would be happy for any type of mobility as long as they can keep their own leg. With that being said, reluctance should be even higher to perform amputations in resource limited environments even if it means the functional result is not as good or if a more complex operation or leg lengthening process is required. However, multiple attempts to repair a useless limb need also to be avoided.

In the situation where fluoroscopy is not available, certain procedures are impossible to perform. Indications may change based on whether or not fluoroscopy may be available in the future or at another accessible location or whether it is not an option at all. The philosophy of not doing an operation unless it can be done well should be respected. Many times the lack of fluoroscopy can be compensated for by making larger incisions and/or performing more traditional operations such as open, instead of closed epiphysiodeses. The Watson Jones anterolateral approach to the hip is another example of this, allowing visualization of the femoral head and neck for cases that could otherwise be done in a more minimally invasive fashion if fluoroscopy were available.

Perioperative and Anesthetic Considerations

In areas where an extensive infrastructure does not exist, operations must be performed with a wider safety margin. This calls for a careful preoperative evaluation, additional intraoperative precautions, and careful postoperative vigilance.

Preoperative evaluation should be done both by the surgeon and anesthesia staff. It should be realized that in areas where nutritional deficiencies and parasitic diseases are endemic that anemia can be common. Even when preoperative lab evaluations are not routinely indicated in first world settings, it may be prudent to do additional work up in other settings. Sometimes in spite of limited resources, local bureaucracy requires seemingly unnecessary testing which has to be respected by volunteer staff. There are a couple of portable devices which can be extremely helpful in these situations. The HemoCue device (Fig. 14.2a) is a relatively inexpensive piece of equipment (approximately $600 USD) and is an efficient and cost effective way to evaluate preoperative hemoglobin levels (cuvettes cost ~$2 per patient depending on quantity purchased). The i-STAT (see Fig. 14.2b) is approximately $10,000 USD, but is able to perform a variety of tests including CBC, chemistry panels, and blood gases (cartridges ~$8–10 for each point of care test).

Informed consent is something that should not be forgotten. Even if it does not involve a piece of paper, it is important to communicate to patients, family members and even local staff the plan of the procedure and the possible risks and complications. This is especially important for ablative procedures, complex surgery that involves considerable risk, and for procedures that involve gradual corrections and limb lengthening which require additional patient and family commitment.

Intra-operative blood loss needs to be minimized and carefully assessed. It is important for the surgeon to know how to calculate the maximum allowable blood loss (MABL). The following formula is used:

$$\mathrm{MABL} = \mathrm{EBV} \times \left(\mathrm{HCL_i} - \mathrm{HCT_f}\right) / \mathrm{HCT_f}$$

EBV—Estimated blood volume (average blood volume [ml/kg] × weight [kg])
Average blood volume
Neonates 85 ml/kg
Infants 80 ml/kg
Adult men 75 ml/kg
Adult women 65 ml/kg
HCT_i—Initial hematocrit
HCT_f—Final hematocrit (lowest acceptable at end of operation)



Fig. 14.2 (**a**) HemoCue. Reprinted with permission of HemoCue America. (**b**) I-STAT. Reprinted with permission from Abbott Point of Care

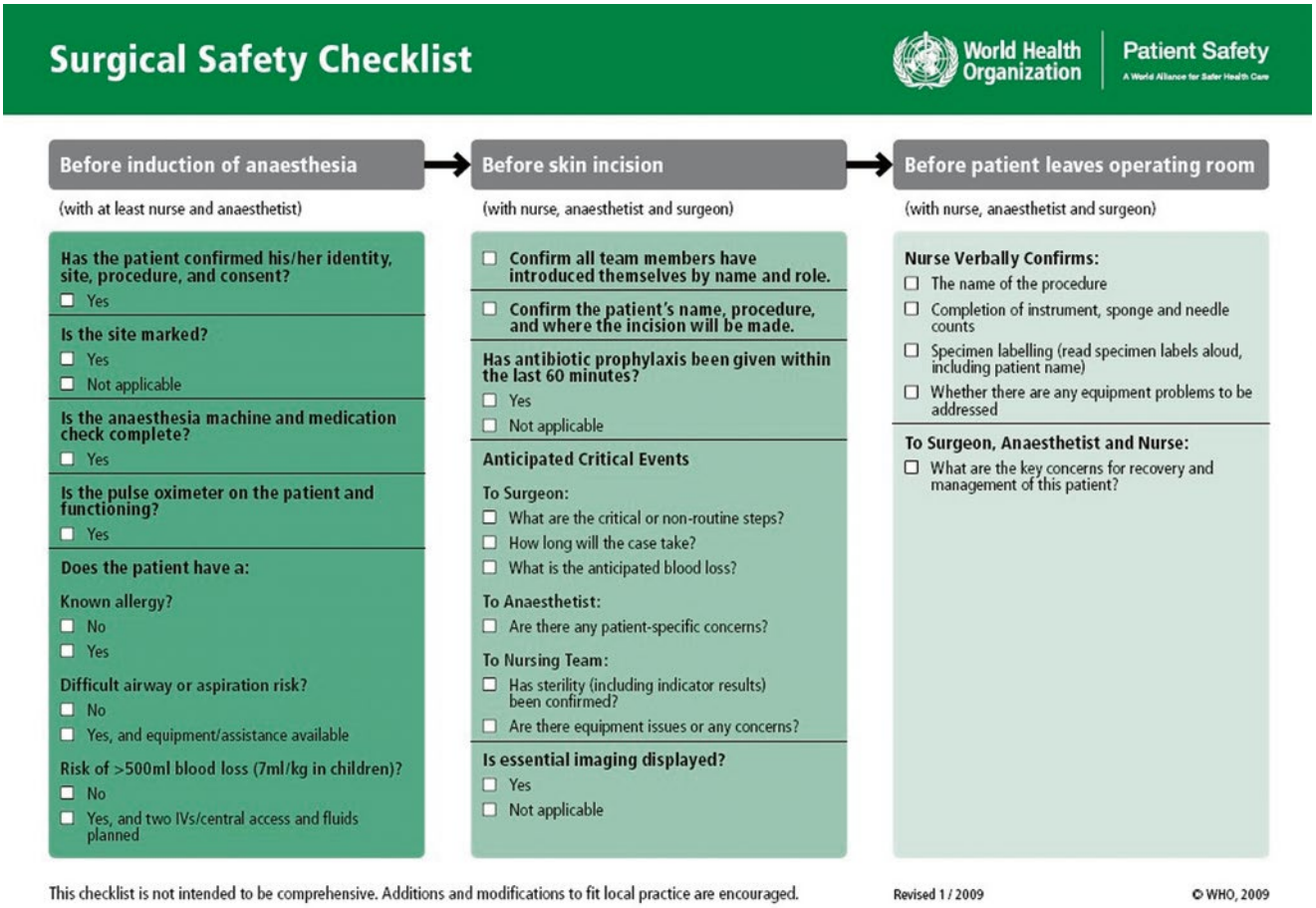


Fig. 14.3 WHO surgical safety checklist. WHO surgical safety checklist, URL: http://www.who.int/patientsafety/safesurgery/en, © World Health Organization 2008 All rights reserved

A healthy patient with a normal starting hematocrit can usually tolerate an acute loss of one third of their blood volume. If you consider that a total estimated blood volume (EBV) is approximately 70 ml/kg (slightly more in infants and less in adult females) then a short way to estimate MABL is to divide 70 ml/kg by three, which is just over 20 ml/kg. *Most patients who are healthy and have normal starting hemoglobin will tolerate 20 ml/kg blood loss without needing transfusion*. In many situations blood may not be available and when it is, often it can take days to obtain. Thus the responsibility lies with the surgeon to make sure that there is a wide margin of safety in regards to surgical blood loss and to always be prepared for the worst possible scenario.

By using these calculations, objective and well informed decisions can be made in regards to performing multiple procedures in one operation versus staging procedures to decrease blood loss. This is especially applicable to children with significant deformities in bilateral lower extremities such as osteogenesis imperfecta, skeletal dysplasias, and rickets. Using the Esmarch bandage as a sterile tourniquet is a safe and effective method of controlling blood loss. Care must be taken to apply it as a wide band tight enough to avoid a venous tourniquet but not overly tight to cause bruising or injury. Any time an Esmarch is used an audible alarm must be set in order to avoid inadvertently leaving it placed beyond 2 h and causing irreversible injury.

The World Health Organization has created a Surgical Safety Checklist (Fig. 14.3) in the interest of reducing surgical deaths and operative morbidity. This is not meant to be a regulatory document but rather a guide to reinforce accepted principles of surgical safety and to promote teamwork and communication. The principles outlined here are basic elements for performing safe operations and should be implemented in every surgical venue.

In the postoperative period, it must be remembered that pain is the fifth vital sign and in North America there is a mindset that less is always better than more. This is not actually in the best interest of patient safety especially in an environment where sophisticated monitors do not line the walls, nurses may not have the latest Pediatric Advanced Life Support credentials, and the code cart is missing or not well stocked. Until an entire infrastructure can be created to remedy these issues then

patient safety should be prioritized over tolerable levels of pain. Fortunately, in these environments patients expect that major surgery will involve some pain and they are much more tolerant of it. As surgeons working in this environment we also need to be trained and updated on resuscitation credentials as we sometimes cannot depend on those around us to do the task.

Sterility

Sterility is a principle that should never be compromised no matter what the situation. A brief discussion is warranted about several options for sterilization, since we as surgeons are not typically involved in the process and do not have the knowledge to assess whether or not various methods provide safe sterilization. In order for something to be considered sterile with autoclave processing, it must reach 121 °C/250 °F with a pressure of 15 psi/100 kPa for 15 min. Indicators are available and should be used routinely. These are an easy and compact addition to the list of essential surgical materials that can be taken on a trip. This can be especially important in situations where aging autoclaves and maintenance schedules may not be strictly adhered to. If a reliable autoclave is not available some portable stovetop units can be acquired and their cost is not prohibitive (Fig. 14.4). These units come in various sizes and can accommodate a variety of surgical instruments; the larger sizes can accommodate larger instruments and implants but are not able to accommodate intact full size instrument trays.



Fig. 14.4 All-American stove-top sterilizer autoclave. Reprinted with permission of Wisconsin Aluminum Foundry

Cidex®

Cidex® is a disinfectant manufactured by Johnson & Johnson that comes in three different variations: Cidex® 14-Day (glutaraldehyde), Cidex® Plus 28-Day (glutaraldehyde), and Cidex® OPA (ortho-phthaladehyde). These provide a fast and effective method of sterilization for a wide variety of instruments and external fixator parts etc. It can be useful for heat sensitive materials that are not destroyed by submersion. When used appropriately Cidex® is bactericidal, fungicidal, tuberculocidal, and sporocidal. Although it can be quite effective, it is very sensitive to dilution, storage methods, expiration dates and duration of use. Similar to opening a carton of milk, it has an expiration date and a limited period of use once opened and poured into a sterilization tray. Due to this variability, it must not be used indiscriminately and we do not use it for implant sterilization. There are indicator strips available which can be helpful to prove efficacy of sterilization.

Ethylene Oxide

Ethylene oxide, known by the trade name Anprolene®, is especially useful for sterilizing heat sensitive materials. It is a highly toxic and explosive gas which was used for making bombs in World War II. It can provide safe and effective sterilization of heat sensitive materials such as gowns, drapes, gloves, Esmarch bandages, Ioban®, cautery pencils, suction tubing, bulb syringes, suture packs (Nylon, Monocryl, Vicryl), and electronic devices (drill, camera, etc.). Most items with the exception of food, drugs, and liquids can be sterilized with ethylene oxide. There are some commercially available processing units that provide ventilation pumps, purge pumps, aeration, and monitors that track cycle progress. Special plastic sterilization bags and ethylene oxide ampules are marketed to be used with these units. For emergencies and austere conditions these plastic sterilization bags and ampules can be safely used without the processing units if appropriate precautions are taken.

Operating Room Set Up

Optimizing every controllable factor includes arranging the operating room furnishings before an operation in order for the operation to proceed as smoothly as possible. Many operating rooms are small and in order to comfortably operate and prevent people from contaminating the field it is important to remove all nonessential pieces of equipment from the

room and arrange the remaining furnishings in an ideal position to work effectively. Don't start operations with the C-arm on the wrong side of the table or the cautery unit inconveniently located. Patient positioning and draping should follow the same philosophy. The surgeon must take the lead and cannot expect all these things to fall into place automatically as may happen with an experienced surgical team at their home institution. It is also essential that equipment outside the operating room is well organized and available at the moment needed. It is not acceptable to have equipment on hand that is either not known about or not available for use at the moment needed. Part of being a good surgeon is to know your equipment and have it prepared to implement the preoperative plan and to also be prepared with a backup plan should the operation take a different direction than anticipated. This sometimes requires spending almost as much time in the storage room organizing equipment and preparing for cases than is spent in the operating room actually doing the cases.

Surgical Modus Operandi in Resource-Challenged Environments

Do not exceed the capacity of the local infrastructure with sophisticated operations that will not be correctly followed up. Placing a Taylor Spatial Frame in an environment where no one knows how to manage it would be an example of this. Also performing highly invasive operations that potentially could result in medical complications that the local infrastructure is not prepared to deal with could be disastrous.

The pros and cons of internal vs. external fixation in a primitive environment have a different dynamic than in the USA. The "one and done" philosophy should be of primary consideration. This means that gradual corrections, which require diligent follow-up and can involve delayed complications, should be avoided when possible. And when internal fixation can be used, it is usually preferable.

Internal vs. External Fixation in Resource-Challenged Environment

Benefits of internal fixation:

- Less postoperative maintenance necessary.
- No pin site infections and less risk of late osteomyelitis.
- Less soft tissue irritation allowing easier range of motion.
- May result in earlier return to function.
- Better for treatment for femoral deformities due to less soft tissue concerns.
- Often does not need to be removed.

Benefits of external fixation:

- Can be applied with minimal incisions.
- If equipment can be reused:
  - Inventory is more easily maintained.
  - Expenses are minimized.
- Enables gradual corrections and bone transport in appropriate situations.
- Facilitates intentional displacement of osteotomies that are performed away from the apex of deformity.
- Postoperative alignment can be more easily adjusted.

Another option is to consider the use of Steinmann pins or K-wires for fixation. This is a very economical approach to osteotomy fixation. In extreme situations bicycle or motorcycle spokes can be sharpened with a small bench grinder, sterilized, and used for fixation. In addition to the biological benefits for making osteotomies in metaphyseal bone, the larger surface area provides more stability and also facilitates stable pin/wire fixation methods. These can be buried under the skin if there is a high risk of infection, but this obligates another procedure for removal. If left in a percutaneous position then a large portion of wire (2 cm or more for most situations) should be left outside the skin and generously bent in order to prevent migration and an otherwise unnecessary return trip to the operating room.

Reuse of Implants and External Fixation Components

The reuse of implants (including plates, screws, intramedullary rods, and bone fixation half pins) is not something that can be universally recommended due to variability in implant strength, size, material, and variability in the magnitude of stress and number of cycles to which it was subjected. Catastrophic failure can occur when cyclic stresses that exceed the endurance limit of a metal are applied to an implant over a given amount of time. The endurance limit is an amplitude of stress below which it appears that even with an indefinitely large number of loading cycles failure will not occur. The endurance limit is usually about half of the ultimate tensile strength of an implant, unless it is notched, which can significantly decrease the endurance limit. Some metals such as aluminum do not have an endurance limit and even small amounts of cyclic stress will eventually cause failure. Titanium and the ferrous alloys like stainless steel are materials that *do* have an endurance limit, meaning that small amplitude (below endurance limit) loading cycles will theoretically not cause failure. Practically speaking, it is not possible to know the magnitude of stress and number of cycles to which an implant has been subjected nor is it possible to visually decipher this with the naked eye. Ultimately, the risks and benefits of reusing an implant must be left up to the judgment of the surgeon. If an implant is reused, it is advisable to carefully examine it, and consider using more robust constructs to decrease the amount of stress on an implant

thus increasing the number of cycles to failure. Using common sense and appropriate reprocessing methods, most implants can be reused without complication.

The implications of reusing external fixators in resource limited environments are different from North America due to perceptions and medicolegal issues. The real question is: What is safe and effective for the patient? In parts of Europe (where resources are not severely limited) and in developing countries fixators are reused on a regular basis. In the USA many external fixators are labeled "single use only." Due to an interest in cost containment and social responsibility many institutions in North America have sought ways to reprocess single use devices (SUDs). The Federal Drug Administration (FDA) has thus classified SUDs into three categories: noncritical risk (level 1), semi-critical risk (level 2), and critical risk (level 3) [5]. The reuse of fixator clamps and bars that do not penetrate the skin are considered level 1 SUDs by the FDA. They have approved various reprocessing protocols for these devices. The reprocessing protocols include cleaning with various solutions to remove all biologic material, dismantling the device, inspection of components for defects or fatigue cracks (usually with magnification or microscope), and re-sterilization.

Biomechanical effects of cyclic loading and patient outcomes with the reuse of fixators have been studied. Matsuura et al. report the effects of various magnitude and quantity of load cycles on the EBI Dynafix external fixator [6]. They loaded one group with 100–450 N of compressive force for three million load cycles to simulate three uses of the device. They found that 17 % of initial stiffness was lost and that some fatigue cracks began to develop at three million cycles. Another group was subjected to higher loads and it was found that fatigue of the device occurred much sooner. Their advice is that external fixators should only be reused a limited number of times and should be carefully inspected. Fixators with more articulating parts certainly are more prone to failure than simple rods and rings. Also fixators that are applied to large patients, highly active patients, and fixators that have been left in place for prolonged periods of time have a higher risk of failure vs. those applied for damage control orthopedics and have only been left in place for a few days or weeks. Several clinical studies have been published showing no loosening of components, no loss of fixation, no increase in pin site infections, and no mechanical failures when reusing fixators [7, 8]. Even if there is a failure of an external fixation device it can usually be resolved with noninvasive and often times nonoperative means.

In spite of these results, commentaries have been published against the reuse of external fixation devices [9] that are not scientifically based, do not apply to limited-resource environments and add fuel to medicolegal fears. The reuse of external fixators can be safely done if the appropriate reprocessing methods and inspection are implemented.



Fig. 14.5 12v cordless variable-speed drill

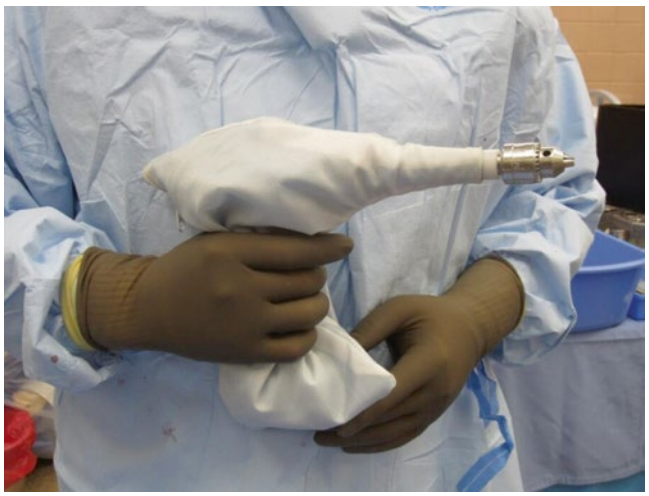
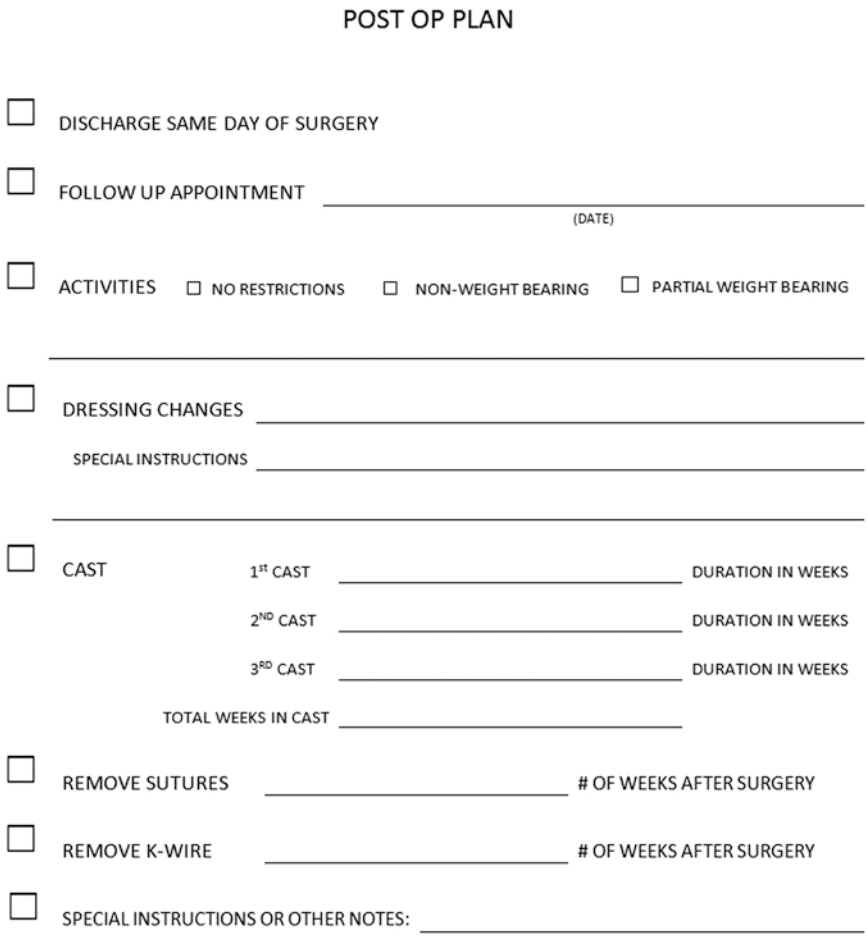


Fig. 14.6 Cordless drill kit shown with sterile cover and surgical chuck

Cordless Power

Many orthopedic operations require a power drill. In limited resource environments the prohibitive costs of the systems that are on the market have led us to develop a safe and effective alternative (Figs. 14.5 and 14.6). We utilize a common hardware store drill with a sterile surgical cover and stainless steel surgical chuck. We use a 12-v cordless variable speed drill with revolutions up to 1,050 rpm. Drills that function at rpms above 1,500 in general are not appropriate for orthopedics as they create excessive heat and can cause osteonecrosis. The stainless steel surgical chuck is custom made in order to facilitate the use of K-wire placement and convenient loading of various drill bits and it can withstand the demands of harsh solutions and repeated cycles of steam sterilization. The system is used with an impervious cover that can also be autoclaved. Theoretically, any drill could be

Fig. 14.7 Post-op plan


POST OP PLAN

☐ DISCHARGE SAME DAY OF SURGERY

☐ FOLLOW UP APPOINTMENT _____ (DATE)

☐ ACTIVITIES ☐ NO RESTRICTIONS ☐ NON-WEIGHT BEARING ☐ PARTIAL WEIGHT BEARING

_____

☐ DRESSING CHANGES _____

SPECIAL INSTRUCTIONS _____

_____

☐ CAST 1ST CAST _____ DURATION IN WEEKS

2ND CAST _____ DURATION IN WEEKS

3RD CAST _____ DURATION IN WEEKS

TOTAL WEEKS IN CAST _____

☐ REMOVE SUTURES _____ # OF WEEKS AFTER SURGERY

☐ REMOVE K-WIRE _____ # OF WEEKS AFTER SURGERY

☐ SPECIAL INSTRUCTIONS OR OTHER NOTES: _____

made sterile using ethylene oxide and thus not require special covers and surgical grade chucks; however, fluids and biomaterials can easily enter vent holes and damage the internal components of the drill. In addition hardware store quality drill chucks used in this fashion do not stand up to the rigors required for appropriate cleaning and reprocessing.

Postoperative Care

At the end of any operation, good documentation is essential. This includes writing clear orders, an operative note with all significant aspects of the operation including implants used, and some clear postoperative instructions. We have found it helpful to use a premade form for a detailed postoperative plan that includes dates of follow-up, physical therapy instructions, casting, dressing, and suture removal instructions as well as any specific needs such as antibiotics (Fig. 14.7). If you are not likely to be present at the time of follow-up, it is even more important to leave detailed instructions that can be easily understood by local staff. Leaving your contact information with the local surgeon or directly on the operating note should be made a routine. Most importantly, the patient must be able to easily access someone who is knowledgeable of the operation and capable of managing any possible complications. Thankfully the ease and accessibility of international communication allows us to follow up remotely in ways never before possible.

Other Aspects of Orthopedics in the "Wild"

Certain surgeons are attracted to a swashbuckling mindset that leads them to perform difficult operations in the most austere of conditions. Performing operations without the appropriate facilities and equipment should not be seen as a badge of honor, as it is mandatory to adhere to some minimum requirements for safely performing operations. These include running water and reliable electricity and lighting, as well as an intact operating room which provides a well-protected barrier to the outside elements. As previously discussed, safe sterilization, safe anesthesia, adherence to basic principles of sterile technique and stable orthopedic fixation are essential. These are factors that the surgeon should have

control over. Surgical risk and the difficulty of performing operations are increased when doing surgery in the "wild." Some of these factors are controllable and others are not. Factors such as biology, patient compliance, and limitations in available resources and personnel are largely uncontrollable. The surgeon must optimize all controllable factors and take extra precautions to assure patient safety and stable fixation in spite of other uncontrollable risk factors This may mean added points of fixation—extra bone pins on a fixator or more robust plating patterns than would otherwise be necessary. Often times, communication is difficult and follow-up is scant; thus, it is necessary to build constructs not prone to failure and take extra precautions in order to avoid complications. This also involves extra efforts communicating with local staff and patients.

Box 14.3. Improvise—Don't Compromise

- Techniques may need to be modified in situations of limited resources but surgical principles and expected outcomes should never be compromised
- Understand the capabilities and resources of your anesthesia team
- Do not perform operations whose complexity and risk exceeds the capacity of the post operative infrastructure
- Be extra careful about surgical safety while working in environments that may not have implemented the safety measures you are accustomed to
- Opioids must be used judiciously in environments lacking appropriate monitoring, resuscitation equipment and properly trained personnel
- Principles of sterility and sterile technique should never be compromised
- Make sure your equipment is prepared and your operating room properly arranged before embarking on an operation
- Principles of stable fixation should not be compromised and constructs should often be made more robust if limited follow-up is expected
- Economize
- Be flexible

Specific Techniques

Clubfoot

Clubfeet seen in resource limited environments are not usually like the ones typically seen by surgeons in North America. In addition, the success of Ponseti casting is making the operative treatment of clubfoot a lost art. Often times in these environments, severe clubfeet are present in older children, and those who have had prior surgery. For this reason a review of the surgical options for clubfoot is briefly described here.

The Ponseti treatment for clubfoot has revolutionized the treatment of clubfoot around the world. The results of this treatment have been shown to be superior to previous methods of casting and/or surgery [10–12]. The most rapid corrections with Ponseti casting are seen in newborns; however, the technique continues to be effective in older children as well. When performing casting, it is important to follow all of the details of the Ponseti technique to get the very best results. These include: supinating the forefoot to lock the metatarsal cuneiform joints in order to concentrate forces at the subluxed talonavicular joint, placing long leg casts in order to provide sufficient abduction of the forefoot, and performing percutaneous Achilles tenotomy in nearly all cases.

Neglected and recurrent clubfeet are a common cause of disability in developing countries. The stigma of this deformity often prevents those who have it from going to school, integrating into society, and having a job. In older children and adolescents the correction of the deformity has been shown to be worthwhile for social reasons in spite of the fact that often times it does not restore normal mobility and shape to the feet [13].

The excellent results and logistics of effectively implementing the Ponseti method add complexity to surgical decision making when patients in limited resource environments are unable to return for weekly casting, or when short term humanitarian surgical trips do not permit the luxury of providing treatment over time. Ultimately, the best outcome for the child is what should be sought. The surgeon must remember that no child has ever died just because they had a clubfoot, and that a poor surgical result is worse than a deformity not previously operated. Thus immediate operative treatment may not be in the best interest of all patients. Every possible effort should be made to cast clubfeet even if an operation will ultimately be necessary. In situations where casting is logistically difficult, an accelerated program has been described where casting is done every 5 days and has been shown to give similar results to the typical 7-day protocol [14]. Even faster protocols have been tried but are not recommended due to the amount of swelling that can occur and diminishing returns in speed of correction.

In cases where operations are required, casting can be a helpful preoperative adjunct to surgery. Sometimes even just a few days of casting can improve operative results, decrease wound complications, and facilitate the ease of an operation. A hybrid approach can be taken where a minimal release (including posterior tibialis tenotomy, plantar fasciotomy, and Achilles lengthening) is combined with preoperative and postoperative casting for cases where extensive casting is difficult or not practical. There is much debate about the

Table 14.2 Surgical options for neglected and recurrent clubfoot[a]

Posteromedial release	Soft-tissue release for cavus, adductus, varus, and equinus	• Cincinnati • 2 incision • Turco
Additional procedures	Treatment of residual adductus	• Calcaneal closing wedge • Lichtblau calcaneal resection • Calcaneo-cuboid wedge resection/arthrodesis
	Treatment of residual equinus	• Excision head of talus • Naviculectomy • Distal tibia osteotomy
	Treatment of residual heel varus	• Calcaneal osteotomy (lateral slide)
	Treatment of residual supination	• Tibialis anterior transfer • Medial cuneiform osteotomy • Trans cuneiform osteotomy
	Wound closure difficulty	• Leave wound open • Immobilize under-corrected then serial casting postoperatively • Dorsal foot rotational flap • Distal tibia shortening osteotomy
Salvage procedures (for severe and recurrent clubfoot)	Older child with stiff/severe deformity	• Triple arthrodesis—Lambrinudi modification • Posterior release with midfoot dorsal closing wedge
	Arthrogryposis and myelomeningocele	• Talectomy
External fixation	Ilizarov or Taylor Spatial Frame for recurrent/residual deformities and severe clubfoot	• Soft-tissue distraction • Distraction osteogenesis (osteoplasty)

[a]Adapted with permission from Penny NJ. The neglected clubfoot. Tech Orthop 2005; 20(2):153–166

maximum age limit for effective casting. In reality there is no specific age at which casting becomes ineffective; although the time and materials required to correct the clubfoot deformity can significantly increase with age and stiffness of the foot. This is due to progressive ossification of the foot bones. As the child becomes older there is less cartilage and less remodeling potential in the foot.

Posteromedial release has traditionally been a standard treatment for clubfoot. Due to the successful implementation of the Ponseti method in North America this operation is becoming less common. However, in severe, recurrent, or neglected clubfoot, especially in older children, it is still a viable option. For children with significant adductus, posteromedial release can be combined with a lateral column shortening osteotomy (a Lichtblau anterior calcaneal resection, calcaneal cuboid wedge resection, or calcaneal shortening osteotomy). Other options for residual deformities including residual equinus, heel varus, or forefoot supination are outlined in Table 14.2. In older children with severe deformities, triple arthrodesis is often necessary. This can be done in children as young as 8 years old, especially in bilateral cases where a foot size discrepancy would not be created. Another option that has been performed with success for severe cases is a posterior release with a dorsal closing wedge midfoot osteotomy. The temptation to correct recurrences and residual deformity, particularly in the 5- to 12-year-old age group, needs to be weighed against expected results, risk, and recuperation time. In many cases it is best to avoid repeated surgery and wait until a definitive operation like triple arthrodesis can be performed at an age closer to skeletal maturity. The reader is referred to the excellent article by Norgrove Penny for a detailed description of the options and techniques for neglected clubfeet [15].

Previously operated and neglected clubfoot can also be managed with the use of external fixation. Two basic methods of foot deformity correction using Ilizarov techniques are: (1) soft tissue stretching (correcting through joints) and (2) bony correction (osteotomies) with distraction osteogenesis. Due to the improved ability to correct rotational deformity with Taylor Spatial Frame (TSF), we have applied this to clubfeet in a fashion that simulates Ponseti casting (correction through soft tissue). Other fixator configurations with or without osteotomies are also effective. Any of these methods require experience and an infrastructure that can support close follow-up and postoperative management. One of the biggest risks with external fixation is the risk of recurrent deformity. The long treatment time, possibility of persistent hyperesthesia, and other complications need to also be considered when entertaining the use of gradual distraction for surgically treating older children.

SIGN Nail

The SIGN nail (Surgical Implant Generation Network, www.signfracturecare.org) is an intramedullary locking nail which can be implanted without fluoroscopy. It is made by a nonprofit organization for use in limited resource environments



Fig. 14.8 Hip spica application. After appropriate anesthesia, the patient's torso is placed supine on the end of the armboard. Three assistants are used to stabilize and position the patient's arms/head region and each of the lower extremities. Standard casting materials, including a temporary towel to pad the patient's back, are used to complete the hip spica cast. Reprinted with permission from Pasque C, Harbach G. Hip spica application using an operating table armboard. J Pediatr Orthop 2000; 20(6)

and made available to surgeons and institutions at minimal cost. This has revolutionized long bone fracture care around the world with more than 120,000 cases performed. SIGN has established programs in more than 55 countries where surgeons have had the necessary training and experience. Accountability, quality control and maintenance of a sustainable inventory are realized through the use of a database that requires uploading of preoperative and postoperative images as well as input of case data and implant use. There is a learning curve to using the system, which has some unique features enabling distal interlocking without X-ray guidance. This system is useful in limb deformity correction when stabilizing osteotomies or even performing lengthening over nails. Fixator assisted nailing is also a useful technique for a variety of long bone deformities. There is a pediatric version of the SIGN nail available; however, the standard nail is suitable for a trochanteric entry and may be used in older children. The pediatric nail is more useful for fractures and has a unique distal interference fit without a distal interlocking option. The standard adult nail comes in sizes as small as 8×200 mm, making it versatile for deformity corrections in relatively small patients.

Spica Cast

Many pelvic and femoral osteotomy surgeries involve the use of a postoperative spica cast. It is mentioned here to highlight the fact that a specialized spica table is not necessary nor may it be available in many resource challenged environments. Spica casts can be applied on a simple arm board (Fig. 14.8) [16]. A plank of wood or other material ideally about ½″ thick and about 4″ wide can be slid under the pad on the operating table and be used for both an arm board and spica table. In order to do this, the board is placed at 90° to the table under a thin mattress and sticks out to the side like a diving board. One person stabilizes the proximal end of the board onto the table as well as the torso of the child who is placed over the side of the bed on the opposite end of the board. Bilateral and unilateral spica casts can be placed on any size child with the appropriate size arm board.

Educational Resources

Having a collection of orthopedic textbooks available on a laptop or iPad is very helpful when performing orthopedics in resource-challenged environments. Many times demands are made to perform difficult operations that one may not commonly perform. The surgeon must first assess his/her own abilities and again remember that a poorly performed surgery is best not performed at all. But a review of operative details and often a consult from a colleague can remind one of the details needed to efficiently perform a quality operation and prepare the proper equipment for a variety of operations that you may not routinely perform. Fortunately, most textbooks are now available in electronic formats which can allow one to maintain an entire orthopedic library on a small electronic device. The Global Help Website www.global-help.org has a wide variety of textbooks and monographs available for free. They have an excellent monograph on the Ponseti treatment of clubfoot which has been translated into more than 20 languages and serves as an educational tool both for surgeons and parents. Other patient specific educational materials are available on this site as well. An excellent resource is their "Pediatric Orthopedic Library," which on a single disc contains the *Atlas of Pediatric Orthopedics Surgery* (3rd Edition) (Morrissy and Weinstein); *Bibliography of Orthopedic Problems in Developing Countries* (Spiegel); *Lovell & Winter's Pediatric Orthopedics* (5th Edition) (Morrissy and Weinstein); *Neuromuscular Disorders; Practice of Pediatric Orthopedics*

(Staheli); *Sequelae of Septic Arthritis of the Hip* (Spiegel, Penny, and Shrestha); *Standards in Pediatric Orthopedics* (Hensinger); *The Easter Seal Guide to Children's Orthopedics* (The Easter Seal Society); *What Parents Should Know About Flatfeet* (Staheli, Staheli, and Mosca). The DVD cost is $1.00 USD. This allows a traveling surgeon to carry a huge library and moreover offer an invaluable gift to the local surgeons for an extremely low cost.

HINARI (Health Inter Network Access to Research Initiative) is a World Health Organization program which provides free or very low-cost online access to up to 35,000 information sources/journals in biomedical and related social sciences to *local, nonprofit institutions* in developing countries. This functions as an institutional subscription to journals referenced by PubMed etc. An application process is required, and in the most resource challenged countries, subscriptions are completely free of charge, whereas in other developing countries a small fee is assessed. More information and application materials can be obtained at www.who.int/hinari/en/.

Summary

Working with limited resources and often times in unfamiliar locations can be some of the most challenging but also most rewarding experiences in life. Many of these operations are life changing for the patients we treat. Basic principles should never be compromised; every effort should be made to obtain outcomes equal to those where ample resources exist. This often requires more robust fixation, extra precautions to avoid complications, and clear communication. As orthopedic surgeons with education and resources, we should consider it our obligation to help those who are in need regardless of their ability to compensate us. This can be in our own country or another. By going beyond stagnate and repetitive routines, new perspectives are gained, new relationships are built and lives are changed.

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Part III

Underlying Conditions

Metabolic Disorders

15

Mehmet Kocaoglu, I. Levent Eralp, and F. Erkal Bilen

General

Deficiency in the shape, strength and structure of bone tissue due to altered bone mineral homeostasis is called *metabolic bone disease* (1). The major factors affecting this homeostasis can be thought of as a three 3's: the intracellular and extracellular levels of *three* ions (calcium, phosphorus, and magnesium), which are controlled by *three* hormones (parathyroid hormone, calcitonin, and 1, 25-dihydroxyvitamin D) and act upon *three* tissues (bone, gut, and kidney) [1, 2]. Common clinical manifestations of metabolic bone disease in children include electrolyte disturbances, fractures, bone deformity, abnormal gait, and short stature.

The most commonly encountered forms of metabolic bone disease in children are the various types of rickets and renal osteodystrophy. Other less common but important pediatric metabolic conditions include osteoporosis, malabsorption syndromes, inherited diseases such as hypophosphatasia, X-linked hypophosphatemia, and various forms of vitamin D-dependent rickets [1, 3] (Box 15.1).

Box 15.1

- Metabolic problems related to vitamin D must be addressed prior, during, and after the surgical treatment.
- There are various forms of metabolic bone diseases. Thus, collaboration with the pediatric/internal medicine department is essential.

Pathophysiology

Although there are several types of *rickets*, the basic pathogenesis is a relative decline in calcium or phosphorus (or both) of large enough amount so that it interferes with physeal growth and mineralization of the bone matrix in the growing child [1, 3, 4]. This decrease in the mentioned serum ion levels may result from inadequate intake or diminished absorption of phosphorus, calcium or vitamin D, reduced conversion of vitamin D to its active form, end-organ insensitivity, impaired release of calcium from bone, or phosphate wasting. In addition, there is evidence that insufficient vitamin D may interfere with mineralization independent of calcium or phosphate levels [1, 3].

In renal osteodystrophy, glomerular damage leads to phosphate retention, and tubular damage causes decreased production of the active form of vitamin D (i.e., 1,25-dihydroxyvitamin D) due to absence of 1-hydroxylase activity. These two factors severely impede intestinal calcium absorption and reduce plasma ionized calcium. The subsequent hypocalcemia generates secondary hyperparathyroidism, which remains ineffective in increasing intestinal absorption of calcium. Consequently, the body's only means of increasing serum calcium levels is by bone resorption. Metabolic acidemia may further deteriorate this condition (Figs. 15.1 and 15.2) [1, 3, 4].

In patients with *osteoporosis*, the bone is structurally normal but is reduced in overall amount. In children, osteoporosis may be idiopathic, as in juvenile osteoporosis, or may be due to disuse or chronic corticosteroid administration. The mechanism is uncertain, but numerous theories include increased bone resorption versus decreased bone formation, possibly due to deficient 1,25-dihydroxyvitamin D or calcitonin

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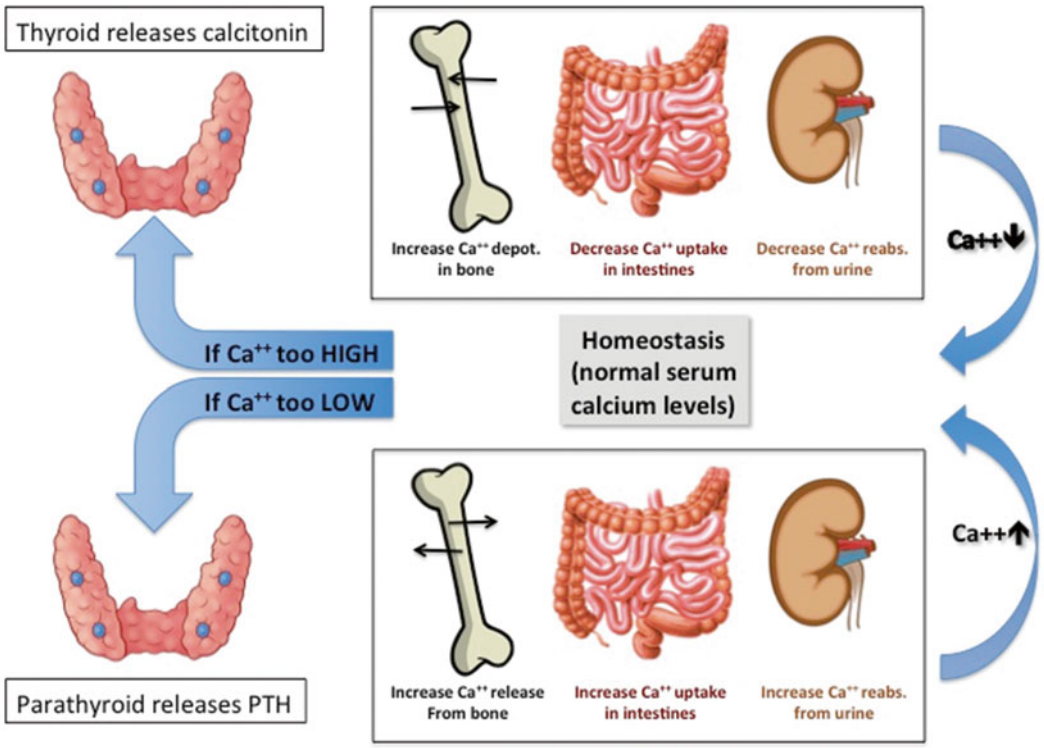
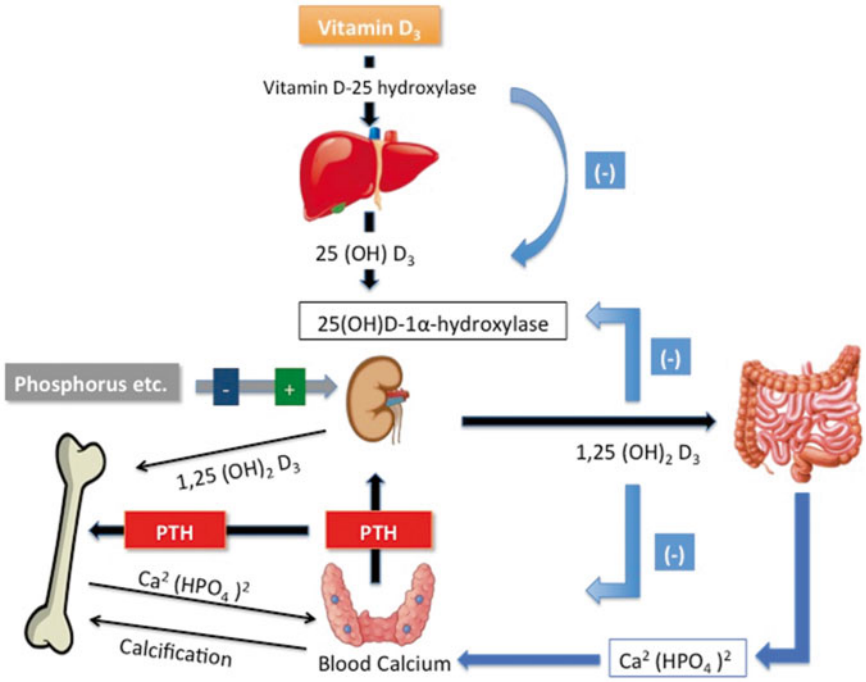


Fig. 15.1 Metabolic control of calcium levels

Fig. 15.2 Vitamin D metabolism



or due to a major interruption in transduction of mechanical forces that stimulate new bone formation [1, 3].

Hypophosphatasia results from a genetic error in the synthesis of alkaline phosphatase, the enzyme necessary for the maturation of the primary spongiosa in the physis. This condition leads to normal production of osteoid tissue but inadequate mineralization, with resultant skeletal deformities that resemble rickets [1, 3, 4].

Nutritional Rickets

General

The primary etiology of this condition is vitamin D deficiency in the diet. This deficiency leads to failure of calcification of cartilage and osteoid tissue. Vitamin D deficiency results in an inability to absorb calcium and phosphorus. Parathormone (PTH) is released in response to hypocalcemia. Subsequently, serum calcium levels become normal or slightly decreased, but phosphate and vitamin D levels remain low (Table 15.1). Thus, radiologically, the physes display an elongation and a hazy appearance related to alterations in the provisional zone of calcification [4, 5]. The widened growth plate differentiates rickets from more common physiologic angular deformities of the lower extremity [6]. With treatment, calcification occurs and radiographic appearance gradually normalizes.

Treatment

The routine treatment for nutritional rickets include the supplemental administration of vitamin D. Radiographs typically show improved mineralization within 2–4 weeks of initiating medical treatment [4, 6]. If the child does not respond to vitamin D therapy, vitamin D-resistant rickets should be suspected. Because residual deformity is very rarely observed after adequate medical treatment of nutritional rickets, there is no specific orthopedic treatment besides follow-up to ensure acceptable lower limb alignment.

Rickets of Prematurity

Premature infants with comorbidities, followed up in intensive care units, sometimes present with pathologic fractures, probably caused by passive motion exercises. With treatment of the underlying rickets, the fractures consolidate with orthopedic immobilization techniques [4].

Drug-Induced Rickets

Certain antiepileptic medications, such as valproic acid and phenytoin, have been known to create rachitic changes in children [7] (Box 15.2). These drugs lower vitamin D levels through a P-450 microsomal enzyme system mechanism in the liver. Patients mostly present with pathologic fractures while on treatment for seizures. Medical therapy with vitamin D supplementation and consultation with the treating neurologist is helpful.

Box 15.2

- Metabolic bone diseases can be caused by certain medications as well.
- Remember that certain antiepileptics such as valproic acid and phenytoin may lower vitamin D levels in the body.

Vitamin D-Resistant Rickets (Familial Hypophosphatemic Rickets)

General

Vitamin D-resistant rickets involves a group of disorders in which normal dietary ingestion of vitamin D is insufficient to accomplish normal mineralization of bone [2, 6]. There are four major forms of vitamin D-resistant rickets, the most common of which is inherited as an X-linked dominant trait, followed in occurrence by an autosomal dominant type [2, 4, 6]. The inherent pathology is the renal tubule's incapability to retain phosphate, which causes hypophosphatemia. The third group is characterized by failure of the kidney to accomplish the second hydroxylation of vitamin D. This condition can simply be treated medically, therefore orthopedic treatment is infrequently needed. In the fourth group, also known as renal tubular acidosis, the kidney excretes fixed base and wastes bicarbonate resulting in wasting of calcium and sodium [3, 6]. Laboratory findings are listed in Table 15.1.

Vitamin D-resistant rickets typically becomes evident between the ages 1 and 2 years, slightly older than nutritional rickets. The major complaints are delayed walking and angular deformities of the lower extremities (Figs. 15.3 and 15.4). Systemic manifestations are generally absent. The deformities are much more severe when compared to nutritional rickets. Once affected children begin to walk, genu varum develops, although genu valgum may occur in some children [4, 6]. Short stature is also a feature of hypophosphatemic rickets, with their standing height often being 2 standard deviations below the mean for their peers [8]. Radiologically, the physes are widened, there is genu varum and coxa valga, Furthermore, a varus deformity of the distal tibia often leads to varus malalignment of the ankle joint (Fig. 15.5). The upper extremities are involved, to a lesser degree, as well [4].

Treatment

Medical Treatment

The standard treatment consists of oral supplementation of large doses of phosphorus along with administration of vitamin D. Studies have revealed that longitudinal growth is superior in children who receive vitamin D treatment [4].

Table 15.1 Biochemical abnormalities in rickets

Biochemical abnormality						
Type of metabolic bone disease	Calcium	Phosphate	ALP	PTH	25 (OH) Vit D	1,25 (OH)2 Vit D
Nutritional	N	N/↓	↑	↑	↓↓	↓
Vitamin D resistant	N	↓	↑	N	N	N
Vitamin D dependent I (inability to hydroxylate)	↓	↓	↑	↑	↑↑	↓↓
Vitamin D dependent II (receptor insensitivity)	↓	↓	↑	↑	N/↑↑	↑↑↑↑
Renal osteodystrophy	N/↓	↑	↑	↑↑	N	↓↓



Fig. 15.3 Photograph of a patient with severe genu valgum deformity



Fig. 15.4 Photograph of a patient with severe genu varum deformity

Furthermore, treatment by growth hormone administration also increases height and has positive influence on bone density [9]. Recently, analogs of vitamin D3 (1,25-dihydroxyvitamin D3) have been proved to be more effective than the previously used supplements [10].

Orthopedic Treatment

The orthotic management of skeletal deformities in patients with vitamin D-resistant rickets has not been successful. If patients complain of increasing pain and difficulty walking, angular deformities should be surgically corrected [4, 11]. The postoperative management should be done in close collaboration with the attending nephrologist or endocrinologist, since calcium levels tend to suddenly increase with postoperative immobilization period. The most common deformity seen in this patient group is a gradual anterolateral bowing of the femur accompanied by tibia vara (Figs. 15.6 and 15.7). In order to reach a physiologic lower extremity alignment, multilevel osteotomies are often necessary [4, 11, 12]. The mechanical axis can be slightly overcorrected during surgery. The suggested fixation modality varies among reports. While external fixation allows fine-tuning of the alignment postoperatively [13] (Fig. 15.8), intramedullary fixation and plate fixation have also been reported [12, 14–16] (Fig. 15.9; Table 15.2). Regardless of type of implants utilized, careful preoperative planning of the surgery for these multiplanar deformities is obligatory to successfully reestablish the alignment of the lower extremity.

Recurrent deformity is a common sequela of osteotomies in patients with hypophosphatemic rickets [4, 12, 14] (Fig. 15.10). As expected, younger patients have a higher risk of recurrence. Therefore, milder deformities should not be operated early in the infancy. At this stage guided growth by hemiepiphyseodesis is an advisable alternative [17]. Only when gait is compromised by a thrust, or symptoms such as pain start, a corrective osteotomy should be done.



Fig. 15.5 X-ray showing distal tibial varus deformity



Fig. 15.7 Photograph of a patient with severe anterior bowing of the femur



Fig. 15.6 X-ray of a patient with a anterior bowing of the femur

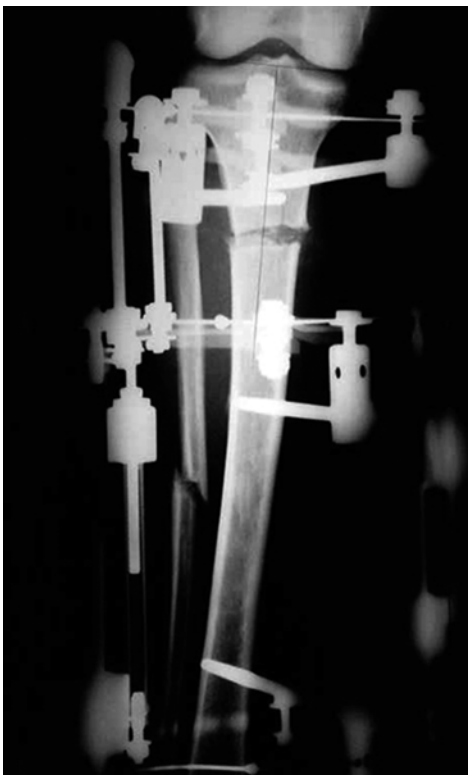


Fig. 15.8 X-ray of external fixation for the tibial deformity



Fig. 15.9 Intramedullary fixation of a tibial deformity correction

Short stature is also noted amongst children with hypophosphatemic rickets. The common indication for long bone lengthening is a shortening of the entire or segment of limb in one or both legs [11]. The procedure can be performed by monolateral or circular external fixators or by lengthening over an intramedullary nail (LON) (Fig. 15.11) [12]. The application of flexible intramedullary nails in limb lengthening for children is also an alternative with multiple advantages [18]. This technique also respects the bone biology that is essential during the limb lengthening.

Author's Preferred Method, Tips and Tricks

The pathological change caused by hypophosphatemic rickets occurs very close to the growth plate, often leading to juxta-articular multiplanar deformities and severe malalignment. Preoperative assessment usually reveals multiple centers of rotation of angulation (CORA) (Fig. 15.12); thus several osteotomies may be required to fully correct the multiapical deformities. Many methods of treatment have been described. The most common involve acute or gradual correction, using either circular or monolateral external fixators, which can secure accurate correction of the deformity and address the limb-length discrepancy. However, these methods are uncomfortable for the patient, especially when both legs are involved. There are further disadvantages, such as the need for daily adjustments, weekly follow-ups, a high rate of pin-track infection, and a long duration of external fixation.

We use the fixator-assisted nailing technique described by Paley and Herzenberg [19], with osteotomies created at the center(s) of the deformity(ies), followed by correction using a monolateral fixator and stabilization with locked intramedullary nailing (Figs. 15.13, 15.14, and 15.15). Removal of the external fixator at the end of the operation reduces postoperative discomfort and avoids pin-track infections, except where patients require additional postoperative lengthening over the nails. However, this technique does not allow residual correction or adjustment postoperatively. Thus, careful analysis of the deformity and preoperative planning are crucial. We recommend using this technique for the patient, who has essentially reached skeletal maturity. Flexible intramedullary nails (elastic nails) can be used in the pediatric age group [18].

Osteotomies in the long bones can be executed through limited incisions percutaneously either by the Gigli saw technique (Fig. 15.16) or by the multiple drill hole technique (Fig. 15.17). This technique combines the accuracy, minimal invasiveness, and safety of external fixation with the patient convenience of internal fixation. The intramedullary nail prevents the recurrence of the deformity, which is especially important in patients with metabolic bone diseases who are prone to recurrence of the deformity as the metabolic problem continues. In addition, the surgeon must be familiar with both external fixation and intramedullary nailing techniques, as well as combined techniques, which can be technically demanding.

Surgical Technique

The patient is placed supine on a radiolucent table and the lower limb is checked for imaging on the radiography table from the hip joint to the ankle joint on both frontal and side views (Fig. 15.18). In patients with genu valgum, resulting both from the femur and/or the tibia, mini-open release of the peroneal nerve is performed prior to correction of the deformity to avoid neurapraxia due to traction. Two 6-mm conical Schanz screws are placed into the proximal and distal segments above and below the osteotomy to maintain the stability, perpendicular to the anatomical axis, taking care to leave enough space for the inserted intramedullary nail without making contact with the screws (Figs. 15.19, 15.20, and 15.21). The level of osteotomy is chosen based on preoperative planning and performed percutaneously using the multiple drill hole technique for the femur or a Gigli saw for the tibia. Following the osteotomies, the deformity is corrected using a monolateral external fixator (Fig. 15.22). The correction is confirmed by obtaining frontal and side view long radiographs. If the desired correction is not achieved, it is adjusted and confirmed with new X-rays. Once the surgeon is satisfied with the correction, the intramedullary nailing is performed through a 2 cm transverse incision over the ligamentum patellae (Fig. 15.23).

Table 15.2 Review of literature

Authors	Journal (year)	Title	Number of patients	Results
Fucentese et al.	J Child Orthop (2008)	Metabolic and orthopedic management of X-linked vitamin D-resistant hypophosphatemic rickets	12	Single bilateral surgical correction was performed in six patients; one patient each had three and five corrections. Bone lengthening was performed in three patients. At last follow-up, the height of seven operated patients was within normal range. In addition, leg axis was normalized in six patients with mild genua vara in two. Bone healing was excellent: surgical complications were rare. In case of bone deformity, surgery can safely be performed, independent of age or bone maturation
Kocaoglu et al.	J Bone Joint Surg Br (2011)	Combined technique for the correction of lower-limb deformities resulting from metabolic bone disease	17 (43 segments)	43 segments in 17 patients with metabolic bone disease underwent surgical treatment by the fixator assisted nailing technique. The deformity correction was achieved with a low complication rate. The use of intramedullary nail prevented recurrence of deformity and refracture
Kanel et al.	J Pediatr Orthop (1995)	Unilateral external fixation for corrective osteotomies in patients with hypophosphatemic rickets	(29 segments)	Corrective osteotomies were performed on 29 bones in nine children with hypophosphatemic rickets. Use of the Orthofix external fixator enabled precise correction of the deformities without interruption of medical management
Rubinovitch et al.	Clin Orthop Relat Res (1988)	Principles and results of corrective lower limb osteotomies for patients with vitamin D-resistant hypophosphatemic rickets	10 (44 osteotomies)	Osteotomies were combined with shortening and compression plating. Recurrence of deformity occurred in 27 % of the cases. While osteotomies were safe and provided dramatic improvement to limb deformity, postoperative control of vitamin D metabolism was the one constant factor for maintenance of correction
Petje et al.	Clin Orthop Relat Res (2008)	Deformity correction in children with hereditary hypophosphatemic rickets	10 (53 segments)	37 corrective operations were performed on ten children. Depending on the patient's age, external fixation was used in 53 segments: Kirschner wires in 18, DynaFix in three, the Taylor patial Frame device in 13, and the Ilizarov device in 19. Internal fixation with intramedullary nailing was performed in 12. Deviation of the mechanical axis and knee orientation lines was increased at the follow-ups conducted during a period of 5–12 months. Additional follow-ups revealed a recurrence rate of 90 % after the first corrective procedure and 60 % after a second procedure
Song et al.	Acta Orthopedica (2006)	Deformity correction by external fixation and/or intramedullary nailing in hypophosphatemic rickets	20 (55 segments)	55 segmental deformities (20 femora, 35 tibiae) from 20 patients were examined retrospectively. Distraction osteogenesis was used in 28 segments and acute deformity correction in 27. External fixation was applied in 24 segments, intramedullary nailing in six, and external fixation and intramedullary nailing in 25. Recurrent deformity or refracture occurred in 10 of 21 segments with distraction osteogenesis by external fixation only, 4 of 6 with acute correction by intramedullary nailing, and 1 of 25 with distraction osteogenesis or acute correction by external fixation and intramedullary nailing. External fixation and intramedullary nailing can be recommended to prevent complications during or after deformity correction in hypophosphatemic rickets
Eralp et al.	J Bone Joint Br (2004)	A correction of windswept deformity by fixator assisted nailing	2 (7 segments)	Seven segments in two patients with vitamin D related metabolic bone disease were treated by fixator-assisted nailing technique. All deformities were accurately corrected and there was no consolidation problem at the osteotomy sites. The total treatment time was found less than with other techniques
Stanitski	Clin Orthop Relat Res (1994)	Treatment of deformity secondary to metabolic bone disease with the Ilizarov technique	8 (18 segments)	Complications were limited to several pin-tract infections and mild translational deformity in two patients. Healing index averaged approximately twice that seen in pediatric femoral lengthening and was 25 % greater than for patients undergoing tibial lengthening. The lack of implants requiring removal, modularity, and reasonable treatment time make this technique an attractive alternative to conventional osteotomy for management of limb-length deformity associated with metabolic bone disease



Fig. 15.10 Note the recurrence of the deformities in a patient despite the intramedullary nails

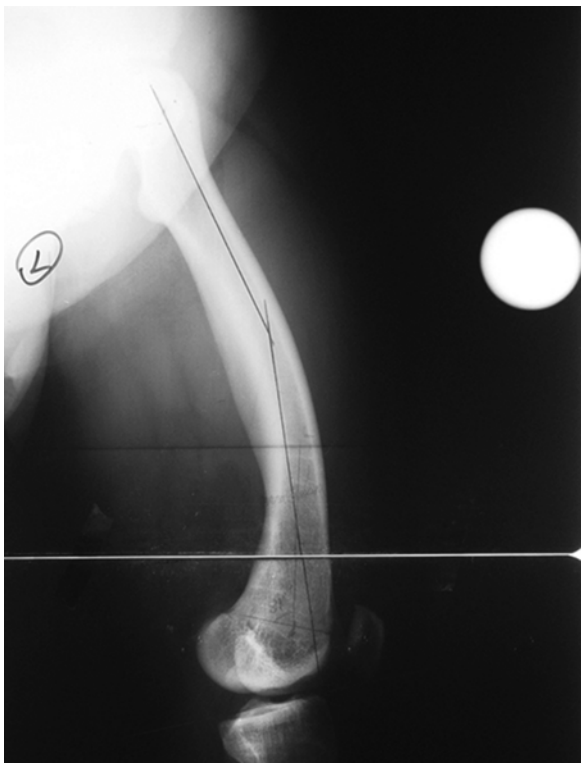


Fig. 15.12 Long sagittal bowing deformity at the femur: note there are multiple centers of rotation in these type of deformities

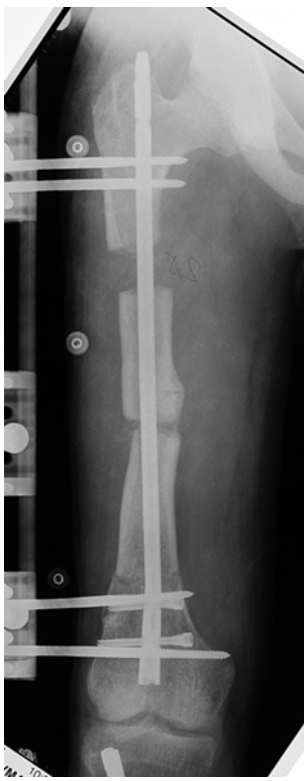


Fig. 15.11 Following deformity correction (fixator assisted nailing) lengthening over nail was performed at the femur



Fig. 15.13 X-ray showing a varus bowing deformity at the femur

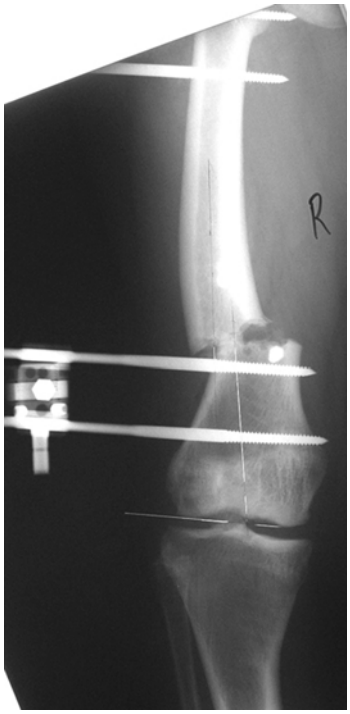


Fig. 15.14 The deformity has been acutely corrected with use of an unilateral external fixator



Fig. 15.15 Once the satisfactory correction was achieved, intramedullary static fixation was performed (FAN procedure)

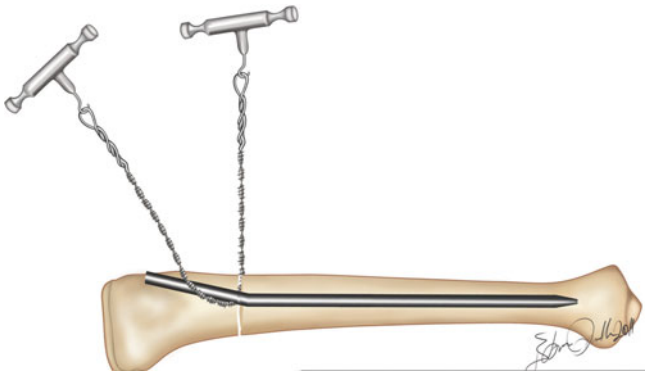


Fig. 15.16 Gigli saw osteotomy technique

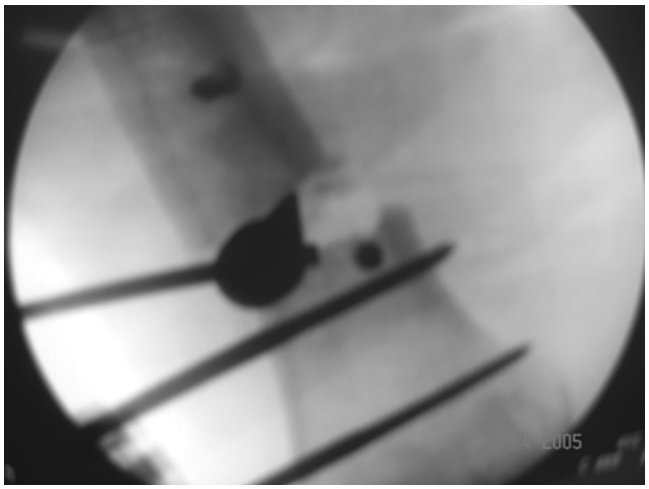


Fig. 15.17 Drill hole technique which is completed by an osteotome

The standard ligament split approach is used to open the tibial or femoral canals under fluoroscopic control. Over a guide wire, the medullary canal is over-reamed 1 mm larger than the diameter of the nail to be used (Fig. 15.24). Interference blocking screws are placed before or after nail insertion to reduce the larger diameter of the medullary canal at the metaphyseal level to prevent motion of the nail (Fig. 15.25). The nail is inserted and locked both proximally and distally if no lengthening is planned. The positions of the nail and the Schanz screws and the deformity correction are once again confirmed under fluoroscopic control, and then the fixator is removed (Figs. 15.26 and 15.27) (Box 15.3).

Follow-Up

Isometric exercises are begun postoperatively and partial weight-bearing is allowed with crutches as tolerated. Full weight-bearing is allowed only after achieving the consolidation of three of four cortices on AP and lateral radiography (approximately 3 months).

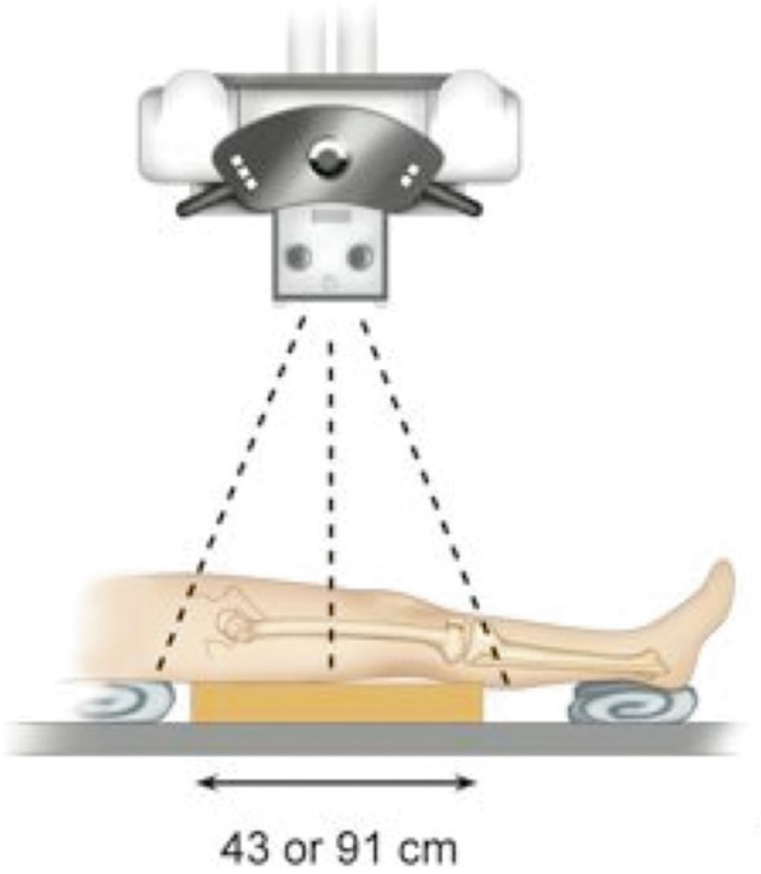


Fig. 15.18 Long X-rays for deformity analysis provided by a radiolucent table



Fig. 15.19 Two parallel Schanz screws distally at the femur



Fig. 15.20 Two parallel Schanz screws proximally at the femur

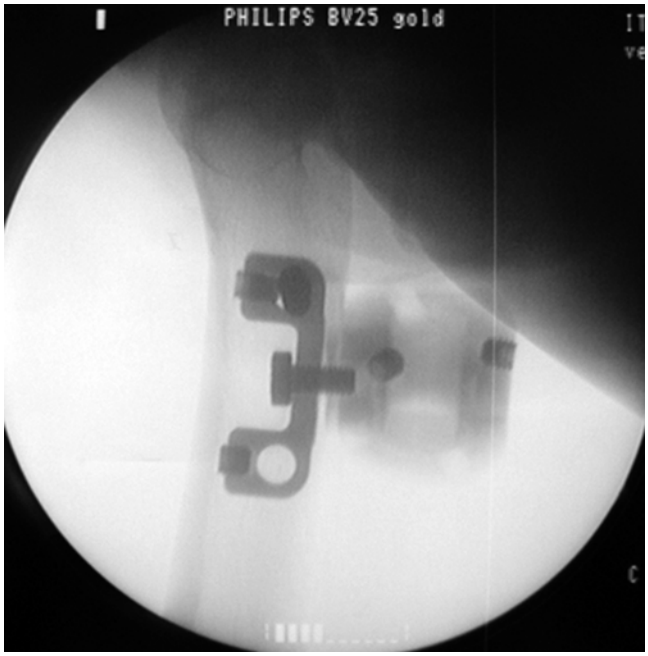


Fig. 15.21 Schanz screws must leave enough space for intramedullary nailing



Fig. 15.23 Transverse skin incision over the ligamentum patella



Fig. 15.22 Following osteotomy the correction is performed and maintained by a monolateral external fixator

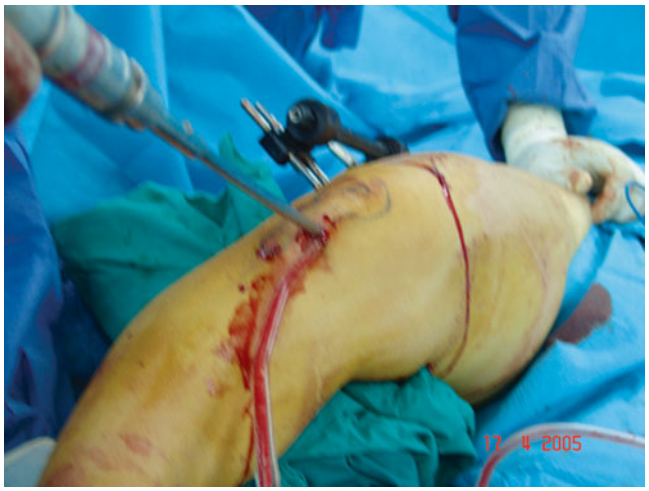


Fig. 15.24 The medulla is overreamed by 1 mm

Tumor-Related Hypophosphatemic Rickets (Oncogenic Hypophosphatemic Osteomalacia)

Certain tumors have been found to secrete phosphatonins that result in phosphaturia. Neurofibromatosis, fibrous dysplasia, less commonly osteoblastoma and hemangiopericytoma tend to produce rachitic changes in bone. The condition generally resolves with excision of the tumor [20].

Renal Osteodystrophy

General

As the use of renal transplantation for treating renal failure in children has increased, the prevalence of renal osteodystrophy has also climbed. Manifestations of renal osteodystrophy are present in 66–79 % of children with renal failure [21].



Fig. 15.25 Interference screws are used to help maintain the correction

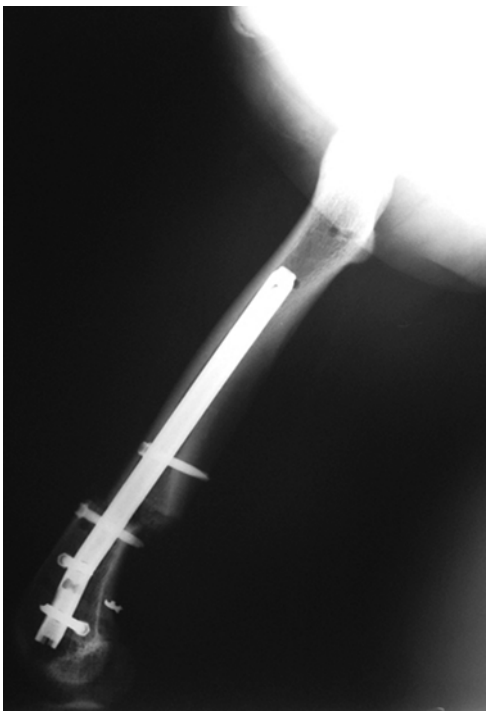


Fig. 15.27 Lateral view following intramedullary static fixation

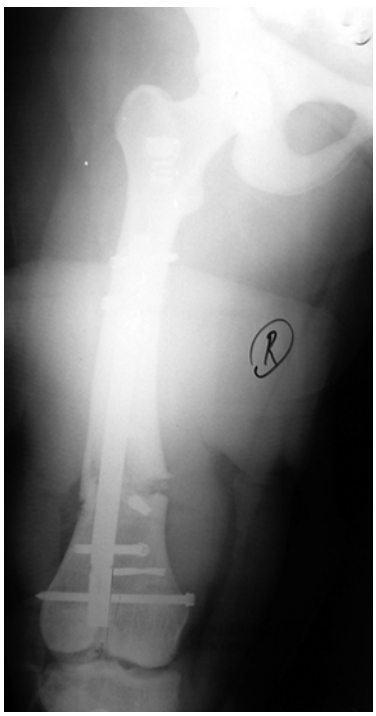


Fig. 15.26 AP view following intramedullary static fixation

Box 15.3

- The deformities are multiapical (long bowing) and tend to recur.
- Intramedullary nailing should be performed always if possible to prevent recurrence.
- Preoperative planning is of paramount importance, and should contain paper-tracing to simulate the surgical procedure. This will increase the precision of the procedure while shortening the surgical time.
- Fixator-assisted nailing is very effective for accurate deformity correction and for prevention of recurrences.

Renal osteodystrophy is noticeably different from either nutritional or hypophosphatemic rickets. It is often driven by the presence of secondary hyperparathyroidism, which leads to activation of osteoclasts and resorption of bone [2, 4].

Features of both rickets and hyperparathyroidism are present in children with renal osteodystrophy. Rachitic changes include lack of calcification in the zone of provisional

calcification of the physis while secondary hyperparathyroidism induces osteoclastic resorption of bone.

Serum vitamin D and calcium levels are usually low, accompanied by increased levels of blood urea nitrogen and creatinine in the blood along with acidosis (see Table 15.1).

Children affected by renal osteodystrophy are often short-statured and have fragile bones. These patients often have bone pain and fractures occur easily. The most common orthopedic manifestations are skeletal deformities, usually genu valgum, periarticular enlargement of long bones, slipped capital femoral epiphysis (SCFE), muscle weakness, and Trendelenburg gait especially if SCFE is present [4]. On radiographs, widespread osteopenia with thin cortices and unclear trabeculae are present. The underlying bone has a ground glass appearance. The physes are increased in thickness, with an indistinct zone of calcification. In severe and persistent renal failure, aggressive lytic areas in long bones may develop (*brown tumor*). Since many patients with renal failure, especially those who undergo renal transplant, are treated with steroids, osteonecrosis can also develop.

Treatment

Medical Treatment

Treatment of the causal renal disease is of crucial importance. Dialysis and renal transplantation are prolonging the survival of these patients. Medical therapy is initiated with the use of 1,25 dihydroxy form of vitamin D. The use of calcitriol significantly decreases serum PTH levels and delays secondary bone changes [22]. The treatment of acidosis with sodium bicarbonate also improves metabolic bone disease.

Decreased skeletal growth and short stature is a significant problem, probably due to disturbances in the growth hormone—insulin like growth factor axis. Recombinant human growth hormone (rHGH) restores growth in these children. However, rHGH, also weakens the physes and may predispose the child to develop SCFE [23]. In patients with renal osteodystrophy, who are refractory renal osteodystrophy to medical treatment, parathyroidectomy may be indicated.

Orthopedic Treatment

Patients with renal osteodystrophy are generally referred to the orthopedic surgeon for the treatment of three pathologies: angular deformity of lower extremity long bones, SCFE, and avascular necrosis [4]. Any surgical intervention in this patient population should be carefully considered, as the perioperative risks are amplified due to associated anemia, hypertension, bleeding tendencies, and electrolyte imbalances. The risk of infection is additionally increased in patients with a renal transplant who are under immunosuppressive therapy.

Angular Deformity

Angular deformity occurs in renal osteodystrophy because the bone is soft, undermineralized, and prone to bend with weight bearing. Genu valgum is the most common deformity, but genu varum or a windswept deformity may also occur [4, 24]. If the beginning of renal osteodystrophy occurs before 4 years of age, varus deformity may develop, because the normal alignment of the leg is in slight varus, which then is accentuated when the bone becomes weak. Similarly, older children are prone to the development of genu valgum because of the physiologic valgus alignment of the lower extremity. Valgus at the ankle may also accompany the genu valgum [25].

Some milder deformities will correct with medical treatment of the renal osteodystrophy [4, 25]. Deformities do not respond well to bracing. If the patient becomes symptomatic, and has had optimum medical treatment without resolution of the deformity, corrective osteotomy should be accomplished [4, 17, 25]. Usually, the greatest deformity is in the distal femoral metaphysis, but sometimes, a supplementary proximal tibial osteotomy is also needed. Internal or external fixation may be used. While external fixators have been successfully applied, taking care of achieving stable constructs and utilize of hydroxyapatite coated Schanz pins, but bone healing may be delayed [26]. Recurrence of the skeletal deformity is common in patients with continuing metabolic pathology, so medical treatment should be adjusted before and continued after corrective osteotomy. Elevation of serum alkaline phosphatase concentration above 500 U/L is a worthy marker of ongoing metabolic bone disease [4, 25]. Milder deformities may respond to hemiepiphysiodesis [17].

Slipped Capital Femoral Epiphysis

The clinical picture of a child with SCFE secondary to renal osteodystrophy differs from the usual cases of SCFE. Often, these patients are younger, and obesity is not a part of the clinical picture. Bilaterality is also very common. Radiologic pathology in the physis is more pronounced, accompanied by physeal widening and generalized osteopenia [27]. The goal of standard management of SCFE is to prevent further deformity and promote early closure of the proximal femoral physeal growth plate. However, cessation of proximal femoral growth may not be desirable in a young child with renal osteodystrophy. Moreover, physeal healing is compromised due to the underlying metabolic pathologic condition. Fortunately, pain and physeal widening resolves with initiation of appropriate medical treatment. If the slip is displaced or symptoms continue despite of medical treatment, fixation with one screw, accomplishing stability cross the physis without closing it, should be performed.

Avascular Necrosis

Prolonged use of corticosteroids is the likely cause. Avascular necrosis occurs frequently bilateral, affecting the hip. Treatment is usually symptomatic [4].

Hypophosphatasia

General

Hypophosphatasia is a rare, genetic defect of alkaline phosphatase production, resulting in pathologic mineralization of bone. There is a wide variation in the severity of the disease and the prognosis depends on the age of onset; perinatal, infantile, childhood and adult hypophosphatasia can occur [4, 28]. The genetic defect for this disorder is determined to be in the tissue-nonspecific alkaline phosphatase gene (*TNSALP*), and many different mutations have been described [29].

The pathology detected in hypophosphatasia closely resembles that seen in patients with rickets. Osteoid production remains undisturbed, but deprived of effective *ALP*, mineralization of osteoid is compromised [4, 28, 29]. As a results, the physes are widened with persistence of the zone of calcification and metaphyseal remnants of cartilage islands. If hypercalcemia is added to the clinical picture, heterotopic calcification *foci* can be formed. In laboratory tests, *ALP* levels are decreased not only in the serum but also in tissues like kidney, bone, etc. [1, 4]. Serum phosphorus, vitamin D, and PTH levels may remain within normal limits, but hypercalcemia is sometimes present. Characteristically, urine pyrophosphate levels are increased [1, 4].

In the severe perinatal form, the infants may be stillborn. If they survive delivery, they frequently die from respiratory infection in early infancy. All bones display severe demineralization on radiographs. The infantile form starts later, usually around 6 months of age. These babies have demineralized bones with severe rachitic changes. Fractures and bowing of the extremities are frequent. If these children can survive infancy, they tend to improve clinically, but they are short statured throughout childhood due to absence of normal endochondral bone growth [1, 4].

Treatment

Although there is no conventional medical treatment of hypophosphatasia, successful bone marrow cell transplantation with improvement of the disease has been described [30].

Fractures and deformities need orthopedic management. Fracture healing is commonly delayed. Multiple osteotomies with intramedullary fixation are often required to correct the bowing and provide structural support to the long bones [4]. Thus, when possible it is advised to utilize an intramedullary nail in all corrected bone segments.

Medical control of the underlying disease is of paramount importance as the deformities tend to recur in various metabolic bone diseases. Therefore, consultation with the endocrinologist should be done preoperatively and continued after the surgical intervention.

Example Cases

Case 1: Figures 15.28, 15.29, 15.30, 15.31, 15.32, 15.33, and 15.34 show the treatment sequences of a patient (hypophosphatemic rickets) with a profound bilateral femoral varus deformities.

Case 2: Figures 15.35, 15.36, 15.37, 15.38, 15.39, 15.40, 15.41, 15.42, and 15.43 show the treatment sequences of a patient (hypophosphatemic rickets) with a profound genu valgum deformity.

Case 3: Figures 15.44, 15.45, 15.46, 15.47, 15.48, 15.49, 15.50, 15.51, and 15.52 show the treatment sequences of a patient (hypophosphatemic rickets) with a profound genu varum deformity.

Case 4: Figures 15.53, 15.54, 15.55, and 15.56 show the treatment sequences of a patient (X-linked rickets) by guided growth technique.

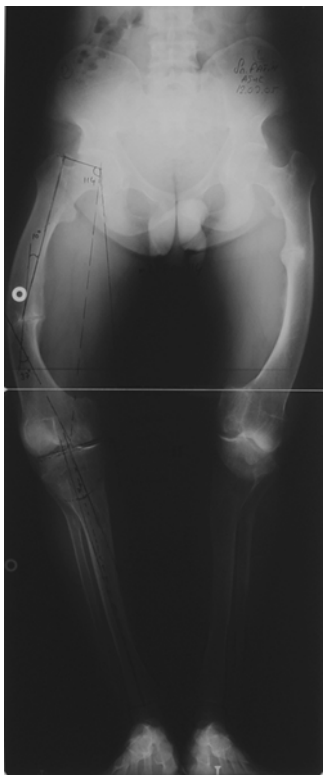


Fig. 15.28 A patients orthoroentgenogram showing severe varus deformities



Fig. 15.29 Right lateral orthoroentgenogram shows anterior femoral bowing deformity. Note there is pending stress fracture at the femoral dyaphyseal level



Fig. 15.30 Left lateral orthoroentgenogram shows anterior femoral bowing deformity

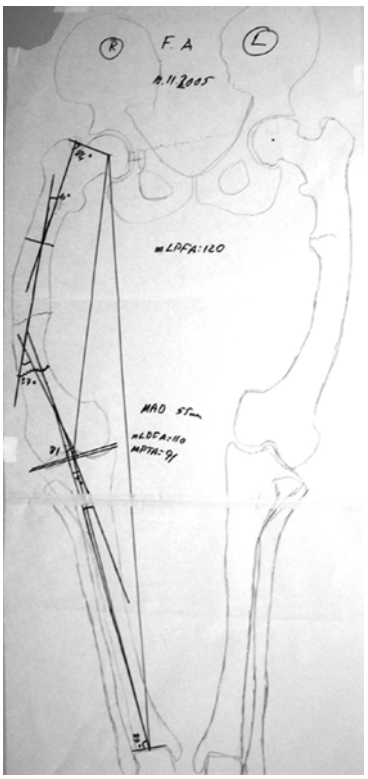


Fig. 15.31 Paper-tracing for deformity analysis

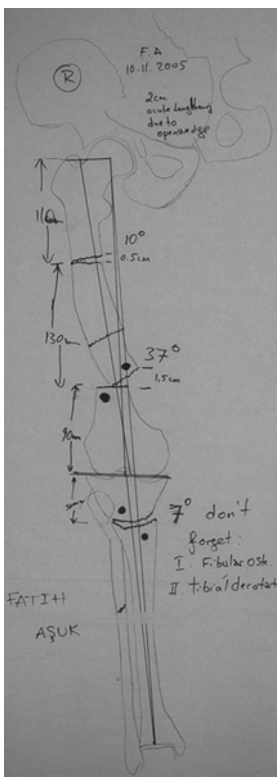


Fig. 15.32 Paper-tracing for surgical simulation



Fig. 15.33 Orthoroentgenogram following FAN procedure at the femur and tibia on the right



Fig. 15.35 Orthoroentgenogram of a patient with severe genu valgum deformity

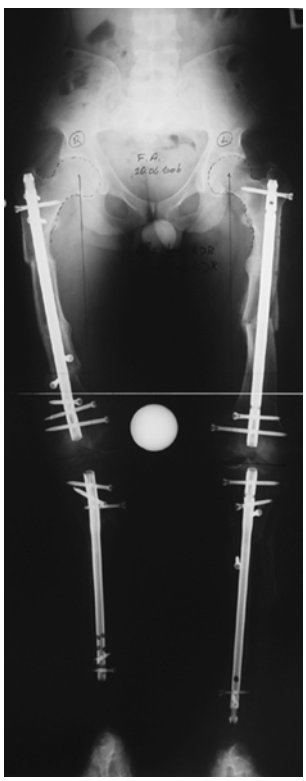


Fig. 15.34 Orthoroentgenogram following final FAN procedure at the femur and tibia on the left



Fig. 15.36 Right lateral orthoroentgenogram of the same patient



Fig. 15.37 Left lateral orthoroentgenogram of the same patient

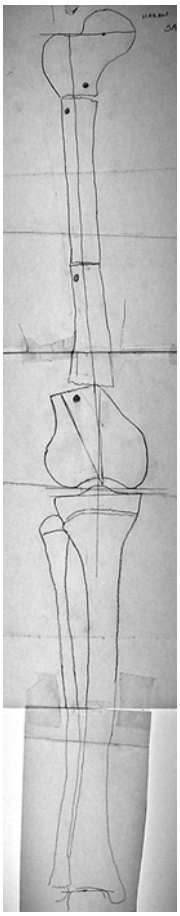


Fig. 15.38 Paper-tracing for surgical planning



Fig. 15.39 X-ray of the right femur following FAN procedure

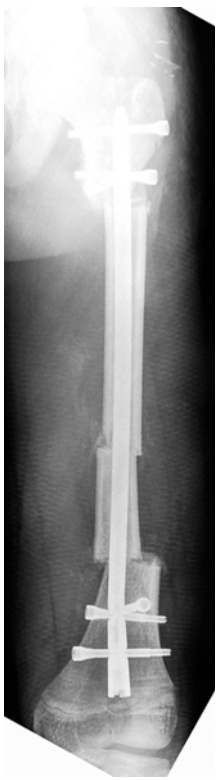


Fig. 15.40 X-ray of the left femur following FAN procedure



Fig. 15.41 Orthoroentgenogram at the end of the treatment



Fig. 15.42 Photographic documentation of the same patient prior to surgery



Fig. 15.43 Photographic documentation of the same patient after the treatment



Fig. 15.44 Right frontal orthoroentgenogram showing severe genu varum

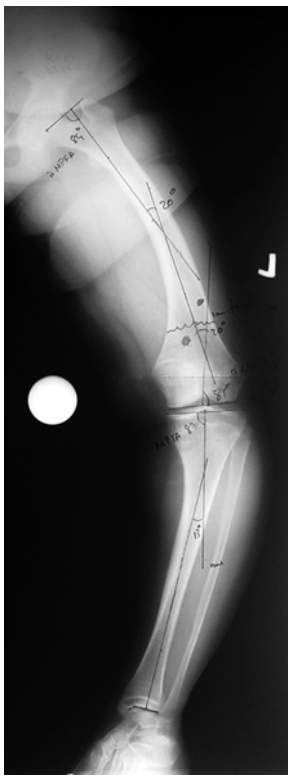


Fig. 15.45 Left frontal orthoroentgenogram of the same patient showing severe genu varum



Fig. 15.47 Left lateral orthoroentgenogram showing anterior bowing (same patient)



Fig. 15.46 Right lateral orthoroentgenogram showing anterior bowing (same patient)

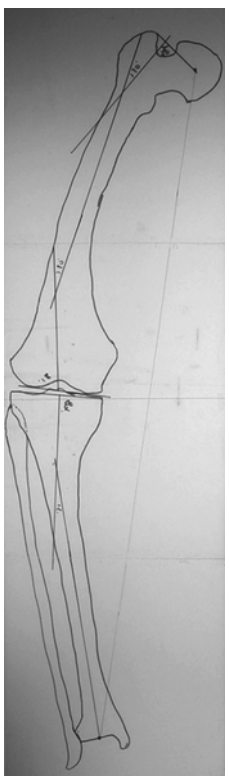


Fig. 15.48 Paper-tracing for deformity analysis on the right

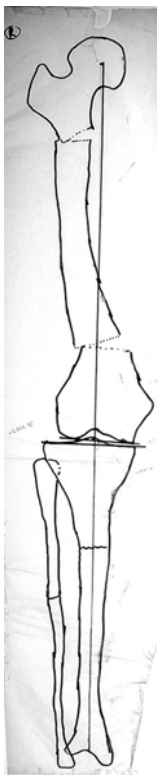


Fig. 15.49 Paper-tracing for surgical simulation



Fig. 15.50 Orthoroentgenogram at the end of the treatment



Fig. 15.51 Photographic documentation of the same patient prior to surgery



Fig. 15.52 Photographic documentation of the same patient after the treatment



Fig. 15.53 Orthoroentgenogram of a patient with X-linked rickets prior to treatment



Fig. 15.55 Orthoroentgenogram of the same patient after 3 months later



Fig. 15.54 Orthoroentgenogram of the same patient after application of hemiepiphyseodesis

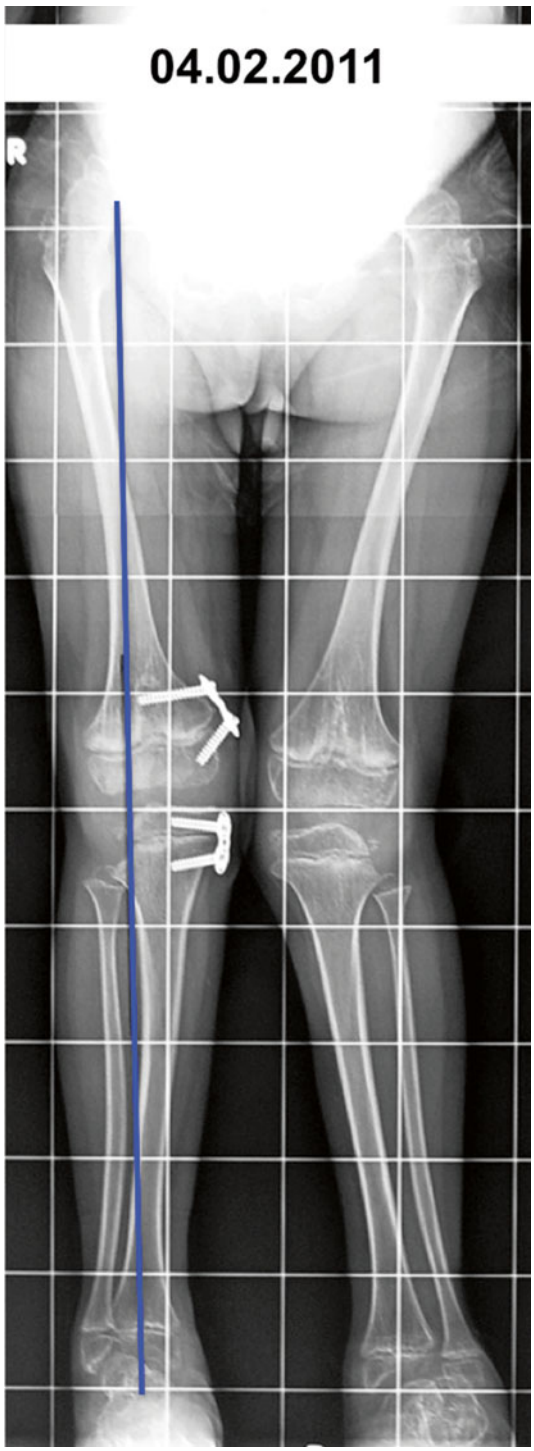


Fig. 15.56 Orthoroentgenogram of the same patient after 7 months later. Please note the correction of the mechanical axis

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Osteogenesis Imperfecta

16

François R. Fassier

Introduction

Until recently, the surgical treatment of osteogenesis imperfecta (OI) was limited to open osteotomies and "Shish Kebab" rodding [1] with multiple complications and high reoperations rate. Since 1992, the use of bisphosphonates has improved the prognosis of these children. Additionally, a team approach (medical treatment, surgery, and postoperative rehabilitation) has led to better functional outcomes [2] even though the medical treatment is not a cure and the bone, though stronger, remains brittle. Therefore, the surgeon and the family must be aware of the necessity for multiple surgical interventions during growth.

The Problems

The preoperative assessment of lower extremities deformities in OI children is crucial.

Bone Deformity(ies)

The structure of the bone is affected: the deformed bone gets thinner in the frontal plane (AP view) and wider in its sagittal plane (lateral view) (Fig. 16.1). As a consequence, the medullary canal is deceptively narrow and the evaluation for the size of an implant is challenging. The cross section of the bones (femur and tibia) is more "rib-shaped" and the triangular aspect of the normal bone has disappeared.

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Consequence: Measuring the size of the implant on the lateral view of the deformed bone may lead to an over optimistic size for a rod and intraoperative problems (fracture).

One fundamental aspect of OI bone surgery is that the child needs more bone than metal: Reaming to enlarge the medullary canal should be limited as it removes bone, and a large implant takes most of the stress in the bone which disappears around the rod. This is a very challenging situation. Prevention is the best way to deal with this problem.

The bone deformity is (like scoliosis) a 3D problem. The 2D pre-op imaging (AP and LAT of the affected bone) gives a limited understanding of the complexity of the deformity. Intraoperatively, prior to making an incision, it is useful to analyze the bone deformity with fluoroscopy: it allows to define precisely the apex (or apices) of the deformity (or deformities) and choose wisely where to perform the osteotomy (or osteotomies).

Soft Tissue Problems

Soft tissues play an important role in the development of bone curvatures in OI. During growth, the soft tissue resists the elongation of the bone. The tether effect leads to a pattern of deformities defined by the most important muscle groups.

In the femur, the posteromedial hamstrings are the most resistant muscles, and therefore the femur bows anterolaterally. In the tibia, most muscles are posterior and lateral, hence the anteromedial bow of the tibia. This "rule" can be modified according to the antenatal position of the fetus: for instance, a child with OI born in a breech position may be born with a recurvatum deformity of the femur due to the extended knee with a flexed hip.

These soft tissues problems can, and will cause difficulties at the time of surgical correction of the deformities. Generally speaking beyond 60° of angulation, it is advisable to shorten the bone (closing wedge resection) to be able to straighten the bone without too much tension in the muscles.

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Fig. 16.1 The medullary canal on the AP view (**a**) is smaller than on lateral view (**b**)

Two important points to check prior to surgery: (1) correction of coxa vara: the amount of free adduction preoperatively gives the amount of possible skeletal correction (adduction test); (2) if the foot is in equinus prior to correcting procurvatum tibial bow, this equinus will be worse at the end of the correction. In such a case, a shortening of the tibia can be done at the time of the osteotomy. A tendo Achilles lengthening can also be done at the end of the procedure.

Before Surgery

At our center, most patients with severe OI receive bisphosphonates soon after birth. Intravenous Pamidronate is the most widely used treatment in North America. Zoledronate is progressively replacing Pamidronate as it is more potent and is administered only twice a year. The acute complications of the bisphosphonates include a flu-like reaction in most children at first cycle treatment as well as risk of hypocalcaemia (significant respiratory distress has been reported in children with preexisting respiratory compromise [3]). Fracture healing does not seem to be affected by the administration of bisphosphonates while osteotomy healing (slow remodeling process) is often delayed. Therefore, no bisphosphonates are given postoperatively (see Box 16.3).

The patient/family must be aware of the risks of complications that can be seen with any type of surgery but also specific complications related to the OI:

- Anesthesia: There is a risk of malignant hyperthermia [4, 5] during surgery, even though a recent paper [6] comparing patients with OI and those without OI did not find a difference between the two groups.
- Difficult intubations (big head, big tongue).
- Ventilation problems (deformed thorax).
- Fracture: The risk is always present during the transfer of patient, while finding an IV access, positioning on the OR table, and during the surgical manipulations.
- Neurologic complications (patients with Type III OI may have hydrocephalus).
- Hemorrhage.

Surgical Correction

Why?

A deformed bone (particularly when weak due to OI) loses its biomechanical properties and is likely to break at the apex of the deformity. Usually the deformity gets worse after a fracture, leading to a vicious circle of fracture/deformity/fracture.

When?

Rodding before walking age is technically more difficult, implant size is reduced and postoperative rehabilitation can be challenging. Since the introduction of bisphosphonates, stronger bones can be obtained in a few months, and therefore there is no need to operate on children before walking age.

How?

"Guided growth" with hemi-epiphysiodesis using non-locking plates or staples is quite popular for managing angular deformities of the lower extremity in children. This technique is very effective when the angulation originates close from the growth plate; and the growth plate has a normal growth potential. It is unlikely to work on a diaphyseal angulation like OI, and with a limited function of the growth plate. Therefore, the indications of hemi-epiphysiodesis in OI are relatively rare. A recent paper from Yilmaz and collaborators [7] demonstrates the safety and effectiveness of "guided growth" in patients with skeletal dysplasias. No case of OI is reported in this series.

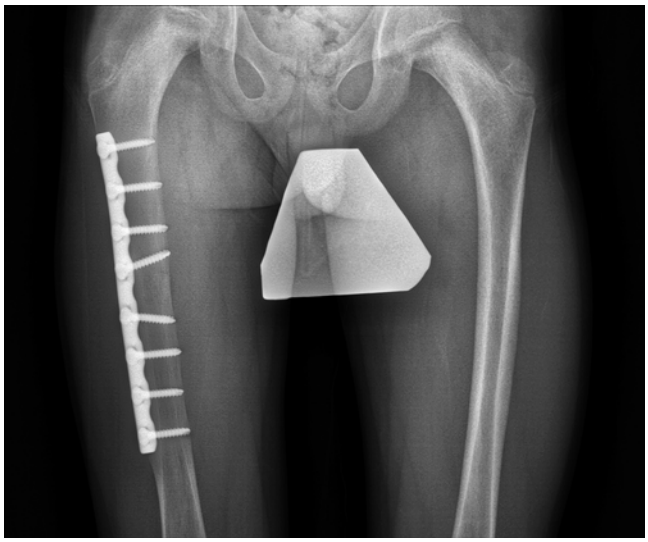


Fig. 16.2 Type I OI, resorption of the bone at the level of the plate, particularly the medial cortex

The use of IM rods was proposed by Sofield and Millar in 1959 [1]. The rod acts as a tutor in the bone, preventing progressive deformity during growth. It reduces the fracture rate, and if a fracture happens, it eases the treatment, preventing the displacement of the bone fragments.

Application of plates is generally not indicated in patients with OI, since the plate creates a rigid segment and the bone is likely to break above or below the implant [8]. Furthermore, under the plate, the bone is not submitted to any stress and resorbs (Fig. 16.2).

Different Types of Rods Can Be Used in OI Bones

Regular rods (such as K-wires, Rush rods, elastic nails) have the advantage of being readily available, relatively inexpensive and come in varying diameters from 1 mm up (K-wires). The Telescopic rods introduced by Dubow-Bailey [9] have been modified by Sharrard [10] Karbowski [11] and more recently by Cho [12]. A new concept of telescopic rod, without the need for arthrotomies has been described by the author [13] with short term promising results. These implants are more expensive, need to be ordered (and currently not available in all countries), and the smallest diameter is 3.2 mm, which is sometimes too large for small children. The major advantage of telescopic (or growing rods) is that the implant gets longer as the child grows, reducing the need for reoperation (which is needed every second or third year with non-telescopic rods because the bone deforms under the tip of the nail during growth) (Fig. 16.3). If a telescopic rod is not available, it is also possible to use "sliding rods": two regular rods (Rush or elastic nails) are inserted: one from proximal epiphysis-distally, and the other rod from the distal epiphysis-proximally. As each of the two rods are inserted from a distinct epiphysis, these implants move away of each other during growth. Unfortunately, they do not provide the same stability as telescopic nails, and it is common to see the bone deform during growth (Fig. 16.4). In their report [14] Laville and Al concluded that sliding rods were not superior to telescopic rods.



Fig. 16.3 Type III OI, deformity of the femur under the tip of the nail with progressive migration through the cortex

Open or Percutaneous Osteotomy?

The growing trend for minimally invasive surgery in orthopedics is beneficial, as it results in less scars and decreased soft tissue trauma (including less blood loss and less postoperative pain); but is technically more demanding. For the femur: before the era of bisphosphonates, OI bone was extremely brittle, but it was easier to do a percutaneous osteotomy, or even an osteoclasis. Since most patients receive bisphosphonate before surgery, the surgeon must be aware that the bone which is harder will break suddenly and not necessarily at the desired place! The amount of preoperative deformity as well as the absence of a medullary canal on X-rays should lead the surgeon to prefer the "open" technique in selected cases.

For the tibia: because of the soft tissue problem (relative equinus deformity) it is better in most cases to do an open, closing wedge osteotomy. It allows appropriate shortening of the tibia so that the foot will be plantigrade following surgical correction.



Fig. 16.4 Sliding rods. (**a, b**) After 4 years, progressive varus deformity aggravated by a proximal femoral fracture (**b**)

Note: Author's preferred method: Whenever possible, I use percutaneous surgery for the femur, but if after 15–20 min, I am not able to reduce the osteotomy and realign the bone, I switch to the open technique.

The relative contraindications for percutaneous femoral osteotomies are as follows:

- Angulation > 60°.
- No medullary canal seen on X-rays.

Osteotomy

The use of an oscillating saw for performing the osteotomy in patients with OI is not recommended: It burns the bone ends, and as these patients receive bisphosphonates (which reduces the number of osteoclasts), the remodeling is slow. It takes time to remove the dead bone and this may lead to delayed union or non-union. It is better to use osteotomes (after preparing the bone with a few drill holes) and clean the bone ends (in the open technique) with a rongeur.

Reduction after osteotomy: the soft tissue tightness is here again a challenge. If there is too much tension at the osteotomy site, do not fight with OI bone! Just shorten as much as necessary to get a good alignment without too much tension.

Note: At the time of reduction, I usually ask the anesthetist to give adequate muscle relaxation to the patient for a few minutes, allowing a smooth manipulation of the bone fragment. If the bone breaks, it may just be a split of one of the fragments: cerclage technique (with resorbable suture) is often sufficient. If the bone bursts in multiple fragments, it may be best to postpone rodding and leave the patient in skin traction for a few weeks.

Do not accept poor alignment as it will likely increase overtime. In the femur, if the nail is too anterior distally, it will perforate the cortex (see Fig. 16.3a). It is easy to do a percutaneous distal metaphyseal osteotomy to be able to center the rod in the middle third of the epiphysis (for telescopic rods) on both frontal and sagittal planes.

In the tibia, the deformity is most exclusively in procurvatum, particularly in the distal tibia. It is advisable to overcorrect to 5–10° of recurvatum to slow down the recurrence of the deformity during growth.

Technique for Femoral Rodding (see Box 16.1)

With an open osteotomy through a posterolateral approach, it is simple to ream the medullary canal to the appropriate size of nail. (Remember that the child needs more bone than metal; avoid excessive reaming!)

If the implant is to be a Rush nail; its introduction through the tip of the greater trochanter (GT) is facilitated with the retrograde insertion of a K-wire that is used as a guide.

Note: The author has modified his own K-wires so that the tip on the blunt end is "cup-shaped" allowing a better contact between the nail and the K-wire guide (Fig. 16.5).

The length of the nail should be calculated from the tip of the GT (or the ossified proximal metaphysis) to the intercondylar notch. Pushing the nail through the growth plate (often the strongest part of OI bone) helps stabilizing the

Box 16.1. Summary of Femoral Fassier-Duval Rod Technique

1. Male rod introduced with male driver.
2. Male rod positioned in the middle third of the distal epiphysis on both AP and LAT intraoperative fluoroscopic views.
3. Remove the male driver, keeping pressure on the nail.
4. Measurement of the female rod, which is cut intraoperatively (Midas Rex®)
5. Female rod introduced: verify that the rod is not too long, as when screwing it in the GT you may push the male distally, into the knee joint.
6. The female rod threads are left proximal to the ossified metaphysis.
7. Cut the male rod above the female and check smoothness of the rod (as it has to slide down into the hollow female nail).

nail which is often "floating" in an empty metaphysis. As long as just one attempt of placing the nail through the growth plate is done, epiphysiodesis is unlikely to occur. For the use of the elastic nails, the original technique was described [15] for normal bones and the entry point is in the metaphysis. The technique has been adapted by Metaizeau for OI bones with a retrograde nail introduced through the epiphysis distally, and an antegrade nail inserted through the greater trochanter. Both nails "slide" away of each other during growth [16]. For sliding rods, care must be taken for the distal rod to be anchored in the epiphysis without disturbing the knee motion. The tip of the rod must be bent and embedded.

The Fassier-Duval telescopic rod is inserted also from the GT and the introduction of the male driver is done first: A K-wire (same diameter as the male nail) is pushed up in the buttock from the osteotomy, and the male driver is pushed down on the K-wire as a guide. When the driver is at the level of the osteotomy, the K-wire is removed and replaced with the nail. After reduction of the osteotomy, the male nail is pushed down in the shaft until it reaches the growth plate, and screwed in the epiphysis. C-arm control of the position in the bone is important: the nail should be in the middle third of the epiphysis both on AP and lateral. It is relatively simple to be centered on AP but in lateral, any mild bow of the femur will drive the nail to be too anterior. In such a case, the nail is withdrawn a few centimeters, a percutaneous metaphyseal osteotomy is done and the nail is pushed in the center of the epiphysis (Fig. 16.6 and Box 16.4). The male driver is removed while maintaining pressure on the nail to avoid dislodging it. The female nail is measured, from the ossified proximal femoral metaphysis to 1 cm above the distal femoral growth plate. The threads of the female nail being left above the ossified metaphysis of the proximal femur (see Fig. 16.6). (The measurement of the nail length does not take the threads of the female nail into consideration.) When the female nail has been cut (Midas Rex®) and the lumen of the nail cleaned to allow a smooth telescoping; the female nail is pushed down over the male nail. At the moment the proximal end of the nail disappears inside the wound, a careful X-rays measurement is done to verify that the distance between the distal end of the female nail and the "aisles" of the male nail ("A" on Fig. 16.7) is greater than the distance between the proximal threads of the female nail and the proximal metaphysis ("B"). If "B" is greater than "A", while screwing the female nail in the non-ossified GT, one will push the male nail into the knee joint. The male nail is cut above the female and the smoothness of the section checked with a little probe to ensure that the male nail slides smoothly distally during growth.

The technique for percutaneous osteotomy(ies) and rodding with the Fassier-Duval is very well described by Paul Esposito [17].

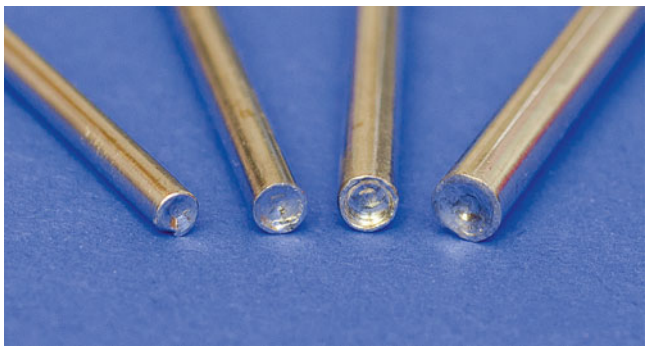


Fig. 16.5 Modified K-wires to serve as guide to introduce a Rush nail

Technique for Tibial Rodding (see Box 16.2)

Whatever the type of rod used, the principles of the technique are the same:

- The osteotomy is done open through a direct anterior approach at the apex of the deformity.
- The nail is introduced through a medial parapatellar tendon incision.
- The entry point of the nail is behind the patellar tendon, on the edge of the proximal tibial epiphysis (which is extra articular). A pre-bent K-wire is useful to find the

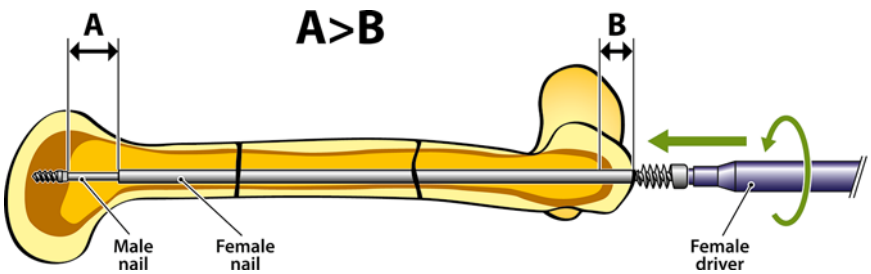


Fig. 16.6 FD rodding. Before screwing the female nail in the non-ossified greater trochanter, verify (via C-arm) that the distance between the distal end of the female nail and the aisles of the male nail A is greater than the distance between the proximal threads of the female nail to the ossified metaphysis B. If B >A the male nail will be pushed distally

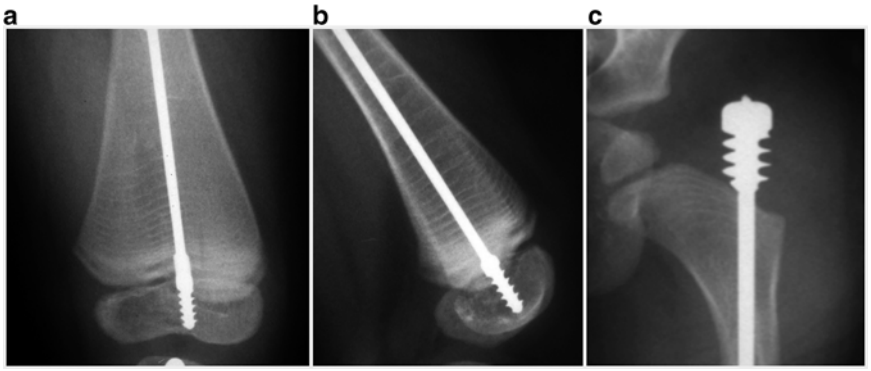


Fig. 16.7 "Ideal" positioning of the FD rod in the femur at the distal (**a, b**) and proximal ends (**c**)

Box 16.2. Summary of Tibial Fassier-Duval Rod Technique

1. Use a bent K-wire to find the medullary canal.
2. Ream over the K-wire (beware of posterior tibial cortex breach if entry point is too anterior.
3. The male driver is pushed down and the male nail positioned in the middle third of the distal tibial epiphysis.
4. The male driver is removed, keeping the male nail in place.
5. Cut the male nail.
6. Measure the female nail.
7. Slide the female nail over the male.
8. Check that the knee extension is not limited by the male nail.

Box 16.3. Surgery and Bisphosphonates

- The medication can be given 48 h before surgery (no bisphosphonates in the blood stream 48 h after infusion).
- No infusion for 4 months postoperatively, or until clear evidence of bone healing is seen on X-rays.

medullary canal. If the tibia is deformed into valgus, it is better to use a lateral parapatellar tendon approach. The direction of the rod will be more in varus, allowing a better correction of the deformity.

Figure 16.8 shows the "ideal" position of the Fassier-Duval rod in the tibia, proximally and distally (see Box 16.4).

Surgical Tips

Long-term immobilization is known to be detrimental to bone health, particularly for children with OI. Therefore,

Box 16.4. Optimal Position of Fassier-Duval Rods

Male Rod

- Middle third of the distal epiphysis both on frontal and sagittal planes.

Female Rod

- The threads should be left above the ossified proximal metaphysis for the femur, and within the proximal epiphysis for the tibia.

when such a child needs to have both femurs and tibiae osteotomized and rodded, different strategies can be used. The most popular is to rod one side at the time, femur first, then the tibia can be done with the use of a sterile tourniquet. The second leg is operated a week later and the postoperative immobilization is 3 weeks (4 weeks for the first side). This surgical approach has the advantage of reducing blood loss and operating time. The disadvantage is to have the child undergo two separate sessions of general anesthesia. Other options depend on patient's needs; if the tibiae are straight and do not need rods; it is common to do both femurs at the same time. It is likely though that the patient will need blood transfusion (except for percutaneous surgeries). Such an option implies that the surgical set-up and the draping are changed between the two sides (see "positioning").

Positioning of the Patient

Most children with OI are smaller than average, and the soft tissue tightness can lead to intraoperative difficulties. A good way to stay out of trouble is to have the patient semi-supine, with a radiolucent cushion (such as a saline bag) under the ipsilateral buttock and another support under the ipsilateral shoulder (Fig. 16.9). The patient is brought to the edge of the table (beware of metallic supports that may interfere with C-arm use). This maneuver has two major advantages: at the beginning of the surgery, the leg can hang off the edge of the table, allowing good control of the proximal femur deformity

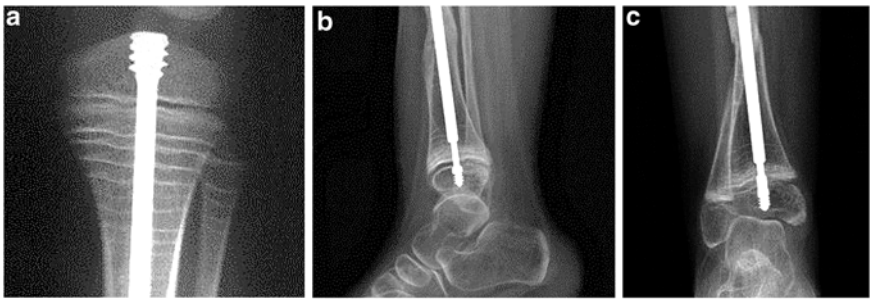


Fig. 16.8 "Ideal" positioning of the FD rod in the tibia at the proximal (**a**) and distal ends (**b, c**)

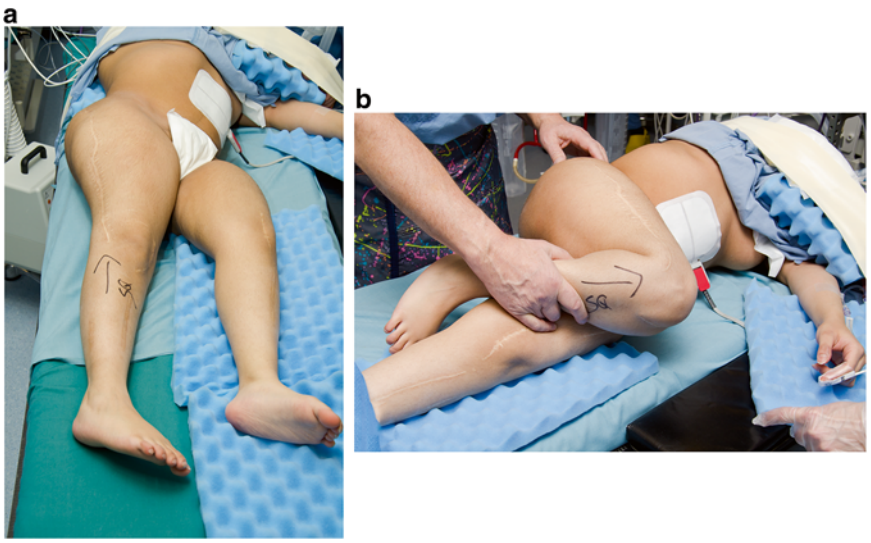


Fig. 16.9 Positioning of the patient on the OR table. The operated side is raised, as is the ipsilateral shoulder. (**a**) This position allows to do lateral shoot through of the knee without changing the C-arm position. (**b**) One must be sure that objects such as the patient's upper extremity, suction tube, cautery cable will not interfere with adequate intraoperative fluoroscopic visualization of the operative field

and rule out a "pseudo coxa vara" (see next section on Coxa Vara). The second advantage is to be able by flexing, adducting, and internally rotating the hip to get a lateral view of the femur without changing the C-arm position. Beware during positioning that the contralateral arm is not left parallel to the trunk, as the hand will be in C-arm field during the surgery. Verify as well that towel clips, suction tube, cautery cable or plate do not prevent a good imaging of the knee during surgery. Another major advantage of this position is at the time of the introduction of the nail through the greater trochanter: it is much easier to manipulate the long tools (male driver in case of Fassier-Duval rodding or the nail itself in other types of rodding) as they can be "outside" the OR table and often lean lower than the level of the table (which would be impossible if the patient was in the middle of the OR table).

Coxa Vara Correction (see Box 16.5)

This deformity is present in about 60 % of Type III OI patients and may progressively develop after IM rodding of the femur [18]. True coxa vara must be differentiated, on X-rays, from "pseudo" coxa vara due to the projection of an anteriorly bowed proximal femur [18]. To do so, the patient is positioned on the edge of the table (semi-lateral as for femoral rodding) (Fig. 16.10), and before surgery, a true AP view of the hip is obtained in letting the leg hang out of the table. In addition, the "adduction test" will give a precise idea of how much correction can be obtained. The surgical technique for coxa vara correction is illustrated in the Fig. 16.11 [19]. The advantages of such a technique are that it is adaptable to varying bone size and the implants (K-wires and cerclage wires) are available in most operating rooms.

Depending on the stability of the fixation in the bone, in most cases, there is no need for postoperative spica cast (which both the patient and parents appreciate). For bilateral

Box 16.5. Coxa Vara in OI

Preoperative examination includes:

- AP view of the hip in extension to eliminate a pseudo coxa vara.
- Adduction test to assess the amount of possible correction.



Fig. 16.10 Before correcting a coxa vara, a true AP view of the hip is obtained with the ipsilateral hip extended off the edge of the table. The anterior bow of the femur is eliminated and a true assessment of the neck-shaft angle is appreciated

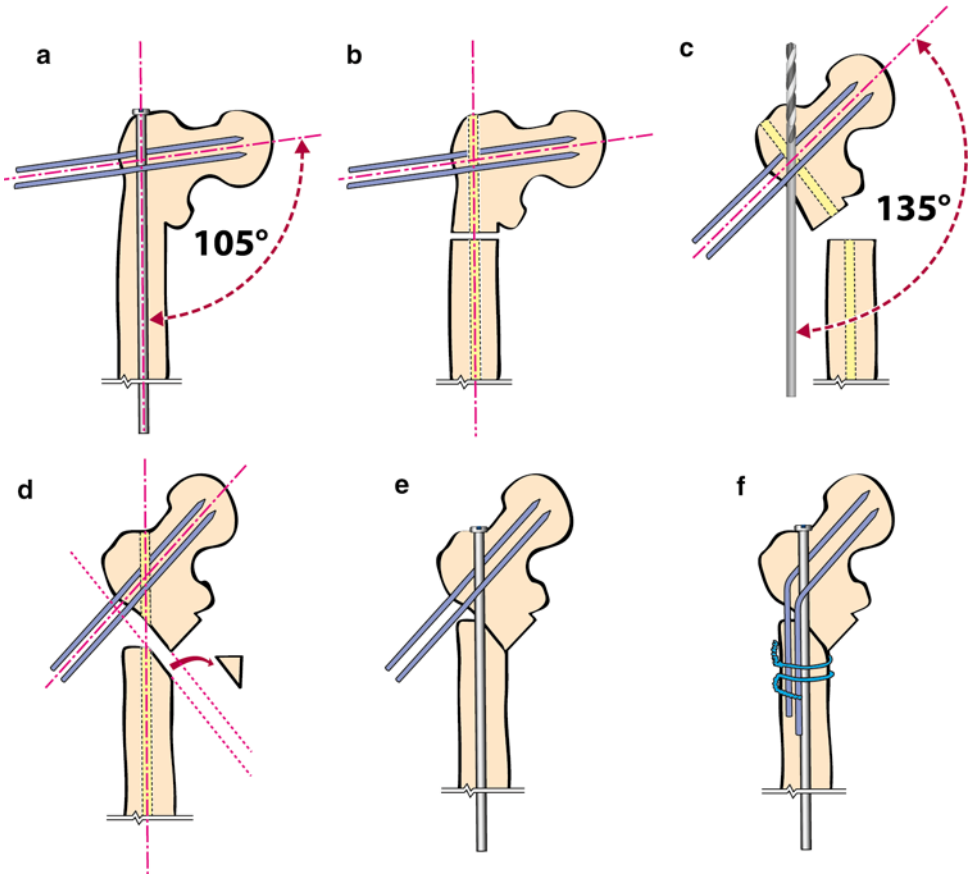


Fig. 16.11 The sequential steps of the coxa vara correction: The two K-wires can be used as a joy stick (**a**–**b**) and the reaming of the proximal fragment is done through the *lateral* cortex (**c**). The angle between the drill bit and the K-wires determines the final neck-shaft angle. The removal of a medial wedge of bone from the distal bone fragment (**d**) allows better stability of the fixation with rodding and cerclage wire (**e**, **f**)

cases (done 1 week apart), an "A" frame can be used, particularly in young patients (Fig. 16.12). No weight-bearing for 6 weeks followed by a week of intensive physiotherapy starting in the pool, allows the patient to recuperate quickly. The results of this technique [19] show that an average 30° of correction of the neck shaft angle can be obtained (Fig. 16.13).

Postoperative Rehabilitation

Third and a crucial part of the team work after medical and surgical treatment is physical rehabilitation. It generally starts 3 weeks postoperatively with the use of braces. Pool therapy and a tilting table are used to progressively help the upright position.



Fig. 16.12 Postoperative immobilization after bilateral femoral and tibial rodding. The tape between the legs avoids spontaneous external rotation of the hips

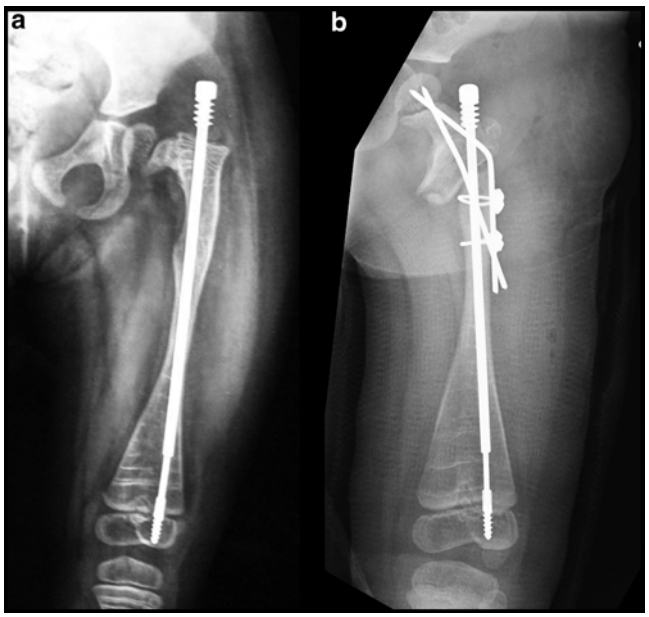


Fig. 16.13 (**a**) Preoperative and postoperative (**b**) radiographs of the femur of a patient with coxa vara associated with Type IV OI

Stretching of Tight Muscles

Most patients with severe anterolateral bow of the femur have a fixed flexion contracture of the hip postoperatively that needs to be stretched. Similarly, the Tendo Achilles needs to be stretched after most tibial rodding. When patients are able to support the standing position, they start using a pediatric walker and progress toward free ambulation depending on the severity of the disease. It must be noticed that the knowledge on muscles in OI is very limited [20].

Use of Braces

The ultimate goal of an OI team is to see their patients walking independently. Nevertheless, because of extreme hyperlaxity seen in few cases, a concern of fracture, particularly at the beginning of the walking age, braces are useful.

- HKAFOs are no longer used by the author.
- KAFOs are used initially postoperatively for patients who have never walked before surgery. As soon as the physiotherapist confirms that the quadriceps strength is sufficient for independent ambulation, the thigh cuff of the KAFO is removed and the knee is unrestrained.
- AFOs: Used at the initiation of walking postoperatively, to help the patient get a good balance. These AFOs must be flexible enough to allow the bones to share the load. The AFOs have an anterior shell to serve as a support as the tibia may deform anteriorly. Patients who have telescoping rods in the tibia, do not need long term use of AFOs.
- SMOs: Very useful to control excessive foot/ankle valgus, due to hyperlaxity.
- Molding of braces can be done at the end of the surgery while the patient is still under general anesthesia. It has several advantages: no pain; no fear for the child; limitations of range of motion easily detectable with less risk of fracture; and finally the braces are ready to be fit on the patient when the cast comes off after 3 weeks.

Results

The combination of medical treatment, surgical realignment-rodding and postoperative rehabilitation has proven to improve "ambulation, gross-motor function, self-care and mobility beyond physiological expectations due to developmental growth" [2]. Such results have also been demonstrated in the long term in adults [21]. Generally speaking, sliding rods or telescopic rods are superior to a single rod with regards to the longevity of the rod [22]. However, complications are frequent and the reoperation rate is high [23] as despite bisphosphonates, the bone remains brittle though harder. The children feel better and their level of activity raises, and with it, the risk of fracture increases.

During growth, the rod can migrate through the cortex (see Fig. 16.3), which is common with non-telescopic nails, and these patients need revision surgery. In other cases, particularly with telescopic rods (if the female rod threads are screwed in the ossified metaphysis), the greater trochanter grows proximally and the tip of the FD rod becomes completely sunk into the bone. The indications for the reoperation in such a case are not clear [24] as the patient may be completely asymptomatic. Retrieving the rod in the bone without an osteotomy is challenging despite rescue tools; and a common solution is to wait until a significant complication (such as fracture) occurs to change the rod. No telescoping may occur after a fracture, but also spontaneously (progressive bowing of the bone and the rod): in such cases, the rod behaves as a regular rod. The indication for revision should then be the same as for solid rods (angulation, fracture, migration of the implant beyond the confines of the bone).

Delayed union/non-union rate was as high as 20 % in OI before the era of bisphosphonates [25]; but with these medications, the risk is much higher [26] and the treatment of such a complication remains challenging. In many patients, the pseudo-arthrosis is mildly symptomatic (recurrent pain after minimal trauma) while in others the pain is debilitating. An innovative solution has been proposed by Dr. Cho [27]: The use of a temporary locking plate, particularly in the subtrochanteric area. However, long term results of this technique are currently not available.

Another challenge for the surgeon is the long and thin bone often seen in adolescents with Type III OI. In such cases, a single Rush rod can be used, but a locking nail is a safer implant with no need for post-op immobilization. Unfortunately, most locking nails are 8.5 mm in diameter and greater, which may be too large for patients with OI. The GAP nail developed by Drs. Galban and Para [28] seems promising in such cases (with diameters between 4.8 and 8 mm).

Surgical correction of the lower limb deformities in OI has become more frequent since these patients benefit from medical treatment, including the use of bisphosphonates. In the future, other drugs such as Denosumab (for OI Type VI), TGF beta-blockers and sclerostin antibody will likely further improve the bone quality in such patients, while awaiting other advances such as gene therapy for this condition. The challenge for the orthopedic surgeon is to continue to develop minimally invasive techniques and better implants until metallic implants will no longer be necessary for patients with OI.

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17 Lower Limb Deformity in Neuromuscular Disorders: Pathophysiology, Assessment, Goals, and Principles of Management

Unni G. Narayanan

Cerebral Palsy

Cerebral palsy (CP) refers to disorders of the development of movement and posture that arise from a disturbance to the developing fetal or infant brain. These motor disorders are permanent but not progressive and can be accompanied by deficits of sensation, perception, cognition, and communication; seizure disorders; and secondarily acquired musculoskeletal deformity [1, 2].

CP accounts for the most common cause of chronic childhood disability, with a prevalence of 2–3 per 1,000, and is associated with a wide range of etiologies [3]. Prematurity is one common risk factor due to the fragility of the cerebral vasculature and immaturity of the autonomic nervous system that controls cerebral perfusion. Some additional risk factors include congenital brain malformations, intrauterine infections, placental malformations, prenatal brain hemorrhages, or strokes; perinatal causes of hypoxic-ischemic encephalopathy and hyperbilirubinemia; and postnatal strokes, infections, or traumatic and other kinds of brain injuries. A number of genetic disorders (Rett syndrome, fragile X syndrome, familial or hereditary spastic paraparesis) and metabolic disorders (mucopolysaccharidoses, phenylketonuria, etc.) can mimic features of CP.

Classification of Cerebral Palsy

CP is best classified by the Gross Motor Function Classification System (GMFCS), which has been shown to be reliable, valid, consistent, and highly prognostic [4, 5]. The GMFCS comprises five levels of severity (Fig. 17.1). Children in level I walk independently without a walking aid and can perform most activities of typically developing children, with some difficulties with speed, balance, and coordination. In level II, children are independently ambulant without a walking aid, but require support in more challenging situations such as on uneven surfaces or while climbing stairs. In GMFCS level III, children rely on walking aids such as crutches or walkers for at least some of their functional mobility. Some may use wheelchairs for longer distances. Children in GMFCS levels IV and V rely entirely on a wheelchair for any functional mobility. In level IV, children can usually sit without support and do some functional weight bearing for transfers. Walking, if possible, is limited to short distances with a walker, for exercise purposes. Children classified as level V are unable to sit without support, and weight bearing, if at all, is limited to supported weight bearing in a stander for exercise rather than functional purposes. These children depend on a caregiver for much of their activities of daily living.

CP is also classified by the type of abnormal tone or movement disorder. The location and extent of brain involvement account for the type and severity of the abnormal tone and movement disorder as well as the topographical distribution of involvement of the trunk and limbs. Lesions involving the pyramidal tract are associated with spasticity or velocity-dependent hypertonia, whereas lesions of the basal ganglia are associated with dyskinesias including dystonia or choreoathetosis. Dystonia is characterized by jerky involuntary movements of the limbs or trunk and neck. Choreoathetosis is characterized by involuntary writhing movements. Cerebellar involvement is associated with ataxia and hypotonia. Some children have features of more than one type or "mixed"-tone abnormalities. The commonly used topographical classification, hemiplegic, diplegic (lower extremities involved more than upper extremities), and quadriplegic or total body involved, has some clinical and prognostic utility, but these terms are not very reliable. Instead, the regional involvement may be categorized as being "unilateral" or "bilateral," which can be symmetric or asymmetric with one side more involved than the other.

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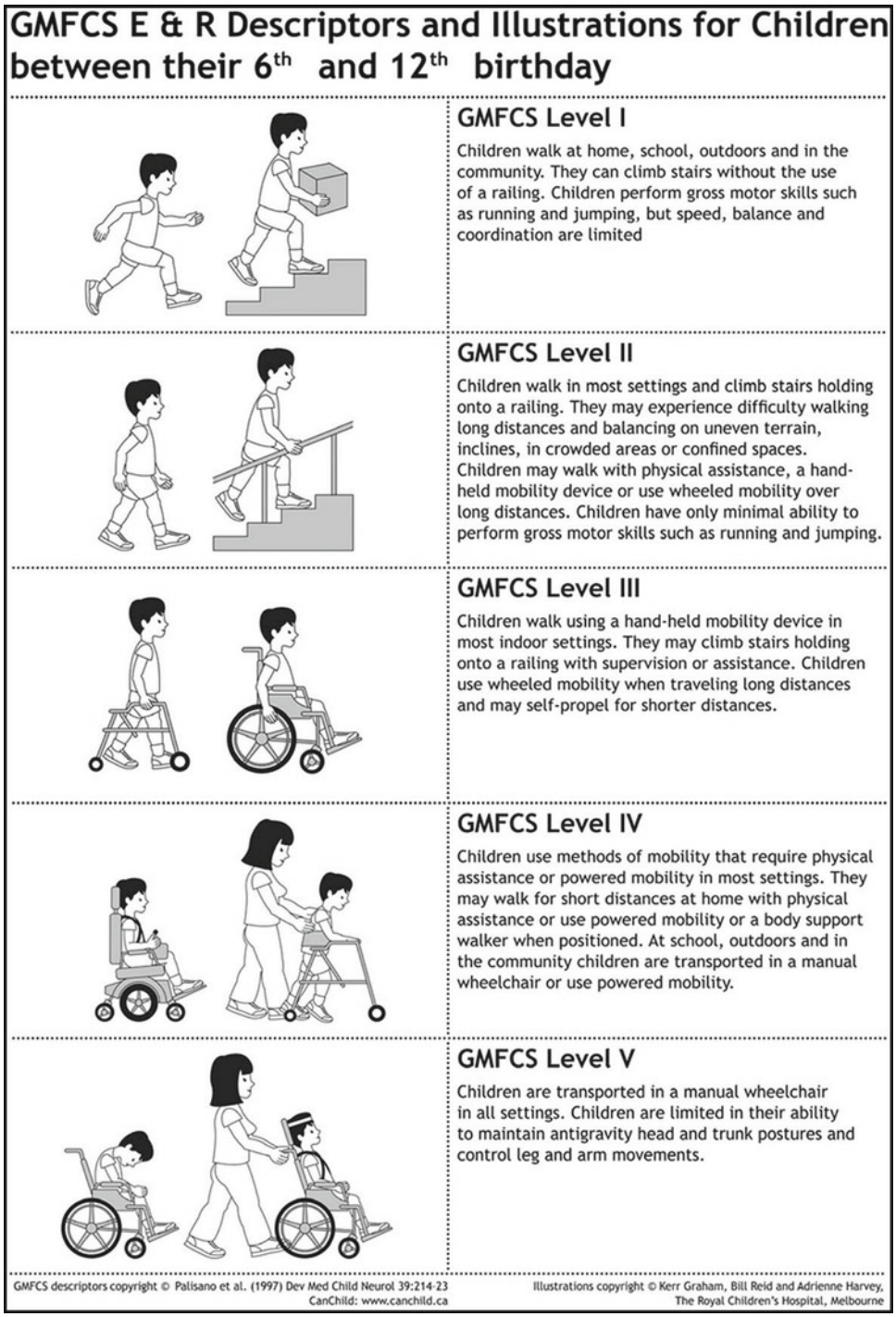


Fig. 17.1 GMFCS levels. Reproduced with permission from copyright © Kerr Graham, Bill Reid, and Adrienne Harvey, The Royal Children's Hospital, Melbourne

Pathophysiology of Musculoskeletal Deformity in Cerebral Palsy

Musculoskeletal deformities in CP are acquired secondarily. Characteristically, upper motor neuron disorders are associated with hypertonia, hyperreflexia, and co-contraction of agonist–antagonist muscles. Other features can include loss of selective motor control, poor centrally mediated balance and coordination, and muscle weakness [6]. These children often have a delay in the development of the motor milestones.

Hypertonia is associated with "dynamic" muscle contractures. Muscles are taut due to the increased muscle tone, but they are of sufficient length. Muscles typically grow in response to the stimulus of stretch. In a child with CP, developmental delay prevents the child from engaging in the typi-

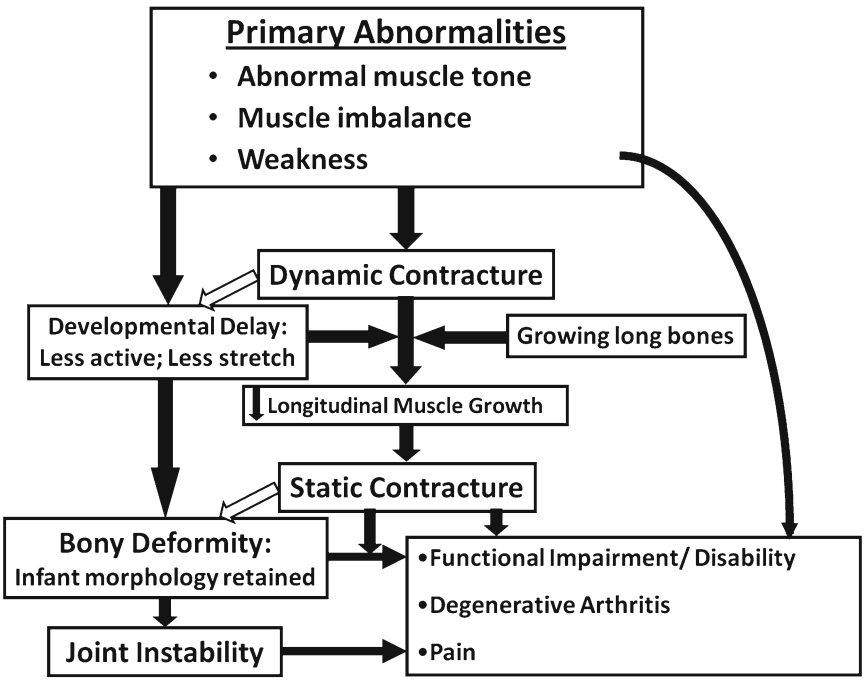


Fig. 17.2 Pathway to secondary musculoskeletal pathology in cerebral palsy. Adapted from Noordin S, Narayanan U. Neuromuscular Disorders, Chapter 64. In: Mercer's Textbook of Orthopedics and Trauma, 10th Edition. Boca Raton: CRC Press; 2012

cal physical activities that would provide the source of stretch. The hypertonia further impedes the extent of any stretch. Consequently, muscle growth is impaired relative to bone growth. This results in the development of "static" contractures where the muscles are both tight and too short [7]. The acquisition of milestones, from rolling over into the prone position, sitting upright, crawling, pulling up to stand, cruising, and walking, typically occurs during the first 12–15 months of a child's life. The growing skeleton undergoes significant changes during this time, in response to the natural stresses applied to the skeleton. The delay in motor milestones, or in some cases failure to achieve them at all, deprives the skeleton of these important corrective forces, resulting in the retention of infantile morphology of the skeleton (e.g., increased femoral anteversion and coxa valga in the proximal femur). Furthermore, the abnormal muscular forces acting on the growing skeleton contribute additionally to the development of bony deformities and joint instability over time. Joint instability in these patients is often a consequence of both the bony deformity and the unbalanced forces of the agonist–antagonist muscles that bridge across a joint. The skeletal deformities result in lever arm dysfunction [8]. In the lower extremity, the interaction of joint contractures, muscle weakness, bony deformities, and joint instability at multiple levels is associated with inefficient (higher energy consuming) pathologic and compensatory gait patterns in ambulant children. This leads to limitation in physical function or increase in symptoms such as pain and fatigue. In nonambulatory children, contractures and deformities of the lower limbs contribute to discomfort; difficulties with care, positioning, and mobility; and poorer quality of life. Although the primary lesion in the brain is not progressive, the secondary musculoskeletal consequences become worse with growth [9–12] (Fig. 17.2).

Specific Deformities of the Lower Extremity

Pelvis and Hip

Across the hip joint, adductors and flexor muscle groups are typically more involved, overpowering their antagonist counterparts, the hip abductors and hip extensors. This muscle imbalance often results in adduction and flexion contractures of the hip in the coronal and sagittal planes, respectively. The hip flexion contracture arising from the iliopsoas can be accompanied by anterior pelvic tilt with associated lumbar lordosis. Anterior pelvic tilt might also be secondary to weak hip extensors or weak anterior abdominals or a combination of these [13, 14]. In contrast, contracted hamstrings can pull down on the ischium resulting in a relative posterior tilt of the pelvis. In the transverse plane, the hip commonly lies internally rotated. This is more likely due to increased femoral anteversion rather than an internal rotation contracture as such [15]. In bilaterally involved children, the muscle contractures and bony deformities may not be symmetric. The more severely involved side might be more adducted and internally rotated. If this is accompanied by relative abduction and/or external rotation of the contralateral side, the deformity is categorized as "windswept" towards the abducted/externally rotated side (Fig. 17.3).



Fig. 17.3 Windswept hips

In CP, the morphology of the proximal femur is often characterized by a relative increase in the neck-shaft angle in the coronal plane (coxa valga) and excessive femoral anteversion in the transverse plane (Fig. 17.4a). Femoral anteversion is the angle in the transverse plane, by which the neck of the femur is directed (forward) from the transcondylar or coronal plane [16] (Fig. 17.4b). The terms femoral anteversion and femoral (medial) torsion are used interchangeably, the latter term suggesting that the orientation of the proximal femur relative to the coronal plane is a consequence of torsion occurring along the shaft of the femur rather than in the neck [17–19]. In the newborn, both femoral anteversion and neck-shaft angle are increased. This might be related to the fetal position. As the growing infant begins to roll over and crawl and pull up to stand and walk, the progressive extension of the hip is believed to push back the femoral neck axis to the physiologically normal values of older children and adults by the age of 1 or 2 years. The developmental delay of CP results in the retention of uncorrected higher degrees of fetal alignment [20]. This is borne out by the finding that the mean neck-shaft angle increases with the severity of CP by GMFCS level from 135.9° (range 130° to 145°) at GMFCS level I to 163.0° (range 151° to 178°) at level V [21]. Femoral anteversion also increases with GMFCS level but seems to plateau at level III to level V, where it averages 40.0° (range 25° to 67.5°). Whether the abnormal proximal femoral geometry is further affected by the abnormal forces associated with adduction/flexion contractures is less clear. The reduced femoral head coverage associated with increased femoral anteversion is compensated for by increased internal rotation of the femur and increased anterior tilt of the pelvis [15].

At birth, the hip joint is typically normal in children with CP. Hip instability is usually acquired secondarily due to the interaction between the adduction and flexion contracture with the increased neck-shaft angle and anteversion, contributing to the (postero) lateral displacement of the femoral head. The magnitude of the hip displacement (subluxation) is quantified using the Reimer's migration percentage (MP) [22] (Fig. 17.5). The "hip at risk" is defined as the hip that is likely to progress to hip dislocation if left untreated. This has been variously defined as the hip that reaches a migration percentage of 30–40 %. The proportion of children who will reach this threshold is directly correlated with increasing GMFCS level in three population-based studies [23–25]. Using 30 % as the definition of hip at risk, the incidence is virtually 0 % for GMFCS level I but increases to 90 % for level V [25]. If, however, a threshold of 40 % is used to define a hip at risk, the incidence is 65 % for GMFCS level V [24]. This is probably a more valid estimate of the risk because it has been observed that approximately one-third of hips with an MP between 33 and 40 % will improve spontaneously or will not deteriorate [24]. Progressive subluxation is very likely if the MP is greater than 40 % in a child still growing (Fig. 17.6a–c). The posterolaterally displaced femoral head pushes up against the superolateral surface of the acetabulum leading to increasing acetabular dysplasia. In association with hip flexion contracture, the acetabular deficiency tends to be more posterior [20]. The acetabular dysplasia is quantified by the acetabular index when the tri-radiate cartilage is open, or by the acetabular angle of Sharp when the tri-radiate cartilage is closed [26]. Initially the subluxated femoral head might reduce in the abducted position, but eventually the dislocation becomes irreducible. The uncovered femoral head is subject to the abnormal forces of the overlying capsule and abductors draped over the superolateral surface of the femoral head (Fig. 17.6d). This leads to loss of sphericity and progressive flattening of this surface leading to increasing femoral head deformity. Ultimately, there is erosion of the articular cartilage and degenerative changes consistent with osteoarthritis (Fig. 17.6e).

Knee and Lower Leg Segment

At the knee, the most common deformity is in the sagittal plane, knee flexion deformity. The knee flexion deformity has a wide range of etiologies. It can arise from contracture

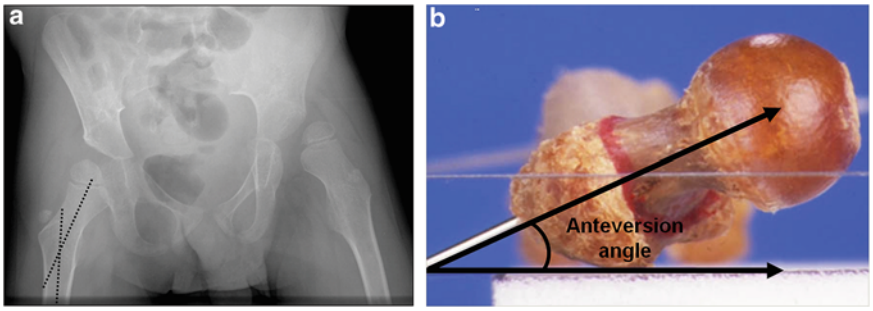


Fig. 17.4 (**a**) Coxa valga. (**b**) Femoral anteversion. Adapted with permission from Narayanan UG. Chapter 26. In: Weisel SW (editor). Pediatrics in Operative Techniques in Orthopedic Surgery. Vol. 2, Part 4. Philadelphia: Wolters Kluwer Health; 2011

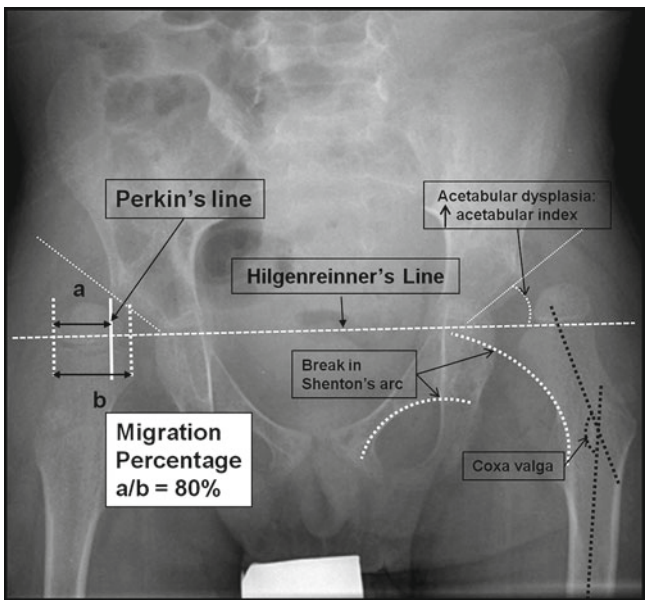


Fig. 17.5 AP radiograph of the pelvis with radiographic parameters quantifying hip displacement and acetabular dysplasia

of the medial and/or lateral hamstrings. Since the medial hamstrings (semitendinosus and semimembranosus) and part of the lateral hamstrings (long head of biceps femoris) take origin on the ischium which is above the hip joint, a "functional" contracture of the hamstrings is possible even in the presence of normal or even excessive length of the hamstrings when there is an associated anterior pelvic tilt that raises the origin of these muscles [13, 14, 27]. Knee flexion deformity might also be consequence of a weak gastrocsoleus complex leading to the forward translation of the tibial shank over the foot, leading to the ground reaction force passing posterior to the knee resulting in a (external) knee flexion moment that has to be counteracted with a corresponding (internal) extension moment using the quadriceps. If the knee extensors are weak, the knee goes into progressively increased flexion. A gastrocnemius contracture can also lead to flexion deformity of the knee to take the tension off the gastrocnemius, especially if the foot is being held plantigrade in a brace. A combination of these factors can be at play. Initially a flexion deformity present during weight bearing or walking may not be associated with a fixed flexion contracture. Over time passive extension is likely to become restricted with increasing magnitude of fixed knee flexion contracture due to inadequate length of the hamstrings and/or a posterior capsular contracture. Weight bearing in the crouch position increases the joint reaction force on the patella-femoral joint which can cause anterior knee, patella alta, and avulsion fractures of the inferior pole of the patella (Fig. 17.7).

Box 17.1. Multiple Etiologies of Knee Flexion Deformity

1. True contracture of the hamstring/s
2. "Functional" contracture of hamstring/s: Increased anterior pelvic tilt due to any cause raises the origin (ischial tuberosity) of the medial hamstrings and long head of biceps, which in turn contributes to flexion at the distal insertion. Excessive anterior pelvic tilt can be due to:
   (a) Hip flexion contracture
   (b) Weak hip extensors
   (c) Weak anterior abdominal muscles
   (d) Compensatory strategy for excessive femoral anteversion
3. Weak plantar flexion—Knee extension couple leads to forward lean of the tibia shank over the foot, leading to the ground reaction force passing posterior to the knee, causing an increased external flexion moment. Reasons for this include:
   (a) Weak gastrocsoleus complex, often iatrogenic due to over-lengthened tendo-Achilles, in a child with spastic diplegia
   (b) Hindfoot valgus with midfoot break and forefoot abduction
   (c) Excessive external tibial torsion
4. Gastrocnemius contracture (dynamic or static): Knee bends to allow foot to be plantigrade by taking the stretch off the contracted gastrocnemius.
5. Posterior capsular contracture of the knee joint.
6. Combinations of above.

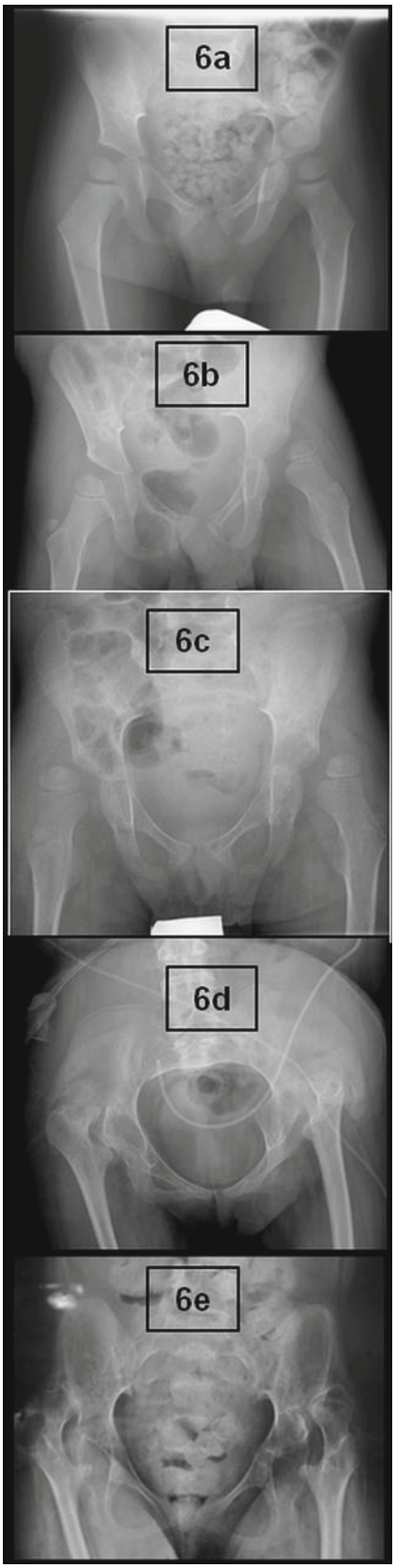


Fig. 17.6 (**a**–**c**) Hip at risk progressing to increased subluxation and acetabular dysplasia. (**d**, **e**) Dislocated femoral head leading to loss of sphericity and advanced degenerative changes

True coronal plane deformity at the knee is uncommon. However, the medial rotation of the hip, due to increased femoral anteversion, which is characterized by a corresponding medial rotation of the knee (patella), leads to an apparent (pseudo) valgus appearance of the (flexed) knee. This appearance is exacerbated if the femoral medial torsion or anteversion above the knee is accompanied by external tibial torsion below the knee. The tibial segment can be associated with normal torsion, and increased external or increased internal tibial torsion. At birth, the torsion of the tibia is relatively internal, with the bimalleolar axis typically being neutral in the coronal plane. The tibia undergoes external rotation maximally during the first 4 years and an average of 1 degree per year until skeletal maturity when it reaches an average of 28 degrees external [28]. In some children with CP, this internal torsion may persist due to the failure of the natural lateral rotation to occur. In others, significantly increased external tibial torsion can occur often in association with valgus hindfoot, flattening of the midfoot, and forefoot abduction. This is associated with a poor lever arm for push off in gait and is often accompanied by crouch position at the knee due to the weak plantar flexion-knee extension couple (Fig. 17.8).

Ankle and Foot

Equinus of the ankle is the most common deformity of the lower extremity in CP. Equinus arises from an imbalance between the plantar flexors, which might be hypertonic and/or contracted, and the dorsiflexors due to weakness of the tibialis anterior. What begins as a dynamic contracture due to spasticity is likely to become fixed when the gastrocnemius-soleus complex fails to keep up with the tibial growth. In bilateral CP, the gastrocnemius is usually more involved than the soleus. Consequently, the ankle might have sufficient dorsiflexion when the knee is flexed but with the knee extended, the ankle equinus becomes evident due to the stretch of the gastrocnemius muscle at its origin above the knee (Fig. 17.9). In unilateral CP (hemiplegia) the soleus tends to be equally involved as the gastrocnemius, such that the equinus contracture is unchanged with the position of the knee. This distinction is important to assess as it will influence which musculotendinous structure should be targeted for intervention: gastrocnemius alone or both the gastrocnemius and the soleus or tendo-Achilles.

In the coronal plane, the hindfoot might be in physiologic valgus position, or deformed into excessive valgus or varus. These coronal plane deformities typically arise in combination (coupled) with ankle equinus. Equinovalgus deformities are more common in bilateral CP (diplegics). With the hindfoot in valgus, the talus and calcaneus are typically plantarflexed. There is a collapse of the subtalar joint (midfoot break) with the prominence of the head of the plantarflexed talus, and the midfoot is everted taking the forefoot with it into an abducted position (Fig. 17.10a, b). There is a flatten-

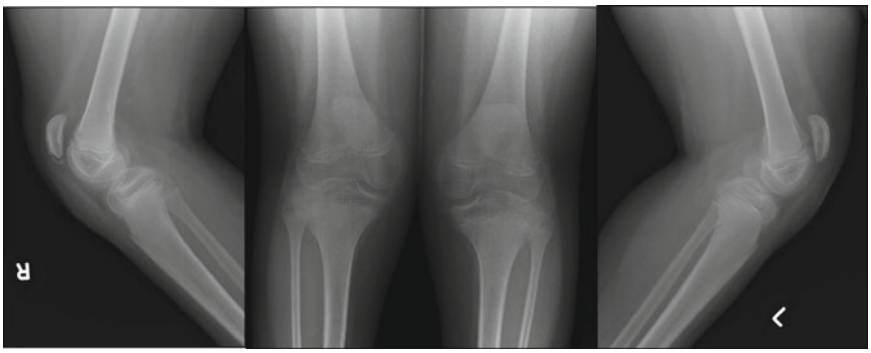


Fig. 17.7 Bilateral patella alta due to long-standing crouch with avulsion fractures of the inferior pole of patella



Fig. 17.8 Torsional malalignment: Pseudo-valgus due to increased femoral internal torsion and increased external tibial torsion

ing of the longitudinal arch and a pronated appearance, although in fact the foot might be relatively supinated to compensate for the hindfoot valgus. Passive correction of the hindfoot out of the valgus position unmasks the supinated position of the foot. The abducted forefoot is often accompanied by a hallux valgus deformity that is acquired over time due to the foot rolling off the medial border during push off. To test the true dorsiflexion of the ankle, the hindfoot valgus must be passively corrected to lock the subtalar joint prior to dorsiflexing the ankle. Failing to do so allows dorsiflexion to occur about the unlocked subtalar joint and provide a spurious estimate of dorsiflexion. The equinovalgus foot with forefoot abduction provides a very ineffective lever arm to generate any power during push off (Fig. 17.11a, b). The tibial shank falls forward over the foot due to dorsiflexion through the subtalar joint, and shifts the ground reaction force behind the knee resulting in a flexed knee or crouched position.

Equinovarus deformities are more common in unilateral CP (hemiplegics). The hindfoot varus might be a consequence of an overactive tibialis posterior and/or tibialis anterior muscle in combination with weak peroneals. The deformity might be flexible or rigid. The hindfoot varus might be accompanied by midfoot cavus due to the tight plantar fascia or foot intrinsic muscles, and forefoot adductus. Weak tibialis anterior along with overactive peroneus longus might lead to increased plantar flexion of the first ray. This might be the primary problem resulting in a compensatory hindfoot varus to maintain a weight-bearing tripod. This can be tested using the simulated Coleman block test. In the prone position, the lateral border is pushed down on to simulate standing on the Coleman block [29]. The hindfoot varus corrects to normal valgus and the increased plantar flexion of the first metatarsal ray becomes apparent relative to the remaining rays, confirming that the hindfoot varus is secondary to the plantarflexed first ray. The hindfoot varus leads to increased weight bearing along the lateral border of the foot leading to callus formation and even stress fractures causing discomfort and difficulties with brace and shoe wear (Fig. 17.12).

Lower Limb Assessment in the Ambulant Child (GMFCS Levels I–III)

The physical examination of the ambulant child begins with a visual assessment of the gait to identify overall abnormal gait patterns and specific problems. This dynamic assessment is followed by an on-table examination of the lower limb tone abnormality, range of motion of the major joints, and skeletal alignment to explain the gait abnormalities identified



Fig. 17.9 Equinus contracture involving the gastrocnemius more than the soleus (Silfverskiold test)

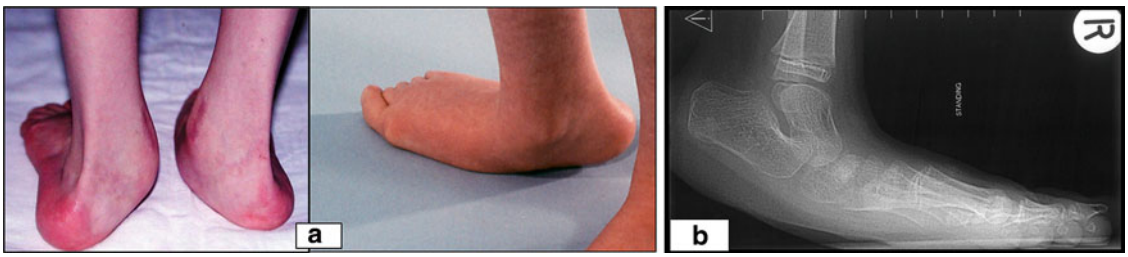


Fig. 17.10 (**a**) Equinoplanovalgus deformity with hindfoot valgus, midfoot break, forefoot abductus, hallux valgus. (**b**) Weight-bearing lateral radiograph of equinoplanovalgus deformity

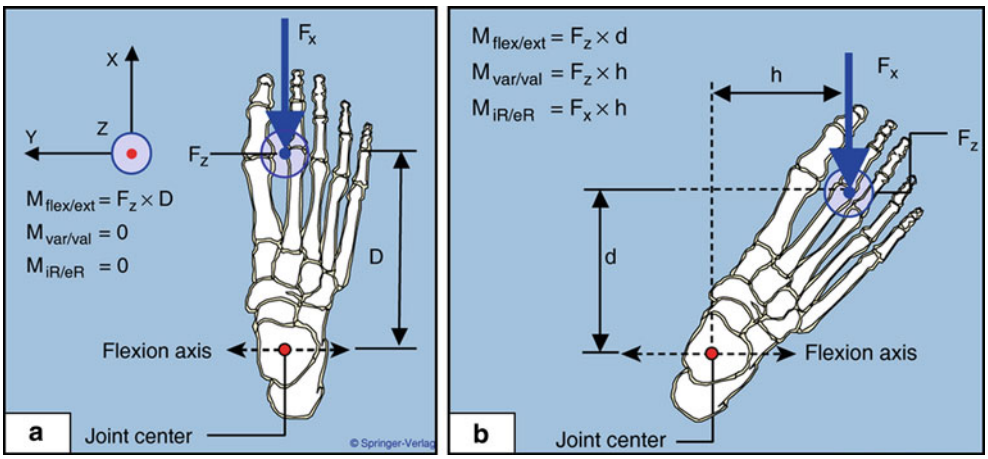


Fig. 17.11 (**a**) Lever arm of the normal foot. (**b**) Lever arm deficiency of the equinoplanovalgus and abducted foot. Figures reproduced with permission from Paley D. Gait considerations. Berlin, Heidelberg: Springer-Verlag; 2003, with kind permission from Springer Science and Business Media



Fig. 17.12 Equinocavovarus deformity in a hemiplegic child. Note the plantarflexed first ray and the stress fracture of the 5th metatarsal due to excessive loading of the lateral border

during walking. These should in turn be linked to specific gait-related problems reported by the child during the elicitation of the history prior to the physical examination.

Box 17.2. Assessment of the Ambulatory Child

1. Generate gait-related problem list from child and parent/s
2. Observational assessment of the gait: Dynamic exam
   (a) With and without orthotics
   (b) With and without walking aids if possible
3. Examination on the table: Static exam
   (a) Muscle tone
   (b) Muscle length: Passive range of motion
      - Fast stretch (R1)
      - Slow stretch (R2)
   (c) Muscle strength
   (d) Selective control
   (e) Bony alignment
4. Radiographic imaging
5. Video and 3-D gait analysis

Box 17.3. Five Prerequisites of Gait

1. Stability of the weight-bearing side during stance
2. Clearance of the foot in swing phase
3. Appropriate pre-positioning of the swing-phase foot for the next step
4. Adequate step length
5. Energy conservation

Observation of Gait

Inspect the child's walk from the front, back, and the side to best estimate the gait abnormalities in all three planes. The child should walk both in their usual orthotics and footwear and barefoot if possible. The assessment is both qualitative and quantitative. A useful framework to evaluate the global quality of the gait is to assess the five prerequisites or attributes of gait [30, 31]: (1) stability in stance; (2) (foot) clearance in swing; (3) correct pre-positioning of foot for the next step; (4) adequate step length; and (5) energy efficiency (minimizing the excursion of the body's center of mass during walking). This is followed by a systematic assessment for specific abnormalities at each level: foot and ankle; knees, hips, and pelvis; and trunk and upper extremities, with due consideration of the coronal, sagittal, and transverse planes to estimate the reasons for any compromise in any five priorities of gait. This quantitative assessment can be greatly enhanced by capturing the gait with video (ideally split screen to see the frontal and sagittal planes simultaneously and in slow motion) [32].

Gait analysis provides a more sophisticated assessment of gait patterns and deviations. Three-dimensional gait analysis generated in a motion laboratory provides kinematic data that describes the dynamic position of each limb segment in each of the three planes; kinetic data which includes moments and powers about the joints; dynamic electromyography which documents the timing of specific muscle activation during the gait cycle; and pedobarographs which provide useful information about the weight-bearing characteristics of the stance-phase foot. 3-D gait analysis provides insight beyond what is derivable from observational analysis alone, and can help distinguish primary abnormalities (which might benefit from treatment) from secondary abnormalities that represent coping responses (which requires no treatment) [32–34]. Gait analysis has the potential to influence or alter treatment decisions for at least some patients [35–37]. Gait analysis has identified recognizable gait patterns which can be classified and used for making treatment decisions [38, 39]. Finally, the effectiveness of these interventions can be assessed objectively using gait analysis as a measure of gait outcomes [40].

Abnormal Gait Patterns in Cerebral Palsy

In bilateral CP, there are broadly recognizable patterns [38] (Fig. 17.13).

True equinus pattern is characterized by the absence of a heel strike and a toe-toe pattern because the ankle remains plantarflexed throughout stance phase. If the equinus is not severe, initial contact may be with the forefoot and the heel might even come in contact with the ground in midstance. The knee can be flexed during stance phase to accommodate the equinus, or it may come to full extension or even hyperextend to allow the plantarflexed foot to remain flat on the ground for more stability (Fig. 17.14a). True equinus pattern can be associated with poor stability due to the narrow base of support and poor clearance if the knee does not flex enough to facilitate clearance of the plantarflexed foot, leading to foot drag and tripping. Step length is typically shortened because the plantarflexed foot leads to an early forefoot strike. The decreased step length might also be a consequence of poor stability in the stance-phase limb forcing the contralateral swing-phase limb to contact the ground sooner.

Jump gait pattern is characterized by toe-toe pattern due to equinus in combination with a pathologically flexed knee and flexed hip that resembles someone preparing to jump. All five priorities of gait are affected in this pattern. Step length is further compromised as the swing-phase knee might not be able to extend if increased hamstring activity prevents the knee from reaching full extension in terminal swing.

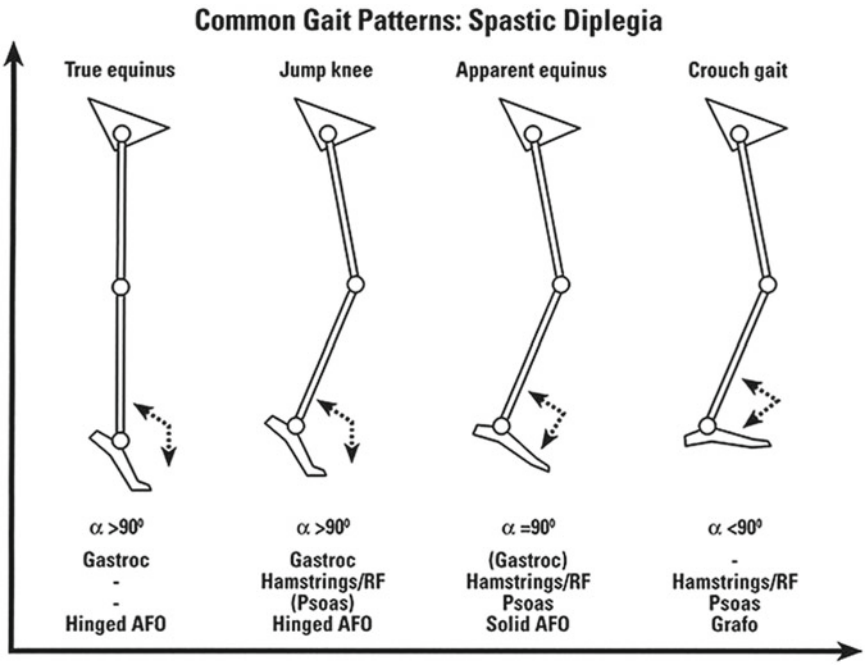


Fig. 17.13 Common gait patterns in bilateral (diplegic) cerebral palsy. Reprinted with permission from Rodda J, Graham HK. Classification of gait patterns in spastic hemiplegia and spastic diplegia: a basis for a management algorithm. Euro J Neurol 2001; 8 (Suppl 5): 98–108

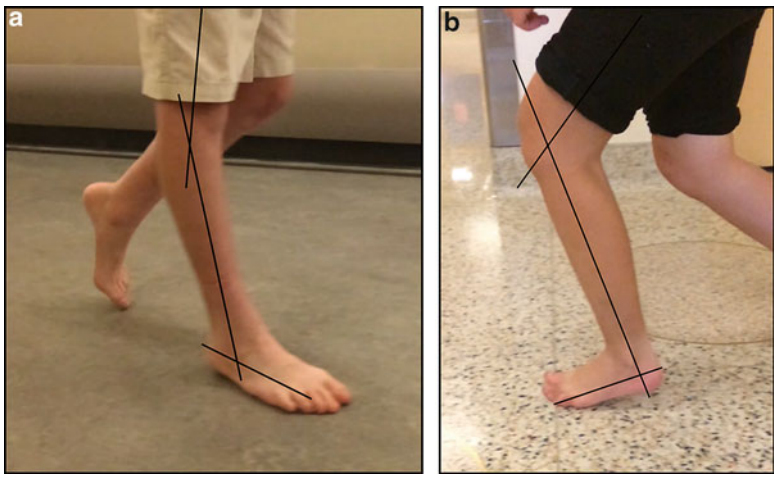


Fig. 17.14 (**a**) Knee hyperextended to accommodate equinus contracture with flatfoot contact despite plantarflexed position of ankle. (**b**) Forefoot contact (apparent equinus) to accommodate fixed flexion deformity of the knee

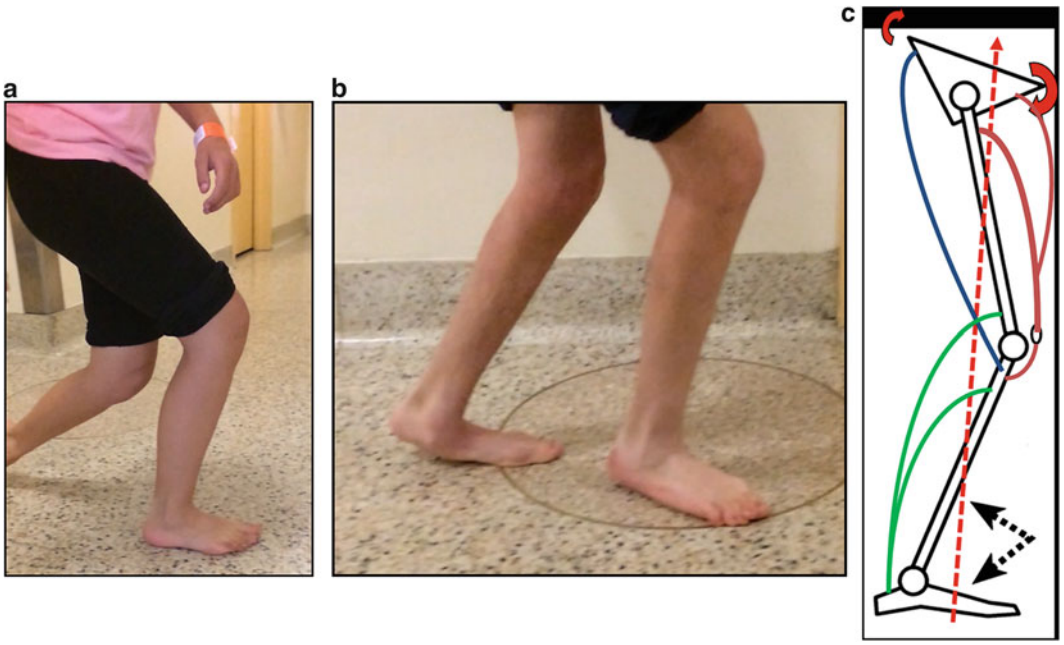


Fig. 17.15 (**a**) Crouch gait pattern: Flexed knee with forward translation of the tibia over the dorsiflexed ankle. (**b**) Crouch gait pattern in the presence of equinovalgus foot. (**c**) When in crouch the ground reaction force passes in front of the ankle, behind the knee and in front of the hip, resulting in ankle plantar flexion, knee extension, and hip extension (internal) moments to counteract the abnormal external moments generated by the ground reaction force

In *apparent equinus* pattern, the child's heels do not come in contact with the ground even though the ankle is not in equinus (Fig. 17.14b). This happens when there is a fixed flexion deformity of the knee. If there was an adequate range of ankle dorsiflexion this fixed knee flexion deformity can be accommodated to keep the center of gravity over the base of support, which is what one finds in the crouch gait pattern. If, however, the ankle has only sufficient range to come to just neutral dorsiflexion, keeping the foot flat on the ground would cause the child's center of gravity to fall well behind the base of support in the presence of an uncorrectable flexed knee. To stop from falling backward the body is forced to shift the center of gravity forward by flexing the hip and increasing forward trunk lean or tipping forward on tiptoe. This pattern is characterized by problems with all five priorities of gait.

Crouch gait is characterized by flexed knee during stance phase accompanied by dorsiflexed foot segment relative to tibial shank. This dorsiflexion can occur through the ankle (true calcaneus) when there is unrestricted range of dorsiflexion of the ankle, which could be iatrogenic following prior over-lengthening and weakening of the gastrocsoleus from tendo-Achilles lengthening (Fig. 17.15a). However, crouch pattern is often present even in the presence of equinus of the hindfoot, when the equinus is accompanied by increased hindfoot valgus (Fig. 17.15b). In this position, the subtalar joint collapses and "dorsiflexion" occurs through the subtalar joint accompanied by midfoot break and a rocker bottom-like foot. In either situation the tibial shank is flexed forward over the foot segment throughout stance. This shifts the ground reaction force well behind the knee, creating an external flexion moment that has to be neutralized by a corresponding internal extension moment by the activation of the quadriceps. In order to keep the center of gravity over the base of the support, the hips must flex (Fig. 17.15c). Crouch pattern seems to be the consequence of the natural history in children with more severe bilateral CP (GMFCS III or greater). All five priorities of gait are affected, and the energy consumption associated with this pattern is substantial. Weight bearing in the crouch position increases the joint reaction force on the patella-femoral joint which can cause anterior knee pain, patella alta, and avulsion fractures of the inferior pole of the patella.

Each of the above patterns can be associated with a *stiff knee* pattern. Stiff knee gait is characterized by limited flexion in initial swing due to tightness of the rectus femoris, which affects clearance of the foot in swing phase. Consequently the feet might drag, slowing the gait, causing tripping and increased shoe wear.

Any of the patterns can also be associated with transverse plane abnormalities that can arise from one or more of the following levels: the forefoot (adductus or abductus), hindfoot (varus or valgus), tibial torsion (internal or external), femoral anteversion (or torsion), hip rotation (internal or external), or pelvic rotation (protraction or retraction). The foot position can be quantified by the foot progression angle



Fig. 17.16 Internal rotation gait pattern

(FPA), normally, an average of 5 degrees external [41]. The knee progression angle should also be assessed as this will help determine the source of rotational abnormality. A normal FPA does not rule out torsional malalignment. In children with bilateral CP it is not unusual to have increased internal rotation of the hips due to increased femoral anteversion characterized by internal knee progression angle (patella points medially) and apparent "scissoring" (Fig. 17.16). Foot progression angle may be internal, normal, or even external if accompanied by increased external tibial torsion and planovalgus abducted foot. Internal rotation of the hip in the face of external rotation of the tibial segment creates a pseudovalgus appearance at the knee (miserable malalignment).

In unilateral CP (hemiplegia) four types of gait have been described [38, 39] (Fig. 17.17). *Type 1* pattern is characterized by equinus in swing phase only. This is a drop foot pattern due to weakness of the tibialis anterior which is unable to adequately dorsiflex the ankle during swing phase. Initial contact might be with the forefoot but the heel comes in contact with the ground early in stance with adequate dorsiflexion in midstance. *Type 2* pattern is characterized by equinus both in swing and stance phases with a toe-toe or toe-heel pattern in stance phase. This is due to a contracture of the gastrocsoleus complex which might be dynamic due to spasticity or fixed due to static contracture. In *Type 3* pattern, the knee is involved as well, showing abnormal flexion (jump) or stiff knee pattern. Finally, in *Type 4* there is additional proximal involvement at the hip, including varying combinations of hip adduction, flexion, and internal rotation. This pattern can be particularly troublesome as it is often accompanied by progressive hip displacement and dysplasia.

These gait patterns are recognizable, but the distinctions are not always clear with many permutations and combinations involving one or both lower extremities.

On-Table Physical Examination

The static examination (on the table) includes assessment of muscle tone and length, bony alignment, muscle strength, and selective control.

Assessment of Muscle Tone and Length

Muscle tone tested is assessed for each muscle group by testing the relevant passive joint range of motion affected by that muscle group. Range of motion is assessed passively with a fast stretch that elicits a catch at an angle (R1) often well before the maximal passive range obtained by a slow, sustained stretch (R2) [42, 43]. This is a feature of spasticity, which is velocity-dependent hypertonia. The larger gap between the R1 and R2 represents the dynamic component of the contracture, and is more amenable to spasticity management. R2 represents the functional muscle length. AS R2 approaches R1, there is increasing static contracture or true shortening of the muscle tendon unit (in addition to tightness). Often, biarticular muscles are more involved than muscles that cross only one joint. It is therefore important to separately test the length of both groups of muscles (when possible) that contribute to range in order to intervene selectively on the biarticular muscles.

The following specific joints (muscle groups) of the lower extremity are tested in the supine position. Hip flexors are tested by extending the hip (Thomas test) while with the opposite hip is held sufficiently flexed to neutralize the anterior pelvic tilt and, but not overcorrect, the lumbar lordosis. Hip abduction is tested first with the knees flexed to test the adductor muscle length. Abduction is then tested with the hips and knees held in extension to assess the additional contribution of the gracilis which is a hip adductor and a knee flexor. The Galeazzi sign assesses the relative lengths of the thigh segment. A shorter thigh segment in combination with an adduction contracture might be indicative of hip subluxation on that side. Knee extension is quantified to rule out a knee flexion contracture. This could be due to one or more of the following: shortened hamstrings, shifted hamstrings due to increased anterior pelvic tilt, shortened proximal

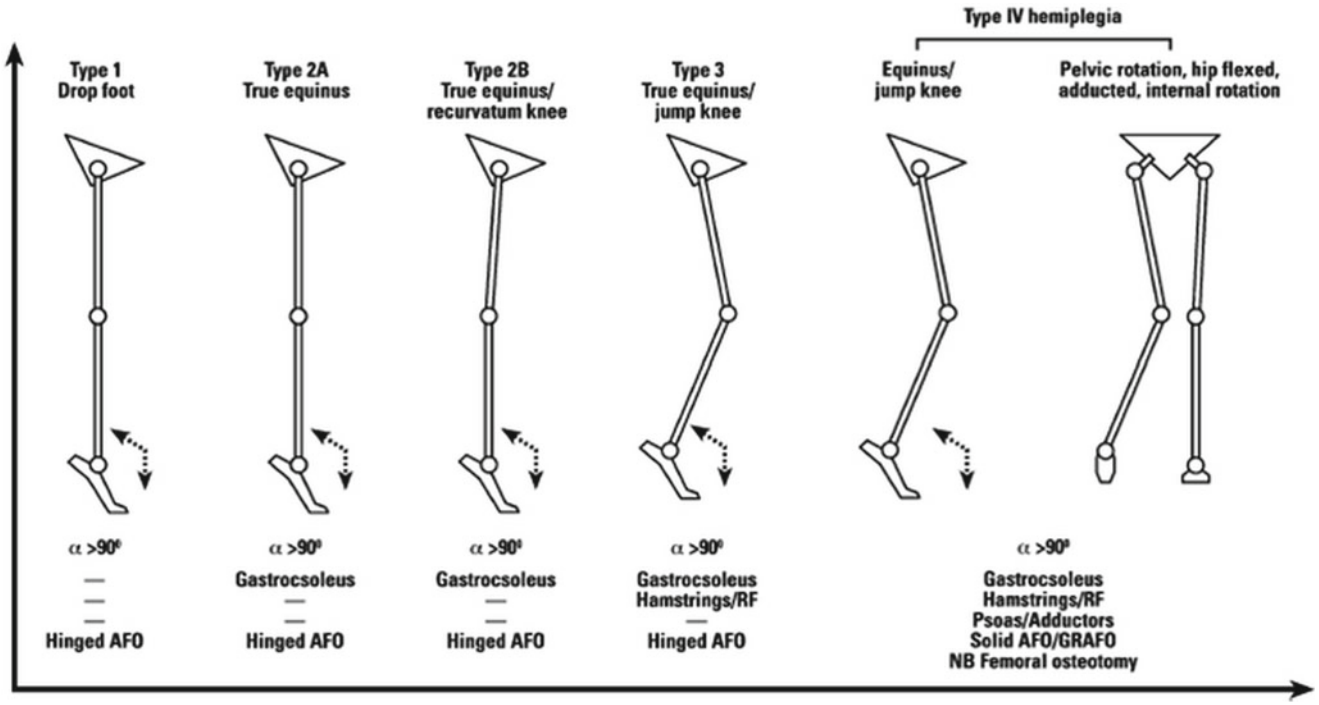


Fig. 17.17 Common gait patterns in unilateral (hemiplegic) cerebral palsy

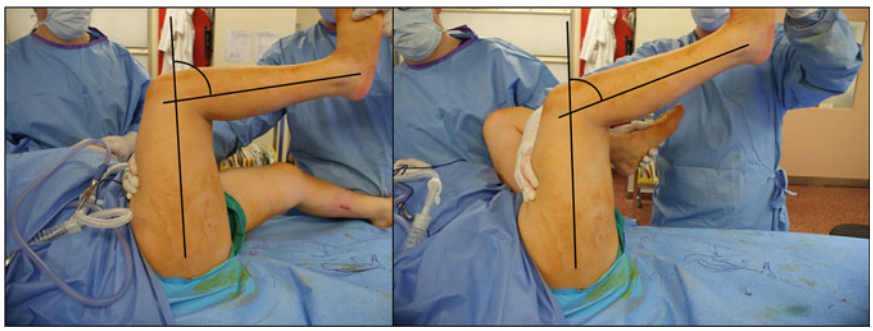


Fig. 17.18 Unilateral and bilateral popliteal angles: The hamstring shift test

gastrocnemius, or a posterior contracture of the knee joint capsule. The popliteal angle tests the functional length of the hamstrings. This is first done unilaterally (one leg at a time) with the hip on the tested side held in 90° of flexion, while the contralateral hip is held in full extension. The bilateral popliteal angle or the hamstring shift test requires testing the popliteal angle with the contralateral hip and knee flexed to neutralize the pelvic tilt similar to the Thomas test [27]. This is a measure of the "true" hamstring contracture (Fig. 17.18). A large difference (reduction) in the popliteal angle (increased available hamstring length) suggests that anterior pelvic tilt might be responsible for the relative elevation of the ischium which is the origin of the medial hamstrings and the long head of the biceps femoris. Ankle dorsiflexion is tested both with the knee flexed and the knee extended (Silfverskiold test) to distinguish contractures arising from the gastrocnemius alone from those where the soleus is involved as well. At the time of testing ankle dorsiflexion, any hindfoot valgus must be corrected to neutral by supinating the foot to ensure that the subtalar joint is "locked" to ensure that dorsiflexion is occurring through the ankle joint rather than spuriously through the subtalar joint (see Fig. 17.9). Hindfoot is tested for its flexibility to correct any valgus or varus. If a cavovarus foot deformity is present, simulate a Coleman block test, as described earlier, to identify whether the hindfoot varus is fixed or not and if not confirm that it is secondary to the plantarflexed deformity of the first metatarsal ray or a primary problem [29]. In a cavovarus foot, the hindfoot might be in equinus, neutral, or even relative calcaneus with the forefoot plantarflexed relative to the hindfoot.

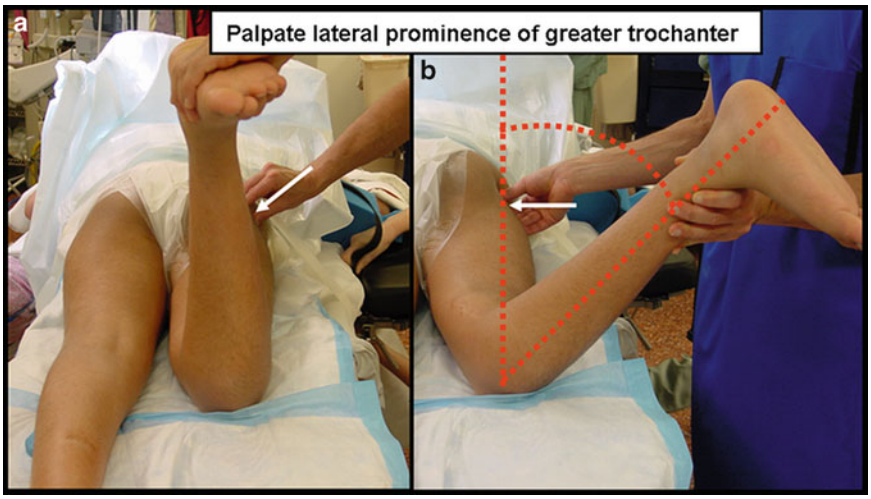


Fig. 17.19 Femoral anteversion with trochanteric palpation test

In the prone position, knee flexion is tested rapidly and then slowly for the Duncan–Ely (DE) sign. A positive sign would be associated with a significant elevation of the buttock off the bed, due to an increase in hip flexion and anterior pelvic treat created when the short and/or spastic rectus femoris is being stretched and pulls down on its origin at the anterior inferior iliac spine causing the hip to flex and pelvis to tilt anteriorly.

Assessment of Bone Alignment

Assessment of the bony alignment includes the examination of the torsional profile of the lower limbs in the prone position to evaluate for excessive femoral anteversion and tibial torsion. The magnitude of femoral anteversion can be estimated using the trochanteric palpation test. In the prone position, the knee is flexed to 90° and held upright or perpendicular to the examination table. In this baseline position, the patella is pointing directly "anterior" into the table. The prominence of the greater trochanter is palpated in the proximal lateral thigh. In this baseline position, the maximum prominence of the greater trochanter sits more posteriorly. The lower leg (tibia) serves as a goniometer. The lower leg is rotated outward which corresponds to internal rotation of the hip, until the prominence of the greater trochanter is palpated most laterally, which corresponds to the femoral neck (axis) being horizontal and parallel to the floor. The number of degrees of outward rotation of the tibia relative to the vertical position at baseline represents the femoral anteversion on that side [44, 45] (Fig. 17.19). Tibial torsional alignment is based on thigh foot angle (TFA) and the transmalleolar (bimalleolar) axis (TMA). With the ankle held in neutral dorsiflexion, the TFA is the angle between the long axis of the foot (heel bisector to second toe) and the thigh axis, which is usually between 10 and 20 degrees external [41]. If the lateral border of the foot is straight, one can infer that there is no deformity arising from the forefoot and that an abnormal TFA is arising from the torsional alignment of the tibia and fibula. This is best quantified by the TMA which is the angle between the trans(bi)malleolar axis and the transverse knee axis, which is usually between 10 and 30 degrees external. TMA less than or greater than this range represent internal and external tibial torsion, respectively [46] (Fig. 17.20).

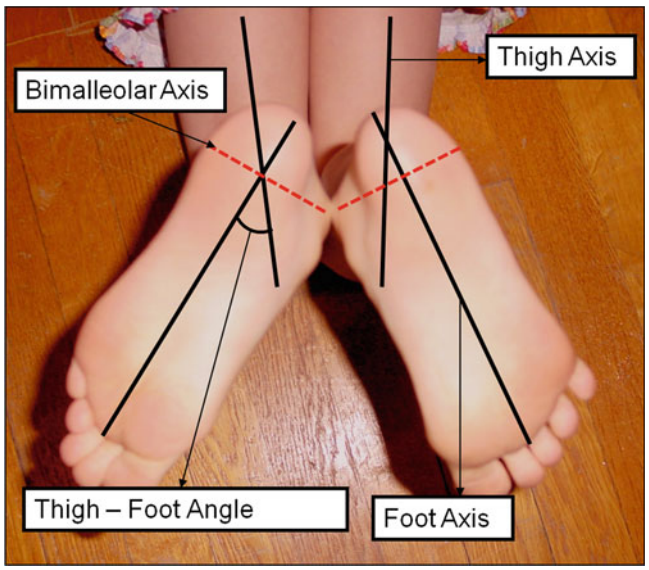


Fig. 17.20 Assessment of tibial torsional profile: The TFA and TMA

Assessment of Muscle Strength and Selective Control

In children with CP, there is increasing evidence that muscles are weak, and that motor function is directly associated with muscle strength [47]. It is therefore important, whenever possible, to assess muscle strength for prognostication and clinical decision making [48]. Muscle strength is tested for each of the lower limb muscle groups using manual muscle testing (MMT) [49]. These tests are somewhat subjective and reliant on the examiner's experience, and are only possible in older

children who are capable of cooperating with the manual testing. In the gait laboratory and in research settings, isometric muscle strength testing using dynamometers or isokinetic evaluations has some value.

Selective motor control is impaired in children with CP and contributes to ambulatory function and patterns [50, 51]. The ability to isolate movements can be graded into three levels of control: (0) no ability; (1) partial ability; and (2) normal ability to isolate movements [52]. These are tested and reported along with the strength for each muscle group tested.

Lower Limb Assessment in the Non-ambulant Child (GMFCS Levels IV–V)

In the non-ambulant child the physical examination should include an assessment of the child seated in the wheelchair or seating system, and assessment lying on the table. In the chair, specific attention is paid to the presence, flexibility, and impact of any spinal deformity in the coronal (scoliosis) and sagittal (kyphosis/lordosis) planes, and pelvic inclination on sitting balance, ischial weight bearing, skin integrity, and hygiene. Particular areas that are vulnerable to skin breakdown are over bony prominences including those due to kypho-scoliosis, sacrococcygeal area, ischial weight-bearing area, and prominences of the greater trochanter or dislocated femoral heads in the lateral proximal thigh. Additionally, in the presence of a large, rigid scoliosis there can be skin breakdown on the side of the concavity where the inferior aspect of the rib cage might rest against the iliac crest and skin crease in the flank might be hard to reach or maintain hygiene due to stiffness of the deformity.

The lower extremity examination of the non-ambulant child is done in the supine position. The examination of the hip joint, knee joint, and foot and ankle is identical to that described for ambulant children. Usually, there is little value in turning the child prone to assess the torsional profile, as these can be reasonably assessed supine.

Goals of Treatment in Cerebral Palsy by GMFCS Level

For ambulatory children (GMFCS levels I–III) the primary goals of treatment are to optimize gait efficiency in order to preserve or improve independence and physical function that will permit increased participation in real-world life activities. Functional problems of gait impact walking distance, speed, balance and stability, endurance, and fatigue, and may lead to increasing discomfort. Improving the appearance of gait and minimizing the reliance on walking aids and orthotics are also identified as important priorities by children with CP and their parents [53].

Box 17.4. Goals of Treatment: Ambulatory CP (GMFCS Levels I–III)

1. Optimize gait efficiency (correct biomechanics) in order to optimize energy conservation
   (a) Preserve or improve physical function
      - Walk longer distance
      - Walk faster
      - Decrease fatigue
      - Better stability: Reduced tripping and fewer falls
      - Keep up with friends
   (b) Pain relief or pain prevention and increased endurance
   (c) Preserve or increase activities and participation
      - More physically active
      - More independence
      - More participation in sports/recreational activity
2. Improve appearance of gait
   (a) Reduced reliance on walking aids
   (b) Reduced use of orthotics
   (c) Feet flat on the ground
   (d) Feet pointing forward
   (e) Reduced dragging of feet
   (f) Stand and walk taller (knees less bent)
   (g) More symmetry

Goals of treatment for nonambulatory children (GMFCS IV and V) are completely different from those for ambulatory children. These more severely involved children are at high risk of developing joint contractures, progressive hip displacement, and spinal deformity that interfere with comfort and caregiving, positioning, transfers, mobility, and seating. The musculoskeletal involvement occurs in the context of other associated comorbidities including seizure disorders, visual and hearing impairments, cognitive delays, swallowing difficulties, drooling, aspiration and pneumonias, gastroesophageal reflux, malnutrition, constipation, and incontinence. The goals of treatment are to prevent or relieve pain, facilitate caregiving, improve health, and optimize the quality of life [54].

Box 17.5. Goals of Treatment: Nonambulatory CP (GMFCS Levels IV–V)

1. Relieve or prevent pain and discomfort
2. Facilitate ease of care
   (a) Dressing, toileting, bathing/hygiene
   (b) Positioning: Seating and lying down
   (c) Transfers and mobility
3. Preserve or improve health
4. Improve quality of life

Management of Lower Extremity Problems: Principles of Treatment and Techniques

In general, the principles of treatment include (1) prevention or slowing the development of progressive joint contractures, bony deformities, and joint instability in the younger child, and (2) correction of these clinically significant bony deformities and joint instability when these have already occurred. These objectives are best achieved using a multidisciplinary approach.

Early intervention focuses on dealing with dynamic contractures in order to delay the need for more extensive surgical procedures (Fig. 17.21). Multiple strategies are used including physical therapy to stretch muscles, orthotics (braces) to maintain stretch or stabilize joints, and serial casting in an attempt to simulate the stretch that would normally arise from typical physical activity. Stretch is facilitated by local or systemic tone management by pharmacologic (botulinum toxin-A, phenol injections) or neurosurgical methods (selective dorsal rhizotomy, intrathecal baclofen). Effective tone management improves range of motion and tolerance of brace wear, and might prevent or delay the onset of static contractures and bony deformities [55–60]. Severe spasticity involving bilateral lower extremities in children with good balance and muscle strength might benefit from more extensive spasticity control from selective dorsal rhizotomy (SDR), which has been shown in randomized trials to produce significant reduction in muscle tone, but only modest benefits on physical function [61–64]. SDR does not necessarily reduce the need for subsequent orthopedic surgery [65–67]. In a systematic review, the functional effects and benefits of SDR in the long term remain uncertain, despite appearing promising in small series of selected patients [68–70]. Baclofen is a GABA agonist that can reduce hypertonia. The relatively large oral doses required for any meaningful effect can be associated with undesirable side effects. When administered intrathecally, very small doses (micrograms) are required to reduce severe generalized hypertonia due to spasticity or dystonia. Intrathecal baclofen administered continuously by an implantable pump is usually reserved for more severely involved children (GMFCS level V), as it can be associated with significant complications [71–73].

Established deformities are most efficiently addressed with single-event multilevel orthopedic surgery (SEMLS) using a combination of fractional muscle or intramuscular or tendon lengthening, tendon transfers, and corrective osteotomies and joint stabilization procedures to address bony deformities and joint instability that contribute to lever arm dysfunction [74, 75]. Surgery is delayed until the gait patterns have matured and any functional gains from the natural history have plateaued [76]. The optimal timing is sometime between 7 and 11 years of age, to reduce the likelihood of recurrence of deformity with further growth necessitating repeat surgery (see Fig. 17.21). In the short term, SEMLS is associated with substantial improvements in gait quality, with more modest corresponding functional improvements [77–79]. In general, the children with more abnormal gait patterns stand the most to gain, but about 25 % show little measurable benefit. It is unclear whether these patients would have deteriorated further without the intervention [80]. There is very little information in the literature about the long-term effects of multilevel orthopedic surgery at skeletal maturity or adulthood [77, 81–83].

Table 17.1 summarizes the most common surgical procedures for cerebral palsy and their indications.

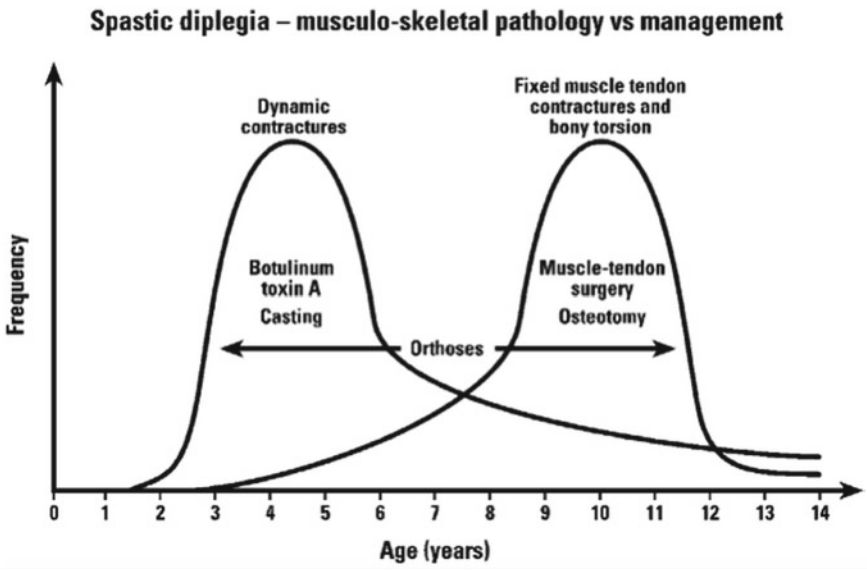


Fig. 17.21 Management strategies and timing principles. Reprinted from Current Orthopedics, 17, Bache CE SP, Graham HK, The management of spastic diplegia, 88-104, Copyright 2003, with permission from Elsevier

Table 17.1 Common soft-tissue and bony procedures performed during multilevel surgery for cerebral palsy

	Indications for surgery	Procedure
Hip flexion contracture: Psoas	*Ambulatory Child* • Reduced hip extension in terminal stance • Excessive anterior pelvic tilt + lordosis • Hip flexion contracture >20° • Hip subluxation • Kinematic: Excessive anterior pelvic tilt with double-bump pattern in diplegia; single-bump pattern in hemiplegia • Kinetics: Delay in transition from hip extensor (internal) moment to hip flexor (internal) moment (beyond the typical 25 % of gait cycle) *Nonambulatory Child* • As part of the management of hip displacement	*Ambulatory Child* Selective intramuscular lengthening of the psoas tendon over the brim of the pelvis (spare the iliacus) Note: Proximal femoral derotational osteotomy done above the lesser trochanter can functionally lengthen the psoas *Nonambulatory Child* Iliopsoas tenotomy or release of the lesser trochanter
Hip adduction contracture: adductors Note: During normal gait the arc of hip motion in the coronal plane is from 10° of abduction to 10° of adduction. 15° of abduction is sufficient for normal gait	*Ambulatory Child* • Decreased abduction in swing phase (scissoring) • Adduction contracture: <15° *Nonambulatory Child* • Adduction contracture interfering with care: dressing, toileting, perineal hygiene • As part of the management of hip displacement	*Ambulatory Child* Percutaneous or open release of adductor longus alone is usually sufficient *Nonambulatory Child* Open release of adductor longus, adductor brevis, gracilis + iliopsoas tenotomy
Hip internal rotation: Anteversion	*Ambulatory Child* • Excessive internal rotation of the hip due to increased femoral anteversion • Increased "scissoring" with knees rubbing • Internal foot progression angle—causing tripping *Nonambulatory Child* • As part of the management of hip displacement	*Ambulatory Child* Proximal femoral (intertrochanteric) derotational osteotomy with internal fixation Alternative: Distal femoral derotational osteotomy *Nonambulatory Child* Combined proximal femoral derotation + varus osteotomy with stable internal fixation
Proximal femur: Coxa valga and hip subluxation	• Increased neck-shaft angle • Migration percentage >30 % • Break in Shenton's arc • Increased anteversion	Proximal femoral (intertrochanteric) varus and external derotational osteotomy (VDRO)
Pelvis: Acetabular dysplasia	• Acetabular index >30° • Subluxated hip • Older than 5 years	Periacetabular pelvic osteotomy (e.g., Dega) to restore posterolateral coverage
Knee flexion: Hamstrings	*Ambulatory Child* • Decreased knee extension in terminal swing • Increased popliteal angle, provided that – Bilateral popliteal angle >30° • Kinematics: – Posterior pelvic tilt – Decreased knee extension in terminal swing *Nonambulatory Child* • Knee flexion contractures due to hamstring contracture interfering with care, dressing, seating, and positioning. Goal is to decrease contracture (and popliteal angle) and not to straighten the knee entirely	*Ambulatory Child* Medial hamstring lengthening: Intramuscular tenotomies of the gracilis, semitendinosus; aponeurotic lengthening (striping) of semimembranosus Lateral hamstring lengthening: Fascial striping of the biceps femoris aponeurosis. This is avoided unless there is posterior pelvic tilt present to prevent excessive weakness of hip extensors. *Nonambulatory Child* Percutaneous or open hamstring lengthening: Medial and lateral
Knee flexion contracture	• Knee flexion contractures >20° despite hamstring lengthening • Hamstring length is sufficient (normal) • Older children (closer to skeletal maturity): >12 years • Significant crouch pattern in gait (GMFCS II–III) or during standing for transfers (GMFCS IV)	Distal femoral extension osteotomy: This must be accompanied by patella tendon shortening or patellar tendon distal advancement to re-tension the elongated knee extensors following the extension osteotomy

(continued)

Table 17.1 (continued)

	Indications for surgery	Procedure
Stiff knee: Rectus femoris	• Stiff knee pattern: Limited flexion at initial swing • Foot drag resulting in poor clearance • Positive Duncan–Ely sign • Kinematic: Peak knee flexion in initial swing is decreased in amplitude and delayed • EMG: RF active throughout gait cycle • Avoid in more severe GMFCS III and IV	Rectus femoris tendon is separated from the underlying vasti and the detached tendon is tenodesed to the semitendinosus, gracilis, or sartorius
Tibial torsion: External	• External torsion: Bimalleolar axis >30° external – Out-toeing: External foot progression angle >30° – Foot drag – With planovalgus/abducted foot – Relatively normal foot progression angle in the presence of increased femoral anteversion that is being corrected by external derotation of the femur	Distal tibial internal derotational osteotomy to correct bimalleolar axis to 10° external (concomitant fibular osteotomy may not be needed for derotations of <30°)
Tibial torsion: Internal	• Internal torsion: Bimalleolar axis >10° internal – Intoeing: Internal foot progression angle >10° internal – Tripping	Distal tibial external derotational osteotomy to correct bimalleolar axis to 10° external (concomitant fibular osteotomy may not be needed for derotations of <30°)
Equinus: Gastrocnemius-soleus	*Ambulatory Child* • Equinus in stance phase: Poor stability • Equinus in swing: Poor clearance • Tripping and falling • Increased shoe wear *Nonambulatory Child* • Contracture interfering with bracing or footwear necessary for weight bearing for transfers (GMFCS IV) • Contracture interfering with bracing or footwear, or resting of feet on the footplate of wheelchair (GMFCS V) Note: These apply to any foot deformity in nonambulatory children	Gastrocnemius contracture alone (Silfverskiold): Isolated lengthening of the gastrocnemius (Strayer technique) Gastrocnemius > soleus contracture (Silfverskiold test): Strayer + soleus fascial lengthening by striping the aponeurosis leaving muscle intact or gastrocsoleus aponeurotic lengthening (e.g., Baker, Vulpius)
Equinus: Tendo-Achilles	• Fixed equinus contracture involving both gastrocnemius and soleus, seen commonly in hemiplegia	Tendo-Achilles lengthening is the most aggressive method of lengthening the gastrocsoleus complex. This can be done by 2 or 3 hemitenotomy sliding lengthening technique or open z-lengthening
Equino (plano)valgus	• Increased hindfoot valgus with collapse of subtalar joint (rocker bottom) and abducted forefoot – Poor push off (tendency to crouch) – Pain with weight bearing – Blistering or callosity over medial prominence of talar head – Loss of flexibility: Unbraceable	Lateral column lengthening through the calcaneus, along with plantar flexion osteotomy of first ray. May need lengthening of peroneus brevis; lengthening of gastrocsoleus as needed to address the unmasked equinus after correction of hindfoot valgus Triple C osteotomy: Medial slide osteotomy of calcaneus; opening wedge osteotomy of cuboid: closing wedge osteotomy of medial cuneiform
Equino (cavo) varus	• To prevent or correct weight bearing on lateral border of the foot which leads to – Poor stability in stance – Pain and callosity – Difficulty with brace and shoe wear – Stress fracture of the fourth or fifth metatarsal – Poor clearance in swing • Flexible hindfoot varus is amenable to tendon transfers • Fixed contractures of tibialis posterior require lengthening • Hindfoot varus driven by the presence of plantar flexion of the first ray (confirmed by the Coleman block test)	Split tibialis anterior tendon transfer to the cuboid if hindfoot varus is flexible and occurs primarily in swing phase Split tibialis posterior tendon transfer to the peroneus brevis when the hindfoot varus is flexible but occurs in stance phase as well Requires lengthening of tibialis posterior: Intramuscular tenotomy. Tibialis posterior tendon lengthening may be combined with the split tibialis anterior tendon transfer Plantar fascia release along with extension osteotomy of the base of first metatarsal or medial cuneiform; consider transfer of the peroneus longus to the peroneus brevis. These may be combined with tibialis posterior tendon lengthening All of these may require gastrocsoleus or tendo-Achilles lengthening as needed to address the equinus contracture

Orthopedic Procedures at the Hip

Iliopsoas Lengthening

The indication for psoas lengthening in ambulatory children is controversial [14, 84]. In the presence of a hip flexion contracture, accompanied by increased anterior pelvic tilt resulting in reduced hip extension in terminal stance, selective intramuscular lengthening of the psoas (sparing the iliacus) can be done over the brim of the pelvis [85]. In nonambulatory children with hip instability, iliopsoas tenotomy is performed at the level of insertion into the lesser trochanter.

Adductor Releases

In ambulatory children adductor lengthening is only necessary if the adduction contracture is severe. During normal gait the coronal plane arc of motion of the hip is limited from 10° of abduction to 10° of adduction. Abduction of 15° with the hip and knee in extension is compatible with normal gait [86]. In the ambulatory child with CP, adductor surgery is usually restricted to percutaneous or open release of adductor longus alone. A proximal tenotomy of gracilis may be added if the range of hip abduction is further reduced when examined with the knee in maximal extension. More extensive adductor releases to include the adductor brevis are seldom indicated in ambulant children unless there is a more severe adduction contracture interfering associated with increased hip displacement [86]. In a non-ambulant child, adductor releases help to relieve adduction contractures that can interfere with care, dressing, and perineal hygiene, and are routinely lengthened when the contracture is associated with increased hip displacement either in isolation or along with proximal femoral osteotomies to center the hip. The adductor longus, adductor brevis, and gracilis are generally released from their origins through a medial transverse groin incision just distal to the groin crease, centered over the adductor longus tendon. The same approach allows access to the iliopsoas tendon insertion at the lesser trochanter. Adductor releases alone are insufficient to stabilize hip subluxation and more than 75 % of the time for GMFSC levels IV and V children, subsequent bony reconstructive surgery is required [87].

Femoral Derotation and Varus Derotational Osteotomy

When there is a clinically significant internal rotation gait due to increased femoral anteversion, the excessive anteversion can be corrected by derotational osteotomy proximally (inter-trochanteric) or distally [88, 89]. The proximal osteotomy allows for the addition of varusization when there is true coxa valga and some subluxation [90] (Fig. 17.22a). The osteotomy is proximal to the insertion of the iliopsoas on the lesser trochanter, resulting in some functional lengthening of the psoas that occurs with external rotation of the lesser trochanter. The proximal osteotomy can be done supine or prone, the latter having the advantage of providing reliable (bilateral) intraoperative assessment of the torsional profile. Internal fixation with a blade plate or locking plate often provides sufficient stability to avoid postoperative casting [91] (Fig. 17.22b, c).

Windswept deformities are not uncommon in more severely involved nonambulatory children. These hips require an approach that addresses the asymmetric hip deformity. The adducted and internally rotated side requires releases of the adductors, flexors, and hamstrings along with an external derotational osteotomy of the femur. On the abducted and externally rotated side, the abductors, iliotibial band, and lateral hamstrings may have to be released, and derotational osteotomies of the femur are based on whether and by how much the femur is anteverted. The "abducted" side may just be less adducted and require similar but less extensive procedures than on the more adducted side.

Periacetabular Pelvic Osteotomy

In the presence of hip subluxation, acetabular dysplasia develops secondarily due to the mechanical pressure of the subluxating femoral head. The acetabular deficiency is usually posterolateral. Following proximal femoral varus derotational osteotomy to redirect the femoral head into the center of the acetabulum, posterolateral coverage is improved with a Dega-type periacetabular osteotomy or one of its adaptations, which comprises a lateral opening wedge osteotomy through the outer table, hinging on or just proximal to

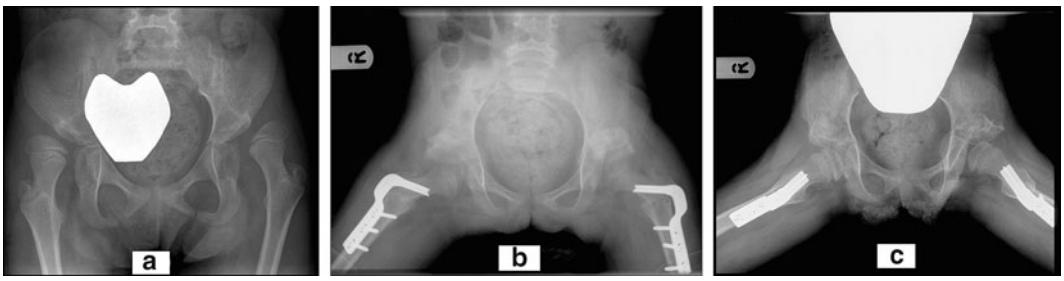


Fig. 17.22 (**a**) An 8-year-old girl with bilateral hip subluxation, coxa valga, and acetabular dysplasia. (**b**) AP radiographs after bilateral proximal femoral varus derotational osteotomy with blade plate fixation, and periacetabular pelvic osteotomy (Dega). (**c**) Frog-leg lateral radiograph after bilateral proximal femoral varus derotational osteotomy with blade plate fixation, and periacetabular pelvic osteotomy (Dega)

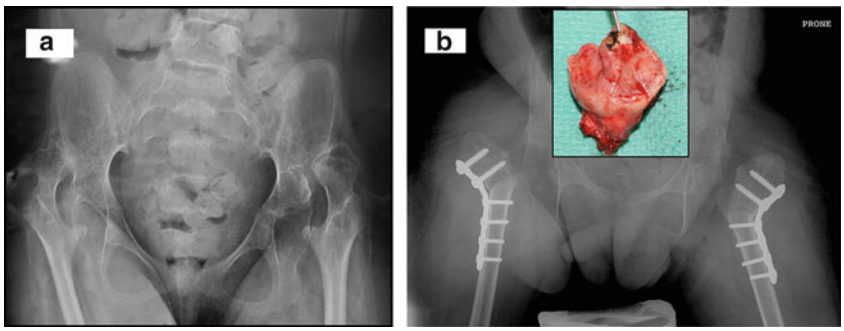


Fig. 17.23 (**a**) Deformation of the dislocated femoral head with loss of sphericity and flattening of superolateral surface. (**b**) Femoral head resection, subtrochanteric valgus osteotomy of the proximal femur

the tri-radiate cartilage [92, 93]. Bone graft is derived from the proximal femoral varus osteotomy and the anterior iliac crest (Fig. 17.22b, c).

Femoral Head Resection and Related Procedures

Salvage procedures are indicated for long-standing hip dislocations that are painful but not reconstructible because of the advanced changes associated with the long-standing dislocation including loss of sphericity of the femoral head and severe acetabular dysplasia. The femoral head typically deforms along its exposed superolateral surface due to the pressure from the overlying abductors (Fig. 17.23a). In general, salvage procedures are not as effective as reconstructive procedures to achieve the goals of pain relief. Proximal femoral head resection at the base of the neck, along with a subtrochanteric valgus osteotomy of the femur, tenodesis of the ligamentum teres to the lesser trochanter, and capsular closure, in addition to the adductor and psoas lengthening is an effective strategy to provide sufficient abduction for perineal hygiene and pain relief from the previous dislocation but not without a significant period of recovery [94] (Fig. 17.23b). Heterotopic ossification or proximal migration is uncommon with this procedure but occurs more frequently with other salvage procedures that involve more extensive proximal femoral resections.

Orthopedic Procedures at the Knee and Lower Leg Segment (Tibia)

Hamstring Lengthening

In ambulant children, just the medial hamstrings are typically lengthened [86, 95]. Using a posteromedial approach (patient supine) or posterior midline approach (patient prone), intramuscular tenotomies of the gracilis and semitendinosus and fractional lengthening of semimembranosus aponeurosis can be performed. Lateral hamstring (biceps femoris) lengthening is generally avoided in ambulatory children due to the increased risk of knee hyperextension and weak hip extension [96]. Transfer of the tenotomized semitendinosus to the adductor tubercle might preserve hip extension and prevent increased anterior pelvic tilt, but the indications and effectiveness of this procedure remain uncertain [86]. In non-ambulant children, hamstring lengthening can be performed percutaneously. Hamstring lengthening is often performed in conjunction with hip reconstructive surgery, to reduce the tension on the relocated hips and to address knee flexion contractures that might be interfering with positioning, dressing, or caregiving.

Distal Femoral Extension Osteotomy

In the presence of fixed knee flexion contractures (15° to 30°), a distal femoral extension osteotomy might be the only way to achieve "full" extension of the knee acutely [97]. An anteriorly based closing wedge osteotomy is performed through a lateral incision using a subvastus approach to the distal femur stabilized with a fixed-angle plate for internal fixation (Fig. 17.24a–e). The sciatic/peroneal nerves are at risk of stretch neuropraxia with acute corrections (extension) of knee flexion contractures (especially with the hip flexed) intraoperatively or in the immediate postoperative period, which can result in dysesthesias in the lower extremity. Postoperative immobilization in some flexion is recommended for a few days following surgery before the knee is stretched to full extension.

Patellar Tendon Shortening or Advancement

This procedure is often done in combination with the distal femoral extension osteotomy to re-tension the relatively lengthened quadriceps following the femoral osteotomy. The patellar tendon realignment can be done by an infrapatellar shortening procedure of the patella tendon [76]. Alternately, patellar tendon advancement via a distal transfer of the tibial tubercle in the skeletally mature child or the anterior portion of the tibial tubercle apophysis in a skeletally immature child may be performed [98].

Anterior Hemiepiphysiodesis of Distal Femoral Physis

Guided growth with the use of anterior staples or extraperiosteal tension-band plates might gradually correct fixed flexion deformities in children who have sufficient growth remaining. The long-term effectiveness of these procedures remains uncertain [76].

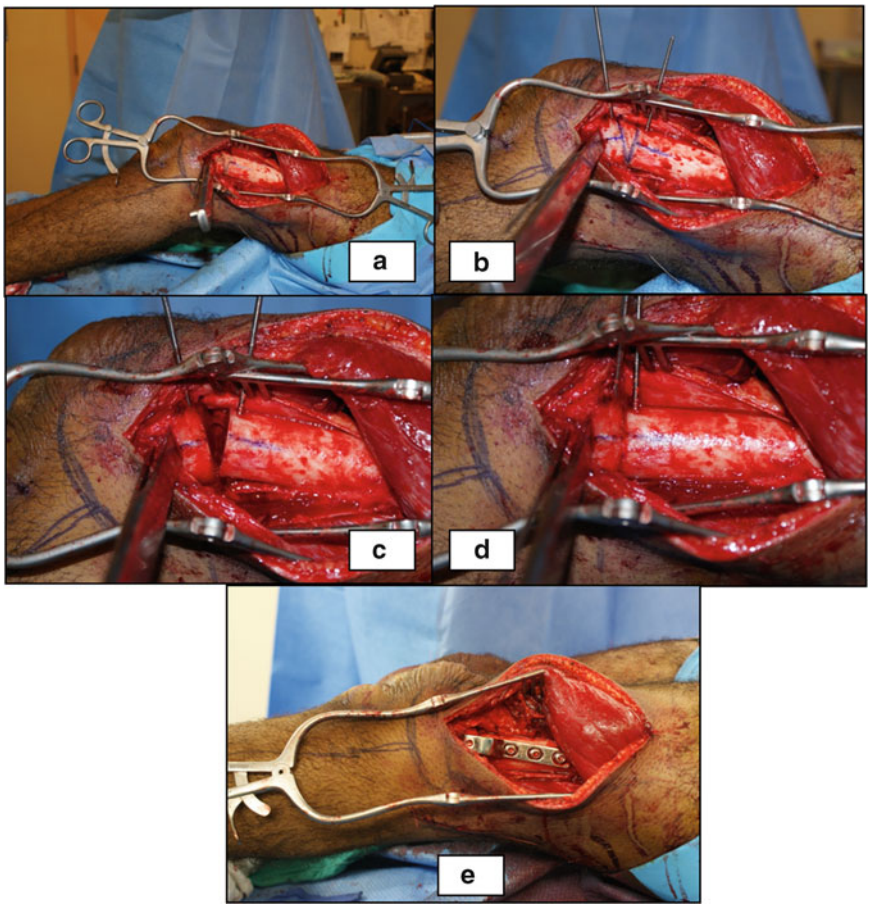


Fig. 17.24 (**a**–**e**) Distal femoral extension osteotomy with blade plate fixation

Rectus Femoris Transfer

Co-spasticity of the rectus femoris and the hamstrings is associated with a stiff knee gait pattern, characterized by decreased knee flexion in early swing phase resulting in poor foot clearance or foot drag [99]. The Duncan–Ely sign is positive on the physical examination. Gait kinematics show decreased amplitude and delay of peak knee flexion in swing. Dynamic EMG shows abnormal activation of the rectus femoris throughout swing. Rectus femoris transfer is indicated to improve knee flexion in early swing in order to improve foot clearance [100]. The rectus femoris tendon is separated from the underlying vasti; the detached tendon is transferred medially over the surface of the vastus medialis and tenodesed to the semitendinosus, gracilis, or sartorius. Early postoperative range of motion (immediate CPM) is important to prevent adhesions. Postoperative rehabilitation including muscle strengthening and retraining exercises is crucial for the success of this procedure. This procedure has been shown to be more effective in higher functioning ambulatory children who are motivated and capable of participating in a rehabilitation program [99, 101, 102].

Distal Tibial (Supramalleolar) Derotational Osteotomies (Fig. 17.25)

Excessive external or internal torsion of the tibia can result in abnormal foot progression angle, associated with foot drag and a poorer lever arm of the foot for push off. Torsional deformities are corrected through distal tibial derotational osteotomies and stabilized with internal fixation (ideally plates/screws) [103]. A concomitant osteotomy of the fibula is usually only necessary when derotation exceeds 30° [104]. No fixation is required for the fibular osteotomy.

Orthopedic Procedures at the Ankle and Foot

Gastrocnemius and Soleus Lengthening

The soleus typically contributes less to equinus than the gastrocnemius in children with bilateral CP, whereas in hemiplegics the soleus is just as involved in the contracture. Based on the relative contributions of both muscles options include selective lengthening of the gastrocnemius alone [105], with complete sparing of the underlying soleus;

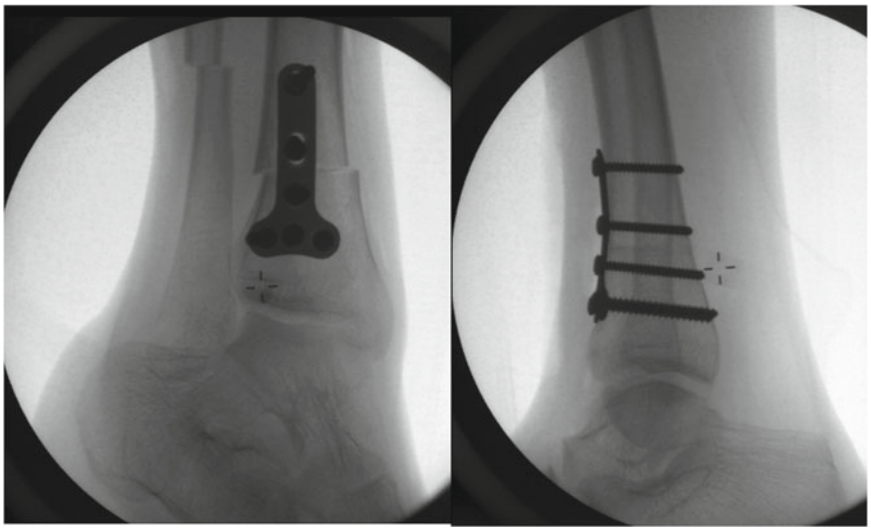


Fig. 17.25 Distal tibial derotational osteotomy

addition of the aponeurotic striping of the underlying soleus; combined gastrocsoleus lengthenings; and tendo-Achilles lengthening. Tendo-Achilles lengthening in bilateral CP has been implicated in development of iatrogenic crouch, and should be avoided unless the contracture is severe and involves both gastrocnemius and the soleus [106–109]. Surgery in children under 8 years is associated with increased risk of recurrence and overcorrection. The gastrocnemius lengthening is performed at the junction of the proximal and mid-calf, at the level of the inferior border of the muscle bellies of the medial and lateral heads of the gastrocnemius, where the aponeurosis of the gastrocnemius is separable from the underlying soleus [105]. In older children with long-standing equinus, there may be additional though milder contracture of the soleus. In these children, it is possible to obtain additional dorsiflexion by dividing the aponeurosis of the soleus immediately deep to the divided gastrocnemius while preserving the underlying muscle fibers of the soleus [76]. In hemiplegics, both gastrocnemius and soleus are usually equally tight and therefore a tendo-Achilles lengthening can be performed by a percutaneous or mini-open sliding technique (Hoke or White) or open z-lengthening [110]. Recurrence of equinus is the most common complication of a tendo-Achilles lengthening in a hemiplegic, and is usually a concern when done in children less than 8 years old.

Tibialis Posterior Tendon Lengthening or Split Tibialis Posterior Tendon Transfer (SPOTT)

Equinovarus deformities are more common in unilateral CP or hemiplegia. Fixed contractures need lengthening of the tibialis posterior tendon. An intramuscular tenotomy of the tibialis posterior at the distal calf is effective and distal z-lengthenings are seldom required. When the contracture is dynamic and the hindfoot is passively correctable to normal, a split tibialis posterior tendon can be transferred to the peroneus brevis [111]. Transfers are more likely to be effective and last longer when done in younger children.

Split Tibialis Anterior Tendon Transfer

If the hindfoot varus deformity occurs primarily in swing phase of gait and sets up the foot to land on the lateral border, a split tibialis anterior tendon transfer to the cuboid can be very effective [112]. A tibialis posterior tendon lengthening can be combined with a split tibialis anterior tendon transfer (SPLATT) and tendo-Achilles lengthening [113]. The goal is to achieve a braceable, plantigrade foot in stance so that weight bearing on the plantar surface rather than along the lateral border and in swing phase prevents difficulties with foot clearance.

Foot Osteotomies

Equinoplanovalgus deformities are more common in bilateral CP. A lateral based opening calcaneal osteotomy just proximal to the anterior process will lengthen the lateral column, correcting the hindfoot valgus and forefoot abductus and restoring the medial arch. Bone graft (iliac crest or allograft) is used to fill the gap at the osteotomy site. Peroneal brevis tendon lengthening is usually necessary as it is tensioned when the foot is swung out of abduction. Gastrocsoleus recessions might be needed to address any accompanying equinus, which will be unmasked when the valgus is corrected [114]. To minimize the chance for recurrent deformity, medial procedures are often added, such as capsulodesis of the talonavicular joint and advancement of the tibialis posterior tendon. Plantar flexion osteotomy of the first ray to restore the normal tripod of the weight-bearing plantigrade foot might be necessary if following the correction of the

hindfoot valgus, and mid/forefoot supination is apparent. An alternative to the lateral column lengthening strategy is the "triple C" method, which includes (1) a closing wedge varus producing osteotomy of the calcaneus along with some medial slide all done from the lateral approach, and (2) an opening wedge osteotomy of the cuboid using a wedge of autologous bone graft obtained from (3) a plantar-based closing wedge osteotomy of the medial cuneiform [115]. More severe or rigid varus or valgus deformities of the hindfoot are best managed by calcaneal osteotomies using closing wedge or sliding osteotomies. Once the hindfoot deformity is corrected, a subtalar (extra-articular) arthrodesis can be performed to correct and hold the deformity permanently [116]. Others recommend a talonavicular arthrodesis to accomplish the same objective.

In the presence of a plantarflexed first ray, plantar fascia release and extension osteotomy of first metatarsal or medial cuneiform will address a midfoot cavus and secondarily eliminate the compensatory hindfoot varus. The peroneus longus tendon is tenodesed to the peroneus brevis to prevent further plantar flexion of the first metatarsal. Hallux valgus deformities are only addressed if painful or are interfering with shoe or brace wear. Correction of the deformity should be accompanied by an arthrodesis of the first metatarsophalangeal joint.

Summary

The lower extremity musculoskeletal consequences of CP are significant both in ambulant and non-ambulant children. The goals for these two groups are quite different (see Boxes 17.4 and 17.5). These goals are best achieved using a multidisciplinary approach, of which orthopedic surgery plays an important role. However, orthopedic surgery deals with the peripheral consequences of CP while the primary pathology in the brain remains unaddressed. There is an imperative for stronger evidence for the effectiveness of many of these interventions based on outcomes that are meaningful to this population [54, 117–119]. The longevity of these outcomes up to and beyond skeletal maturity into adulthood needs to be established [81]. Improved understanding of the biology of brain development and repair and epidemiological research might provide the means to prevent or mitigate many of the etiological causes of CP. Until that time orthopedic surgery will continue to play an important role in the management of the deformities associated with CP.

Myelomeningocele (Spina Bifida)

Defective closure of the neural tube in the first month of embryonic life leads to a spectrum of neural tube defects, which affects about 0.5–1.0/1,000 pregnancies in the USA [120]. The overall incidence has declined with oral folic acid supplementation and termination of pregnancy that is facilitated by maternal prenatal screening with ultrasound and elevated levels of α-fetoprotein and β-human chorionic gonadotrophin levels. The neurologic deficit has both motor and sensory components and the specific musculoskeletal deformities of the lower extremity depend on the level of the lesion and the resulting imbalance between the effects of neurologic deficit present at the level (mixed upper and lower motor neuron) and below the level (lower motor neuron) of the lesion and the relatively unopposed action of the uninvolved neurologic levels above the lesion (Table 17.2). Unlike in cerebral palsy, where the deformities are absent at birth, many of the musculoskeletal deformities associated with spina bifida might be present at birth. However, similar to CP, such deformities usually become worse with growth.

Table 17.2 Musculoskeletal deformities by level of lesion in myelomeningocele[a]

Level	Motor examination	Orthopedic manifestations	Functional manifestations
Thoracic	• Flaccid lower extremities	• Scoliosis (neuromuscular, congenital) • Kyphosis • Possible clubfeet, hip dislocations	• Household ambulation with HKAFO bracing when small • Wheelchair when bigger
Upper lumbar	• Hip flexion/adduction partially intact • Knee flexion/extension, ankle power, hip extension/abduction absent	• Hip dislocation from unbalanced muscles common • Clubfoot deformities	• Ambulation with HKAFO bracing when small • Often wheelchair when bigger
Lower lumbar	• Knee extension intact (L4—quadriceps) • Ankle dorsiflexion possible (L5—tibialis anterior)	• Clubfeet • Calcaneus (L5) • Hip dislocation or subluxation sometimes	• Ambulation with KAFO bracing
Sacral	• Hip flexion/adduction, knee flexion/extension, ankle power present • Hip extension/abduction partially intact • Gastroc and foot weakness	• Cavovarus foot deformity • Hip dislocation much less common • Ulcers may lead to osteomyelitis	• Very good ambulation with AFO bracing

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Hip Dysplasia

Hip dysplasia is most common with higher level lesions (thoracic and high lumbar) and is often present at birth and generally more resistant to reduction than idiopathic developmental hip dysplasia. The management of hip dysplasia in this patient population is controversial. Reconstructive approaches to relocate and stabilize the hips are unnecessary for the non-ambulant children as pain is generally not a feature of hip dislocation in spina bifida. In ambulant children, gait function may be more affected by the presence of contractures than the instability of dislocated hip. Furthermore, complications associated with surgery (decrease in strength and increased fracture risk) may outweigh the benefits of reduction [121].

Knee Flexion Deformity and Tibial Torsion

Functional walking is reliant on the presence of some quadriceps action (present in lower lumbar level lesions or more distal neurologic level). In the presence of knee flexion deformity, the excessive external flexion moment at the knee cannot be adequately counteracted by an extension moment generated by the weak knee extensors. The use of knee-ankle-foot orthoses (KAFO) might facilitate some walking in such children. Fixed flexion deformities are amenable to radical posterior capsular releases which might provide some functional benefits in ambulatory children with low lumbar or sacral lesions who are in crouch [122–124]. There may also be a role for addressing knee flexion contractures in nonambulatory patients with higher level lesions, to facilitate standing. Marked external tibial torsion may contribute to gait-related difficulties in ambulatory children who can benefit from derotational tibial osteotomies.

Ankle and Foot Deformities

A variety of specific foot deformities are associated with neurologic level of the neural tube defect. Some deformities are present at birth (e.g., equinovarus or clubfeet) while others are acquired over time. Common deformities include equinovarus (clubfoot) most common with the higher level lesions; cavovarus is often seen with L-4 lesions; and calcaneus and calcaneovalgus are seen with L-5 lesions. Pure equinus might arise from isolated reflex segments of the cord causing spasticity. A late-onset cavus deformity that is progressive with growth might be associated with tethered cord.

Goals of management in children with myelomeningocele are to maintain a stable, plantigrade foot for weight bearing while minimizing the risk of ulceration of insensate areas of the weight-bearing areas of the foot. Neurogenic equinovarus deformities are stiffer than idiopathic clubfeet, but can still be successfully managed with serial casting using the Ponseti method in some cases, with careful attention paid to the health of the skin during casting. A well-balanced but floppy foot does not need surgery but can be successfully managed with a brace as can a passively correctable deformity. Surgery is indicated for persistent or recurrent deformity in children who can walk. In general, proximal deformities should be corrected before distal ones and soft-tissue releases are done before corrective osteotomies. Equinus is often correctable with a tendo-Achilles tenotomy. Severe equinovarus deformities often require extensive posteromedial releases, which might be facilitated by excision of the talus. There is little role for tendon transfers in this patient population. Recurrent deformities might benefit from talectomy or triple arthrodesis. Cavovarus deformities can be managed by plantar fascia releases, calcaneal osteotomy, and osteotomy of the bases of the metatarsals. Ankle valgus can be managed by supramalleolar osteotomy or distal medial tibial hemiepiphysiodesis with a screw [122, 123].

Poliomyelitis

Poliomyelitis is an acute enteroviral infection affecting the anterior horn cells of the spinal cord and the brain stem motor nuclei. The musculoskeletal deformities associated with polio provided the mainstay of clinical practice of pediatric orthopedic surgeons in the last century. With the advent of widespread vaccination since the 1950s, polio is on the brink of eradication, with a few remaining endemic areas in South Asia, Nigeria, and Afghanistan, where the penetration of vaccination has been suboptimal.

This pure motor condition manifests in three stages. The *acute stage* is characterized by initial gastrointestinal symptoms and fever, followed by meningeal symptoms and an asymmetric paralysis within 48 h, which affects the lower extremity more commonly than the upper extremity, particularly the glutei, hip flexors, quadriceps, tibialis anterior, and medial hamstrings. During the acute phase, management involves bed rest, positioning, and range of motion to avoid contractures. During the *convalescent stage* muscle strength improves, much of it in the first month, but this can last for up to 2 years. During this time, treatment includes range of motion, splinting and bracing (orthoses) to prevent fixed contractures, and strengthening of the affected muscle groups. However, overactivity of muscles in the convalescent phase has the potential to inhibit recovery of function. The *chronic stage* begins 2 years from the onset of the disease, and no further muscle recovery is expected. The paralysis is associated with flaccid joints or active joints with muscle imbalance. Flaccid joints will develop contractures only if

Table 17.3 Common deformities of the lower extremity in poliomyelitis, and their surgical management

Deformity	Surgical management
Quadriceps femoris paralysis	Biceps femoris and semitendinosus transfer to patella
Flexion contracture of the knee	Soft-tissue release or femoral osteotomy
Genu recurvatum	Tibial osteotomy
	Soft-tissue triple tenodesis
Flail knee	Orthosis with drop-lock knee and arthrodesis
Equinus	Achilles tenotomy
Cavus	Plantar fascia release
	Calcaneal osteotomy
	Base of metatarsal osteotomies
Ankle valgus	Medial distal tibial hemiepiphysiodesis
	Supramalleolar osteotomy
Hindfoot (subtalar) valgus	Calcaneal osteotomy (medial slide)
	Extra-articular arthrodesis of the subtalar joint (Grice)
	Triple arthrodesis
Convex pes valgus (rocker bottom foot)	Open reduction with transfer of tibialis anterior and peroneus brevis and lengthening of the tendo-Achilles

left immobile in abnormal positions for long periods of time. These joints are best treated with stretching exercises and orthoses to maintain the joint in neutral position. Active joints, on the other hand, are prone to develop fixed contractures over time and secondary bony deformities. These are treated by dynamic braces to counteract the unopposed muscle actions. Surgical treatment includes muscle-balancing procedures, such as tendon lengthening and transfers [125], and corrective osteotomies to address fixed deformities and to stabilize joints (Table 17.3). These are ideally done when the child is older, so that optimal growth potential has been achieved [125–127].

Box 17.6. Principles of Tendon Transfer

1. The joint of interest must be free of fixed deformity, with an acceptable range of passive motion.
2. Any deformity will require correction prior to transfer.
3. Muscle strength of tendon to be transferred must be at least grade 4, as the transfer is associated with a decrease in strength by one grade.
4. The line of excursion and strength of transferred muscle should be similar to the one being replaced. Linear line of action is preferable.
5. Synergistic muscle (in-phase transfer) is preferred over an antagonistic muscle (out-of-phase transfer).
6. Paratenon of the transferred tendon should be preserved.
7. Neurovascular supply to the transferred muscle should be persevered.
8. Preferable to run in one muscle compartment with a smooth gliding channel.
9. Transfer is applied under adequate tension.
10. Firm fixation to bone is preferable to tendon.

Hereditary Motor Sensory Neuropathy

These are a group of hereditary neuropathies with sensory and motor involvement of the peripheral nerves. These can usually be diagnosed by one or more of the following investigations: blood tests for genetic testing, nerve conduction studies, and nerve biopsies. The most common such neuropathy is hereditary sensory motor neuropathy (HSMN) type 1 or Charcot-Marie-Tooth (CMT) disease type 1. CMT type 1A is autosomal dominant and associated with demyelination of the peripheral nerves due to a duplication of peripheral myelin protein (PMP22) that resides on chromosome 17. The typical clinical presentation is usually not obvious until the second decade characterized by progressive atrophy, muscle weakness, loss of balance, and contractures involving the intrinsic muscles of the foot which are the first nerves to become involved. Sensory loss occurs first in the distal legs followed by the distal upper extremity. Sensory and motor involvement gradually worsens. Nerve conduction velocities are slowed in children even before the clinical onset of disease. Motor amplitudes decrease gradually. Nerve biopsy typically demonstrates age-related loss of myelinated axons and disability correlates with axonal loss [128]. There are numerous additional subtypes of CMT 1 caused by different mutations resulting in demyelination of the peripheral nerves. In contrast, HSMN type 2 or CMT type 2 (CMT2) is an axonal (non-demyelinating) peripheral neuropathy which is characterized by distal muscle weakness and atrophy, and mild sensory loss, clinically similar to CMT1 but less severe, and nerve conduction velocities are typically normal. Peripheral nerves are not enlarged or hypertrophic. There are many subtypes of CMT2 (15 genetic mutations) which are clinically indistinguishable and differentiated only by genetic testing [129].

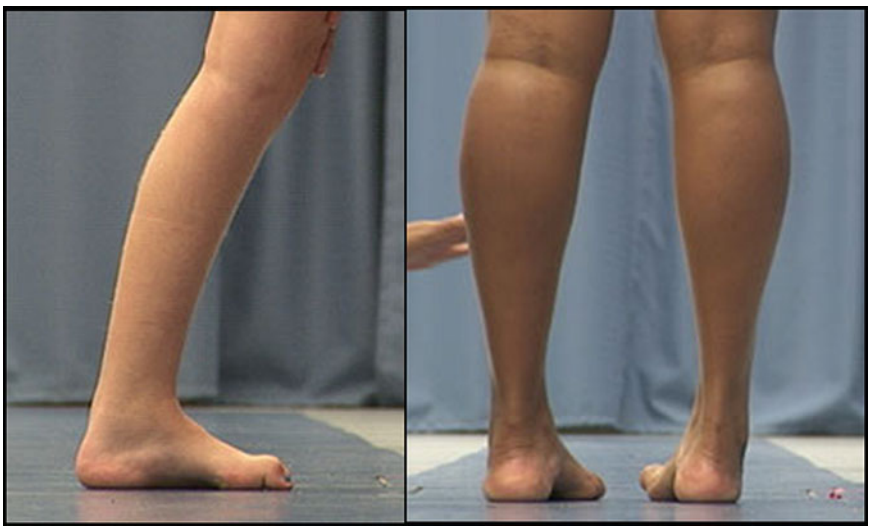


Fig. 17.26 Cavovarus foot deformity in Charcot-Marie-Tooth disease

Foot Deformities (Fig. 17.26)

The earliest and most common deformities involve the intrinsic muscles of the foot leading to clawed toes. This is followed by weakness of the peroneus brevis and tibialis anterior, while the peroneus longus and tibialis posterior are spared. This pattern of weakness results in cavus and hindfoot varus, associated with weight bearing on the lateral border of the foot resulting in callosities and difficulties with shoe wear. A high stepping gait facilitates clearance of a drop foot. Examination reveals mild sensory disturbances and absent reflexes [130].

Treatment is initially symptomatic and supportive, and custom orthotics or AFOs can play a useful role in the management of flexible deformities. Surgery is indicated when orthotics fail to manage symptoms. For cavus deformities that are not fixed, an open plantar fascia release is done, but usually in combination with an extension osteotomy of the first metatarsal (or medial cuneiform) to address the plantarflexed deformity of the first ray. Flexibility of the hindfoot varus is preoperatively assessed using the Coleman block test and reveals the plantar flexion of the first ray that is the driving force of the hindfoot varus, obviating the need for a calcaneal osteotomy. A rigid cavus associated with forefoot equinus might require extension osteotomies of additional metatarsals. More severe cavus might require a midfoot or tarsal osteotomy. Rigid hindfoot varus may need a calcaneal osteotomy (lateral based closing wedge). Claw toe deformities can be addressed with transfers of the extensor hallucis longus to the neck of the first metatarsal and the common extensor tendon to the lateral cuneiform along with flexor tenotomies or transfer of the toe flexors to the extensor hood. Triple arthrodesis is ideally avoided because of longer term risk of arthritis in the ankle but may be necessary for severe fixed deformities [131].

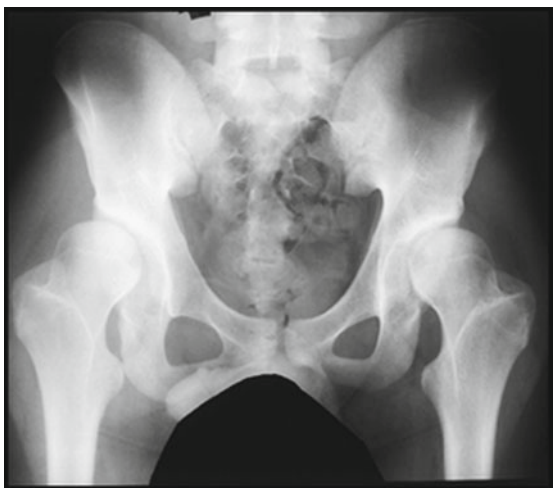


Fig. 17.27 Older onset hip dysplasia in Charcot-Marie-Tooth disease

Hip Deformities

The onset of hip dysplasia occurs relatively late in HSMN, most commonly seen in patients affected with CMT1, typically presenting first in adolescence [132, 133]. Since the development of dysplasia is insidious and asymptomatic, annual radiographs are recommended to monitor the hips once a diagnosis of CMT1 has been established. Radiographs usually reveal a dysplastic acetabulum characterized by smaller lateral and anterior center-edge angles; decreased femoral head coverage; increased acetabular anteversion; and abnormal proximal femur characterized by increased coxa valga [134] (Fig. 17.27). Pelvic osteotomies to correct the acetabular dysplasia and proximal femoral osteotomies are indicated when hips become symptomatic or the acetabular dysplasia becomes progressive. Stable internal fixation is

recommended to avoid prolonged immobilization which can cause profound weakness. Postoperative nerve palsy is reported more commonly in HSMN [132].

Muscular Dystrophies

These are a group of more than 30 genetic diseases characterized by progressive skeletal muscle weakness. Duchenne muscular dystrophy (DMD), the most common form, is an X-linked recessive condition due to an absence of dystrophin, a protein involved in the stabilization and integrity of the cell membrane. Cell membrane becomes permeable to creatine kinase (CK), which is elevated in the blood, and muscle fibers degenerate and are replaced with fibrofatty tissue. Becker muscular dystrophy is associated with faulty or decreased dystrophin and is similar to DMD but much less severe. CK levels are elevated but are not as high as in DMD.

DMD presents sometime between 2 and 5 years of age in boys. Initial presentation is characterized by a history of mildly delayed walking, with abnormal stiff knee gait, or toe walking, lumbar lordosis, and difficulty in going up stairs. Physical examination often reveals pseudohypertrophy of the calf, and a positive Gower sign, due to the greater weakness of the proximal muscles (pelvic girdle and hip extensors) than the distal lower extremity muscles. Toe walking and lumbar lordosis are essentially compensatory strategies to minimize the moments about the weight-bearing joints and maintain balance while standing and walking. Over time, fixed lower extremity contractures including equinus or equinovarus deformities of the foot can develop. The natural history is one of rapid decline with most boys unable to walk by 12 years of age, progressive scoliosis, and cardiorespiratory decline until death in the early twenties. The use of corticosteroids has been shown to stabilize the cell membrane and slow the rate of deterioration of muscle strength, prolong independent ambulation, decrease the risk of scoliosis in childhood, and preserve pulmonary function by several years [135, 136]. Lower extremity joint contractures are managed with bracing or surgically with tendon lengthening. The surgical indications for performing lower extremity tendon surgery must serve to preserve function, assist seating, or improve comfort with footwear.

Summary

This chapter provides an overview of the lower limb musculoskeletal consequences of the various neuromuscular disorders. Although the etiology, pathophysiology, and natural history vary widely across these conditions, there are some common principles of management that apply to most of these conditions. Orthopedic surgery plays an important role in management of these conditions, most effectively in the context of a multidisciplinary approach along with physical therapists, developmental pediatricians, physiatrists, neurologists, orthotists, and other specialists. Although our interventions have been shown to make substantial differences, much work remains to be done to define the effectiveness and longevity of the outcomes, in terms that are relevant and meaningful to these populations. These conditions are chronic and permanent and currently have no cures. Future advances will stem from basic research to understand the embryology and biology of the neural axis and the musculoskeletal system, and the pathophysiology of these heterogeneous disorders; and epidemiologic research that will unravel the risk factors will eventually provide the strategies to mitigate or reverse the effects of these conditions, or prevent them from occurring in the first place.

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Arthrogryposis

18

Reggie Hamdy and Noémi Dahan-Oliel

Arthrogryposis is a group of disorders characterized by nonprogressive, multiple joint contractures and muscle weakness, present at birth. The word "arthrogryposis" is derived from the Greek language (arthro = joint and gryp = curved), referring to the multiple congenital contractures inherent to this category of disorders [1]. Most individuals with arthrogryposis have normal or above normal intelligence and normal sensation. Arthrogryposis can be seen in isolation or in association with other congenital abnormalities as part of a syndrome with or without visceral and/or central nervous system involvement [2]. There are over 300 different syndromes associated with congenital contractures [3], the most common being amyoplasia affecting all four limbs [4]. Contractures typically affect two or more areas of the body and limit children's mobility, self-care, and daily activities.

Prevalence

The overall prevalence of arthrogryposis has been estimated at 1 in every 3,000 live births [5]. Recently, the prevalence of arthrogryposis was reported to range from 1 in 2,000 in Leicester, UK [6], to 1 in 5,100 in Alberta, Canada [7].

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Etiology

The main cause of arthrogryposis is diminished fetal movements, resulting from either fetal or maternal abnormalities. The decreased fetal movements ultimately lead to the development of multiple joint contractures, as connective tissue around the joint limits joint mobility, leading to contractures [1, 6]. During pregnancy, a limb will not achieve optimal growth when in utero movement is restricted for several months [1]. There are numerous causes of diminished fetal movements, including neuropathic, muscle or connective tissue abnormalities, space limitations of the fetus, intrauterine vascular compromise, and maternal diseases [1, 6]. Prenatal diagnosis may be possible by using repeated ultrasound studies to evaluate fetal movements and characteristic fetal positions.

Classification

Two main classification systems for arthrogryposis exist. The first classification system by Dr. Judith G. Hall was first published in 1997 and classifies arthrogryposis into three main groups, according to the presence or absence of associated visceral and central nervous systems [2] (Table 18.1). A more recent classification system by Dr. Bamshad and colleagues [4] divides arthrogryposis into three main entities, including amyoplasia, distal arthrogryposis, and syndromic (Fig. 18.1). The main difference among those two classification systems is the inclusion of different types of distal arthrogryposes together, as in the newer classification, or as separate entities depending on involvement of body areas, as in the Hall classification. In this chapter, the different disorders encompassed by the umbrella term arthrogryposis are presented using Hall's classification [2].

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Group 1: Disorders Affecting Mainly the Four Limbs

The most common disorder of this group is amyoplasia (a = no, myo = muscle, plasia = growth) meaning no muscle growth. It is the most common form of arthrogryposis, and represents one third of all cases. Most affected individuals have all four limbs involved. It has not been observed to recur in siblings [1].

Distal arthrogryposes (DA), as the name indicates, involve the distal joints, and do not involve a primary neurological and/or muscle disease. Distal arthrogryposes share a pattern of hand and foot involvement, limited involvement of proximal joints, and variable expressivity [4]. These disorders are inherited as autosomal dominant traits. Currently, distal arthrogryposes are subdivided into 10 types, depending on the number and nature of additional features [4]. Of these, type 5 can be further divided into several subtypes based on additional phenotypic features [8, 9]. Type I (DA1) is characterized by a typical positioning of the hands with medially overlapping fingers, clenched fists, ulnar deviation of the fingers and camptodactyly, and clubfoot. The degree of joint deformity is highly variable. The other types of distal arthrogryposes involve other body parts (e.g., eyes, mouth), and hence are presented as part of group 2 arthrogryposes.

There are other types of arthrogryposis involving mainly the limbs and which are included in group 1, such as contractural arachnodactyly and symphalangism [1].

Both amyoplasia and distal arthrogryposis type I have been found to respond well to physical therapy [10, 11] and these disorders are usually seen by orthopedic surgeons.

Table 18.1 Hall classification of arthrogryposis

Group	Type of involvement
Group 1	Mainly limb involvement (four limbs, lower limbs or upper limbs)
Group 2	Limbs are affected with involvement of other parts of the body
Group 3	Limbs are affected with central nervous system involvement

Group 2: Disorders Affecting the Limbs with Involvement of Other Parts of the Body

Distal arthrogryposes (DA) other than type 1 are included in this group as in addition to contractures of the hands and feet, they involve other body parts. Type 2 can be further separated into two subtypes, Freeman-Sheldon syndrome (FSS or DA2A) and Sheldon-Hall syndrome (SHS or DA2B) [4]. The defining characteristic in FSS is that oropharyngeal abnormalities, scoliosis, and a very small oral orifice, puckered lips, and dimple in the chin, hence the name "whistling-face syndrome" (Fig. 18.2). SHS has similar congenital contractures present in DA1, with the addition of more prominent nasolabial folds, palpebral fissures, and a small mouth [4]. DA3 and DA4 are very rare. DA5 is unique in that ocular abnormalities are present in addition to muscle contractures. Other types of DA are very rare and have distinguishing features, such as very short stature and cleft palate in DA6, and inability to fully open the mouth and pseudo-camptodactyly in DA7 [4].

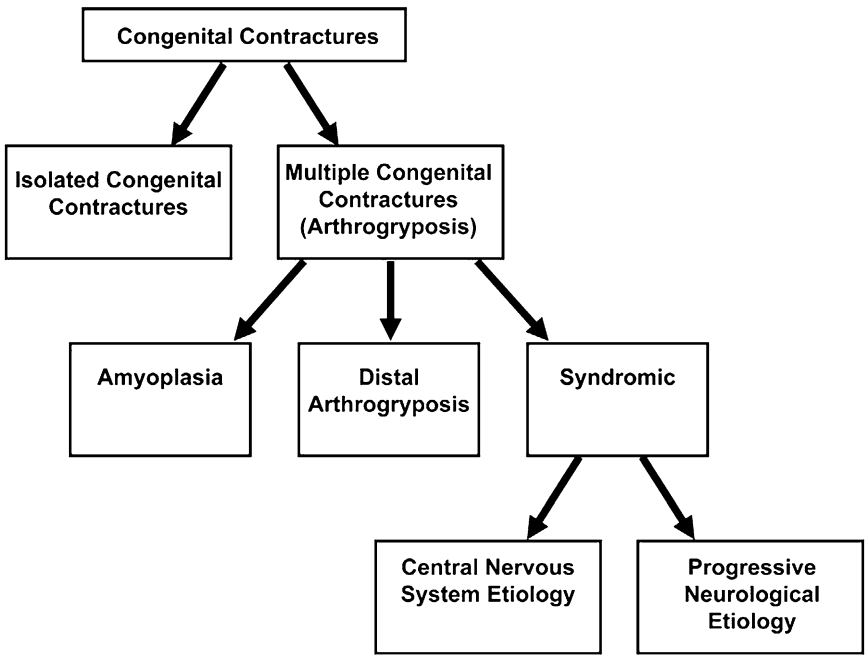


Fig. 18.1 Types of congenital contractures [4]. Reproduced with permission from Bamshad M, Van Heest AE, Pleasure D. Arthrogryposis: a review and update. J Bone Joint Surg Am. 2009 Jul;91 Suppl 4:40–6

In addition to DA, there are many different syndromes involving muscle contractures and other body areas [2], such as multiple pterygium syndromes, diastrophic dysplasia, and Larsen syndrome. Multiple pterygium syndromes involve a web or triangular membrane that forms across a joint [2]. Different pterygium syndromes have different forms of inheritance and features. Popliteal pterygium syndrome is the most common of the multiple pterygium syndromes, but is very rare and affects 1 in 300,000 live births. It is inherited as autosomal dominant with a wide pattern of expression. Key features include genitourinary, craniofacial, and extremity malformations in association with popliteal webs that vary greatly in severity. The fixed knee and ankle deformities render ambulation difficult (Fig. 18.3). Diastrophic dysplasia involves contractures of the shoulders, elbows, interphalangeal joints, and hips [1, 2]. This type of dwarfism has an autosomal recessive inheritance. Other features include cystic masses involving the external ear and cleft palate [1]. Mortality rate is increased during infancy, but life expectancy is normal past infancy. Larsen syndrome involves a distinctive facial appearance with flattening of the face, prominent forehead, depressed nasal bridge, widely spaced eyes, and ligamentous hyperlaxity, as well as flexed hips and hyperextended knees [1]. Often, there are multiple joint dislocations (hips, knees, shoulder, and elbows), hand deformities with long cylindrical fingers, feet deformities in the form of equinovarus or equinovalgus, and a high incidence of spine anomalies, specifically cervical kyphosis caused by marked hypoplasia of one or two vertebral bodies. Congenital cardiac and respiratory anomalies may also be present. The management of the orthopedic problems is usually surgical.



Fig. 18.2 Photograph of a toddler with Freeman-Sheldon syndrome showing typical facies. Reproduced with permission from Bamshad M, Van Heest AE, Pleasure D. Arthrogryposis: a review and update. J Bone Joint Surg Am. 2009 Jul;91 Suppl 4:40–6

Group 3: Disorders Affecting the Limbs with Involvement of the Central Nervous System

Numerous syndromes with involvement of the central nervous system can have associated congenital contractures [2]. These may be caused by developmental abnormalities of the forebrain in utero, chromosomal deletions or rearrangements,

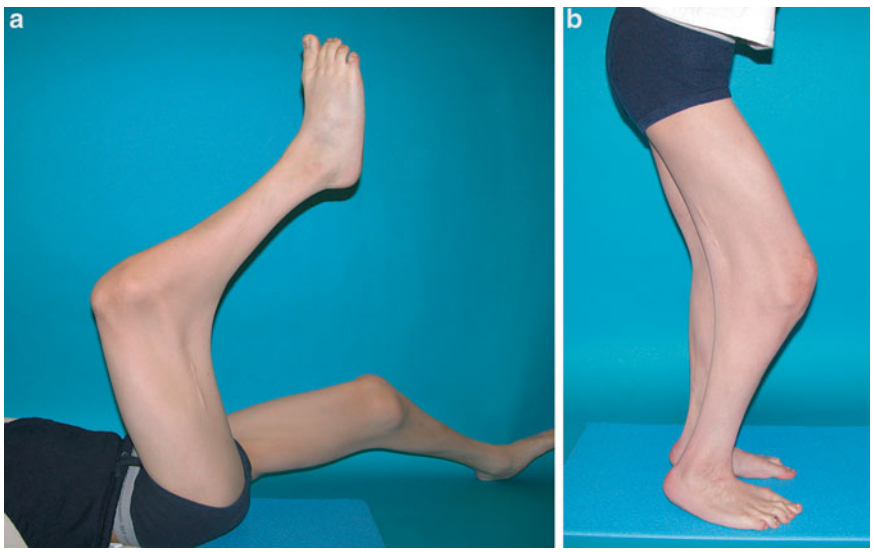


Fig. 18.3 (**a**, **b**) Eleven-year-old boy with popliteal pterygium syndrome

or genetic mutations. These disorders are the most common cause of severe arthrogryposis [4]. Nerve conduction studies and EMG may be indicated in the case of neuromuscular disorders associated with arthrogryposis [4].

Genetic Aspects of Arthrogryposis

Multiple congenital contractures are present in a number of genetic syndromes. Many specific types of arthrogryposis have been mapped to loci in human chromosomes [1, 2]. The disorder may be caused by a single gene, in which case the disorder can be autosomal dominant, autosomal recessive, or X-linked. Rarely, disorders have a much higher risk of recurrence, such as mitochondrial inheritance. The presence of chromosomal abnormalities is prominent in cases of multiple congenital contractures with intellectual disability. Recent studies have identified several genetic mutations responsible for different forms of arthrogryposis [12–15]. Some types of arthrogryposis are sporadic, such as amyoplasia, in which genetics does not play a role in its inheritance. Prenatal testing and genetic counseling are indicated for parents with a higher risk of having another affected child.

Intellectual Skills

Intelligence is normal in many forms of arthrogryposis, including amyoplasia, distal arthrogryposis type I, diastrophic dysplasia, and Larsen syndrome. Patients with certain forms of arthrogryposis associated with chromosomal abnormalities or with CNS involvement may have decreased cognitive function [1].

Workup of a Child with Arthrogryposis

Any infant or child seen with multiple contractures should have a thorough clinical evaluation including a comprehensive family history, physical examination, and if necessary, further workup. In the physical examination, it should be determined first if arthrogryposis involves only the limbs or if it involves also other parts of the body, specifically the central nervous system. Second, it is important to determine which part of the limbs is involved, and to what extent. The range of motion of each joint should be carefully recorded. Radiographs of the spine, hips, and feet should be obtained. Bone mineral density appears to be lower in children with arthrogryposis compared to age-matched peers [16], and should be investigated.

It is essential to determine if this condition is genetic or nongenetic, not only for management purposes but also for counseling the parents on the risk of future pregnancies. Genetic investigations should include both blood cells and skin fibroblasts as, in some cases, mosaicism may exist (when chromosomal studies on blood cells are normal but an abnormality is detected in the skin fibroblasts). The most common form of arthrogryposis, amyoplasia, is not a hereditary condition and no specific genetic diagnosis can be made in this condition.

The focus of this chapter is on lower limb deformities as it pertains to amyoplasia, as it is the most common form of arthrogryposis.

Clinical Picture

A child with amyoplasia presents with a typical clinical picture. In the upper limbs, the shoulders are internally rotated and adducted; the elbows are extended, the forearms are pronated, the wrist and fingers are flexed (Fig. 18.4). This pattern of upper extremity involvement is often described as a policeman's hand or a waiter's tip hand (Fig. 18.5). In the lower limbs, the foot is most commonly affected (about 90 % of cases), followed by knee deformities (about 70 % of cases) and then hip deformities in about 40 % of patients. Flexion, abduction, and external rotation contractures of the hips are typically observed (Fig. 18.6) (Box 18.1). Often, there is loss



Fig. 18.4 Photograph of a 5-month-old baby with arthrogryposis showing bilateral clubfeet, extension contractures of the knees, and upper limb involvement with extended elbows and flexed wrists



Fig. 18.5 Photograph of a 5-year-old child with arthrogryposis showing typical upper and lower limb deformities, including policeman tip position with flexed wrist and extended elbows in the upper limb, clubfeet and knee flexion contractures in the lower limb



Fig. 18.6 Photograph of a 2-year-old child showing typical flexion abduction and external rotation contractures of the hips

Box 18.1. Diagnostic Approach for the Lower Limbs in a Child with Amyoplasia

- The deformities are present at birth.
- All four limbs are usually affected.
- Sensation is normal.
- Muscular atrophy and weakness is present.
- Normal intelligence.
- The feet are most commonly affected.
- Knee flexion contractures are more common than extension contractures.
- Hip flexion, external rotation, and abduction contractures of the hip, hip subluxation, or dislocation may be present.

of skin creases across the joints and dimpling at the sites of the joints. The muscles are severely atrophied. Children with amyoplasia have normal IQ and normal sensation [17].

Prognosis

The functional long-term prognosis is usually very good for most patients with amyoplasia, unlike other neuromuscular conditions. The contractures are usually most severe at birth and then gradually improve with life. The overall good prognosis should be clearly explained to the parents. Long-term ambulatory status and functional outcome at skeletal maturity are not necessarily correlated with the severity of arthrogryposis at birth, warranting early intensive treatment and rehabilitation in these children [18, 19].

General Management

Early intervention, in the form of aggressive physiotherapy including stretching, joint mobilization, and range of motion exercises, as soon as possible after birth, can facilitate optimal long-term functional outcome [4, 17–19]. Surgical interventions may also be indicated during the first year of life, as outlined below. Regular and frequent follow-up (every few weeks) is recommended during the first year of life to monitor progression and plan surgical interventions. This is followed by regular visits, typically every 6 months.

Children with arthrogryposis should be followed in a multidisciplinary clinic. The overall goal in the management of these children is to improve function and ambulation while fostering optimal development. It is often recommended that the foot deformities be treated first, followed by the knee and finally the hip deformities [17].

Lower Limb

Involvement of the lower limb is common in arthrogryposis [11], and presents a challenging problem to the pediatric orthopedist. Lack of joint motion may be due to inadequate muscle development, lack of normal and mobile skin and tissue, and contractures with fibrous connective tissue. These issues must be considered when devising the treatment plan, and include the affected joints (e.g., foot, knee, and hip) (Box 18.2). Range of motion of the knee and hip may be improved with treatment, thereby facilitating ambulation [20].

Foot

Clubfoot deformity or talipes equinovarus is the most common foot deformity in arthrogryposis and is present in over 90 % of infants with amyoplasia [21]. Talipes equinovarus is much more common and is more rigid than the standard idiopathic clubfoot. It may be associated with knee contractures and hip contractures, subluxation, or dislocation (Fig. 18.7).

The goal in the management of feet deformities is to obtain a painfree, plantigrade foot. Treatment should start immediately after birth with the Ponseti technique. Previous studies have reported that clubfeet in children with arthrogryposis are usually stiff and resistant to standard treatment and may require talectomy as a first-line treatment [22, 23]. However, more recently, it has been shown that tenotomy of the tendo Achilles before the start of the Ponseti technique may facilitate the manipulations and may yield good results with the Ponseti technique [24]. We recommend this approach as the initial treatment. In cases of failure of the Ponseti technique, a formal posteromedial release is recommended [25] even though the recurrence rate is high (Fig. 18.8). In cases where a posteromedial release is not sufficient to obtain a plantigrade foot, then a talectomy is advisable [22, 23]. In cases of recurrent deformities, repeat soft tissue releases could be dangerous as the anatomy is completely distorted and neurovascular damage may ensue. Consideration should be given to correction through multiple osteotomies. The use of gradual correction with external



Fig. 18.7 Photograph of a 3-week-old infant with arthrogryposis showing right knee extension contracture, left knee subluxation, and bilateral clubfeet

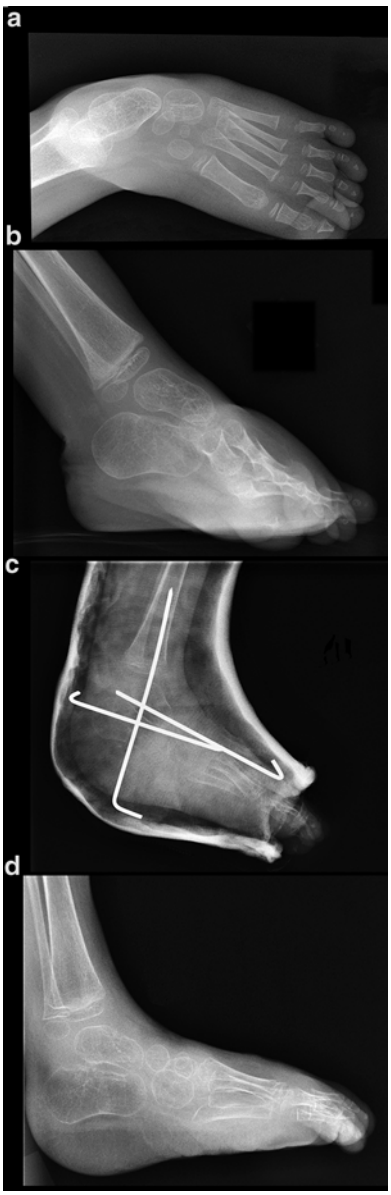


Fig. 18.8 (**a**, **b**) Radiographs of a 4-year-old child with arthrogryposis showing left clubfoot. (**c**) Postoperative radiograph following radical posteromedial release of the left foot after a failed Ponseti technique. (**d**) Three-month postoperative radiograph showing adequate correction



Fig. 18.9 Photographs (**a**, **b**) showing severe deformities of the right foot in a 3-year-old girl. The right foot is severely supinated at age 9 (**c**, **d**). Treatment of deformity with Ilizarov external frame (**e**, **f**). (**g**, **h**) Satisfactory alignment of the right foot post-Ilizarov removal at age 11

fixators is another viable option (Fig. 18.9). A standard Ilizarov circular fixator could be used [26] or a Taylor Spatial Frame with the foot program [27]. In patients near or at skeletal maturity presenting with residual or recurrent foot deformities, triple arthrodesis may be the best option [28].

Conservative treatment in the form of the Ponseti technique, as described above, should be considered first. In case of recurrence or substantial residual deformity, a radical posteromedial release may be necessary around the age of 1 year followed by prolonged splinting to prevent or minimize recurrence of deformities [24, 27]. It should be clearly explained to the family that while such surgical treatment will eventually correct the deformity, the foot remains stiff, with a high likelihood of needing subsequent surgeries as the recurrence rate is very high [22, 29].

Congenital vertical talus is reported to occur in approximately 2–12 % of patients with arthrogryposis [30]. Because many of these patients have the potential to ambulate, the

congenital vertical talus should be treated early, before walking age. Immediately after birth, the modified Ponseti technique should be initiated as described by Alaee, Boehm, and Dobbs [31]. This may be successful in the correction of the deformity. However, if the congenital vertical talus is rigid, then more aggressive treatment in the form of a one-stage release is usually recommended. A tibialis anterior transfer to the talar neck with or without Grice subtalar fusion is also recommended [32]. In very stiff feet and recurrent deformities, surgical options include excision of the navicular, talectomy, multiple osteotomies, and gradual correction with an external fixator (Fig. 18.10). If the patient is near skeletal maturity and has severe deformities, then a triple arthrodesis may be considered [32].

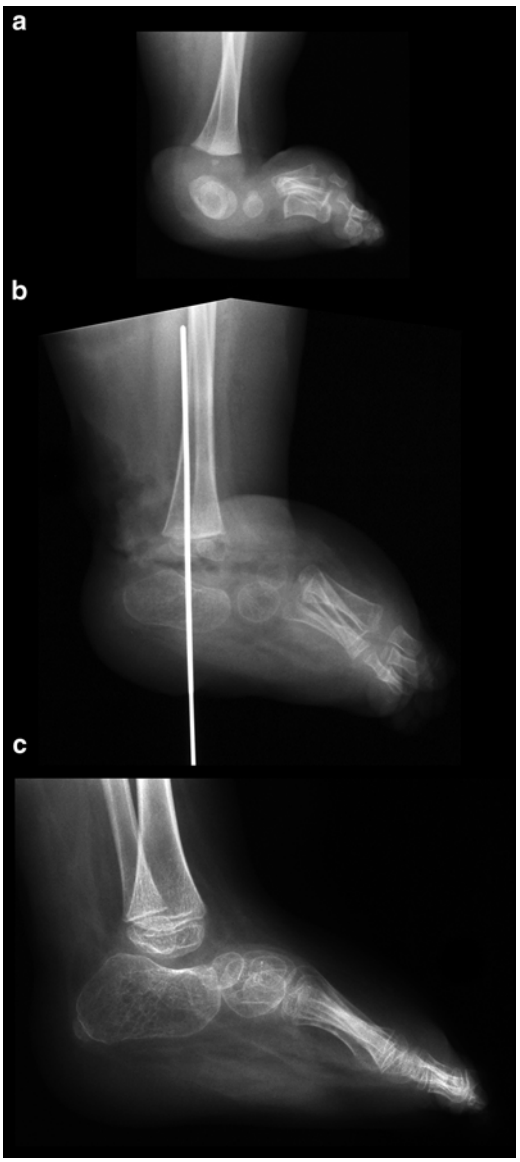


Fig. 18.10 (**a**) Radiograph showing vertical talus in a 2-year-old child with arthrogryposis. (**b**) Radiograph obtained after talectomy was performed shows good position of the calcaneus under the tibia. (**c**) Radiograph obtained 4 years postoperatively

Knee

The knee is the second most commonly affected lower extremity joint in amyoplasia (38–90 % of patients) [33]. Knee contractures can be the most disabling deformity as they prevent motion and anatomical limb alignment [34]. Among knee deformities, flexion contractures are the most common in arthrogryposis [35, 36] and are also more disabling than extension contractures [35]. The goal in the management of flexion deformities is to facilitate ambulation, sitting, and standing (Table 18.2). Knee deformities may be more difficult to treat than other deformities and, more importantly, present some of the most challenging problems causing gait disturbances and may render ambulation very difficult [33, 34]. Contrary to extension deformities that may not interfere with walking, flexion contractures of more than 20–30° usually interfere with ambulation and should be treated. In fact, the presence of knee flexion contractures is the single most important indicator of the potential for ambulation. We and others have shown that correction of knee flexion deformities has a direct positive impact on the ambulation potential of these children [19, 36]. In our own series of patients where knee flexion contractures were treated with supracondylar osteotomies (Fig. 18.11) or with gradual correction using an Ilizarov circular frame, we reported that ambulation gains were maintained in most patients despite some loss in correction with time in many patients [19]. However, quadriceps strength should also be taken into account in the management of knee deformities as quadriceps weakness (noted in about 60 % of patients) [37] may preclude any gains in ambulation despite correction of knee flexion deformity [33].

The fact that there are several options for the treatment of knee flexion deformities emphasizes the challenge that these deformities present [34]. Treatment of knee deformities should start at birth with intensive stretching exercises and bracing. If the flexion deformity exceeds 30° and is not responsive to physiotherapy, serial casting or braces with hinges may be used in order to gradually stretch the contracture. If these conservative measures fail to decrease the contracture to 20° or less, more aggressive techniques should be used such as soft tissue releases, femoral supracondylar osteotomy with or without shortening, anterior hemiepiphysiodesis of the distal femur, and gradual correction of the

Table 18.2 Indications of management for knee contractures

Knee contracture severity	Degrees	Management
Mild	0–20	• Physiotherapy • Stretching • Serial casting
Moderate	20–40	• Anterior distal femoral hemiepiphysiodesis • Soft tissue release
Severe	40–60	• Femoral extension osteotomy
Very severe	60+	• Gradual distraction

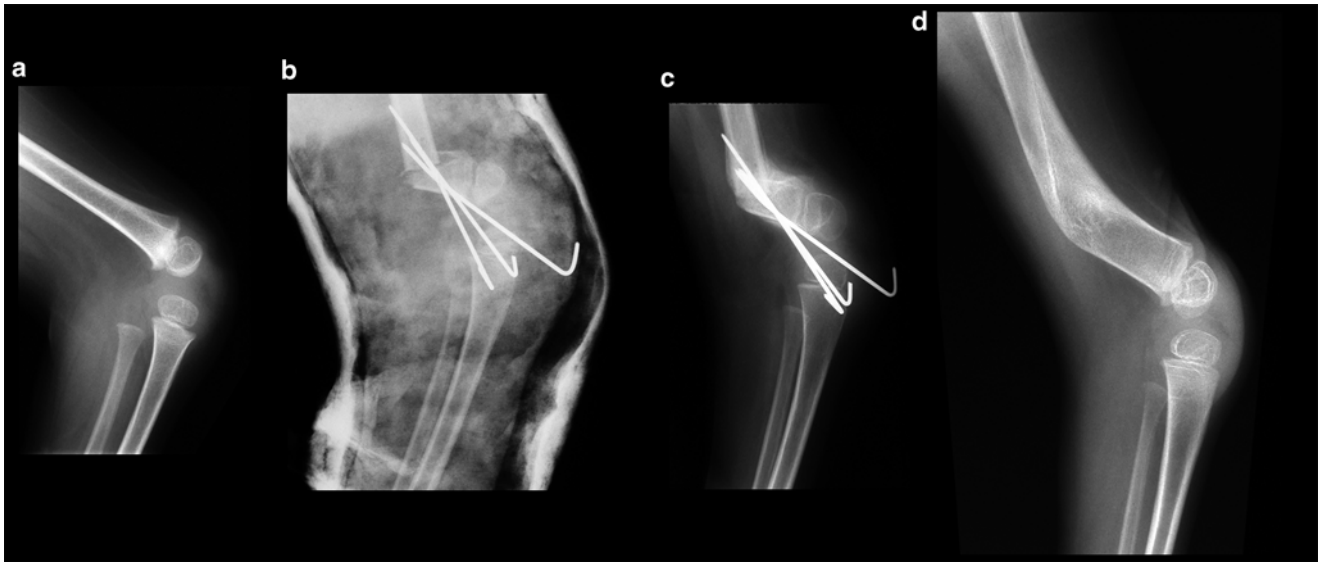


Fig. 18.11 (**a**) Preoperative radiograph showing a severe knee flexion deformity in a 5-year-old child with arthrogryposis. (**b**) Postoperative radiograph after supracondylar osteotomy was performed. (**c**) Radiograph obtained 2 months postoperatively. (**d**) Radiograph obtained 1 year postoperatively showing remodeling

deformity using external fixators. However, some of these techniques have a high rate of recurrence in the skeletally immature patient and the natural history of knee flexion deformity is typically one of progression [38].

Soft tissue releases of the hamstrings alone are usually not enough to correct the flexion deformity, and the release should include the posterior capsule of the knee joint. The surgical dissection is not easy as much of the muscle is replaced by fibrous tissues and it may be difficult to identify the neurovascular structures. The posterior capsule has to be released in most cases as it is very thick and is believed to be one of the main factors in recurrence of the deformity [33]. Anterior hemiepiphysiodesis of the distal femur is another excellent option that has been recently described [38]. This technique is minimally invasive and should be used as a first-line treatment of flexion deformities in the skeletally immature patient with mild to moderate deformities—less than 45° [33, 39]. Care should be taken in the placement of the tension-band plates so as not to interfere with the patellofemoral joint. Supracondylar extension osteotomy with distal femoral shortening is another excellent option in the treatment of knee flexion deformities as it avoids extension soft tissue dissection around the knee. The main problem with this technique is that it does not increase the range of motion of the knee—it only changes the arc of motion. Therefore, an increase in extension of the knee leads—at the same time—to a decrease in flexion, and this may interfere with the sitting position and getting in and out of a car [33]. Hence, careful planning of the treatment intervention is necessary, as well as a discussion with the patient and family preoperatively regarding the expectations of the surgery. Because of the rapid remodeling that occurs following these extension osteotomies (about 1° a month) [40], this technique has a high recurrent rate of flexion deformities and it is therefore preferable to reserve that technique for patients closer to skeletal maturity [34].

In severe knee flexion deformities of more than 60°, it may be safer to use gradual correction with external fixators [36] (Fig. 18.12). This has the advantage of protecting the neurovascular structures from sudden stretching, unless a concomitant shortening is performed with the supracondylar osteotomy. Posterior soft tissue release could be performed before the application of the external fixator [36]. Furthermore, gradual distraction may lead to an increase in the arc of motion of the knee. Attention to details of placement of the hinges when standard Ilizarov circular fixator is used is important in order to prevent subluxation of the knee or compression of the articular cartilage [33]. Gradual distraction may be associated with several complications, including fractures, exacerbation of the osteoporosis already present in these patients, knee subluxation, pin tract infection, pain, and recurrence of the deformity [36].

Knee extension contractures are less common [33] and may vary from full extension of the knee with no flexion, knee recurvatum, and anterior knee subluxation to frank knee dislocation [33]. Long-standing deformities may lead to abnormal loading on the articular cartilage with subsequent damage and deformities of the epiphysis. Treatment of these contractures and/or dislocations should also start immediately after birth and include passive stretching, casting, and bracing. If knee extension contracture is associated with a clubfoot deformity, then corrective casting could be



Fig. 18.12 (**a**) Photograph shows a severe knee flexion deformity in a 10-year-old patient with arthrogryposis (scar shows previous soft tissue releases). (**b**, **c**) Photographs show that the patient had difficulty standing with this degree of knee flexion contracture. (**d**) Photograph shows gradual correction with a circular external fixator. (**e**) Photograph shows full correction has been obtained

Box 18. 2. Management of the Lower Limb Deformities in Amyoplasia

- Early aggressive physiotherapy
- Feet
  - Early tenotomy of the tendo Achilles plus Ponseti technique
  - If Ponseti technique fails, consider posteromedial release
  - In severe and recurrent cases, talectomy or multiple osteotomies with acute or gradual distraction are indicated
- Knee flexion deformities
  - Stretching
  - Bracing
  - Casting following posterior soft tissue releases
  - Anterior hemiepiphysiodesis of the distal femur
  - Distal femoral extension osteotomies
  - Gradual soft tissue distraction
- Knee extension contractures
  - Stretching
  - Casting
  - Quadricepsplasty
- Hip flexion contractures
  - Physiotherapy
  - Soft tissue releases
  - Subtrochanteric extension osteotomy

- Hip instability
  - Reduce unilateral hip dislocation
  - In bilateral hip dislocation, surgical treatment depends on mobility of the hip
- The potential for ambulation is not related to the severity of deformities at birth; warranting early intervention for optimal long-term function

performed on both the foot and knee simultaneously [33]. If surgical correction of the foot is required, then sufficient amount of knee flexion is necessary in order to maintain the position of the foot, and hence knee surgery could be performed at the same time as the foot surgery [33]. In cases of failure of conservative treatment for knee extension contractures, a quadricepsplasty is indicated when less than 35° flexion is present despite nonoperative measures [41]. Most authors suggest that surgery should be performed early, between 3 and 6 months of age [41]. Numerous techniques of quadricepsplasty have been described, including percutaneous, mini-incision, and open techniques [33]. Care should be taken not to over lengthen the quadriceps tendon [33]. Knee dislocations usually require surgery and should be reduced before 6 months of age [33].

Hip

Hip involvement is common in patients with amyoplasia and has been reported to range from 50 to 90 % [18, 42]. Contractures may occur as isolated flexion or external rotation contractures, or as a combination of flexion, external rotation, and abduction contractures. Stretching exercises should start immediately after birth. It is difficult to apply orthoses for hip flexion contractures. An increased lumbar lordosis may compensate for a hip flexion deformity. However, if there is persistent hip flexion contracture of more than 30° despite physiotherapy, surgical treatment is usually required as such contractures may interfere with walking [43]. Soft tissue release should be considered, although it is very difficult to obtain complete correction of the contracture with soft tissue release only. Alternatively, subtrochanteric extension femoral osteotomy may be considered, but the recurrence rate is high [44]. For these reasons, it is preferable to wait until skeletal maturity before performing a subtrochanteric extension osteotomy. External rotation contractures may be beneficial as they widen the base of stance and in most of the cases do not need special treatment [17].

Hip dislocations occur in about 10–30 % of patients with amyoplasia [45] and are almost always teratologic. Almost all authors agree that a unilateral hip dislocation needs to be treated in order to prevent pelvic obliquity, scoliosis, and unequal sitting pressure. On the other hand, the management of bilateral hip dislocation in children with amyoplasia is controversial. Some authors believe that attempts at surgical

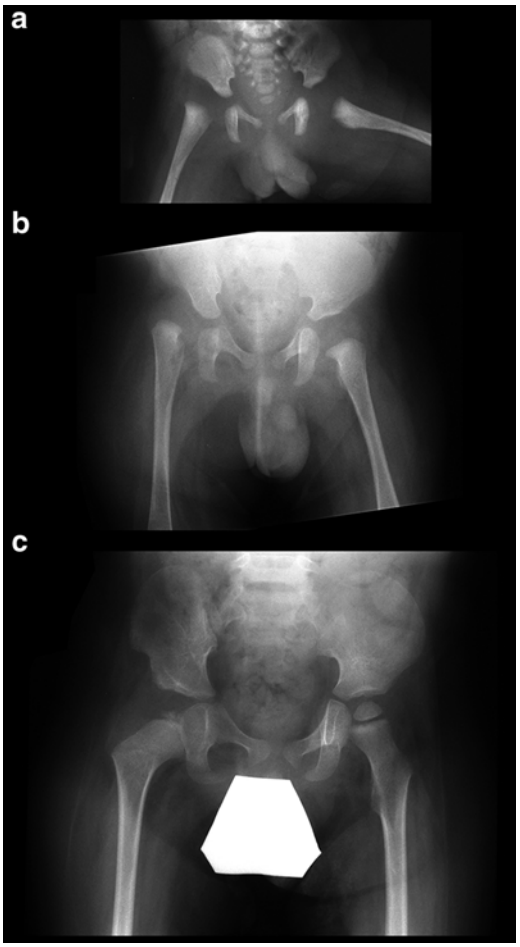


Fig. 18.13 (**a**) Radiograph shows a unilateral hip dislocation in a neonate with arthrogryposis. The contralateral hip shows an abduction contracture. (**b**) Radiograph shows failed closed reduction of the right hip and the abduction contracture of the contralateral hip has been successfully treated with physiotherapy. (**c**) Postoperative radiograph obtained after open reduction showing avascular necrosis of the right hip

reduction of bilateral dislocations have a high rate of complications, including increased stiffness, avascular necrosis, and the possibility of redislocation and hence recommend no treatment. On the other hand, we and others favor reduction of bilateral dislocation as this may help function and ambulation [25, 42].

Closed reduction or the use of Pavlik harness is often unsuccessful [46, 47] and open reduction is usually necessary (Fig. 18.13). We prefer to use a Smith-Peterson approach, although a medial approach is advocated by others [21]. A complete capsulotomy near the acetabular brim is necessary, together with a pelvic osteotomy, usually an acetabuloplasty. A high rate of avascular necrosis, up to 70 % in some series, has been reported [46]. Postoperatively, a hip spica should be kept for 8–12 weeks, followed by part-time bracing for another 12 weeks [17]. Bilateral stiff hip dislocations with limited abduction may interfere with toileting, and in such cases bilateral valgus osteotomies are recommended (Fig. 18.14).

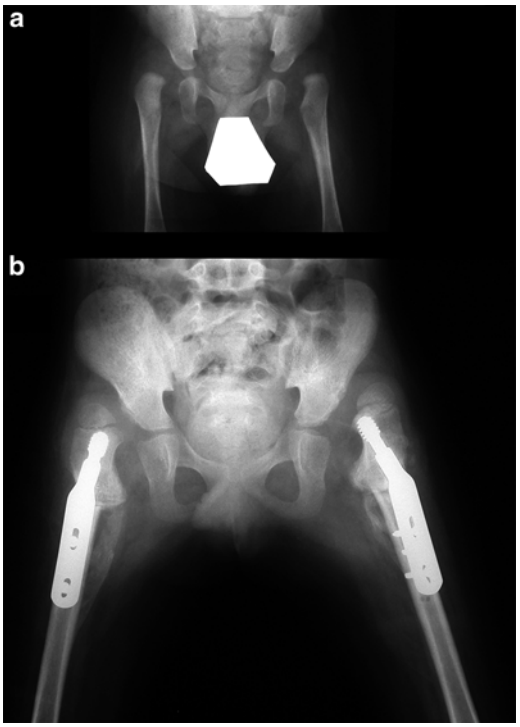


Fig. 18.14 (**a**) Radiograph of a young girl with arthrogryposis showing a bilateral abduction contracture of the hips. (**b**) Radiograph showing bilateral valgus osteotomies with increased abduction

Upper Limb

The classical upper limb deformities in arthrogryposis include adduction and internal rotation of the shoulder, extension of the elbow, flexion and ulnar deviation of the wrist, and stiff fingers with thumb-in-palm deformity. The goal in the management of upper limb deformities is to maximize functional independence by positioning the limbs for optimum use. For a long time, it was believed that the best position of the upper limbs should be one limb in full extension and one in flexion. However, absence of a strong unilateral grasp renders the use of bimanual function necessary, and therefore a bimanual use pattern of the upper limbs is recommended. Preservation of available elbow and wrist motion is essential [48]. Early surgical treatment is warranted if bony changes of normal elbow joints occur early on, and if development and independence in daily activities may be promoted through correction of the upper limbs [49]. Ideally, the upper limbs should be positioned at tabletop level in order to facilitate self-care, self-feeding, and the use of computers.

Scoliosis

The reported incidence of scoliosis in arthrogryposis is 20–30 % [50]. About one third of children with amyoplasia have scoliosis. There is no single curve that is typical in these patients; congenital, paralytic, and idiopathic-like curves have been reported, and spinal deformity varies from minimal to severe [50]. Scoliosis is typically not present at birth but is usually detected during the first few years of life. Early onset, a paralytic curve pattern, and pelvic obliquity are poor prognostic patterns for curve progression. Bracing is usually ineffective in controlling the curve and most children with severe scoliosis will require surgical treatment. Combined anterior and posterior fusion seems to give the best curve correction with the least loss of correction after surgery. Children with arthrogryposis should have a physical examination of the spine early on with regular follow-ups, including upright radiographs to avoid an undetected severe deformity that is difficult to treat.

Orthopedic Management of Specific Conditions

Popliteal Pterygium Syndrome

Conservative treatment in the form of physiotherapy, traction, and casting is usually unsuccessful. Careful preoperative planning with MRI examination is essential in order to delineate the exact anatomical position of the sciatic nerve. When the popliteal webbing is mild, multiple skin Z plasties, excision of the posterior fibrotic band, hamstring lengthening, posterior capsulotomy, and tendo Achilles lengthening are recommended. When the flexion deformity is severe, femoral shortening and extension osteotomy with soft tissue release may be one option [51]. Another option that seems to give good results in severe deformities is gradual correction using a circular fixator, excision of the dense fibrotic band and multiple Z plasties of the skin.

Larsen Syndrome

Conservative treatment is typically unsuccessful in stabilizing the knees and surgery, including lengthening the quadriceps is recommended. The treatment of bilateral hip dislocations is controversial. Equinovarus deformities are treated with posteromedial release. Conservative methods are recommended for the correction of hip dislocations, knee deformities, and clubfeet, and operative correction should be performed once the child's general health is stable [52]. Because of the potential morbidity and mortality associated with cervical kyphosis, early posterior spinal fusion is indicated for patients with mild and flexible cervical kyphosis. Children with Larsen syndrome should be screened with radiographs at their first visit for early detection of cervical kyphosis to enable timely surgical management with posterior fusion [53].

Timing for the Management of Lower Limb Deformities

Many patients will have involvement of the feet, knees, and hip joints. The treatment should be individualized. However, early aggressive treatment, starting with the feet, then progressing proximally to the knees and then the hips is recommended.

Intensive physiotherapy throughout the treatment process is strongly emphasized. We do not recommend simultaneous correction of multiple joint deformities at the same time because of the lengthy surgical intervention involved with such a procedure and the different rehabilitation programs for each joint.

Ambulation in Children with Amyoplasia

Most of the lower limb deformities are worst at birth and several studies have shown that many children with amyoplasia are able to ambulate, despite having had numerous surgeries [18, 43]. As previously mentioned, correction of severe knee flexion deformities facilitates ambulation. Muscle strength, specifically that of the quadriceps, may affect ambulation to a greater extent that the severity of the deformities [43] and hence the importance of muscle strengthening exercises. The importance of adequate bracing and its role in helping children ambulate cannot be overemphasized (Fig. 18.15). Other factors that may affect ambulation include upper limb function and the child's and caretakers' motivation. Although amyoplasia is not a progressive disorder, there is a tendency for recurrence of the deformities after correction.



Fig. 18.15 Bracing and the use of canes to facilitate functional ambulation as well as transfers from a wheelchair

Rehabilitation

Multiple joint contractures usually affect the child's capacity of performing daily activities, such as feeding, dressing, grooming, toileting, mobility, and transfers. Decreased function in daily activities had a linear relationship with ambulatory level in 30 children aged 5–18 years of age with amyoplasia or nonsyndromic arthrogryposis, but normal scores in happiness and pain/comfort were reported [16]. Participation in sports and physical activity was found to be reduced in children with arthrogryposis compared to peers [16, 54, 55], demonstrating the need for rehabilitation in these children. The main objective of rehabilitation professionals is to maximize the child's potential and autonomy at home, at school and in the community. Stretching and bracing starting early in infancy is critical to optimize range of motion. Physical therapists focus on improving gross motor skills and mobility. Assistive devices (e.g., cane, walker, and wheelchair) are recommended depending on the severity of contractures and involvement of joints in order to promote independent mobility. Technical aids and other customized adaptations such as adapted utensils, raised working surface and adapted toys, and access to computers are provided to compensate for decreased function and facilitate autonomy in activities of daily living (e.g., feeding, writing, play, computer use). Occupational therapists focus on fine motor skills and activities of daily living. Splints or braces may be indicated to improve or maintain range of motion and joint alignment, to prevent contractures, and to maximize function in everyday activities such as eating, writing, or walking. Speech therapy, nutrition, and other rehabilitation areas may be warranted depending on the child's underlying needs. An individualized treatment plan delivered throughout the lifespan of the affected child with arthrogryposis via a multidisciplinary team is essential to promote each child's full potential during infancy, pre-school, school-age, and adolescence (Table 18.3). Adequate transitioning to adult health care services and preparation for independent living is recommended when the child is nearing discharge from the pediatric hospital center.

Summary

Arthrogryposis is a general term that encompasses a multitude of different disorders with the commonality of multiple congenital contractures. The lower limbs are often affected, requiring surgical interventions and intense and long-term rehabilitation. Clubfoot deformity or talipes equinovarus is the most common foot deformity in arthrogryposis. The knee is the second joint most commonly affected. Knee contractures

Table 18.3 Goals in rehabilitation during the lifespan

Lifespan years	Rehabilitation goals	Frequency of therapy
Infancy	• Increase range of motion and maintain gains	Weekly
	• Positioning	
	• Educate and reassure parents and family about handling and caring for their child	
Pre-school and school-age	• Maintaining range of motion	Weekly or monthly for follow-up
	• Strength training	
	• Promoting independence in functional activities	
	• Assistive devices to assist mobility • Technical aids and customized adaptations to maximize function in self-care, handwriting, computer, and leisure activities	
Adolescence	• Therapy following surgery to improve range of motion and strength	Intensive therapy postsurgery
	• Maximize independence in school and community	Consultation and monitoring as needed (e.g., driving)
	• Transitioning to adult health care	
	• Preparation for independent living	

can be the most disabling deformity as they prevent motion and anatomical limb alignment. Treatment must be individualized and all the affected joints of the upper and lower limbs and spine should be comprehensively evaluated. Multidisciplinary treatment of the child with arthrogryposis with a focus on early and continued clinical evaluation is warranted for a positive prognosis and to optimize the affected child's integration in society and improve his or her quality of life.

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Limb Lengthening and Deformity Correction in Patients with Skeletal Dysplasias

19

Mihir M. Thacker, Ellen Dean Davis, Colleen P. Ditro, and William Mackenzie

Introduction

Skeletal dysplasias include a wide and varied array of disorders, all characterized by an abnormality in the formation of bone and cartilage. There are many (>400 types) skeletal dysplasias and most are caused by specific mutations in genes involved in skeletal development [1] (Table 19.1). These disorders are often associated with short stature (proportionate or disproportionate), angular deformities of long bones, joint surface irregularity, and ligamentous laxity/instability [2–5]. Patients with skeletal dysplasias may have spinal deformities and/or instability which can be a significant cause of morbidity. In addition to skeletal manifestations, they may also have significant systemic involvement, which often is the cause of premature mortality [2, 5] (Table 19.2). Extremity involvement can be quite disabling and affect quality of life. Treatment of these children must involve a multidisciplinary team familiar with both skeletal and extraskeletal concerns. There is a paucity of literature regarding the natural history of many of these disorders, but there has been a recent surge in interest in many of these disorders, particularly mucopolysaccharidoses [6–9]. With improved understanding of the mechanisms involved in the genesis of these disorders, better medical treatments are being formulated.

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Skeletal abnormalities in patients with skeletal dysplasias can be a source of considerable morbidity and often need surgical correction [5, 10]. These may result from:

1. Shortening of specific limb segments (rhizomelic, mesomelic, and acromelic) characteristic of many disproportionate skeletal dysplasias with short stature.
2. Unequal growth rates of paired long bones in the forearm and leg (e.g., relatively long fibula in achondroplasia associated with genu varum; and relatively short fibula in chondro-ectodermal dysplasia associated with genu valgum).
3. Joint contractures, deformities, and instability: Joint contractures result from angular deformation centered at the joints rather than in the adjacent physeal and metaphyseal regions of the developing bones. These are almost always in flexion and are particularly characteristic of diastrophic dysplasia. Joint instability can result from a combination of generalized ligamentous laxity and structural epiphyseal abnormality (e.g., spondyloepiphyseal dysplasia, Morquio syndrome).

Lower Extremity Deformity in Skeletal Dysplasia

Individuals with skeletal dysplasias are not only shorter in stature than the average population, but each dysplasia has its own set of unique skeletal abnormalities (Box 19.1). In addition, the natural history of these dysplasias is highly variable and influences decision making. Characteristic deformities at various bones and joints are seen in skeletal dysplasias (Table 19.3).

The challenges of deformity correction in skeletal dysplasia patients include:

- Multifocal, and often multiplanar deformities (sagittal, coronal, rotational, and translational).
- The abnormalities in ossification can impair visualization of the chondro-osseous structures and joint lines on plain radiographs, adding another level of difficulty to

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Table 19.1 Site of genetic mutation and mode of inheritance of certain bone dysplasias

Dysplasia	Gene defect	Mode of inheritance
Diastrophic dysplasia	DTDST gene (5q32-q33.1)	AD
Multiple epiphyseal dysplasia (MED) Type I Type II	COMP (Chr19) COL9A2	AD, less commonly AR
Pseudoachondroplasia	COMP (chromosome 19q13)	AD
Ellis-van Creveld syndrome	EVC gene (chromosome 4p16)	AR
Achondroplasia	FGFR3	80 % Sporadic mutation 20 % AD
Hypochondroplasia	FGFR3/IGF-1	Spontaneous mutation, less commonly AD
Spondyloepiphyseal dysplasia (SED)	Type II collagen	Congenita: AD Tarda: x-linked recessive
Kniest syndrome	Type II collagen	AD
Metaphyseal chondrodysplasias Schmid type Jansen type McKusick	Collagen type X A1 chain deletion PTHRP (chromosome 3p22-p21.1) RMRP (chromosome 9)	AD AD AR
Cleidocranial dysostosis	RUNX2/CFBA1gene (chromosome 6)	AD
Morquio syndrome (type IV Mucopoly-saccharidosis)	GALNS/GLB1 gene defect	AR

AD autosomal dominant, *AR* autosomal recessive, *DTDST* diastrophic dysplasia sulfate transporter, *COMP* cartilage oligomeric protein, *COL* collagen, *EVC* Ellis-van Creveld, *FGFR3* fibroblast growth factor 3, *IGF1* insulin like growth factor 1, *PTHRP* parathyroid hormone related peptide, *RMRP* RNA component of mitochondrial RNA processing endoribonuclease, *GALNS* *N*-acetylgalactosamine-6-sulfatase, *GLB* beta galactosidase, *RUNX2* runt related transcription factor 2

Table 19.2 Selected systemic abnormalities in patients with skeletal dysplasias

System involved	Pathologic condition	Skeletal dysplasia
Head	Hydrocephalus	Achondroplasia
Eye	Cataracts Retinal detachment	Morquio syndrome SED congenital, Kneist
Ear	Recurrent otitis media Hearing loss	Achondroplasia, Kneist Kneist
Cardiac	Congenital heart defects Acquired (cardiomyopathy, valvular disease)	Ellis-van Creveld dysplasia Morquio syndrome
Respiratory	Laryngotracheomalacia Central +/− obstructive sleep apnea Restrictive lung disease	Campomelic dysplasia, diastrophic dysplasia Achondroplasia Metatropic dysplasia, thanatophoric dysplasia, Ellis-van Creveld dysplasia
Immunologic	T-cell impairment, neutropenia, anemia	McKusick metaphyseal chondrodysplasia

Table 19.3 Characteristic deformities of various skeletal dysplasias

Deformity	Skeletal dysplasia
Acetabular dysplasia	Diastrophic dysplasia MED Morquio syndrome
Coxa vara	Cleidocranial dysplasia MED Pseudoachondroplasia SED—congenita Schmid metaphyseal chondrodysplasia
Coxa valga	Morquio syndrome
Hip degenerative disease	Pseudoachondroplasia MED SED
Genu varum	Achondroplasia McKusick metaphyseal chondrodysplasia (mild) Pseudoachondroplasia Schmid metaphyseal chondrodysplasia
Genu valgum	Diastrophic dysplasia Ellis-van Creveld syndrome MED Morquio syndrome Pseudoachondroplasia SED - congenita
Internal tibial torsion	Achondroplasia Schmid metaphyseal chondrodysplasia
External tibial torsion	
Ankle varus	Achondroplasia McKusick metaphyseal chondrodysplasia (mild) Schmid metaphyseal chondrodysplasia
Talipes equinovarus	SED (supple) Diastrophic dysplasia (rigid)

MED multiple epiphyseal dysplasia, *SED* spondyloepiphyseal dysplasia

planning the deformity correction. This may necessitate additional imaging studies such as MRIs (often needing sedation or anesthesia, which is not without its difficulties) and CT scans as well as liberal use of intraoperative arthrograms.

- Deformity at one site (and its correction) may influence the development and/or progression of deformities at adjacent sites. Other challenges include deciding which deformities need to be managed surgically and which can be accommodated by adjacent correction.

Skeletal dysplasias can affect the quality of bone and cartilage. Therefore, it is imperative to have an understanding of the pathophysiology and natural history of these conditions. These differences can affect the types of correction and fixation used. The size and quality of bone often times may influence implant selection and necessitate creativity in the operating room in order to accomplish the desired surgical goals.

Box 19.1

- Each skeletal dysplasia is unique with specific skeletal and systemic manifestations. Evaluation by an experienced multidisciplinary team is essential.
- Deformities in skeletal dysplasias are usually multifocal and multiplanar.
- Deformities may arise from bony, articular, as well as soft tissue abnormalities. These may need to be addressed individually for appropriate correction.

Patients with certain skeletal dysplasias have upper cervical instability and need appropriate evaluation.

Surgical Considerations

Preoperative Evaluation

Evaluation of a child with skeletal dysplasia is best performed by a multidisciplinary team (including orthopedic surgery, genetics, physical and occupational therapists, and other necessary specialists such as otorhinolaryngology, neurosurgery, ophthalmology, and dentistry) familiar with these conditions [5, 11]. A thorough history, including questions regarding pain, endurance, gait abnormalities, activities of daily living, and recreation, provides clues to functional limitations. A detailed systemic evaluation is also indicated and may identify major, sometimes life-threatening, issues (see Table 19.2). The extremities should be examined for range of motion, ligamentous laxity, joint contractures, and malalignment (in all planes). Examination of the spine for sagittal and coronal plane deformities and a thorough neurological examination are needed because of the frequent incidence of spinal deformities, instability, and neurological compromise in certain dysplasias. Cervical spine instability is common in mucopolysaccharidoses like Morquio syndrome, spondyloepiphyseal dysplasia (SED), diastrophic dysplasia, pseudoachondroplasia, and chondrodysplasia punctata. Flexion-extension radiographs are useful in evaluating such patients who may be at risk for cervical instability, especially if they have symptoms of neck pain and long track signs. We use flexion-extension radiographs in most dysplasias, except for achondroplasia, at presentation and also intermittently thereafter, especially in dysplasias such as SED or Morquio syndrome, where instability may develop over time. These, however, may be somewhat difficult to interpret in this population given the delayed ossification, odontoid hypoplasia, limited neck motion, and abnormal anatomy seen in many of these patients. Additionally, sometimes the bony landmarks used for measurement may be obscured by overlapping shadows cast by the mandible, occiput, and foramen magnum. Any patient with back or radicular pain, neurogenic claudication, incontinence, or objective neurologic findings (such as hypertonia, brisk reflexes, clonus, motor weakness) should be evaluated with an MRI [12]. The child's gait is assessed by observing the child walk, run, and play [13]. Gait analysis can be helpful in these patients. Discussion with the family is critical to establish the goals of treatment as well as to manage expectations appropriately.

Imaging

A skeletal survey is often performed at the initial visit to identify the skeletal features that aid in diagnosis. At our institution, skeletal survey for skeletal dysplasia typically includes anteroposterior (AP) and lateral radiographs of the cervical, thoracic, and lumbar spine and AP view of the long bones of the upper and lower extremities as well as of the pelvis, hands, and feet. Cervical flexion-extension views are obtained in most dysplasias (except for achondroplasia). We add further radiographs as clinically indicated. AP and lateral full-length radiographs of the lower extremities (preferably weight-bearing views) are mandatory prior to surgical intervention. These allow for an evaluation of the overall alignment and location of deformities. It is also imperative to obtain dedicated weight-bearing radiographs of the specific joints to allow for appropriate deformity analysis (please refer to Chap. 2). The full-length radiographs alone will not always allow for an accurate quantitative determination of the magnitude of deformity. For example, the magnitude of ankle deformity can be misjudged based on full-length radiographs centered at the knee because of the parallax of the X-ray beam. Additional three-dimensional studies such as computed tomography and/or magnetic resonance imaging may be performed for detailed evaluation of bone and cartilage anatomy, measurement of torsional abnormalities, assessment of joint condition, etc. However, these are static studies and may not always reflect the alignment of the limb in conditions of dynamic loading (gait). We often perform a three-dimensional gait analysis in these patients as an adjunct to X-rays to aid in preoperative decision making. Such dynamic evaluation helps to quantify the forces across the various joints and also helps us compare pre- and postoperative function.

Anesthesia

Preparation of the patient for the surgical procedure is at least as important as the surgical procedure itself (Box 19.2). Given the fact that many of these patients have small and "difficult" airways, preoperative anesthesia consults and discussion of the various issues specific to that patient are very useful. There may be minor surgical procedures but there is no "minor

anesthesia" for these patients. Anesthesia should be performed by an anesthesiologist familiar with various medical comorbidities associated with this patient population. Our institution has described a "difficult" airway management in these patients as one which requires two anesthesia providers when ventilating with a facemask. One provider applies the face mask while maintaining anterior mandibular displacement and cervical stabilization while the other ventilates with positive pressure [14]. Care must be taken to protect the neck in patients with cervical instability by using in-line cervical traction [15]. In addition, specialized tools such as video laryngoscope and fiberoptic intubation should be readily available in these situations. Given the fact that each anesthetic session poses a certain risk to the patient, we prefer to combine as many procedures as feasible under the same anesthetic.

Regional anesthesia is often used for pain control and to reduce the amount of narcotic needed during and after surgery. Caudal epidural analgesia is preferred over lumbar epidurals in patients with known vertebral anomalies, prior fusions, and thoracolumbar kyphosis that can co-exist with these conditions. Typically, caudal epidural may be administered to those patients 10 years of age and younger who undergo lower extremity osteotomies. Careful dosing of the epidural medications and regular neurovascular checks are essential to avoid missing the diagnosis of an early compartment syndrome.

Positioning

Positioning these patients is also critical as patients with severe kyphosis are at risk for developing paraparesis/paraplegia with prolonged supine position. The pathogenesis of this phenomenon is poorly understood at this time. Neuromonitoring should be considered in long procedures in such high-risk patients undergoing lower extremity surgery.

Box 19.2

- Careful anesthetic technique, protecting the cervical spine and minimizing the number of anesthetic episodes, is important.
- Consider neuromonitoring during nonspinal procedures in the supine position in patients with severe kyphosis to minimize risk of adverse neurologic events.
- Careful preoperative planning and attention to addressing the bony and soft tissue deformities is key to improving outcomes.
- Sequence of correction of deformities may vary in individual circumstances, but is usually proximal to distal.

Implant Size and Design

Implant size and design needs to be considered in preoperative planning. Some of these patients are extremely small (primordial dwarfism) and appropriately sized implants are critical to the successful execution of the preoperative plan. The popular commercially available precontoured implants will rarely accommodate the morphology of bones in patients with skeletal dysplasias. Given the poor bone quality in some of the patients, locking plate and screw constructs may be useful when performing acute corrections. Even while planning deformity correction with external fixators, implant selection should be kept in mind. For example, the appropriate fit of a Taylor Spatial Frame (Smith & Nephew; Memphis, TN, USA) may necessitate use of extra-extra short struts (which may need to be ordered preoperatively). Frequently, given the multifocal nature of the deformities, more than one osteotomy and therefore fixation options may be needed in the same bone. Having an appropriate inventory of instruments and implants saves the surgeon from frustration and improves patient outcome.

Methods of Deformity Correction

The principles of deformity correction in the dysplasia population are no different from those without dysplasias. However, the multifocal and multiplanar nature of the deformities, issues with size and quality of the bone, ligamentous laxity, and growth potential make deformity correction extremely challenging in these patients. Many of these patients have substantial delay in ossification, making planning and correction of deformities even more challenging. It has been demonstrated that the choice of osteotomy often times changes based on the results of intraoperative arthrography, particularly in patients under 8 years of age [16]. We use arthrography liberally during surgery to mitigate some of these issues. This helps us not only to visualize the cartilaginous anatomy but also to evaluate the dynamic stability of the joint with stress views in the operating room.

Deformity correction in these patients can be accomplished via acute or gradual methods.

Acute Correction

Acute methods of deformity correction include use of osteotomies with immediate correction and fixation via various methods. In general, these techniques follow the same principles as those patients without skeletal dysplasias. Care should be taken to be familiar with the various deformities associated with particular conditions (see Table 19.3). For example, genu varum is present in the majority of achondro-

plastic children by 5 years of age and 90 % of achondroplastic adults [17]. The genu varum can be secondary to distal femoral varus, lateral joint line opening, proximal and distal tibial varus and is nearly always associated with internal tibial torsion. Progressive varus, symptomatic gait abnormalities, lateral thrust, and occasionally cosmesis are our typical indications of genu varum correction. One or all of these deformities may be present, so careful preop planning must be done while planning the correction. The deformity in young children usually is due to proximal tibial varus, lateral joint line opening, and internal tibial torsion. These children can be successfully treated with a proximal tibial osteotomy with fibular shortening osteotomy via acute correction. Care must be taken to displace the proximal tibia in order to maintain a neutral mechanical axis and correct the typical internal tibial torsion. With this method, acceptable correction can be attained [18]. Recurrence is common in very young children, especially if there is undercorrection. It has been suggested that the preferred treatment for skeletally mature individuals with achondroplasia and distal tibial varus is an acute opening distal tibial osteotomy combined with a shortening distal fibula. Displacement usually is required to correct the mechanical axis [18]. The authors have typically used a shortening, derotational osteotomy to manage this deformity. With acute deformity correction, one can re-establish normal mechanical alignment, but not perform additional limb lengthening. In addition, acute corrections, particularly of the proximal tibia, have been associated with complications including peroneal nerve palsies, vascular injuries, and compartment syndromes [19].

Gradual Correction with Guided Growth

Gradual deformity correction is a powerful tool for angular realignment and/or lengthening in the skeletal dysplasia population. This can be done by way of guided growth techniques or external fixation methods. There is a paucity of literature that addresses the application of guided growth techniques in this unique population. However, we have demonstrated that growth modulation using a tension band plate and screw system or staples is an effective way to provide deformity correction in a variety of skeletal dysplasias [20]. It is a relatively simple surgery that has a low risk of damage to the physis or mechanical failure. We have also found that despite an abnormal epiphysis and metaphysis, the screw purchase has been reliable. An exception is pseudoachondroplasia where it can be difficult to achieve epiphyseal fixation. This technique can be used in very young patients [20]. Guided growth via hemiepiphyseal stapling in patients with multiple epiphyseal dysplasia (MED) has also been studied. In these children, stapling has been effective for angular deformity correction. However, the response of the physis after staple removal is unpredictable, necessitating avoidance of excessive overcorrection and close monitoring until skeletal maturity [21]. There are some skeletal dysplasias that are associated with extremely slow growth or have significant joint laxity, which can affect correction. Slow growth and a significant deformity is a combination that will likely lead to failure of correction when using guided growth treatment in such patients (Box 19.3).

Gradual Correction with External Fixation

Gradual deformity correction can also be accomplished using external fixators [22, 23]. This approach has two main advantages. Primarily, the multifocal and multiplanar deformities that are associated with skeletal dysplasias can be addressed at one surgery. Some of the gradual correction devices also allow for simultaneous multiplanar correction, addressing coronal, sagittal, and axial deformities without the need for complex osteotomies that are prone to error. Secondly, this method also allows for concurrent limb lengthening in this patient population with varying degrees of short stature. Patients must be counseled about their care and management preoperatively as limb lengthening can be associated with several complications [24, 25]. However, with appropriate instruction and care, external fixators are extremely useful for correction of most complex deformities. Currently, we prefer to use the Taylor Spatial Frame (Smith & Nephew; Memphis, Tennessee) for more complex deformities and a uniplanar external fixator for some of the less complex deformities.

Box 19.3

- Growth modulation around the knee is a useful technique for deformity correction in patients with skeletal dysplasia, but the results are dependent on the growth potential and degree of associated ligamentous laxity.
- Special inventory of implants of appropriate size is essential.
- Plan procedures keeping in mind the natural history of the underlying skeletal dysplasia.

Controversies in Lengthening for Stature

Limb lengthening (without the correction of angular deformities) in short-statured individuals has been a subject of debate [24–29]. The achondroplastic population has been the most studied to date. The motivation to lengthen limbs in the

skeletal dysplasia population has mainly been to improve functional disabilities associated with difficulty in locomotion [30]. Other concerns, particularly in the idiopathic short-statured patient, have been related to aesthetic, social, and psychological issues.

Medical and surgical treatments to address short stature in achondroplasia have been studied. Growth hormone (GH) administration has led to variable results [31]. In trials, early gains in height after commencement of GH treatment have been seen to varying degrees. However, the effects have not been as promising with continued administration. There are also concerns of aggravating the disproportionate nature of limb segment shortening. Surgical techniques for stature lengthening in achondroplasia have addressed the femora, tibiae, and humeri via distraction osteogenesis. This method has reliably been able to provide increases in height, but at a cost [24, 25, 27, 29].

The average gain in height through surgical limb lengthening in achondroplasia has been reported to be 18–23 cm, resulting in an average total patient height of 140 cm [24, 30, 32]. This does not completely ameliorate the disproportionate trunk height. There are several drawbacks to limb lengthening purely for increased stature, including prolonged treatment time and high complication rate (reported up to 100 %). It is unclear as to whether or not such bilateral limb lengthening improves the patient's quality of life. Kim et al. showed that there was an improvement in self-esteem (using the Rosenberg self-esteem questionnaire), but there was no difference in physical or functional scores (using the SF-36 and AAOS lower extremity scores) when comparing achondroplastic patients who underwent lengthening versus those who did not undergo surgical treatment [24]. They studied 22 patients with achondroplasia who had tibial and femoral lengthening and compared them to a matched cohort with achondroplasia who did not undergo lengthening. They had 123 complications in 88 lengthened segments but reported that may of these were minor and did not need intervention. They felt that lengthening was a viable option given the improved self-esteem. The same group also looked at humeral lengthening in the achondroplasia population and reported that humeri tend to consolidate faster and have less fractures and angular malalignment compared to femoral lengthening. Extensive (more than 50 % of the length of the femur) lengthening of the femur results in higher incidence of fractures and joint issues. Extensive lengthening is also associated with physeal growth retardation, both femoral and tibial. Surgical strategies include bilateral tibial lengthening, lengthening of the femur and tibia on one side, crossed femur and tibia or all 4 segments simultaneously. Time in external fixators may be shortened by lengthening over an intramedullary nail or use of intramedullary lengthening techniques. The authors rarely perform stature lengthening.

There is a role for stature lengthening in the carefully selected patient who is well aware of the long and arduous nature of the process and the potential for significant complications. However, the patient must be counseled that the gain in height may not correlate to an improvement in quality of life [25].

Postoperative Considerations

There are several aspects of the postoperative course specific to the short-statured patients that should be taken into consideration. This is particularly true when utilizing external fixation for deformity correction. These children typically require more support because of their limited upper extremity reach. It is important to work with the physical and occupational therapists to provide appropriate assistive devices and strategies for activities of daily living.

The psychosocial aspect of deformity correction should not be overlooked in patients with skeletal dysplasias. These children may have preexisting self-image concerns because of their underlying conditions. With external fixators in place, these self-conscious feelings can be amplified. It may be useful to involve mental health professionals in the preoperative evaluation and postoperative care of these patients.

The skeletal dysplasia population often travels to referral centers for these complicated deformity corrections. We prefer to keep these patients locally during the timeframe of distraction and/or correction. This allows us to follow them in clinic on a weekly or biweekly basis and to appropriately address any complications that may arise. This oftentimes entails working with local or national charities, such as the Ronald McDonald House, for lodging and other necessities. In addition, the caregivers of these children need ample time to plan for the required postoperative stay if they are not locally based.

Planning for the Future

When planning these corrections, care must be taken to consider the long-term effects of such surgical interventions. Many individuals with skeletal dysplasias, such as multiple epiphyseal dysplasia (MED), spondyloepiphyseal dysplasia (SED), pseudoachondroplasia, diastrophic dysplasia, and Morquio syndrome (MPS IVA), are predisposed to premature osteoarthritis. These patients often need total joint arthroplasty (TJA) at a relatively young age. This must be taken into account when performing osteotomies that may make TJA more difficult. The surgeon needs to consider various obstacles when performing TJA in these patients. For instance, in total knee arthroplasty, customized implants and

constrained designs are often needed because of patient's short size and a lack of ligamentous constraints associated with the underlying pathologies [33, 34]. When considering total hip arthroplasty, the surgeon needs to evaluate the need for potential acetabular augmentation secondary to bony deficiency, the possibility of extensive soft tissue releases because of contractures, presence of tortous and narrow proximal femoral intramedullary canals, and the potential for needing custom implants due to small stature [35]. When proper preoperative planning is executed, good results can be achieved with total joint arthroplasty in patients with skeletal dysplasia, though complication rates can be high [33, 35].

Specific Skeletal Dysplasias and Their Associated Deformities

Achondroplasia

Achondroplasia is the most frequently encountered form of skeletal dysplasia with an incidence of approximately 1 in 10,000 live births [36]. This results from a mutation in the FGFR3 gene. This over-activity of FGFR3 results in abnormalities in endochondral ossification, whereas intramembranous ossification is not affected. This therefore results in involvement of the base of the skull (resulting in foramen magnum stenosis) and the extremities. There are predictable patterns of deformity in this population. In the upper extremity elbow flexion deformities are common and often associated with distal humeral recurvatum. The severe rhizomelia seen in these individuals can result in marked limitation in reach and humeral lengthening can improve function in these patients. In the lower extremity, hip flexion and knee deformity are often noted. The most commonly addressed deformity is genu varum, which is often associated with internal tibial torsion and occasionally genu recurvatum. A common misconception is that uncorrected genu varum in children with achondroplasia results in knee osteoarthritis later in life. In fact, osteoarthritis is rare and the knee joint is usually not a source of functional limitation in these patients. Spine problems such as central stenosis of the lumbar segments often result in limited function. The etiology of genu varum is unclear. Several mechanisms have been proposed, including ligamentous laxity, obesity, asymmetric growth of the proximal tibial physis, and asymmetric growth of the fibula relative to the tibia [37–39]. As previously mentioned the varus deformity can exist in the distal femur, the knee joint, and the proximal and distal tibia. Indications for surgical correction of genu varum and internal tibial torsion in these children include knee pain, progressive deformity, lateral thrust during gait, and rarely, cosmesis. Genu valgum, though uncommon, may also be seen in some patients. However, the valgus deformity is rarely severe enough to warrant surgical correction.

Several techniques for correction of genu varum in achondroplastic patients have been described previously. Fibular epiphysiodesis or shortening may be useful in young patients (5–8 years old) with mild deformity [18, 40]. However, long-term data on this technique is lacking. Acute correction in young children with an oblique proximal tibial osteotomy (Fig. 19.1a–k), simultaneously correcting the tibial varus and internal rotation, has achieved reliable results with a low recurrence rate as long as the joint line and mechanical axis are brought to a neutral position [41, 42].

In older children and adolescents, we prefer gradual correction of the proximal tibia with a circular fixator for severe deformities. Concurrent varus deformity in the distal femur and distal tibia is corrected as needed. The authors tend to use acute correction of these associated deformities. This approach allows for simultaneous correction of multiple sites of deformity as well as multiple planes of deformity [22, 43]. Gradual tensioning of loose lateral collateral knee ligaments has been described but the authors do not use this technique.

When considering isolated distal tibial varus correction, we do a very comprehensive preoperative analysis to confirm that there is no significant proximal tibial deformity. Concurrent untreated proximal tibial varus, even if mild, will result in poor patient satisfaction.

Bifocal tibial osteotomies (Fig. 19.2a–d) can be utilized to correct both the proximal and distal tibial varus deformities and internal rotation and also simultaneously gain length with an acceptable complication risk (mostly equinus contractures with more than 15 % lengthening through the distal tibial osteotomy) [44].

Spondyloepiphyseal Dysplasia Congenita (SEDC)

Spondyloepiphyseal dysplasia congenital (SEDC) results in a short trunk type of disproportionate dwarfism (Fig. 19.3). As the name suggests, both the axial and appendicular skeleton are involved. The genetic cause for this condition involves an abnormality in type II collagen [45]. Lower extremity problems include coxa vara, femoral retroversion, hip flexion contracture, genu valgum, and foot deformities, such as equinovarus and planovalgus. As the genetic defect is in type II collagen, articular cartilage is affected. This results in premature osteoarthritis that manifests in the second to third decade of life [46].

Severe coxa vara with neck-shaft angles less than 90° is often present in patients with SEDC. This deformity can be difficult to judge radiographically because of delayed ossification of the capital femoral epiphysis [47]. We have described the use of the Hilgenreiner-Trochanteric angle for assessment of coxa vara in these children [48]. The epiphyseal deformity of the hip can result in femoral head flattening,

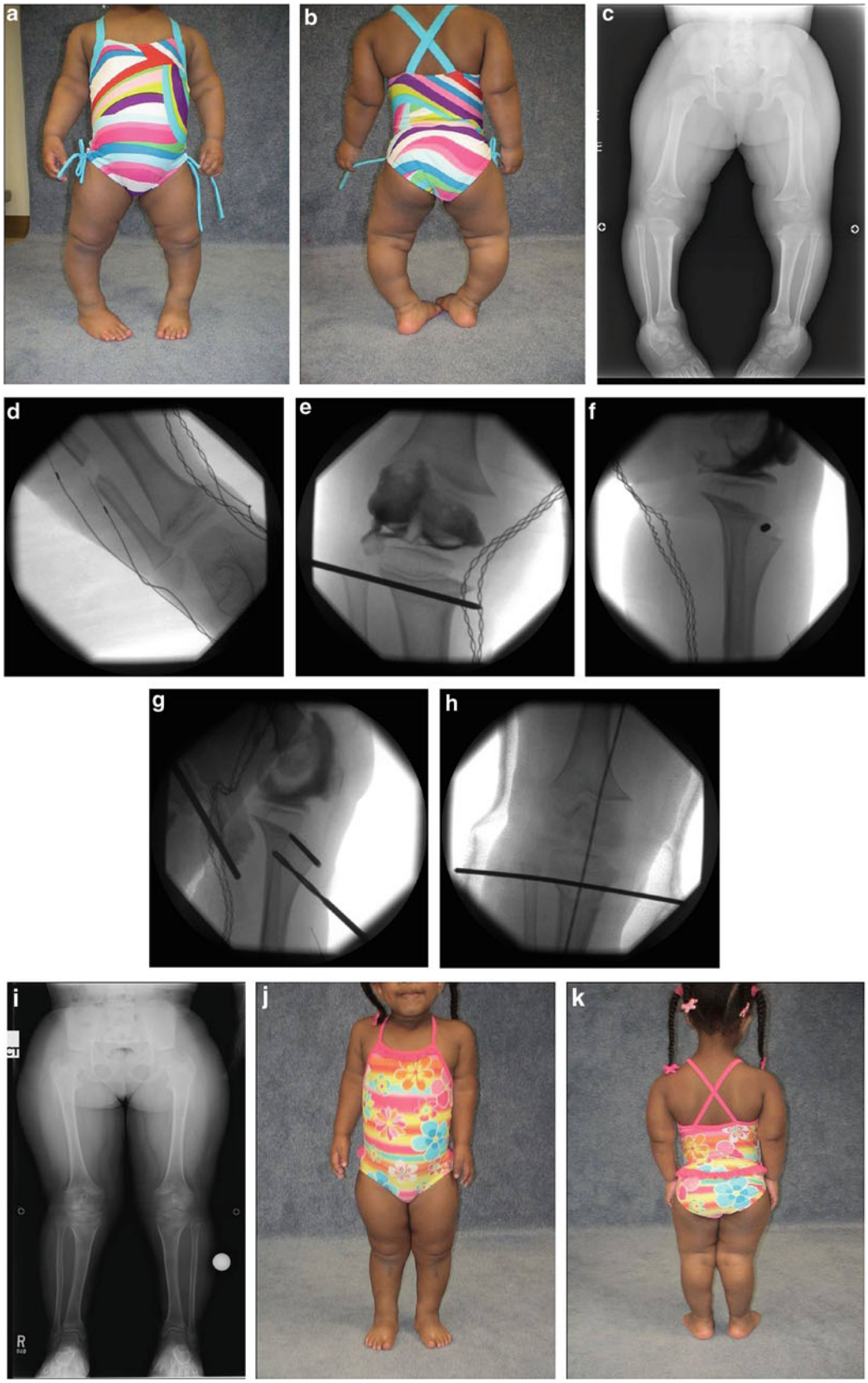


Fig. 19.1 Achondroplasia. (**a**, **b**) Clinical photographs of a patient with achondroplasia with bilateral genu varum and internal tibial torsion. (**c**) Anteroposterior (AP) radiograph of bilateral lower extremities, showing characteristic skeletal changes with genu varum, long fibula. (**d**) Acute correction of proximal tibial varus and internal rotation. The first step is a fibular shortening osteotomy. (**e**, **f**) The next step is an arthrogram to delineate the joint surface and placement of a K-wire parallel to the joint surface. (**g**) Oblique plane osteotomy of the proximal tibia. (**h**) Intraoperative fluoroscopy, confirming adequate correction using the bovie cord technique. (**i**) Postoperative radiographs after healing and cast removal demonstrate excellent alignment. (**j**, **k**) Clinical photographs of the patient in (**a**, **b**) after correction of genu varum deformity

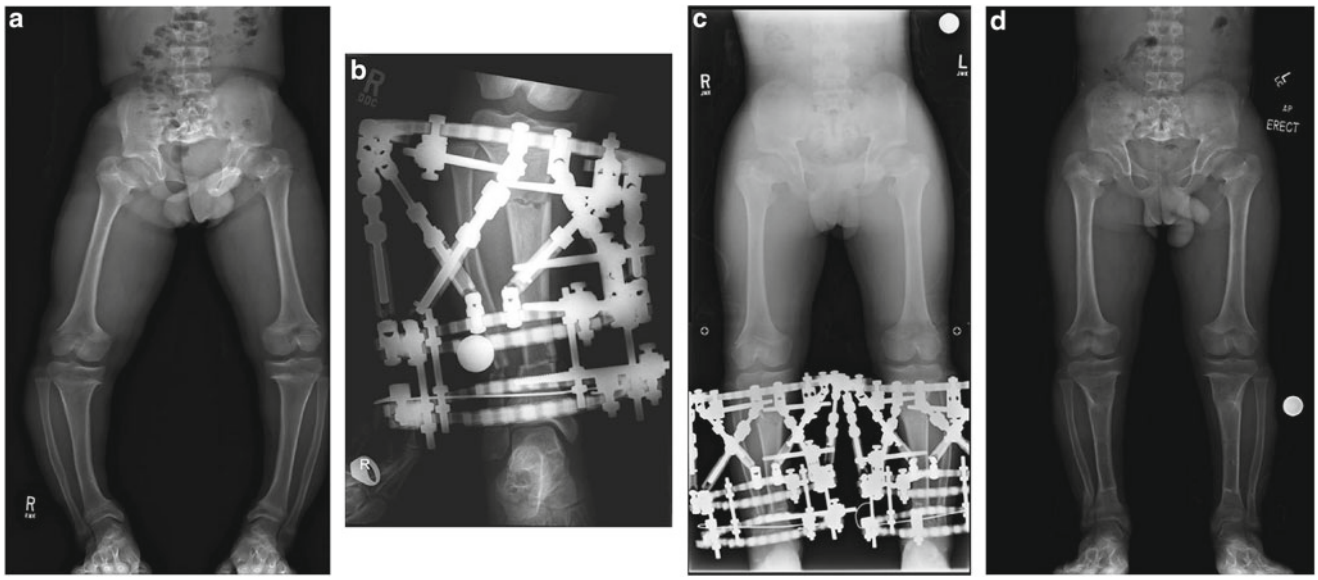


Fig. 19.2 (**a**) Thirteen-year-old boy with achondroplasia. There is varus in both the proximal and distal tibia. (**b**) Bilevel tibial osteotomies with application of a circular frame for gradual correction of the proximal tibial deformity and acute correction of the distal tibial deformity. (**c**, **d**) Anteroposterior radiograph of the lower extremities showing correction of the deformities bilaterally, before and after frame removal



Fig. 19.3 (**a**, **b**) Clinical photographs of a girl with spondyloepiphyseal dysplasia congenita (SEDC) showing characteristic changes such as kyphoscoliosis, lumbar hyperlordosis, pectus carinatum, genu valgum

hinge abduction, and lateral subluxation [47]. Indications for correction of coxa vara include hip pain and reduced function, Trendelenburg gait, a neck-shaft angle of less than 100°, progressive varus, Hilgenreiner-epiphyseal (H-E) angle greater than 60°, and Hilgenreiner-Trochanteric (H-T) angle of less than 0° [48–50]. Correction is performed through a proximal femoral valgus osteotomy. Femoral retroversion and hip flexion contracture (which are almost

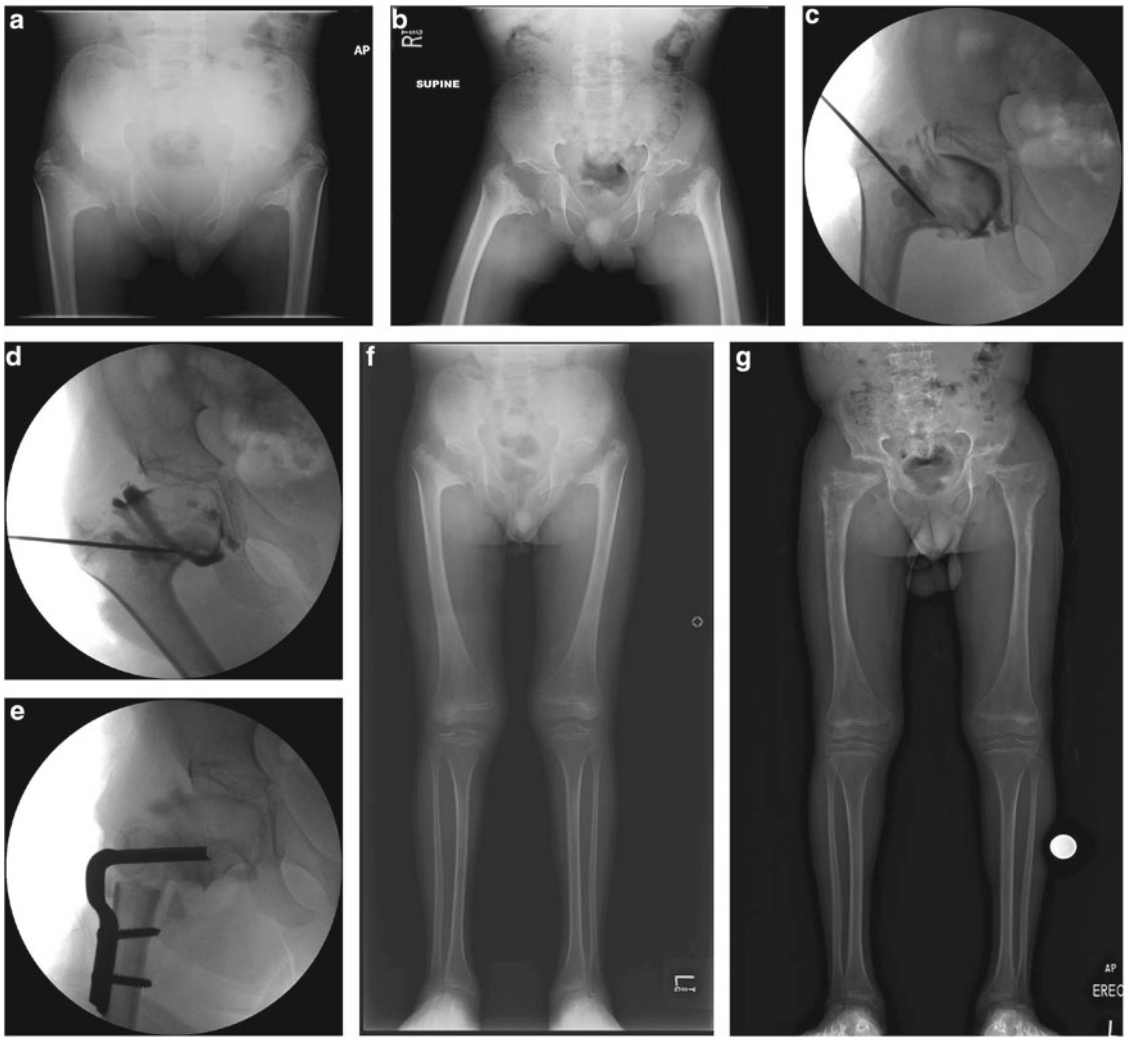


Fig. 19.4 (**a**, **b**) AP and frog leg lateral views of the pelvis in a patient with SEDC show delayed ossification of the head and neck of the femur. The high riding trochanters result in decreased abduction. (**c**) Intraoperative radiographs (arthrogram) demonstrate severe coxa vara. The contrast outlines the cartilaginous femoral head. Guide wire is placed through the trochanter to shut down the trochanteric physis. (**d**) The degree of correction based on the maximum adduction view. (**e**) The appearance after the proximal femoral osteotomy and fixation with a blade plate. (**f**, **g**) Preoperative and postoperative long leg radiographs after correction of hip deformities and removal of hardware

always present) can be addressed concurrently through this osteotomy. Fixation can be achieved with a blade plate or locking plate system (Fig. 19.4a–g). Ideally the fixation is placed across the trochanteric apophysis to achieve physeal closure. If there is a triangular metaphyseal fragment, we try to achieve fixation of that fragment. Radiographic measurements associated with low recurrence of coxa vara after proximal femoral valgus osteotomy are H-E angle less than 38° and H-T angle greater than 13° [48] and progressive proximal femoral epiphyseal ossification. Open reduction of the hip may be necessary at the same surgical procedure, but this is rare. Correction of the hip deformity should precede correction of knee deformity. It is important for patients and their parents to be aware that correction of this proximal femoral deformity may result in an aggravated appearance of genu valgum [51].

Genu valgum is another common deformity seen in SEDC. The valgus can be attributed to the distal femur, proximal tibia, or both segments. The magnitude of genu valgum is often related to the severity of coxa vara. This deformity can often be corrected by distal medial femoral and/or proximal medial tibial hemiepiphysiodesis. If inadequate growth is remaining to achieve correction through growth modulation, then realignment osteotomy at the appropriate level can be performed (Fig. 19.5a–d). This can also address any knee flexion and/or tibial torsion at the same time. Note that the proximal femoral alignment has been corrected prior to final correction of the knee deformity (see Fig. 19.5d).

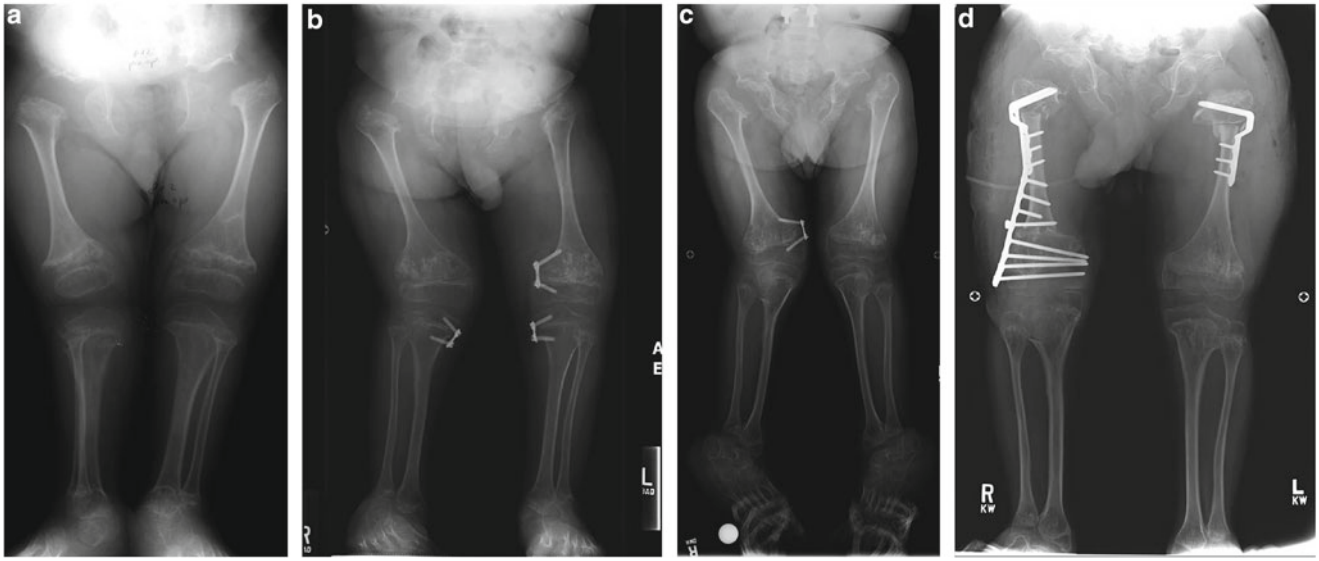


Fig. 19.5 (**a**) Patient with SED with bilateral genu valgum. (**b**) Correction of the genu valgum on the left with growth modulation. (**c**) Residual genu valgum on the right side, despite distal medial femoral hemi-epiphysiodesis. (**d**) Use of a locking plate to achieve appropriate translation during acute distal femoral valgus correction. This patient has also undergone proximal femoral varus correction bilaterally

Symptomatic valgus deformities of the ankle and hindfoot can be treated with supramalleolar tibial and fibular osteotomies and hindfoot procedures as needed. The symptomatic, flexible foot deformities in patients with SEDC are typically managed with orthotics. Stiff equinovarus feet are managed by techniques similar to idiopathic clubfeet.

Multiple Epiphyseal Dysplasia (MED)

Multiple epiphyseal dysplasia is a skeletal dysplasia with wide variations in clinical presentation and skeletal manifestations [52, 53]. It is caused by mutations in multiple genes including those for cartilage oligomeric protein (COMP), type IX collagen, and matrilin-3 and results in symmetric changes of the epiphyses of long bones [52]. Spine involvement is absent in these patients. They have mild short stature and often present with lower extremity joint pain, angular deformities, gait deformities, or stiffness [54, 55]. The epiphyses in these patients are often small and fragmented. Avascular necrosis may complicate fragmentation of the femoral head in up to 50 % of patients with MED [56, 57]. Individuals with MED are predisposed to premature osteoarthritis, particularly in those with severe involvement of the proximal femoral epiphyses [58]. Hip deformity usually mimics that seen in Legg-Calve-Perthes disease. Treatment of these deformities is similar to that for Perthes disease [47].

Joint pain and stiffness is very common in these children. Anti-inflammatory medications, exercise in warm water, stretching, and physical therapy can help to improve these symptoms.

The deformity at the knee is usually genu valgum [59]. Prior to skeletal maturity, this can be addressed effectively with guided growth techniques (Fig. 19.6a–e). After or nearing skeletal maturity, osteotomy at the site of deformity (distal femur and/or proximal tibia) should be undertaken. Ankle valgus can be managed with guided growth as well [20].

Diastrophic Dysplasia

Diastrophic dysplasia is an autosomal recessive condition caused by a mutation in DTDST gene which encodes for a sulfate transporter protein [60, 61]. There is a wide variation in the phenotype of this condition. Common lower extremity bony deformities include hip dysplasia with or without hip dislocation, genu valgum, and rigid foot deformities (Fig. 19.7a, b). Soft tissue problems include hip and knee flexion contractures as well as lateral patellar dislocation [5, 62].

Hips in these patients are notoriously difficult to reduce and reconstruct. They are also difficult to assess radiographically because of a marked delay in ossification of the proximal femur. The severity, deformity, and degree of hip flexion contracture increase with age [63]. The deformities present can include severe acetabular dysplasia, a shortened femoral neck, a wide, irregular femoral head, and (relative) trochanteric overgrowth. These deformities often lead to painful and restricted motion along with hinge abduction. Premature arthritis requiring total joint replacement in young adulthood-middle age can be expected.

Open reduction of these hips is technically difficult because of the severe acetabular dysplasia and short femoral

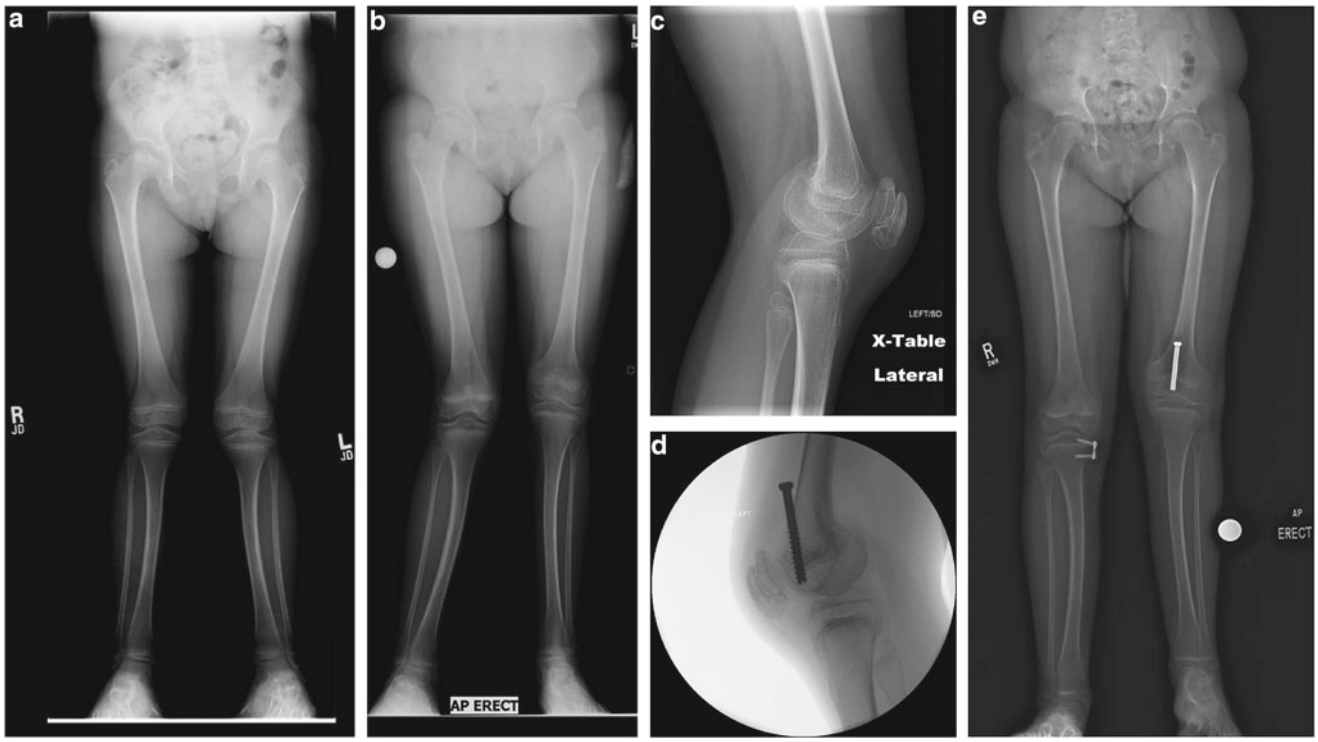


Fig. 19.6 (**a**, **b**) Progressive genu valgum, over 6 years, in a patient with multiple epiphyseal dysplasia. (**c**) Double layered patella is typically associated with patients with multiple epiphyseal dysplasia who have a homozygous mutation of the diastrophic dysplasia sulfate transporter gene (one of the less common, autosomal recessive, variants of MED). This may be a source of symptoms if there is instability between the two fragments. (**d**) Anterior distal femoral epiphysiodesis for correction of distal femoral procurvatum deformity. (**e**) Genu valgum corrected after proximal medial tibial hemiepiphysiodesis

neck. Function with hip dislocation is usually satisfactory in the short term but osteoarthritis and joint replacement is inevitable. Surgical hip dislocation and femoral head reduction have been successful in the short term for painful irregular movement in the authors' experience.

Knee problems in children with diastrophic dysplasia are universal. Genu valgum is present, oftentimes with a laterally dislocated patella (25 % of patients) [5] (Fig. 19.8a, b). Progressive knee flexion contracture is often a clue as to the presence of abnormal patellar mechanics. The authors prefer to reduce the patella at a very young age to allow for remodeling and to alleviate valgus force at the knee. Patellar realignment procedures that we utilize include an extensive lateral release with lengthening of the iliotibial band and biceps femoris with medial plication and a vastus medialis obliquus (VMO) advancement (Fig. 19.9a–f). Medialization of the insertion of the patellar tendon insertion is often required. Anatomical variations include a shallow, irregular intercondylar grove and the medial femoral condyle can be covered with synovium.

Rigid foot deformities are typically seen in diastrophic dysplasia [64]. Deformities described in these children vary from tarsal valgus and metatarsus adductovarus in 43 %, equinovarus and adductus in 29 %, equinus in 8 %, metatarsus adductovarus in 13%, and normal in 7 %. These feet are not similar to idiopathic clubfeet. The main difference is that the navicular is displaced laterally [64]. Serial manipulation and casting using the Ponseti technique can be effective in infancy but these feet often need surgical correction which must be customized to the deformity [65]. Recurrent equinus is the most common long-term complication. The feet are often scarred and further soft tissue release doomed to failure. The authors have been managing equinus with supramalleolar extension tibial and fibular osteotomies (Fig. 19.10a–c). Recurrence in the growing child is common.

Morquio Syndrome

Mucopolysaccharidosis type 4, also known as Morquio syndrome, is an autosomal recessive condition attributable to an abnormality of the *N*-acetylgalactosamine-6-sulfatase (GALNS) gene or the B-galactosidase (GLB1) gene [66–68]. Deficiency of these enzymes leads to accumulation of keratin sulfate and chondroitin sulfate in lysosomes. Onset of clinical symptoms is between 1 and 3 years of age, with severe early growth limitation and genu valgum being the most apparent features. A waddling gait and flat feet are seen around 3 years of age (Fig. 19.11).



Fig. 19.7 (**a**) Child with diastrophic dysplasia with hitch hiker thumbs. (**b**) Rigid clubfeet and hallux varus in a patient with diastrophic dysplasia

The hips have incomplete ossification of the proximal femoral epiphyses and acetabular dysplasia. This leads to progressive hip subluxation and femoral head deformation (coxa valga, coxa magna, and coxa brevis) [69, 70]. The end result of their untreated hip pathology is oftentimes premature osteoarthritis in early adulthood [71].

In order to adequately assess the complex deformities of the hip joint that may warrant operative reconstruction, preoperative computed tomography scanning [6] and intraoperative arthrography are very useful. Isolated proximal femoral varus osteotomies or redirectional acetabular osteotomies have not prevented progressive dysplasia and subluxation. The acetabular bony roof is short with a large limbus. The limbus does not ossify normally, likely secondary to the accumulation of proteoglycans. Realignment procedures include a proximal femoral osteotomy with an acetabular procedure to improve coverage [7, 70, 72]. The authors use a large shelf harvested from the inner wall of the ilium (Fig. 19.12a–d).

Bilateral genu valgum is a prominent feature of Morquio syndrome. This is a result of progressive deformation due to incomplete ossification of the distal femoral and proximal tibial epiphyses, in combination with generalized ligamentous laxity [7]. Intraoperative knee arthrography is very helpful in planning corrective surgery about the knee. A distal femoral and/or proximal tibial realignment osteotomy has been advocated [3, 70]. The authors have found success using guided growth techniques in the young child (preadolescent) (Fig. 19.13a, b) [20].

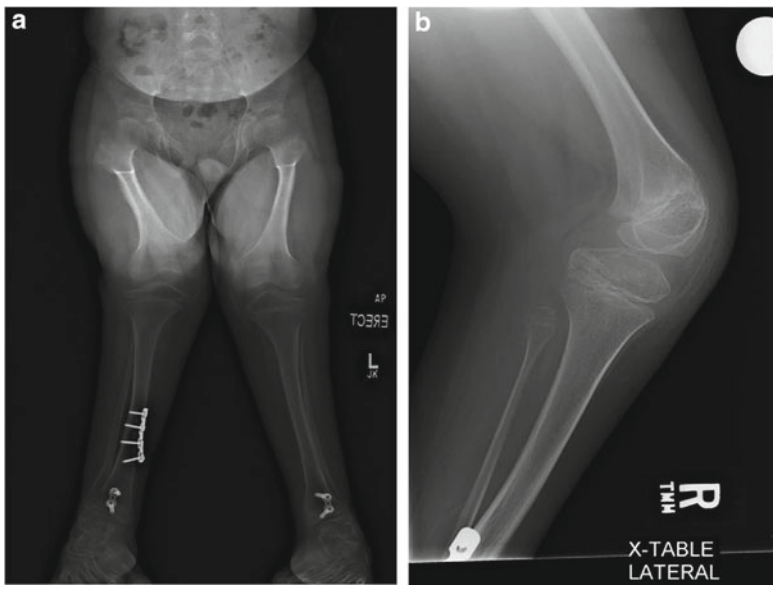


Fig. 19.8 (**a**, **b**) Flexed knees secondary to dislocated patellae in diastrophic dysplasia

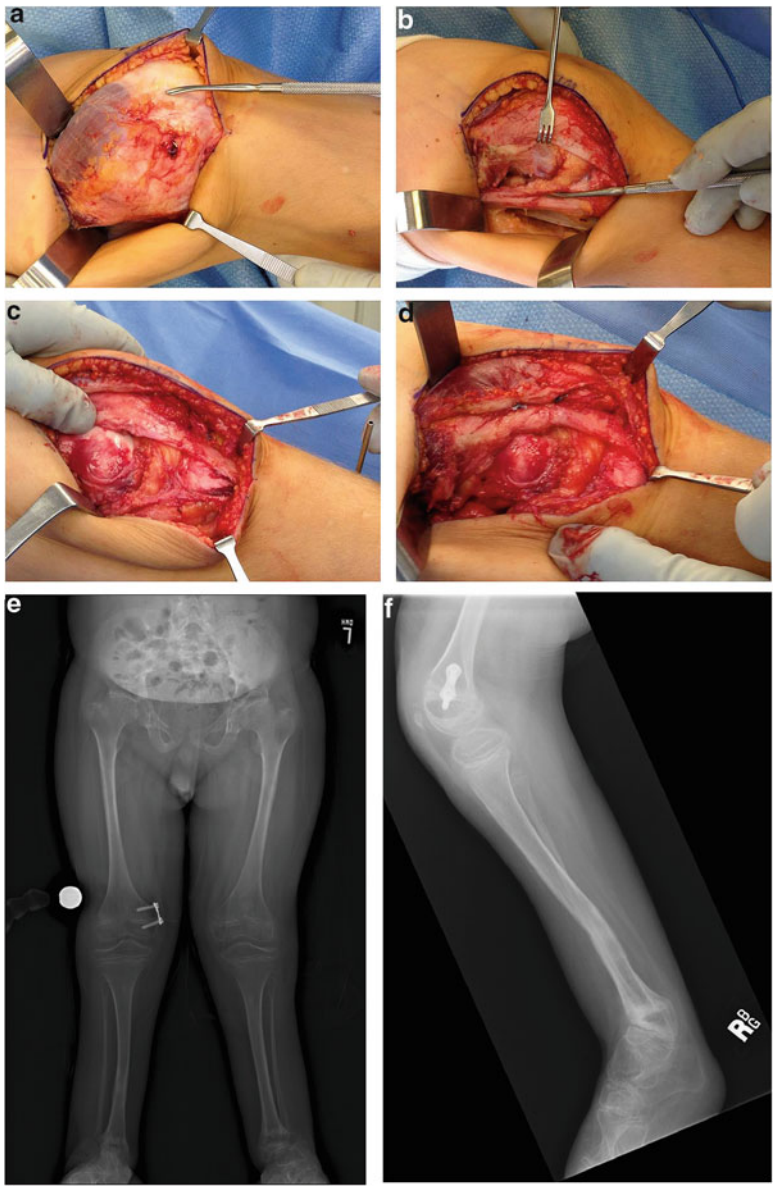


Fig. 19.9 Operative correction of patellar dislocation in a patient with diastrophic dysplasia. (**a**) Patella dislocated laterally with the vastus medialis draped over the anterior surface of the knee. (**b**) Lateral release. Freer elevator pointing to thickened, tight iliotibial band. (**c**) Patella relocated after extensive lateral release. (**d**) Patella stabilized with vastus medialis obliquus advancement. (**e**, **f**) Postoperative radiographs demonstrating correction of genu valgum with growth modulation and the patella is appropriately located on both views

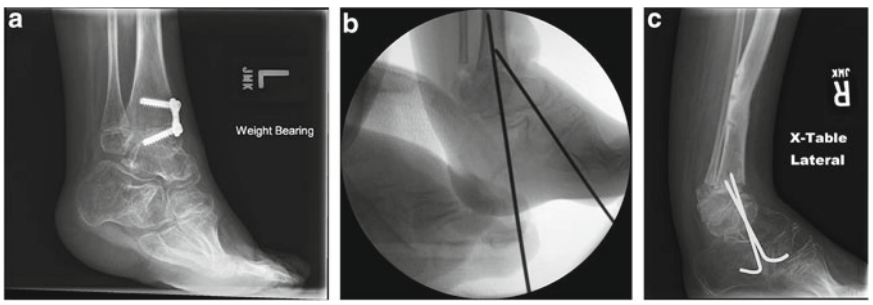


Fig. 19.10 (**a**) Rigid equinovarus foot in a patient with diatrophic dysplasia. A previous attempt at anterior hemiepiphysiodesis had been made at an outside facility, without success. (**b**) Patient was treated with supramalleolar osteotomies with acute correction. Note the fibular shortening and posterior translation of the distal fragment. (**c**) Healing in plantigrade position after acute correction. These patients are at a high risk of recurrence

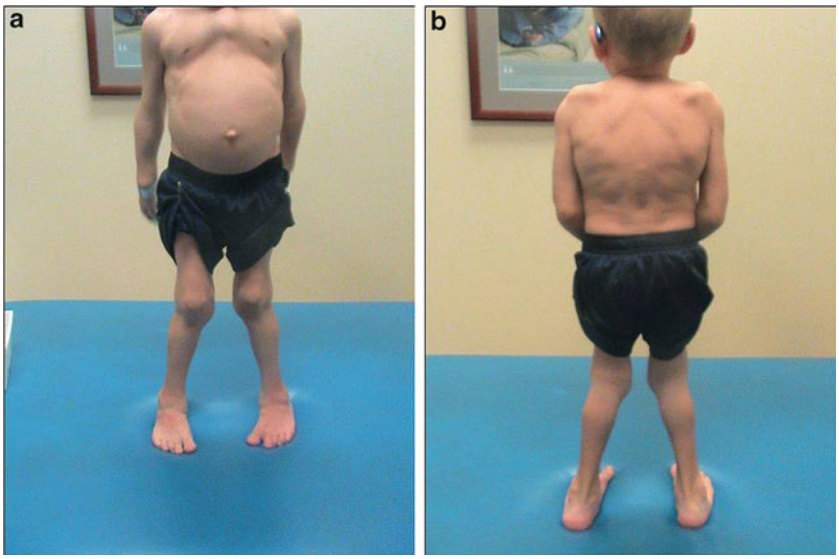


Fig. 19.11 (**a**, **b**) Clinical photographs of a boy with Morquio syndrome showing the characteristic pectus carinatum, thoracolumbar kyphosis, genu valgum, planovalgus feet

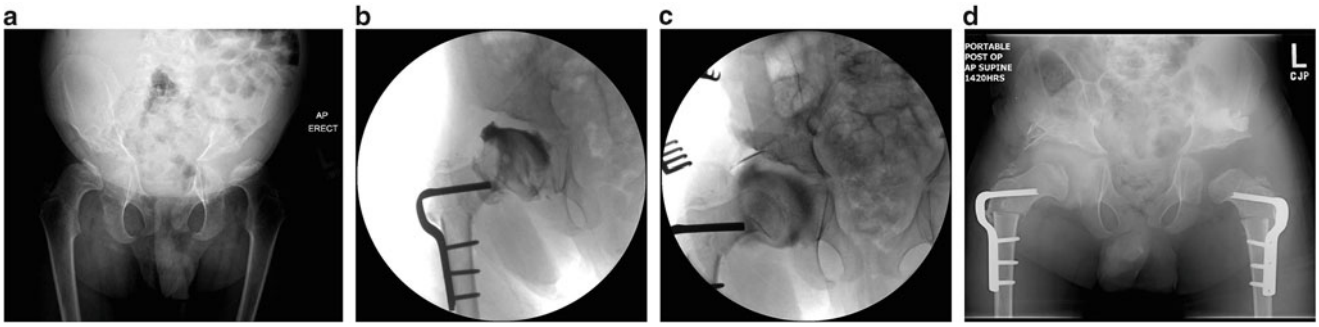


Fig. 19.12 (**a**) AP pelvis radiograph in a patient with Morquio syndrome, demonstrating coxa valga, subluxated hips, small epiphyses, and dysplastic acetabule. (**b**) Arthrogram shows residual acetabular dysplasia and femoral head uncoverage after proximal femoral varus osteotomy. (Large unossified cartilaginous labrum/acetabular anlage makes a Pemberton osteotomy less appealing.) (**c**) Improved coverage with shelf acetabuloplasty. (**d**) Postoperative AP pelvis radiograph demonstrates good femoral head coverage bilaterally with a shelf on the right and a Pemberton osteotomy on the left

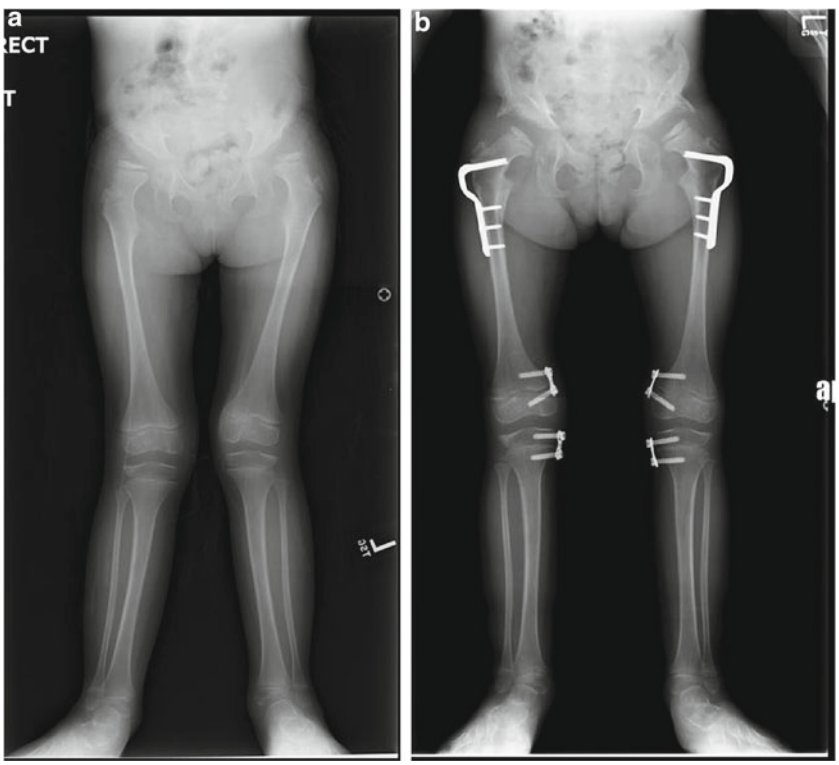


Fig. 19.13 (**a**, **b**) Genu valgum correction in a patient with Morquio syndrome using growth modulation. Recurrence may be seen in younger patients

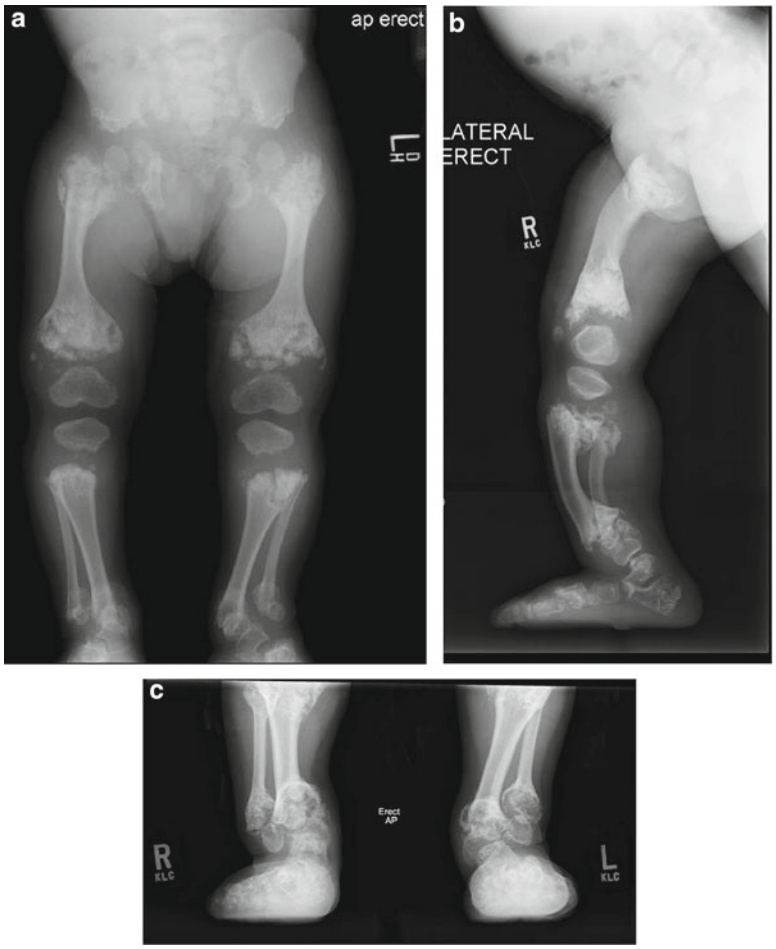


Fig. 19.14 (**a**–**c**) Lower extremity deformities in a patient with Jansen metaphyseal dysplasia. Characteristic severe widening of the physes and bowing of long bones are seen

The pes planovalgus feet seen universally in those children with Morquio syndrome can usually be managed very well using functional foot orthoses. If conservative treatment with these orthoses fails, lateral column lengthening or subtalar fusion in combination with medial column reconstruction is an option [7]. However, long-term results of these procedures are not known.

Metaphyseal Chondrodysplasia

These are a group of dysplasias resulting from failure of uniform ossification of the cartilage columns, with persistence of cartilage islands, underdevelopment of the cartilage, resulting in deformities of the affected bones. The epiphyses are normally preserved and the metaphyses are the most severely affected. The differential diagnoses include rickets and hypophosphatasia. These are usually associated with genu varum, ankle varus, and coxa vara of variable severity. There are three major types of metaphyseal chondrodysplasia.

- *Schmid type* is the most common type of metaphyseal chondrodysplasia. It is an autosomal dominant condition which results from a heterozygous mutation of the COL10A1 gene on chromosome 6q21-q22.3. This causes a defect in the α1 chain of type X collagen (which is essential for calcification of the hypertrophic zone of chondrocytes) [73].
- *McKusick type* of chondrodysplasia (also known as *cartilage hair hypoplasia*) is an autosomal recessive disorder due to a defect in the RMRP gene on chromosome 9p13.3. This results in milder genu varum and coxa vara and is also associated with decreased immunity, varying degree of anemia, and characteristic cone-shaped epiphyses in the phalanges [74, 75].
- *Jansen type* (Fig. 19.14a–c) is a rare but severe, autosomal dominant, form of chondrodysplasia resulting from a mutation (at 3p21.31) resulting in ligand independent activation of parathyroid hormone related protein receptor I (PTHRP1). This results in hypercalcemia and hypophosphatemia [76, 77].

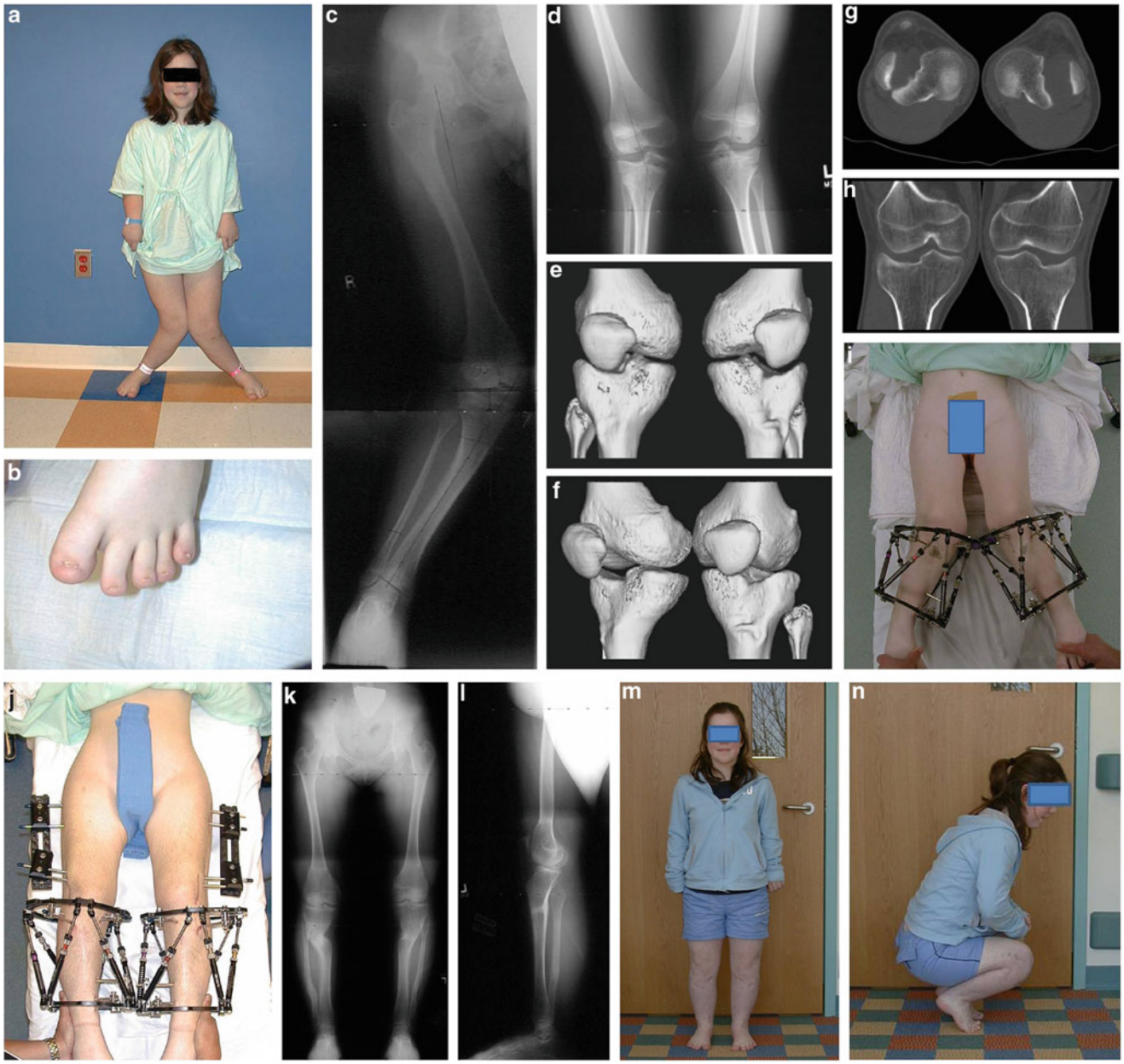


Fig. 19.15 Ellis-van Creveld syndrome (EvC). (**a**) Clinical photograph of a girl with EvC showing severe genu valgum. (**b**) Characteristic dystrophic nails seen in EvC, also called chondro-ectodermal dysplasia. (**c**) AP radiograph of the right lower extremity demonstrates multifocal valgus in the lower extremity. (**d**) AP radiograph of both knees demonstrates characteristic intra-articular deformity with depression of the lateral tibial plateau. (**e**–**h**) CT scans (axial and coronal views as well as 3D reconstructions) highlight the depression of the antero-central portion of the lateral tibial plateau. (**i**, **j**) Clinical photographs after application of a Taylor Spatial Frame™ and then after correction of distal femoral valgus as well as a lateral release for correction of patellar instability. (**k**, **l**) Postoperative AP and lateral radiographs demonstrating restoration of the mechanical axis after distal femoral and proximal tibial correction. (**m**, **n**) Clinical photographs after correction demonstrating improved alignment as well as function

Treatment of deformities in these patients varies by type and severity. Patients with Schmid type dysplasia may show some spontaneous improvement in the first decade but persistent or severe deformities may need correction using guided growth or osteotomies. Patients with McKusick type usually have mild deformities and rarely need treatment. Patients with Jansen type dysplasia often have severe deformities and need earlier and more aggressive treatment and also have the greatest risk of recurrence.

Ellis-van Creveld Syndrome (EvC) or Chondro-ectodermal Dysplasia

Ellis-van Creveld (EvC) syndrome is a rare autosomal recessive skeletal dysplasia that is characterized by short ribs and disproportionate short stature, ectodermal dysplasia (including characteristic nail and tooth changes), postaxial polydactyly, and congenital cardiac defects (most commonly atrial septal defects) (Fig. 19.15a–n). This results

from a homozygous mutation of the EvC gene located on chromosome 4p16. Orthopedic issues include postaxial polydactyly, genu valgum, patellar instability, as well as a characteristic depression of the lateral tibial plateau. The genu valgum in these patients comes from both the distal femur and the proximal tibia and frequently needs correction on both sides of the knee. The accompanying external tibial torsion, which can be fairly severe, makes the use of guided growth less appealing. Correction of angular deformities (often gradual), and soft tissue releases, including peroneal nerve decompression are important components of correcting and maintaining appropriate patellar tracking [78].

Summary

Lower limb deformities are a common source of morbidity and pain in patients with certain skeletal dysplasia. These patients need a comprehensive evaluation of both their skeletal and their extraskeletal abnormalities by a team of providers who are familiar with these issues. Correction of limb deformities is frequently needed in these patients and despite adequate correction, recurrences are fairly common. Administering general anesthesia can be quite a challenge in these patients and appropriate equipment and expertise are paramount to safely performing their orthopedic procedures. Prolonged procedures, especially in patients with severe kyphosis, may place the spinal cord at risk for stretch and neuromonitoring must be considered during these procedures (even if they do not involve the spine). Combining procedures to minimize the episodes of anesthesia should be strongly considered to decrease anesthesia risk. The cervical spine should be carefully evaluated preoperatively, especially in patients at risk for cervical instability (most dysplasias except achondroplasia). Lower limb deformities should be carefully evaluated as most are multifocal and multiplanar. Appropriate equipment and size appropriate fixation devices must be available during deformity correction surgery for these patients. Deformity correction in these patients can offer relief of symptoms and improvement in their mobility. Premature arthritis may be seen in some dysplasias and methods of deformity correction should not compromise the subsequent arthroplasty once the joint is irreversibly damaged.

Enzyme replacement and bone marrow transplant have shown promise in treatment of some dysplasias (particularly the mucopolysaccharidoses). Improvement in understanding of the molecular mechanisms and pathways involved in the genesis of dysplasias may enable us to intervene at a molecular level early on and hopefully prevent/blunt some of the musculoskeletal manifestations of the disease and hopefully this will translate into less morbidity and better quality of life for our patients.

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Lower Extremity Benign Bone Lesions and Related Conditions

20

Lori Karol

Introduction

Benign bone lesions comprise a group of diseases where the presence of abnormal tissue within or on the surface of the growing bone may create skeletal deformities of the lower limb. In conditions such as fibrous dysplasia, loss of mechanical strength of the bone can lead to angular deformity or pathologic fracture. In the growing child, angular deformity may also result from abnormal physeal growth as seen in multiple hereditary exostoses. Limb length discrepancy may result from disturbances in physeal function as seen in such conditions as enchondromatosis. This chapter will discuss the most common pathologic bony lesions seen in children that are associated with lower limb deformity.

Nonossifying Fibroma

A nonossifying fibroma is the most common benign bone lesion found in children. It is a solitary lytic lesion which is characterized by replacement of the normal bone with bland fibrous tissue. The lesions are usually asymptomatic and are diagnosed either coincidentally on a radiograph obtained following an injury or at the time of pathologic fracture [1, 2]. They are usually diagnosed in the first two decades of life [3].

Nonossifying fibromas appear as lytic lesions that are eccentric in the metaphyseal regions of growing bones (Fig. 20.1a). They may appear lobulated, have distinct sclerotic margins, and do not create periosteal reaction. They are contained within the bone and do not typically expand the bone but do erode the cortex from within. The appearance within the lesion is lytic, without spicules of calcification. They are most often solitary. They occur most frequently in the long bones, with the most common site being the medial aspect of the distal femur.

Biopsy is often not needed since the radiographic appearance is characteristic and does not resemble malignancy. Specimens from nonossifying fibromas show sheets of bland fibroblasts with small nuclei without pleomorphism.

Treatment of nonossifying fibromas is nonsurgical in the vast majority of patients. Size greater than 50 % of the width of the bone and greater than 3.3 cm in length has been shown to be predictive of pathologic fracture, but smaller lesions have been noted to lead to stress fractures too, albeit rare [4, 5]. Pathologic fractures heal with immobilization and may lead to resolution of the lesion. If the nonossifying fibroma is in a weight-bearing bone, is sufficiently large, and has undergone pathologic fracture, curettage and bone grafting can eradicate the lesion and allow for full athletic participation [6, 7] (see Fig. 20.1b) The natural history of nonossifying fibromas is that they regress during adulthood.

Box 20.1. Nonossifying Fibroma
Age at presentation: 1st or 2nd decade.
Location: Eccentric metaphyis.
Number: solitary.
Presenting symptoms: usually none, occasional fracture.
Rx: none unless large.
Malignant degeneration: none.

Fibrous Dysplasia

Fibrous dysplasia is a noninherited condition defined by the replacement of normal trabecular bone by fibrous tissue and immature disorganized osteoid [8]. When monostotic, the condition shows no gender predilection. Fibrous dysplasia may be diagnosed in childhood, adolescence, or adulthood.

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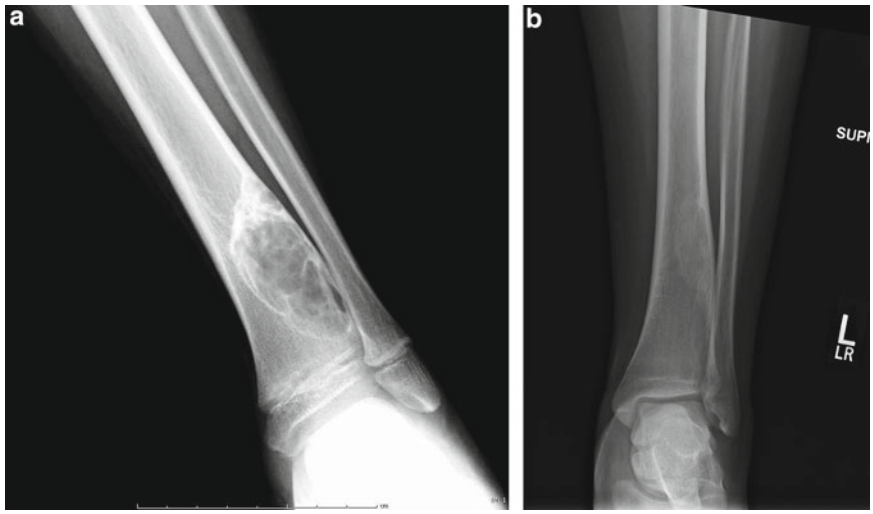


Fig. 20.1 (**a**) Radiograph of the ankle of a 7-year-old boy with ankle pain. Note the eccentric lobulated contained lesion which thins the surrounding cortex. Biopsy at the time of surgery confirmed the diagnosis of a nonossifying fibroma. (**b**) Radiograph 8 years following curettage and bone graft shows resolution of the lesion. The boy is asymptomatic and participates in contact athletics

Lesions may be present in one bone, known as monostotic fibrous dysplasia, or in many bones, known as polyostotic fibrous dysplasia. When associated with endocrine abnormalities such as precocious puberty, the diagnosis of McCune Albright syndrome is made. Mazabraud's syndrome is the association of fibrous dysplasia with muscular myxomas and is even more rare. Polyostotic fibrous dysplasia is more common in girls than in boys, especially when associated with endocrinopathy.

Fibrous dysplasia and McCune Albright syndrome have been found to result from a somatic mutation in the guanine nucleotide-binding protein, Alpha-stimulating activity polypeptide gene, known more succinctly as the Gnas pathway [9, 10]. Because this is a somatic mutation, the disease is not hereditary. This mutation is specific to fibrous dysplasia and is not seen in other lytic processes of bone.

Presenting symptoms of fibrous dysplasia include limp, pathologic fracture, or bone pain [11]. Patients may present with angular deformity less frequently. Those children with McCune Albright syndrome present with precocious puberty, which usually manifests as painless vaginal bleeding in young girls. They also have characteristic café-au-lait spots on their skin, present from birth or infancy, with irregular borders described as "coast of Maine" (Fig. 20.2). Other endocrinopathies associated with McCune Albright syndrome include hyperthyroidism, growth hormone excess, and Cushing's disease [12, 13].

The radiographic appearance of fibrous dysplasia is one of one or more ill-defined lytic lesions. The disease may be present in the metaphysis or diaphysis of the bone, but is typically not epiphyseal. The margins of the lesions are somewhat indistinct, lacking the sclerotic rim frequently present in patients with simple bone cysts. Owing to the lack of margins, the lesions of fibrous dysplasia are described as "flame-shaped," tapering off into the normal bone. As the bony trabeculae are replaced by fibrous tissue, the appearance within the lesions is blurred, opaque, and gray on radiographs. This has been termed "ground glass" appearance. The lesions can be expansile and may thin the cortices of the native bone. They do not create a periosteal reaction (Fig. 20.3). Large lesions in weight-bearing bones may result in bowing of the bone, with coxa vara of the femur and anterior bowing of the tibia being most common.



Fig. 20.2 Clinical photograph of a girl with McCune Albright syndrome. Note the skin macule which is hyperpigmented and has irregular borders. The child has spinal involvement and scoliosis

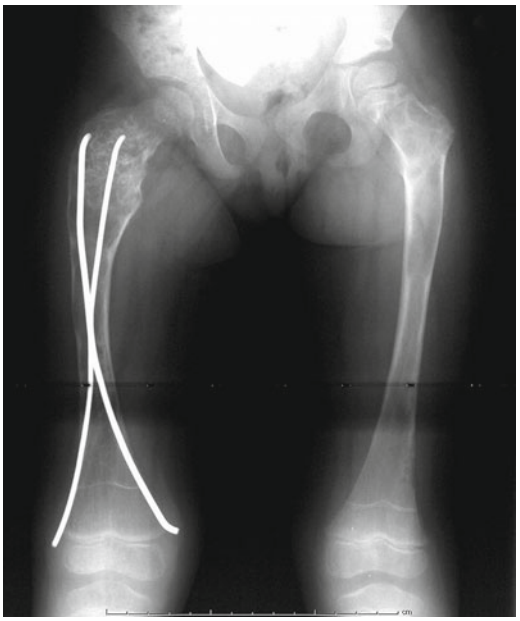


Fig. 20.3 Anteroposterior radiograph of a 3-year-old female with polyostotic fibrous dysplasia. Poorly defined lytic areas are present throughout both femora and the right hemipelvis. The right femoral neck is expanded, and an early shepherd's crook deformity is present. Intramedullary fixation is present due to previous pathologic fracture

The most common sites for fibrous dysplasia are within the long bones, but any bone may be involved. Vertebral involvement is seen less frequently. The skull may be affected, which can lead to disfigurement over time.

The differential diagnosis in patients with monostotic fibrous dysplasia includes simple and aneurysmal bone cysts, eosinophilic granuloma, and when the tibia is involved, osteofibrous dysplasia. Lesions due to fibrous dysplasia can usually be differentiated from bone cysts as the margins of the lesion are less distinct, there is no fallen leaf sign (seen in patients with simple cysts following pathologic fractures), and the matrix radiographically is more opaque in fibrous dysplasia. Upon presentation, it has been recommended to obtain a technetium bone scan to identify the presence of polyostotic disease. By the age of 6 years, it has been found that all affected sites can be identified on bone scan [11]. Due to the lack of need for treatment of asymptomatic lesions, it has not been the practice of my institution to obtain a bone scan for identification of all bony lesions.

The diagnosis is usually made both clinically and radiographically (especially in the polyostotic form); therefore biopsy is only rarely required. When biopsy tissue is obtained, firstly, the lesion in fibrous dysplasia is filled with tissue rather than fluid on aspiration, differentiating it from bone cysts. On gross examination, tissue from fibrous dysplasia lesions appears firm and gray. There may be areas that appear cystic. The specimen will feel gritty to palpation. Microscopic examination shows the abundant fibrous tissue with limited cellularity. Islands of immature osteoid are present, but do not form mature trabeculae. These spicules of immature bone take on abnormal irregular shapes, which have been termed "Chinese Letters." The islands of osteoid are not rimmed in osteoblasts, in distinction to osteofibrous dysplasia. Giant cells and osteoclasts may be scattered in the microscopic field.

The orthopedic management of fibrous dysplasia is aimed towards correcting deformity and the treatment of pathologic fractures. The natural history of fibrous dysplasia in the skeletally immature patient is that of slow expansion of the bony lesions. Deformity of weight-bearing bones appears as the mechanical strength of the bone is lost due to progressive replacement with fibrous tissue. In polyostotic disease, gait becomes more difficult, with the median age at which patients require assistive devices to walk being 7 years [14]. Characteristic deformities include coxa vara due to the "shepherd's crook" deformity of the femur, angular deformity at the knee, and procurvatum of the tibiae. Pain in the extremity may occur as the bone weakens and deforms secondary to microfractures that occur due to the stress of weight bearing. Limb length discrepancy may develop.

Bony lesions do not require operative intervention unless associated with pathologic fracture or deformity. Curettage and bone grafting of fibrous dysplasia leads to recurrence of the lesions, and the bone graft (allograft or autograft) is eventually replaced again by the fibrous tissue characteristic of fibrous dysplasia. Cortical bone grafts have been used, with hopes of less resorption and greater improvement of mechanical strength of the bone [15]. Unfortunately, these grafts also resorb, although in a more delayed fashion compared to morselized bone graft.

Pathological fractures occur most frequently in the first decade of life between the ages of 6 and 10 years [16]. The femur is the most common bone to sustain a fracture. Fractures occurring through dysplastic bone may be treated nonoperatively in the upper extremity, but usually benefit from surgical treatment in the lower extremities. Load-sharing intramedullary implants allow earlier weight bearing, which is important due to the underlying bone fragility (Fig. 20.4) [17–19]. Deformity should be fully corrected, as residual angulation will predispose to future fractures.

The shepherd's crook deformity of the proximal femur is particularly challenging to treat [15, 20]. Progressive coxa vara leads to a waddling gait, hip pain, and eventual pathologic stress fractures when the neck-shaft angle is less than 120°. Proximal femoral valgus osteotomy can correct the deformity, but fixation in the dysplastic bone is extremely challenging. Plate fixation frequently leads to pathologic fracture at the ends of the plate where stress concentration occurs or to loss of fixation due to screw pullout. Hip screw and side plate fixation is typically performed; however the screw over time may migrate out of the femoral neck as the

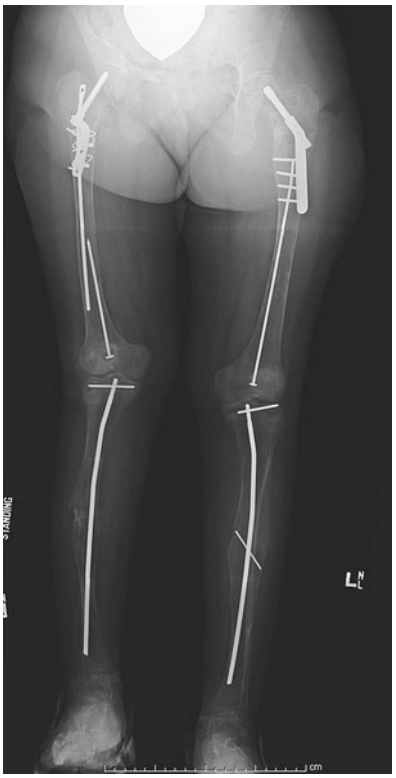


Fig. 20.4 Anteroposterior radiograph of the lower extremities of an 11-year-old girl with McCune Albright syndrome. She has undergone intramedullary fixation of the femora and tibiae as treatment of repetitive pathologic fractures. Note the proximal femoral fixation for treatment of coxa vara and shepherd's crook deformities. There is also a leg length discrepancy

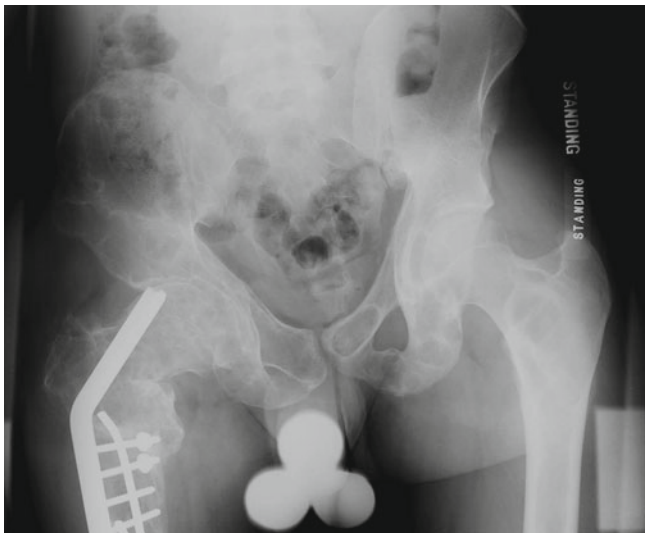


Fig. 20.5 Pelvis radiograph of a 21-year-old male who has undergone multiple surgeries for treatment of right femoral fractures and coxa vara. Note the expansion of the femoral neck, the lytic appearance of the femoral neck and head, and the superior migration of the implant in the femoral neck. There is shortening of the length of the limb on the right

dysplastic bone remodels and the deformity recurs. Our preference is to overlap a hip screw and side plate device with an intramedullary flexible nail, with the goal of achieving reasonable proximal femoral fixation to allow the osteotomy to heal, yet providing load-sharing distal to the device by virtue of the intramedullary nail. Customized proximal femoral nail-screw devices have been used as well, but again the proximal femur typically remodels despite the presence of the implant (Fig. 20.5).

Osteotomies for deformity correction require meticulous preoperative planning. The deformities are typically multiplanar, and more than one osteotomy may be required for passage of intramedullary devices. There is frequently no patent intramedullary canal in the dysplastic bone, so insertion of flexible or rigid nails must be undertaken carefully to avoid penetration with the nail or further pathologic fracture. The intramedullary canal may require drilling to allow nail passage. Additionally, large amount of blood loss should be anticipated and prepared for, as these lesions are hypervascular. Unfortunately, skeletal growth leads to recurrent deformity, and revision surgery should be expected. Progression of the fibrous dysplasia can lead to enlargement of the bone, recurrent angulation, and increasing bone pain due to bone fragility (Fig. 20.6a, b).

Nonunion is infrequent in fibrous dysplasia. Bony callous can develop, but the bone softens with time due to replacement with fibrous tissue.

Limb length discrepancy is seen most frequently in children with severe polyostotic disease. Lengthening is usually fraught with complications due to the poor mechanical properties of the dysplastic bone. Contralateral epiphysiodesis may be offered, but is often rejected by some families due to the patient's underlying short stature.

Scoliosis is seen with greater frequency in patients with fibrous dysplasia [21, 22]. Vertebral involvement with scoliosis has been found in one-third to over 50 % of patients with polyostotic disease. As would be expected, spinal fusion surgery may be difficult due to the bony fragility and excessive bleeding. Yet successful results of posterior spinal fusion have been published [22]. The efficacy of scoliosis bracing in fibrous dysplasia has not been proven, and the concurrent presence of rib lesions precludes its use in most patients.

Medical treatment with bisphosphonate agents has been used in patients with polyostotic fibrous dysplasia [23–26]. Bisphosphonate use has not been shown to affect the bony lesions in fibrous dysplasia, but has been found to decrease the pain patients experience. New clinical studies using receptor activator of nuclear factor kappa-B ligand (RANKL) inhibitors have shown promise in influencing the progression of bone disease [27]. Patients with McCune Albright syndrome require referral at the time of diagnosis to pediatric endocrinologists for medical management [28, 29]. Those children with



Fig. 20.6 (**a**) AP radiograph of the lower extremities of a 9-year-old girl with polyostotic fibrous dysplasia. She has undergone a right proximal femoral allograft replacement due to marked bony enlargement suspicious for malignancy, a left proximal femoral osteotomy, and right tibial osteotomy with intramedullary fixation. (**b**) Two years later, the tibia has undergone great expansion and angular deformity has worsened with recurrent bone pain with weight bearing

skull involvement should be referred as well to craniofacial specialists, as bony enlargement and overgrowth can lead to cosmetic disfigurement and optic foraminal sclerosis may progress to blindness.

The development of cystic lesions most closely resembling aneurysmal bone cysts may occur within the dysplastic bone. Rapid enlargement of the lesions coupled with increasing pain is usually present. Malignant degeneration has been documented in rare patients with fibrous dysplasia [30, 31]. Malignant degeneration is most common in adulthood and has been associated with previous irradiation, which is now contraindicated in patients with fibrous dysplasia. Painful enlargement of the dysplastic bone may also be present. In such instances, evaluation by an orthopedic oncologist should be sought.

Box 20.2. Fibrous Dysplasia

Age at presentation: 1st or 2nd decade.
Location: Long bones most frequent, metaphyis or diaphysis.
Number: solitary or polyostotic.
Presenting symptoms: limp, pathologic fracture, bone pain.
Rx: intramedullary fixation of fractures, deformity correction, bisphosphonates for pain.
Malignant degeneration: rare in adulthood.

Osteofibrous Dysplasia

Osteofibrous dysplasia, also known as Campanacci's disease, is a peculiar bone dysplasia which is typified by both sclerotic and lytic lesions most commonly present in the tibia in children and adolescents [32, 33]. It is usually not genetically transmitted. Osteofibrous dysplasia is bimodal in presentation. It can present in early infancy with apparent anterior bowing in the tibia [34, 35]. In such cases, it must be distinguished from anterolateral bow of the tibia as associated with congenital pseudarthrosis of the tibia and neurofibromatosis. The fibula in patients with infantile osteofibrous dysplasia is usually uninvolved, and pathologic fracture is rare. The condition is nearly always unilateral and does not involve the rest of the skeleton.

The second form of osteofibrous dysplasia presents in late childhood or adolescence. It also is most common in the tibia and involves only one bone. It has been genetically linked to adamantinoma.

Osteofibrous dysplasia is not typically genetically transmitted, but a rare form has been published in which several family members were affected [36, 37].

Radiographs of osteofibrous dysplasia show bubbly lytic lesions scattered throughout the anterior diaphysis of the tibia [38]. There is usually anterior bowing, and the limb may be somewhat shorter than the contralateral leg. The cortex may appear thickened in areas (Fig. 20.7).

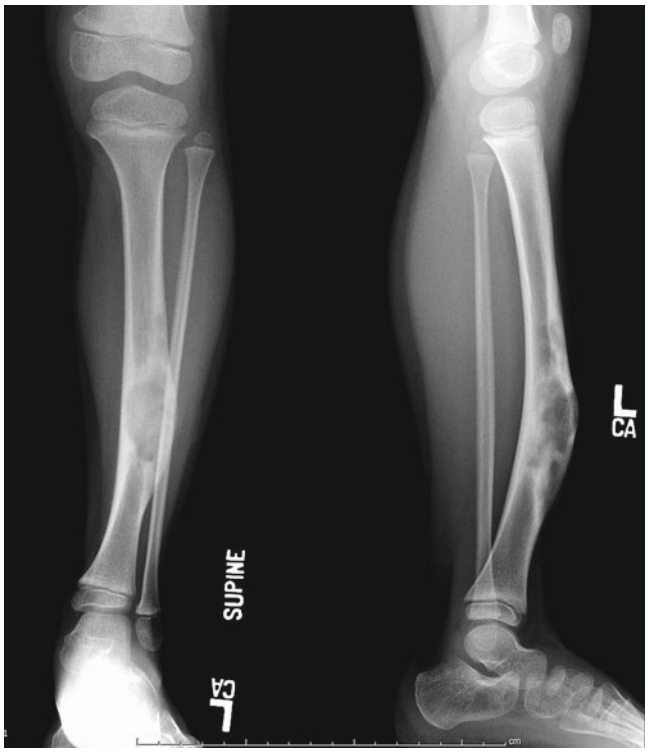


Fig. 20.7 AP and lateral radiograph of a 7-year-old female with progressive anterior bowing of the tibia. The lateral view shows loculated lytic lesions in the anterior cortex of the tibia typical for osteofibrous dysplasia

The differential diagnosis of osteofibrous dysplasia includes monostotic fibrous dysplasia, congenital pseudarthrosis of the tibia, and adamantinoma.

Pathological examination of tissue from osteofibrous dysplasia shows fibrous tissue with areas of immature osteoid, as is seen in fibrous dysplasia. The histological distinction from fibrous dysplasia is that the osteoid is rimmed in osteoblasts in osteofibrous dysplasia, which is its characteristic histologic finding. Cells stain positively for cytokeratin, which is also present in adamantinoma, supporting a relationship between the two lesions [38–42].

Treatment of osteofibrous dysplasia is controversial. Spontaneous resolution of osteofibrous dysplasia in young babies has been described, so treatment in these infants should be observation in the absence of pathologic fracture [43]. If fracture does occur, intramedullary fixation such as is performed in congenital pseudarthrosis of the tibia, combined with bone grafting the fracture site, usually is successful in obtaining and maintaining union [44].

Older children with osteofibrous dysplasia have undergone various forms of treatment. Observation of radiographically stable lesions may be chosen when the patient is asymptomatic. Marginal resection with curettage has been shown to lead to recurrence and is not recommended. Those surgeons who have concern that osteofibrous dysplasia may evolve into malignant adamantinoma have performed aggressive resection of the affected tibial area, with reconstruction as needed. Bone segment transport with proximal corticotomy and external fixation and treatment with free vascularized fibular graft have met with success in very rare instances [45, 46].

Box 20.3. Osteofibrous Dysplasia
Age at presentation: infantile or 1st or 2nd decade.
Location: anterior tibial diaphysis.
Number: solitary.
Presenting symptoms: bowing, enlargement, pain.
Rx: controversial, possible en bloc excision.
Malignant degeneration: associated with adamantinoma.

Enchondromatosis

Enchondromas are benign hyaline cartilaginous lesions present within the metaphysis. Enchondromas may be solitary or multiple, in which case the condition is named Ollier's disease. Solitary enchondromas are common and rarely symptomatic, where Ollier's disease is rare. Maffucci's syndrome is defined as the presence of multiple enchondromas with soft tissue hemangiomatosis and carries a much bleaker prognosis. Enchondromatosis is not genetically transmitted but may be the result of a somatic mosaic mutation in isocitrate dehydrogenase 1 or 2 [47]. The incidence of Ollier's disease is estimated at 1 per 100,000.

Presenting complaints in multiple enchondromatosis are usually bone pain due to mechanical insufficiency, pathologic fracture through the lesions, angular deformity at the knee or fingers, or limb length discrepancy [48]. The condition typically presents in the first or second decade of life.

The radiographic appearance of multiple enchondromatosis is quite distinct (Fig. 20.8). Streaky lytic lesions are seen in the metaphysis of the bones. At the knee, the elongated areas of enchondroma give a "fanlike" appearance to the metaphysis. There are areas of fine calcification within the lesions due to the presence of calcification in the cartilaginous lesions. The lesions seem to stop at the physis. It is common that multiple enchondromas are seen, and that they tend to present unilaterally. The cortex of the bone is intact in the absence of pathologic fracture. Angular deformity at the knee and limb length discrepancy can be appreciated (Fig. 20.9) [49].

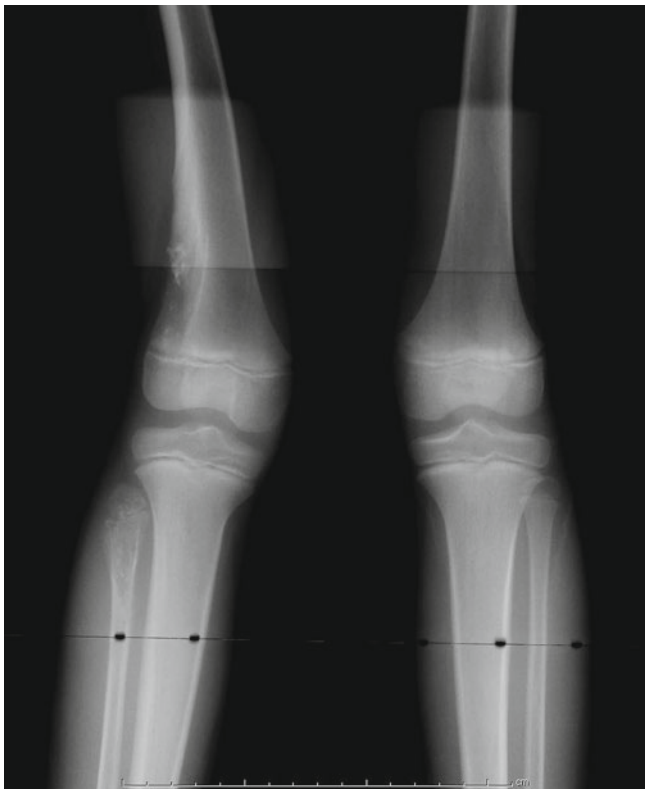


Fig. 20.8 Radiographic appearance of enchondromatosis of the distal femur and proximal tibia in a 7-year-old male. Note the eccentric location of the metaphyseal femoral lesion, and the presence of intralesional calcification which is typical of cartilage tumors. The right proximal fibula is also involved

Biopsy reveals a pearly white to bluish-white specimen. Microscopic examination shows bland cartilaginous tissue, with nests of plump chondrocytes. The nuclei are uniform, and mitotic figures may be seen in children but should be few in number. "Chicken-wire" calcification, which are areas of punctate calcification within the lesion, can be appreciated, which renders the specimen slightly gritty.

Treatment of children with solitary enchondromas is merited if there is angular deformity. Small asymptomatic lesions in the lower extremities may be observed. Multiple enchondromatosis presents for orthopedic treatment more frequently. Curettage and bone grafting of the enchondromas is in large part ineffective due to the extensive nature of the disease, but may be of some benefit in the hand. Children with Ollier's disease have significant angular deformities, with a predisposition towards distal femoral varus [49]. The angular deformity is usually accompanied by varying amounts of limb shortening which may measure up to 25 cm at maturity (Fig. 20.10) [48]. In the lower extremity, angular correction with or without lengthening may be undertaken. Standard fixation with internal devices such as plates and screws may be compromised by poor bony integrity due to the large cartilaginous lesions. Gradual deformity correction using external fixation devices with fine wire bony fixation has been successful [50–54]. When pins or wires are inserted intralesionally, fixation can usually be maintained; however it should be expected that the regenerate will contain enchondromatosis tissue (Fig. 20.11a, b) [53]. Lengthening with intramedullary devices has also been reported [55].



Fig. 20.9 (**a**) Asymmetric genu valgum and limb length discrepancy in a young boy with Ollier's disease. (**b**) A close-up of the left knee shows the streaky lytic lesions characteristic of enchondromatosis



Fig. 20.10 Severe limb length discrepancy measuring greater than 15 cm and varus deformity of the left distal femur in 7-year-old male with Ollier's disease. Note the presence of enchondromas in bilateral femora and tibias as well as the pelvis. The extent of involvement is asymmetric

Malignant transformation has been described in enchondromatosis in up to 20–30 % of patients by the age of 40. Patients with significant expansion of a lesion or with increasing pain should be suspected. Chondrosarcoma is the most frequent malignant tumor seen in up to 25 to 30 % of adult patients with enchondromatosis [56, 57]. Histologic overlap between benign enchondromas seen in Ollier's disease and low-grade chondrosarcoma makes the diagnosis of malignant degeneration challenging. Remote non-bony malignancies are associated with Maffucci's syndrome, which carries a nearly 100 % incidence of the development of a malignant tumor [58]. These children must be carefully monitored throughout their lifetime for the appearance of cancerous lesions.

Box 20.4. Enchondroma

Age at presentation: 1st or 2nd decade.
Location: metaphyis, predisposition to asymmetry.
Number: solitary or multiple (Ollier's disease).
Presenting symptoms: bone pain, pathologic fracture, angular deformity, limb length discrepancy.
Rx: osteotomy for deformity correction, lengthening, in hand may curettage and bone graft.
Malignant degeneration: chondrosarcoma in 25–30 % of Ollier's disease.

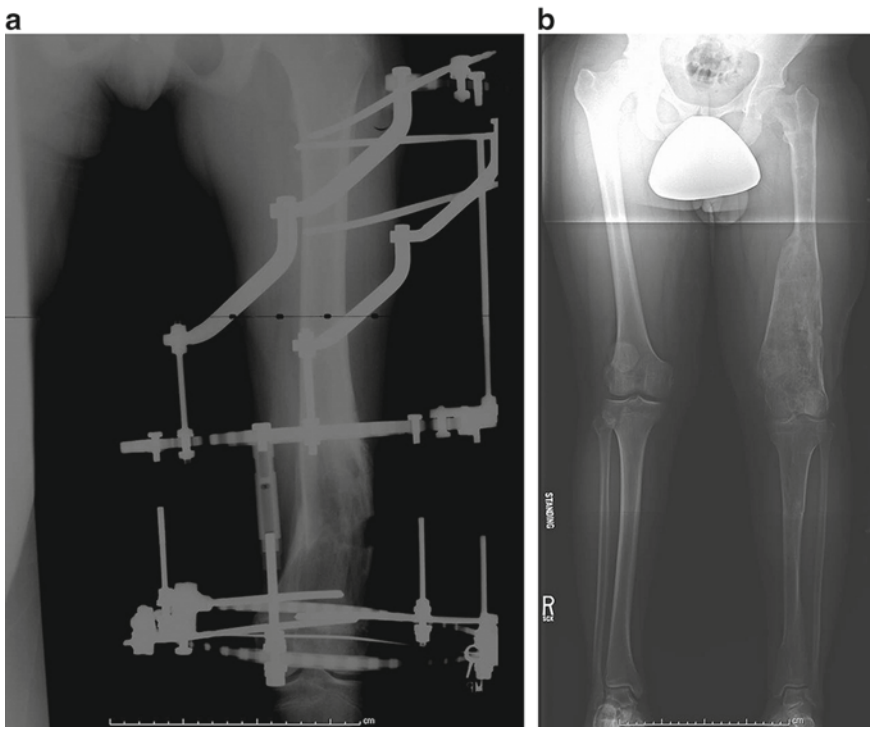


Fig. 20.11 (**a**) Varus osteotomy of the distal femur in patient with Ollier's disease. External fixation with half-pins and fine wires was successful in stabilizing the osteotomy. (**b**) Standing radiograph 1.5 years following removal of the external fixator shows satisfactory alignment but a persistent limb length discrepancy

Solitary Osteochondromas and Multiple Hereditary Exostoses

An osteochondroma is a common benign bone growth which protrudes from the surface of the native bone and is capped with cartilage. Osteochondromas occur in the metaphyseal area and have been hypothesized to be the result of sequestration of a fragment of physis which causes the outgrowth. The bone of an osteochondroma is normal bone and is in continuity with the host bone. Osteochondromas may occur in any bone, including the pelvis, spine, and fingers, but are most common in the distal femur, proximal tibia, and proximal humerus [59].

Multiple hereditary osteochondromatosis or exostoses (MHE) are an inherited condition typified by the presence of multiple osteochondromas scattered throughout the skeleton. It is genetically transmitted as an autosomal dominant trait with very high (96 %) penetrance but variable phenotypic expression. The number and location of osteochondromas can vary among family members. The tumor suppressor genes EXT1, EXT2, and EXT3 for MHE have been localized on chromosomes 8, 11, and 19 [60–62]. Patients with MHE have a loss of function of these genes, thereby allowing growth of the osteochondromas. Patients with EXT1 mutations generally have more severe phenotypical expression and shorter stature than those with EXT2 mutations [63–67]. It is believed that patients with solitary osteochondromas have somatic, but not generalized, mutations in the EXT gene. The incidence of multiple osteochondromas is 1 per 50,000. Ninety percent of patients with MHE have a positive family history and demonstrable mutations in the EXT 1 or 2 genes [68].

Patients with a single osteochondroma often present in the first or second decade for evaluation of a painless periarticular mass. Patients with MHE usually present in early childhood with multiple small masses. The median age of presentation in MHE is 3 years [69]. The presence of a positive family history generally created a heightened awareness of the condition in the parents and therefore an earlier presentation. Patients may complain of pain if there has been trauma in the area of the osteochondroma, or if the mass is sufficiently large to create symptoms due to impingement of overlying muscle or tendons. In many cases, the patient is asymptomatic, but the lesion has been identified coincidentally on a radiograph obtained for another reason. Patients with MHE tend to be of mildly short stature. Stature less than the 25th percentile was seen in 58 % of patients with MHE [66]. Additionally, the extremities will appear shorter than the trunk does, with the amount of shortening being linked to the size of osteochondromas present in the limb segment [70]. Angular deformity, such as genu valgum and ankle valgus, may be present.

Radiographs show a bony protrusion in the periarticular area originating from the metaphyseal bone (Fig. 20.12).



Fig. 20.12 Radiograph of a 10-year-old boy with multiple hereditary exostoses. Note the numerous sessile lesions, such as that seen at the distal tibia, and the pedunculated lesions as seen at the distal femur and proximal tibia. Ankle valgus is present at the left ankle, and an intraosseous osteochondroma from the right tibia deforms and thins the fibula

Osteochondromas are not epiphyseal. The cortex of the osteochondroma is in continuity with the native bone, and the trabeculae within the osteochondroma are similarly in continuity with the metaphysis. Osteochondromas may be pedunculated, with a distinct bony stalk, or sessile, where the lesion is broad-based. When pedunculated, osteochondromas typically point away from the joint. Osteochondromas may vary in size. Large osteochondromas in two-bone segments, such as the lower leg, can cause deformity of the smaller bone, in this case the fibula. Osteochondromas in the distal femur, proximal tibia, and/or proximal fibula are seen in 94 % of children with MHE. Proximal humerus masses are present in 50 % of cases, the scapula and ribs in 40 %, the distal radius and ulna in 30 %, the proximal femur in 30 %, the phalanges in 30 %, the distal fibula in 25 %, and the distal tibia in 20 % of MHE patients [69].

The radiographic appearance of an osteochondroma is sufficiently distinct that biopsy is not needed. When osteochondromas are excised for relief of symptoms, pathologic evaluation of the mass should be obtained. The bone of an osteochondroma is histologically normal bone. The junction of the bone and the cartilage cap shows endochondral ossification. It is important to evaluate the cartilaginous cap of the

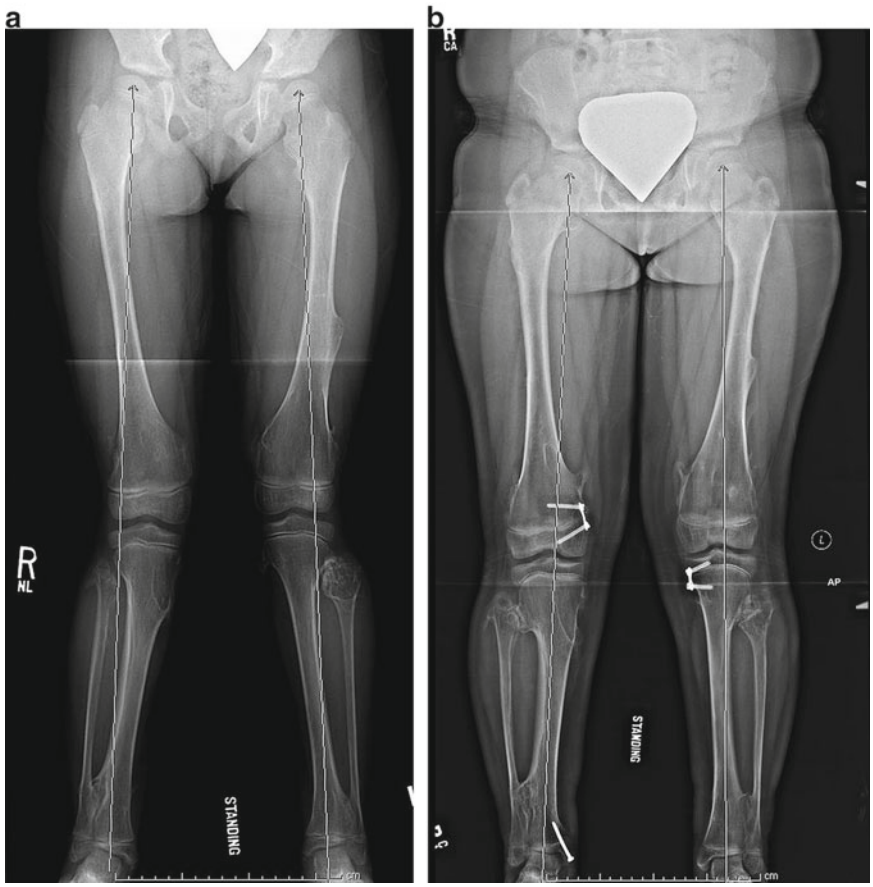


Fig. 20.13 (**a**) Valgus deformity in an 8-year-old male with multiple hereditary exostoses. (**b**) Radiograph 4 years following growth modulation shows correction of the valgus deformity. Note the medial malleolar screw placed for treatment of ankle valgus

lesion. In a benign osteochondroma, the cartilage cap is thin (<2 cm) and the chondrocytes histologically bland, without mitotic figures or nuclear pleomorphism. In some instances, osteochondromas can undergo malignant transformation into low-grade chondrosarcomas. The incidence of malignant transformation is less than 1 % and is more likely in mature patients and those with pelvic lesions. Yet an increase in pain and rapid increase in size should merit excisional biopsy. The cartilage cap in an osteochondroma that has become malignant is thickened, and the nuclei of the cartilaginous region pleomorphic. It is the combination of the histological appearance of the cartilage cap combined with its thickness that leads to the diagnosis of chondrosarcoma [71].

Treatment of a solitary osteochondroma is excision if the lesion is causing pain or mechanical symptoms due to its size. Recurrence is unlikely with complete excision. Patients with MHE should be offered excision of symptomatic lesions, and observation if the masses are asymptomatic. Due to the number of osteochondromas that many of these children have, excision of all the lesions is impractical and unnecessary; however recent validated outcome questionnaires have highlighted the complaints of musculoskeletal pain in children with MHE [72–74]. Masses that are creating secondary deformities, such as large distal tibial lesions that are producing fibular deformity (see Fig. 20.12), may benefit from excision. Advanced imaging of larger lesions prior to surgery may be helpful in identifying the location of neurovascular structures that may be displaced by the osteochondromas. In particular, the peroneal nerve is frequently thinned or displaced by proximal fibular lesions, and should be identified and protected if the proximal fibular osteochondroma is to be excised. Even so, postoperative peroneal nerve palsy frequently results from excision of fibular lesions [75, 76], and preoperative counselling of the patient and family about the possibility of a foot drop necessitating use of an ankle-foot orthosis should be performed. Additionally, venous thromboembolism following excision of lesions behind the knee has been seen in rare cases [77].

Angular deformity of the lower extremities may be present in patients with MHE. Growth modulation in skeletally immature patients may be quite useful, especially at the knee. When preoperative planning growth modulation, consideration for inaccuracies in bone age should be made [78]. Most often, the knee assumes a valgus position due to proximal tibial involvement [70]. Patellar instability may be associated with genu valgum in some children [79]. Medial proximal tibial and distal femoral growth modulation plating can be useful (Fig. 20.13a, b). Skeletally mature patients may rarely require osteotomy. In such cases, the peroneal nerve is prone to postoperative palsy, so fibular osteotomy should be approached with great caution.

Ankle valgus is also frequently present in patients with MHE due to distal tibial and fibular lesions [80]. Radiographs show relative shortening of the distal fibula and distal tibial valgus tilt. Gradual deformity correction via insertion of a staple or fully threaded medial malleolar screw in skeletally immature patients has been shown to be of some utility [81, 82], but this author finds that correction of the ankle is frequently very slow and often incomplete. Rarely is the ankle deformity sufficiently symptomatic, however, to merit osteotomy. Other children with MHE may complain of ankle deformity due to large distal tibial osteochondromae which exert pressure on the distal fibula, leading to erosions and bowing of the fibula. Excision of the osteochondroma can relieve symptoms, and the fibula will most likely remodel following excision of the tibial mass. A long-term study of adults with ankle valgus secondary to osteochondromas linked persistent excessive ankle valgus with limitations in recreational activities and with the development of arthritis [83].

Progressive hip dysplasia due to the presence of proximal femoral osteochondromas has been seen in a minority of patients with MHE [84, 85]. Surgical reconstruction via excision of the osteochondromas combined with proximal femoral osteotomy as needed for containment can be useful.

There have been published studies which have raised awareness of the presence of spinal osteochondromas in patients with MHE [86, 87]. The incidence of clinically important spinal osteochondromas remains quite rare, but it is imperative that the lower extremity surgeon be aware of their occurrence. The patient who presents with numbness or tingling in the lower extremity, or with difficulty with ambulation, should have a careful neurologic assessment. It is enticing to explain the symptoms by the presence of osteochondromas either in the popliteal fossa or at the proximal fibula, but consideration for a spinal etiology of the symptoms should be given. Spinal osteochondromas are readily seen via either CT or MRI scanning. Excision of a space-occupying lesion in the spinal canal should be offered.

As mentioned above, malignant transformation of an osteochondroma may occur in up to 1–2 % of patients [71, 88–90]. Warning signs include a crescendo of pain, pain at rest, and unusually rapid growth of a lesion, particularly in a skeletally mature patient. Lesions in the pelvis and shoulder girdle are most prone to malignant degeneration. Preoperative imaging of the cartilaginous cap with MRI can support the suspicion of malignancy if the cap is greater than 2 cm in maximal thickness. Referral to an orthopedic oncological surgeon is merited in such cases. Treatment is typically wide surgical resection.

Finally, there are several case reports of popliteal artery pseudoaneurysms in association with large osteochondromas in the posterior knee area. In such cases, vascular repair is needed in conjunction with excision of the mass [91, 92].

Box 20.5. Osteochondroma
Age at presentation: 1st or 2nd decade.
Location: metaphyseal.
Number: solitary or multiple.
Presenting symptoms: painless mass, mechanical symptoms.
Rx: excision of symptomatic lesions.
Malignant degeneration: chondrosarcoma in ≤1 %.
Under treatment: correction of angular deformity.

Dysplasia Epiphysealis Hemimelica (Trevor's Disease)

Dysplasia epiphysealis hemimelica is a condition where intra-articular osteocartilaginous lesions develop, originating from the epiphysis in skeletally immature children. Lesions are solitary in one-third of patients and multiple in two-thirds. Lesions in those patients with multiple epiphyseal involvement tend to be unilateral and to involve only one side (i.e. either the medial or lateral) of the epiphyses (Fig. 20.14). The disease is not genetically inherited but is more common in boys than girls [93, 94].

Patients present either in the first or second decade of life with limited range of motion of involved joints which lead to gait disturbances. The most common sites are the distal femur, distal tibia, talus, and navicular. Lower extremity involvement is more common than upper extremity involvement. When the condition affects the distal tibia or talus, the presentation is usually of fixed equinus, while at the knee, a flexion position is adopted. Range of motion may be somewhat painful owing to the presence of the intra-articular mass. When present in the hip, the lesions are typically from the acetabulum and can lead to subluxation if sufficiently large.

Radiographs may initially be normal, as the cartilaginous masses in the very young child are difficult to detect. As the child grows older, the lesions resemble those of osteochondromas, but the masses originate from the epiphysis and point towards the joint rather than away. Small lesions may be difficult to fully appreciate on plain radiographs, but MRI shows the cartilaginous extent of the lesion [95].

Histologically, lesions are made up of normal bone and varying amounts of overlying cartilage. The cells in both the bone and cartilage are benign in appearance and mitotic figures are not seen.

The intra-articular lesions grow in the skeletally immature patient. Treatment consists of excision of symptomatic lesions [96]. Unfortunately, due to the intra-articular nature of the masses, complete excision is difficult. While MRI may show delineation between the lesion and the native epiphysis,

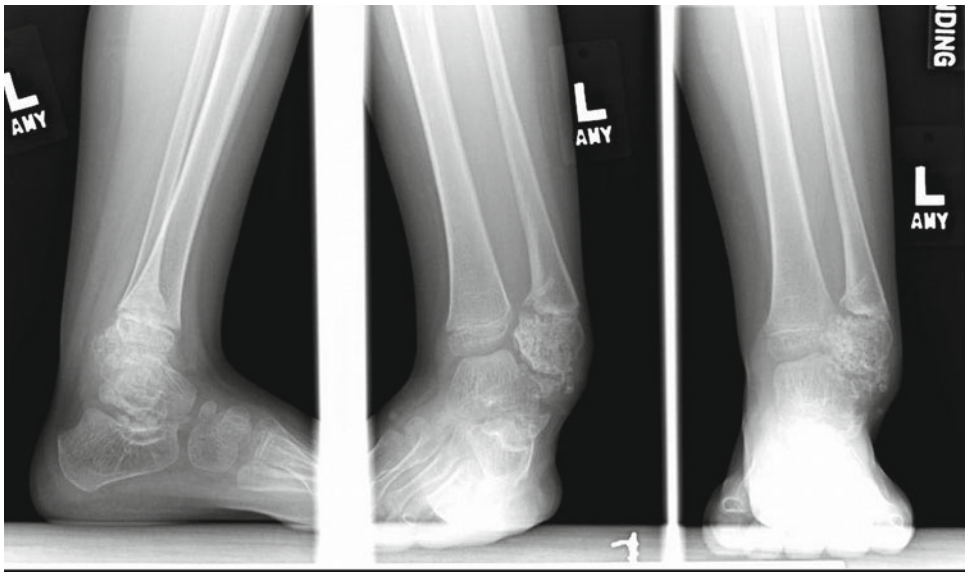


Fig. 20.14 Dysplasia hemimelica epiphysealis (Trevor's disease) affecting the distal fibula and lateral midfoot. Note the enlargement of the distal fibular epiphysis and intra-articular exostoses

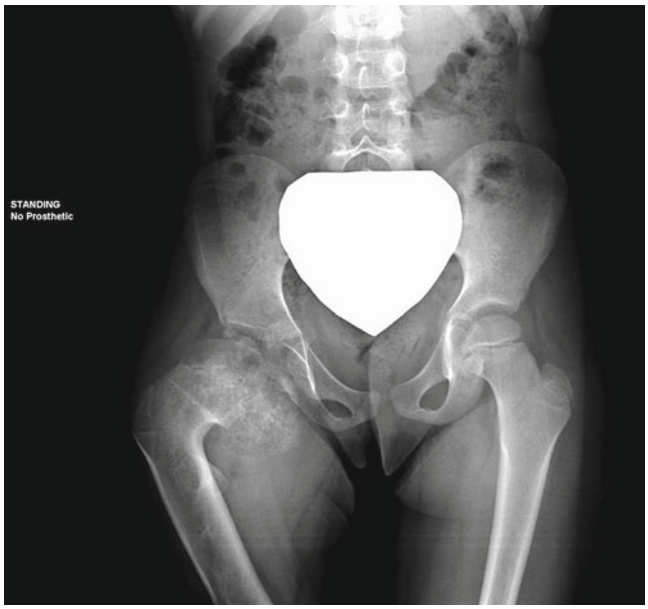


Fig. 20.15 Trevor's disease of the right hip of a 5-year-old male. Note the exostoses emanating from the femoral head and acetabulum leading to lateralization of the hip

there is most often no visible distinction intraoperatively between the mass and the native articular cartilage, which is problematic for the surgeon in deciding on the margins of the excision. Recurrence is common. Residual joint incongruity leads to early degenerative arthritis. Excision of hip lesions is particularly challenging but may be successful via surgical hip dislocation [97–100] (Fig. 20.15). Limb length discrepancy owing to premature physeal closure may be present and require treatment.

Malignant transformation in dysplasia epiphysealis hemimelica has not been described.

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21 Management of Juxtaphyseal Malignant Bone Tumors Around the Knee Joint: New Concepts in Limb-Sparing Surgery

Hidenori Matsubara and Hiroyuki Tsuchiya

Introduction

There are various kinds of malignant bone tumors in pediatrics. Osteosarcoma is the most common primary malignant bone tumor in children. Prior to the 1970s, amputation or joint disarticulations were the standard treatment and the only choice of treatment. However, now over 90 % of osteosarcoma resections are limb-sparing surgeries due to advancements in chemotherapy, medical imaging, and implant technology [1–3]. Options for limb salvage reconstruction after wide resection include osteoarticular allografts, allograft prosthetic composites, recycled autograft, and modular or custom-made endoprostheses. However, the limb function often remains limited and deteriorates over time. Complications such as infections, nonunion of grafts, and bone resorption could eventually lead to amputation. Therefore, the aim of biological reconstruction is to achieve better limbs and almost normal limb function, and to reduce complications.

We first performed transepiphyseal osteotomy and resected a malignant bone tumor followed by reconstruction using the distraction osteogenesis technique (DO method) [4–6] and massive frozen tumor-bearing bone treated with liquid nitrogen (LN method) [7, 8]. In this chapter, we introduce our two original biological reconstruction procedures for malignant tumors.

Indication

To facilitate decision making for reconstruction in cases of juxta-articular malignant bone tumor around the knee joint, we developed a new system for classifying tumor excision (Table 21.1, Fig. 21.1) [9]. Cases indicated for the DO method should have tumors of Types I to IV according to our classification system, of no more than a 15-cm defect after the tumor resection (resulting in a treatment time of less than 1 year), and with a good response to chemotherapy. Theoretically, the DO method can be applied even in cases with Types V and VI tumors combined with arthrodesis. Currently, however, it has been replaced with the LN method. The LN method can be indicated in all cases. For chemotherapy responders, this enables preservation of the maximum amount of healthy tissue, such as soft tissue, growth plates, and joint surfaces, to provide excellent limb function [3].

Surgical Techniques

Type I: DO Method (Fig. 21.2)

In Type I, the tumor is diaphyseally or metaphyseally located over 2 cm away from the epiphyseal plate. The operative technique of the DO method consists of en bloc tumor excision with preservation of the epiphyseal plate and articular surface. An external fixator is applied and osteotomy is performed for postoperative bone transport. After docking bone transport, we usually perform bone graft at the docking site. Shortening-distraction is indicated if acute shortening is possible during the operation. Optional intramedullary nailing, if feasible, or plate conversion, is also used to shorten the external fixation time [4, 6].

Type I: LN Method (Fig. 21.3)

One-site osteotomy and pedicle freezing method are applied in the Type I LN method. After transdiaphyseal osteotomy, soft tissue is removed, and the tumor is curetted after thorough isolation to prevent fracture during freezing. Bony lesions connected to the limb are then rotated and frozen in

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Table 21.1 Decision making for reconstruction based on location and extent of tumor

Type I: Tumor at diaphyseal location or metaphyseal location over 2 cm from the epiphyseal plate
Type II: Tumor at metaphyseal location extending to less than half of the epiphyseal growth plate
Type III: Tumor at metaphyseal location extending to the whole growth plate
Type IV: Metaphyseal tumor extending through the growth plate into part of the epiphysis at least 10 mm from the articular surface
Type V: Tumor extending into less than half of the epiphysis less than 1 cm away from the articular surface
Type VI: Tumor extending into more than half of the epiphysis less than 1 cm away from the articular surface

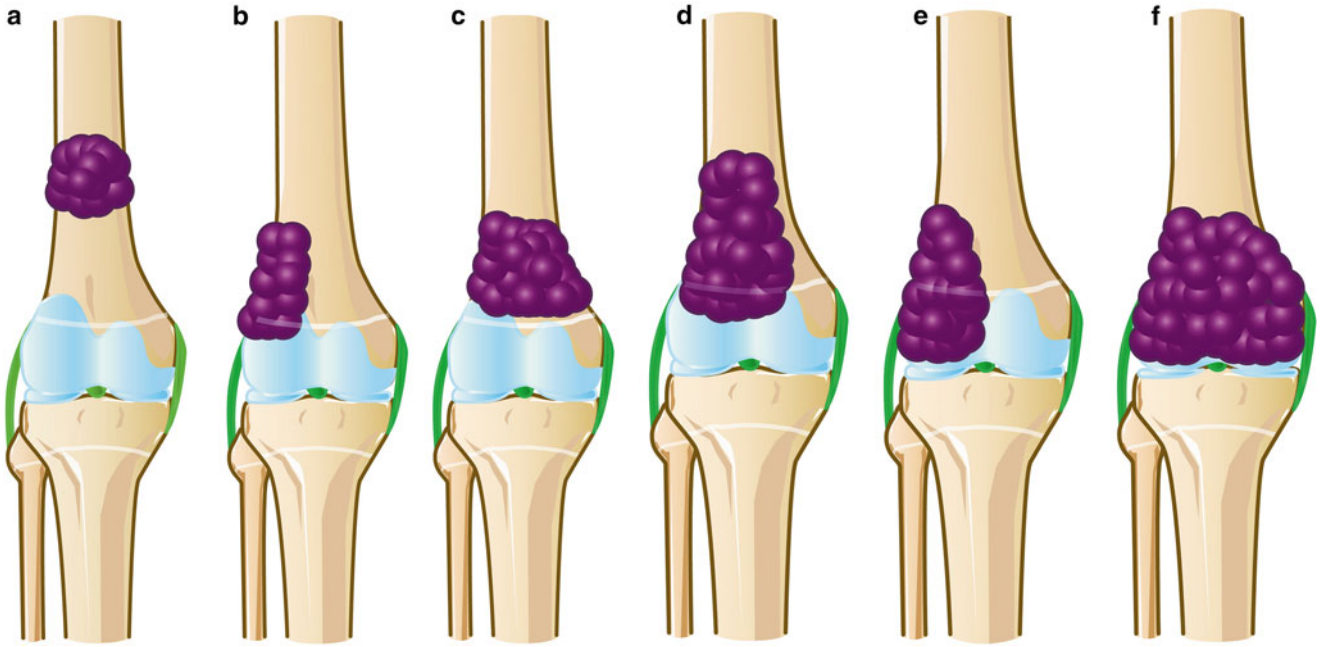


Fig. 21.1 Classification of tumor excision. (**a**) Type I. (**b**) Type II. (**c**) Type III. (**d**) Type IV. (**e**) Type V. (**f**) Type VI

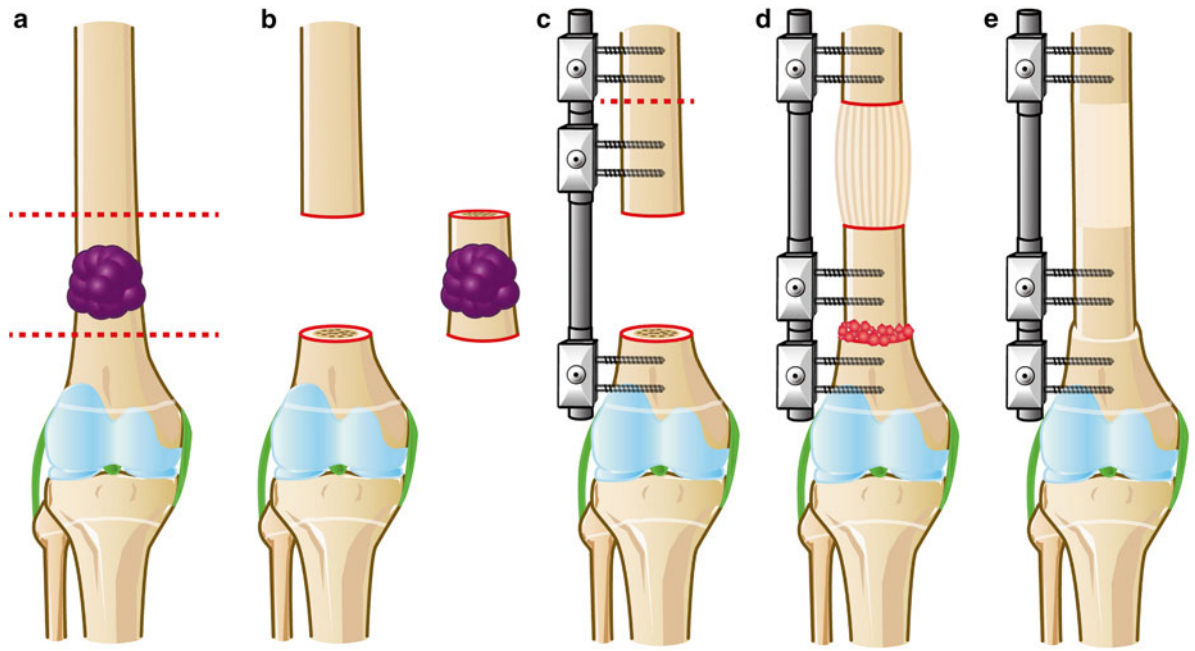


Fig. 21.2 Type I: DO method. (**a**) Resection line. (**b**) Tumor resection. (**c**) Application of external fixator and osteotomy for lengthening. (**d**) Bone grafting at docking site. (**e**) After bone consolidation

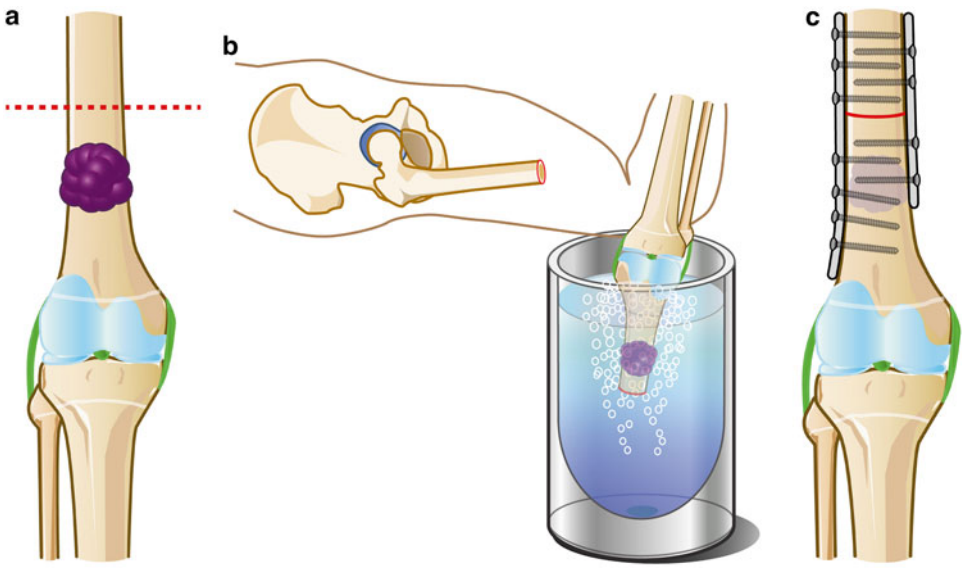


Fig. 21.3 Type I: LN method. (**a**) Resection line. (**b**) Pedicle freezing. (**c**) Reconstruction with double plates

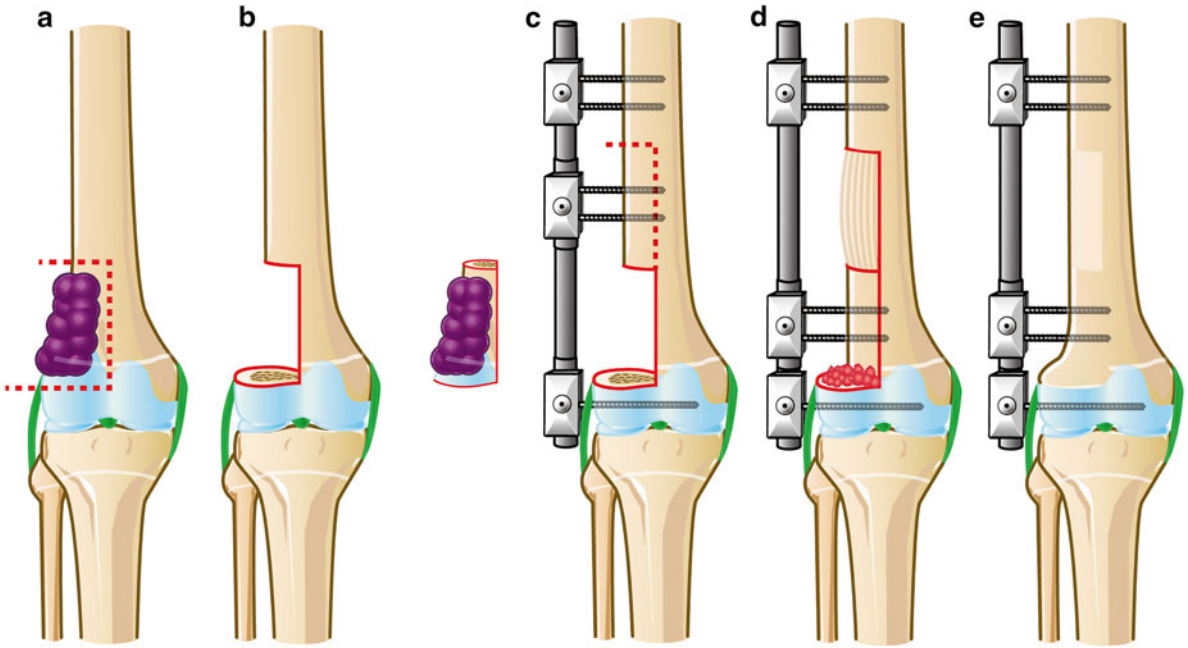


Fig. 21.4 Type II: DO method. (**a**) Resection line. (**b**) Tumor resection. (**c**) Application of external fixator and osteotomy for lengthening. (**d**) Bone grafting at docking site. (**e**) After bone consolidation

liquid nitrogen for 20 min, thawed at room temperature for 15 min, and then thawed in distilled water for 10 min. Reconstructions are performed by osteosynthesis using double plates [8].

Type II: DO Method (Fig. 21.4)

In Type II, the tumor is located metaphyseally and extends to less than half of the epiphyseal growth plate. There is also a possibility of joint deformity due to unequal growth after reconstruction. The operative technique of the DO method consists of hemicortical en bloc tumor excision. An external fixator is applied, and proximal hemicortical osteotomy is performed for postoperative bone transport. Epiphysiodesis of the other side of the growth plate is one option to avoid future angular deformities for young patients. After docking bone transport, we usually perform a bone graft at the docking site. At that time, plate conversion is optionally used to shorten the external fixation time. If a deformity occurs in the future, correction with or without lengthening is performed.

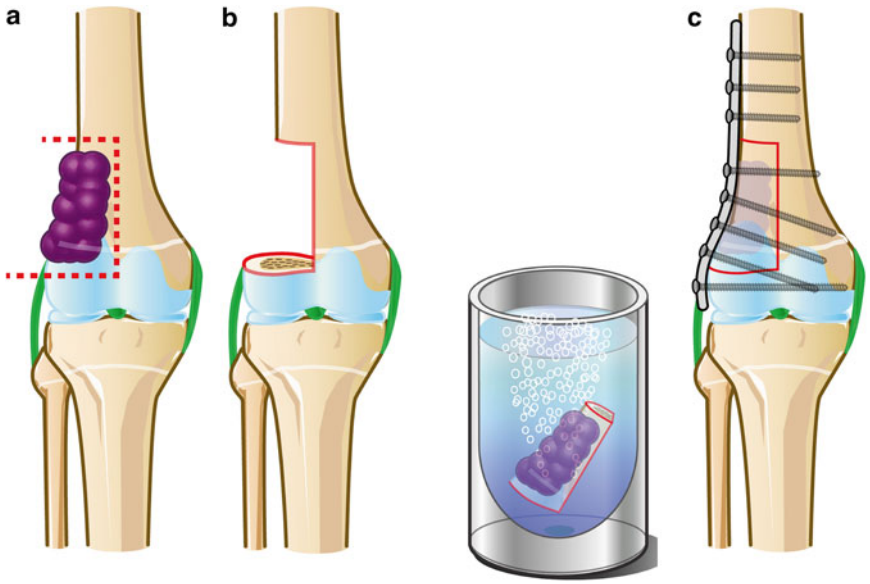


Fig. 21.5 Type II: LN method. (**a**) Resection line. (**b**) Freezing tumor-bearing bone. (**c**) Reconstruction with single plate

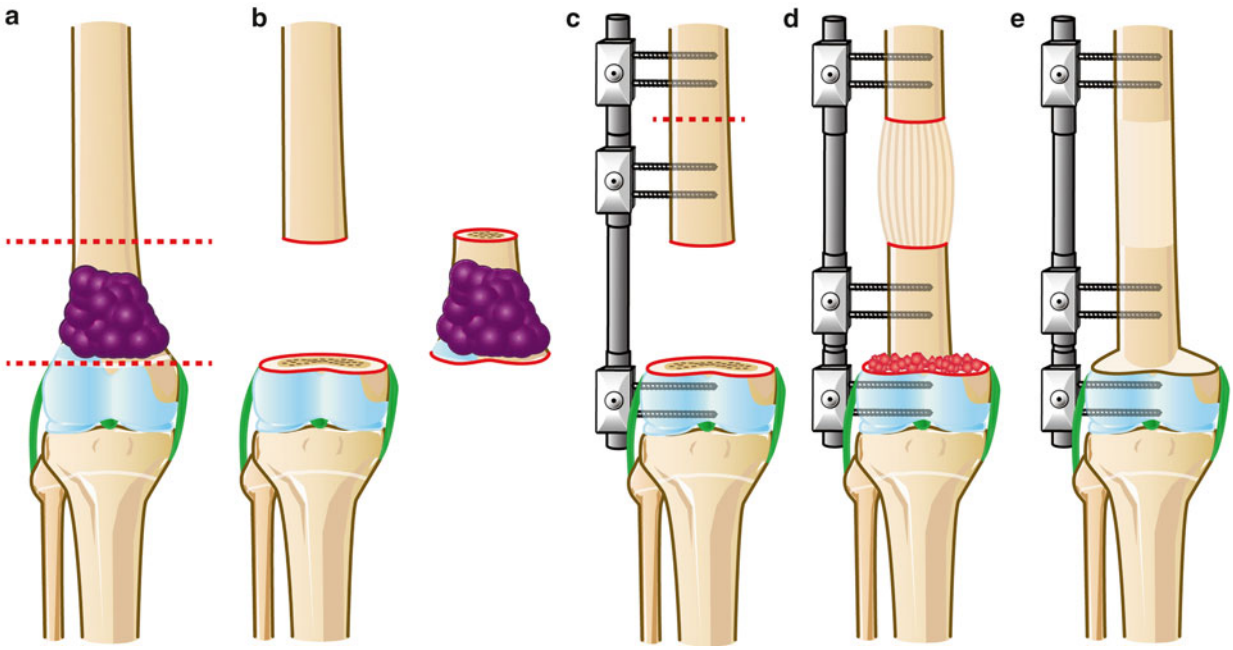


Fig. 21.6 Type III: DO method. (**a**) Resection line. (**b**) Tumor resection. (**c**) Application of external fixator and osteotomy for lengthening. (**d**) Bone grafting at docking site. (**e**) After bone consolidation

Type II: LN Method (Fig. 21.5)

Hemicortical resection and a free-freezing method are applied for Type II resections. After transepiphyseal and hemicortical resection, soft tissue is removed, and the tumor is curetted after thorough isolation. The tumor-bearing bone is frozen in liquid nitrogen and thawed as usual. Reconstruction is performed by osteosynthesis using a single-locking plate [7].

Type III: DO Method (Fig. 21.6)

In Type III, the tumor is metaphyseally located and extends to the whole growth plate. Limb length discrepancies are anticipated in immature patients, which may be managed surgically if necessary. The operative technique of the DO method consists of transepiphyseal and intercalary en bloc tumor excision with the growth plate. Surgical technique is basically same as in the Type I DO method.

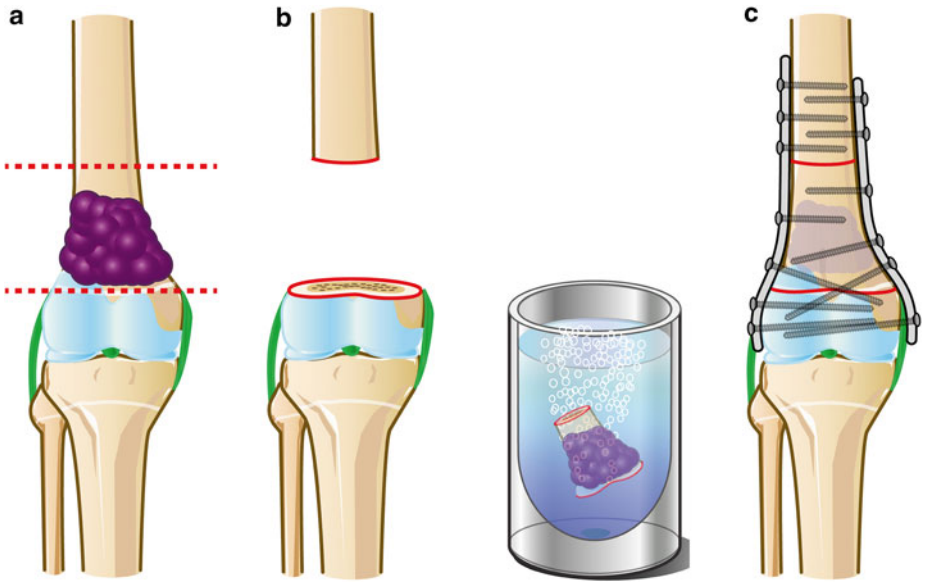


Fig. 21.7 Type III: LN method. (**a**) Resection line. (**b**) Freezing tumor-bearing bone. (**c**) Reconstruction with double plates

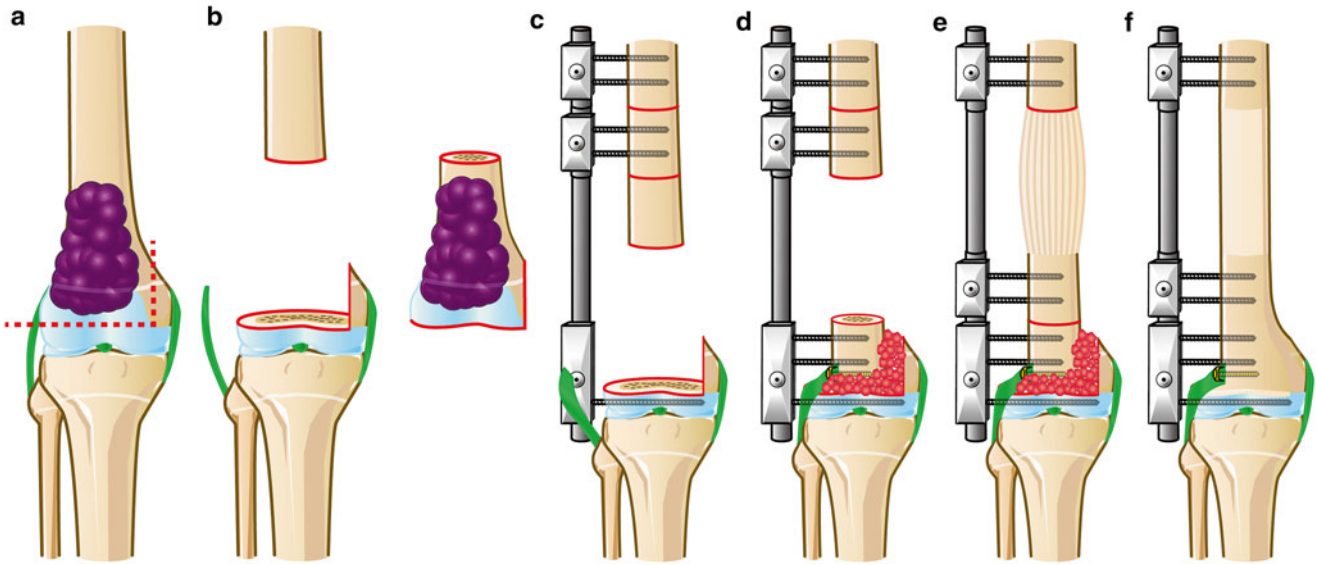


Fig. 21.8 Type IV: DO method. (**a**) Resection line. (**b**) Tumor resection. (**c**) Application of external fixator and osteotomy for lengthening. (**d**) Reconstruction of periarticular structure with diaphyseal bone cylinder. (**e**) After docking. (**f**) After bone consolidation

Type III: LN Method (Fig. 21.7)

Intercalary resection and the free-freezing method are applied for Type III resection. After transepiphyseal and intercalary resection, free freezing is performed in the same way as in the Type II LN method. Reconstruction is performed by osteosynthesis using double plates.

Type IV: DO Method (Fig. 21.8)

In Type IV, the tumor is metaphyseally located and extends to the whole growth plate into part of the epiphysis at least 10 mm away from the joint line. Limb length discrepancies are anticipated in immature patients, which may be managed surgically if necessary. The operative technique of the DO method consists of transepiphyseal and intercalary en bloc tumor excision with the growth plate, and with preservation of the articular surface. The periarticular structure should be stabilized immediately to prevent joint contracture. If only a thin epiphysis or articular surface is preserved, a free bone cylinder should be taken from the diaphysis and fixed to the remaining epiphysis with a bone graft and a spike washer and screw, in order to stabilize and reconstruct the periarticular structure. The newly created diaphyseal defect is restored by bone transport. If feasible, optional plate conversion is also used.

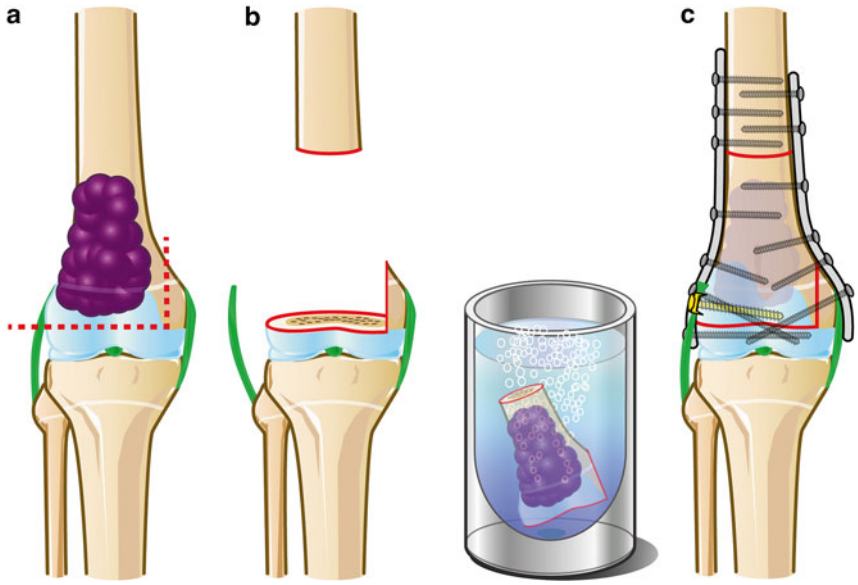


Fig. 21.9 Type IV: LN method. (**a**) Resection line. (**b**) Freezing tumor-bearing bone. (**c**) Reconstruction with double plates and spike wisher

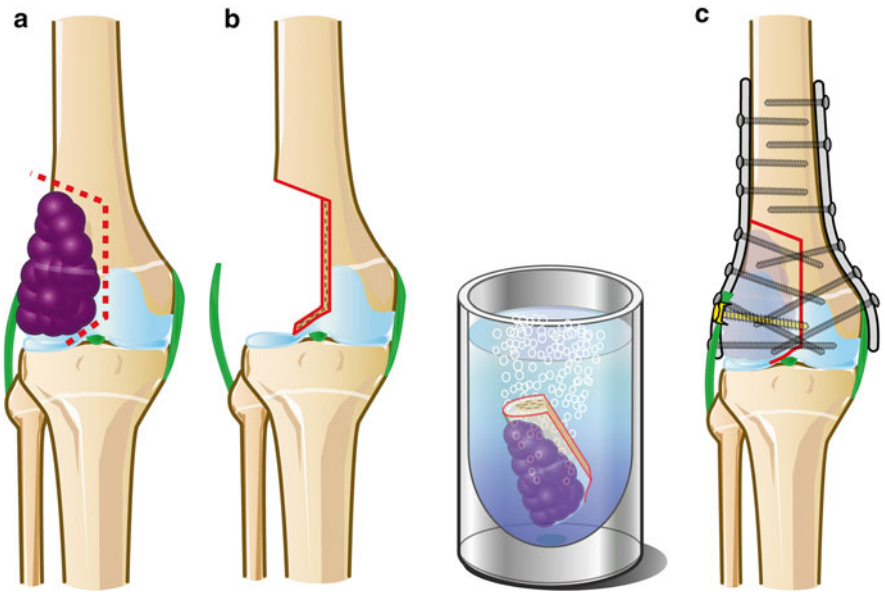


Fig. 21.10 Type V: LN method. (**a**) Resection line. (**b**) Freezing tumor-bearing bone. (**c**) Reconstruction with double plates and spike wisher

Type IV: LN Method (Fig. 21.9)

Intercalary resection and free-freezing method are applied for Type IV resection. After transepiphyseal and intercalary resection, the tumor-bearing bone is frozen and thawed in the same way as above. Reconstruction is performed with double plates. When the ligament is detached from tumor-bearing bone, it is reconstructed with a spike washer and screw.

Type V: LN Method (Fig. 21.10)

In Type V, the tumor extends into less than half of the epiphysis. There is a possibility of joint deformity due to unequal growth after reconstruction. The operative technique of the LN method consists of hemicortical resection with the osteoarticular surface, and the free-freezing method is applied for Type V. After resection of the hemicondyle, soft tissue is removed, and the tumor is curetted after thorough isolation.

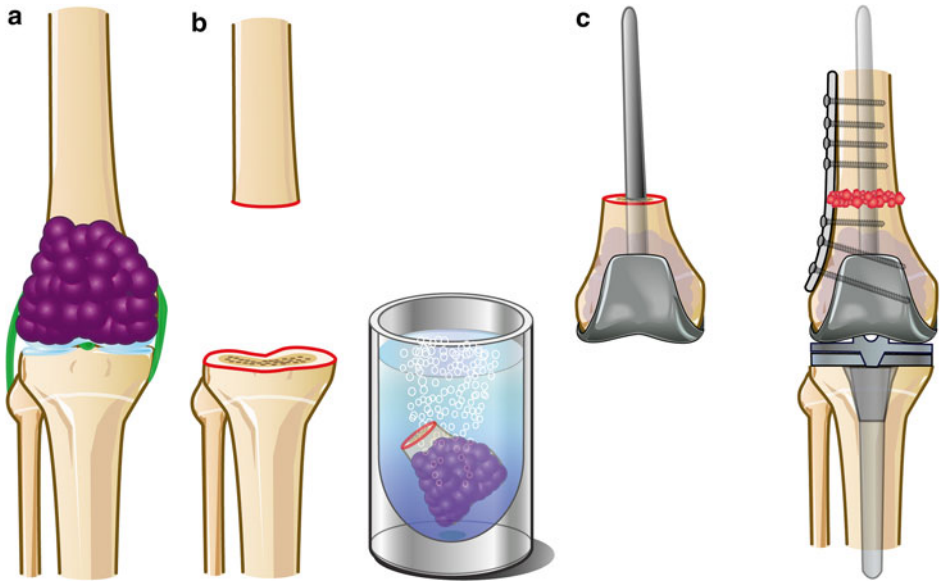


Fig. 21.11 Type VI: LN method. (**a**) Resection line. (**b**) Freezing tumor-bearing bone. (**c**) Reconstruction with surface arthroplasty with or without a locking plate and bone graft

The tumor-bearing bone is frozen in liquid nitrogen and thawed in the same way as above. Reconstructions are performed by osteosynthesis using single or double plates, a spike washer, and screws. Frozen osteoarticular grafts tend to undergo osteoarthritis changes; therefore, an optional composite autograft indicated for Type VI is also applied to avoid future operations. Otherwise, UKA or TKA is carried out for degenerative arthrosis.

Type VI: LN Method (Fig. 21.11)

In Type VI, the tumor extends into more than half of the epiphysis. There is no possibility of preservation of the whole joint. There is also a possibility of joint deformity and limb length discrepancy after reconstruction. The operative technique of the LN method consists of en bloc resection with the osteoarticular surface, and the free-freezing method is applied for Type VI. Reconstruction is performed by a combination of frozen grafts and surface arthroplasty with or without a locking plate and bone graft. Strictly speaking, these are partially biological reconstructions; however, they can preserve bone stock.

Case Examples

Case 1 (Type I: DO Method) (Fig. 21.12)

A 17-year-old boy presented with osteosarcoma of the proximal tibia. Neoadjuvant caffeine-potentiated chemotherapy was administered, and the clinical effect was a complete response with significant ossification of the soft-tissue mass of the lateral tibia. We planned intercalary resection and reconstruction with the DO method (bone transport). After the operation, gradual lengthening was started and bone transporter docking was completed. As of the 7-year follow-up, the patient is free of disease, and can run without any pain.

Case 2 (Type IV: LN Method) (Fig. 21.13)

A 10-year-old boy presented with osteosarcoma of the distal femur. Neoadjuvant caffeine-potentiated chemotherapy was administered, and the clinical effect was a complete response with significant shrinkage of the soft-tissue mass of the medial femur. We planned transepiphyseal and intercalary resection and reconstruction with the LN method. Reconstruction was performed by osteosynthesis using plates and cannulated screws. The resection margins were tumor free, and pathological evaluation indicated grade III/IV on the Rosen and Huvos evaluating system. As of the 22-month follow-up, the patient is free of disease, and can play football.

In summary, we introduced limb-salvage surgery for reconstruction after tumor resection using distraction osteogenesis and a frozen autograft method utilizing liquid nitrogen, according to the tumor site, size, and response to chemotherapy. The advantages of the DO method include regeneration of living bone with sufficient strength and durability, biological affinity, resistance against infection, and lifelong restored function. The disadvantages include delayed union at the docking site and pin- or wire-tract

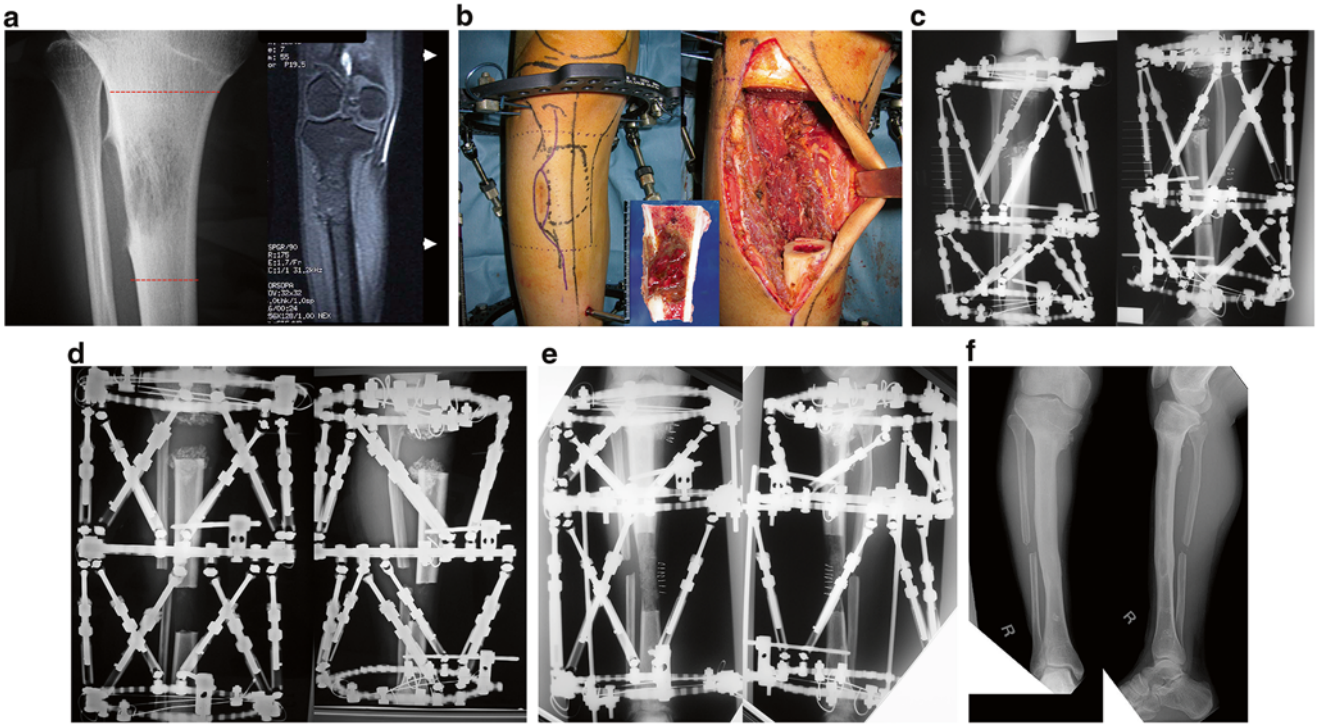


Fig. 21.12 Case (Type I: DO method). A 17-year-old boy with an osteosarcoma of the proximal tibia. (**a**) Preoperative X-ray and MRI scan. Osteotomy line (*broken lines*). (**b**) After tumor resection with intercalary osteotomies. (**c**) X-ray after the operation. (**d**) X-ray during bone transport. (**e**) X-ray at the complication of docking of bone transporter. (**f**) X-ray at 26 months after the operation

infection, as well as the fact that the procedure is time consuming (Table 21.2). The advantages of the LN method are simplicity, osteoinduction, osteoconduction, a short treatment time, a perfect fit, sufficient biomechanical strength, and easy attachment of tendons and ligaments. The disadvantage is basically dead bone at the time of reconstruction. Moreover, revitalization takes a long time, and degeneration of the preserved cartilage matrix occurs over time (Table 21.3). Possible complications with the DO method include pin-site infection managed with antibiotics or removal of pins, docking-site nonunion or delayed union, and delayed consolidation of regenerated tissue treated with bone graft. Complications with the LN method are late infection and nonunion of frozen bone, which are managed with bone graft combined with rigid fixation. There are two important points in obtaining better functional results when either method is selected: firstly, the surgeon should try to preserve the epiphysis if possible; and secondly, it should be attempted to perform biological reconstruction as much as possible.

In future, there may be greater use of biological reconstruction methods to obtain better and earlier functional results, because a natural limb is the best option. To achieve this, we have to aim to facilitate bone regeneration and revitalization much faster, and to adopt biological soft-tissue reconstruction, through advances in tissue engineering, such as with regard to osteogenic and angiogenic growth factors, or with stem cell implantation.

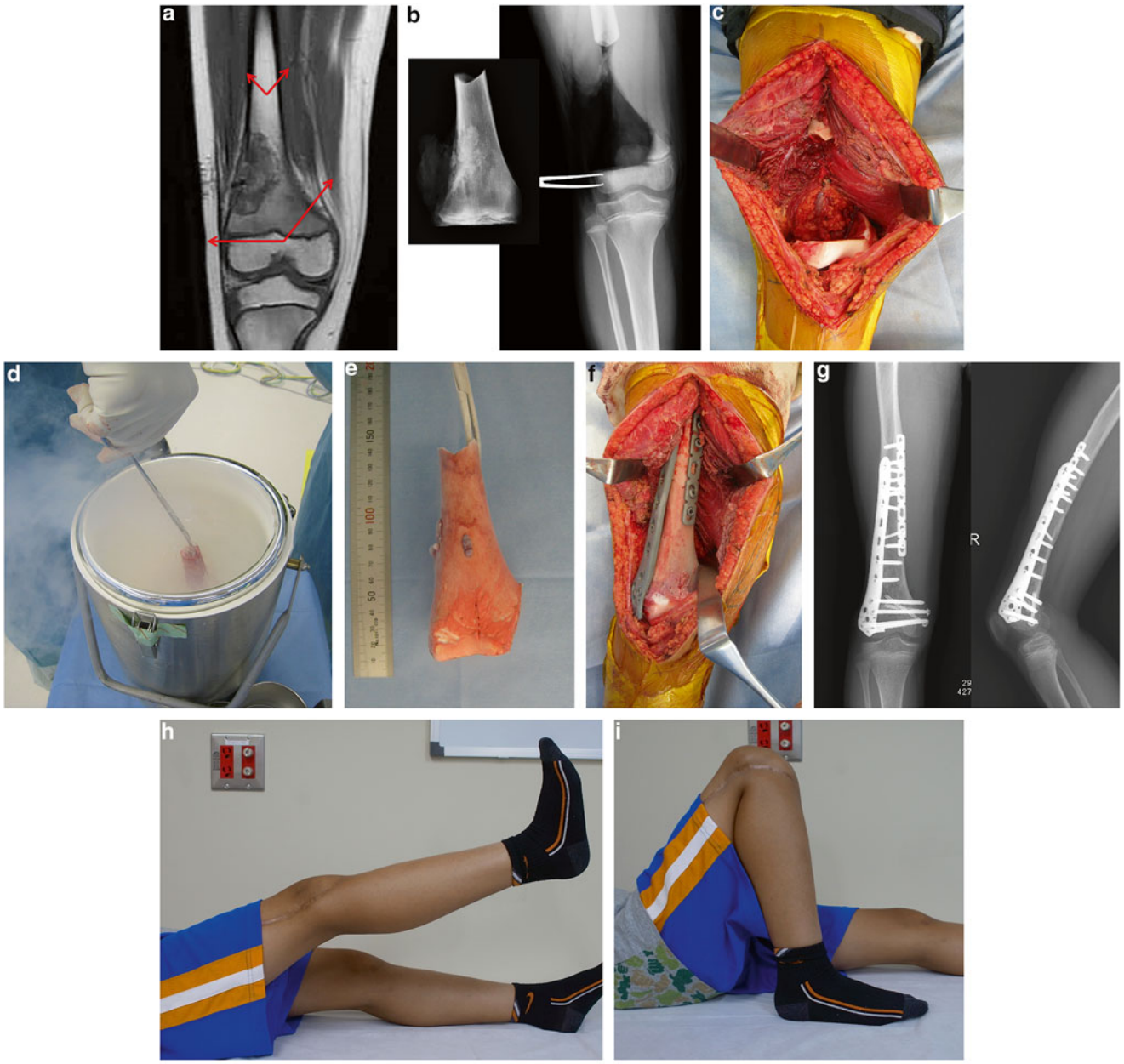


Fig. 21.13 Case (Type IV: LN method). A 10-year-old boy with an osteosarcoma of the distal femur. (**a**) Preoperative MRI scan. Osteotomy line (*arrows*). (**b**) Intraoperative radiograph. Transepiphyseal and intercalary osteotomy was performed. (**c**) After transepiphyseal and intercalary osteotomy. (**d**) Freezing in liquid nitrogen. (**e**) After freezing. (**f**) Osteosynthesis with plates and screws. (**g**) Radiograph 22 months after reconstruction. Host-graft junctions achieved complete union. (**h**) Twenty-two months after reconstruction, no extension lag. (**i**) The range of motion of the knee was 0–140°

Table 21.2 Advantages and disadvantages of the DO method

Advantage	Disadvantage
Living bone regeneration	Lengthy external fixation
Sufficient strength	Patient's burden
Biological affinity	Delayed consolidation
Resistance against infection	Delayed consolidation
Lifelong restored function	Delayed union of docking site

Table 21.3 Advantages and disadvantages of the LN method

Advantage	Disadvantage
Simple procedure	Dead bone reconstruction
Reattachment of soft tissue (tendon and ligament)	Long time to be revitalized
Osteoinduction and osteoconduction	Cartilage degeneration over time
Perfect fit	Delayed union at host-graft junction
Sufficient biomechanical strength	Bone absorption
No contagion	
No harmful denatured protein	

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Part IV

Congenital and Developmental Disorders

Congenital Femoral Deficiency Reconstruction and Lengthening Surgery

22

Dror Paley, David Y. Chong, and Daniel E. Prince

Introduction

Demographics

The often-cited incidence of congenital femoral deficiency (CFD) is 1 in 52,029 based on a review of the Edinburgh Register of the Newborn by Rogala et al. published in 1974 [1]. In retrospect, this may not be accurate, as the incidence is based on the only case identified during the 4.5-year collection period from 1964 to 1968, notably, after thalidomide use was discontinued. This survey is also representative of a relatively homogenous population, of which no information is given regarding race, age, environmental exposure, medication usage, or socioeconomic status of the population.

Embryology

Fetal growth and development of the lower extremities are controlled by a complex cascade of a multitude of growth factors that are expressed in a particular sequence and at various concentrations during development. In the process of limb patterning, mesenchymal cells in the limb bud integrate positional information from the three axes [2, 3].

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Embryogenesis of the extremities occurs between 4 and 8 weeks after fertilization, and the majority of congenital anomalies occur during this period of time. Limb bud development begins with the lateral migration of two layers of mesoderm and outgrowth into the overlying ectoderm. Cells from the underlying somitic mesoderm ultimately form the muscle tissue of the limb, while cells from the lateral plate mesoderm form cartilage and bones. Distinct but coordinated molecular pathways primarily control each axis. In the process of limb patterning, mesenchymal cells in the limb bud integrate positional information from the three axes, indicating a complex interplay of the responsible factors [4, 5].

Development of the proximal-distal axis is at least partially controlled by fibroblast growth factors (FGF) secreted by the apical ectodermal ridge (AER) [4]. The AER is a thickened layer of the ectoderm that forms over the distal edge of the limb bud. FGF secreted by the AER stimulates proximal-distal growth of the limb via differentiation of the underlying mesoderm. As part of a complex positive feedback loop, the signal to produce FGF is supplied by the underlying mesoderm. The AER signaling center is responsible for the differentiation of the underlying mesoderm and the development of the limb in the proximal-to-distal direction. Removal of the AER results in arrest of limb outgrowth. Furthermore, ectopic implantation of the AER results in formation of an extra limb. The AER also contributes to interdigital necrosis, allowing separation of the initially webbed hand [6]. Defects in the AER lead to anomalies such as limb truncation, transverse deficiencies, and syndactyly [7]. The genetics of the apical ectodermal ridge have been further elucidated as a complex interplay of related genes. Five paralogous gene families, named Hox 9 through Hox 13, from Hox A and Hox D gene groups, come together co-linearly (HoxA-9 to HoxA-13 and HoxD-9 to HoxD-13) and play important roles in the AER.

Development in the anteroposterior axis is patterned by secretion of sonic hedgehog (SHH) from the zone of polarizing activity (ZPA), a collection of cells along the posterior

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aspect of the limb bud. Transplantation of the ZPA from the posterior aspect to the anterior aspect of the limb bud causes creation of a mirrored duplication of the ulnar aspect of the hand. In addition to its primary role in development of the anterior-posterior axis, the ZPA also contributes to maintenance of proximal-to-distal limb development and participates in the feedback loop of the AER [4]. Mutations in Indian hedgehog, SHH, and PITX1 in humans have been implicated in lower extremity polydactyly, while mice studies have shown an interaction between the expression of each factor [2, 8].

Development in the dorsal-ventral axis is regulated by the Wingless-type (Wnt) signaling pathway within the dorsal ectoderm [3]. The Wnt pathway induces the underlying mesoderm to develop dorsal characteristics and is blocked in the ventral ectoderm, allowing the development of ventral characteristics. In mice, inactivation of the Wnt signaling pathway results in the development of biventral limbs. The Wnt pathway also contributes to regulation of SHH, reflecting the complex interaction and coordination among the 3-dimensional pathways responsible for limb development [4, 6].

It is unlikely that CFD is caused solely by a defect in the overall function of the AER, as the distal portion of an extremity affected by CFD is often normal. It is more likely that the defect is in the underlying mesodermal layer, within the complex interplay of developmental proteins at a point more downstream in the differentiation of the limb.

Pathophysiology and Genetics

A single underlying genetic cause for CFD has not been elucidated. In the senior author's personal experience (over 1,000 patients with unilateral CFD) there was only one unilateral case that had a parent with unilateral CFD. In contrast, in a smaller group of multi-limb congenital deficiencies including CFD on at least one limb, it was not uncommon to find a history of a first- or second-degree relative with a congenital limb anomaly. Several authors have postulated a genetic cause given that the presence of the condition at birth, its association with other abnormalities that have known genetic conditions, occasional bilateral involvement, and reports of familial cases are supporting evidence for an underlying inheritable genetic defect with incomplete penetration or an autosomal recessive pattern of inheritance [9–11]. It should be noted that phenotype has been used to diagnose and classify patients with CFD, despite quite varied clinical and radiographic presentations. As genetic and molecular understanding of the disease progresses, it is likely that grouping all patients into a single diagnosis of CFD will not be sufficient. Some of the reported abnormalities associated with CFD are fibular hemimelia, femoral-fibular-ulnar complex, patella aplasia/hypoplasia, absence of the fourth or fifth ray, unusual facies, Pierre-Robin sequence, and syndactyly of the toes. Considering the other congenital conditions that present in select individuals with CFD, the underlying pathophysiology and, therefore, genetic causes are likely to be distinct in this group of patients. In other words, one must be careful to assume that all patients *diagnosed* by today's criteria with "CFD" will have the same underlying causality.

An inheritable genetic defect or susceptibility to other risk factors or exposures at a particular stage in lower extremity development of the fetus would commonly present with bilateral involvement; however, bilateral involvement is rare, and associated with other known genetic defects, such as Pierre-Robin sequence. The Pierre-Robin sequence is primarily an anatomical defect of mandibular outgrowth that impacts on oropharyngeal volume and patency of the palate. No single genetic abnormality has been identified in Pierre-Robin, though recent evidence shows that Sox9 may be a crucial step in the pathway [12]. Similarly, it is likely that a variety of inheritable genetic defects are responsible for a common resultant phenotype with similar but varied congenital abnormalities. Conversely, a recent report of a child with Pierre-Robin and bilateral CFD found that array comparative genomic hybridization analysis showed no abnormalities at 1,543 loci while whole-exon analysis identified no mutations suspected to be causative for the patient's condition [13].

A subset of bone morphogenetic proteins (BMPs), known as growth/differentiation factors (GDFs), affect several skeletal processes, including endochondral ossification, synovial joint formation, and tendon and ligament repair. Studies in mice have suggested that GDF-5 deficiency affects the composition and material properties of cortical bone tissue in the femur, but the detailed mechanisms by which this occurs remain to be determined [4]. Paley et al. reported radiographic mineralization of the pseudoarthrotic region of the femur in patients with CFD with the application of exogenous BMP-2 directly to the area [14].

Other investigators have supported a teratogenic cause for CFD as the phenotype is similar to the effects of thalidomide exposure in utero [15]. These effects imply that at a particular stage in fetal growth an exposure in utero occurs causing a downstream effect on the remaining growth of the affected lower extremity. The exact stage in development at which the abnormality occurs could determine the severity of involvement. While a teratogenic exposure is possible as the underlying cause, given the embryology of the limb, the explanation would have to involve a teratogen with transient effect only at the time of exposure. Thus, fetuses exposed to the teratogenic agent at one time point might have only mild CFD, exposed only at the time of limb bud formation of the proximal femur, while those fetuses with more severe involvement have persistent exposure to the teratogenic agent.

Endogenous retinoids, including vitamin A and retinoid acid (the active form of vitamin A), have long have been implicated in limb development and abnormalities [5–7]. Embryological studies of quail, rats, and mice demonstrate a role for retinoic acid in the organization of the dorsoventral axis [7, 16]. These results suggest that a combination of maternal vitamin A supplementation and excess vitamin A in the neonatal period could potentially be unfavorable for cartilage development. It can be speculated that neonatal vitamin A supplementation may be beneficial for bone only if the mother's vitamin A status is low.

In summary, the embryologic and genetic data point towards a somatic mutation disorder of cells within the developing limb bud as opposed to a germ cell mutation (hereditary, familial) disorder for unilateral CFD. In contrast, multi-limb cases may be related to a germ cell mutation.

Deformity and Pathoanatomy

Osseous Deformities

CFD was initially grouped with cases of coxa vara, as this is the most obvious deformity in the majority of cases [17]. Cases were often separated into congenital short femur and proximal femoral focal deficiency (PFFD), for mild versus severe cases, respectively. Recognition that these were a spectrum of pathology of the same congenital deficiency leads to a consensus name change to CFD [18]. Although the femoral deformity and deficiency are the most obvious, non-femoral structures are also involved. These include the acetabulum, musculature, vessels, ligaments of the knee, tibia, fibula, and foot. In utero ultrasounds have shown both a shortened femur and a shallow acetabulum [19]. As with other pediatric proximal femur pathologies, dysplasia of the acetabulum whether initially present or not is progressive [20].

The deformities of the femur vary from coxa valga in very mild cases to severe coxa vara in more severe cases (Fig. 22.1). There is a flexion and retroversion deformity of the upper femur. The distal femur demonstrates a valgus deformity often referred to as a hypoplastic lateral femoral condyle since the distance from the physis to the knee joint line is smaller laterally than medially. In other words, there is a greater valgus convergence between the joint line and the distal femoral physis. The deformity of the proximal femur will be discussed in greater detail in a later section.

The ratio of growth in length of the short limb compared to the long limb remains relatively unchanged throughout growth [21–23]. This enables the final discrepancy in leg length to be predicted from the initial radiographs [24]. On this basis the Paley multiplier method is able to accurately calculate the predicted limb length discrepancy (LLD) at skeletal maturity [25, 26].

Ligamentous Structures

Anterior-posterior instability of the knee is common in CFD, but its severity is variable. Multiple modern era studies utilizing MRI have evaluated the ligamentous structures at the knee in patients with various degrees of CFD. The results support the conclusion that like the hip, the distal femur and proximal tibia require interaction of bony and soft tissues to develop normally.

Manner et al. [27] performed a radiographic study of 34 knees associated with CFD with radiographs and MRI. The anterior cruciate ligament (ACL) was affected in all knees studied, with 15 % hypoplastic and 85 % absent. The PCL was hypoplastic in 21 % and absent in 24 %. The most common type (14 knees, 41 %) was aplasia of the ACL and a normal PCL. The cruciate ligament dysplasias were differentiated into three groups (Fig. 22.2). Type I had a hypoplastic or absent ACL and a normal PCL. The intercondylar notch width and height were decreased compared to the normal side, and the lateral tibial spine was hypoplastic. Type II knees had aplasia of the ACL and hypoplasia of the PCL. They had even narrower and shorter intercondylar notches, and both tibial spines were hypoplastic. Two cases had lateral femoral osteochondral lesions. Type III knees had aplasia of both cruciate ligaments. The intercondylar notch was essentially absent and covered with hyaline cartilage, and both tibial spines were aplastic. The distal femoral joint surface is concave, matching the convex tibial plateau like a ball and socket. Three of these cases had discoid meniscus.

Overall, the authors found that on the tunnel-view radiographs, narrowing or absence of the femoral notch and flattening of tibial eminences corresponded to a hypoplasia or deficiency of the cruciate ligaments. Thus, the shape of the distal femur and proximal tibia can be used to predict, with relative accuracy, the presence or absence of the ACL and PCL ligaments.

These radiographic studies have been validated with the use of arthroscopy to directly confirm the presence or absence of the ACL and PCL. Johansson and Aparisi [28] published a case series of six patients with cruciate ligament dysplasia. Three patients had both an anterior and posterior drawer sign, and arthroscopy confirmed aplasia of both the ACL and PCL. Three patients had an isolated anterior drawer, and had ACL aplasia on arthroscopic examination. All knees were found to have hypoplastic tibial spines on radiographs.

Chugiak [29] looked at a larger series of 21 patients with clinical and arthroscopic examinations. All patients were found to have an anterior drawer sign, and nine patients (43 %) had a posterior drawer sign. Four patients (19 %) had medial instability, including one patient who also had lateral instability. The instrumented and clinic drawer tests were not found to be reliable enough, leading the authors to recommend imaging of the cruciate ligaments prior to lengthening

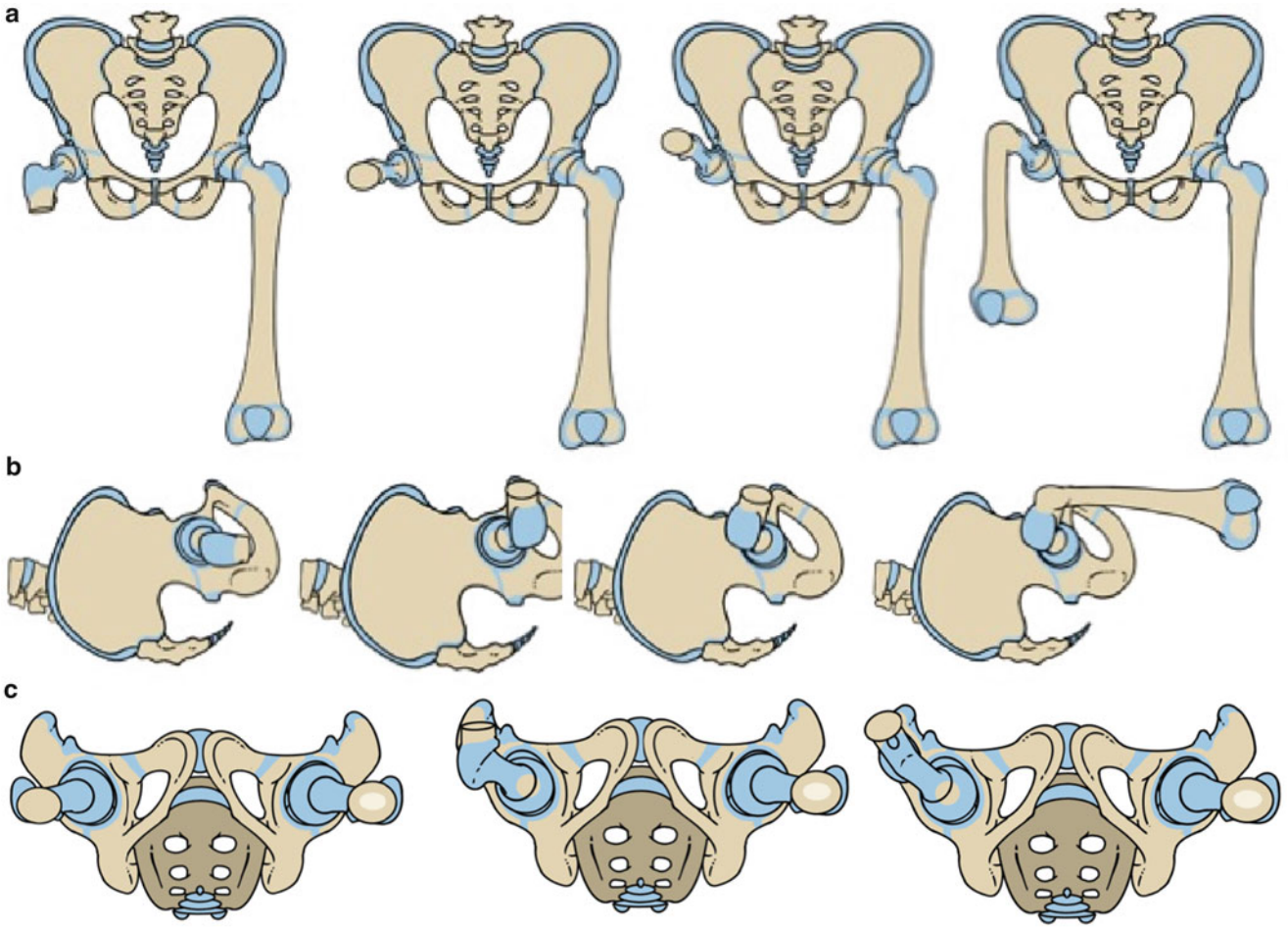


Fig. 22.1 Anteroposterior (**a**), lateral (**b**), and inferior-superior (**c**) views of pelvis and hip joints. These illustrations serve to simulate and recreate the proximal femoral deformity seen with severe CFD on the right hip. The model is based on a normal proximal femur with a 130° neck-shaft angle. First, flex the proximal femur 90° relative to the pelvis. Next, abduct the flexed proximal femur 45° relative to the pelvis. Now reconnect the distal femur to the proximal femur. The distal femur should be placed in 45° of external rotation relative to the pelvis. The resulting deformity is the CFD femur deformity typically seen in most type 1b and some type 1a cases. Note that the femoral neck appears to be retroverted due to the 90° hip flexion of a 130° neck-shaft angle femur. Just the flexion makes it appear to have 50° of retroversion. Since the distal femur is fixed to the proximal femur in external rotation this retroversion is increased even more. Also note that the varus deformity is caused by the abduction of the proximal femur with the hip flexed. The hip flexion places the greater trochanter facing posteriorly. The proximity of the greater trochanter to the ilium and to the sacrum in this position explains why the gluteus medius and minimus, and piriformis muscles, are short

to avoid knee dislocation. The majority of patients (38 %) were found to be completely deficient in both the ACL and PCL. The ACL was completely deficient in 16 (71 %) and hypoplastic in 3 (14 %) patients. The PCL was completely deficient in 10 (48 %) and hypoplastic in 3 (14 %) patients. Only one patient had an intact ACL and PCL.

In the majority of patients, both menisci were intact, with only three (14 %) hypoplastic and unrelated to the cruciate ligaments. Changes in the intercondylar notch and tibial eminences were noted in some patients but it was not specifically studied, as the authors felt that they were not as appreciable in patients under 6 years of age. They did note that the femoral intercondylar notch developed in some patients that had aplasia of both cruciate ligaments.

Muscle Pathoanatomy

Despite the wide spectrum of CFD, the underlying muscular anatomical differences appear relatively consistent. Pirani et al. showed that in patients with Aitken types A through D, the muscles were all present but altered in their size, structure, and location [30]. The study used MRI to qualitatively assess the musculature. The majority of muscles were found to be smaller: gluteus maximus, gluteus medius, gluteus minimus, quadriceps, adductor magnus, adductor longus, adductor brevis, pectineus, semimembranosus, semitendinosus, and biceps femoris. The exception is the sartorius muscle that was found to be hypertrophied, to which Pirani et al. and Panting et al. attributed as the underlying cause of the deformities of the proximal femur, given the sartorius'

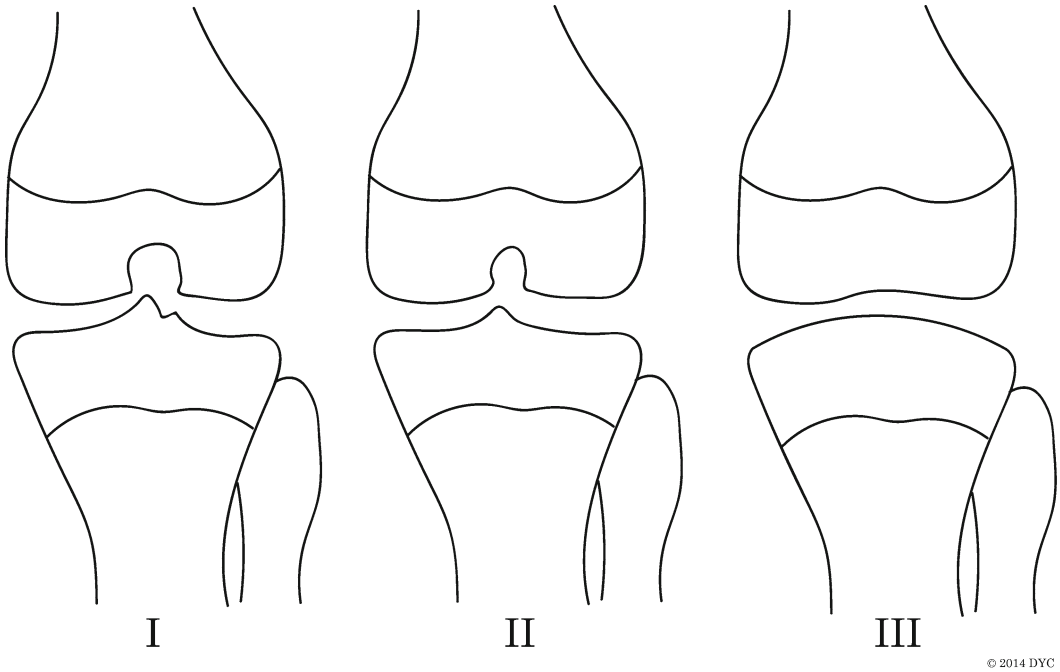


Fig. 22.2 Manner et al. [27] classification of intercondylar notch using the tunnel view correlates to deficiency of the cruciate ligaments. Type I: The intercondylar notch width and height are decreased compared to the normal side. The lateral tibial spine is hypoplastic. This corresponds to a hypoplastic or absent ACL and a normal PCL. Type II: Narrower and shorter intercondylar notch with hypoplasia of both tibial spines. This corresponds to aplasia of the ACL and hypoplasia of the PCL. Type III: The intercondylar notch is absent and covered with hyaline cartilage, and both tibial spines are aplastic. The distal femoral joint surface is concave, matching the convex tibial plateau. This corresponds to aplasia of both cruciate ligaments

orientation [31]. The obturator externus was found to be elongated and the muscle belly extended almost entirely to its insertion. The short external rotators had a larger cross-sectional diameter and were found to insert on the posteromedial greater trochanter. Overall, the course of the muscles was more perpendicular and proximal than normal, inserting onto the proximally migrated femur.

We have also found this to be true in our surgical experience: the musculature is present, the muscle bellies extend nearly to their insertion, and the course of the muscles is proximal. Panting and William had previously noted similar findings, including the hypertrophied sartorius muscle, the relatively normal gluteal muscles, and a hypoplastic quadriceps muscle [31]. Biko et al. also used MRI in seven patients to primarily evaluate the osseous structures, and reported a qualitative decrease in size of the musculature in general. At the level of the triradiate cartilage, the cross-sectional size of the gluteus muscles was significantly smaller in comparison to the uninvolved contralateral limb [32].

Pirani et al. postulated that the muscles became the primary stabilizers of the deformed hip joint, since the osseous structures did not impart inherent stability. However, in our experience the majority of untreated patients with CFD do not report ipsilateral hip pain, even after knee fusion and with prosthetic use and weight bearing.

Vascular Pathoanatomy

The normal embryological development of the arteries of the lower extremity parallels the formation of limb buds, occurring between the 4th and 8th weeks of gestation. The limb begins with a single axial dorsal artery that continues as the ischiadic artery and on to the popliteal artery. The external iliac artery arises at 5 weeks of gestation and bifurcates into the inferior epigastric artery and the femoral artery, which then bifurcates at 6 weeks gestation into the lateral and medial branches. The medial branch becomes the deep femoral artery and gives rise to the medial and lateral circumflex arteries and connects to the ischiadic artery, while the lateral branch develops into the femoral artery. By 7.5 weeks the femoral artery caliber is larger than the ischiadic artery and becomes the major blood supply to the popliteal artery and leg. After full development, the dorsal axial artery remains as the inferior gluteal artery while the ischiadic artery remains as the arteria comitans nervi ischiadici, running as the vasa vasorum of the sciatic nerve [33].

The vascular anatomy is also usually abnormal in an extremity affected by CFD. Chomiak et al. reported the results of 21 patients with various degrees of CFD studied with computed tomography (CT) angiograms to identify vascular abnormalities [34]. The authors found that more severe cases of CFD had a smaller diameter and a shorter length of

the femoral artery; however, the severity of osseous abnormalities did not directly correlate with the topographical vascular anatomy abnormalities. All patients, at the least, had differences compared to the contralateral unaffected extremity: mainly with smaller vessel caliber, decreased number of vessels to the thigh, and more proximal bifurcation of the external iliac into femoral artery and deep femoral artery. Despite this, 19 of the 21 patients had the blood supply to the femur and pseudoarthrosis from branches of the deep femoral artery, which originated from the external iliac artery.

Notably, Chomiak found 2 of the 21 patients with a persistent ischiadic artery as the dominant vascular supply to the leg and a diminutive femoral artery that supplied solely the medial thigh [34]. This is an extremely unusual finding in otherwise normal humans, but is seen in lower mammals [35].

It is critical to identify these vascular anomalies preoperatively when reconstruction using rotationplasty is considered. The senior author has identified several cases with a dominant ischiadic artery and absent superficial femoral artery while performing rotationplasty. In our opinion, an MR angiogram (MRA) is indicated in all such cases prior to a rotationplasty.

Box 22.1. CFD Pathology

- The incidence of CFD is approximately 1 in 50,000.
- Limb bud formation is affected by a complex interplay of signaling, including AER, ZPA, and Wnt.
- CFD is likely caused by a somatic mutation during the development of the limb bud, but some cases may be caused by an inherited germ cell mutation.
- CFD usually presents with proximal coxa vara and distal femoral valgus.
- Cruciate ligament deficiency is common and can often be identified on radiographs.
- Generally, most muscles around the pelvis are present and hypoplastic, though the sartorius may be hypertrophied.

  An ischiadic artery may be present and more dominant than the femoral artery in some patients with CFD.

Evaluating the Child with Unilateral CFD

History

Most children born with unilateral CFD have no family history of this or other congenital anomalies. Nevertheless, inquiry should be made into family history, exposure to drugs, medications, radiation, or infectious diseases during the first trimester. Many cases of CFD are now identified with prenatal ultrasound early in the pregnancy by measuring the lengths of the two femurs. In such cases the predicted leg length discrepancy at birth and maturity can be calculated using the multiplier method [36].

Physical Exam

There is an obvious leg length discrepancy. Associated fibular hemimelia (FH) and ray deficiency may be present. The hips, knees, and ankles should be examined for range of motion and flexion contractures. Neonates and young infants normally have such contractures for the first 3–6 months. At the hip, patients may have external rotation and fixed flexion deformity of the hip, as well as limited abduction when coxa vara is present. At the knee, patients may have flexion contractures, hypoplastic or subluxation-dislocation of the patella, AP, or rotary instability of the knee.

Finally, muscle length tests are recorded to identify preexisting limits to muscle lengthening. These tests include the Ely test for rectus femoris tightness, popliteal angle measurement for hamstring tightness, and Ober sign for fascia lata or iliotibial band tightness (Fig. 22.3).

Box 22.2. Physical Examination Findings Often Seen in Patients with CFD

- Hip: External rotation (ER) deformity or increased ER versus internal rotation (IR); fixed flexion deformity (FFD) of hip; limitation of abduction (when coxa vara is present).
- Knee: Fixed flexion deformity of knee; no limitation of knee flexion; hypoplastic patella; lateral tracking or subluxated or dislocated patella; anteroposterior instability of knee; rotary instability of knee; anterior dislocation of tibia on femur with knee extension followed by reduction of knee with attempted flexion; hypermobile meniscal clunks; temporary locking of the knee during flexion.
- Ankle: Limitations of ankle dorsiflexion; obligatory eversion with dorsiflexion; hypermobility of ankle with increased eversion; lateral malleolus more proximal than medial malleolus.

Imaging

Radiographic Examination

Radiographs should include a full-length anteroposterior (AP) standing legs with the patellas pointing forward. In children who are unable to stand, a pull-down X-ray may be



Fig. 22.3 Muscle lengthening tests: (**a**) The Ely test checks for rectus femoris tightness, demonstrating pelvic flexion with prone knee bend; (**b**) the popliteal angle is the angle between the vertical and the line of the tibia with the hip at 90° flexion and the knee maximally extended; (**c**) the Ober test checks for fascia lata tightness; a positive sign is the thigh going into an abduction position when the hip is hyperextended and the knee flexed to 90°

performed. It allows measurement of the length of the femurs and tibia, though it does not include foot height. Long lateral leg radiographs in maximum extension allow for evaluation of knee flexion contractures and more accurate length measurement. A supine AP pelvis allows more accurate measurement of the center-edge angle (CEA) of both hips to assess for hip dysplasia. It is also a better quality X-ray to assess for ossification of the femoral neck. It is important that the pelvis be level for more accurate measurement.

Magnetic Resonance Imaging

Magnetic resonance imaging (MRI) is useful to assess the integrity of the proximal femur. It can help determine whether the femoral head is joined to the shaft of the femur via a cartilaginous femoral neck. In case of a femoral neck pseudoarthrosis, the MRI can help determine if the cartilage of the femoral head is fused to the acetabular cartilage. For optimal imaging, the cuts of the proximal femur should be reformatted in an oblique plane to see the entire proximal femur as a single image. MRI can also help outline the intra-articular pathology of the knee, identifying deficiency of the cruciate ligaments and outlining the shape of the joint surfaces in the frontal and sagittal plane.

Computerized Tomography (CT)

CT is only useful at an older age when the acetabulum and proximal femur are nearly fully ossified. Three-dimensional CT reconstruction is useful to compare the normal acetabulum with the dysplastic side. In older children 3D CT can show the pathologic anatomy.

Box 22.3. Evaluating the Child with CFD

- CFD can be bilateral; check the contralateral side as well.
- The hip, knee, and ankle should all undergo thorough physical examinations.
- Fibular hemimelia is commonly associated with CFD.
- Radiographic imaging should include a full-length AP legs (standing or pull-down), long leg laterals in maximum extension, and a supine AP pelvis.
- MRI is useful to evaluate soft tissue, ligaments, and a cartilaginous femoral neck.

Classification Systems

Multiple classification systems have been described over the years in attempts to categorize the pathology and direct surgical treatment (Figs. 22.4, 22.5, and 22.6). Most older classification schemes were based on plain radiographs. Newer classification systems incorporate modern imaging modalities, specifically MRI, which allows detailed evaluation of osseous, cartilaginous, and soft-tissue structures, including the non-ossified femoral head, non-ossified acetabulum, labrum, pseudoarthrosis, and musculature [32]. The ability to perform MRI under anesthesia for infants allows for interpretation of these images with better reliability. As a result, MRI dramatically improves the ability to correctly categorize patients over conventional radiographs alone [37]. Table 22.1 shows a comparison of several of the classification schemes further elucidated below.

Aitken, 1959

In 1959 Aitken proposed a four-class schema for PFFD (see Fig. 22.4) [17]. This became the most commonly utilized classification system for PFFD. Aitken focused the classification system on the development of the femoral head, neck, and acetabulum, with progressive dysplasia from one class to the next. The Aitken classification does not include the congenital short femur group discussed earlier.

Aitken initially proposed that Class A patients were not technically PFFD, as all the structures were present, albeit deformed. These patients had a femoral head, a cartilaginous femoral neck, an adequate acetabulum, and a very short femoral segment. Severe subtrochanteric femoral varus and femoral shortening were appreciated. A pseudoarthrosis of the proximal femur was appreciated between the femoral shaft and head, either between shaft and trochanteric component (subtrochanteric pseudoarthrosis) or between trochanteric component and head (femoral neck pseudoarthrosis). Aitken noted that the pseudoarthrosis did not spontaneously resolve in all patients by skeletal maturity, but did eventually heal in some patients.

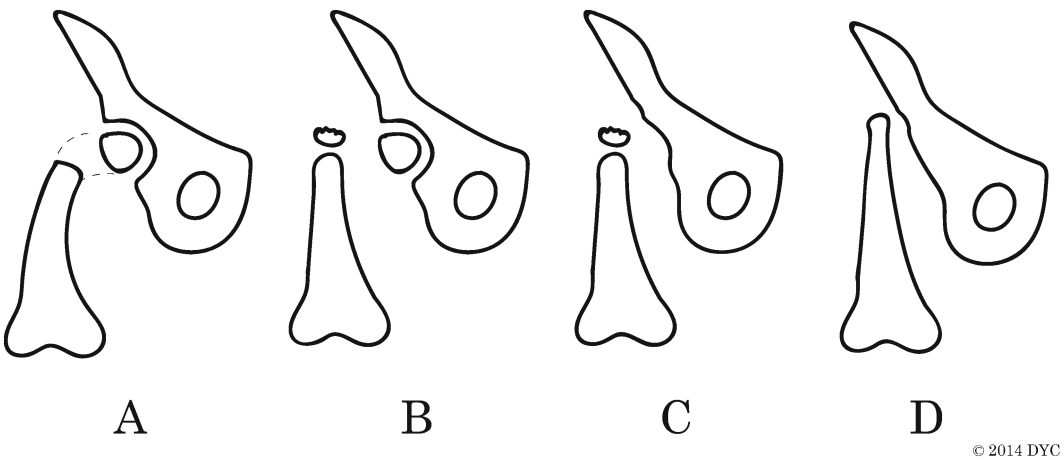


Fig. 22.4 Aitken classification of congenital femoral deficiency. The Aitken classification is based on the relationship of the acetabulum and the femoral head: (**a**) Adequate acetabulum, femoral head is present and attached to shaft; (**b**) adequate acetabulum, femoral head not connected to shaft, which instead has a proximal ossified tuft; (**c**) dysplastic acetabulum, minimal or absent femoral head, disconnected femoral shaft with tuft; and (**d**) no acetabulum, no femoral head, shortened proximal femoral shaft with no tuft

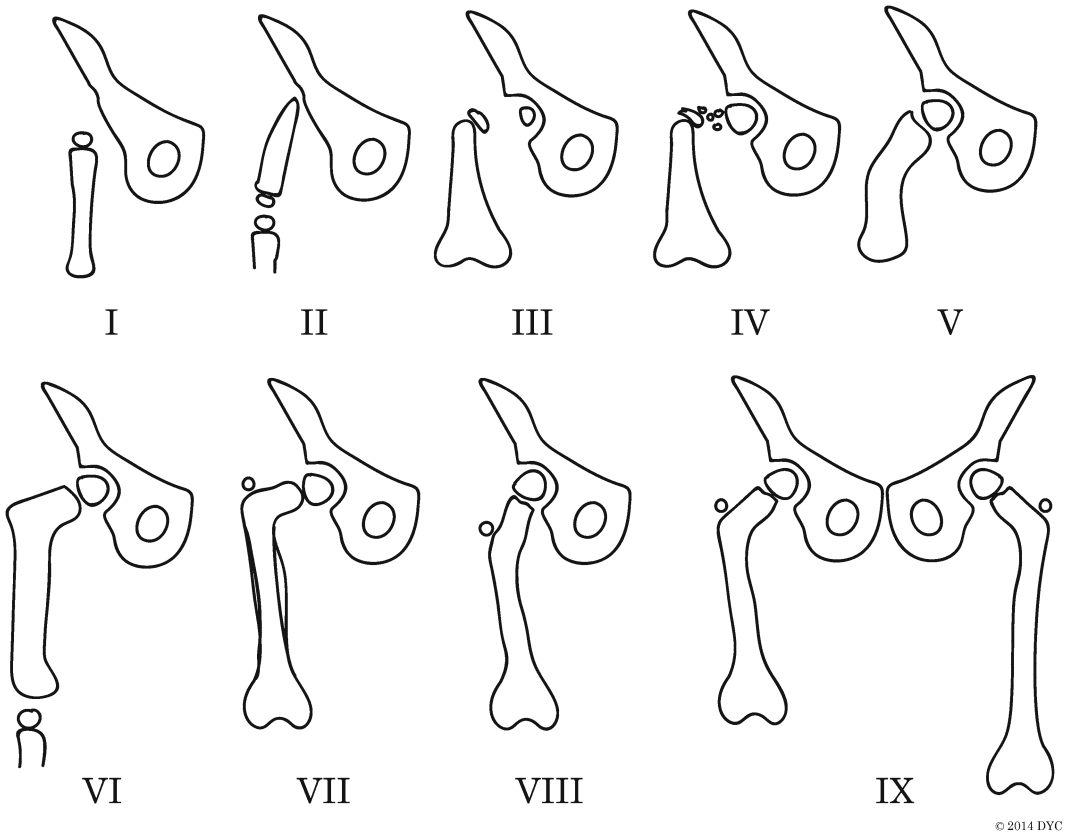


Fig. 22.5 Pappas classification of congenital femoral deficiency. The Pappas classification is divided into nine classes: (I) congenital absence, (II) proximal femoral and pelvic deficiency, (III) proximal femoral deficiency with no osseous connection between femoral shaft and head, (IV) proximal femoral deficiency with disorganized fibrosseous disconnection between femoral shaft and head, (V) midfemoral deficiency with hypoplastic proximal and distal development, (VI) distal femoral deficiency, (VII) hypoplastic femur with coxa vara and sclerosed diaphysis, (VIII) hypoplastic femur with coxa valga, and (IX) hypoplastic femur with normal proportions

The hallmark of the Aitken Class B PFFD was a lack of connection, either bony or cartilaginous, between the femoral shaft and head that persists into skeletal maturity. These patients have a femoral head within an adequate acetabulum and a shortened, deformed femoral shaft. The proximal portion of the shaft often has an ossified tuft. The femoral head and shaft do not move as a unit.

Aitken Class C is defined by a severely dysplastic acetabulum. The femoral head does not ossify and shaft of the femur is short with an ossified tuft at the proximal end of the shaft. There is no cartilaginous or osseous femoral neck or trochanteric component.

Aitken Class D is the most dysplastic form of PFFD. This class is characterized by the absence of the acetabulum and femoral head and a deformed, shortened femoral shaft without tufting on the proximal shaft of the femur.

Pappas, 1983

The Pappas classification (see Fig. 22.5), published in 1983 [38], was based on the anatomy of 139 cases evaluated in Massachusetts. Pappas describes nine classes based on the femoral deficiency and associated deformity, with diminishing severity as the class number increased. This was the first classification scheme to separate middle and distal femoral deficiencies into their own class. Recommendations were given for treatment for each class; however, treatment was depen-

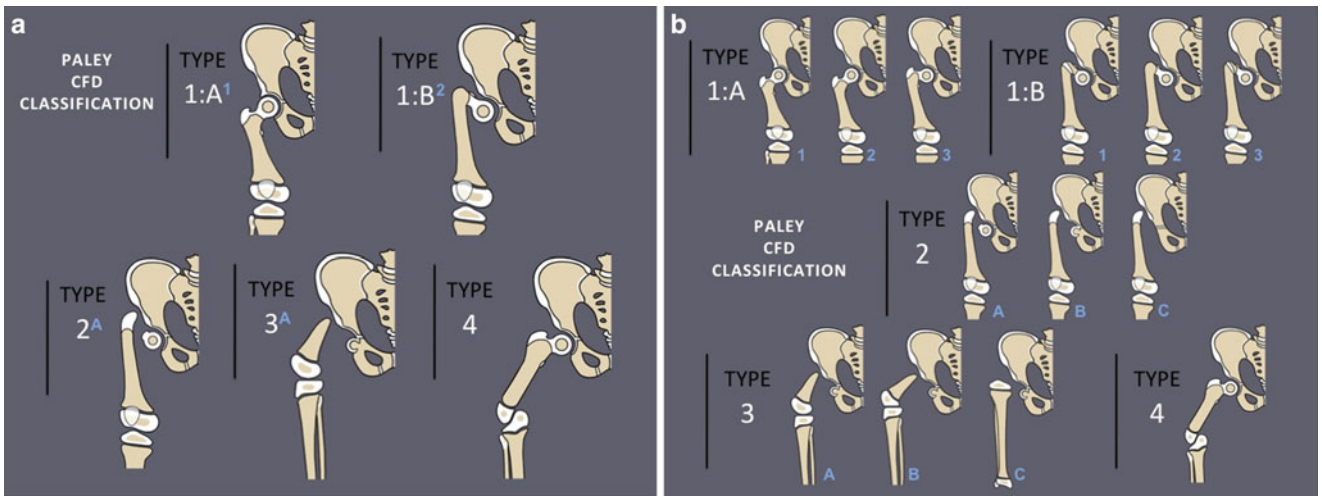


Fig. 22.6 (**a**) Paley classification of congenital femoral deficiency. There are four types of congenital femoral deficiency: (1) intact femur normal ossification (**a**) or delayed ossification (**b**); (2) mobile pseudoarthrosis; (3) diaphyseal deficiency; (4) distal deficiency. (**b**) Each Paley type has several subtypes: $1a_1$: Normal-shaped short femur with mild genu valgum; $1a_2$: normal-shaped short femur with mild genu valgum and retroversion; $1a_3$: coxa vara, retroversion, and mild genu valgum; $1b_1$: subtrochanteric type; $1b_2$: neck type; $1b_3$: combined neck and subtrochanteric type; 2a: mobile femoral head; 2b: partially fused femoral head; 2c: completely fused or absent femoral head; 3a: mobile knee joint with greater than 45° of motion; 3b: stiff knee with less than 45° of motion; 3c: absent femur or fused knee joint

Table 22.1 Comparison of different classification schemes

	Paley [42]	Pappas [38]	Gillespie and Torode [39]	Hamanishi [15]	Aitken [17]
Normal anatomy/minimal deformity/ hypoplasia	1A	7,8,9	Group 1	Type 1, type 2	Class A
Subtrochanteric delayed ossification	1B subtrochanteric	4, 5	Group 1	Type 3a	Class A
Femoral neck delayed ossification	1B femoral neck	3	Group 1	Type 3b	Class A
Combined delayed ossification	1B combined	3		Type 4a	
True pseudoarthrosis with no cartilaginous or osseous femoral neck	2a, 2b	3	Group 2	Type 4b	Class B
Dysplastic acetabulum, no femoral head	2b, 3	2	Group 2	Type 4c	Class C
No proximal femur, short distal femur only	3a,b	2	Group 2	Type 4c	Class D
Femoral absence	3c	1		Type 5	
Distal femoral deficiency only	4	6			

dent on the patient's joint stability and function, which are not part of the classification system. Therefore treatment could not be based directly on the classification schema of Pappas.

Gillespie and Torode, 1983

In 1983, Gillespie and Torode proposed a straightforward division of CFD into two categories: congenital short femur and true PFFD [39, 40]. Each of the two groups had both radiographic and clinical criteria, but were ultimately defined by the function of the hip and knee joints, rather than the type of surgery each would necessitate.

Group 1 was defined as those patients who could have good hip and knee function with or without surgical correction. Radiographically, group 1 patients had coxa vara, lateral femoral bowing, and hypoplasia of the knee with valgus but the predicted leg length discrepancy was no greater than 30 % of the normal femur and femoral deficiency was 40–60 % of the contralateral femur. Clinically this group had laxity in the sagittal plane and mobile joints without flexion deformities at hip or knee.

Group 2 patients had hip and knee joints that were not adequate to support the patient without pain. The appropriate treatment recommendation for this group of patients was amputation and/or rotationplasty of the lower limb. These

patients had markedly short femurs. Radiographically, these patients had less than 40 % of the femoral length of the normal side and deficiency or absence of the femoral head and/or neck. The authors defined deficiency of the femoral head as a tenuous cartilaginous connection between femoral shaft and head. Clinically, these patients had fixed flexion deformity at hip and/or knee.

The overall goal of the Gillespie and Torode classification was to simplify the decision making. The authors rationalized for families and surgeons a "life plan" regarding treatment of the affected limb based on each of the two groups. This classification aims to aid families and physicians communicate more effectively and correlate early clinical signs with long-term treatment plans. The value of this classification schema is its simplicity and its guidance in surgical treatment. The drawbacks to this classification scheme are its inability to adapt to improved surgical techniques based on a better understanding of pathoanatomy, as well as advances in biology and technology.

Hamanishi, 1980

Hamanishi [15] published a five-tiered classification system for congenitally short femur. This schema involved a scoring system that included the entire lower limb and upper limb involvement, as opposed to other classification systems, which focus only on the femur. However, the schema does not include specifically the acetabulum, as the author believed that this component is directly related to the femoral head. It was intended to differentiate those patients with the teratologic sequelae of thalidomide exposure from those patients with idiopathic CFD. The diagnosis was based on a series of 60 total patients, 14 of which were known to have had in utero thalidomide exposure. It is an observational classification system and does not aim to guide treatment or prognosis. The study was quite comprehensive as the authors included socioeconomic class, an extensive family history, parental age, birth history, and in utero history in their review. The age of the study subjects at the time of evaluation varied from 6 months to 50 years.

Hamanishi identified several differences between the two groups: overall, the thalidomide exposure group had femora that were more severely affected than the idiopathic group; the thalidomide exposure group more often had concomitant abnormalities of the tibia and radius as opposed to the fibula and ulna in the idiopathic CFD group; the thalidomide group had lower extremity preaxial (tibial) polydactyly, whereas the idiopathic group had lower extremity postaxial (fibular) deficiency. The author defined five groups with subgroups in some utilizing radiographic measurements of the affected femur to quantify the degree of deformity.

Group 1 was defined by hypoplasia of the femur, either without deformity type 1a or with minimal angulation and cortical thickening, type 1b. The type 1a was associated with ipsilateral below the knee deformities, while the type 1b was associated with severe, symmetric bilateral below the knee deformities. The second group was defined by hypoplasia and femoral varus angulation. Type 2a had a normal neck-shaft angle but type 2b had a decreased neck-shaft angle. A transverse subtrochanteric ossification defect was noted in this type as well, but all ossified later in life.

The type 3 group had marked coxa vara from delayed and inconsistent ossification of the femoral neck and subtrochanteric region. This group also has marked retroversion of the femoral neck and distal femur external rotation. Type 3a with a straight femoral shaft was considered "stable" as the proximal femoral varus would not progress, while the "unstable" type 3b with an increased epiphyseal head angle >60° was reported to progress with time.

The type 4 femurs had absent or stunted and tapered proximal femoral shafts with the femoral head attached directly to the shaft, when present. The femoral head was connected to the shaft by a pseudoarthrosis and the entire shaft was proximally migrated. The type 4a had a pseudoarthrosis of the femoral neck and subtrochanteric region. The type 4b had an absent proximal femur with the femoral head connected directly to a tapered proximal shaft. The type 4c had a complete absence of the proximal femur.

Type 5 femurs indicated a rudimentary femur, which ossified later, or complete absence.

One major flaw in this classification system is that it divides the groups based on measurements with arbitrarily chosen cutoffs, such as the neck-shaft angle and femoral varus which change with time. Thus, young patients could move from one group to another without intervention and the treatment plan would also therefore be variable. Additionally, this classification scheme does not include the stability of the hip or knee, and the presence of knee ligaments, and does not guide surgical or nonsurgical treatment.

Some important remarks from this study are that the leg length discrepancy was consistent, when stated as a percentage of the unaffected normal side, which was consistent with older retrospective reports [21, 22] and newer mathematical methods to predict limb leg discrepancy at maturity [26].

Kalamachi, 1985

The Kalamachi classification (1985) [41] attempted to relate the femoral deficiency directly to the limb function. Type I patients had congenital shortening of the femur with a normal hip and knee morphology and function. Type II patients had a congenitally short femur with coxa vara, with some irregularity of the growth plate, a located hip, and a well-developed acetabulum.

Type III patients had a developed acetabulum but a deficient proximal femur. With time, they were separated into two groups. In the IIIA group, the radiographically apparent proximal femoral defect ossifies into a bony bridge, often in varus. In the IIIB group, the defect does not ossify, and develops into a pseudoarthrosis. Type IV patients had no acetabulum and no connection between the pelvis and the distal femoral fragment, which often was a spike proximally. Type V patients had no hip joint and no femur. For all five classification types, the most common treatment was a Boyd or Syme amputation. Knee fusions were used to correct deformity as well as provide a large lever arm for a severely shortened femoral segment. Femoral osteotomies were used mostly in type II and IIIA patients to correct coxa vara. Limb equalization procedures were done in nine patients (four epiphysiodesis, five lengthening) of type I and II. The rest were treated non-operatively or with orthotic and prosthetic fitting.

Paley, 1998

Paley proposed a classification system for CFD (first presented at a symposium in Dallas in 1996 documented in a book published from that symposium [42]) based on the degree of development and integrity of the femur (see Fig. 22.6). Unlike the previous classifications, Paley wanted to create a classification where the type would not change with the degree of ossification of the femur (age dependent). A longitudinal follow-up of different classification systems by Sanpera and Sparks [43] showed that the existing classification systems were inaccurate in predicting the final femoral morphology based on the initial radiographs. Paley also wanted a classification that was oriented towards reconstruction and not amputation. As such, the classification types and subtypes each have a separate surgical reconstructive prescription.

Box 22.4. Femur Deficiency Classification (Paley, 1998) [42]

Type 1: *Intact femur* with mobile hip and knee:
(a) Normal ossification proximal femur
(b) Delayed ossification proximal femur (neck, subtrochanteric, or combined neck subtrochanteric types)

Type 2: *Mobile pseudoarthrosis* (greater trochanteric apophysis present), knee usually mobile:
(a) Femoral head mobile in acetabulum
(b) Femoral head partially fused to acetabulum
(c) Femoral head and acetabulum completely fused or absent

Type 3: *Diaphyseal deficiency* of femur (greater trochanteric apophysis absent):
(a) Distal physis present; knee motion ≥45°
(b) Distal physis present; knee motion <45°
(c) Complete deficiency of distal femur or fusion of distal femoral remnant to tibia (distal physis absent)

Type 4: *Distal deficiency of femur* (proximal end normal)

Treatment Options for Congenital Femoral Deficiency

Rotationplasty

Rotationplasty has been a commonly accepted treatment for patients with CFD with significant proximal deficiency, which is deemed "unreconstructable" [44–48]. The most commonly employed surgical technique is the Van Nes rotationplasty and variations of this technique. In the Van Nes rotationplasty, the lower limb is rotated 180° to use the ankle and foot as a functional knee joint with a prosthesis. The rotationplasty can be combined with a knee fusion when the knee is unstable, or as in the Brown modification the knee is maintained after 180° rotation and the distal femur is fused to the pelvis to substitute for a deficient hip allowing flexion and extension [44]. This will be discussed in more detail in a later section.

Gillespie and Torode reported their surgical recommendations along with their classification schema in 1983 [39, 40]. Prior to the advent of the Wagner device in 1971, patients classified as Group 1 with congenital short femur underwent rotationplasty. After the Wagner technique demonstrated the feasibility of lengthening up to 20 cm, femoral lengthening became the standard recommendation for Group 1, but to a maximum of 20 % of the femur length because the risk of hip and knee subluxation or dislocation was considered too high. Of the five Group 1 patients that underwent rotationplasty, three had good function but two required a Syme amputation and fitting with an above-the-knee prosthesis. Patients classified by Gillespie and Torode as Group 2 had true PFFD and received rotationplasty, Syme amputation for prosthesis fitting, and/or knee fusion. Of 43 Group 2 patients with instability of the hip and knee, 21 underwent rotationplasty, and of those, 8 required de-rotational surgery and 2 required a Syme amputation. They strongly advocated knee fusion for Group 2 patients to correct the valgus and flexion deformities at the knee and

tibial rotationplasty to restore "knee flexion" with the ankle dorsiflexion.

Brown reported on three patients with severe CFD who underwent a modification of the Van Nes rotationplasty with mean of 6-year follow-up who had satisfactory results and range of motion of both the hip and knee to 90° of flexion [44]. The Brown modification includes resecting the deficient proximal femur, externally rotating the limb 180°, fusing the femoral remnant to the pelvis, and leaving the muscles distal to the knee undisturbed. Using this technique there was no derotation as previously recurred with the Van Nes rotationplasty which incorporated a midtibial osteotomy for rotation.

Ackman et al. reviewed 12 patients who underwent Van Nes rotationplasty for CFD to determine the long-term outcome [48]. At a mean of 21.5 years (11–45) after their rotationplasty, a total of 12 prosthetic patients were compared with 12 normal age- and gender-matched controls. The authors found no differences between the groups in overall health and well-being on the SF-36, but significant differences were seen in gait parameters in the CFD group. Patients who had undergone Van Nes rotationplasty had a high level of function and quality of life at long-term follow-up, but presented with significant differences in gait and posture compared with the control group.

Syme Amputation

Alman et al. reviewed the results of treatment of 16 patients who had had an isolated unilateral PFFD; 9 were managed with a rotationplasty and 7 with a Syme amputation combined with an arthrodesis of the knee [49]. The perceived physical appearance, gross motor function, and metabolic energy expended in walking were assessed. The mean duration of follow-up was 9.9 years (range, 4–14 years). The mean age of the patients at the time of the study was 13.9 years (range, 8–18.4 years) in the rotationplasty group and 14.8 years (range, 9.5–19.9 years) in the Syme-amputation group. There were three female patients in each group. Roentgenograms showed that the femoral head was in the acetabulum (Aitken class A or B) in four of the seven patients in the Syme-amputation group and in five of the nine patients in the rotationplasty group; the remaining patients did not have this finding (Aitken class C or D). There was no difference in gross motor function or perceived physical appearance between the groups. Rotationplasty was associated with a more energy-efficient gait than was Syme amputation.

Fowler et al. measured lower limb kinematics and kinetics during preferred and fast speeds of walking in persons with PFFD to compare outcomes after Syme amputation (nine subjects) with those after Van Nes rotationplasty (ten subjects) [50]. Subjects with a Van Nes rotationplasty and full tibial rotation (seven subjects) demonstrated prosthetic knee function during stance as they were able to support a flexed-knee posture at both speeds and produced greater knee-extensor moments at preferred speeds as compared with the Syme group. Non-prosthetic limb compensatory mechanics were significantly exacerbated in subjects with a Syme amputation compared with the Van Nes group: (1) stance-phase vaulting, resulting in greater inappropriate ankle-power generation at both walking speeds; (2) excessive hip-extensor moments at fast speeds; (3) excessive hip-power absorption and generation at both speeds; and (4) excessive knee-joint power generation at both speeds. The improved gait after Van Nes rotational osteotomy is one factor that should be considered when making clinical decisions for children with PFFD.

Other studies have found some potential issues with Syme amputation in the long term: heel pad migration, skin sloughs, and problems with prosthetic fitting. Anderson et al. reviewed 69 Syme amputations performed in 62 children with the major indication of leg length discrepancy, due to either paraxial fibula hemimelia (33 cases) or proximal focal femoral deficiency (19 cases) [51]. The average age at amputation was 5.6 years, with an average follow-up of 10.5 years (range 1–25 years). Although the results were assessed by a combination of chart review, patient recall examinations, and questionnaires, satisfaction in adulthood was found to be high. Early complications included three skin sloughs and one infection. Late complications included 2 retained os calcis apophyses, 1 exostosis, and 16 cases of heel pad migration. Only one of the heel pad groups required revision; prosthetic adjustment resolved symptoms in the remaining patients. However, prosthetic knees were often too low because of failure to limit the length of the stump appropriately, though this finding was not as problematic in the CFD group as the residual limb was significantly shorter.

Limb Lengthening

Aston et al. published a series of 27 patients with Paley type 1 CFD (Pappas grades VII, VIII, and IX) who underwent a total of 30 lengthening procedures, with 3 patients undergoing a second femoral lengthening [52]. All patients underwent femoral lengthening with a multiplanar Ilizarov-type external fixator. The mean increase in length was 5.8 cm (3.3–10.4 cm) and 18.65 % of the total length of the femur (9.7–48.8 %). The mean time in the frame was 223 days (75–363) with a mean distraction index of 1.28 months per cm. The authors initially performed the osteotomy distal, but after 17 distal osteotomies changed to a proximal osteotomy for lengthening, noticing a significant increase in mean range of knee motion from 98.1° to 124.2° ($p = 0.041$) and a trend

towards a reduced requirement for quadricepsplasty, although this was not statistically significant ($p=0.07$). The overall incidence of regenerate deformation or fracture requiring open reduction and internal fixation was similar in the distal and proximal osteotomy groups (56.7 % and 53.8 %, respectively). However, in the proximal osteotomy group, preplacement of a Rush nail reduced this rate from 100 % without a nail to 0 % with a nail ($p<0.001$). When comparing a distal osteotomy with a proximal one over a Rush nail for lengthening, there was a significant decrease in fracture rate from 58.8 % to 0 % ($p=0.043$). This supports the premise of Paley and Herzenberg that CFD patients should be lengthened over a Rush rod or have a Rush rod inserted after frame removal to reduce the risk of regenerate bending or fracture [53].

Aston et al. concluded that lengthening of the femur with an Ilizarov construct should be carried out through a proximal osteotomy over a Rush nail and lengthening to a maximum of 6 cm during one treatment, or 20 % of the original length of the femur, to reduce the risk of complications [52]. Paley and Standard have published on a distal osteotomy for CFD to avoid lengthening through the abnormal, sclerotic bone often seen in the proximal femoral shaft as much as possible with good success [54, 55].

Saghieh and Paley studied a group of 95 CFD patients who had undergone lengthening between the years 1988 and 2000 (Table 22.2). These patients did not undergo the Superhip or Superknee procedures that are described later in this chapter. All femoral lengthenings were with the Ilizarov device with extension of the fixator across the knee to the tibia with hinges. The postoperative result based on knee and hip range of motion score, gait score, lengthening goal score, and alignment score, pain score, and activity level score was excellent and good in over 93 % in all groups, with a mean overall lengthening of 6.0 cm (range, 2.2–12.5). Complications included femur fracture, pin site problems requiring surgery, premature consolidation, nerve irritation/palsy, delayed union, knee subluxation, and hip subluxation. The fracture rate in the older age group was significantly lower than the other groups, but it included several patients that were undergoing lengthening over nails. There was no significant difference in unplanned surgery rate between groups 1 and 2 (see Table 22.2). The difference with group 3 was related to refractures. Prophylactic rodding was not performed at the time of fixator removal in this series of patients, but is now part of the standard treatment algorithm, given the high rate of fractures [56].

The authors have previously presented their results on femoral lengthening after a hip stabilization procedure [57]. A retrospective review was performed of 35 patients with CFD, Paley types 1a and 1b, who underwent femoral lengthening after a pelvic osteotomy, proximal femoral osteotomy, or a combination. Patients underwent a hip stabilization procedure at a mean age of 2.4 years (2–5.5). The mean age at first femoral lengthening was 3.7 years (3–10.7). The mean overall limb length difference prior to femoral lengthening was 64.8 mm (47–100), with a mean postoperative difference of 8.1 mm (−14 to 32). The average amount of time in the external fixator was 186.1 days (107–311), or 1.1 months/cm. Preoperatively, average knee ROM was 2°–131° and hip ROM was −2° extension to 115° flexion. Follow-up at a mean of 14.4 months (5.6–45.2) showed return to baseline ROM in knee of 2°–119° and hip from −1° extension to 91° flexion. The overall rate of obstacles and complications was 37.1 % and 32.4 %, respectively [58].

Table 22.2 Limb lengthening results and complications in 95 patients

		Group 1 (<6 years)	Group 2 (6–13 years)	Group 3 (>13 years)
Number of patients (*n*)		30	40	25
Amount of length achieved				
	Length (cm)	5.4 (2.8–8.5)	6.2 (2.5–11)	6.3 (2.2–12.5)
	Relative (%)	39 (12–71)	24 (7–54)	20 (5–58)
	Lengthening index (month/cm)	1.0 (0.5–1.8)	1.1 (0.5–2.2)	1.1 (0.5–2.2)
Good/excellent outcome score		28/30 (93 %)	37/40 (93 %)	24/25 (96 %)
Problems, obstacles, and complications				
	Femur fracture	13	14	4
	Pin-site problems requiring surgery	4	3	1
	Nerve irritation/palsy	3	3	1
	Premature consolidation	0	3	3
	Delayed union	2	4	3
	Knee subluxation	5	5	3
	Hip subluxation	3	3	1
	Total unplanned surgery	21 (70 %)	22 (55 %)	9 (36 %)

Treatment of CFD has always been divided between amputation and lengthening. In the past, the indications for amputation were much more liberal. Currently, the dividing line can be based more on pathoanatomy. Treatment varies, depending on the severity of the underlying deficiency. The less involved patients, such as Paley type 1a, usually only require leg length equalization surgery, consisting of limb lengthening and/or contralateral epiphysiodesis at the appropriate time. The current treatment of more severely affected patients, such as Paley type 1b and type 2, consists of complex surgical interventions consisting of reconstruction of the hip and knee joints, correction of femoral deformity and length, ossification of the pseudoarthrosis, and reconstruction of lax or absent ligaments. This is discussed in the subsequent sections. The most severely affected patients, Paley type 3, usually undergo surgery to improve prosthetic use, as regaining satisfactory function with limb lengthening and reconstruction is often not feasible.

Box 22.5. Classification and Treatment

- Multiple classification schemes exist, mostly based on their anatomic deformity, though sometimes they are age dependent or have arbitrary cutoffs.
- The Paley classification separates into types and subtypes based on their surgical treatment for reconstruction.
- Van Nes rotationplasty has been used historically on severely shortened femurs with good results.
- Patients undergoing Syme amputation with knee arthrodesis often have more altered gait mechanics with greater energy expenditure compared to those undergoing a rotationplasty.
- In the properly selected patient, limb lengthening often has good outcomes, but can have significant complications. However, these untoward events can be lessened with prophylactic joint stabilization and intramedullary rodding techniques.

Recommended Surgical Reconstructive Strategy for Paley Type 1 CFD

Outlining a "Life Plan" for the Family

At the initial consultation, a surgical reconstructive strategy or "life plan" projected to skeletal maturity should be outlined for the child and the family. It is helpful to write this out on the back of a printed radiograph for the family to put in the child's scrapbook and to refer to in the ensuing years. The strategy is based on the type of CFD, the projected LLD at maturity, and the reconstructive potential of the hip and knee. In cases with combined CFD and FH, the strategy for FH must be combined with that of the CFD [18].

Step 1: Preparatory Surgery for the Hip and Knee

Prior to lengthening one must determine whether the hip and knee joints are stable and/or deformed and whether surgical procedures for these joints are required before initiating limb lengthening. At the hip, if the acetabulum has an acetabular index with comparable slope to the normal contralateral side, a center edge angle (CEA) $\geq 20°$, and a neck-shaft angle (NSA) $\geq 110°$, no separate hip surgery is required before the first lengthening [59]. If the acetabulum shows signs of dysplasia then a pelvic osteotomy should be performed prior to lengthening. Increased slope of the sourcil (acetabular roof) or acetabular index compared to the other side is a subtle but sensitive sign of acetabular dysplasia. Coxa vara should be corrected prior to lengthening if the NSA is less than 110°. Similarly, external rotation deformity of the hip is a factor to consider for correction at the same time as the acetabular dysplasia. If a Dega-type osteotomy is chosen, then there is usually a gain of about 1 cm in leg length. Associated hip deformities of retroversion, hip flexion contracture, and hip abduction contracture should be simultaneously addressed. The flexion contracture of the hip is treated by recession of the psoas tendon and release of the rectus femoris tendon. A bony flexion deformity may be treated by a proximal femoral extension osteotomy. The abduction contracture is treated by lengthening or resection of the fascia lata and, if necessary, an abductor muscle slide at the iliac crest. When all of these deformities are present together and especially with higher degrees of angulation, the reconstructive procedure is called the Superhip procedure [60].

Other factors should be examined prior to lengthening. The proximal femur should be normally ossified for the patient's age. Often, severe cases of CFD have delayed ossification of the femoral neck or subtrochanteric region, and bone morphogenic protein (BMP) may be added to the femoral neck to promote ossification [14]. The fascia lata is a thin but very tough limiting membrane that resists lengthening and applies pressure across the knee joint and the distal femoral growth plate. Thus, it should always be removed or released before lengthening. It can also be used to reconstruct absent knee cruciate ligaments if procedures are done concurrently. A hemiephysiodesis may also be performed to correct frontal plane deformities. The ideal age for the preparatory procedure is between ages 2 and 3 years.

Step 2: Serial Lengthenings of the Femur and/or Tibia

A prediction of total leg length discrepancy at skeletal maturity helps determine the approximate number of lengthening surgeries required. This can also be done using the Paley

multiplier method [26]. The majority of CFD cases will require at least two lengthenings. The goal of each lengthening depends on the total discrepancy at maturity. The safe range of lengthening is from 5 to 8 cm, as long as a good physical therapy program is available. This amount seems to be independent of the initial length of the femur and age of the child, and can be performed safely even in a toddler. In most cases, the maximum of 8 cm is attempted because of a large total discrepancy, as long as the patient maintains adequate knee range of motion.

The first lengthening of the femur can be performed 12 months after the preparatory surgeries, assuming that the femoral neck has ossified. If the preparatory surgery is performed between the ages of 2 and 3 years, then the first lengthening can follow between the ages of 3 and 4 years. However, if the femur is excessively short (total femoral length <75 mm) for an external fixator or if the femoral neck fails to ossify, then it would be beneficial to wait for a year or two for more growth. By beginning lengthening at a young age, the level of prosthetic or orthotic need is reduced earlier in a child's life. In the senior author's experience, the complication rate for limb lengthening is no different for the younger age group [61]. Lengthening of the femur in children younger than 6 years may be associated with sustained growth stimulation [62].

Between 4.5 and 7 years, children may have more difficulty psychologically dealing with limb lengthening. This is related to the normal cognitive stages of children at this age. Children at this age group seem to understand too little and too much at the same time. They are beginning to be more independent and may appear to be mature enough to handle the process. They understand that they have one short leg. They do not connect the recognition of a short leg with the solution of limb lengthening. Their cognitive level is insufficient to understand why their parents allowed someone to do this to them. The younger children do much better because their cognitive level accepts everything their parents decide without questioning. Beyond the age of 6 or 7 years, the child enters the age of reason and begins to understand that there is a problem with a solution. Their cooperation is voluntary rather than coerced. One way to explain this to children is to focus on the size of their shoe lift. It is easier for them to understand that they can wear a normal shoe after the operation instead of understanding that their leg lengths will be equal.

The frequency of lengthening should be spread out to no less than every 3 years and preferably 4 years or greater. The rule of 4 is a good guiding strategy: one lengthening every 4 years starting by 4 years. Assuming that a preparatory surgery is done between ages 2 and 3 years, the first lengthening can be done between ages 3 and 4 years. The second lengthening would occur around age 8 years and a third lengthening around age 12 years. For psychosocial reasons, it is preferable to complete all the lengthenings by age 14 when the child starts high school education. If a fourth lengthening is required it can be done around age 16 years. Patients may also have lengthenings performed later in life as an adult. One severe case treated by the author equalized a 25-cm discrepancy with two lengthenings over a nail. If the tibia is also short, a combined femoral and tibial lengthening can allow for greater total lengthening amount in a shorter time frame. However, a combined lengthening may not be able to achieve as much total length as two separate lengthenings, and growth inhibition in the tibia has been reported [62]. In addition, with the advent of smaller and more reliable implantable nails, in the author's experience, internal lengthening can be started as early as 7 years of age.

Step 3: Hemiepiphysiodesis and Epiphysiodesis

Contralateral epiphysiodesis around the knee is used as an adjuvant method to equalize LLD. It should be calculated into the total strategy of equalization surgeries, and can be used for up to 5 cm of LLD equalization. Judicious use in some cases may avoid the need for one lengthening. For example, a predicted discrepancy of 12 cm may instead be treated with only one 7-cm lengthening and a 5-cm epiphysiodesis around puberty. Calculation of the timing of epiphysiodesis can be achieved quickly and accurately using the multiplier method [26].

Ipsilateral hemiepiphysiodesis is very useful to correct the valgus deformity of the knee from distal femoral or proximal tibial origins. The 8-plate, developed by Peter Stevens, or a similar device, is a simple way to temporarily arrest the one side of physeal growth [63, 64]. Correction of the valgus deformity of the femur permits future implantable lengthening of the femur since there is no angular deformity.

Box 22.6. Planning Surgery for the CFD Patient

- Determine the classification and total discrepancy of the deficiency to plan out a timeline of surgeries.
- Have a low threshold to perform preparatory surgeries of the hip and knee to prevent subluxation or dislocation, which can be disastrous.
- Do not lengthen until the femoral neck has ossified. BMP can be used to promote ossification.
- The fascia lata must be released or resected before lengthening.
- The safe range of each lengthening is 5–8 cm.
- The Rule of 4: First lengthening at 4 years of age and then every 4 years thereafter as needed.

  Contralateral epiphysiodesis can be used to avoid an extra lengthening.

The Superhip Procedure

Is the proximal femur bony deformity (see Fig. 22.1) caused by the hip joint contractures, or vice versa? The net effect in the frontal plane is that the insertion of the abductor muscles (gluteus medius and minimus) on the greater trochanter is abnormally close to the pelvis. This leads to several problems, including impingement of the trochanter on the iliac bone and contracture of the gluteal muscles since the distance between their origin and insertion is short. The gluteus maximus and fascia lata, with its iliotibial band extension to the tibia, are the most lateral of the soft-tissue structures and therefore contribute the greatest to the abduction contracture of the hip. They also contribute significantly to flexion contracture of the hip. The abduction contracture is not obvious, since hip adduction is preserved due to the femoral bony varus deformity, and hip abduction is limited by iliotrochanteric impingement. If the bony coxa vara is corrected by osteotomy, without soft tissue releases, the abduction contracture will be uncovered and will prevent the hip from coming back to a neutral position relative to the pelvis, producing a fixed pelvic tilt. An abduction pelvic tilt on the short leg makes the LLD appear less than before surgery. In the face of an open growth plate or a non-ossified neck or subtrochanteric segment, as in type 1b cases, the abduction contracture leads to recurrence of the coxa vara after osteotomy. The mechanism for this recurrence may be differential growth of the physis, bending at the non-ossified tissues, or slipped capital femoral epiphysis.

To correct these deformities, a three-dimensional osteotomy combined with a series of selective soft tissue releases was developed by Paley in 1997. It is now known as the *SUPER hip procedure* because of its complexity.

Box 22.7. The Evolution of the Superhip Procedure
Dror Paley, MD

The Superhip name originally arose as a billing code to avoid writing down the multiple CPT codes that comprised this multistep conglomerate procedure. While there was no intention to call this operation the Superhip procedure, the name stuck. To avoid the name being misperceived the SUPER prefix was then made into an acronym: Systematic Utilitarian Procedure for Extremity Reconstruction. Other SUPER joint reconstructive procedures for congenital knee and ankle deformities developed by Paley were subsequently renamed the SUPER knee and SUPER ankle procedures. John Birch of Dallas has expressed an aversion to the SUPER prefix and refers to these

(continued)

Box 22.7. (continued)
procedures as Paley hip, knee, and ankle procedures. As I learned from my mentor Dr. Robert Salter from his "no name no fame" comment regarding the innominate osteotomy now called the Salter osteotomy, it is not for a surgeon to name this procedure after themselves. I have given this procedure a generic name. If others choose to name it after me, then so be it.

My understanding of CFD began with my experience at the Hospital for Sick Children, Toronto, in 1984. I worked with Drs. Robert Gillespie and Ivan Krajbich, who treated the severe CFD cases by rotationplasty [49]. In 1986 and 1987, I witnessed reconstructive options for these deformities in Kurgan, USSR. Drs. Popkov and Maltzev showed me how to do pelvic support osteotomy to bypass the tethered and deformed hip. The message of these experts was that the CFD deformed hip did not lend itself to anatomic reconstruction.

After beginning my practice in 1987 at the University of Maryland, I tried to find alternative reconstructive solutions to the CFD hip. I performed many valgus, extension, and internal rotation osteotomies on young children with this deformity. Initially, they had excellent bony correction with reduction of the leg length difference due to a fixed pelvic obliquity that developed after surgery. Gradually, I watched the deformity recur, presumably through their proximal femoral physis or unossified femoral neck. I realized that the reason for this recurrence was the hip flexion and abduction contractures that existed prior to the surgery. In 1996, I recognized that in order to succeed, I needed to untether the proximal femur from the hip abductors and flexors and then realign the proximal femur to a neutral position at the hip joint. This was a radical concept, and I was left with the challenge of how to safely release the hip abductors. Borrowing from the concept of the Hardinge approach to the hip, which took down the anterior third of the hip abductors as a sleeve with the quadriceps muscle, I decided to take down the entire hip abductors with the vastus lateralis as one sleeve. In 1997, I performed the first such procedure.

While the psoas, rectus, and TFL were all lengthened as they are today, treating the hip abductors by releasing the gluteus and vastus tendons off of the greater trochanter together turned out to be a bad idea. While this freed up the proximal femur to rotate into neutral by extension, adduction, and internal rotation of the hip joint, it also changed the muscle-tendon length ratio, thus permanently weakening the hip abductors.

(continued)

Box 22.7. (continued)
This is discussed in more detail below. I initially fixed the femur using a Rush rod and a tension band wire. There was no fixation up the femoral neck. When this procedure was performed for a subtrochanteric type Paley 1b or for a Paley 1a, no recurrence of the deformity occurred since the delayed ossification part was resected. When it was performed for a neck type Paley 1b, the varus usually recurred and the neck did not ossify.

In 2001, I switched to using a fixed-angle plate. I used the 130° sliding hip screw (Smith and Nephew, Memphis, TN). Some of the cases ossified, but most did not. Some of the plates broke and the deformity recurred, while in others the plate began to cut through the head as the varus recurred. I decided that a blade plate would be the best implant to avoid cutout as well as to control flexion and extension forces. With the engineers at Smith and Nephew, I designed an infant and pediatric 130°-angle cannulated blade plate, to correspond to the normal neck-shaft angle. Since 2004, I have been using this new plate. This minimized recurrent deformities, but incomplete ossification of the femoral neck was the usual result.

It was apparent that while we could correct this complex deformity, we could not get the delayed ossification of the cartilage to ossify. To get some of these failed cases to ossify, I resorted to insertion of BMP up the femoral neck into the non-ossified cartilage. The result was dramatic: the recalcitrant-delayed ossification cases ossified. Since BMP-2 (Infuse, Medtronic, Memphis, TN) was not FDA approved to use in children, we were initially hesitant and reserved in its application. We only used it to salvage previously failed Superhips. With the unexpected success that we saw from such application, I decided to apply it to new cases in 2006. The results were equally remarkable. All of the necks ossified and there were almost no recurrent deformities. Clearly, we needed to combine a mechanical with a biologic solution to solve the CFD deformity puzzle.

The soft tissue part of the operation also went through an evolution to its present state. In the first 5 years, we not only employed release of the conjoint tendon off of the greater trochanter to treat hip abduction contracture, but we also released the hip capsule off of the greater trochanter. This was done in an extra-articular fashion so that the joint was never exposed. No initial consequence of this was observed. However, when we started to lengthen femurs that had previously had a Superhip procedure, we encountered two new complications: hip dislocation and slipped capital

(continued)

Box 22.7. (continued)
femoral epiphysis (SCFE). In retrospect, we realize that both were related to the release of the superior capsular ligament from the pelvis to the greater trochanter. Our selective release of the superior capsular ligament demonstrated the importance of this band. This superior capsular pelvic-trochanteric band is essential to prevent the femoral head from moving laterally relative to the acetabulum. After recognizing this, we stopped releasing this band and we no longer experienced dislocations or slips following a Superhip.

After 10 years of performing the Superhip, we had solved the recurrent varus deformity problem, the delayed ossification problem, and the dislocation/SCFE problem. We noticed, however, that the children walked with a marked lurch or Trendelenburg gait. We attributed this to the abductor tendon release. By detaching the hip abductors from the greater trochanter, we had changed the muscle-tendon length ratio of the gluteus medius and minimus muscles, which was difficult to recover. Essentially, we had added tendon length to the glutei due to the quadriceps tendon moving proximally. To solve this problem in 2008, I began to perform the abductor muscle slide off of the ilium instead of the distal tendon lengthening. This preserved the muscle-to-tendon length ratio, keeping the muscle tendon length and tension constant after surgery. The slide is achieved by shortening the height of the iliac crest. As an added benefit, the glutei are a peculiar group of muscles since they have a growth plate connected to them that grows in a direction that lengthens the muscles. After the repair of the abductor slide, the iliac apophysis can increase the tension and re-elevate the origin of this muscle. Since adding the abductor slide, patients do not have the previously noted lurch or Trendelenburg gait.

Superhip Surgical Technique (Figs. 22.7, 22.8, 22.9, 22.10, and 22.11)

1. *Positioning, prepping, and draping*. An epidural is placed by the anesthesia service with a catheter running up the back on the non-operative side. A Foley catheter is placed and also routed to the non-operative side. The patient should be moved to the edge and foot of the radiolucent table in a supine position. The ipsilateral arm should be appropriately padded and placed across the patient's chest. A radiolucent bump (usually a folded towel or sheet) is placed beneath the ipsilateral ischium to roll the pelvis 45° towards the opposite side.

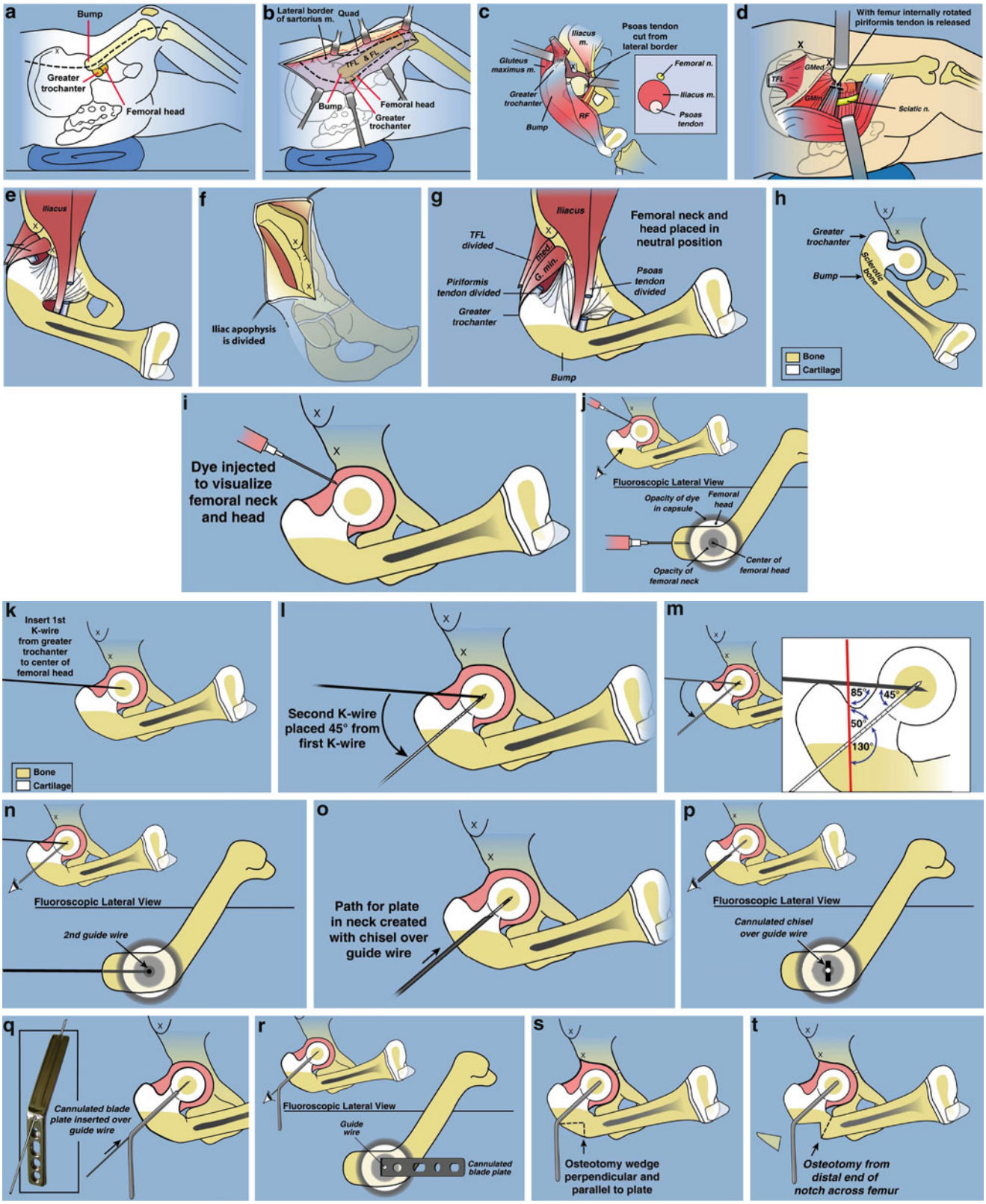


Fig. 22.7 Superhip surgical technique illustrations. (**a**) Straight mid-lateral incision from top of iliac crest to below tuberosity of knee. (**b**) Reflect fascia lata-iliotibial band distally and leave attached at Gerdy's tubercle. (**c**) Release rectus femoris tendon, decompress femoral nerve, and recess iliopsoas tendon. The TFL muscle remains attached to the ASIS. (**d**) Decompress sciatic nerve and release piriformis tendon at greater trochanter. (**e**) Adduction is limited by hip abductor muscles. (**f**) Split apophysis and do abductor muscle slide. (**g**) All extra-articular tethers released. Hip can be placed in neutral position. (**h**) The quadriceps muscle is elevated to expose the lateral and anterior femur. (**i**) Hip arthrogram to visualize femoral head. (**j**) Lateral view shows concentric rings of ossific nucleus, femoral neck, and femoral head, which should be positioned in a bull's-eye pattern. (**k**) Insert first guide wire from tip of greater trochanter to center of femoral head. (**l**) Insert second guide wire at 45° to first. (**m**) Geometric rationale behind 45° angle. Neck-shaft angle is 130° and medial proximal femoral angle is 85°. Therefore the angle between these must be 45°. (**n**) The second guide wire is centered in the femoral head and neck on both views. (**o**) Insert a cannulated chisel up the femoral neck. (**p**) The chisel should be perpendicular to the posterior aspect of the greater trochanter.

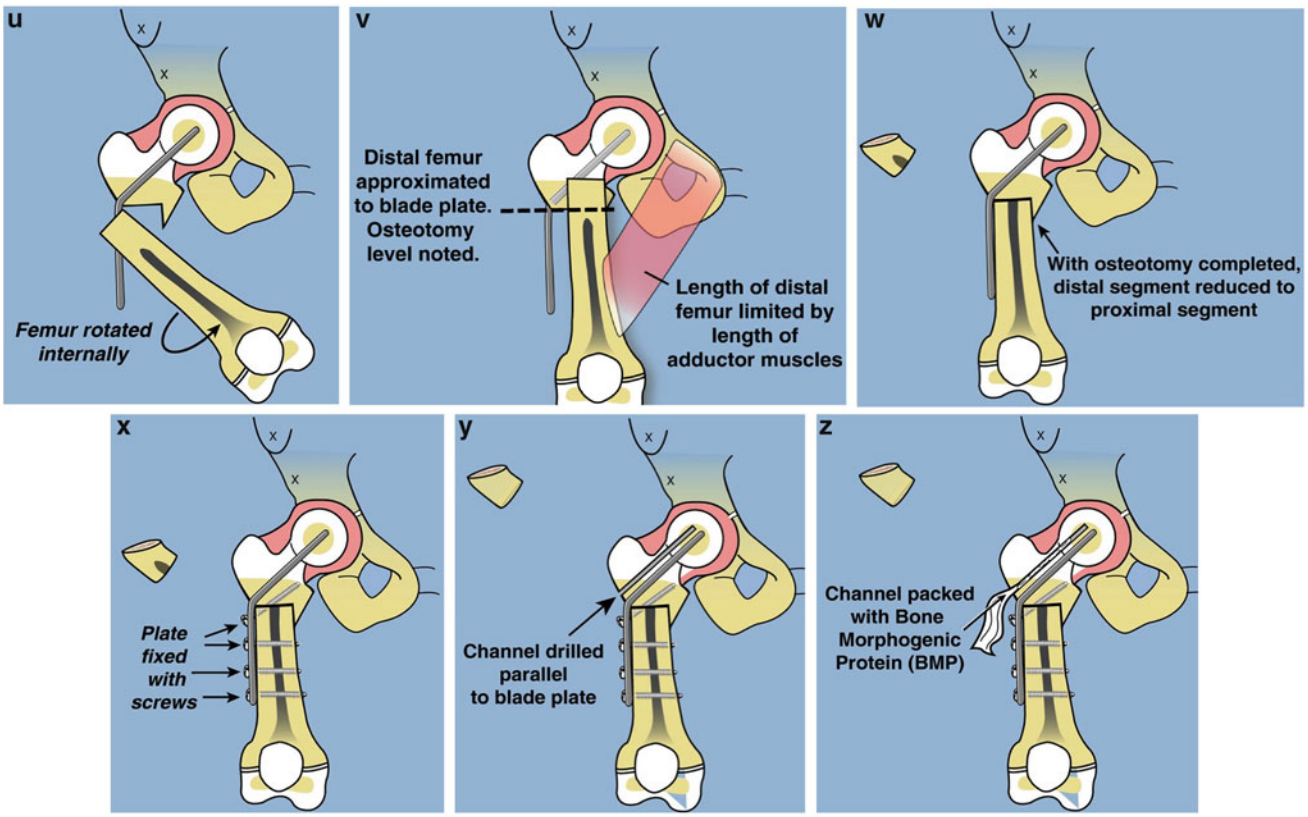


Fig. 22.7 (continued) (**q**) Replace the chisel with a 130° blade plate. (**r**) The blade plate is in place. Note the flexion deformity of the femur shaft relative to the plate. (**s**) Cut perpendicular and parallel to the plate. (**t**) Perform a second osteotomy oblique to the femur. More recently this osteotomy starts inside the notch that was removed eliminating the medial lip. (**u**) Rotate the distal femur internally and abduct it to realign it with the proximal femur. (**v**) Overlap the bone ends and mark the level of overlap. The femur cannot be reduced due to the tethering medial and posterior soft tissues. (**w**) Perform a shortening osteotomy of the femur and reduce the femur to the plate. (**x**) Fix it with three diaphyseal screws and one interfragmentary neck screw. (**y**) Drill a channel parallel to the blade. (**z**) Insert BMP-2 collagen sponges up the femoral neck

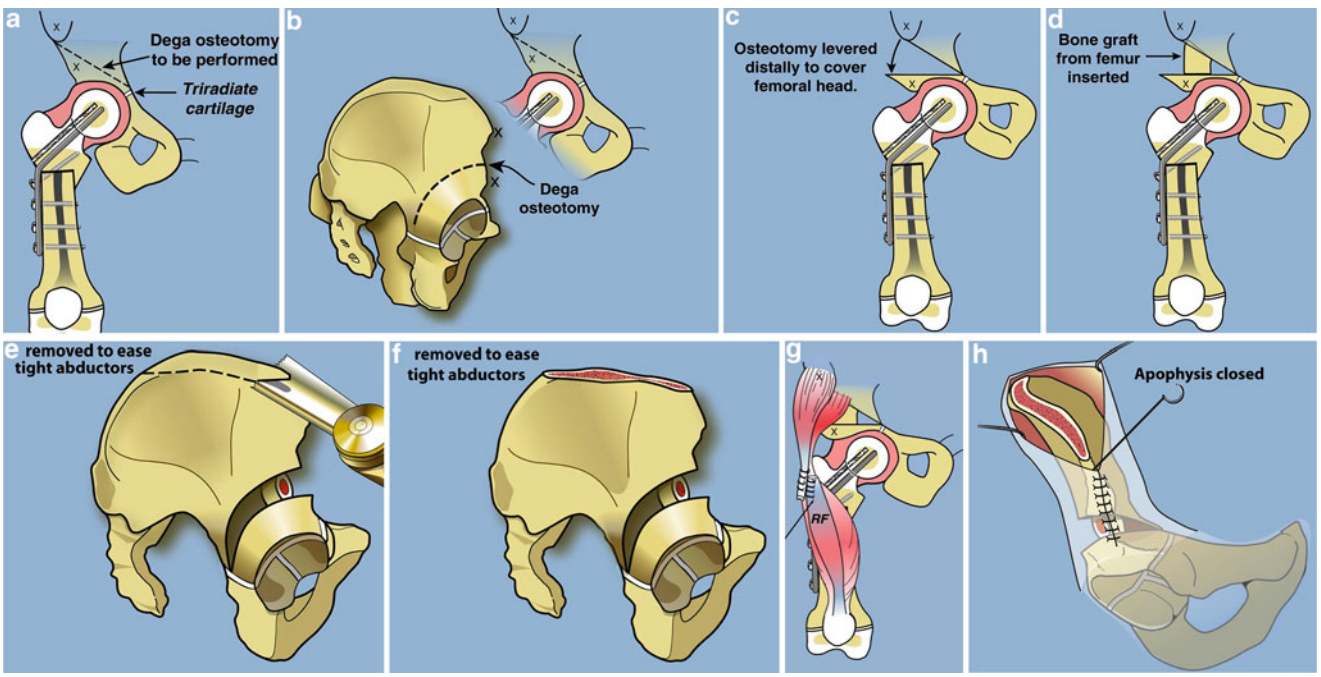


Fig. 22.8 Paley modification of Dega osteotomy. (**a**) The cut should start 2–3 cm proximal to the lateral acetabulum converging on the triradiate cartilage medially. (**b**) The cut should be parallel to the acetabulum circumferentially stopping at the triradiate between the ilium and the ischium. (**c**) The roof of the acetabulum is levered down to cover the femoral head such that the sourcil is horizontal. (**d**) Fashion the bone resected from the femur to fit into the opening wedge space. (**e**) Resect the top of the iliac wing to reduce the tension on the hip abductors so that the apophysis can be sutured closed. This is part of the abductor slide. (**f**) After the resection. (**g**) Transfer the rectus femoris to the TFL. (**h**) Close the apophysis

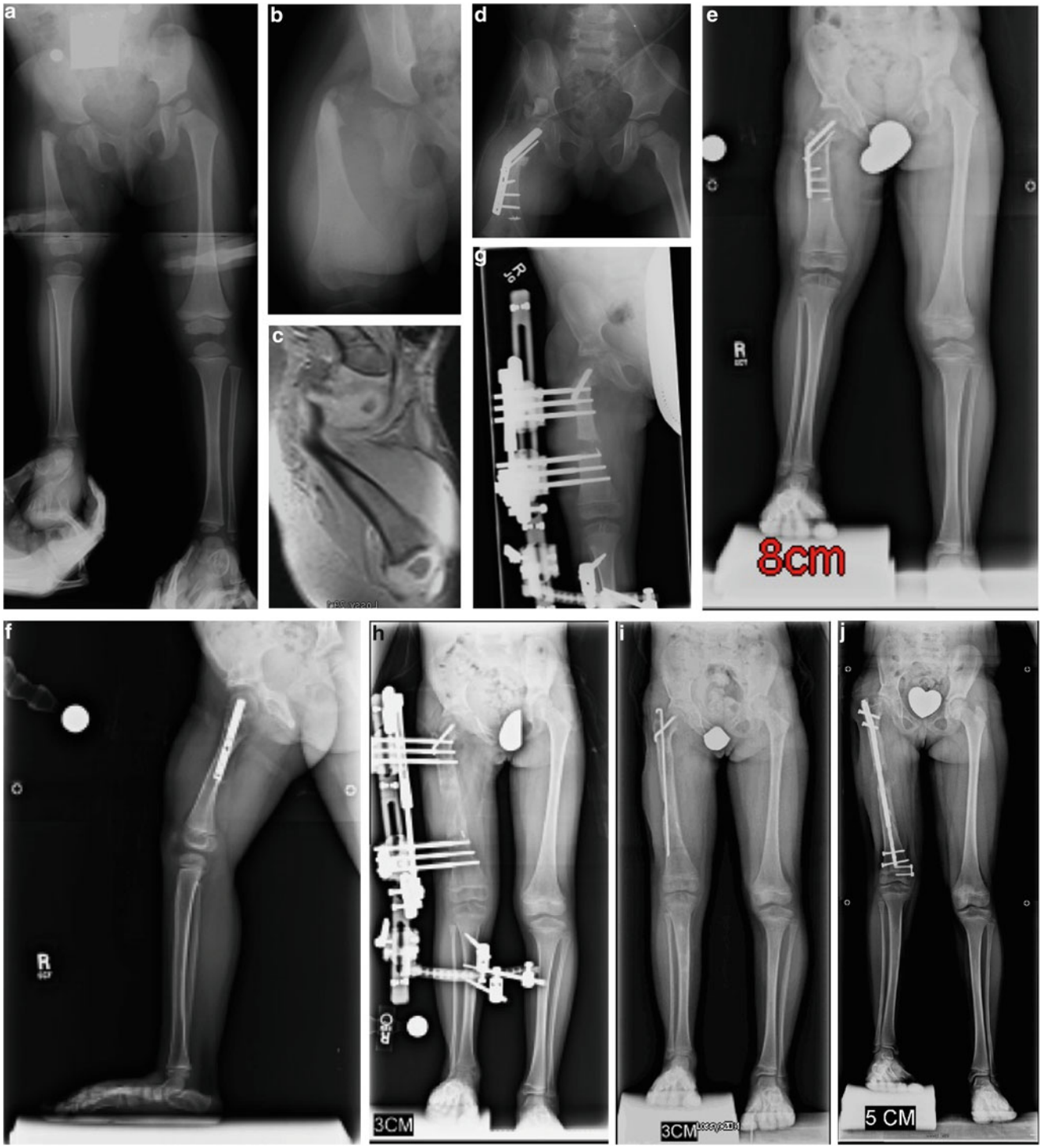


Fig. 22.9 CFD Paley type 1b with delayed ossification of femoral neck (**a**, **b**). MRI showing the cartilaginous femoral neck that remains unossified at 2 years of age (**c**). Superhip with Superknee procedure at 2 years of age, including insertion of BMP in femoral neck (**d**). The neck is fully ossified by 3 years of age. Note the significant growth that has occurred since the year before (**e**, **f**). First lengthening performed at age 4 years with Smith & Nephew Modular Rail System (Memphis, TN) external fixator with articulation across the knee joint (**g**). Eight centimeters of lengthening achieved (**h**). Removal of external fixator with Rush rodding of bone to prevent fracture (**i**). At the age of 8 years she underwent a second lengthening, this time using the PRECICE (Ellipse Technology, Irvine CA) 8.5-mm-diameter lengthening nail (**j**)



Fig. 22.10 Two-year-old girl with CFD Paley type 1b with delayed ossification and severe angulation of the subtrochanteric level of the left femur (**a**). MRI showing the delayed ossification of subtrochanteric region. The neck is normally ossified (**b**). The deformity is fully corrected and the femur is healed after the Superhip surgery. The knee has full extension following posterior capsulotomy and Superknee stabilization. This X-ray is 1 year after the Superhip and Superknee showing significant growth and remodeling which is why the screws are protruding medially (**c**). Lengthening of the femur was performed at age 4 years (**d**). X-rays after lengthening of the femur 6 cm and insertion of Rush rod at the time of removal surgery (**e**)

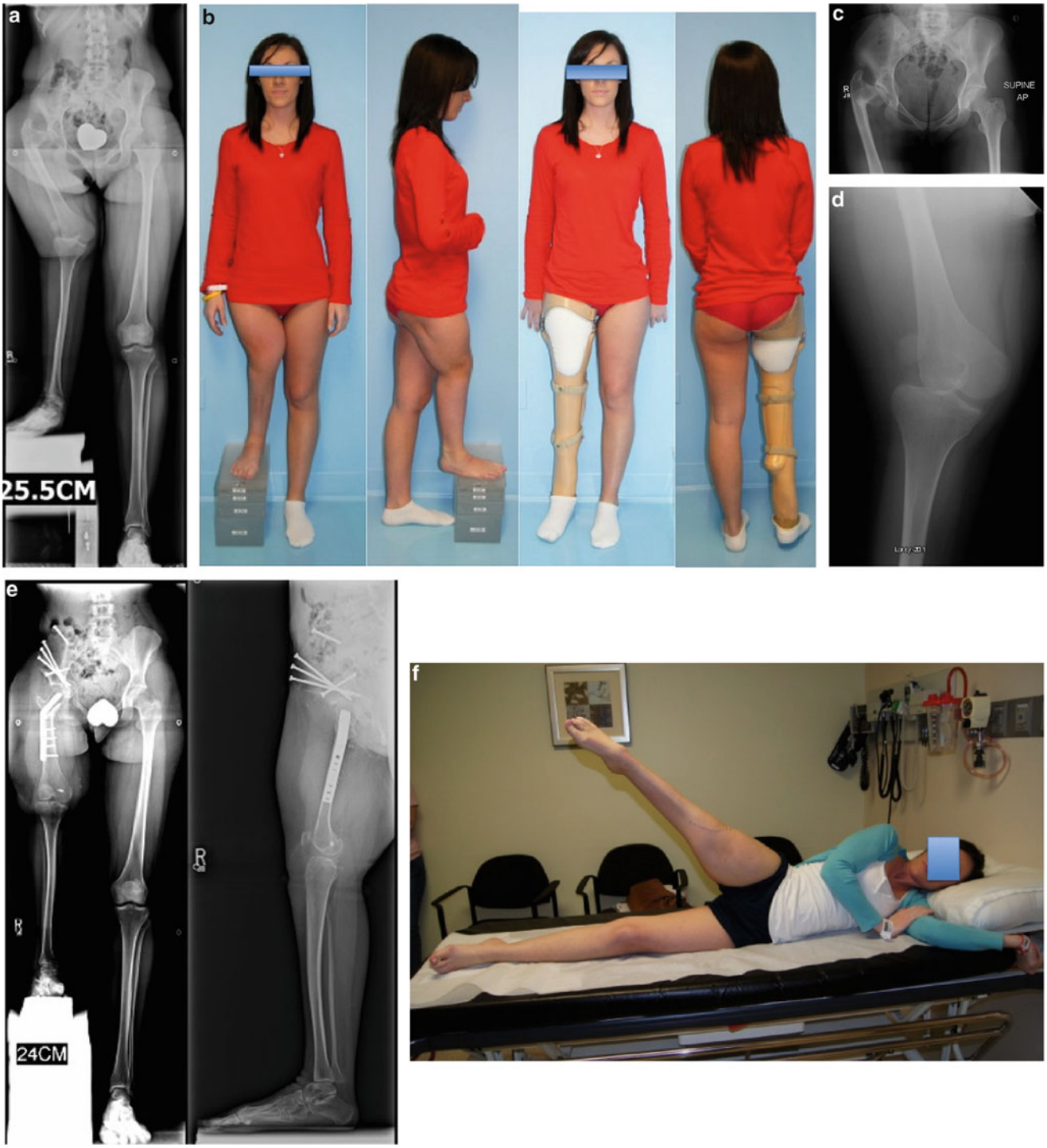


Fig. 22.11 (**a**) Erect leg radiographs of a 22-year-old woman with previously untreated CFD and 25-cm discrepancy. She has marked coxa vara and acetabular dysplasia as well as rotatory subluxation of the knee joint and dislocation of the patella. (**b**) Clinical picture before any surgery with and without her prosthesis. (**c**) AP pelvis X-ray showing the hip deformities. (**d**) AP knee X-ray showing the rotatory subluxation of the knee. (**e**) AP and lateral radiographs after Superhip procedure with Ganz periacetabular osteotomy (PAO) and Superknee procedure for reduction and stabilization of the knee and patella. (**f**) Active hip abduction after recovery. (**g**) Active hip flexion and knee extension after recovery. (**h**) Ganz PAO modified for Superhip procedure. Start by osteotomizing the iliac crest and peeling off the abductor muscles to do an abductor muscle slide. (**i**) The five bone cuts of the Ganz seen from the lateral side. (**j**) The acetabulum is internally rotated to gain posterior coverage. (**k**) The acetabulum is then abducted to gain lateral coverage. (**l**) The iliac wing is shortened, and then the crest is reattached using the same screws that are used to fix the PAO, 3 antegrade and one retrograde. (**m**) Radiograph after first lengthening of femur and tibia (6-cm femur and 5-cm tibia). The femur and tibia were both rodded using Rush rods at the time of fixator removal to prevent fracture

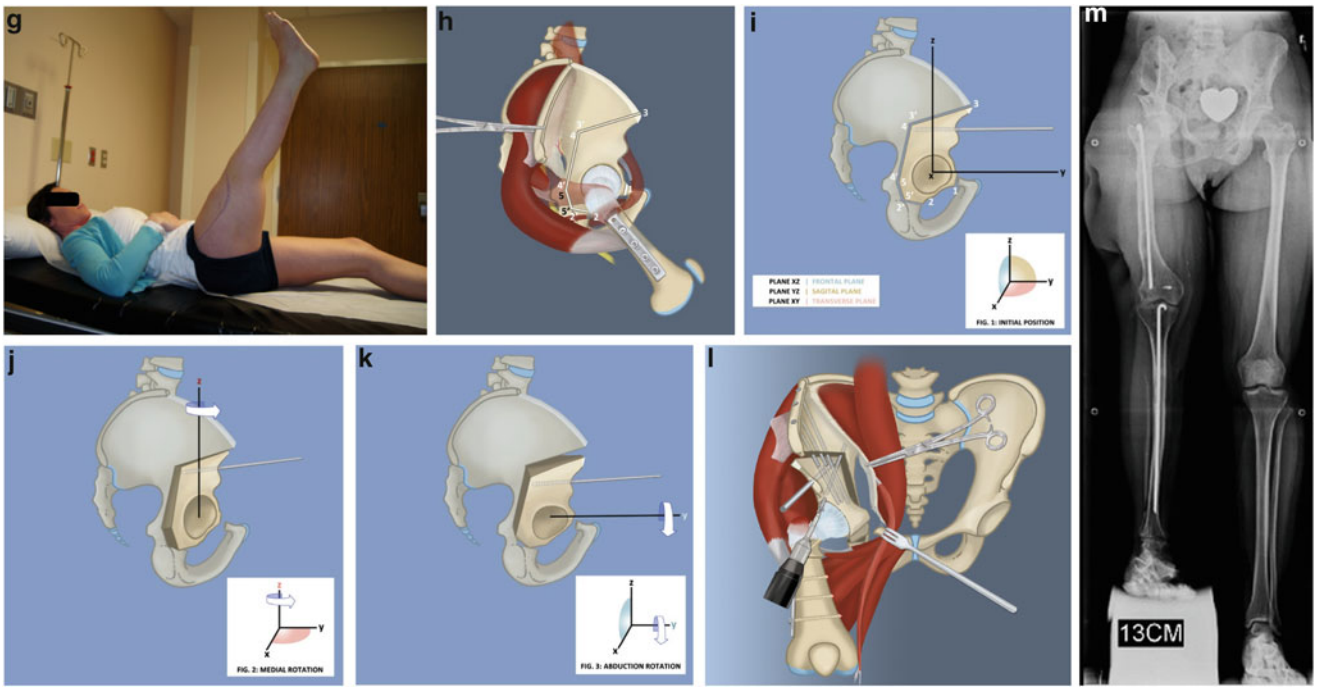


Fig. 22.11 (continued)

The bump should not be beneath the iliac crest or lower back. The entire side should be prepped and draped free from the nipple to the toes. The drapes should extend from the crack of the buttocks to the fold between the scrotum or labia and the thigh. The lower limb should be completely free of the drapes. This is called a forequarter prep.

2. *Incision.* With the leg fully extended, a long mid-lateral incision is made from the top of the iliac crest to the tibial tuberosity. The incision is kept as straight as possible, passing over the proximal femoral "bump" and continuing longitudinally towards Gerdy's tubercle before curving slightly anteriorly towards the tibial tubercle. The incision is carried down to the depth of the underlying fascia lata and iliotibial band.
3. *Flap elevation.* The subcutaneous tissues and skin are elevated as one large flap anteriorly and posteriorly off the fascia of the thigh and pelvic region. The fat is adherent to the fascia and should be dissected preferably with an electrocautery. The electrocautery should be held flat, parallel to the plane of dissection, and can be quite technically difficult. It is important not to incise or damage the fascia if it is being used for knee ligament reconstruction. Dissection may also be carried out with scissors. Anteriorly, the flap is extended medial to the Smith-Peterson interval (tensor fascia lata (TFL) and sartorius) proximally. Posteriorly, the subcutaneous flap is elevated to just posterior to the intermuscular septum. Distally, reflect the flap to the patella if no ligament reconstruction is to be done and all the way to the medial side if ligament reconstruction is to be done. The fascia lata is now fully exposed from the patella to a couple of centimeters posterior to the intermuscular septum distally and from the anterior edge of the TFL to the midgluteus maximus proximally.
4. *Fascia lata release.* The fascia is incised at the TFL-sartorius interval making sure to stay on the TFL side in order to avoid injury to the lateral femoral cutaneous nerve. The fascial incision is extended distally to the lateral border of the patella ending at the tibia. The posterior incision of the fascia lata starts distally and posterior at the intermuscular septum and extends proximally to overlie the gluteus maximus in line with the incision. The gluteus maximus (GMax) should be separated from the overlying fascia anterior to the posterior fascial incision. The fascia should be retracted anteriorly and away from the underlying muscle, while the GMax should be dissected off of the fascia and the intermuscular septum that separates it from the TFL. The GMax should not be split in line with the fascial incision to avoid denervating the muscle anterior to the split. It can now be reflected posteriorly to allow exposure of the greater trochanter, piriformis muscle, and sciatic nerve.

   If knee ligamentous reconstruction is planned, the fascia lata is cut proximally and anteriorly at the musculotendinous junction. The fascial cut should be a step cut

or sloped posteriorly and proximally to include a longer fascia segment posteriorly from the fascia that was dissected off of the GMax. The fascia lata is reflected distally to Gerdy's tubercle. The TFL can be left in place without further dissection. It does not have to be separated from the underlying gluteus medius (GMed). The two muscles are often adherent to each other, and it may be difficult to differentiate the muscle fibers. The distinguishing feature is that the GMed fibers insert on the greater trochanter while the TFL does not. The distal fascia lata becomes the iliotibial band and blends with the underlying lateral knee capsule, which may be partially reflected with the iliotibial band. The fascia should be mobilized all the way until Gerdy's tubercle. The fascia can then be divided into two halves using a straight pair of scissors. It should be kept moist while the rest of the surgery proceeds. The two limbs of the fascia are ready for later use in the Superknee procedure.

5. *Hip flexion contracture releases.* The dissection is carried beneath the sartorius to find the rectus femoris tendon. The rectus femoris tendon insertion is identified at the anterior inferior iliac spine. The constant ascending branch of the lateral femoral circumflex artery and vein is cauterized prior to cutting the tendon. The conjoint rectus femoris tendon (distal to the split into reflected and direct heads) is cut and allowed to retract distally. Care should be taken not to go too distal on the rectus femoris to avoid injury to its innervating branch of the femoral nerve. Just medial to the rectus is the iliopsoas muscle. The iliocapsularis muscle (capsular origin head of iliopsoas muscles) can also be seen here. The femoral nerve lies on the anteromedial surface of the iliopsoas muscle. Before looking for the psoas tendon, the femoral nerve should be identified and decompressed below the inguinal ligament. The posterior aspect of the iliopsoas muscle belly is now elevated from lateral to medial. The psoas tendon is located on the posteromedial surface in the substance of the muscle. The tendon is exposed and cut. Any remaining flexion contracture of the hip is due to the gluteus medius and minimus (the part of these muscles originating anterior to the center of rotation of the femoral head in the sagittal plane), and the anterior fascia of thigh. The release of the gluteus medius and minimus muscles is accomplished by the abductor muscle slide technique (see step 7). If the anterior thigh or sartorius fascia is still tight, it can also be released, taking care not to injure the lateral femoral cutaneous nerve, which should be identified and decompressed. It runs inside the fascia covering the sartorius muscle just medial to the anterior superior iliac spine.
6. *External rotation contracture release*. The piriformis tendon is contracted and prevents internal rotation of the hip. It should be released off of the greater trochanter. The greater trochanter should be identified by palpation. The gluteus medius muscle posterior border is very distinct and proceeds down to the greater trochanter where it inserts. Deep to the medius is the gluteus minimus, and just distal to the minimus is the piriformis muscle. Its tendon can be palpated through its muscle. It may be difficult to identify the piriformis from the minimus. Care should be taken to avoid dissection at the distal border of the piriformis tendon. This is where the medial femoral circumflex branch anastomoses with the inferior gluteal artery branch. The entire piriformis is transected about 1 cm from its insertion onto the trochanter. The sciatic nerve can be identified and if necessary decompressed. It is more posterior to the trochanter and runs deep to the piriformis.
7. *Abductor muscle slide.* The abductors may not appear to be tight on first inspection because of the coxa vara. Adduction into a true AP of the hip, with the neck oriented normally in the acetabulum, is now restricted by the gluteus medius and minimus since the fascia lata has already been cut. Furthermore, the Dega osteotomy, which lengthens the height of the ilium, makes the abductors even tighter. As noted previously, the abductors should be detached at their origin and not their insertion. This avoids changing the muscle-tendon length ratio and avoids weakening the hip abductors, avoiding a lurch or Trendelenburg gait.

The subcutaneous tissue flaps should be elevated to provide adequate exposure to the iliac crest apophysis. There is a tendency to have inadequate posterior exposure, and the flaps should be elevated just beyond the highest lateral point of the apophysis. The anterior extent is the anterior superior and inferior iliac spines, which has been exposed for the release of the rectus femoris tendon. The abdominal external oblique muscles are partially released off of the entire length of the apophysis. Once the apophysis is bare, it can be split from the ASIS to the AIIS, and then split from anterior to posterior along the iliac crest. This should be done with a #15 blade. To know where to split, pinch the apophysis between thumb and index finger of the hand not holding the knife. Stay in the middle of the apophysis along its entire length, pushing down hard with the knife blade until one feels bone. Using a periosteal elevator, pop off the apophysis from the ilium. This should be done at multiple sites to get the entire apophysis to peel back as a unit from the ilium. The apophysis and lateral periosteum are reflected distally, thus relaxing the abductor muscles. The medial half of the apophysis is reflected medially with the iliacus muscle. Since some of the abductors act as flexors of the hip, the abductor slide helps eliminate any remaining flexion deformity of the hip. Furthermore, the iliacus muscle slide also relaxes any residual tension in this muscle.

8. *Elevation of quadriceps.* The quadriceps is now elevated off of the femur in a subperiosteal fashion. Since the femur is so short, the exposure may extend as far as the distal femoral physis. The perforator vessels need to be cauterized as the quadriceps is detached from the linea aspera. Proximally, the vastus lateralis should be elevated off of part of the cartilage of the greater trochanteric apophysis by sharp dissection.
9. *Arthrogram.* A hip arthrogram is now performed using a 20-gauge spinal needle. With the trocar inside, the needle is placed into the hip joint from the anterolateral side. Traction of the femur may facilitate placement. Once the needle appears to be in the joint on the image intensifier, the trocar is removed and normal saline is injected. If the needle is in the joint the saline should go in with little pressure, and when the syringe is removed from the needle, the saline should drip back out. These signs confirm that the needle is in the joint space. The arthrographic dye can now be injected into the hip joint to outline the femoral head, acetabulum, and femoral neck.
10. *Guide wire insertion.* Since the abduction, flexion, and rotation contractures have all been released, the femoral head and neck can now be placed in a neutral orientation to the pelvis by extending and maximally adducting the lower limb across the opposite leg. A guide wire should now be drilled up the center of the femoral neck to guide the insertion of a fixed-angle fixation device. Since the femoral neck is unossified and short, it is very difficult to drill a guide wire at the correct angle up the femoral neck. The goal is to create a 130° neck-shaft angle and a medial proximal femoral angle (MPFA) of 85°. In the normal femur, the angle between the neck-shaft line and the tip of the greater trochanter to center of femoral head line is 45°.

    The first guide wire is inserted from the tip of the greater trochanter to the center of femoral head. Since the tip of the trochanter is cartilaginous in young children, it cannot be seen radiographically and the tip of the trochanter is located by palpation using the wire tip. From this point, the wire is then drilled towards the center of the femoral head as shown in the arthrogram. The image intensifier is placed into the lateral view and the leg rotated until a bull's-eye of three concentric circles is seen. This is formed by the overlapping dye shadows: the outermost circle is the femoral head, the middle circle is the femoral neck, and the inner circle is the ossific nucleus of the femoral head. A second wire should be drilled into the center of the bull's-eye at a 45° angle to the first wire. Using a depth gauge or another wire of the same length as the second wire, measure the amount of wire inside the femoral neck by placing it alongside the second wire and measuring the difference in length between the two wires. This will be the length of the blade of the blade plate to be used. The position of the neck wire can also be confirmed by flexing the hip to 90° and looking at a frog leg lateral of the neck.
11. *Blade plate insertion.* The cannulated chisel for the blade plate should now be hammered up the femoral neck guided by the second guide wire. The chisel should be rotated until it is perpendicular to the posterior aspect of the greater trochanter. This will guide it to the correct angle in the sagittal plane. Tap the chisel out of the femur and reinsert the guide wire in its previous position. Insert the appropriate length 130° blade plate over this wire to the depth of the bend of the plate. Make sure on the image intensifier that the tip of the blade is not too deep into the femoral head. Check its position on AP and lateral planes, as well as using the approach-withdrawal technique with live fluoroscopy to ensure that the blade is not advanced too close to the articular surface of the femoral head. If the blade is suspected of being too long, then replace it with one with a shorter length. Furthermore, if plate placement is off center, there is greater risk of protrusion into the joint.
12. *Subtrochanteric osteotomy.* The femur should be cut perpendicular to the shaft of the plate, starting just below the bend in the plate. This cut should be below the level of the greater trochanter cartilage. To guide this cut, drill a wire perpendicular to the plate. Keep the plane of the saw blade perpendicular to the plate in all planes. The width of the perpendicular cut surface should be as wide as the femoral diaphysis. If the deformity angle is very large, it may be parallel to the lateral cortex. A second subtrochanteric osteotomy should be made oriented less than 90° to the first osteotomy towards the base of the femoral neck to remove the bone protruding medially. The femoral head and neck can be manually tested for any residual impingement, removing any blocks to achieving 90° of hip flexion. The anteromedial corner often needs to be excised to prevent impingement.
13. *Periosteum release.* After the second osteotomy, the distal femur is easily exposed and the surrounding periosteum is peeled off circumferentially. Medially, it is very thick and restricts correction of the varus and rotation deformity. Cut the periosteum transversely around the femur, carefully separating it from the surrounding muscle. Be careful to avoid injury to the profunda femoris and its perforators, which pass immediately under the periosteum. Cutting the periosteum allows the thigh to stretch longitudinally, reducing the amount of shortening required of the femur.
14. *Femoral shortening.* The distal femur is now mobile and can be corrected into valgus and rotated internally. The distal femur is too long to fit end to end with the proximal femoral cut. The two ends should be overlapped,

and a mark should be made at the point of overlap. The distal femur should be shortened at this level. A wire is drilled perpendicular to the femur at the level of the osteotomy and a saw is used to cut, using frequent irrigation to prevent heat necrosis. The segment of bone is kept moist on the back table for use as bone graft for the Dega osteotomy. This is usually a trapezoid-shaped piece about 1–1.5 cm long.

15. *Distal femur fixation.* The femur is now brought to the plate. The bone ends should oppose without tension. The femur is internally rotated to correct the external torsion deformity. To adjust the femur to the correct anteversion, the guide wire should be reinserted into the cannulation of the plate to show the orientation of the femoral neck. The knee should be flexed to 90° and the angle between the wire and the frontal plane of the femur as judged by the perpendicular plane to knee flexion is observed. This wire should appear at least 10° anteverted, relative to the knee. The most distal hole in the plate can now be drilled with the femur held in this rotation. The drill hole should be made at the distal edge of the hole to compress the osteotomy with screw insertion. Two more screw holes are drilled and screws are inserted. The most proximal hole in the plate is designed to drill parallel to the blade of the plate and secures the plate to the proximal femur. The wire in the cannulated hole can be used to guide the drill bit. In type 1b cases, the blade of the plate and the oblique screw goes across the proximal physis and into the femoral head. In type 1a cases with a horizontally oriented growth plate, neither the blade nor the screw should cross the growth plate of the upper femur. In type 1a cases with a vertically oriented growth plate, the blade but not the screw should cross the physis.
16. *BMP insertion.* In type 1b neck cases, BMP should be inserted into the upper femur to stimulate ossification of the cartilaginous neck of the femur. A wire is drilled proximal and parallel to the guide wire in the cannulation of the plate. A 3.8-mm hole is then drilled over this guide wire. The drill hole should extend all the way into the ossific nucleus. BMP-2 (Infuse, Wright Medical) is then prepared on collagen sponges that are manually pushed into the hole with a 3.2-mm drill and forceps. A more recent method of insertion of the BMP is to use a cannula, such as the Craig needle biopsy tool, which is 4 mm in its outer diameter. The BMP sponge is labeled by applying radiographic contrast dye onto its surface. The sponge is then inserted into the end of the cannula, and the cannula is inserted into a 4-mm predrilled hole in the femoral neck. Once the cannula is located at the desired depth, a blunt trocar is used to push the sponges out of the cannula into the femoral neck. Since the sponges are labeled by contrast dye, they can be seen radiographically after the cannula is removed. It should be emphasized that such use of BMP (Infuse implant) is an off-label use of this product.
17. *Pelvic osteotomy*. Surgical technique is covered later in this chapter. The type of pelvic osteotomy depends on the age of the patient and the degree of dysplasia. In most patients under age 6 years, the Dega osteotomy is preferred. In hips with severe dysplasia, a periacetabular triple osteotomy or a Ganz periacetabular osteotomy is used depending on the closure of the triradiate cartilage. With both periacetabular osteotomies, the iliac wing shortening osteotomy should be performed prior to fixation of the acetabular fragment. Consider doing the pelvic osteotomy prior to shortening the distal femur segment, as the amount of shortening needed may change after the pelvic osteotomy. Furthermore, the pelvic osteotomy changes the rotation of the hip, since it rotates the capsule. Thus, the amount of rotation changes if the femur fixation is done after the pelvic osteotomy.
18. *Iliac wing osteotomy.* After the pelvic osteotomy, the apophysis can be sutured back together. Due to the abductor muscle slide, the lateral apophysis cannot reach the top of the iliac crest. Part of the crest has to be resected to allow repair of the apophysis. Putting traction on the lateral apophysis, one can see and mark the level to which it reaches. A saw is used to cut and remove the proximal part of the iliac wing, effectively a shortening osteotomy. The removed bone is used as graft for the acetabular osteotomy or the subtrochanteric femoral osteotomy. The apophysis is then repaired with a running #1 Vicryl suture. The medial and lateral halves of the apophysis should be well opposed to avoid a bifid iliac wing. The external abdominal muscles should be advanced and repaired over the apophysis after the closure.
19. *Muscle repairs and transfers.* The TFL muscle is repaired to the rectus femoris tendon. This essentially elongates the rectus femoris while allowing it to maintain its origin on the pelvis. The interval between the TFL and the sartorius is closed, carefully avoiding the lateral femoral cutaneous nerve. The quadriceps is sutured to the region of the linea aspera. Finally, the gluteus maximus is advanced back to the posterior border of the TFL.
20. *Closure.* If no knee releases or reconstruction is required, the fascia lata should be resected from Gerdy's tubercle. The wound can now be closed. Since there is no fascia lata, the deepest layer is the subcutaneous fat layer, called the "underlayer." One or two medium-sized drains are placed, exiting anterosuperiorly. The second drain is laid medially if a Superknee procedure is performed. The drains are usually secured with a clear adhesive sterile dressing (e.g., Tegaderm, 3 M, Minnesota).

It is important to close the wound in a fashion that the opposite layers get sutured at the same level. The deep edges of the subcutaneous underlayer are brought together with a running #1 braided absorbable suture. The Scarpa's fascia is closed with a running 2-0 braided absorbable suture. The deep dermal layer is closed with a running 3-0 braided absorbable suture, and the skin is closed using a subcuticular 4-0 monofilament suture. Dermabond™ may be used, and sterile dressings are applied.

Final radiographs are taken: an AP pelvis and lateral knee, both including the femur. The patient is then placed into spica cast. The operative limb should be placed in full hip and knee extension with the foot and ankle left free. The opposite limb can be in a flexed, abducted, and externally rotated position, with the cast stopping short of the knee. Alternatively, a one-leg spica with a molded pelvic band can be used. The cast should be bivalved before leaving the operating room.

Box 22.8. Superhip Tips and Tricks

- Pay careful attention to soft tissue handling, making sure that the subcutaneous flaps are cleanly dissected to preserve their integrity and prevent fat necrosis.
- The lateral femoral cutaneous nerve is usually found just under the sartorius fascia and originates from deep to the inguinal ligament.
- The femoral nerve lies on the anteromedial side of iliopsoas muscle. The tendon lies posteromedial.
- Do not dissect distal or deep to the piriformis tendon to avoid interrupting the inferior gluteal anastomosis with the medial femoral circumflex artery.
- The apophysis should be split with a single continuous cut, pushing hard down to bone. Multiple passes will piecemeal the apophyseal cartilage.
- Palpate the back of the greater trochanter while placing the chisel and blade plate to make sure that they stay perpendicular to its posterior border in the sagittal plane.
- The extent of the flexion and adduction deformity should be fairly evident by the orientation of the blade plate to the uncut femur. Greater than 45° of flexion deformity and up to 90° of varus deformity are not uncommon.
- Place the guide wire in the cannulated plate to help guide the rotational correction and BMP hole placement, and determining femoral anteversion.

Postoperative Course

Parents are initially educated on transfers, cast care, and hygiene. The bivalved spica cast can be converted into a removable cast in about 1 week. The patient can then start gentle passive flexion and extension of the hip from 0 to 90°, as well as passive abduction. The spica cast is discontinued after 6 weeks, and if radiographs show adequate bony healing, then the patient is progressed to full weight bearing, gait training, strengthening, and active range of motion. The end goal is to restore the child to normal function before they proceed with limb lengthening.

Pelvic Osteotomies

Paley Modification of Dega Osteotomy (Paley-Dega Osteotomy) (See Fig. 22.8)

In the Superhip procedure, the iliac apophysis has already been split and the periosteum from the lateral and medial aspects of the ilium has already been reflected back with its overlying muscles. The lateral periosteum is reflected back to the edge of the acetabulum and to the sciatic notch. The periosteum should be dissected off the anterior wall of the sciatic notch, feeling for the soft cartilage of the triradiate cartilage where it separates the ilium from the ischium.

Using the image intensifier, a guide wire is drilled approximately 2 cm proximal to the lateral edge of the acetabulum towards the triradiate cartilage medially. On the lateral wall of the ilium, the osteotomy is a curved line running perpendicular to the triradiate cartilage, separating the ilium from the ischium. It runs parallel to the anterior wall of the sciatic notch and extends proximally to the level of the guide wire. The cut curves to make a 90-degree bend heading towards the superior border of the anterior inferior iliac spine. In the frontal plane, the osteotomy is inclined from proximal to distal, parallel to the guide wire. The osteotomy ends at the triradiate cartilage medially. It is easier to start distally and work proximally up to the guide wire. At the most posterior distal part of the osteotomy, it is important to stay very near the sciatic notch to avoid entering the hip joint inadvertently.

The osteotomy is levered distally to bring the roof of the acetabulum down. A laminar spreader is used to distract the osteotomy. The vertical limb of the osteotomy extending down to the triradiate cartilage at the ischium allows for greater bending and greater lateral coverage. It is a myth that the Dega osteotomy gives posterior coverage. It cannot do so because it cannot extend distal or posterior than the posterior limb of the triradiate. The original Dega, as well as subsequent publications of this osteotomy, goes through the inner

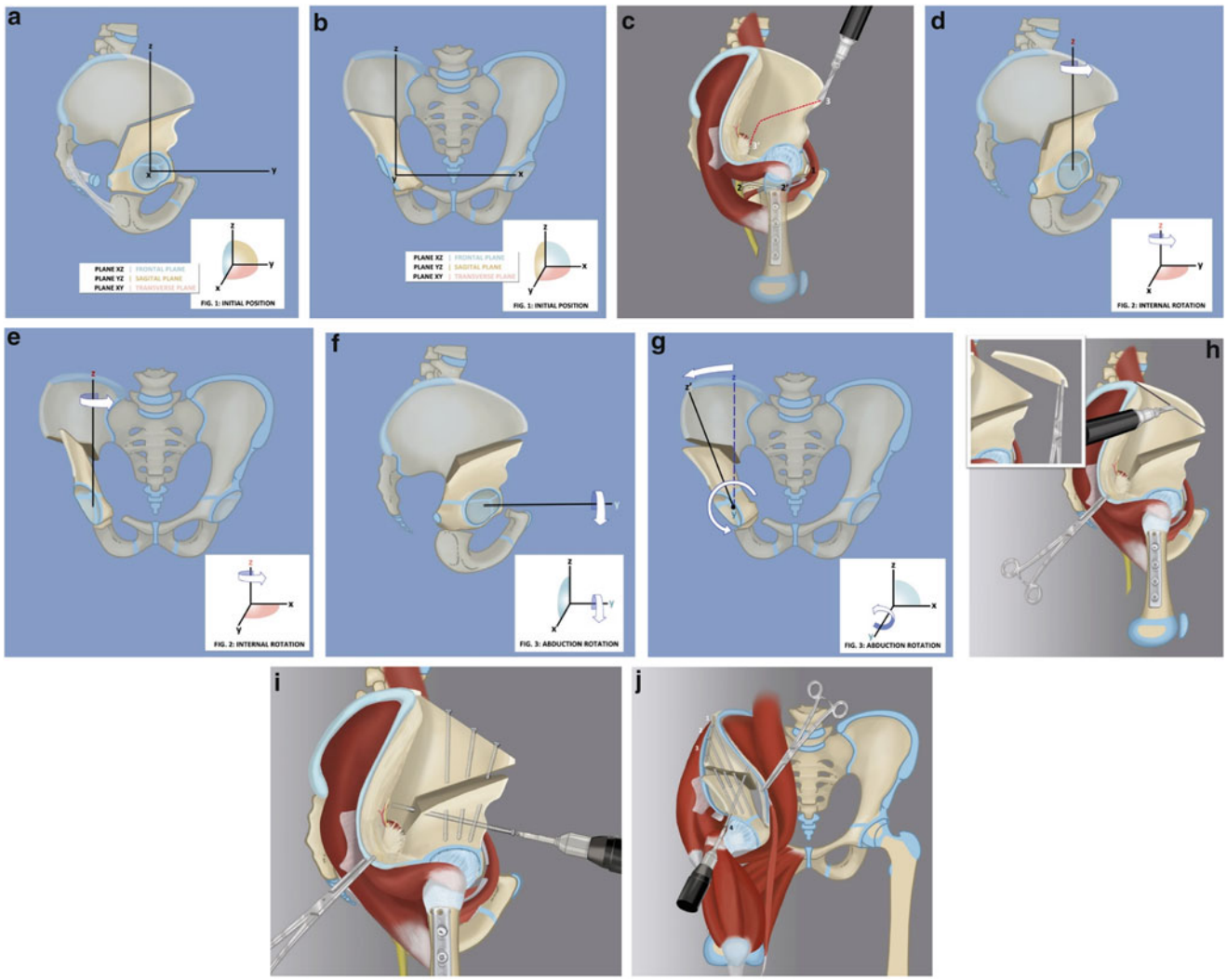


Fig. 22.12 (**a**) Periacetabular triple osteotomy lateral view (Paley). The bone cuts are outlined. The sacrospinous ligament is also released to untether the acetabulum. (**b**) Periacetabular triple osteotomy, AP view (Paley). (**c**) The ischium is cut under direct vision after identifying and decompressing the sciatic nerve. The nerve is not disturbed since the bone is deeper. (**d**) Internally rotate osteotomy to gain better posterior coverage and treat the retroversion of the acetabulum. Lateral view. (**e**) AP view of internal rotation of osteotomy. (**f**) Abduct the osteotomy to gain lateral coverage. (**g**) Abduct the osteotomy to gain lateral coverage. (**h**) Resect the upper end of the iliac wing to allow closure of the hip abductors. (**i**) Fixation is with several antegrade and one retrograde screw. (**j**) Front view of coverage and fixation

table of the pelvis, especially anteriorly. The Paley-Dega modification avoids doing so and also extends farther posterior and distal than other publications of the Dega. In this manner, there is true hinging in a three-dimensional way on the triradiate. Even the anterior part of the cut does not exit the cortex and extends to the triradiate junction between the ilium and pubis.

The bump under the buttocks should be removed at this stage. This allows one to assess the coverage of the femoral head in the true frontal plane. The laminar spreader can be distracted as needed to gain additional coverage and reorientation of the acetabular sourcil. The laminar spreader should be kept posterior to avoid increasing the anterior coverage. In this manner, predominantly lateral coverage is augmented. At this point, the femoral shortening should be carried out and the excised bone segment cut to fit the opening wedge gap of the pelvic osteotomy. Additional bone from the iliac crest, which is resected during the abductor slide, may be inserted to fill the remaining space.

Paley Modification of Periacetabular Triple Osteotomy (Paley PATO) (Fig. 22.12)

1. Pubis: The pubis is exposed subperiosteally by reflecting the medial iliac apophysis and periosteum more medially. The triradiate cartilage arm at the pubis is exposed and the dissection carried out just distal to it. The superior pubic

ramus can be cut under direct vision under the protection of two Hohmann retractors with an osteotome or a saw.
2. Ischium: The ischium is exposed by returning to the back of the femur and finding the sciatic nerve again. The nerve is followed to the ischium. To avoid stretching the nerve, do not place a retractor between the nerve and the ischium. Subperiosteal elevation of the ischial periosteum is carried out. The ischium should be cut under direct vision near the junction where it forms the inferior wall of the acetabulum, distal to the triradiate cartilage.
3. Sacrospinous ligament release: For large corrections, it is important to release the tether of the sacrospinous ligament. This ligament can be palpated after dissecting the medial iliac periosteum off of the quadrilateral plate inside the true pelvis. A finger can be placed on the tip of the ischial spine, and the ligament is then felt. Using proprioception, a finger from the other hand can be used to palpate the finger that is on the ligament. Make sure that the sciatic nerve, which can be visualized laterally, is not in the way. With the ligament isolated in this manner, it can be cut with scissors from the lateral side.
4. Ilium: The ilium is cut with a saw from the anterior superior iliac spine towards the junction of the true and false pelvis. On the lateral side of the ilium, it should be osteotomized towards and into the sciatic notch. On the medial side it should be cut with an osteotome into the sciatic notch. The osteotomy on the medial side has an angle of about 120°. This bend in the cut rounds off the corner of the distal segment and creates a spike of bone on the proximal segment. This increases the surface area and creates a posterior buttress while making the distal segment rotate more easily with less distraction.

To gain posterior and lateral coverage, the acetabular segment should first be rotated internally along the long axis of the body. This gives posterior coverage. It should then be rotated laterally. This gives lateral coverage. To rotate internally, a lion-jaw bone clamp is placed anteroposteriorly, superior to the acetabulum and the bone fragment rotated internally. It can also be placed around the pubis to lever the acetabulum laterally. If the acaetabular fragment does not move sufficiently, then each osteotomy site should be checked for completeness. The ischial osteotomy is often the culprit, and the periosteum around the ischium may need to be divided. Fixation of the triple osteotomy is achieved using a long 3.5-mm or 4.5-mm screw from the ilium to the acetabular fragment. However, because of the abductor slide, the proximal part of the iliac wing must be resected prior to screw fixation. The description of how to do this is described in a later step. Once complete, multiple drill bits can be placed from the remaining iliac wing towards the acetabular fragment, from proximal lateral to distal medial. One drill bit is placed from the anterior inferior spine, retrograde towards the base of the iliac wing from anterior to posterior. A same sized drill bit is used to compare lengths with the drill bits inside the bone to determine the screw lengths. One by one, the drill bits are removed, and replaced with solid, non-cannulated screws, making sure that they do not enter the triradiate cartilage or the hip joint.

Ganz Periacetabular Osteotomy (PAO) (See Fig. 22.11)

The technique for this has been previously published, though there are some variations when performed with a Superhip procedure. This osteotomy is indicated for patients who are skeletally or near-skeletally mature, usually with quite severe acetabular dysplasia.

To do the abductor slide, an osteotomy of the iliac crest is done from lateral to medial. There is no iliac apophysis to be split, so the proximal 1 cm of crest is cut with a saw after first elevating the medial periosteum with the iliacus muscle. The periosteum of the medial side of the ilium is present at every age and dissects off the bone easily, even in older patients. The periosteum on the lateral side of the ilium will not strip off easily after childhood. The glutei fix to bone with Sharpey's fibers, making subperiosteal dissection impossible in adulthood. Therefore, the glutei are not detached from the top of the crest and are stripped laterally after an osteotomy of the crest of the ilium. This includes the TFL. The sartorius and the iliacus muscles are stripped medially. A small cube of bone is cut with a saw and reflected with the sartorius.

The Ganz PAO consists of five osteotomies of the ilium, ischium, and pubis:
1. The first osteotomy is of the pubis. The pubis is exposed subperiosteally and cut with a saw and an osteotome. A sharp Hohmann retractor can be driven into the distal pubis and retracted to provide better exposure.
2. The second osteotomy involves part of the ischium. The ischium is cut from anterior to posterior through half of its thickness. A special angled osteotome is used for this. A space is created just distal to the pubis between the femur and the ischium. The hip is kept partially flexed at all times to help protect the femoral nerve. A blunt instrument can be inserted to feel for the ischium, which is quite deep. The curved osteotome is inserted down to the ischium. It is then cut part way through the bone.
3. The third osteotomy is partway through the ilium. The periosteum should be dissected off of the quadrilateral plate inside the true pelvis. With a finger, the surgeon can feel and find the ischial spine and feel the sacrospinous ligament. An iliac osteotomy is made from just distal to the anterior superior iliac spine to the brim of the true

pelvis. A mark is placed at the junction of the true pelvis with the false pelvis in line with the sciatic notch. An imaginary line can be drawn from this point parallel and just anterior to the anterior wall of the notch, ending at the level of the ischial spine. This osteotomy can be made with a saw.

4. The fourth bone cut is made with an osteotome from the pelvic brim into the true pelvis to end anterior to the ischial spine. To avoid stretch to the femoral vessels the hip should be flexed at all times during this osteotomy. An osteotome is inserted with its lateral part posterior and its medial part anterior. It should be inclined with its handle towards the belly. Either by feel or using the image intensifier, we need to orient this parallel to the anterior wall of the sciatic notch and stop opposite the ischial spine.
5. The fifth and final osteotomy is the completion of the ischial osteotomy. The same curved osteotome used for the anterior ischium is used here to complete the ischial cut. The osteotome is oriented towards the quadrilateral plate. It should be inserted to start at the end of the fourth cut. Once the ischial cut is complete the acetabular segment is mobilized.

A lion-jaw bone clamp is placed on the pubis and the acetabulum is rotated laterally outwards. The segment may need to be internally rotated due to retroversion. If it will not move sufficiently, there is often a tether at the pubis due to the attached muscles. Releasing these frees the acetabular segment to move. Once the segment is placed in the desired position, screw fixation is used to secure it. Again, the wing of the ilium should be shortened first to allow the abductors to slide distally. Three antegrade screws and one retrograde screw (as in the above periacetabular triple osteotomy) are inserted to secure the fragment. The abductors are secured back with screws through the crest segment into the iliac wing.

Knee Considerations in CFD

The knee in CFD may range from a normal, stable, undeformed knee to a contracted, unstable, deformed joint. The most common deformity of the knee is valgus, which is usually nonprogressive. The distal femoral physis is usually closer to the knee joint on the lateral side, often due to hypoplasia of the lateral femoral condyle. CFD cases often have a variable degree of anteroposterior and rotatory instability of the knee related to absent or hypoplastic cruciate ligaments. In some cases, the tibia dislocates anterior or posterior on the femur during extension or flexion, respectively. Furthermore, there may be rotatory instability present. One study has related the radiographic appearance of the tibial spines to the degree of hypoplasia of the ACL [27]. The patella is usually hypoplastic and may maltrack laterally. In some cases, the patella dislocates with flexion. Finally, many cases of CFD have a fixed flexion deformity of the knee.

Indications for Preparatory Surgery of the Knee Prior to Limb Lengthening

Isolated anteroposterior instability is not necessarily an indication for reconstructive knee ligament reconstructive surgery in children with CFD. However, grade 3 ligamentous instability with no endpoint on anterior and posterior drawer tests will usually become symptomatic as the child gets older. If the child is going to undergo a Superhip procedure or Dega osteotomy prior to lengthening, instead of excising the fascia lata, it can be used to reconstruct the knee ligaments. In some children, there is a catching or locking sensation in the knee when going from extension to flexion. It may be painful and require a trick motion to release it. This is due to contracture of the iliotibial band, combined with aplasia of the ACL. In more severe cases, the tibia actually subluxates or dislocates anteriorly on the femur and reduces at about 30° of flexion. In older patients, the posterior aspect of the tibia may be rounded, contributing to anterior dislocation of the tibia on the femur. It is not clear whether this is a secondary change due to chronic dislocation or a primary deformity, since the tibia is not ossified posteriorly in infancy.

Patellar hypoplasia and instability are also very common. The patella frequently maltracks laterally with progressive knee flexion. In some cases, the patella may dislocate with knee flexion. This is due to a combination of factors: valgus distal femur, hypoplastic or absent patellar groove, contracture of the lateral retinaculum with the tight iliotibial band, and external rotatory instability of the tibia on the femur due to cruciate ligament deficiency, which lateralizes the patellar tendon insertion. Patellar maltracking or subluxation/dislocation should be corrected prior to limb lengthening.

Flexion contracture of the knee is another congenital deformity that may be present and should be corrected before proceeding with limb lengthening. When the femur is very short, the acute angle created by the posterior thigh muscles gives the appearance of a flexion contracture. The definition of a flexion contracture is a flexed angle between the anterior cortical line of the femur and tibia in maximum extension. When the contracture is greater than 15°, it should be corrected surgically. Knee flexion deformity can be due to bony or soft tissue causes. In CFD, it is commonly an intra-articular capsular contracture. There may be some extra-articular contribution due to contracture of the hamstring muscles and gastrocnemius muscles. Release of these muscles alone rarely corrects the contracture while capsular release without complete hamstring release corrects the contracture. In some cases there is a true bony flexion of the distal femur that may need to be corrected by osteotomy.

Box 22.9. History of Superknee Procedure

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The knee reconstruction that I developed in 1994 [42] is called the Superknee procedure. The Superknee is a conglomerate procedure combining two or more of the following five procedures, three of which were previously described by other authors, and two of which were developed by me: (1) the Langenskiöld procedure [65] for congenital dislocation of the patella; (2) the MacIntosh procedure [66] for ACL deficiency including extra and intra-articular ACL reconstruction using the fascia lata; (3) the Grammont procedure [67] for recurrent dislocation of the patella; (4) the Paley procedure also referred to as the reverse MacIntosh [66] to prevent external rotatory instability and to act as an extra-articular posterior cruciate ligament; and (5) the Paley anterior approach to posterior capsulotomy of knee. Combinations of two or more of these five procedures are referred to as the Superknee procedure and can be performed at the same time as a pelvic osteotomy and/or Superhip procedure.

The Superknee procedure is a combination of some or all of these components including extra- and intra-articular knee ligament reconstruction, patellar realignment, posterior capsulotomy, and knee flexor tendon releases. Typically, the Superknee consists of the MacIntosh extra- and intra-articular ACL reconstruction, the reverse MacIntosh (Paley) PCL extra-articular reconstruction, the Grammont patellar tendon realignment, lateral release of the patella, and in some cases the modified Langenskiöld (Paley) procedure for patellar reduction. If performed with a Superhip procedure, the incision is a distal extension of the Superhip incision. If performed as an isolated procedure, it can be done through one midline anterior incision or one medial and one lateral incision. If the Superknee is performed without the Superhip procedure, the entire surgery can be performed under tourniquet control. My personal preference is to use the Hemaclear tourniquet (HEMACLEAR, New Jersey) because of its narrow width. There is typically not enough room in a young patient with a congenital short femur for a pneumatic tourniquet to be placed proximal to the surgical incision. The release of the posterior capsule is performed only when there is a significant knee flexion contracture of greater than 15°.

David MacIntosh first described an extra-articular ligament reconstruction for the ACL-deficient knee and later a combined extra- and intra-articular reconstruction with the "over-the-top" technique. This method, although no longer used in sports medicine, is a very

(continued)

Box 22.9. (continued)

useful technique for congenitally deficient knees. In CFD, the instability pattern is different than in an isolated tear of the ACL. There is more of a rotary instability in CFD. Therefore, a purely intra-articular ligament reconstruction is insufficient and can lead to recurrent instability. Thus, the combination of extra- and intra-articular ACL ligament reconstruction is ideal. Having studied under MacIntosh and having learned this procedure directly from him, it was natural for me to think about its application in the CFD cruciate-deficient knee. I have modified this procedure slightly to adapt it to the skeletally immature knee. In my experience, it can now be safely done as young as 2 years of age.

To prevent the tibia from rotating externally, which leads to subluxation of the patella and of the tibia on the femur posteriorly, a medial extra-articular ligament can be created using the medial half of the fascia lata. This creates a strap going around the medial tibia tethering it to the medial distal femur. This is the opposite direction of the extra-articular ligament created with the posterior half of the fascia lata, which is described in the next step. This lateral extra-articular ligament was described by Dr. David MacIntosh for ACL reconstruction. In his honor and memory (he was one of my professors and mentors in Toronto in 1985) and in recognition of his idea of a lateral extra-articular ligament, I have referred to the medial extra-articular ligament procedure as the reverse MacIntosh, or extra-articular PCL procedure.

Superknee Surgical Technique (Ligamentous Reconstruction Only) (Figs. 22.9 and 22.13)

1. *Fascia lata harvest.* The knee is exposed through a long lateral incision ending just distal to the tibial tuberosity distally and midlateral proximally. The anterior margin of the fascia lata (iliotibial band) and the posterior margin where it blends with the intermuscular septum are incised longitudinally. The fascia lata is transected as at its musculotendinous junction with the TFL and reflected distally until its insertion onto the tibia (Gerdy's tubercle).
2. *Fascia lata preparation.* The fascia lata should be split into two longitudinal strips to make two ligaments. If the fascia is not very wide, bias the posterior limb to be larger. The posterior half is tubularized using nonabsorbable suture (#2 Tiger loop™) and a Krackow whipstitch from distal to proximal. The medial half is left flat.

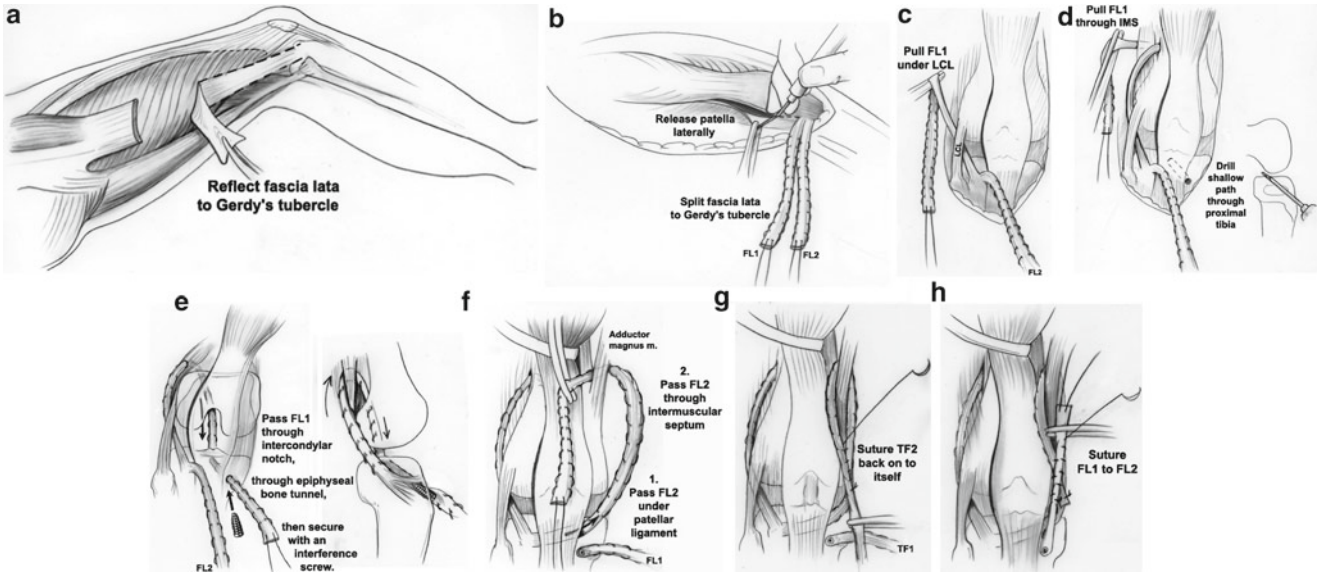


Fig. 22.13 Superknee surgical technique including MacIntosh and Paley reverse MacIntosh illustrations. (**a**) Harvest iliotibial band. (**b**) Divide the ITB into two halves. Do lateral release if maltracking patella. (**c**) Pass the posterior half under the LCL. (**d**) Pass it over the top of the intermuscular septum. (**e**) Drill a hole in the epiphysis of the proximal tibia and pass the ITB through this hole. Secure with interference screw with the knee in extension (MacIntosh). (**f**) Pass the anterior half of the ITB under the patellar tendon and around the adductor magnus and (**g**) suture it back to itself with the knee in 90° flexion (Paley reverse MacIntosh). (**h**) Suture the two new ligaments together

3. *Lateral release and Grammont patellar tendon realignment*: If the patella maltracks laterally, but it is not dislocated or dislocating, a lateral release and patellar tendon transfer should be performed to move the tendon medially. The lateral capsule and edge of the vastus lateralis should be cut, but not through the synovium. The vastus lateralis is elevated from the lateral intermuscular septum and distal femur. If it is still acting as a major lateral tether of the patella, its tendon should be detached from the patella and then transferred medially at minimal tension.

   The lateral release is extended distally to the lateral aspect of the patellar tendon. This longitudinal deep incision should be extended past the tibial tuberosity along the crest of the tibia incising the proximal tibial periosteum. A parallel periosteal, para-tendinous deep incision is made medial to the patellar tendon. Using sharp dissection with a knife blade, the patellar tendon is elevated off the tuberosity trying not to remove any cartilage if possible. Once the tendon is detached the periosteal extension of the tendon is elevated with the tendon so that the detached tendon remains tethered distally. This procedure (described by Grammont in German [67]) is the pediatric equivalent of the Elmslie-Trillat [67] procedure in adults. The patellar tendon can now be displaced medially and sutured medial to the tuberosity with an absorbable suture.
4. *Extra-articular PCL reconstruction (reverse MacIntosh).* To anchor the ligament to the medial femur, it is necessary to elevate the skin flap in a medial direction. The anterior skin flap is kept as thick as possible and is reflected medially until the posterior border of the vastus medialis can be visualized. The medial intermuscular septum (very rudimentary) and the adductor magnus tendon are located posterior to the vastus medialis muscle. Care should be taken dissecting in this area to isolate and not damage the saphenous nerve, as it exits from the quadriceps as the terminal branch of the femoral nerve.

   A subperiosteal tunnel is created around the adductor magnus tendon. The anterior limb of the fascia lata is now passed under the patella tendon, through a tunnel of superficial medial retinaculum, looped around the adductor magnus insertion from posterior to anterior, and then sutured to itself with nonabsorbable suture. This extra-articular ligament should always be tensioned with the knee in 90° of flexion. If it is tensioned in extension it may restrict flexion. The excess "ligament" is saved and sutured to the remaining end of the reconstructed ACL as it exits the tibia in the next step.
5. *MacIntosh extra + intra-articular ACL reconstruction.* The lateral collateral ligament (LCL) is identified using Grant's test. This is performed by putting the leg in a figure-four position, which allows the tensioned LCL to be easily palpated. Dissect the LCL under tension, identifying its anterior and posterior borders. An extra-articular tunnel is created under the LCL. The posterior limb of the fascia lata is passed through the LCL tunnel from anterior to posterior. A subperiosteal dissection is done in the over-the-top region, preserving the intermuscular septum

attachment. A curved tonsil clamp introduced through the "over-the-top" tunnel is used to perforate the posterior knee capsule centrally.

Next, a drill hole needs to be made in the anterior tibial epiphysis to anchor the reconstructed ligament to bone. Drill a guide wire through the anterior tibial epiphysis using image intensifier guidance. The wire should start proximal to the proximal tibial physeal line, lateral to the patellar tendon. The wire should be directed proximally and posteriorly, exiting into the center of the knee about halfway back on the tibia. As CFD patients lack a true notch and a notch-plasty is not an option, it is better to keep the ligament more posterior on the tibia than the normal knee. This avoids damage to the ligament with knee extension.

Once the wire is confirmed to be in the correct position in both the AP and lateral views, a hole is drilled with a cannulated ACL reamer. The diameter of the reamer chosen is matched to the diameter of the tubularized fascia lata ligament, which can be measured with a sizer. A suture passer is then passed through the tibial epiphyseal tunnel and out the capsular perforation through the "over-the-top" tunnel laterally. The suture connected to the fascia lata is looped through the suture passer and pulled through the knee to exit anteriorly through the epiphyseal tunnel. The tubularized fascia lata is now pulled through the knee capsule and out the epiphyseal tunnel.

A transosseous suture is placed into the supracondylar region to help anchor the extra-articular ligament and to support the over-the-top point. The extra-articular ligament should be tensioned in full knee extension. Once it is secured to the anchor, the rest of the graft is anchored to the bony tibial epiphyseal tunnel using an absorbable headless interference screw. Again, tension the ligament in full knee extension to prevent a flexion contracture of the knee. As the patient's epiphysis grows, the ligament will become more taut. Therefore, mild laxity in the ligament is acceptable. The ligament can now be sutured to the anterior limb graft for additional fixation.

Superknee Procedure with Patellar Realignment for Dislocated/Dislocating Patella (Figs. 22.11 and 22.14)

Langenskiöld Patellar Realignment

If the patella is dislocated or dislocating, a modified version of the Langenskiöld [65] procedure is performed before the ligament reconstruction. First, the capsule is incised and separated from the patella and synovium both medially and laterally. On the medial side, the two layers are separated all the way to the medial gutter. The medial capsule is cut transversely at its distal end. The patellar tendon and the quadriceps are also separated from the synovium distally and proximally, respectively.

The synovium is now incised circumferentially around the patella, separating the patella from the synovium completely. The quadriceps tendon is left attached to the patella proximally, and the patellar tendon remains attached to the patella distally. The synovium is separated from these structures. The synovium now has a patella-sized hole in it, which is sutured closed in a longitudinal direction. This leaves the patella temporarily as an extra-articular bone. The patellar tendon is elevated from the apophysis by sharp dissection after circumscribing a medial and lateral incision extending distally into periosteum (Grammont procedure).

After the tendon is elevated, it is shifted medially at least a centimeter. In the original Langenskiöld, the patellar tendon is detached from the tuberosity. A longitudinal incision is made in the synovium, the center of the patella is inserted into this new hole, and the synovium is sutured to the patella circumferentially. The medial capsule with the vastus medialis is now advanced over the top of the patella and stitched to its lateral border. The lateral capsule is left open. If the Paley reverse MacIntosh procedure is used, the fascia lata should not be fixed in place until after the Langenskiöld repair is completed.

Superknee Procedure with Knee Flexion Deformity (Figs. 22.10 and 22.15)

Knee Flexion Contracture Release

If there is a knee flexion deformity >15°, it can be treated by posterior capsular release. This is often done in combination with a Superhip procedure or one of the knee reconstruction procedures described above. To avoid direct surgical and indirect stretch injury to the peroneal nerve, this nerve should be identified and decompressed at the neck of the fibula (see surgical technique below, peroneal nerve decompression).

Next, the biceps tendon should be Z-lengthened. Extend the decompression of the common peroneal nerve under the biceps muscle. With the nerve protected and visualized, Z-lengthen the biceps muscle and tendon. In congenital cases, there is little tendon and mostly muscle. The biceps muscle consists of two parts: the short and long heads. To avoid damage to this muscle, the short head should be dissected off of the periosteum of the femur. Try and keep as much of the muscle together and reflect this part proximally. Isolate as much of the tendon as possible with sharp dissection off of the muscle and reflect this part distally.

The lateral head of the gastrocnemius should be released from the femur. It has a very broad insertion of muscle and tendon. The lateral capsule can now be identified. Dissect the contents of the popliteal fossa away from the posterior capsule. To confirm that it is the posterior capsule, incise it

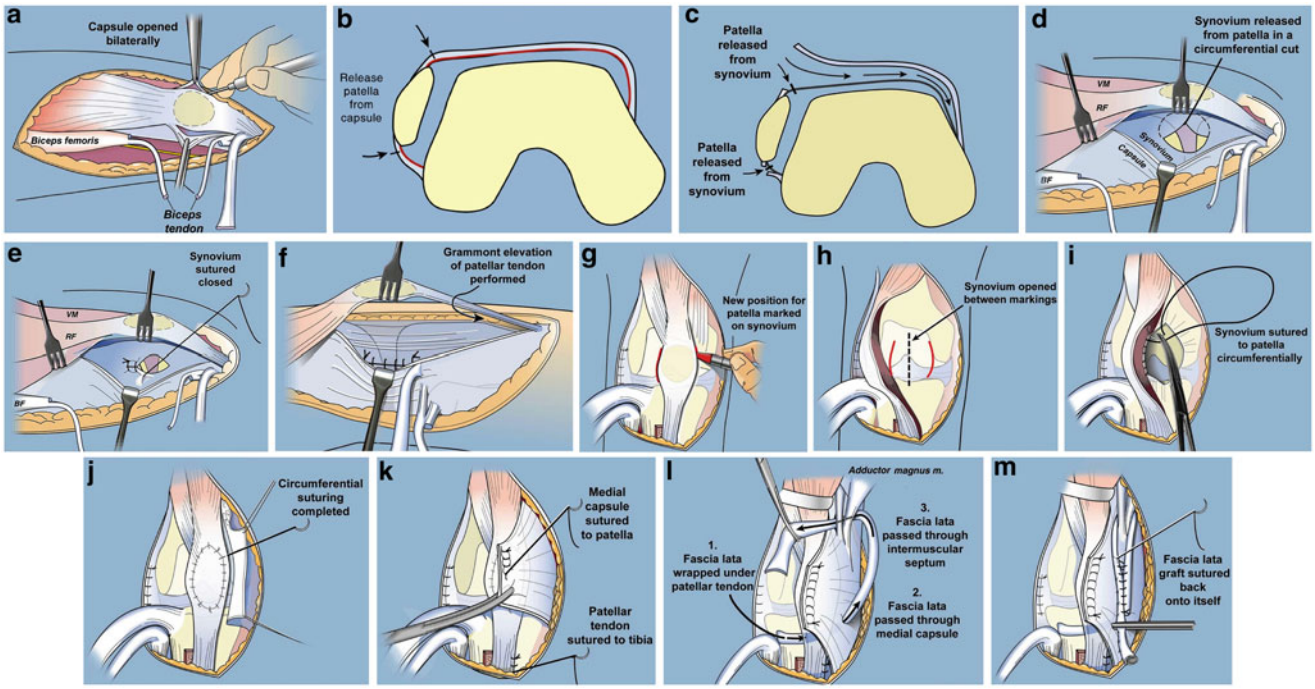


Fig. 22.14 Superknee surgical technique including Grammont, modified Langenskiöld, and Paley reverse MacIntosh procedure illustrations. (**a**) Release capsule on the lateral and medial sides of the patella. Medially cut through the vastus medialis and separate it from the rest of the quadriceps. (**b**) Do not cut through the synovium. (**c**) First separate the capsule from the synovium extensively and circumferentially around the patella and then cut the synovium to release the patella. (**d**) The synovium is also released from the underlying muscle and tendon. This leaves a patella-sized hole in the synovium. (**e**) Suture the hole in the synovium longitudinally. (**f**) Sharply dissect the patellar tendon off of the cartilaginous apophysis extending the dissection distally with a strip of periosteum (Grammont procedure). (**g**) Centralize the patella and mark this new position on the synovium with the knee in extension. (**h**) Incise the synovium between the marks. (**i**) Suture the lateral border of the synovium to the patella and continue circumferentially (Langenskiöld). (**j**) Once the entire synovium is sutured to the edge of the patella the patella will track correctly. The medial capsule with the vastus medialis can now be advanced. (**k**) The medial capsule is advanced and sutured to the lateral side of the patella. The medial patellar tendon is also sutured to secure the medial advancement of the patellar tendon (Grammont). (**l**) Pass the anterior half of the ITB under the patellar tendon and under the medial retinaculum and loop it around the adductor magnus tendon (Paley reverse MacIntosh). (**m**) Secure it back to itself with the knee in flexion. If ACL reconstruction is needed the MacIntosh procedure can be done using the posterior half of the ITB and secured in extension as in Fig. 22.13e

posterolaterally and enter the knee joint. With the knee flexed, the only vascular structures that one should see at the level of the knee joint are the central geniculate artery and vein. These can be dissected free and cauterized. Care should be taken to make sure that the dissection does not inadvertently go distal to the level of the knee joint. If the dissection is behind the tibia instead of the knee joint line, the anterior tibial vessels may be encountered or injured.

The rest of the popliteal soft tissues can be carefully dissected off of the capsule all the way to the medial side. The capsule can be cut under direct vision. A head lamp may be useful for this part of the procedure. Once the posterior capsule is open, try and extend the knee joint. If there is still too much resistance, then the medial side will need to be exposed.

If performing a Superknee with ligament repair, a medial skin flap will need to be elevated anyway. If the posterior capsulotomy is being performed without the rest of the Superknee, then the dissecting scissors can be inserted from lateral to medial until they can be seen under the medial skin. A separate small skin incision can be made medially to visualize the medial capsule. The medial head of the gastrocnemius is identified as the only structure with a muscle inserting proximally. The medial head should be released from the femur, being careful not to injure the femoral vessels that lie immediately lateral to the muscle. The posteromedial capsule is dissected free from the popliteal fossa to communicate with the lateral dissection. The posterior capsule can then be cut under direct vision from both sides. The knee should now be able to fully extend.

The collateral ligaments are left intact. If the medial hamstrings are felt to be tight after the capsulotomy, they can be lengthened through the medial part of the dissection. I prefer to recess the aponeurosis of the semitendinosus and semimembranosus tendons rather than doing a Z-lengthening. After doing the knee capsular release, the knee is examined for instability. If it is unstable, then the ligamentous reconstruction of the Superknee procedure is carried out as described above. Occasionally, there is posterior instability that requires an intra-articular PCL reconstruction. This can be easily done using the iliotibial band by passing one limb

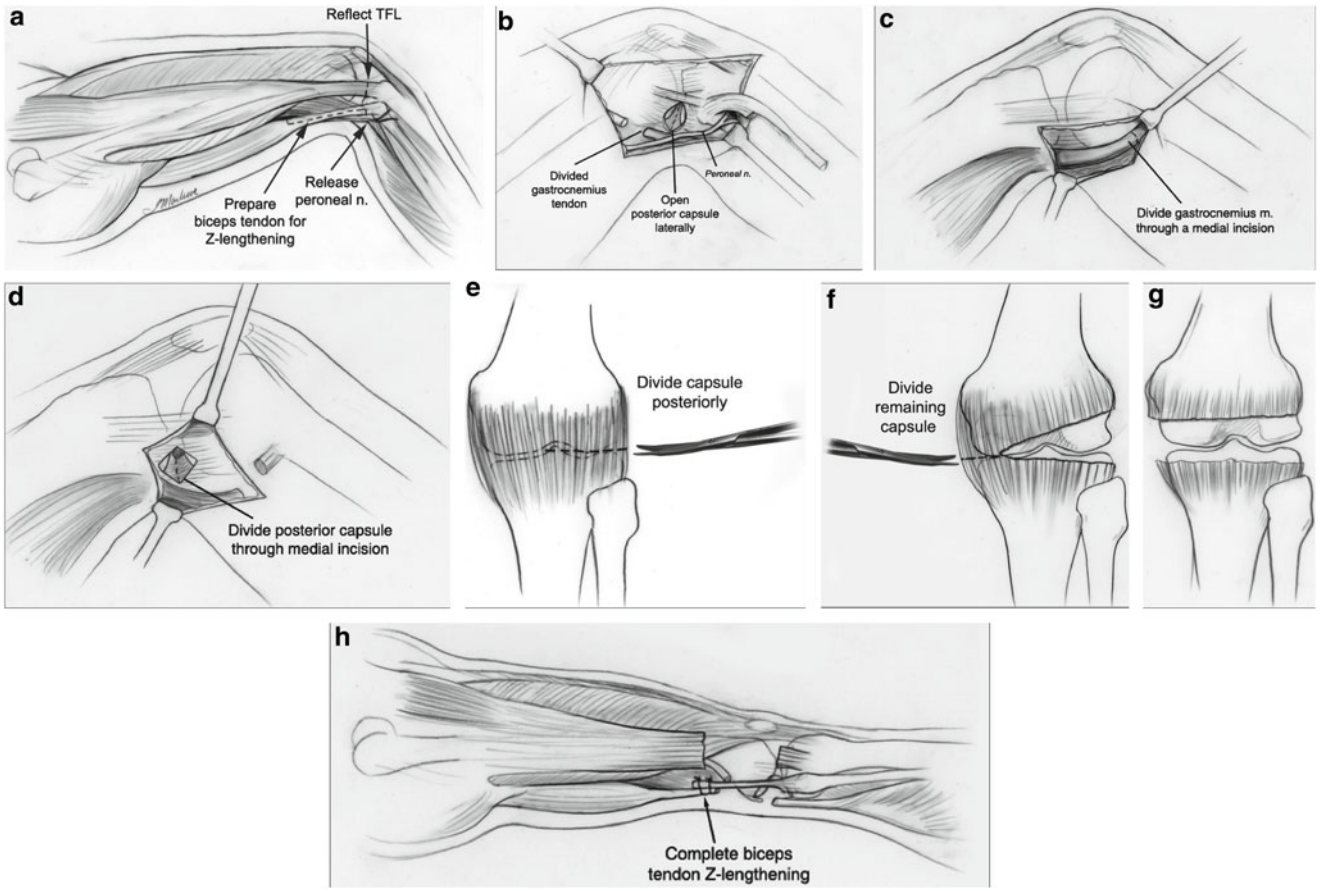


Fig. 22.15 Superknee surgical procedure including posterior capsulotomy, fascia lata, biceps and gastrocnemius tendon lengthening, and peroneal nerve decompression illustrations. (**a**) After lengthening the fascia lata (TFL) the biceps tendon is lengthened in a Z fashion. The peroneal nerve is decompressed as detailed in Fig. 22.16. (**b**) The lateral head of gastrocnemius is released off of the lateral femur. The lateral capsule posterior to the LCL is incised. (**c**) Through a medial incision the medial head of the gastrocnemius is released off of the femur. (**d**) The medial capsule is incised posterior to the medial collateral ligament. (**e**) The popliteal contents are dissected free of the posterior capsule from lateral and medial sides to connect together with the knee in flexion. The central geniculate vessels are cauterized. (**f**) The capsule can now be safely cut from the lateral side. (**g**) The capsular incision is completed from the medial side. (**h**) The biceps tendon is repaired in a lengthened fashion. (**i**) The fascia lata can be used for ligamentous reconstruction as previously described if needed

of it through an epiphyseal drill hole to the back of the knee. The band is then passed from posterior to anterior through a drill hole in the medial femoral epiphysis. It only needs to be secured at the femoral end.

Peroneal Nerve Decompression (Fig. 22.16)

Paley identified two sites of entrapment and described a surgical technique to decompress both tunnels [68]. In a cadaver study, Nogueira demonstrated that nerve tension increases after varus osteotomy and decreases to normal levels after peroneal nerve decompression [76].

The first peroneal nerve tunnel is located at the neck of the fibula. The common peroneal nerve leaves the undersurface of the biceps muscle to enter the lateral compartment of the leg. In order to enter the lateral compartment, the common peroneal nerve must perforate the otherwise intact fascial coverings of this compartment. The lateral compartment fascia folds over itself to form an arcade over the common peroneal nerve. The common peroneal nerve then splits into superficial and deep branches. The superficial peroneal nerve continues unimpeded through the lateral compartment, innervating the peroneal muscles.

The second tunnel is the intermuscular septum. The deep branch passes under the intermuscular septum to enter the anterior compartment of the leg, proximal to the intraosseous membrane. Anteriorly, the septum is confluent with the lateral and anterior compartment fascia. Posteriorly, it abuts but does not become part of the intraosseous membrane.

Surgery can be performed with or without a tourniquet, but without long-acting neuromuscular blockade. The patient is positioned supine, with a bump under the ipsilateral buttock.

The common peroneal nerve can be palpated in most people over the neck of the fibula. To palpate the nerve, roll it with the thumb or finger over the neck of the fibula. Make a 3–5 cm

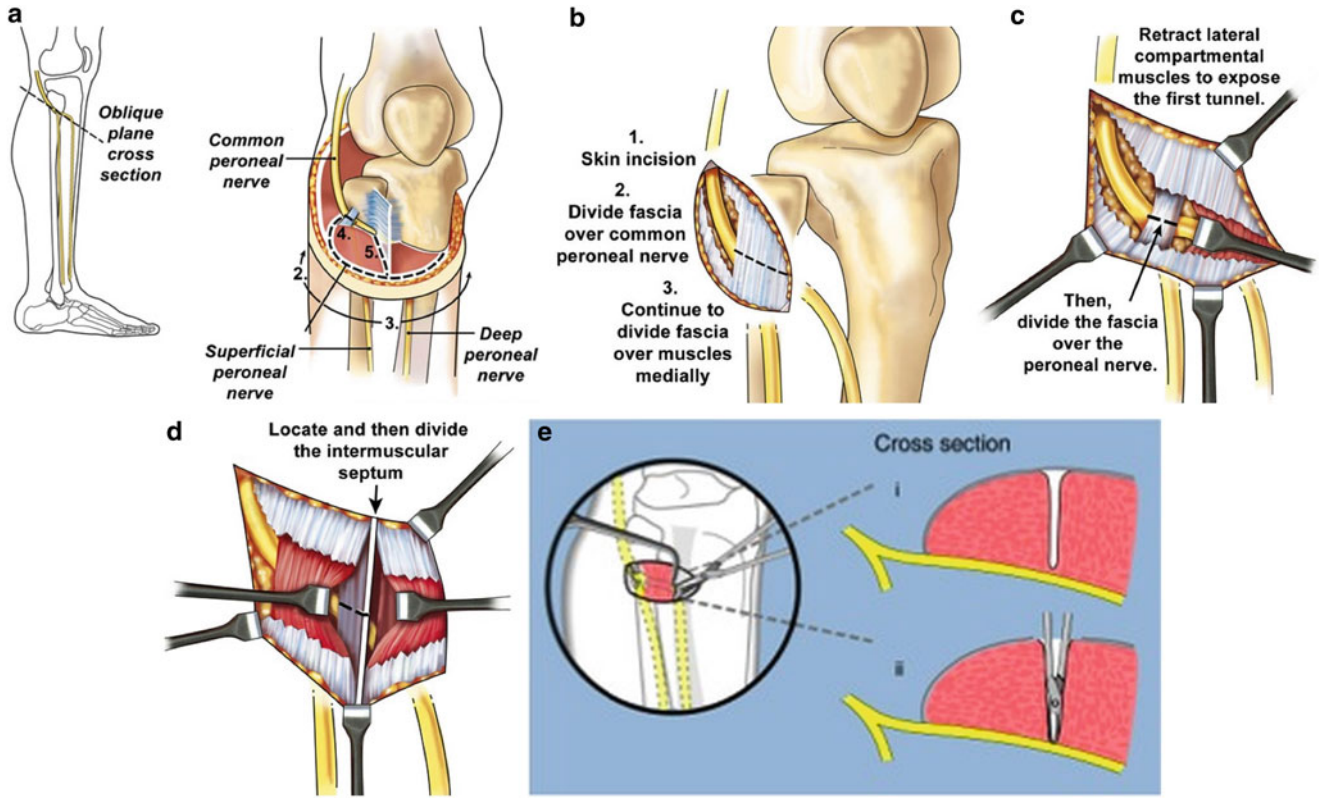


Fig. 22.16 Peroneal nerve decompression. (**a**) The peroneal nerve goes through two very tight fascial tunnels. The first is at the entrance to the lateral compartment and involves the entire nerve. The second is between the anterior and lateral compartments and involves the deep nerve only. (**b**) Start by incising the superficial fascia and identifying the borders of the common peroneal nerve proximal to the first tunnel. Incise the lateral compartment fascia in line with the nerve. (**c**) After incising the fascia retract the peroneal muscles to expose the first tunnel fascia covering the nerve. This fascia can now be cut under direct vision in a retrograde fashion. (**d**) Extend the transverse fasciotomy to the anterior compartment and expose the intermuscular septum between the anterior and lateral compartments. Cut this fascia to complete the decompression of the nerve. (**e**) The intermuscular septum comes down over the deep peroneal nerve like a guillotine. This narrow space under this septum is the second tunnel

oblique incision parallel to the course of the nerve over the neck of the fibula. Cut through skin and subcutaneous tissues down to the fascia, posterior to the lateral compartment. Feel for and identify the nerve again. Dissect and expose the nerve proximally and distally. It is not necessary to remove the fat over the nerve. Follow the common peroneal nerve distally until it disappears into the lateral compartment. Make a transverse incision over the lateral compartment fascia and expose the muscle of the lateral compartment. Retract the muscle of the lateral compartment medially to expose the underlying arcade of fascia covering the common peroneal nerve. This is the first tunnel, which should be released. The nerve can then be seen dividing into two branches, the superficial running down in the lateral compartment, and the deep branch going in the direction of the anterior compartment of the leg.

Extend the transverse compartment fasciotomy across to the anterior compartment. Notice the intermuscular septum separating the two compartments, confluent with the fascia overlying both compartments. Dissect the muscle off both sides of the septum. Be careful not to cut any superficial sensory branches of the nerve that may ascend the septum and innervate the skin overlying the compartments. With the septum exposed on both medial and lateral aspects, cut it from anteromedial to posterolateral. Be careful to stop as soon as the septum ends. Immediately below the septum, a band of fat can often be seen, which contains the deep peroneal nerve. This completes the decompression of the second tunnel of the deep peroneal nerve.

Let down the tourniquet at this point. The nerve can be stimulated with a disposable nerve stimulator. If the tourniquet time exceeds 20 min, the nerve may not initially respond to electrical stimulation due to the tourniquet ischemia. If the nerve was paralyzed before decompression, it can be stimulated before the decompression at the common peroneal nerve and then again after the decompression to evaluate if there is any immediate change.

Box 22.10. Superknee Tips and Tricks

- The stability of the knee cannot be fully assessed until the fascia lata has been released.
- Take a small portion of biceps fascia and intermuscular septum for a wider graft. If the graft is still narrow, bias the more important posterior limb of the graft to be larger.
- Send the ACL reamer all the way into the knee to make sure that the tunnel is adequately drilled for passing the ACL graft.
- Tighten the reverse MacIntosh with the knee in flexion, and the ACL graft with the knee in extension.
- Suture the ends of both limbs of the graft together at the end for additional fixation.

When cutting the posterior capsule, do not stray distal to the knee joint.

Femoral Lengthening for Paley Type I CFD

Choice of Osteotomy Level

Distal femoral osteotomy for lengthening has the advantage of a broader cross-sectional diameter for better bone formation and less deforming forces from the adductors and hamstrings. If distal femoral valgus is present, it can be corrected using a distal lengthening osteotomy. However, the distal osteotomy applies greater forces to the knee joint and increases the risk of knee stiffness and subluxation. These risks can be reduced by articulating across the knee joint and extending fixation to the tibia. Proximal femoral osteotomies have less effect on the knee, but have a greater effect on the hip joint. They produce a much higher risk of adduction contracture, hip subluxation, and dislocation. Extension of the external fixator across the hip is more complex and less desirable than articulated distraction of the knee. Proximal lengthening osteotomies are also more prone to poor bone formation and prolonged consolidation phase in patients with CFD. There is also a higher rate of fracture after removal of external fixation in CFD. For these reasons the author prefers to lengthen at the distal femur in patients with CFD.

However, the external rotation deformity of the femur with CFD should be corrected only by using a proximal osteotomy. The quadriceps muscle is in a normal relationship to the knee joint, and because most of the quadriceps muscle originates distal to the level of a proximal femoral osteotomy, a proximal femoral internal rotation osteotomy does not change the orientation of the quadriceps relative to the knee joint. A distal osteotomy leaves the bulk of the quadriceps muscle attached proximally in a lateral position and rotates the knee medially, thus increasing the effective Q angle and further increasing the tendency to lateral subluxation/dislocation of the patella in these patients. Varus deformity of the hip or proximal femoral diaphysis is corrected using a proximal valgus osteotomy, whereas valgus deformity of the knee is corrected using a distal varus osteotomy.

If the patient has previously undergone preparatory hip surgery, such as a Superhip reconstruction or proximal femoral osteotomy and Dega pelvic osteotomy, the proximal femoral deformities will already have been corrected and no proximal osteotomy is required at the time of lengthening. If hip preparatory surgery was not required, but there is external femoral torsion and proximal varus, then a proximal internal rotation and valgus osteotomy of the femur are carried out together with a distal lengthening osteotomy.

A distal femoral lengthening osteotomy can be used to gradually (ring fixator) or acutely (monolateral fixator) correct the valgus and any mild flexion deformity of the knee (see Figs. 22.10 and 22.11). For valgus correction alone, no peroneal nerve decompression is required. However, if a flexion deformity is to be corrected acutely, the peroneal nerve should be decompressed just prior to the correction at the same surgery. As noted above, this distal region of the femur has a wider cross-sectional area than the proximal femur and is not in the zone of poor healing sclerotic bone. Therefore, the regenerate bone in the distal femur is wider, stronger, and subject to more axial deviation muscular forces than in the proximal femur.

In older children with a wider medullary canal, implantable limb lengthening (Figs. 22.9 and 22.17) or lengthening over nail (LON) can be performed (see Fig. 22.17) [69]. A proximal osteotomy can be used for lengthening over nails because there is little risk of refracture. The internal rod supports the bone until consolidation is complete. Intramedullary nailing in children risks disturbing the growth of the trochanteric apophysis and avascular necrosis of the femoral head. To avoid the latter, we use a greater trochanteric starting point and a nail with a proximal bend (e.g., trochanteric entry femoral, humeral, or tibial nail). To avoid a coxa valga deformity, we prefer to use this technique in patients with coxa vara. A theoretical epiphysiodesis, created by the nail, may lead to gradual correction of residual coxa vara, although this has not been observed. Fixator-only lengthening is typically used for the first lengthening. LON or implantable lengthening is used for subsequent lengthenings if the anatomic dimensions of the femur permit. In the author's experience, there is a higher risk of deep infection in patients who have had previous external fixators. It is unclear if this is related to the number or degree of previous pin infections. It is suspected that latent bacterial forms remain in the bone in a dormant state and are activated at the time of reaming and nail insertion. During external fixator removal, we recommend aggressive curettage of all granulation tissue in the soft tissues and bone of the pin tract in an effort to reduce such latent infection.

Soft Tissue Releases for Limb Lengthening

In patients with CFD, soft tissue releases in conjunction with femoral lengthening are essential to prevent subluxation and stiffness of the knee and hip. If there is no contracture or tightness at the time of the index procedure, the soft tissue releases can either be performed at the index procedure or delayed until these soft tissues become contracted, approximately 6 weeks later. Usually, these are done at the index procedure to avoid an additional anesthetic. If a preparatory hip or knee surgery has already been performed, then the fascia lata, rectus femoris, and biceps femoris may have already been lengthened, and there is no need to repeat them. However, if soft tissue releases have not been performed, then they should be carried out at the time of the lengthening surgery.

Before surgery, the range of motion of the hip and knee should be evaluated using muscle lengthening tests and the presence of contractures or limitation identified. The muscle lengthening tests are the prone knee flexion test (Ely test) for the rectus femoris (see Fig. 22.3a), the straight leg raising test (popliteal angle) for the hamstrings (see Fig. 22.3b), hip abduction range of motion for adductor contractures, and hip adduction range of motion with the knee and hip in extension for the fascia lata. The Ober test is helpful for identifying fascia lata tightness (see Fig. 22.3c).

The rectus femoris is evaluated with the Ely test (see Fig. 22.3a). A positive Ely test demonstrates pelvic flexion at the hip during prone knee bend. The rectus femoris should be released through a small anterior inguinal incision. Even with a negative Ely test, the rectus may be released from anterior inferior spine, as it still tends to tighten with lengthening.

Hamstrings are evaluated by the straight leg raise, measuring the popliteal angle (see Fig. 22.3b). If the knee can fully extend with the hip flexed to 90°, then the popliteal angle is 0° and the hamstrings are not tight and thus such a patient would require no treatment before femoral lengthening. If there is a popliteal angle >0°, the hamstrings are already tight and will lead to contractures during lengthening. The author prefers lengthening the biceps femoris with the iliotibial band through a lateral approach. If needed, the medial hamstrings can be lengthened from the medial side.

If hip abduction is limited, percutaneous adductor tenotomies of the adductor longus and gracilis tendons should be performed. If hip subluxation or more severe adduction contractures develop during lengthening, then an open, more extensive adductor release, including the adductor brevis, is indicated. The adductor release is very important in proximal lengthening, but for distal lengthening, it is usually not necessary. Distal adductor magnus release has been described

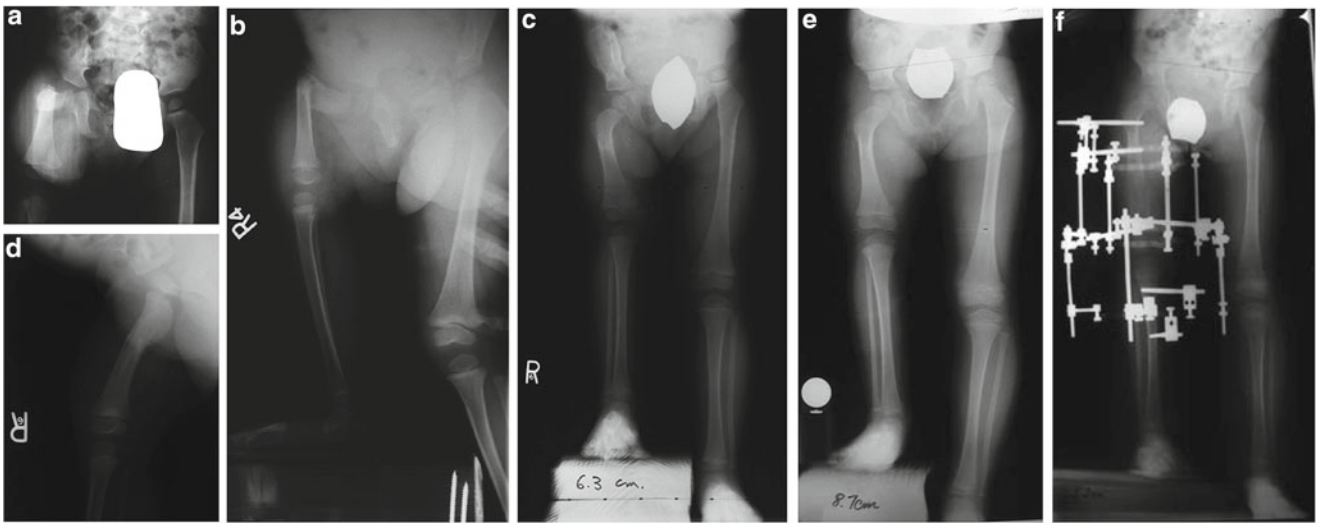


Fig. 22.17 Two-year-old girl born with Paley type 1b subtrochanteric type. AP and lateral radiographs (**a**, **b**) showing characteristic deformity as previous illustrated in Figs. 22.1 and 22.7. After the Superhip procedure performed at age 2 years, the femur appears to have a normal anatomy as shown on AP and lateral views (**c**, **d**). Standing X-ray prior to first lengthening. A 1-cm gain was achieved due to the Dega osteotomy (**e**). First lengthening performed at age 4 years, using Ilizarov apparatus (Smith & Nephew, Memphis, TN) with knee hinges and fixation to the tibia (**f**). After 8-cm lengthening (**g**). Full recovery of hip and knee range of motion and strength following first lengthening (**h**, **i**). Second lengthening performed at age 8 years, using a multiplanar monolateral external fixator (Orthofix LRS and Sheffield components, McKinney, TX). The knee is able to flex and extend with a lateral knee hinge connected to an arch fixed to the tibia (**j**). The femur was lengthened 7 cm (**k**) over a Rush rod similar to Bost's lengthening over nail method [69]. Third and final lengthening of 5 cm at age 15 years (**l**) was performed using an implantable lengthening nail (PRECICE, Ellipse Technologies, Irvine, CA). Note that prior to this a contralateral distal femur epiphysiodesis (5-cm equalization) was performed at age 10 and pelvic osteotomy and distal femoral varus osteotomy were performed at age 14. Total limb length equalization was 26 cm. The hardware was removed 1 year after the lengthening (**m**)

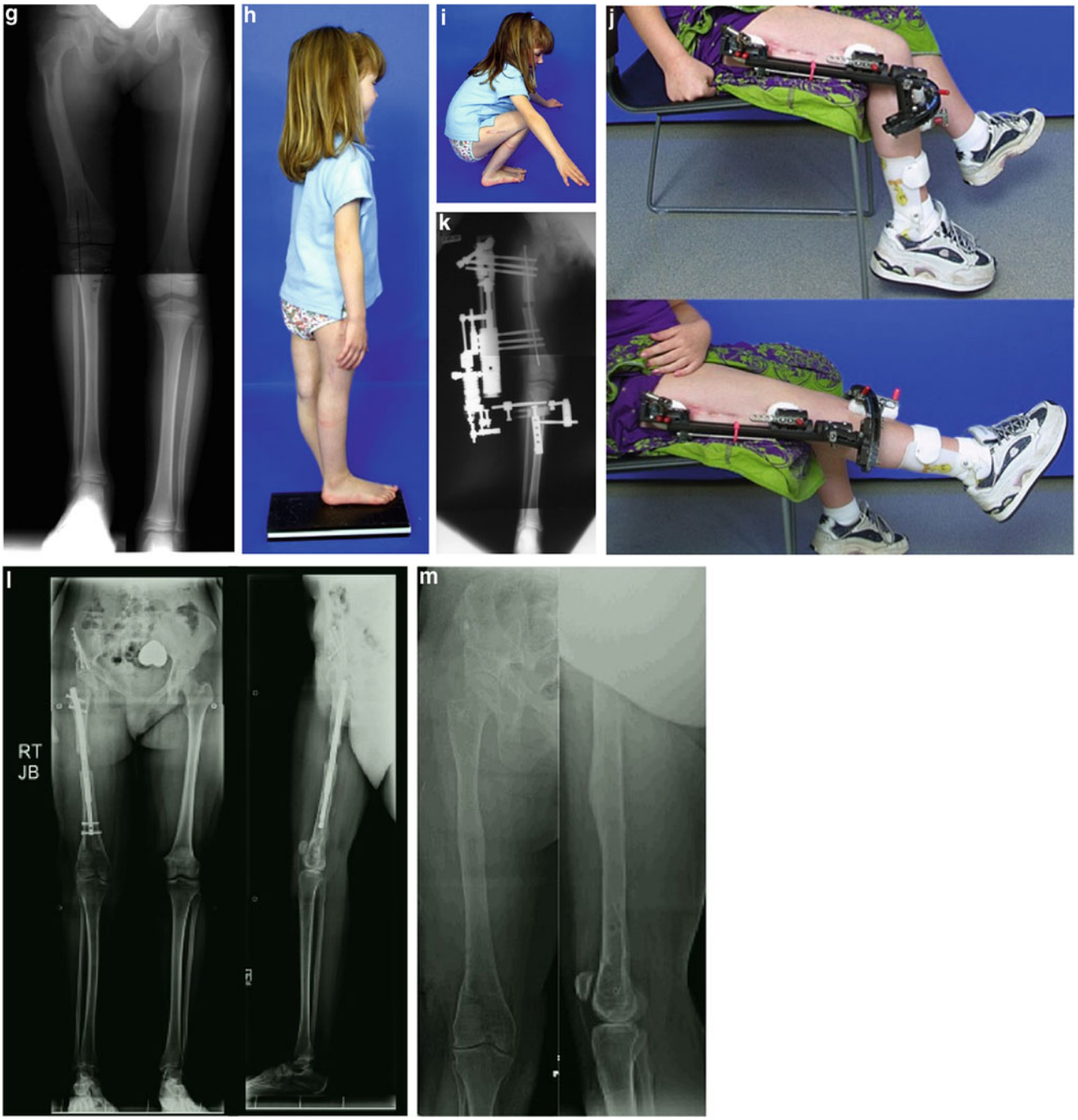


Fig. 22.17 (continued)

for congenital femoral lengthening [70], but this has not been found to be helpful in the senior author's opinion.

To confirm that the fascia lata is tight, perform an Ober test (see Fig. 22.3c). The patient lies on his or her side, and the knee is flexed and hip extended by pulling back on the foot and pushing forward on the buttock. The knee will stay up in the air and not fall down due to contracture of the fascia lata. This is called a positive Ober sign and indicates that the fascia should be excised or cut. In every case of CFD lengthening, if it has not already been excised, the fascia lata should be lengthened. A 3-cm longitudinal incision is made over the posterior edge of the fascia lata, where it connects with the intermuscular septum. The entire fascia lata is transected at the level of the proximal pole of the patella by dissecting anterior to the incision. If the incision is made more posteriorly, the biceps femoris can be easily exposed on the posterior side and the lateral biceps fascia can be safely recessed.

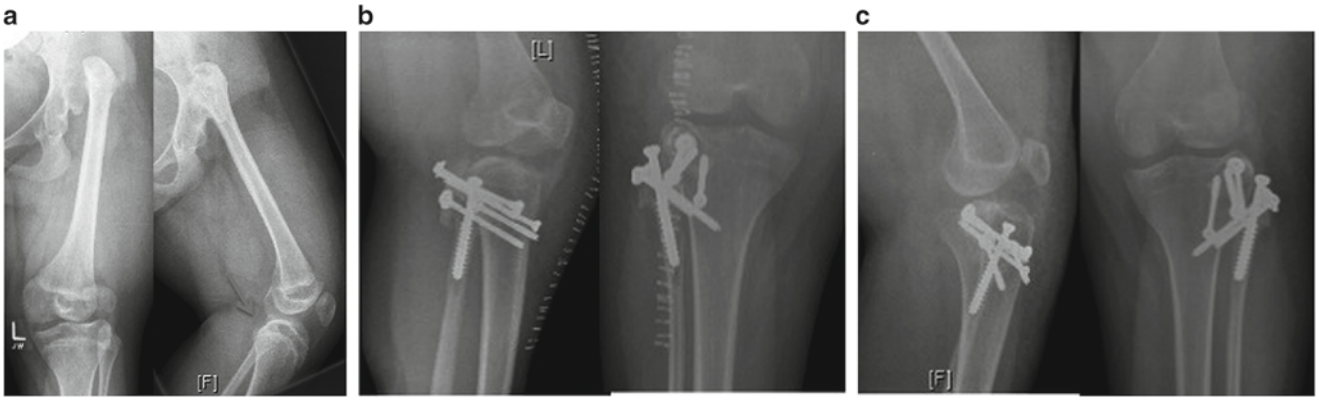


Fig. 22.18 Intra-articular osteotomy. (**a**) CFD Paley type 2 with rotatory subluxation of the knee joint. The posterolateral tibial plateau is round (*arrow*) and contributes to the subluxation. (**b**) The Superknee procedure was performed with decompression of the peroneal nerve and taking down the proximal tibiofibular joint to expose the lateral plateau. A hemi-plateau osteotomy of the posterior plateau was performed, with allograft and internal fixation. (**c**) After complete bone healing the lateral plateau is now flat and not round. The subluxation was eliminated by the Superknee procedure

Patients may develop hyperlordosis or a hip flexion or abduction contracture secondary to the femoral lengthening. If so, the proximal fascia lata is cut transversely through a small lateral incision just distal to the greater trochanter.

Botulinum Toxin Injection

The effect of botulinum toxin has been shown to be helpful in prevention of muscle contractures [71, 72]. In the author's experience, it decreases muscle stretch pain due to muscle spasm, which is primarily in the quadriceps muscle during physical therapy. Botulinum toxin can be injected into the quadriceps up to a limit of 10 U/kg body weight. The added volume of the saline is limited to avoid systemic toxicity, so injection is usually done with a total of 3–4 mL.

Knee Instability

Almost all cases of CFD can be assumed to have hypoplastic or absent cruciate ligaments, with mild to moderate anteroposterior instability. Some patients also have medial-lateral and torsional instability. Despite this, the knee tracks normally preoperatively, and there is no indication to perform ligamentous reconstruction in most cases. The significance of knee instability is the tendency of the knee to subluxate with lengthening. The subluxation is usually posterior, or posterolateral (posterior plus external rotation of the tibia on the femur), but it can also be anterior. Knee extension usually reduces posterior subluxation before lengthening. Therefore, to prevent posterior subluxation, some surgeons recommend splinting the knee in extension throughout the distraction phase [42]. This promotes knee stiffness while protecting the knee from subluxation. The author prefers to protect the knee by extending the external fixation to the tibia with a hinge. The hinge permits knee motion while preventing posterior as well as anterior subluxation. It also prevents pressure being transmitted to the knee joint cartilage. Hinges are an integral part of circular external fixators such as the Ilizarov device, as well as some monolateral external fixators such as the Orthofix Limb Reconstruction System (LRS), Smith and Nephew Modular Rail System.

Less common is anterior subluxation or dislocation of the tibia on the femur. This occurs as the knee goes into extension. It is important to document the angle of flexion the knee relocates or, conversely, at which angle short of full extension the knee dislocates. These dislocations are more commonly due to a posterior deficiency with rounding of the tibial plateau, but they can also be due to an anterior deficiency of the distal femur, in which the lateral radiograph of the knee shows a lack of the anterior protuberance of the femoral condyles. If the posterior condyle is rounded but the anterior part of the knee is straight, an intra-articular osteotomy is required to elevate the posterior condyle: intra-epiphyseal in children and metaphyseal in adults (Fig. 22.18). This can also be combined with a PCL reconstruction as noted above.

Surgical Technique

There are currently a multitude of different devices used for external fixation and distraction osteogenesis. The original Ilizarov fixator technique requires the use of rings or arches. It allows multiplanar fixation in small segments of bone and can incorporate hinges to correct angular deformity gradually or acutely. It allows for hinged articulation across the hip and knee joints, thus protecting these joints from subluxation during distraction without restricting joint motion. However, it is bulkier than a monolateral external fixator and forces the legs to be abducted due to the ring protruding medially.

Monolateral external fixators traditionally lacked the ability to span across joints. They have evolved to permit articulated fixation to the tibia and pelvis. They also allow locking of the hinge to prevent contracture. Deformities can also be corrected acutely. Monolateral fixators are less bulky and easier to wear for the patients. Nevertheless, they are more constrained in their pin location options and are therefore less space efficient than the circular fixators. The Orthofix LRS (Orthofix, Lewisville, TX) and the Smith & Nephew Modular Rail System (MRS, Smith & Nephew, Memphis, TN) are two examples of currently available monolateral external fixators that allow articulated spanning of joints. Their application for CFD lengthening together with the application of the Ilizarov device will be described.

In the femur, half-pin fixation is the rule, even with the circular fixator. Half-pin size is determined by the diameter of the bone, with 6.0-mm pins used for most older children and 4.5-mm pins for younger children with smaller bones. For very young children, small tapered 3.5- to 4.5-mm pins can be used. The author prefers to use hydroxyapatite-coated pins to decrease the chance of loosening and to lower the risk of pin-site infection [73].

Distal Femoral Lengthening: Ilizarov™ Fixator Technique (See Fig. 22.17)

All the acute soft tissue releases are performed first. If soft tissue releases are to be performed on a delayed basis, proceed directly with the frame application. If a second-level, proximal femoral derotation, valgus, and/or extension osteotomy is needed, the proximal pin is inserted into the proximal femur with the hip in the position in which it will lie after the correction. For example, for an internal rotation osteotomy, the proximal pin should be inserted with the knee in external rotation. For a valgus osteotomy, the proximal pin should be inserted with the hip adducted. For an extension osteotomy, the proximal pins should be inserted with the hip flexed. For correction of varus, flexion, and external rotation, the femur should be externally rotated and crossed over the other thigh to adduct and flex the hip. This places the hip in the true neutral position.

The proximal arch is attached parallel to a line from the tip of the greater trochanter to the center of the femoral head, the middle ring is perpendicular to the mechanical axis of the shaft of the femur (7° to the shaft), and the distal ring is parallel to the knee joint line. After the osteotomies, when the rings and arch are parallel, the mechanical axis of each segment will be aligned and the joint orientation of the hip and knee will be parallel. The first half-pin is from lateral to medial in the frontal plane, parallel to a line from the tip of the greater trochanter to the center of the femoral head. A second proximal half-pin is inserted on the proximal arch from 30° anterolateral to the first pin. The proximal arch is perpendicular to the floor with the leg crossed over and rotated as described above for correction of deformity.

Two Ilizarov rings, properly sized for the distal femur, are applied to a distal femoral reference wire, which is parallel to the knee joint. For young children, we obtain an arthrogram to better outline the cartilaginous femoral condylar line and visualize the posterior femoral condyles for hinge placement. Conical washers or hinges are used between the two distal rings because of the valgus of the distal femur. The rings are at a valgus deformity angle to each other. A lateral half-pin is inserted into the mid-segment of the femur. This pin is at 7° to the shaft of the bone.

At this point, the proximal subtrochanteric osteotomy can be performed. This is done percutaneously by making multiple drill holes and then using an osteotome. The osteotomy is internally rotated, laterally translated, and then angulated into valgus and extended to correct all components of the proximal femoral deformity. The order of correction is important to achieve the necessary displacement without loss of bone-to-bone contact and stability.

Two additional half-pins are inserted and fixed onto the distal ring, one from posteromedial and the other from posterolateral, between the quadriceps and the hamstring muscles. One more middle pin is inserted. In small children, all half-pins are inserted by using the cannulated drill technique. This involves insertion of a wire first, then a cannulated drill, and then a half-pin. This technique permits very accurate placement of large-diameter pins in narrow bones to avoid eccentric placement. Eccentric placement of drill holes and half-pins in the femoral diaphysis, especially anteriorly, can lead to fracture. The distal femoral osteotomy is performed percutaneously, with multiple drill holes and an osteotome. The only wire used is removed to avoid tethering of the quadriceps and fascia lata.

Knee Hinges

The last step is to extend the fixation to the tibia using hinges. The center of rotation of the knee is located at the intersection of the posterior femoral cortical line and the distal femoral physeal line in the plane where the two posterior femoral condyles are seen to overlap on the lateral view (Fig. 22.19) [42]. For younger children, it is helpful to inject arthrographic dye into the knee to visualize the posterior femoral condyles. It is important that the distal femoral ring, which is parallel to the distal femoral condyles, appears to be perpendicular to the X-ray beam. The medial and lateral skin is marked at the location of the planned hinge placement. A single half-ring is attached to two threaded rods from the hinges. This half-ring is oriented perpendicular to the tibia with the knee in full extension. The first half-pin is inserted from anterior to posterior into the tibia. After fixing this pin to the proximal tibial half-ring, the knee is flexed and extended through a range of motion. A drop test is performed to see if the tibia flexes through the hinge without catching. If this range feels



Fig. 22.19 Lateral fluoroscopic view of knee arthrogram. The center of rotation knee axis pin has been inserted at the level of distal physis and the intersection with the posterior cortex. The medial and lateral femoral posterior femoral condyle outlines can be seen because of the radiographic dye. They both overlap each other

frictionless, a second and third tibial half-pin are added. Finally, a removable knee extension bar is inserted anteriorly between the distal femoral and the tibial half-ring.

Box 22.11. The Drop Test

- A half-pin is placed in the proximal tibia from anterior to posterior.
- The half-pin is attached to the proximal tibia ring and secured.
- The tibia is raised up in full extension with the hip flexed.
- While supporting the femur, the tibia is released to flex with gravity.
- A positive drop test and a well-placed knee hinge allow the tibia to flex to 90° without catching.
- If there is significant friction, make adjustments to the half-pin.
- Securing the half-pin with the knee in full flexion or full extension or 45° may improve the drop test.
- Do not add any more pins until the drop test demonstrates no friction.
- The success of the drop test depends on the initial accurate placement of the hinge.

Perform the drop test after each pin is added. Use a total of three pins in the tibia.

Distal Femoral Lengthening: Orthofix™ Fixator Technique (See Fig. 22.17)

1. Start by identifying the center of rotation axis of the knee joint (see description of this above). A 1.8-mm wire is drilled into the lateral edge of the physis at the intersection of the posterior cortex of the femur with the physis in line with the plane of overlap of the posterior femoral condyles. The Orthofix LRS comes in pediatric and adult sizes. The hinge axis is lined up through its most distal hole.
2. A commercially available "sandwich" clamp is used, or an extra lid is used in the pin clamp to create a second layer of pin holes more anteriorly. The fixator bar is lined up with the shaft of the femur and the most proximal half-pin inserted.
3. The most distal pin is then drilled one hole proximal to the center of rotation pin. The LRS without sandwich clamps is then used to place the rest of the pins, three proximal and three distal. If the distal femoral valgus is to be corrected acutely, a swivel clamp is used for distal pin placement. When using a pediatric LRS, the three-hole pin clamp contains two half-pins only, since one hole is used up for the knee center of rotation. A third pin is added using Ilizarov cubes connected via these two pins.
4. Before reapplying the sandwich clamps, the osteotomy is performed and the distal valgus corrected acutely. After the correction, the fixator can be exchanged for one with straight clamps and with the sandwich attachments. All the pins should be in the upper deck of the double-decker sandwich clamps. The only pin in the lower deck is the knee axis pin. This pin does not enter the patient's leg. It is a 6-mm segment of pin that protrudes laterally. A Sheffield clamp from Orthofix™ is applied to this pin to act as a surrogate knee hinge. It is locked in place by putting a Rancho™ cube laterally with a set screw to prevent it from moving outward. Conical washers are used between the Sheffield clamp and the LRS to prevent friction. The Sheffield clamp is left partially loose to permit motion.
5. A 1/3 Sheffield arch is attached to the clamp arching towards the tibia. An anteroposterior pin is inserted and the drop test (see above) is performed. If there is friction, the Sheffield clamp should be loosened. If friction persists adjust the connection of the pin to the Sheffield arch. If friction persists the axis pin may need to be bent slightly to alter the axis of rotation. Once the drop test is negative for friction, two more oblique pins are inserted into the tibia and connected to the Sheffield arch using cubes.
6. A removable knee extension bar is fashioned from Ilizarov parts to be used especially at nighttime. If there is an unstable hip, an axis pin for the hip can also be fashioned and attached from the proximal clamp. The same Sheffield clamp and arch arrangement are used.

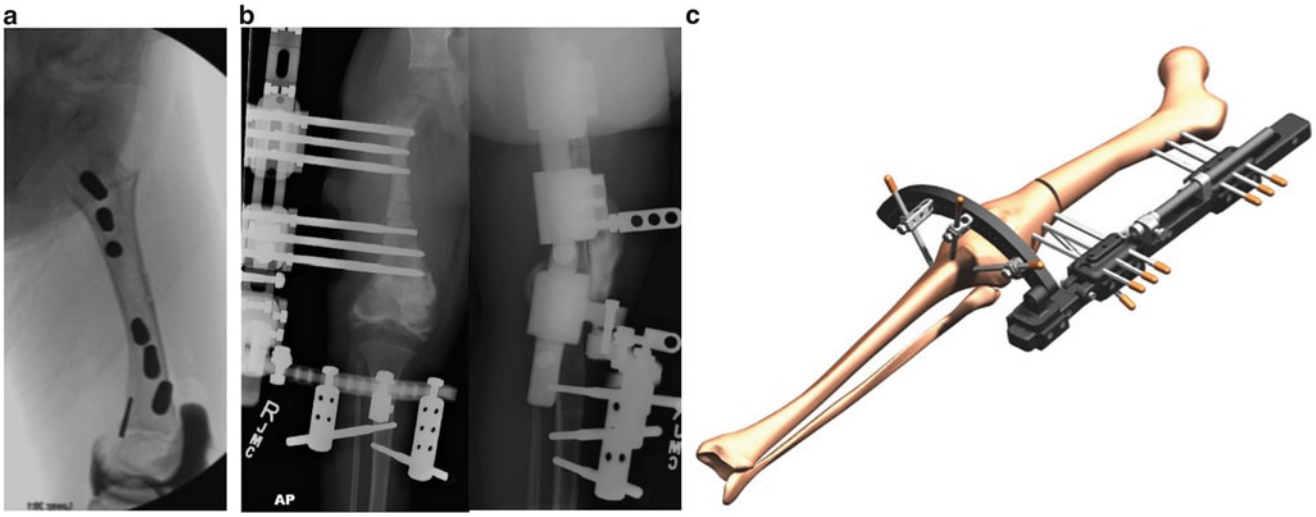


Fig. 22.20 Modular Rail System (MRS, Smith & Nephew, Memphis, TN) technique. (**a**) The external fixation pin is inserted parallel to the knee axis pin. The external fixator is used as a template. (**b**) AP and lateral radiographs showing the external fixator in place and the articulated spanning across to the tibia. The arthrographic dye is in place. The fixator hinge line corresponds to the knee hinge center of rotation axis. (**c**) Illustration of the Modular Rail System external fixator articulated across the knee joint. Note location of knee hinge

Two pins are placed in the pelvis from the anterior inferior and superior spines extending posteriorly. These are fixed to the Sheffield clamp to prevent proximal subluxation of the hip during lengthening. Thus, the same principles are applied when using monolateral as with circular fixation, i.e., hinge fixation across joints when there is a joint at risk.

Distal Femoral Lengthening: Modular Rail System (Smith & Nephew, Memphis) Technique (Figs. 22.9, 22.10, and 22.20)

This external fixator was designed specifically for the CFD patient. It is able to articulate and span across the hip and knee joint either separately or at the same time. It also has clamps that allow fixation in the oblique plane in addition to the frontal plane, producing delta fixation.

1. Pre-construct the external fixator using pediatric or adult rail segments with two pin clamps, knee hinge, and tibial rail segment.
2. Inject radio-contrast solution into the knee joint and obtain a true lateral projection, rotating the femur until the posterior aspect of the medial and lateral femoral condyles overlap.
3. Insert a 2-mm Steinmann pin into the center of rotation of the knee joint. This is defined as the point of intersection of the posterior femoral cortex with the distal femoral physis. In adults, it is the intersection of the posterior cortex of the femur with Blumensaat's line. Confirm on the AP view that the pin is parallel to the joint line.
4. If there is no valgus or flexion deformity, proceed to drilling a 1.8-mm wire into the proximal femur parallel to the Steinmann pin. This wire should be as proximal as possible in the femur but distal to the trochanteric apophysis. Confirm the location of this wire making sure that it is located in the mid-diaphysis or slightly posterior to the midline. Drill a hole using a cannulated drill appropriate to the pin size preferred (e.g., 4.8 mm for a 6-mm pin, 3.8 mm for a 4.5-mm pin, and 3.2 mm for a 3.5- to 4.5-mm pin).
5. Insert a pin of the appropriate thread length corresponding to the diameter of the bone at that level. The bending strength of the pin is related to the smallest diameter protruding from the near cortex. It is better not to leave threads outside of the near cortex to obtain the maximum bending strength out of each pin.
6. Mount the pre-constructed MRS onto the Steinmann pin and the proximal half-pin. The Steinmann pin goes through the cannulated hinge of the fixator.
7. Drill a 1.8-mm wire through the distal most pin hole of the clamp. An overhang clamp can be used to minimize the distance from the most distal pin to the hinge. This pin should be just proximal to the physis. Overdrill the wire with a cannulated drill and insert the pin.
8. Insert the other two pins into the clamp, making sure that the holes line up with the bone. All the pins should be parallel to each other. Make sure that the distal pin clamp is locked down to the fixator with 7-mm red bolts. Slide off the fixator from the pins.
9. Make a 7-mm incision just proximal to the most proximal distal pin. Drill the femur at this level with multiple drill holes. Complete the osteotomy using a sharp osteotome.
10. Reapply the fixator to the pins. Make sure that the osteotomy does not displace or translate, making

adjustments as needed. If AP clamps are being used, drill at least one hole for an oblique posterolateral proximally and distally. There should be three or four total number of pins per segment, depending on the size of the femur and the thigh being lengthened.

11. Drill and place an anterior to posterior pin in the proximal tibia, making sure that it is distal to the tip of the proximal tibial apophysis. Connect the tibial pin to an arch mounted off of the tibial rail extension using a cube. Test the motion of the knee to confirm that the knee moves freely. By placing the first tibial pin in the sagittal plane, anteroposterior adjustment can be done if there is subluxation. Apply a set screw to the pin once the knee motion is free. Confirm with a drop test: if the leg drops with no friction from 0° to 90°, then the hinge is perfect. Apply two more pins with a different orientation in the proximal tibia.
12. Apply a distractor bar between the two clamps, making sure that it turns freely and that the osteotomy is distracting appropriately. The bar distracts 1 mm for every full turn of 360°.
13. Apply a knee extension bar constructed from Ilizarov parts. This includes a post proximally and a twisted hinge distally. Off of each of these, insert a post and fix with a 20- or 30-mm socket. Connect a threaded rod between the two posts. The knee extension bar should be placed in maximum extension of the knee and locked down so that the rod length cannot be changed. The patient uses the bar full time at night and half time during the day. It is easily removable for exercise.
14. Incisions are closed and sterile dressings applied around the pins. Consider Botox to the quadriceps muscles to diminish muscle spasm. Final radiographs are taken prior to leaving the operating room.

Box 22.12. Lengthening Tips and Tricks

- A knee arthrogram is helpful in young children to visualize the posterior condyles or center of rotation.
- The first half-pin or the hinge pin sets up the rest of the pins. Spend extra time getting the first one perfectly right, and the rest will follow.
- For smaller bones, use the cannulated wire technique to ensure proper placement of pins. Eccentric cortical drilling can lead to fractures or pin pullout.
- Use off-axis pins for stronger fixation if possible.

Make sure that the osteotomy is completed and distracts appropriately without too much resistance before leaving the operating room.

Modification for Valgus ± Flexion Deformity of the Distal Femur

If there is a valgus or flexion deformity of the distal femur, changing the pin placement will provide correction. The first wire is placed into the distal femur, using the MRS as a guide to keep the wire parallel to the knee hinge pin. A cannulated drill is then used and a half-pin is inserted. The most proximal pin is then placed into the proximal femur at about 7° of valgus in relation to the shaft. In reference to the distal pin, it should be in valgus angulation and in the same rotational orientation as the distal pin. A distal femoral osteotomy is performed at the distal femur at the planned level, leaving enough room to add two more pins distal to the osteotomy. Once the pins are connected to the external frame and parallel to each other, the valgus deformity is corrected. If there is also a flexion deformity of the knee, extend the knee with the tibia to level the distal segment into extension before adding the remaining frontal plane pins.

Rehabilitation and Follow-Up During Lengthening

Femoral lengthening requires close follow-up and intensive rehabilitation to identify problems and maintain a functional extremity. Clinically, the patient is assessed for hip and knee range of motion, nerve function, and pin-site problems. Radiographically, the distraction gap length, regenerate bone quality, limb alignment, and joint location are assessed. If the lengthening rate is 1 mm per day, then follow-up is usually every 2 weeks. If there is delayed healing of the regenerate bone, or if knee motion is decreasing, then the rate is often decreased. If the lengthening rate is decreased to 0.75 mm or 0.5 mm per day then the follow-up can be increased to 3 or 4 weeks, respectively. Rarely, if there is concern for premature consolidation and the lengthening rate is increased, then follow-up is more frequent.

Patients are allowed to weight-bear as tolerated with their external fixator. Physical therapy is begun within 1 or 2 days after surgery and continues daily throughout the distraction and consolidation phase. In the author's opinion, the amount of therapy is directly related to a better functional result and faster rehabilitation after removal. During distraction, patients have one to two formal physical therapy sessions daily (45–60 min each). In addition, at least two home sessions (30 min each) are recommended to the parent. Patients undergoing lengthening are at their best immediately after surgery and have difficulty with muscle tightness and range of motion only after lengthening has started. It is not until the consolidation phase that the usual orthopedic pattern of rehabilitation and recovery occurs. Thus, lengthening should be thought of as a

prolonged procedure that lasts until the end of the distraction period, and not even be considered in the absence of therapy.

The majority of therapy time is spent obtaining knee flexion and maintaining knee extension. Knee flexion should be maintained as close to 90° as possible. It is important to start pushing the extremes of passive knee flexion (especially prone) during the 1-week latency period after surgery. Once distraction begins, there is more resistance from muscle spasm. If knee flexion begins to diminish (<75°), then lengthening should be stopped or slowed to rehabilitate the knee better, sometimes waiting for improvement before resuming lengthening. Function should never be sacrificed for length, and preserving the knee joint and motion is paramount. A knee extension bar is used at night and part-time during the day to prevent a knee flexion contracture [42]. Posterior subluxation can occur with a fixed flexion deformity and is suspected clinically based on a change in the shape of the front of the knee. The patella becomes very prominent and the tibia appears depressed (ski hill sign).

Weight bearing is encouraged throughout the lengthening as long as the patient has normal proprioception. In younger children, walking without support can be achieved immediately because the fixator is very stiff relative to the patient's weight. When the child is older and heavier, crutches or a walker is preferred until there is sufficient consolidation.

During the distraction phase, therapy is focused on passive range of motion exercises to maintain hip and knee range of motion. During the consolidation phase, the priority shifts to active range of motion and strengthening to regain any motion lost during distraction. Hip abduction and extension exercises help prevent adduction and flexion contractures that can lead to hip subluxation. Release of the adductors, rectus femoris, and TFL during lengthening may need to be considered to allow further lengthening and to prevent hip subluxation.

After the lengthening regenerate is healed and the external fixator removed, recovery is limited by two factors: joint range of motion and muscle weakness. In most cases, joint motion is regained faster than muscle strength. The three antigravity muscles are the limiting factor to full recovery: the hip abductors, quadriceps, and gastro-soleus muscles. To minimize disability, these muscles should have resistance training throughout the lengthening program to prevent atrophy from prolonged external fixation and lengthening. Other modalities such as electrical stimulation or aquatic therapy may be helpful. Please also refer to Chap. 12 for more details regarding physical therapy in such patients.

Fixator Removal and Rodding of Femur (See Figs. 22.9 and 22.10)

The external fixator can be removed once the regenerate bone is fully healed radiographically. The biggest risk after removal is fracture of the femur, which can manifest as painless bending of the femur or as a completely displaced fracture. The use of a spica cast does not reduce the fracture rate. A study, performed by the author in 1999, showed a fracture rate of 34 % after CFD lengthening, compared to 9 % for other etiologies [84]. The regenerate bone should show no gaps, the interzone should be closed, and based on AP and lateral views there should be evidence of corticalization on at least three sides of the lengthened regenerate. Since this refracture rate was so high and unacceptable, Paley started prophylactic rodding of the femur with a Rush rod in 2000. The fracture rate dropped to almost zero in the senior author's personal experience and somewhat higher in a multi-surgeon series [53]. In the senior author's personal series, the rate of infection after such prophylactic rodding was 4 %. These were easy to eliminate by removal of the rod and insertion of an antibiotic cement rod [74]. This was considered a reasonable trade-off: trading 34 % fracture rate for 4 % infection rate.

Surgical Technique for Prophylactic Rodding of Femur at Time of External Fixator Removal

1. The external fixator is removed under general anesthesia, ensuring that the patient is deeply asleep (to avoid involuntary muscle spasms which could lead to fracture of the regenerate at the time of removal). Slightly loosen all of the pins while they are still supported by the frame, which is especially important in hydroxyapatite-coated pins. Remove the middle pins while the fixator is still on. The most distal and proximal pins are removed while an assistant provides bimanual support of the thigh above and below the lengthening zone.
2. After prepping the thigh, curettage any loose granulation tissue from the pin holes. The pin holes in the bone are also curetted in the event of osteolysis or infection. Cover and seal the pin holes with an adherent plastic dressing (e.g., Tegaderm, 3 M, Minneapolis, MN). In the event of a severe infection, the surgeon must weigh risks of a fracture and loss of length versus hardware infection and secondary surgeries.
3. Prep and drape the lower limb free, being careful not to manipulate the leg. A bump may be placed under the ischium to raise the femur for lateral viewing on image-intensifier radiography.
4. Insert a 1.8-mm bayonet-tipped Ilizarov wire at the tip of the greater trochanter. Advance the wire by tapping, rotating the curved tip of the wire to bounce off the inner cortex. Use the drill only if the wire will not advance. A slight bend in the wire may be necessary to direct the wire. The wire is passed through the center of the intramedullary canal and regenerate bone, all the way to the distal femoral physis if possible. A second wire of

equal length can help measure the length by inserting it to the greater trochanter and measuring the difference in length of the two wires.

5. Drill over the wire with a cannulated drill bit. The 3.2-mm, 4.8-mm, and 6.4-mm drill bits correspond to the 1/8″, 3/16″, and 1/4″ Rush rods, respectively. Long cannulated drills are available from Pega Medical (Montreal, Canada) and are preferred since they slightly overdrill above the designated size making rod insertion easier. They are also longer and can reach the full length of almost any femur.
6. Insert an appropriate length Rush rod. As an alternative, a stainless steel flexible nail can be used instead, but they are more difficult to insert and direct. The end of the Rush rod is better designed for directing the implant tip. Recently, a lockable rod (the SLIM, Pega Medical, Montreal, Canada) has been developed for this purpose with a tip similar to the Rush rod but with a threaded proximal part similar to the Fassier-Duval nail (Pega Medical, Montreal, Canada).
7. After rodding, the patient goes home with sterile, dry dressings covering the pin sites and a 10-day course of oral antibiotics, usually a first-generation cephalosporin. Weight bearing and physical therapy are restricted for 1 month. Gentle range of motion of the knee is allowed at home.

Box 22.13. Fixator Removal Tips and Tricks

- Do not remove fixator until three sides have corticalized.
- Use a rush rod or intramedullary device to prevent fracture through the regenerate bone.
- Slightly turn or "crack" all the half-pins while in the fixator prior to removing the frame. The hydroxyapatite bond can be quite strong.
- Curettage out any hypertrophic granulation tissue. Loose pins warrant more thorough curettage of the bone.
- It may not be possible to direct a thin Ilizarov wire all the way down the femur. Drill just the proximal portion and then proceed to the Rush rod.
- If needed, put a small bend in the Rush rod in line with the bevel to augment guided insertion.

Hold off on physical therapy for a month after fixator removal.

Specific Complications and Their Treatment for Congenital Femoral Deficiency Lengthening

Nerve Injury

Nerve injury from surgery or distraction is unusual with femoral lengthening. To avoid peroneal nerve injury from the pins, the posterolateral pin should not enter posterior to the biceps tendon. During distraction, if the patient complains of pain in the dorsum of the foot or asks for frequent massage of the foot, this is most likely referred pain from stretch entrapment of the peroneal nerve. More advanced symptoms include dysesthesia in the distribution of the peroneal nerve or extensor hallucis longus weakness. A nerve conduction study may show evidence of nerve injury, but most cases will be negative since too many fibers are conducting normally. Quantitative sensory testing using the pressure-sensitive sensory device (PSSD), if available, is the most sensitive test to assess for nerve involvement [75]. Near-nerve conduction using very fine electrodes is also very accurate.

If a nerve problem is identified early, it can be treated by slowing the rate of distraction. However, if symptoms persist or motor signs develop, then peroneal nerve decompression should be performed [76, 77]. The timing of nerve decompression affects the rate of recovery of neuropraxia, and an early decompression leads to faster recovery. The peroneal nerve should be decompressed at the neck of the fibula, including transverse fasciotomy of the anterior and lateral compartment and release of the intermuscular septum between these compartments [68] (see Fig. 22.16). When peroneal nerve decompression was performed on patients undergoing limb lengthening, intraoperative findings included hemorrhage, nerve flattening, narrowing of the nerve at the entrance of the fascial tunnel, and reduction of the perineural vascularization at the site of compression. These findings are typical of nerve entrapment and not of stretch injury. In addition, no relationship was found between nerve injury and the amount or percent of lengthening.

Poor or Failed Bone Formation

Hypotrophic regenerate formation requires slowing of the distraction rate. The rate can be slowed to ¾, ½, or ¼ mm per day. If the regenerate bone does not improve, a decision needs to be made to continue lengthening. If the defect in the distraction gap does not fill, it will need bone grafting. Bisphosphonate infusion (e.g., zoledronic acid) can be used to prevent bone resorption while permitting bone formation [78, 79].

Complete failure of bone formation is very unusual. Partial defects, especially laterally, are common. Dynamization of the fixator and bone growth stimulators (e.g., Exogen™) can be used. Resection of the fibrous tissue in these defects and cancellous bone grafting may become necessary to reduce the external fixation time and prevent fracture after frame removal.

There are two methods of bone grafting the regenerate:

1. *Autogenous cancellous bone grafting*. The distraction gap should be opened and the fibrous tissue excised, leaving behind any regenerate bone. Once all of the fibrous tissue has been removed, cancellous autogenous bone graft can be harvested and inserted. In most children, this is obtained from the iliac crest. In young children there may not be enough bone from the standard anterior iliac crest approach. More bone graft can be obtained by splitting the tables of the iliac crest and taking graft from above the acetabulum. This allows harvesting of a large cache of bone that is normally not available for grafting, providing enough bone to fill the distraction gap.
2. *Cortical allograft*. This technique is used when the distraction gap is very long and exceeds the available autogenous cancellous bone graft. The technique is credited to Wasserstein [80], originally from Latvia and later from Germany. He described inserting a slotted allograft into a distraction gap to create early structure and bridging of the gap. A non-slotted allograft can be used instead, stabilized with an intramedullary nail. BMP-2 can be added for both cancellous and cortical grafts.

Incomplete Osteotomy and Premature Consolidation

If there is a lack of separation of the osteotomy site after a week of distraction, it may be due to an incomplete osteotomy or a periosteal hinge that will not separate. Continued distraction can lead to an acute separation of the bone ends, which is usually very painful and may have an audible pop. The pain will continue unabated until the bone is acutely shortened by a few millimeters. It is important to advise the patient of this possibility. If the bone does not separate, or if the patient or parents wish to avoid a painful separation, a repeat osteotomy at the same site should be performed.

Overabundant bone formation may lead to premature consolidation. Watch for a mismatch between the distraction performed and the radiographic distraction. Increasing the distraction rate for a few days may prevent a premature consolidation in such cases. Patients should be checked weekly during this time. If premature consolidation occurs, a repeat osteotomy should be performed at a new location, as a repeat osteotomy through the original site is more likely to lead to failure of bone formation. Leave the fixator in tension, as the osteotomy will separate easier when cut under tension. After the osteotomy, the tension in the frame should be reduced to normal.

Hip Subluxation/Dislocation

There is often mild or moderate acetabular dysplasia in patients with CFD. The pattern is different than DDH; the femoral head is usually uncovered laterally rather than anterolaterally. If the CE angle is less than 20°, the hip joint is considered at risk for dislocation [59]. The orientation of the sourcil should also be horizontal. If it is inclined superiorly, then the hip joint is potentially unstable even with a normal CE angle. It is always safer to err on the side of performing a pelvic osteotomy prior to femoral lengthening than to end up with a hip subluxation.

Hip joint subluxation or dislocation during lengthening is a dreaded complication. It is diagnosed radiographically. The earliest sign is a break in Shenton's line or increased medial head-teardrop distance indicates subluxation of the hip. The hip usually has an adduction and flexion contracture and may also exhibit stiffness with flexion and extension. Hip subluxation does not usually occur if there is adequate coverage of the femoral head, but they can occur if there are adduction and flexion contractures.

If hip subluxation occurs, distraction must stop. The patient should be taken to the operating room to reduce the hip. Release the adductor longus and gracilis, and the TFL and rectus femoris if needed. Abduction should reduce the hip subluxation. If not, the distraction gap should be shortened to loosen the hip joint. The external fixator should be extended to the pelvis with or without a flexion-extension hinge. Pelvic fixation consists of at least two anterolateral pins between the two tables above the anterior inferior iliac spine followed by two off-axis lateral pins. The femur should be in 15–20° of abduction to the pelvis to maintain the reduction. A hip extension bar should be added to prevent flexion contracture with a hinge. Lengthening may resume with caution (Figs. 22.21, 22.22, and 22.23).

If a pelvic osteotomy is performed after lengthening, care must be taken not to force the hip into the acetabulum. Since the femoral head is osteoporotic, it can be easily crushed with attempts at reduction. If an acute reduction is quite difficult, then it may be safer to do a gradual reduction with the external fixation if the external fixator is still in place. A hip dislocation that occurs after frame removal should be treated with an open reduction, capsulorrhaphy, and femoral shortening and pelvic osteotomy. The femur should be osteotomized after the capsulotomy and the femoral head reduced into joint. The bone ends of the femur are overlapped to determine the amount of shortening required. It is essential to shorten. There must be no tension on the femoral head if it is to remain in the acetabulum. It is important to preserve the superior capsule; if released, it will lead to recurrent dislocation. If the hip is difficult to stabilize despite all of these measures, it can be tethered in place using a suture anchor in the cotyloid notch, with the suture passing through the fovea and out the lateral neck. This creates a mobile tether that

prevents dislocation while limiting motion. This technique was developed by the senior author and has been used since 2002 (see Fig. 22.23).

Knee Subluxation/Dislocation

Most cases of CFD usually have hypoplastic or absent cruciate ligaments in the knee. The tendency towards flexion contracture during lengthening predisposes the tibia to posterior subluxation, which is the most common. Posterolateral or external rotatory subluxation may occur with a tight fascia lata and biceps. Anterior or anteromedial subluxation can result from extension contractures and patella alta.

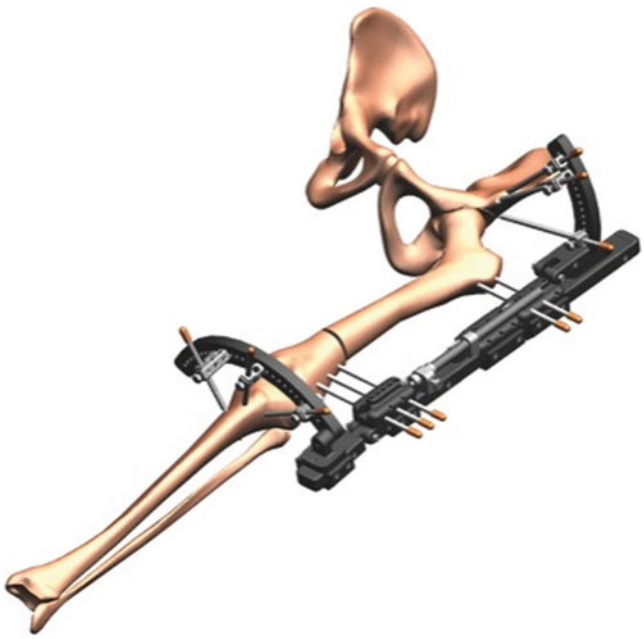


Fig. 22.21 The MRS articulated across both the hip and the knee with fixation to the pelvis and tibia, respectively. Both the knee and hip external fixation hinge axes correspond to the knee and hip center of rotation axes in the frontal plane

A more proximal femoral lengthening is less likely to cause knee subluxation, but it often has narrower and less well-formed regenerate bone, leading to higher fracture risk.

To prevent knee subluxation, the distal fascia lata should be released and the external fixator should span the knee and include the tibia. If the fascia is not released, then the knee can begin to subluxate after only 2–3 cm of lengthening. Without tibia fixation, the knee will often start to subluxate after 4 cm of lengthening. The spanning fixator also protects the knee cartilage and physis from increased forces. An articulated hinge can be used to allow the patient to maintain knee range of motion. Subluxation may be prevented by keeping the knee in full extension throughout lengthening, but it will result in a stiff knee. In the knee, posterior or anterior subluxation can be monitored on a lateral knee radiograph in full extension [81]. If the knee begins to subluxate, the rate of lengthening may need to be decreased as well as more intensive physical therapy for stretching.

If the knee subluxates or dislocates after removal of the external fixator, it should initially be treated by aggressive physical therapy, traction, and splinting. Physical therapy measures include pulling the knee forward and rotating it inwards during knee extension and flexion range of motion. If the knee remains subluxated despite an adequate trial of several months of therapy, it requires a version of the Superknee procedure called a "rescue knee." This involves lengthening of the biceps femoris and the iliotibial band if it is intact. In some cases, the Langenskiöld is done to reduce the patella into joint. If there is a fixed flexion deformity, the femur must be shortened proximally to avoid creating a quadriceps lag. In the most severe cases, a posterior capsulotomy is required. If the knee also has an extension contracture, this can be combined with a Judet quadricepsplasty (Fig. 22.24).

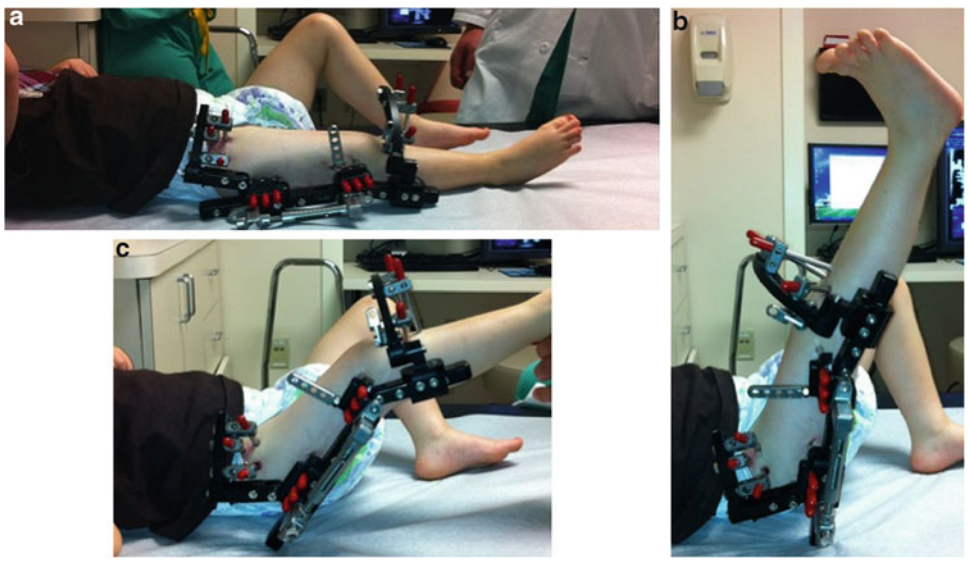


Fig. 22.22 MRS device in place on a 2-year-old boy being lengthened for CFD. The external fixator spans the hip and knee with a hinge connection to the femoral rail. This construct is shown with the hip and knee in extension (**a**), hip flexion (**b**), and hip and knee flexion (**c**)

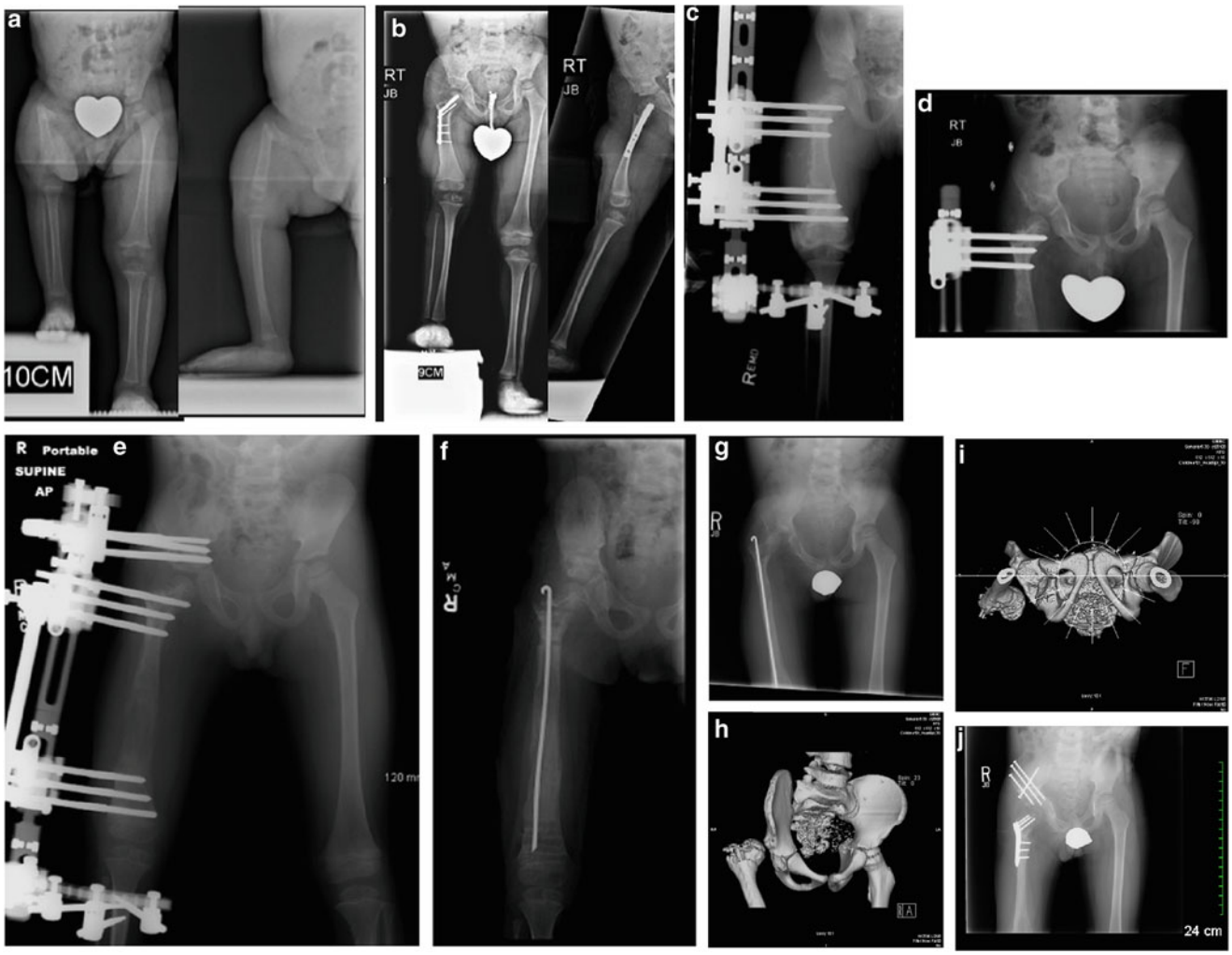


Fig. 22.23 Radiographs of a 2-year-old boy with CFD Paley type 1b subtrochanteric type deformity (**a**). Following Superhip and Superknee reconstruction including Dega pelvic osteotomy (**b**) at age 2. Lengthening of the femur at age 3 years, using the MRS device articulated across to the tibia (**c**). Radiograph after 4 cm of lengthening demonstrating early lateral subluxation of the hip. Note the increased femoral head-tear drop space (**d**). Extension of the external fixator to the pelvis with articulated spanning fixation (**e**). The femoral head is well reduced and stabilized permitting continued lengthening up to 8 cm. Radiograph at the time of removal of the external fixator with Rush rodding to prevent fracture. Note that the hip appears well reduced and stable (**f**). Radiograph 3 months later showing dislocation of the hip joint (**g**). Three-dimensional CT scan showing that the femoral head is dislocated posteriorly (**h**, **i**). Note on the axial view (**i**) that the posterior wall of the acetabulum is hypoplastic. Thus, the acetabulum is retroverted despite excellent superior and lateral coverage from the previous Dega. This confirms the statement that the Dega osteotomy cannot improve the posterior coverage of the hip. A "rescue hip" procedure was performed. The femur was shortened 2 cm and fixed with a plate. An open reduction of the hip was performed. To tether the femoral head into the acetabulum a suture anchor was used from the cotyloid notch through the femoral head and secured over a washer laterally. This washer can just be seen protruding at the upper end of the plate. A periacetabular triple osteotomy (according to Paley's modification, Fig. 22.12) was performed and secured with screws. The hip remains stable and mobile. Illustration of the open reduction, shortening, and tethering technique (**k**–**t**). After the pelvic osteotomy, capsulotomy, and subtrochanteric osteotomy the femoral head can be reduced into joint. The quadrilateral plate is drilled (**k**). The perforation seen from the inside of the pelvis (**l**). The femoral head is drilled through the fovea (**m**). After drilling both the femur and the acetabulum (**n**). The suture anchor is introduced into the medial wall of the acetabulum (**o**). The author's current preference is to use the Juggerknot (Biomet, Warsaw, IN). This has a soft anchor made of nonresorbable suture material that bunches up when pulled back through the hole (**p**) locking on the inside of the quadrilateral plate of the pelvis. A Keith needle is used to pull the suture through the femoral head and neck and greater trochanter (**q**, **r**). The suture is tied over a washer to secure the reduction of the femoral head. The hip remains mobile but not dislocatable. The distal femur can be overlapped on the proximal femur to determine how much shortening is required (**s**). The distal femur is shortened and fixed to the plate

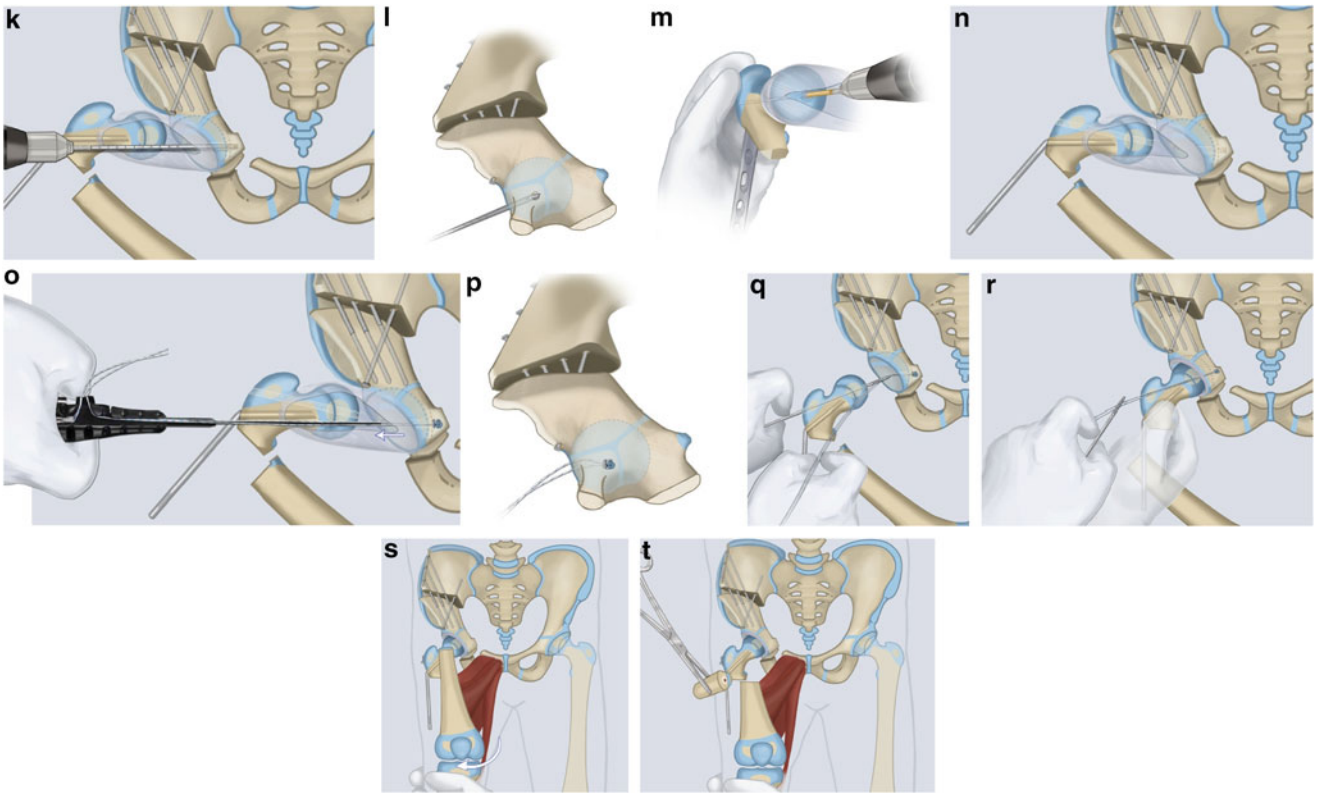


Fig. 22.23 (continued)

Limb Malalignment

Limb length equalization should be based on full-length standing radiographs. Limb alignment is assessed for the femur and tibia both separately and in combination. The joint orientation of the knee should be measured using the malalignment test [82].

Axial deviation from lengthening (procurvatum and valgus for distal femoral lengthening and procurvatum and varus for proximal lengthening) is identified and corrected at the end of the distraction phase, when the regenerate bone is still malleable. When there is malalignment of the femur and tibia, the femoral malalignment is corrected to a normal distal femoral joint orientation. The femur is not over- or under-corrected to compensate for the tibial deformity. The tibia should be corrected separately, either during the same treatment or at a later treatment. Residual malalignment after the external fixator is removed and can usually be addressed with hemiepiphysiodesis or repeat osteotomy.

Fractures

Fractures associated with limb lengthening can occur throughout and after the lengthening process. Fractures can occur through pin holes or be pathologic secondary to disuse osteoporosis. A fall on a frame can cause a femoral neck fracture or a pin hole fracture, usually the second most proximal pin. Physical therapy manipulation can cause flexion buckle fractures of the proximal tibia or distal femur. An unexpected muscle spasm during frame removal can cause flexion failure of the femur through a pin hole or the regenerate. After frame removal, fractures can occur through pin holes or the regenerate bone, usually at the mid-regenerate area or host bone junction. The incidence of all these type of fractures associated with CFD lengthening was 34 % compared to 9 % for non-congenital femoral lengthening [83–85]. In many cases, this was despite the use of a spica cast after removal.

By prophylactically rodding the femur at the time of removal, the risk of refracture is much lower [53]. There is still a concern for intramedullary infection, so proper irrigation, debridement, and curettage before nail insertion are warranted. Patients are given intraoperative antibiotics and a 10-day course of oral antibiotics.

Prophylactic rodding permits continuation of knee mobility after removal. To protect from osteoporotic stress fractures through the tibial pin holes we hold formal physical therapy for a month but permit the patient gentle range of knee motion. Rodding also permits weight bearing with a removable spica cast right after frame removal. Fractures may still occur after rodding if the rod is too small or too short. The appropriate diameter rod should be used in each case. In most small children the 1/8″ (3.1 mm) Rush rod is used. In larger children the 3/16″ (4.65 mm) rod is preferable and in older children the ¼″ (6.2 mm) Rush rod is chosen. In general, the Rush rod is passed as distal as possible to the edge of the distal femoral physis.

Alternatively, a prophylactic submuscular plate can be used when the pin sites are too infected to allow safe rodding (Fig. 22.25).



Fig. 22.24 Thirteen-year-old girl with CFD undergoing lengthening over nail using Orthofix LRS device articulated across to the tibia to prevent knee subluxation (**a**). After the nail was locked and the fixator removed, the knee dislocated by an external rotatory subluxation including dislocation of the patella. Note that this radiograph is a true AP of the femur but an oblique of the externally rotated tibia (**b**). This radiograph is a true AP of the tibia but an oblique of the femur, which is relatively internally rotated to the tibia (**c**). AP and lateral radiographs of the knee after a Superknee procedure was performed to "rescue" the knee. AP and lateral radiographs of the knee (**d**) after a Superknee procedure was performed including the Paley reverse MacIntosh procedure, which in this case was secured with two medial suture anchors in the medial femoral condyle. Because she was skeletally mature an Elmslie-Trillat transfer of the tuberosity was performed instead of a Grammont. Note that the tibia is congruently reduced to the femur. The patella is back in place using the Langenskiöld procedure. This surgery was performed by the author in 1994. The patient maintains excellent stable and painless range of motion to the present date (**e**, **f**)

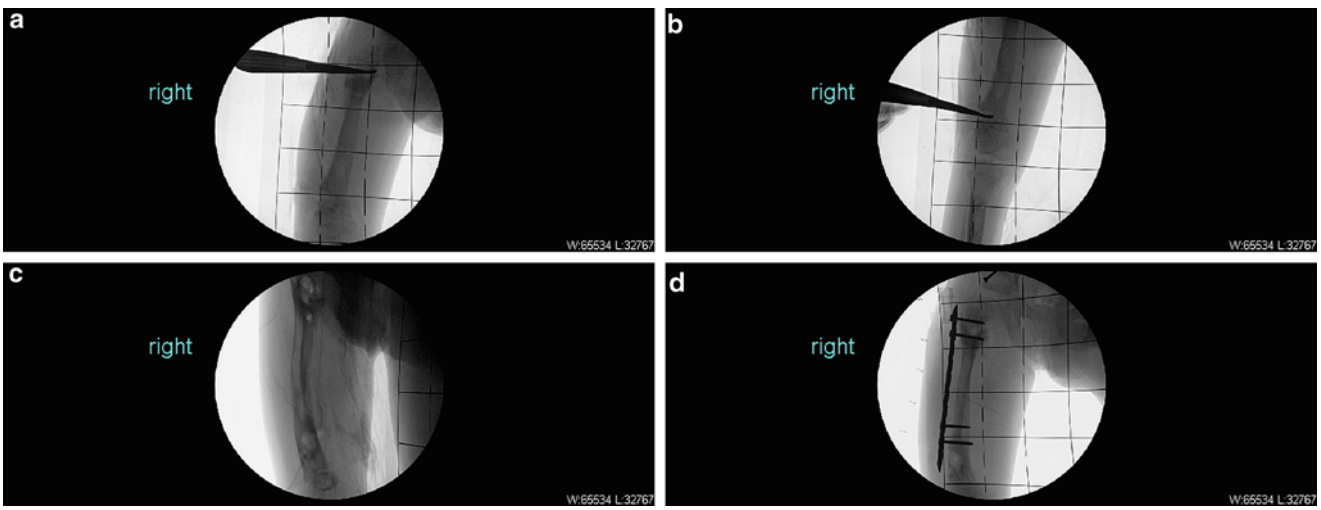


Fig. 22.25 Significant pin-site osteolysis after 8-cm femoral lengthening in a 3-year-old boy after previous Superhip and Superknee procedures. The pin sites are debrided including curettage of the bone, at the time of removal of the external fixator (**a**, **b**). Lateral radiograph showing the holes in the bone that make it at high risk of fracture. Due to the contamination of the bone by the pins the risk of intramedullary infection with Rush rodding was considered to be too high in this case. To prevent fracture a submuscular locking plate was used instead. This avoids contact with the pin sites

Joint Stiffness and Contracture

The knee can develop both flexion and extension contractures during lengthening. Although transfixing pins or wires may contribute to difficulty in flexion, even lengthening with internal distractors may still lead to loss of knee flexion with increased length. However, stiffness of the knee is preventable. Surgical release and lengthening of specific soft tissues (fascia lata, rectus femoris) reduce the joint reactive forces on the knee due to lengthening. They can be done acutely with the index procedure, but the soft tissues may heal together prior to the end of distraction. A delayed release at about 6 weeks allows more soft tissue distraction since the release occurs when the soft tissues are taut from lengthening.

Dynamic splinting can be done by using commercially available products or office-customized splints such as developed by Bhave [86]. The custom knee device consists of two fiberglass casting material cuffs for the thigh and lower leg connected by cast brace hinges. The elastic forces are applied using elastic bandages anteriorly off of various towers suspended proximal and distal to the knee. This is a very efficient and inexpensive type of dynamic splinting.

Physical therapy is essential to successful CFD lengthening. This is especially true for the knee, and one should not consider lengthening a CFD case without outpatient PT. If the knee gets stiff in flexion despite adequate rehabilitation, then a quadricepsplasty should be performed. Rozbruch recommends doing this at or shortly after frame removal [87]. He cuts the central tendon of the quadriceps while leaving the medial and lateral musculature intact. He claims that this does not lead to a quadriceps lag. This author prefers to do a full Judet quadricepsplasty [88] instead. With a concomitant flexion contracture, either an external fixator with gradual distraction or an open posterior capsule release can be performed.

Difference in Treatment of Types 1a and 1b

In general, Paley CFD type 1a (normal ossification) has less deformity, deficiency, and discrepancy in the hip and knee, in comparison to type 1b (delayed ossification). The distinction between types 1a and 1b should be made while the patient is in infancy, because the natural history of type 1b is to ossify. Therefore, adult type 1b cases may appear to be severe type 1a cases. Most type 1a cases do not require the complex Superhip reconstruction. In the author's experience, approximately half do require pelvic osteotomy before lengthening, and all cases should have extension of the fixator across the knee to protect the knee joint.

The strategy of treatment for type 1b is to correct all the associated deformities, which will allow the proximal femur and hip to be more normally oriented and accept more axial loading. The response to the anatomic change is ossification of the proximal femur, converting the CFD from type 1b to type 1a. This conversion usually occurs within 2 years of the Superhip procedure, and no lengthening is attempted until conversion. The first lengthening can be performed between the ages of 2 and 4, once the femur is of adequate size. If initially seen in infancy, patients with type 1a CFD typically undergo their first lengthening at age 2 years, whereas patients with type 1b CFD typically undergo lengthening closer to age 4 years.

Treatment of Paley CFD Type 2 (Fig. 22.26)

In comparison to the Paley type 1b neck type, which is sometimes referred to as a stiff pseudoarthrosis of the femoral neck, the Paley type 2 CFD is a mobile pseudoarthrosis of the femoral neck. In type 2 there is a fibrous anlage of the femoral neck present extending from the base of the greater trochanter to the femoral head. There may be a well-formed synovial joint present with a mobile femoral head (type 2a), partially formed synovial joint with a partial fusion of the femoral head to the acetabulum at one spot (type 2b), or an absent or circumferentially fused femoral head (type 2c). The defining anatomic feature that characterizes all three of these is the presence of a greater trochanteric apophysis, which is missing in Paley type 3. Radiographically, the apophysis can be recognized when the proximal end of the femur appears rounded, while in most Paley type 3 cases, the upper end of the femur appears sharp and pointed. An MRI makes this distinction much more definitive.

There are three approaches for treatment for Paley type 2: (1) rotationplasty versus Syme amputation and prosthetic fitting, (2) serial lengthenings with pelvic support osteotomy at skeletal maturity, or (3) Superhip 1.5 or Superhip 2 procedures for selective cases of types 2a and some type 2b.

Superhip 1.5 Procedure for Treatment of a Mobile Proximal Femoral Pseudoarthrosis with a Mobile Femoral Head with Cartilaginous Remnants of the Femoral Neck Present (Fig. 22.27)

This procedure was developed in 2010. It is used when there is a type 2b with part of the femoral neck present on the femur and the femoral head. It is carried out similar to the Superhip 1, but with exposure of the anterior hip capsule. An arthrotomy is made, and the ends of the femoral neck are identified. A blade plate is used as described previously,

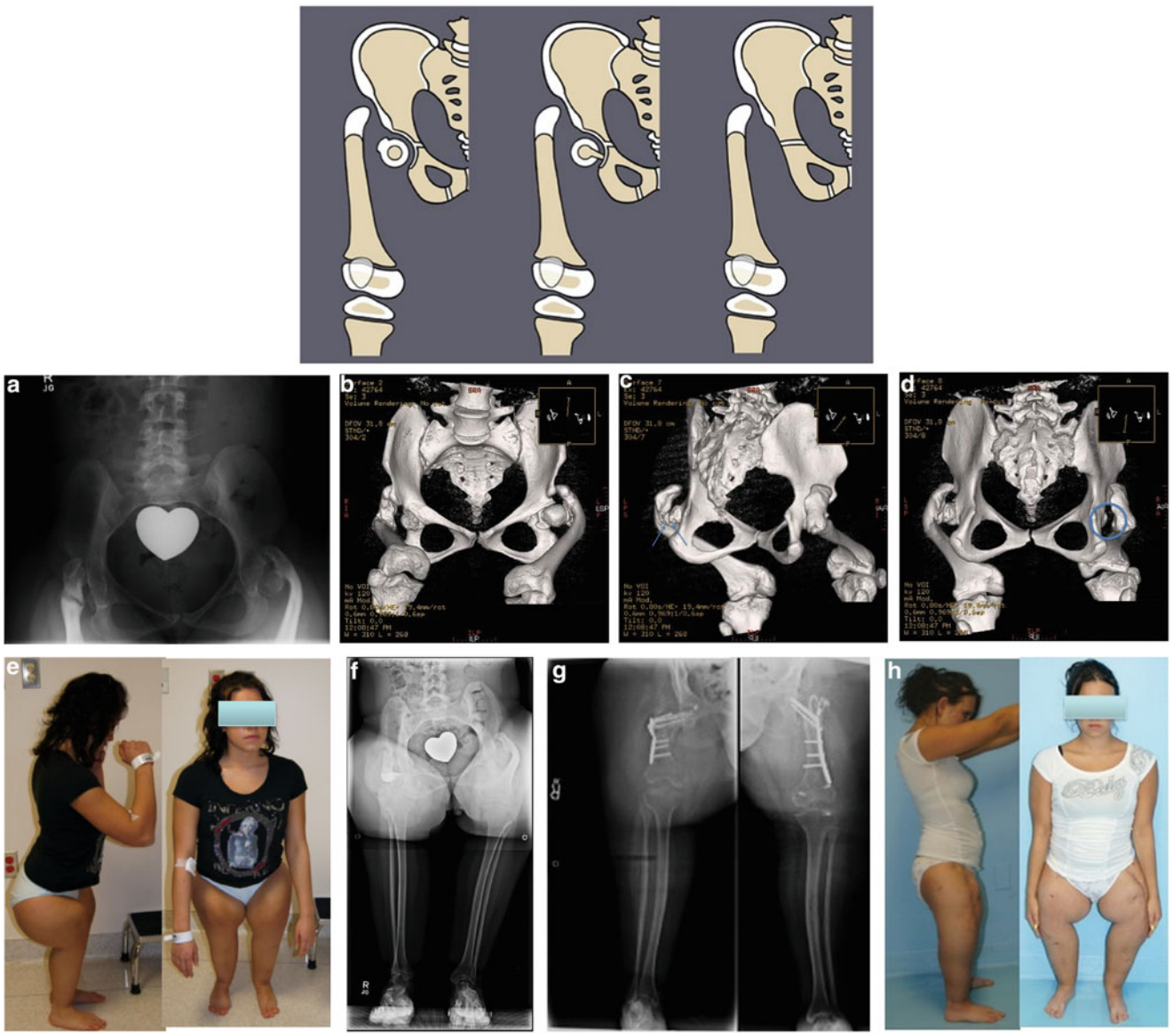


Fig. 22.26 An 18-year-old girl with bilateral Paley type 2a CFD. AP radiograph of pelvis shows femoral neck remnants on either side of the pseudoarthrosis while on the left side there is no femoral neck (**a**). Three-dimensional CT scan cuts show the femoral neck remnants from the front (**b**), oblique (**c**, *arrows*), and back (**d**) views of the right hip compared to a pseudoarthrosis without any neck remnant on the left (**d**, *circle*). Her standing posture is shown clinically (**e**) and radiographically (**f**). She has fixed hip and knee flexion contractures and very disproportionately short thighs. Note that her hands reach down below her knees. Due to the fixed flexion deformities, she fatigues when walking in a crouch gait. The goal of treatment was to make her stand and walk erect by eliminating the flexion deformities of both hips and knees. This was achieved by the Superhip 1.5 on the left and, the Superhip 2 on the right, and the Superknee with capsulotomy bilaterally. The posttreatment radiographs showing the new erect posture and the healed femoral neck pseudoarthrosis are also seen (**g**). Her clinical photograph after recovering from bilateral Superhip procedures. She now walks longer distances without fatigue (**h**). Since she stands and walks without a crouch gait her upper extremities hang to a more normal level than before

except that the cannulated chisel is introduced to each segment independently. The blade plate is then inserted across to stabilize the two parts. BMP-2 is used to induce the two parts of the cartilaginous femoral neck to ossify and unite. This is referred to as "cartilage welding." This is a unique feature of BMP-2 that was first identified by the senior author when treating the unossified femoral neck of type 1b [14] (see Fig. 22.24) [14].

Superhip 2 Procedure for Treatment of a Mobile Proximal Femoral Pseudoarthrosis with or Without Partial Fusion of the Femoral Head (Figs. 22.28 and 22.29)

The same approach as the Superhip is used. The operation remains the same until the subtrochanteric osteotomy. Before performing the osteotomy, the region of the femoral neck is

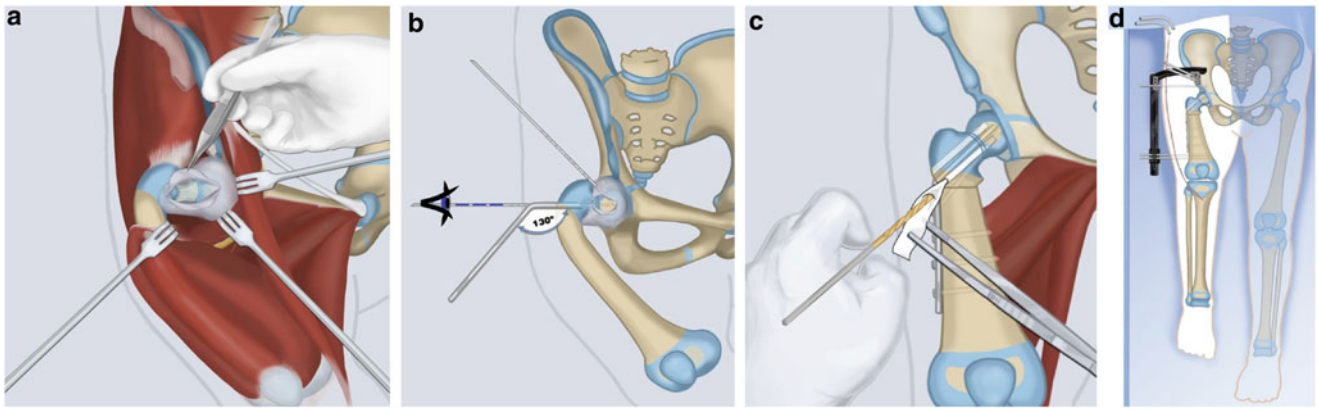


Fig. 22.27 Superhip 1.5 technique. (**a**) This is indicated for Paley type 2a when there is a remnant of cartilaginous femoral neck remaining on the proximal femur and femoral head. Through the same surgical exposure of the Superhip procedure (see Fig. 22.7) the anterior capsule of the hip is exposed by first separating the quadriceps from the gluteus medius. The capsule is then opened in line with the femoral neck. (**b**) The fibrous anlage is resected and the cartilaginous parts of the femoral neck are approximated. (**c**) The femur is osteotomized, shortened, and corrected as for the Superhip procedure and fixed with a blade plate. BMP-2 is inserted into the superior and inferior neck across the cartilaginous pseudoarthrosis. It is used to "weld" the cartilage together by inducing it to form bone. This is an off-label use of BMP-2. (**d**) The forces on the femur are neutralized by a spanning external fixator from the pelvis to the femur

dissected. The level of the acetabulum is identified with the image intensifier and the femoral head and acetabulum are identified. The capsule of the femoral head is opened inferiorly. The femoral head is moved in the acetabulum with forceps. If it is type 2a, it will be mobile. If it is type 2b, it will not move. If there is a cartilaginous or bony bridge, it is always located between the femoral head and the ischium. The cartilage of the head is cut back to the ossific nucleus to expose bone. Small 1-mm slices are cut back until the ossific nucleus is widened to about 2/3 of the femoral head. Dental wire may be passed through the nucleus for additional fixation.

The femoral neck is made from the proximal femur and greater trochanter. Posteriorly, the interval between the vastus lateralis and the abductors is found, and all of the abductors are released off of the proximal femur. The vastus lateralis and medial femoral circumflex artery serve as the pedicle for this bony segment. The external rotators are also removed. The proximal lateral cortex is partially decorticated, as it will eventually be oriented inferiorly and attached to the femoral shaft. The first cut is made about 4–5 cm long, perpendicular to the femoral shaft, creating the new femoral neck. This segment is rotated 135° on its soft tissue pedicle; the greater trochanter moves distal and lateral, and the distal cut end of the subtrochanteric osteotomy is rotated superiorly to fix to the ossific nucleus of the femoral head.

The trochanteric segment is predrilled with a 1.5-mm drill in four places around its cortical periphery, and 5/64″ threaded K-wires are run retrograde through the femoral neck and set to the same length. This bone is then drilled at 45° to its long axis for insertion of a Rush rod. This is drilled in a way to miss the path of the four wire holes and into the area previously decorticated. The distal femur intramedullary diaphysis is prepared with a 3.2-mm drill bit. Dental wire is also placed around this segment underneath the soft-tissue pedicles, in preparation for fixation.

The new femoral neck is then attached to the head. The four threaded K-wires are advanced carefully, holding the head with the dental wire. Verify that the wires are not intra-articular with fluoroscopy and a Freer elevator. After the threaded K-wire fixation, carefully tighten down the dental wire, compressing the femoral neck to head junction.

The second subtrochanteric osteotomy is then made at 45° to the shaft of the femur, removing a trapezoidal piece to provide adequate shortening of the femur. The cut should be made with the leg internally rotated to create femoral neck anteversion. The Rush rod is then placed through the trochanteric piece into the femoral shaft, and the dental wire tightened down. A cannulated 4.0 screw may be placed perpendicular to the shaft cut at 45° to augment fixation if there is adequate room. The abductor slide is performed as needed, and the abductors are repaired to the greater trochanter. To neutralize the forces on the femoral head and neck, an external fixator is applied from the pelvis to the tibia for 3 months.

Pelvic Support Osteotomy

A pelvic support osteotomy is used to treat most type 2c and some type 2b cases, combined with a distal femoral lengthening and realignment osteotomy (Fig. 22.30). This combination is called the *Ilizarov hip reconstruction* [89]. Pelvic support osteotomy is usually enough to prevent proximal migration of the femur during lengthening for non-congenital pathologies.

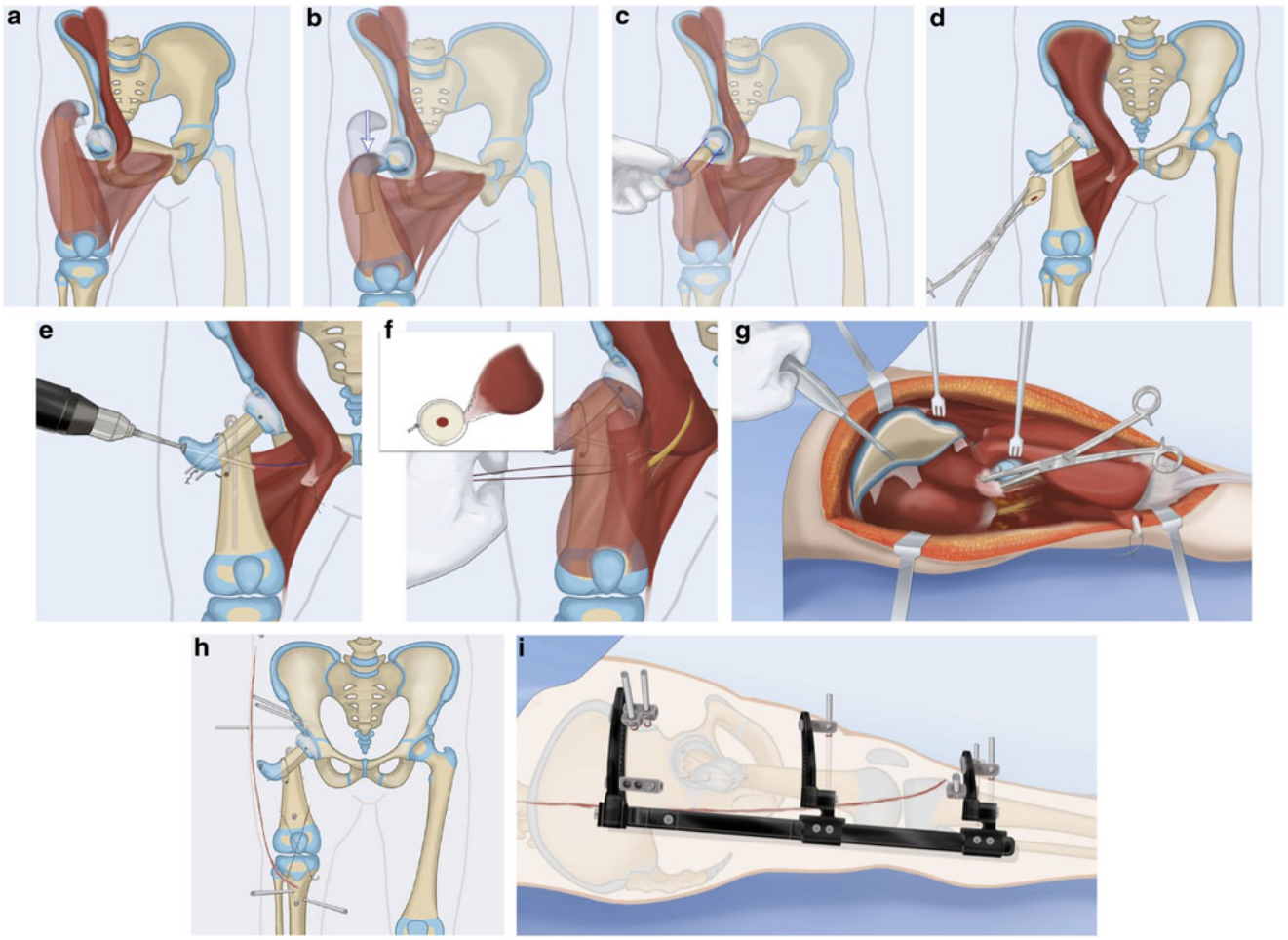


Fig. 22.28 Superhip 2 (SH2) technique. (**a**) After the same exposure as in the Superhip (see Fig. 22.7). The capsule of the hip is exposed without taking down the quadriceps muscle. The psoas tendon is released from the lesser trochanter. The femoral neck anlage is resected. The inferior capsule is opened. (**b**) All of the muscles inserting into the proximal femur are detached including all of the glutei, external rotators, and psoas. The muscles that originate on the proximal femur such as the quadriceps are not disturbed. The quadriceps becomes the vascular pedicle for the proximal femur and greater trochanter. The femur is osteotomized in the subtrochanteric region. It is then mobilized and moved distally. (**c**) The proximal femur is rotated 135° to form a femoral neck. (**d**) The femoral neck is fixed to the head using threaded K-wires and a cerclage wire. The second osteotomy of the femur is at 45° with the distal femur rotated 15° internally for anteversion. The femur is shortened according to the soft-tissue tethers. (**e**) The femur is fixed using a Rush rod, tension band wire, and compression screw technique. (**f**) The psoas tendon is reattached. (**g**) The hip abductors and rotators are reattached. If the abductors are too short, then the apophysis is split and an abductor slide is carried out. (**h**) A spanning external fixator from the pelvis to the femur to the tibia is used. In most cases there is a fixed flexion deformity of the knee. A posterior capsulotomy Superknee procedure is performed and pinned with cross K-wires to hold the extension correction. The capsulotomy and knee extension should be performed after the first osteotomy but prior to the shortening osteotomy of the femur. (**i**) MRS device used to span the hip and knee joints and neutralize the forces on the femoral neck for 4 months

Fig. 22.29 (continued) contraindication for the Superhip 2 procedure. Modification of the technique has made it a viable option in the author's current clinical practice. The critical consideration is rotating the fused area of the femoral head to face and join to the "new" femoral neck. Intraoperative fluoroscopic image (**g**); during this procedure, after fixation with threaded K-wires, cerclage wire and Rush rod are in place. After application of the spanning external fixator (**h**, **i**). Knee capsulotomy and acute extension of the knee flexion contracture were carried out and temporarily fixed with cross K-wires (**h**, **i**). Final radiograph after removing the external fixator (**j**). Note that the Rush rod reaches to the distal femoral physis. Radiograph 2 years later showing that there has been a lot of growth of the distal femur as seen relative to the end of the Rush rod (**k**). Hip and knee range of motion after recovery from Superhip 2 procedure (**l**, **m**). First lengthening of the femur with MRS and articulated fixation spanning the knee joint. The hip joint was left free (**n**). The femur was successfully lengthened 8 cm with no loss of hip or knee motion

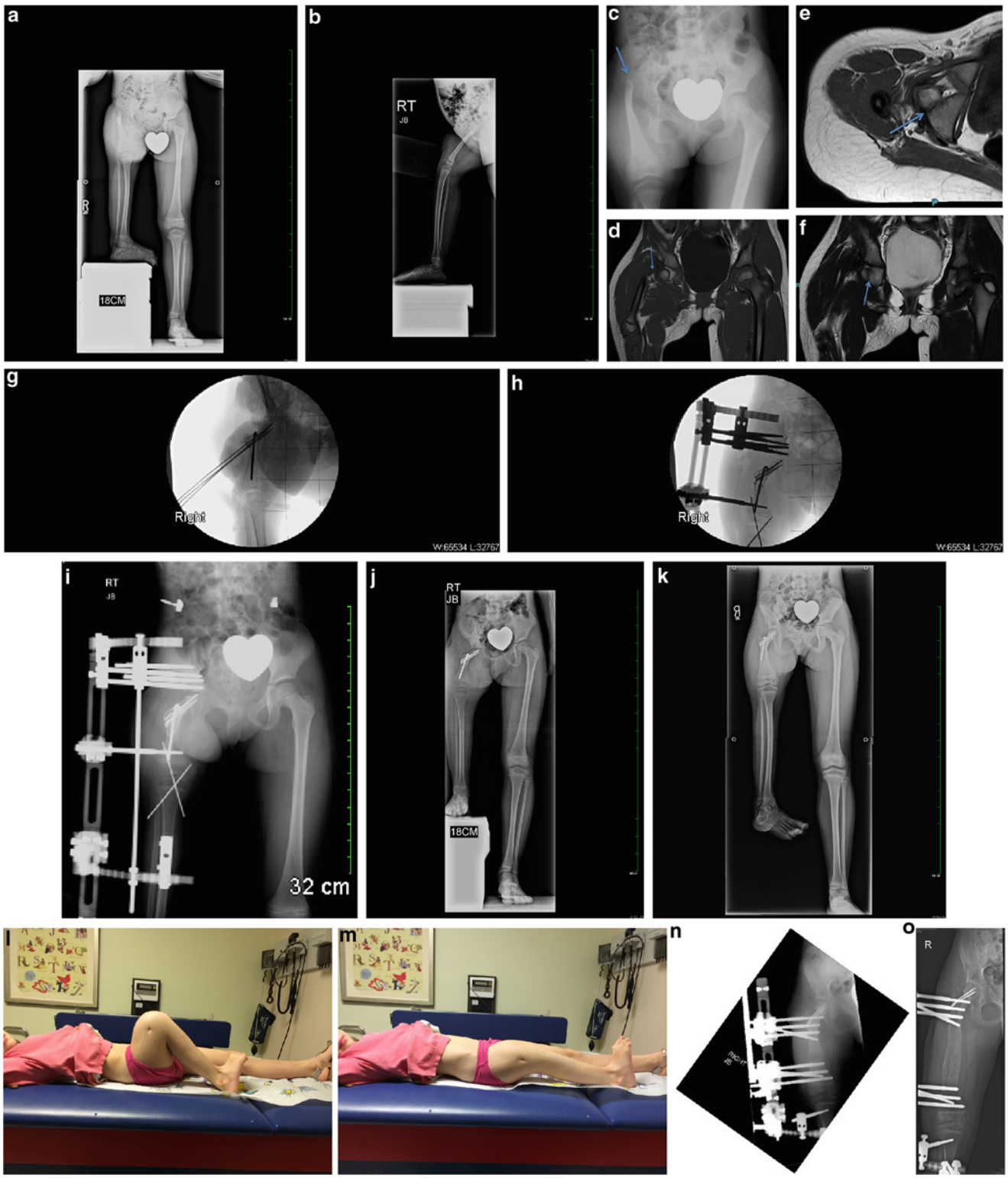


Fig. 22.29 Standing AP and lateral radiographs of patient with Paley type 2b CFD (**a**, **b**). AP pelvis radiograph showing the rounded proximal end of the femur indicating that it has an apophysis. It is proximally migrated. There is an obvious femoral head ossific nucleus with a well-formed acetabulum present. The *arrow* is pointing to the greater trochanteric apophysis (**c**). MRI coronal cut, showing the ossific nucleus with no bony or cartilaginous connection to the proximal femur (*arrow*). The greater trochanter is out of plane for this cut (**d**). MRI transverse cut showing the femoral head in the acetabulum (**e**). Note the bridge of bone posteriorly between the femoral head and the ischium (*arrow*). The rest of the femoral head is covered with cartilage and remains unfused. The same can be seen on the coronal cut of the posterior femoral head (*arrow*) (**f**). This is therefore classified as a partial fusion of the femoral head (Paley type 2b). Type 2b was originally thought to be a

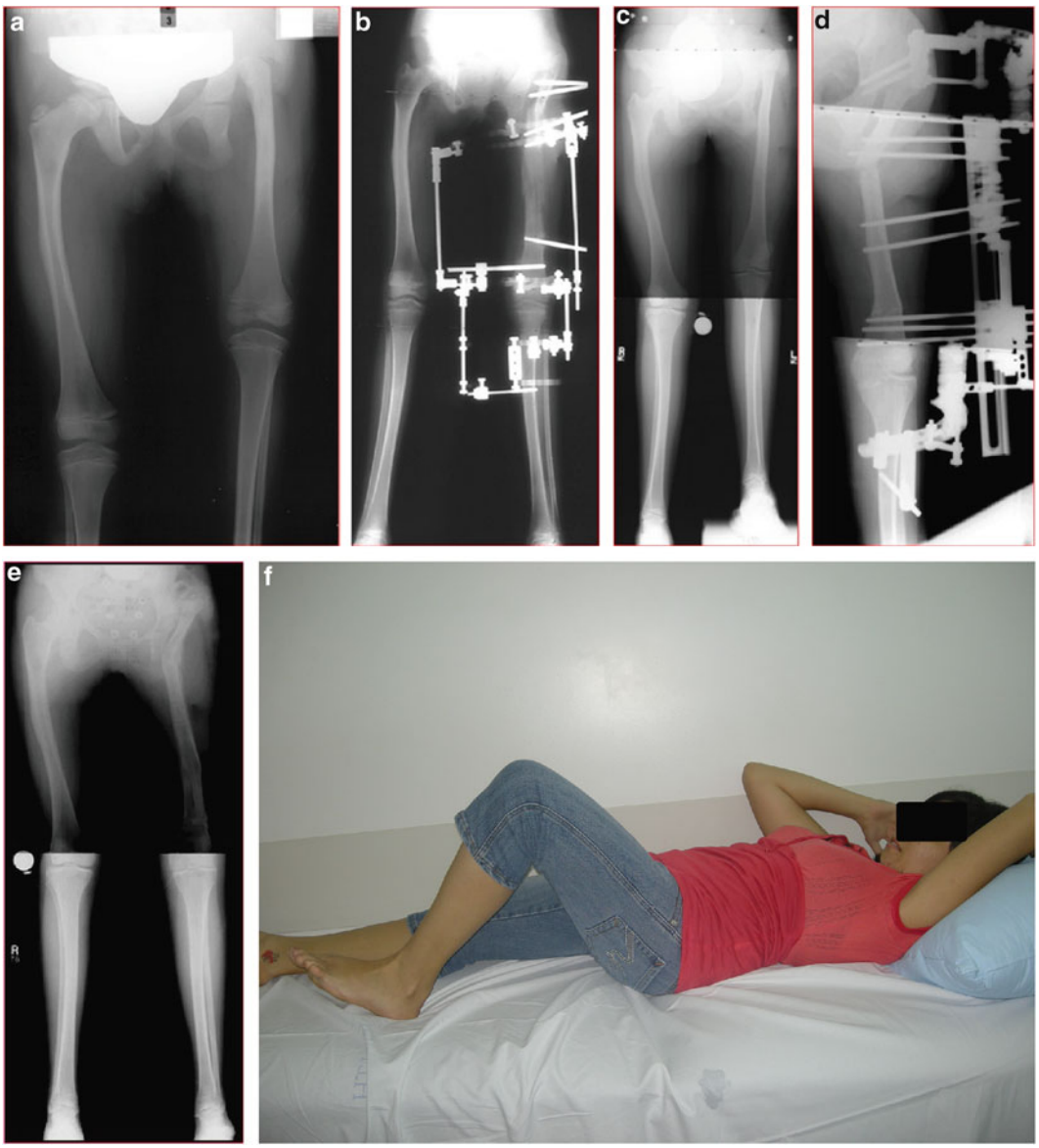


Fig. 22.30 AP radiograph (**a**) of 10-year-old girl with Paley type 2c CFD left side (absent femoral head). She was treated by pelvic support osteotomy (**b**) and distal femoral lengthening (13 cm). This is an unusual and extreme amount for one lengthening. The only reason we exceeded the 8-cm limit is that she was able to maintain knee range of motion to 90° and for financial reasons was not sure if she would ever be able to afford travelling for another lengthening. We therefore continued lengthening as long as she was able to maintain knee motion. The pelvic support angulation remodeled and the leg length difference was 10 cm at skeletal maturity (**c**). At age 16 she underwent a second pelvic support osteotomy with 10-cm lengthening (**d**). Final radiographic result showing limb length equalization (**e**). She has excellent function (**f**) and walks with no appreciable limp. She is now 20 years from her original surgery

For congenital pathologies, the fixation should be extended to the pelvis to prevent proximal migration of the femur. In young children with very short femora, the femur may be too small to perform both the pelvic support and the distal lengthening osteotomies. In addition, the valgus component of the pelvic support osteotomy may remodel straight.

The proximal osteotomy is performed at the level at which the proximal femur crosses the ischial tuberosity in maximum cross-legged adduction. The amount of valgus is equal to the total amount of adduction of the hip plus 15° of overcorrection. The proximal osteotomy should also be internally rotated and extended. The amount of rotation is judged by the position of the knee relative to the hip in maximum adduction. The amount of extension depends on the amount of hip fixed flexion deformity.

The level of the distal osteotomy is determined by the intersection of the proximal and distal mechanical axes. The proximal mechanical axis passes through the proximal

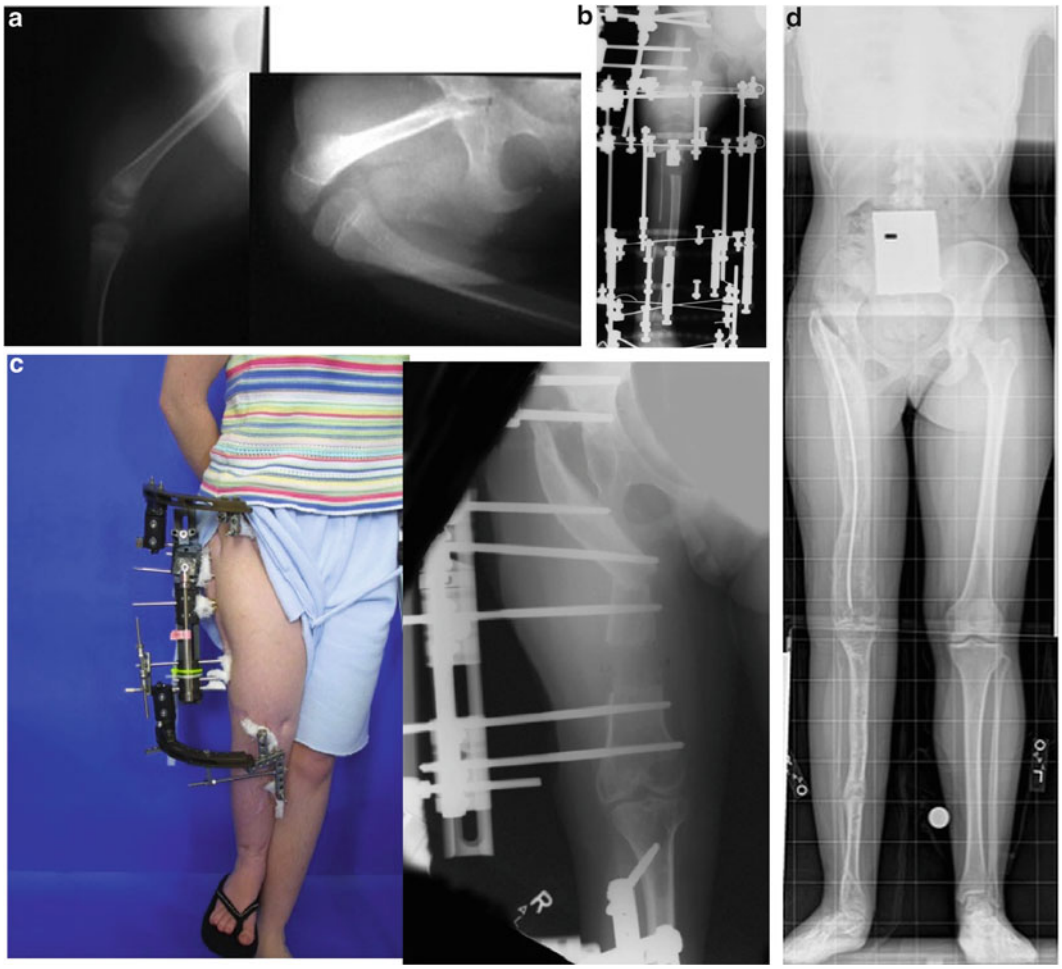


Fig. 22.31 (**a**) CFD Paley type 3A. Note the initial knee flexion and extension for an arc of motion of 70°. Parents refused to have a Syme's or a rotationplasty. She was treated at age 7 with a lengthening of the femur and tibia for a total of 12.5 cm (**b**). At age 10 she underwent the same treatment again and achieved another 12.5 cm of lengthening. At age 14 when she was skeletally mature she underwent a pelvic support osteotomy together with a 10-cm femur lengthening (**c**). At age 18 she underwent her fourth femoral plus tibial lengthening of 12.5 cm. She had a final tibial lengthening of 2.5 cm to correct deformity and equalize her leg lengths (**d**). Her total lengthening was 50 cm. She maintains 70° of knee motion and good ankle and hip motion. She has excellent clinical function, has no pain, and walks with minimal limp. She is now 18 years following her first procedure

formal osteotomy site and is perpendicular to the horizontal line of the pelvis. The distal axis is from the center of the ankle to the center of the knee. Varus correction can be made through the distal osteotomy. The external fixator must still be extended to the tibia with hinges, as previously discussed.

Treatment of CFD Type 3

Deficiency of the proximal femur with absent femoral head, greater trochanter, and proximal femoral metaphysis results in a mobile pseudoarthrosis and a very short femoral remnant. The Paley type 3a CFD has a mobile knee with at least 45° of motion, usually with a 45° knee flexion deformity. The type 3b has a stiff knee with less than 45° of motion and usually has a greater flexion deformity. The most predictable and reliable treatment option in these cases remains prosthetic reconstruction surgery with either rotationplasty or a Syme amputation. Limb reconstruction surgery has a role in these cases and can equalize an LLD. Because the number of these patients treated this way is small, the ultimate functional result for these cases is still not predictable.

Limb Reconstructive Surgery for Type 3

Limb reconstructive surgery is most applicable to type 3a cases that have functional knee range of motion (Fig. 22.31). Many of these patients have knee and hip flexion contractures. These are treated by soft tissue releases, using the same approach as described above for type 1 CFD. The fascia lata is reflected proximally, and the quadriceps and abductors are elevated off the proximal femur. The psoas tendon is absent, but the rectus and sartorius are present.

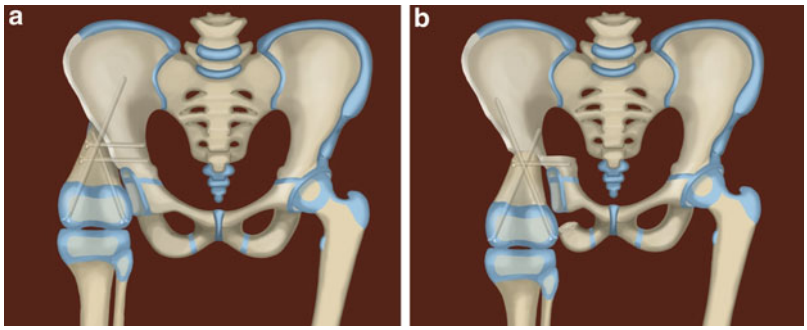


Fig. 22.32 Rotationplasty for CFD type 3. (**a**) Brown rotationplasty. The femur is fused to the side of the pelvis. It is difficult to align the femur perpendicular to the pelvis. The hip protrudes laterally due to its lateral fusion position. (**b**) Paley-Brown rotationplasty. A Chiari osteotomy is used to fuse to the femur. The ischium is notched to make room for the sciatic nerve

Any fibrous femoral anlage is resected, and frequently, some of the cartilaginous femoral anlage may need to be trimmed. The proximal femur is freed from these attachments, including the hip capsular remnants, permitting it to move proximally without a soft-tissue tether. This is important for the acute correction of the knee contracture. The long lateral incision is extended distally and the peroneal nerve is decompressed and protected. Because the femur is so short, it is advisable to explore the peroneal nerve and follow it proximally to the hip joint region prior to significant release. This will prevent injury to the sciatic nerve, as it passes very near the dissection around the hip capsular remnant.

The lateral approach to knee flexion contracture release of the posterior capsule is performed, and the knee joint is fully extended. A 2-mm Steinmann pin may be drilled across the knee joint from the femur into the tibia to maintain full knee extension. During the same operation, a monolateral fixator is placed from the pelvis to the femur and tibia. The most important and strongest pelvic pin is one from the anterior inferior iliac spine oriented towards the greater sciatic notch, using the cannulated drill technique. Another pin from the lateral side in the supra-acetabular region is attached to the fixator. One pin is used in the distal femur and two in the tibia. The fixation is kept in place for 6 weeks to maintain knee and hip extension. Once fixation is removed, range of motion exercises are started to regain knee motion.

After the above procedures, the femur can be lengthened. The external fixator is placed from the pelvis to the femur to the tibia. The femur is lengthened up to 8 cm. The knee joint is distracted, but since no knee or hip hinges are used when the femur is too short, there is little need for physical therapy during this first lengthening. The goal of this first lengthening is to convert a type 3a femur to a type 2b femur. The rest of the treatment is the same as a type 2b: serial lengthenings and eventually a pelvic support osteotomy (Fig. 22.31) combined with a final lengthening. Since the predicted discrepancy ranges between 30 and 40 cm, at least four lengthenings and an epiphysiodesis are required to equalize limb lengths.

Prosthetic Reconstructive Surgery for Type 3

Prosthetic fitting is possible without any surgery. However, this can be difficult with a hip and knee flexion contractures, as well as a foot not in equinus. Surgical releases as described above may help with prosthetic fitting. A Syme or Boyd amputation may also help create a residual limb that is fitted easily. In some cases, even a pelvic support osteotomy may be considered to decrease limp and stabilize the hip joint.

The other approach to prosthetic reconstruction is with rotationplasty [49, 90]. Torode popularized the Van Nes rotationplasty for patients with CFD. They used a long oblique incision. The goal was to fuse the residual knee and rotate the limb 180°, such that the foot points backward and the ankle can function as a knee joint. This was most applicable to cases in which the ankle was already at the level of the opposite knee joint. Brown [44] modified the rotationplasty approach, using a racquet-like incision, performing the rotation between the femoral remnant and the pelvis (Fig. 22.32a). He then fused the femur to the lateral aspect of the ilium, thus converting the knee into a hip joint and the ankle into a knee joint. This provided improved hip stability over that provided by the Van Nes rotationplasty. However, one problem with the Brown method is excessive shortening of the hip muscles and lateralization of the hip and lower limb. It is also challenging to adjust the alignment correctly.

Paley modified the Brown technique in 1997 by adding a Chiari osteotomy of the pelvis and fusing the femoral remnant to the cancellous roof of ilium (Figs. 22.32b and 22.33). The detached muscles are not removed and are instead transferred distally as much as possible. Care is taken to appropriately shorten the knee muscles so that they can function adequately as hip flexors and extensors. The fascia lata connected to the gluteus maximus is reattached to the tibia to serve as a hip abductor. Posterior capsule release of the knee is performed as the knee is often contracted as much as 90°. The peroneal nerve is decompressed to prevent injury and

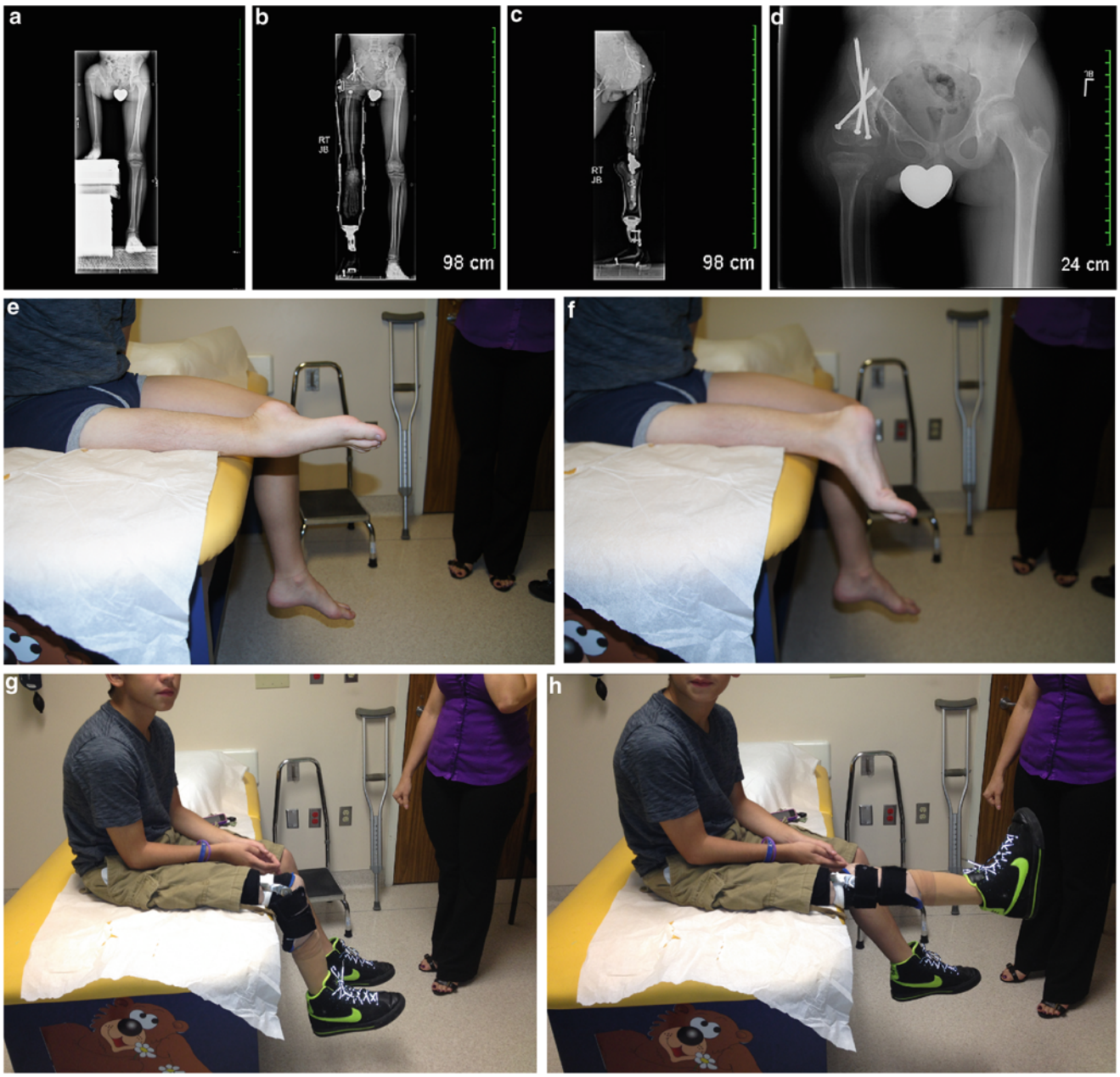


Fig. 22.33 (**a**) Preoperative radiograph of a 10-year-old boy with CFD Paley type 3A. Note how the knee is fully flexed and the hip is rotated into a frog leg position. Standing AP (**b**) and lateral (**c**) radiographs after Paley-Brown rotationplasty using the prosthesis. The ankle is at the level of the opposite knee. Closeup radiograph (**d**) showing the fusion to the pelvis and the closure of the distal femoral physis. Sitting posture with excellent hip flexion and active "knee extension" (via ankle plantar flexion) (**e**), and "knee flexion" (via ankle dorsiflexion) (**f**). Active extension (**g**) and flexion (**h**) of the knee wearing the prosthesis

allow greater rotation. A supramalleolar osteotomy may be performed to correct the internal rotation deformity of the tibia. Epiphysiodesis is done with the same screws used for fixation of the femur to the pelvis. This modified Paley-Brown rotationplasty provides good functional results with better hip function and stability than the Van Nes and the Brown rotationplasties.

The senior author has recently developed a new rotationplasty variant, which for labeling purposes will be called the Paley rotationplasty (Fig. 22.34). The steps are the same as in the Brown, but instead of fusing the femur to the pelvis, the femoral segment is fixed to a mobile femoral head. This gives better hip reconstruction with active abduction motion present. Even if the femoral head is fused to the acetabulum, as in

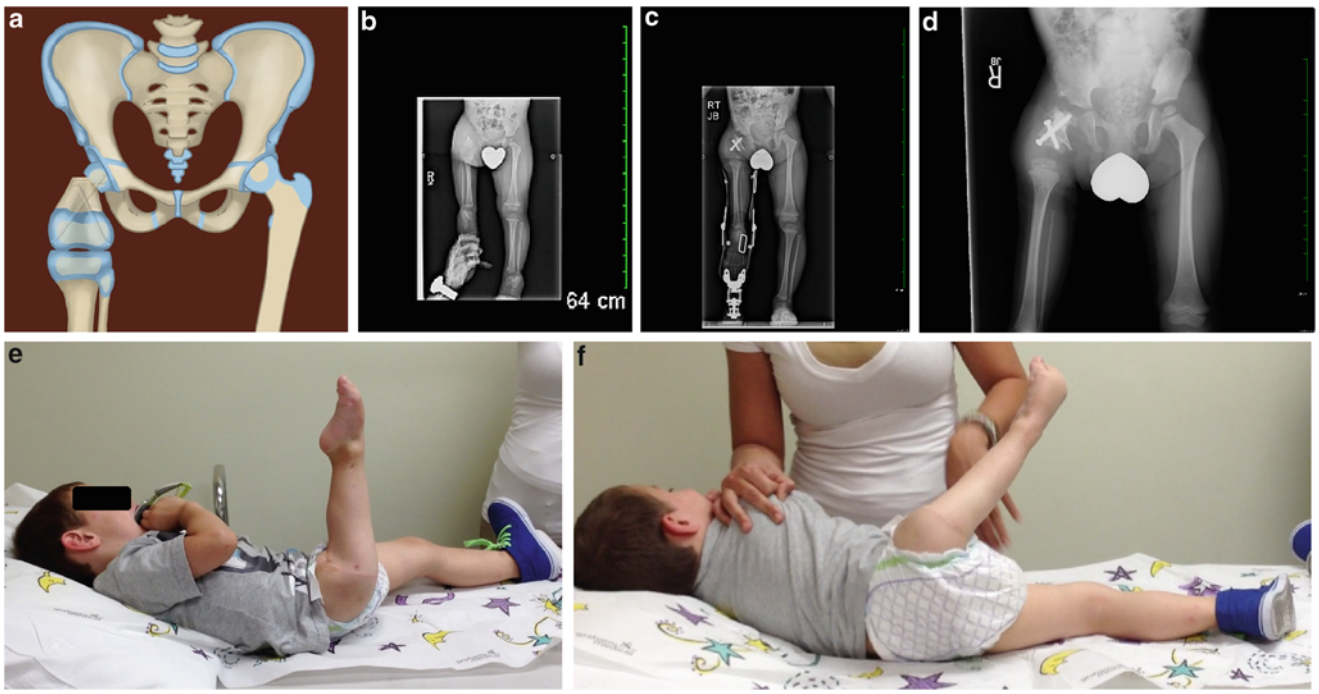


Fig. 22.34 (**a**) Paley rotationplasty. The femoral remnant is fused to the femoral head, which is made mobile in the acetabulum. (**b**) 2-year-old boy with type 3b. (**c**) Paley rotationplasty was performed and the femoral head was fused to the femoral head (**d**). He has excellent active hip flexion (**e**) and abduction (**f**) of the hip joint. Hip flexion is usual but hip abduction is not usually possible after a Brown rotationplasty

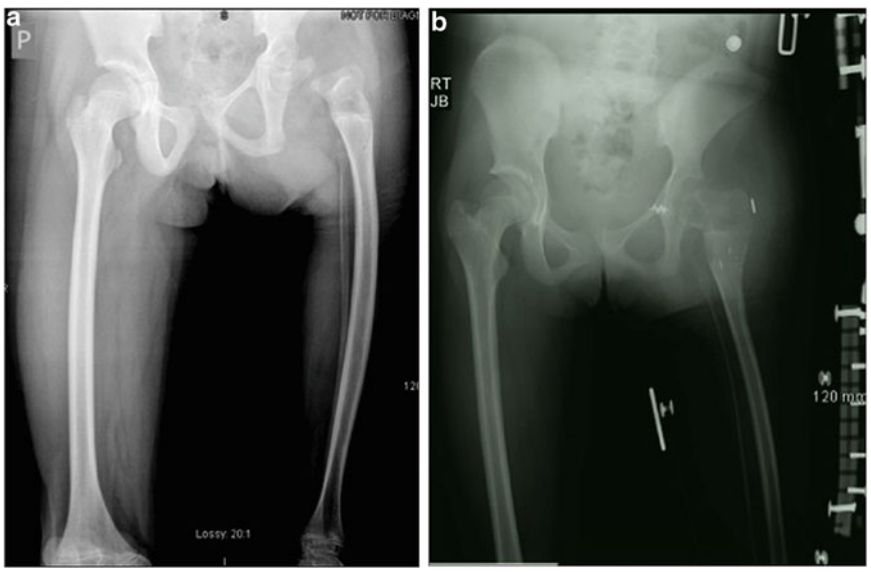


Fig. 22.35 Twelve-year-old girl with CFD Paley type 3c (**a**). She had a rotationplasty performed with insertion of the proximal medial tibia into the acetabulum (**b**). This gave her a stable hip with excellent function of the hip and knee joints

type 2b, it can be freed up similar to the Superhip 2 and then fused to the femoral remnant (see Fig. 22.34). Finally, in cases where there is no knee joint present (type 3c), the proximal tibia or fused distal femoral remnant can be fused to the residual femoral head. If there is no femoral head, then a roof can be created by a Chiari osteotomy or by creating a depression into the pelvis (Fig. 22.35). This is stabilized by a suture ligament tether technique as previously shown (see Fig. 22.23).

Summary

CFD is a spectrum of congenital deficiency, deformity, and discrepancy of the femur that involves not only the osseous part of the femur, but also the surrounding musculature, ligaments, and joints. All of these components must be addressed if one is to be successful in the treatment of CFD. Though the

definitive origin of CFD is still not known, it is a problem that can affect the function of a patient for his or her entire life. Depending on the severity, there are several treatment strategies, reconstructive surgeries, and prosthetic options, all of which can improve a patient's gait, function, and quality of life. As each deformity is different, so also is each patient and family, and therefore treatment should be tailored to their individual needs and cultural expectations. Advances in surgical techniques, new biologic agents, and new technologies have expanded the reconstructive indications and options in lieu of amputation. Though more research of newer treatments and their long-term outcomes are needed, there are now more options and hope for a brighter future for a child with CFD.

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Fibular Hemimelia: Principles and Techniques of Management

23

John E. Herzenberg, Lior Shabtai, and Shawn C. Standard

Introduction

Congenital fibular deficiency or fibular hemimelia (FH) is a rare congenital disorder in which the fibula is partially or completely absent. It is the most common congenital long bone deficiency, and the incidence in the USA is between 7.4 and 20 cases per million live births (up to 1 per 50,000 live births) [1–3]. It is primarily associated with lower extremity anomalies such as femoral shortening, hypoplastic lateral femoral condyle, genu valgum, anteromedial bowing of the tibia, limb length discrepancy (LLD), absent or deficient anterior cruciate ligament (ACL) or posterior cruciate ligament (PCL), ball-and-socket ankle joint, equinovarus or equinovalgus foot, tarsal coalition, and absence of lateral rays of the foot (Fig. 23.1). Upper extremity manifestations may include syndactyly and ulnar hemimelia [4, 5]. Rodriguez-Ramirez et al. [6] examined the prevalence of associated congenital osseous anomalies in patients with fibular hemimelia and found that lateral femoral condyle hypoplasia was the most common associated anomaly (93 %) followed by ball-and-socket ankle joint (80 %), congenital short femur (72 %), tarsal coalition (51 %), and forefoot ray deletion (44 %). Equinovalgus is much more common than equinovarus. Roux and Carlioz [7] demonstrated absence of the anterior cruciate ligament in 95 % of the cases. In conclusion, FH can present with a broad spectrum of severity, from mild cases to very severe manifestations.

The treatment of FH changes according to the severity of deformity and the associated skeletal manifestation with a broad spectrum of choices: from simple contralateral epiphysiodesis in mild cases to limb reconstruction and lengthening in more severe cases, the latter composed of multiple surgical procedures during the patient's childhood and adolescence. The common alternative to extensive lengthening is amputation and prosthetic rehabilitation [5, 8–16]. The challenge in limb lengthening is to achieve normal limb alignment and length with a functional, painless, plantigrade foot. This undertaking is often characterized by repeated surgical procedures and a high rate of complications and sequelae such as pin tract infections, residual LLD, delayed union, ankle and knee stiffness, refracture, knee subluxation, and residual foot deformities. Deciding which treatment to apply (lengthening versus amputation) can be challenging for the surgeon and the family [8–12].

Parents must be aware of all the options and the associated potential complications before making treatment decisions. Fortunately, very little can be done during the first year of life in most cases (unless the child has an associated club foot), so there is ample time to meet with the parents, discuss the options, and have them carefully consider which option is best for their family and their child.

The aim of this chapter is to discuss in detail the principles and techniques of management of lengthening reconstruction for fibular hemimelia by understanding and addressing each deformity in this complex, multifaceted disorder. A detailed description of the amputation options (Syme or Boyd) are covered in the amputation chapter (Chap. 13) by J.A. Herring.

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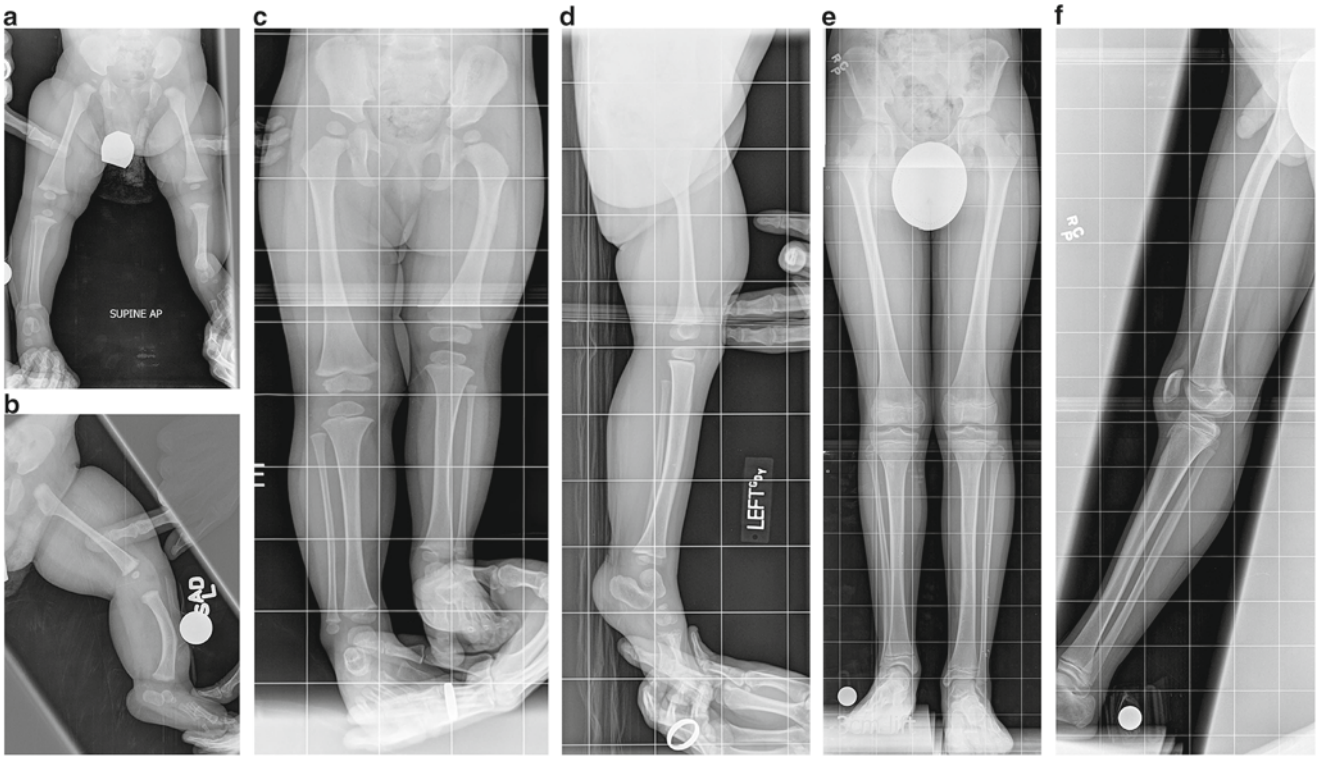


Fig. 23.1 AP (**a**) and lateral (**b**) view radiographs show 10-month-old infant with fibular hemimelia, concurrent congenital femoral deficiency, a typical anterior bow of the tibia, and equinovalgus foot deformity. AP (**c**) and lateral (**d**) view radiographs show a 2-year-old child with fibular hemimelia and congenital femoral deficiency as well as mild genu valgus and ankle valgus. AP (**e**) and lateral (**f**) view radiographs of a 12-year-old patient with fibular hemimelia who has genu valgus, mild leg length discrepancy, a ball-and-socket ankle joint, and talocalcaneal coalition. Reprinted with permission from the Rubin Institute for Advanced Orthopedics, Sinai Hospital of Baltimore

Box 23.1. Typical Deformities Associated with FH

- Tibial shortening
- Equinovalgus foot
- Missing all or part of fibula
- Missing one or more lateral rays
- Talocalcaneal coalition
- Anteromedial bowing of tibia
- Cruciate ligament insufficiency
- Genu valgum
- Hypoplasia of lateral femoral condyle
- Limb length difference
- Syndactyly
- Ball-and-socket ankle joint
- Equinovarus foot

Classification

Several classifications exist for fibular hemimelia. The earliest published classification is the Coventry and Johnson [17] classification from 1952 (Fig. 23.2). They divided patients with FH into three types:

Type I: Partial unilateral absence of fibula; normal or slight bowing of the tibia with some shortening of the limb; the foot is normal or slightly deformed.

Type II: The fibula is completely or almost absent; anterior bowing of the tibia with skin dimpling; deformed ankle joint; deformed foot with absent rays.

Type III: Includes type I or type II but is associated with other congenital deformities or bilateral involvement.

The Achterman and Kalamchi classification, from 1979 [18], is the most commonly used (Fig. 23.3). They divided the patients into two groups with one subdivision:

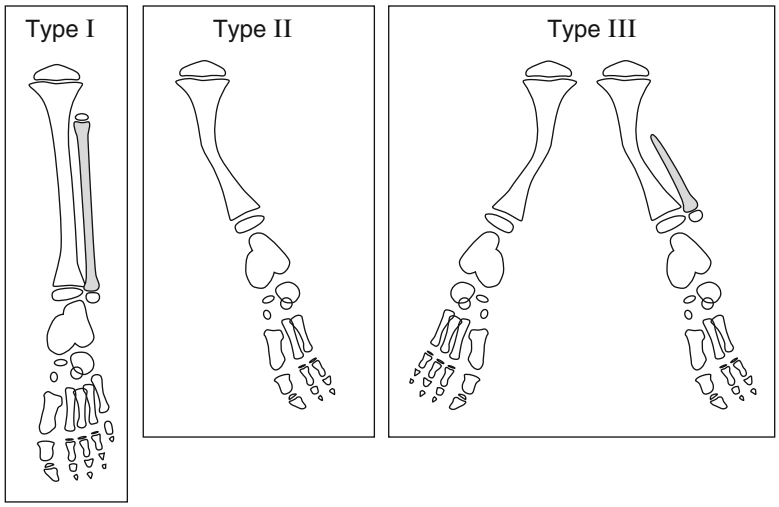


Fig. 23.2 Coventry and Johnson classification of FH: Type I: partial unilateral absence of fibula, normal or slight bowing of the tibia with some shortening of the limb, and the foot is normal or slightly deformed. Type II: fibula is completely or almost entirely absent, anterior bowing of the tibia with skin dimpling, deformed ankle joint, and deformed foot with absent rays. Type III: includes type I or II associated with other congenital deformities or bilateral involvement. Reprinted with permission from the Rubin Institute for Advanced Orthopedics, Sinai Hospital of Baltimore

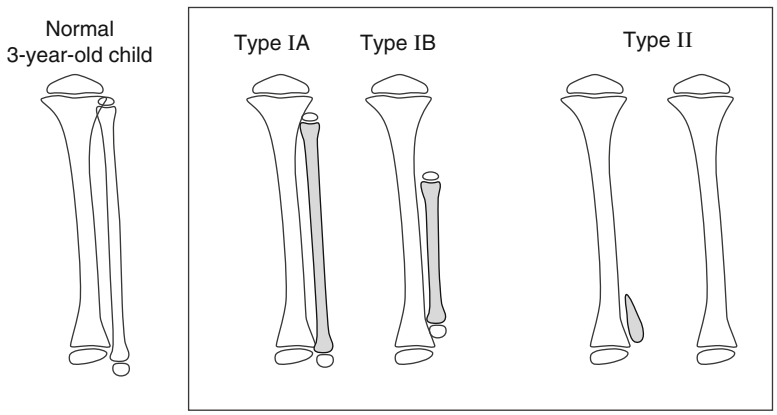


Fig. 23.3 Achterman and Kalamchi classification of FH: Type I: incomplete fibular deficiency, subdivided into I-A and I-B. Type I-A: proximal fibular epiphysis is distal to the level of the tibial growth plate, distal fibular growth plate is proximal to the dome of the talus. Type I-B: 30–50 % partial fibular absence, fibula does not articulate with the talus. Type II: complete fibular deficiency, with or without a tiny distal fibular remnant. Reprinted with permission from the Rubin Institute for Advanced Orthopedics, Sinai Hospital of Baltimore

Type I: Incomplete fibular deficiency; Type I is subdivided into: Type I-A: The proximal fibular epiphysis is distal to the level of the upper tibial growth plate and the distal fibular growth plate is proximal to the dome of the talus; and Type I-B: 30–50 % fibular shortening and the fibula does not articulate with the talus.

Type II: Complete fibular deficiency, with or without a small distal fibular remnant.

The Stanitski classification, from 2003 [19] (Fig. 23.4), is a morphologic description that divides FH into four categories based primarily on radiographic presentation:

1. Fibula: Normal, partially absent, or completely absent.
2. Ankle joint: According to the morphology of the tibiotalar joint: horizontal, valgus, or ball-and-socket joint.
3. Tarsal bones: Tarsal coalition present or absent.
4. Foot: Numbers of rays.

Birch et al. [20] in 1998 presented a new classification system based on the clinical status of the foot and the magnitude of limb shortening as a percentage of the contralateral limb on radiographs in order to anticipate the extent of deformity at maturity and to recommend the appropriate treatment required (Fig. 23.5). In 2011, Birch later [5, 21] modified the treatment proposed: fewer amputations and more reconstruction procedures. Cases are divided into two categories:

Type 1: Functional foot, in which the foot has at least three rays and can provide a stable weight-bearing platform. Type 1 is divided into four subtypes according to the percentage of total limb shortening compared with the contralateral side.
Type 1A: <6 % leg length inequality; the treatment is orthosis or contralateral epiphysiodesis.

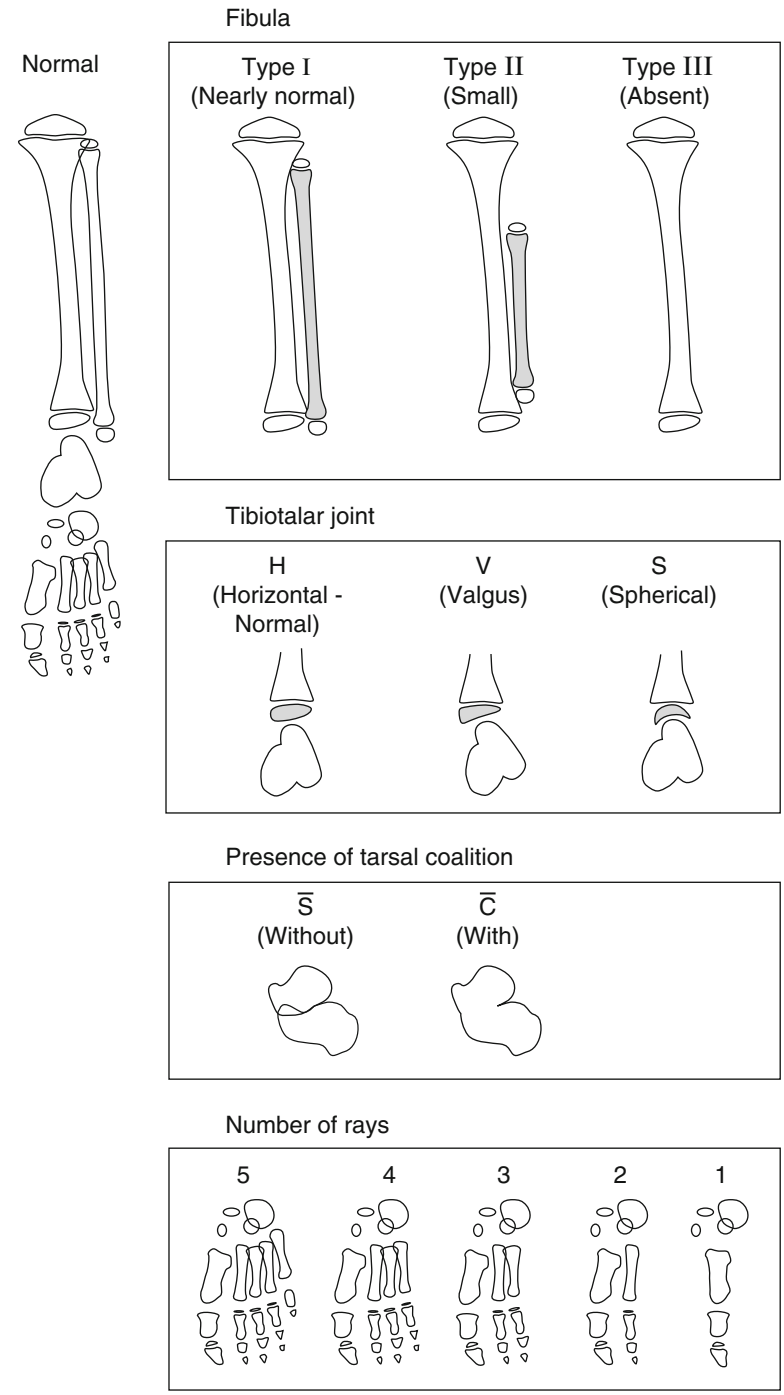


Fig. 23.4 Stanitski classification of FH according to four parameters: *1*. Fibula: normal, partially absent, or completely absent. *2*. Ankle joint: according to the morphology of the tibiotalar joint: horizontal, valgus, or spherical (ball-and-socket joint). *3*. Tarsal bones: tarsal coalition present or absent. *4*. Foot: numbers of rays. Reprinted with permission from the Rubin Institute for Advanced Orthopedics, Sinai Hospital of Baltimore

Type 1B: 6–10 % leg length inequality; the treatment is epiphysiodesis ± lengthening.

Type 1C: 11–30 % leg length inequality; the treatment is one or two lengthening procedures ± epiphysiodesis or extension orthosis.

Type 1D: >30 % leg length inequality; the treatment proposed is more than two lengthening procedures or amputation or extension orthosis.

Type 2: Nonfunctional foot. Type 2 is subdivided into Types 2A and 2B according to the functionality of the upper extremity.

Type 2A: Upper extremities are functional; therefore, the treatment proposed is early amputation.

Type 2B: Upper extremities are nonfunctional, and so foot amputation is contraindicated, as the foot must act as a replacement for the upper extremity.

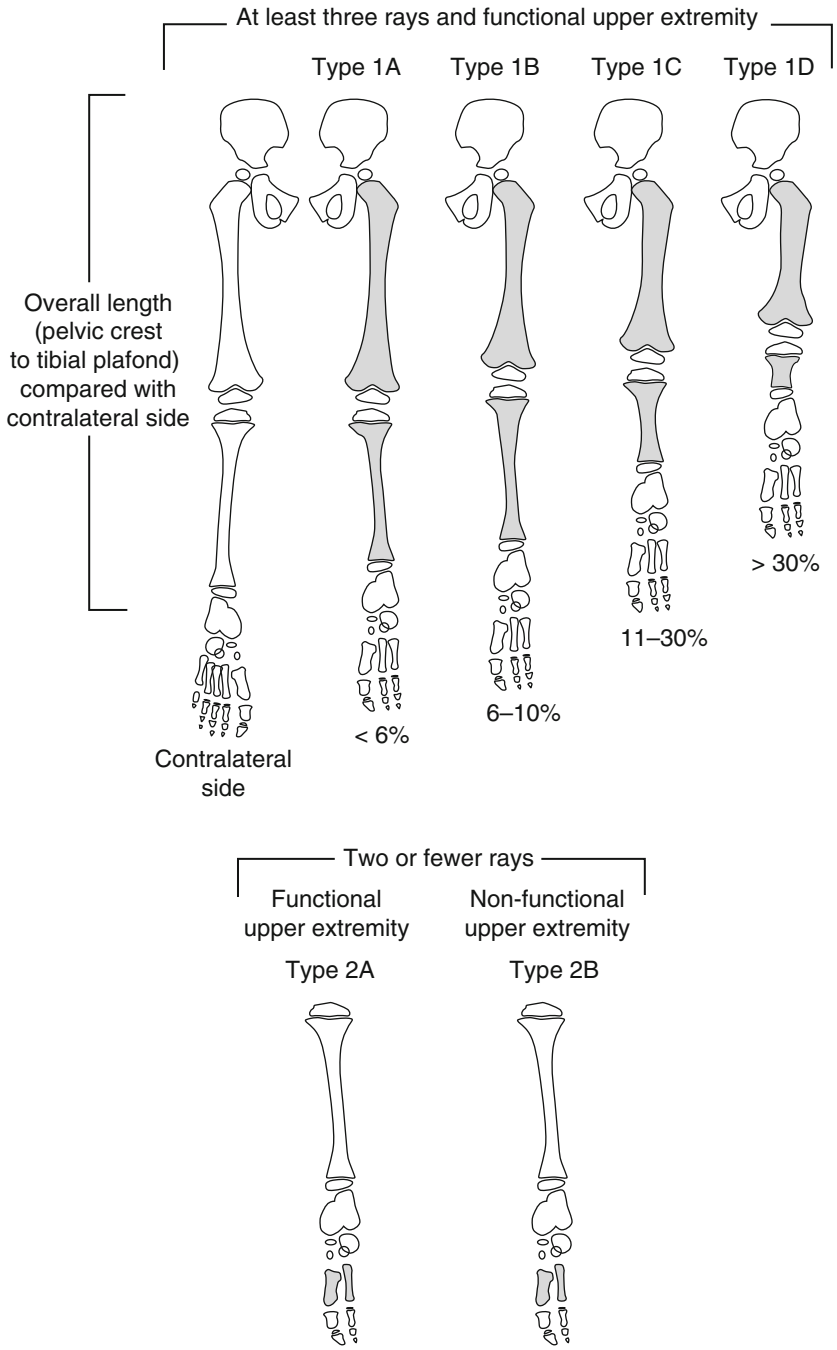


Fig. 23.5 Birch classification of FH. Type 1: Functional foot, foot with at least three rays that can provide a stable weight-bearing surface. Type 1 is divided into four subtypes according to the limb length difference compared with the normal side. Type 1A: <6 % limb length inequality; Type 1B: 6–10 % limb length inequality; Type 1C: 11–30 % limb length inequality; Type 1D: >30 % limb length inequality. Type 2: Nonfunctional foot (fewer than three rays). Type 2 is subdivided into Types 2A and 2B according to the functionality of the upper extremity. Type 2A: Functional upper extremity; Type 2B: Nonfunctional upper extremity. Reprinted with permission from the Rubin Institute for Advanced Orthopedics, Sinai Hospital of Baltimore

Box 23.2. Classification Schemes Used in North America for FH

- Coventry and Johnson
- Achterman and Kalamchi
- Stanitski
- Birch
- Paley

A more recent classification proposed by Paley [16] is surgically oriented (reconstruction, not amputation) (Fig. 23.6). He classified FH into four types, based on the ankle pathology:

Type 1: Stable normal ankle.
Type 2: Dynamic valgus ankle.
Type 3: Fixed equinovalgus ankle. Type 3 is divided into four subtypes according to the location of the valgus deformity.

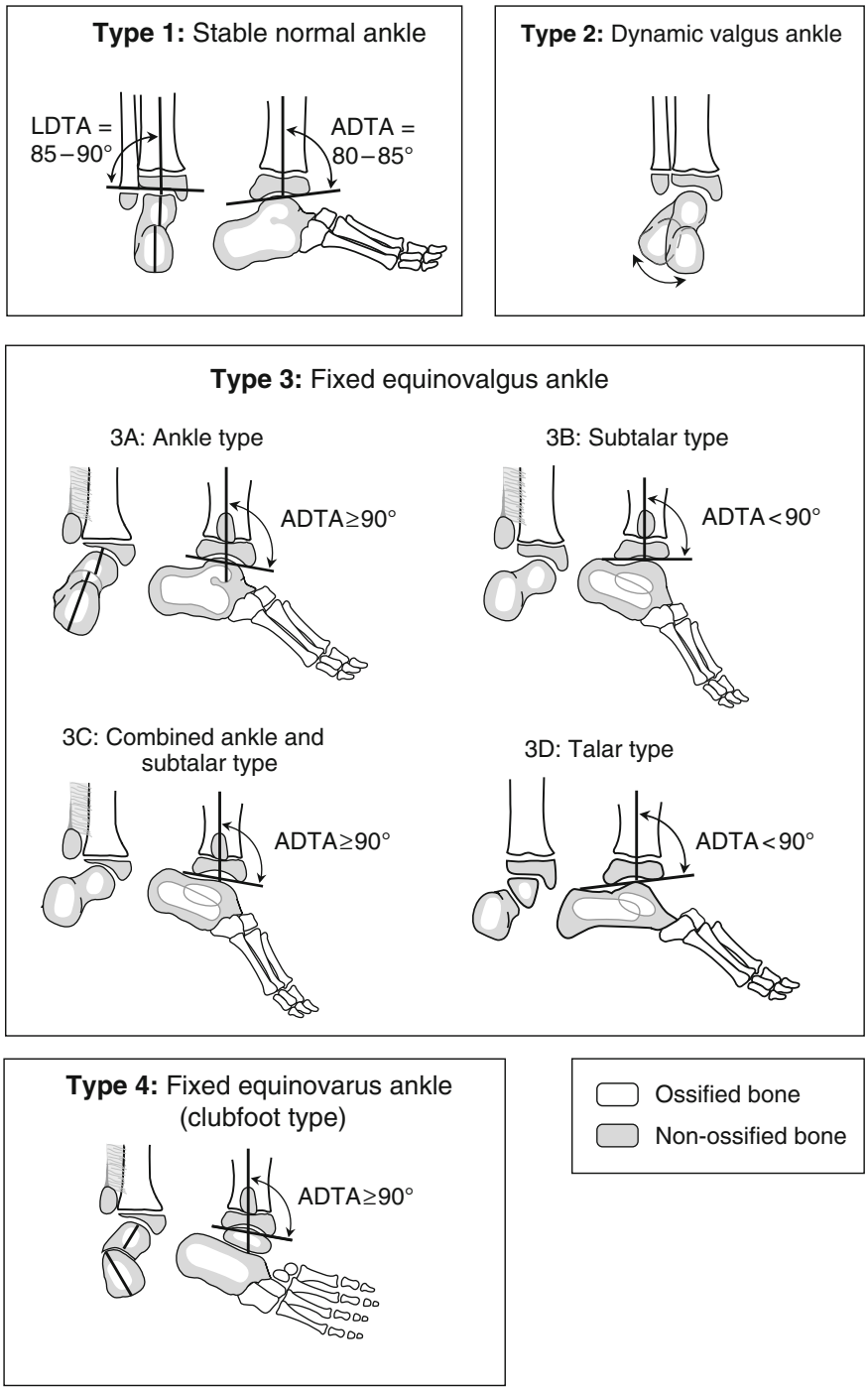


Fig. 23.6 Paley classification of FH. Type 1: Stable normal ankle. Type 2: Dynamic valgus ankle. Type 3: Fixed equinovalgus ankle. Type 3 is further subdivided according to the etiology of the valgus into 3A (ankle), 3B (subtalar), 3C (combined ankle and subtalar), and 3D (talar). Type 4: Fixed equinovarus ankle (clubfoot). Reprinted with permission from the Rubin Institute for Advanced Orthopedics, Sinai Hospital of Baltimore

Type 3a: Ankle type.
Type 3b: Subtalar type.
Type 3c: Combined ankle and subtalar type.
Type 3d: Talar body type.
Type 4: Fixed equinovarus ankle (clubfoot).

For Type 1, tibial lengthening is recommended, along with tendo Achilles lengthening. For Type 2, tibial lengthening is recommended, along with tendo Achilles lengthening and a supramalleolar reorientation osteotomy. For Type 3, soft tissue lengthening (peroneal tendons and tendo Achilles), resection of the fibrous anlage and interosseous membrane, and a reorientation osteotomy is recommended. This has been nicknamed the "Super-ankle procedure" and will be described later in the chapter. The osteotomy site varies according to whether the deformity is classified as Type 3a, 3b, 3c, or 3d. Type 3a needs a supramalleolar osteotomy, type 3b needs a subtalar osteotomy, Type 3c needs both

supramalleolar and subtalar osteotomy, and Type 3d, which is very rare, may be treated with an opening wedge osteotomy of the body of the talus. Type 4 (clubfoot type) is treated initially by applying Ponseti casts and performing an Achilles tenotomy followed by a "Super-ankle procedure" at age 12–24 months. The casting treatment typically converts the foot position from equinovarus to equinovalgus, which can then be addressed with the Super-ankle procedure. While the more severe Type 3 cases should also be considered candidates for amputation, Paley's classification is geared towards reconstruction, with the subtype designation dictating the specific reconstructive procedure.

Box 23.3. Paley Classification of FH

- Type 1: Normal ankle
- Type 2: Dynamic valgus ankle
- Type 3a: Fixed equinovalgus ankle, ankle type
- Type 3b: Fixed equinovalgus ankle, subtalar type
- Type 3c: Fixed equinovalgus ankle, combined ankle/subtalar type
- Type 3d: Fixed equinovalgus ankle, talar body type
- Type 4: Equinovarus type (clubfoot)

Clinical Assessment of a Child with FH

In our practice, a patient with fibular hemimelia is usually seen during the first year of life. In less than one-third of cases, the diagnosis has already been made using prenatal ultrasonography [22]. During the initial visit, full-length standing anteroposterior (AP) and lateral view radiographs are obtained to evaluate the exact configuration of the skeletal anatomy of the lower legs, to determine the amount of LLD, and to look for concurrent deformities such as congenital femoral deficiency. The child's height predication based on the long limb should be recorded. Based on the radiographic LLD (or the clinically measured LLD), the ultimate LLD at maturity can be predicted. We use the Multiplier Method [23, 24] to predict the ultimate LLD and ultimate overall height (based on the long leg) at skeletal maturity. This is easily done with the Multiplier mobile app (available for free on the Apple iTunes store for the Apple iOS platform and Google Play for the Android platform).

These two predictions (LLD and height) allow for formulation of a multistage treatment plan for the family. The family should leave the first visit with a clear picture of the pros and cons of the two main treatment arms: amputation versus reconstruction. Knowing the predicted adult height (based on the current length of the long leg) and the predicted LLD at skeletal maturity allows the surgeon to propose various treatment strategies. For example, if the Multiplier Method predicts an adult height for a boy to be 5′7″ (170 cm) with an LLD of 6 cm, then most families would choose limb lengthening as opposed to epiphysiodesis. However, if the same boy had a predicted height of 6′2″ (188 cm) with an LLD of 6 cm, then epiphysiodesis would become a more attractive option. For the reconstruction option, the family needs to be told if it will be necessary to perform foot/ankle reconstruction and how many lengthening procedures are anticipated. External fixation is the mainstay of lengthening in young children with FH [21]. In skeletally mature patients, it is possible to use the newer magnetic telescopic intramedullary (IM) nails for tibial lengthening [25, 26]. Combinations of lengthening and epiphysiodesis can also be done to minimize the number of lengthening procedures required.

The initial clinical exam should include a complete orthopedic examination of the child to look for associated anomalies. In the lower extremity, the number of rays and position of the foot and ankle are noted. Specifically, the ankle is examined to determine if it is mobile and well aligned or contracted into equinovalgus or equinovarus. The range of motion and stability of the hip, knee, and ankle are assessed. The knee joint is often somewhat unstable (ACL or PCL deficiency) and may be in excessive valgus alignment.

If the involvement of the hip and knee is severe (as in combined FH and congenital femoral deficiency), then the overall reconstruction plan is either concurrent hip/knee reconstruction or sequential reconstruction. Most commonly, the significantly involved hip and knee would be addressed first between the ages of 18 and 24 months (this is discussed at length in Chap. 22). The ankle reconstruction would then typically occur 6–12 months after the hip and knee reconstruction, although in some cases, it may be feasible to perform simultaneous hip, knee, and ankle reconstruction (Super-hip, Super-knee, and Super-ankle procedures).

Even if the femur is not involved or minimally involved, there still may be instability of the knee due to an absent ACL. In such cases, the preoperative full-length standing lateral view radiograph shows anterior subluxation of the tibia on the femur (Fig. 23.8). Consideration should be given to perform an intra-articular ACL substitution procedure prior to tibial lengthening. If the PCL is also deficient, then an extra-articular PCL reconstruction should be performed at the same time.

Radiographic Assessment of a Child with FH

The initial radiographic assessment should include plain films: full-length standing AP view radiograph depicting both legs with a lift under the short leg (Fig. 23.7) and a full-length standing lateral view of the affected leg with the knee in maximum extension (Fig. 23.8). The typical appearance in severe cases is a mild valgus deformity of the knee secondary

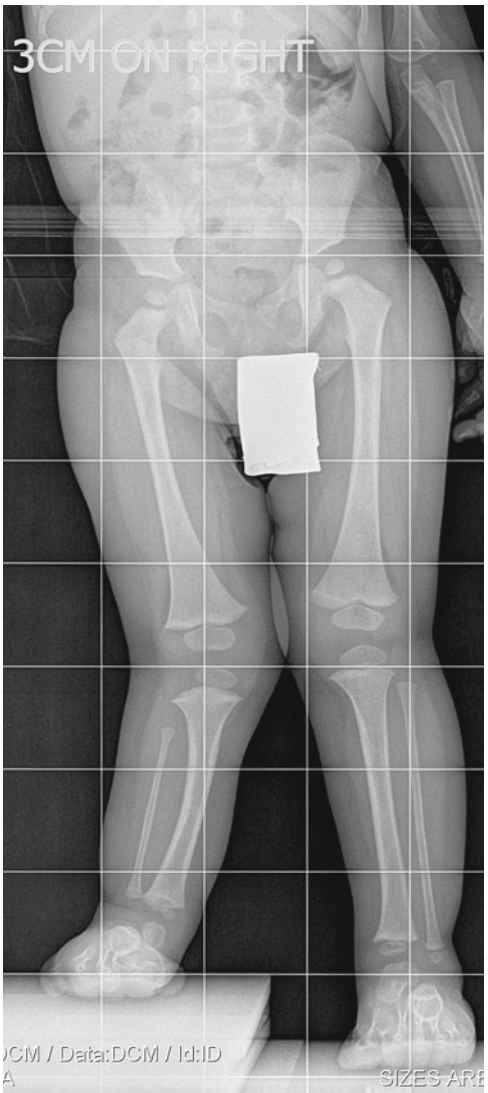


Fig. 23.7 Full-length standing AP view radiograph shows a typical child with FH who has knee valgus and tibial valgus bow. Reprinted with permission from the Rubin Institute for Advanced Orthopedics, Sinai Hospital of Baltimore

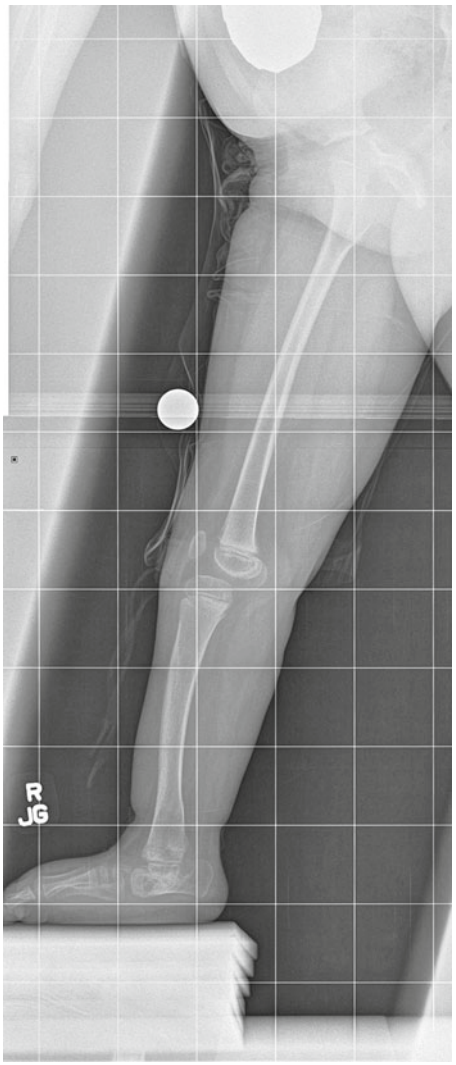


Fig. 23.8 Full-length standing lateral view radiograph shows a typical child with FH who has an apex anterior bow of the tibia and congenital instability of the knee due to the lack of functional intra-articular ligaments (ACL/PCL). Reprinted with permission from the Rubin Institute for Advanced Orthopedics, Sinai Hospital of Baltimore

to a hypoplastic lateral femoral condyle. This creates a mechanical lateral distal femoral angle (mLDFA) of less than 85°. Additional valgus may come from an apex anteromedial bow in the tibia. A dimple on the skin is usually visible over the apex of this bow (Fig. 23.9). A lateral view radiograph of the foot may show overlap of the talus and calcaneus or a talo-calcaneal tarsal coalition. If the talus and calcaneus are on top of one another, the ankle valgus is typically from a tilt in the distal tibial plafond. The talus and calcaneus may be stacked on top of each other on the lateral view, which points toward the ankle as the source of valgus (Fig. 23.10). If the talus and calcaneus are overlapped on the lateral view (Fig. 23.11) or side by side on the AP ankle (Fig. 23.12) view giving the appearance of a "double barrel shot gun," then the valgus component is more likely subtalar in origin. Most patients with FH have a talo-calcaneal coalition, which may not be evident on the initial radiographs obtained during infancy (Figs. 23.13 and 23.14) but become more obvious as the coalition ossifies with age.

In addition to plain films for cases of Paley Type 3 FH, it can be useful to obtain a magnetic resonance imaging (MRI) evaluation of the foot and ankle just prior to performing the Super-ankle procedure. Since much of the hindfoot and distal tibia is unossified during the first 2 years of life, it can be difficult to accurately classify the exact Paley Type 3 subtype. In such cases, an MRI can help differentiate between Type 3a, b, and c, thus allowing better preoperative planning (Figs. 23.15 and 23.16). Also, during surgery an arthrogram of the ankle joint can help delineate the valgus orientation at the ankle joint (Fig. 23.17).

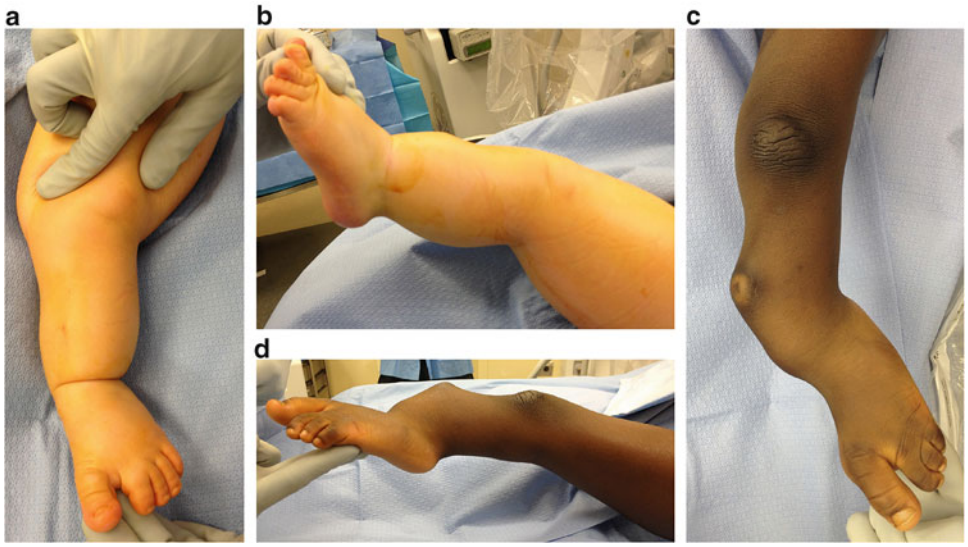


Fig. 23.9 (**a**, **b**) Clinical photos show a child with moderate deformity secondary to FH. The typical anterolateral bow, equinovalgus foot, and dimple at the apex of the tibial bow are shown. (**c**, **d**) Clinical photos show a child with severe deformity secondary to FH. Reprinted with permission from the Rubin Institute for Advanced Orthopedics, Sinai Hospital of Baltimore



Fig. 23.10 Lateral view radiograph of the foot shows the talus and calcaneus stacked on top of each other, suggesting the hindfoot valgus is supramalleolar in origin. Reprinted with permission from the Rubin Institute for Advanced Orthopedics, Sinai Hospital of Baltimore



Fig. 23.11 Lateral view radiograph of the foot shows the talus and calcaneus overlapped on top of each other, suggesting the hindfoot valgus is subtalar in origin. Reprinted with permission from the Rubin Institute for Advanced Orthopedics, Sinai Hospital of Baltimore

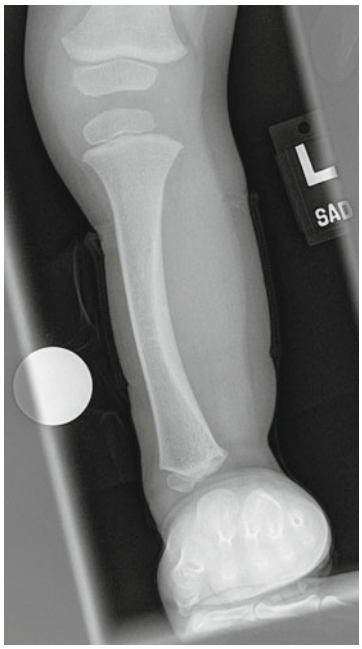


Fig. 23.12 AP view ankle radiograph shows the talus and calcaneus side by side, suggesting the hindfoot valgus is subtalar in origin. Reprinted with permission from the Rubin Institute for Advanced Orthopedics, Sinai Hospital of Baltimore

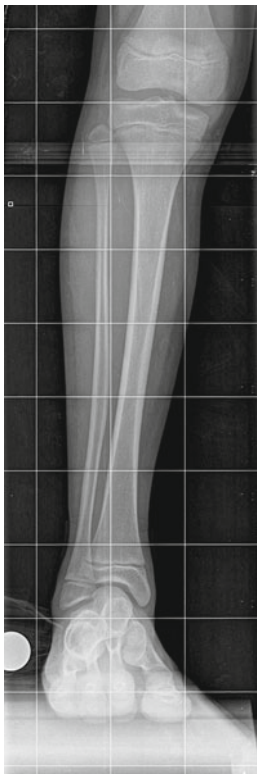


Fig. 23.13 AP view radiograph of the tibia, ankle, and foot in a typical child with FH shows a ball-and-socket ankle with a talocalcaneal coalition. As an infant, the talocalcaneal block is cartilaginous, but the talocalcaneal coalition becomes more radiographically evident as the child ages and the bones ossify. Reprinted with permission from the Rubin Institute for Advanced Orthopedics, Sinai Hospital of Baltimore



Fig. 23.14 Lateral view radiograph (same child depicted in Figure 23.13) shows the classic "C" sign of the talocalcaneal coalition and a concurrent talonavicular coalition. Reprinted with permission from the Rubin Institute for Advanced Orthopedics, Sinai Hospital of Baltimore

Principles of Treatment

One must understand that the initial reconstruction is a very complex set of procedures that is unique for each child. The deformity related to fibular hemimelia is the combination of contracted soft tissues and abnormally formed joints of the ankle and foot. The treatment of FH should address several issues such as limb shortening, lower leg deformity, and an abnormally positioned foot with or without absence of lateral rays. The goals of treatment are to create a lower limb with a stable hip, knee, and ankle by:

1. Correcting the foot (usually equinovalgus) into a plantigrade position.
2. Equalizing lower limb lengths by skeletal maturity.
3. Correcting lower limb malalignment.

In many of the more severe cases, it may not be possible to improve the limited ankle mobility. Furthermore, the treatment itself (multiple lengthening procedures or Super-ankle reconstruction plus multiple lengthening procedures) may result in further loss of ankle motion compared to the amount present initially. The success of reconstruction is not necessarily determined by the final ankle motion. The amount of final ankle motion is usually predetermined by the amount of motion initially present at the ankle joint.

The amount of LLD predicted by the end of growth does not necessarily determine whether a successful reconstruction is possible. There are potentially no set limits to the

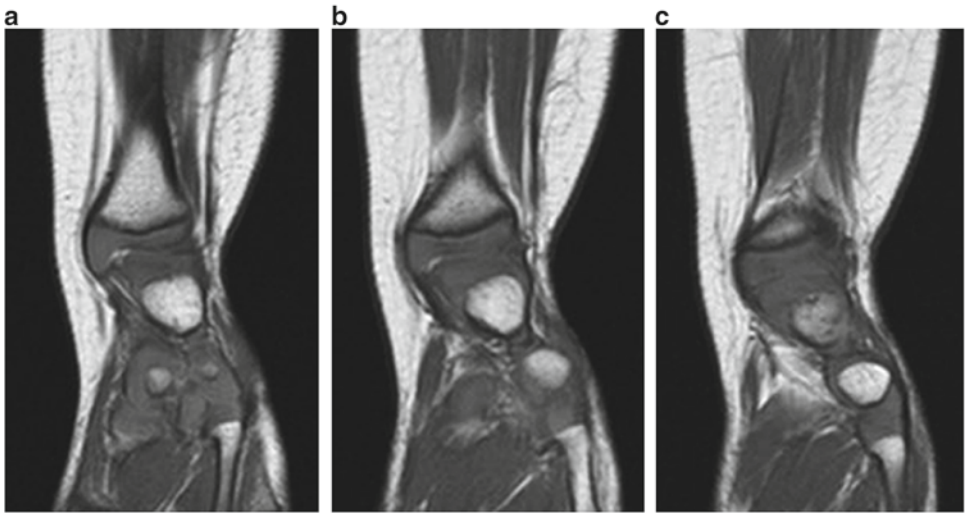


Fig. 23.15 (**a**–**c**) Sequential MRI scans of the left lower extremity in a patient with fibular hemimelia show that the valgus originates in the supramalleolar region (Paley Type 3A—ankle type). Reprinted with permission from the Rubin Institute for Advanced Orthopedics, Sinai Hospital of Baltimore

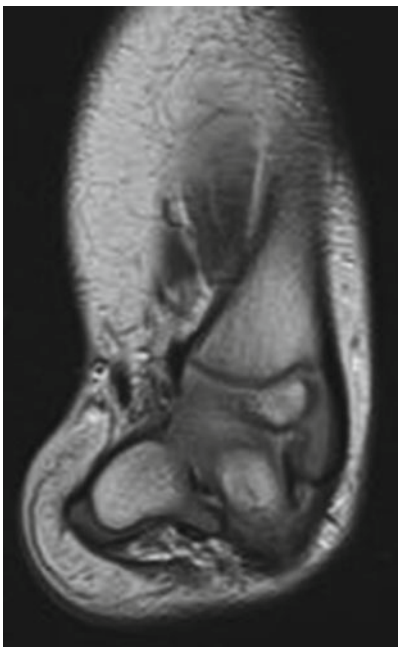


Fig. 23.16 MRI scan of the right lower extremity in a patient with fibular hemimelia shows that the hindfoot valgus originates in the subtalar joint (Paley Type 3B). Reprinted with permission from the Rubin Institute for Advanced Orthopedics, Sinai Hospital of Baltimore

amount of overall lengthening that can be performed, especially when divided into small steps of approximately 5 cm for each treatment. However, the overall predicted LLD does help when estimating the number of lengthening treatments that will be needed to equalize the limb lengths before skeletal maturity. Dividing the amount of lengthening into smaller amounts might be less traumatic for the muscles, nerves, and adjacent joints than trying to perform a heroic lengthening during one treatment. For example, if the predicted discrepancy is 15 cm, this may be better addressed with three 5-cm lengthening treatments instead of two 7.5-cm lengthening treatments. Better yet, two 5-cm lengthening treatments combined with one 5-cm epiphysiodesis may yield the optimum outcome.

For children with Paley Type 3 (fixed equinovalgus deformity), the decision to lengthen during the first reconstructive surgery is determined by the amount of ankle motion present. If the ankle is intrinsically stiff, then a 4-to-5-cm lengthening is performed at the same surgical setting as the Super-ankle procedure. If the ankle is relatively mobile, then another option is for the initial surgery to concentrate on positioning the foot and ankle in a stable or corrected position while concurrently correcting the bowing in the lower leg with a fixator for a minimal amount of lengthening (just enough to provide correction of the apex anteromedial bow). This strategy may help preserve ankle mobility because the wires used for the Super-ankle procedure cross the ankle joint for approximately only 3 months for this limited lengthening instead of 6 months for a more typical 4-to-5-cm lengthening.

The number of subsequent lengthening treatments is determined by the overall predicted lengthening goal. The subsequent lengthening treatments are performed at intervals of 4–6 years apart for a total of up to three or even four lengthening treatments for the most severe grades of FH. The first lengthening usually achieves a 4 to 5 cm gain in length. Subsequent lengthenings can achieve between 5 and 7 cm of length each.

The amount of length gained during a single lengthening treatment is determined by the rate and quality of regenerate bone formation and by motion and stability of the joints above and below the lengthening site. Bone formation is better in the proximal metaphysis rather than in the diaphysis. This is another reason why it may be preferable not to perform a large amount of lengthening at the time of the initial Super-ankle procedure. Development of a knee flexion contracture during lengthening may dictate the need to stop further lengthening. The amount of lengthening will also be limited if the patient has poor regenerate bone quality or

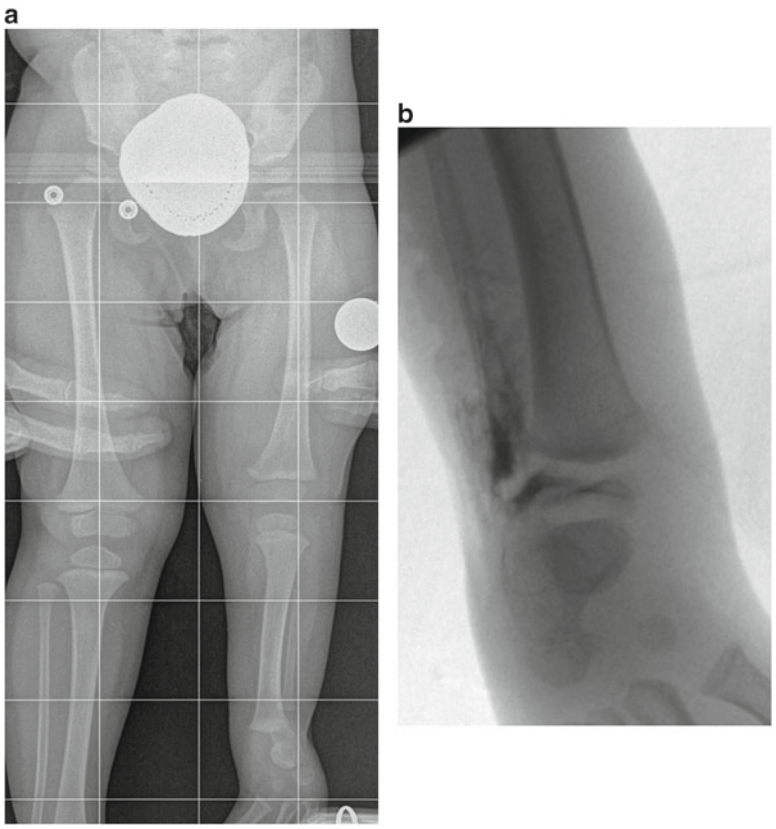


Fig. 23.17 (**a**) Supine full-length radiograph of an 18-month-old girl with left fibular hemimelia shows typical equinovalgus of the foot and ankle. (**b**) Intraoperative AP ankle arthrogram of the same patient shows the distal tibia as the source of the ankle valgus. Reprinted with permission from the Rubin Institute for Advanced Orthopedics, Sinai Hospital of Baltimore

stiffness of the joints with loss of motion. In patients younger than 6 years of age, the maximum length typically gained during a single lengthening is 5 cm. The total amount of lengthening in a younger patient is limited to avoid excessive pressure on the growth plates that can cause premature closure of the growth plates and loss of potential natural growth. Older patients can often tolerate gaining more length in a single treatment (between 5 and 7 cm).

Box 23.4. FH Treatment Options

- Amputation
- Lengthening with external fixator
- Lengthening over nails
- Lengthening with internal telescopic nails
- Foot/ankle reconstruction for dynamic valgus
- Super-ankle for rigid equinovalgus

Surgical Techniques for FH

Lengthening for Paley Type 1

Mild cases of FH may be lengthened using standard limb lengthening methodology, which is described in Chap. 7 on limb lengthening by Dr. Feldman and coauthors.

All children (and adults) with FH have a certain degree of tightness of the Achilles tendon, which will be made worse with limb lengthening. Therefore, we recommend performing a triceps surae lengthening procedure at the time of tibial lengthening. We recommend an open Vulpius procedure (Fig. 23.18) and a prophylactic anterior compartment fasciotomy (Fig. 23.19). The fasciotomy decreases the risk of compartment syndrome (a known complication of tibial osteotomy). An additional benefit of anterior compartment fasciotomy (especially if the longitudinal incision in the fascia is combined with a short cruciate incision [transverse]) is that it allows the anterior compartment musculature to bulge out, thus creating the impression of a larger diameter calf. This is desirable, as all patients with FH have a smaller ipsilateral calf due to combined bone and soft tissue hypoplasia.

The fixator should be extended to include the foot to prevent equinus deformation of the ankle during lengthening. Once the lengthening is complete, the foot frame may be removed provided there is no flexion contracture of the knee. We favor circular external fixators over monolateral external fixators for their superior ability to correct deviations that may occur during lengthening (typically procurvatum and valgus) and for the ability to insert fixation pins and wires from multiple directions, creating a more stable construct. Circular frames are generally well tolerated in the tibia and are easy to bridge across the ankle to the foot. Figure 23.20

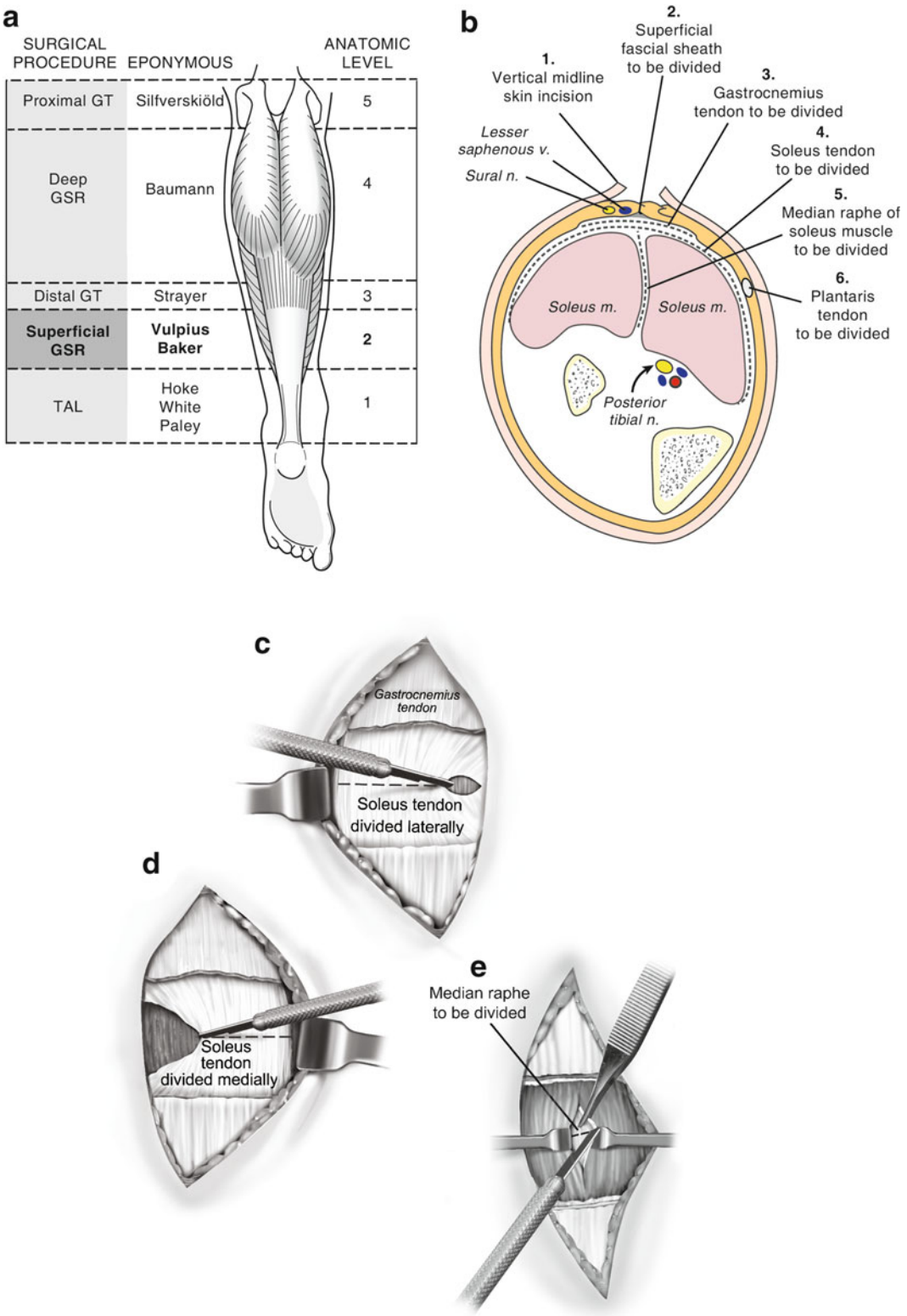


Fig. 23.18 Open Vulpius procedure is recommended for all Paley Type 1 and 2 FH lengthening procedures. (**a**) Posterior leg can be divided into five anatomic levels. (**b**) Cross-sectional view of anatomic level 2 with steps to perform gastrocnemius–soleus recession numbered sequentially. The *broken lines* represent the tendon portion of the gastrocnemius and soleus muscles and the median raphe of the soleus muscle. (**c**) After midline vertical incision, vertical division of the superficial fascia, and horizontal release of the gastrocnemius tendon, then the soleus tendon is released. (**d**) Soleus tendon is also released horizontally to the opposite edge (medially) of the tendon using deep retraction. (**e**) The median raphe (central tendon) of the soleus muscle is located and released. *GSR* gastrocnemius–soleus recession, *GT* gastrocnemius tenotomy, *m* muscle, *n* nerve, *TAL* tendo Achilles lengthening, *v* vein. Figure and legend reprinted with permission from the American Podiatric Medical Association. Lamm BM, Paley D, Herzenberg JE. Gastrocnemius soleus recession: a simpler, more limited approach. J Am Podiatr Med Assoc 2005; 95:18–25

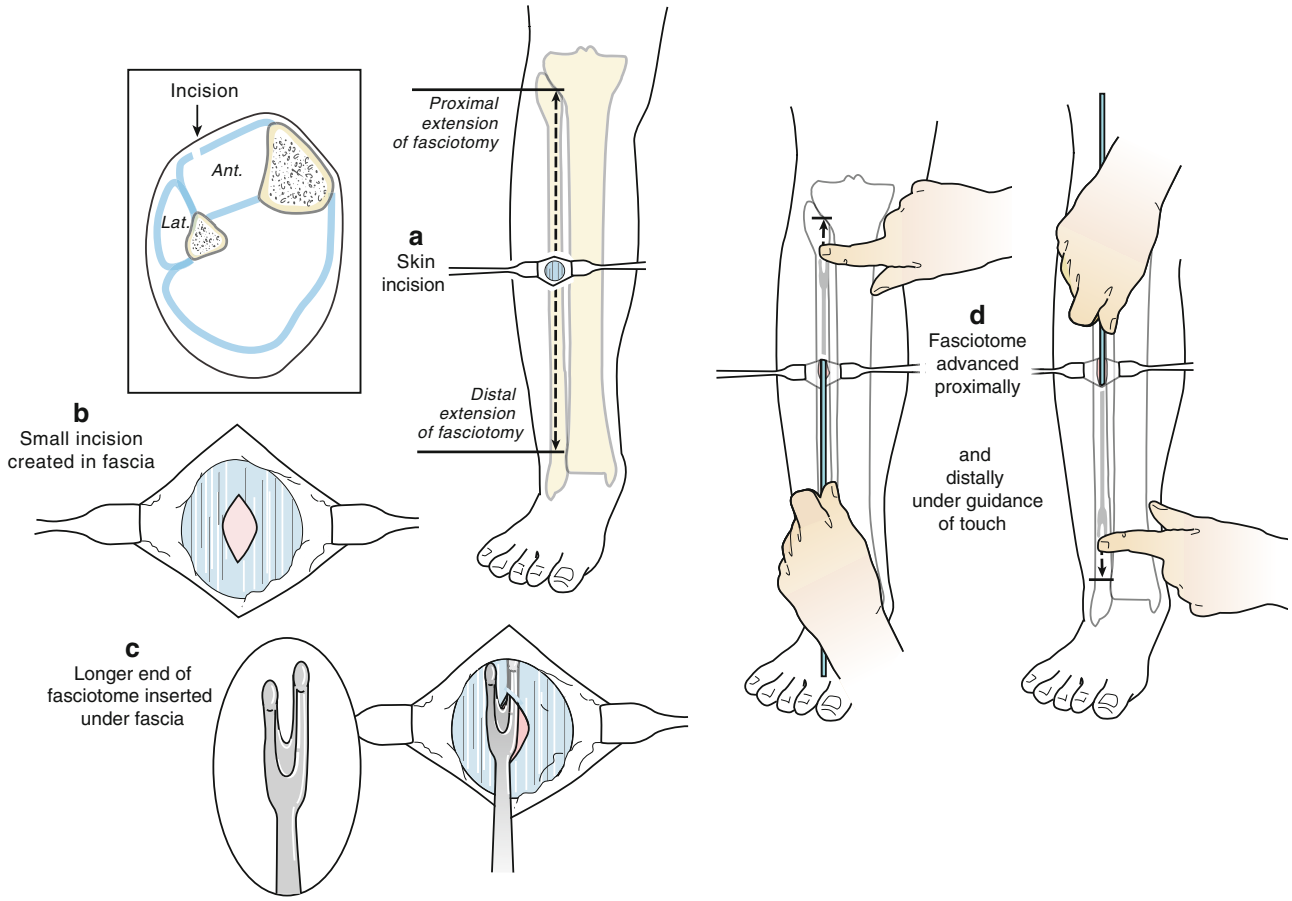


Fig. 23.19 Prophylactic anterior compartment fasciotomy is recommended for all FH lengthenings. This decreases the risk of post-osteotomy compartment syndrome and also causes a bulge of the anterior compartment musculature, increasing the circumference of the calf (a positive effect for a hypotrophic calf). (**a**) Small midline anterior longitudinal incision is made just lateral to the tibial crest. (**b**) Fascia is exposed using blunt dissection, and a small incision is created. (**c**) Fasciotome is then inserted under the fascia. (**d**) Fasciotome is advanced proximally, and care is taken not to invade the muscle. Note that the opposite index finger follows the leading edge of the fasciotome. *Ant.* anterior compartment, *Lat.* lateral compartment. Figure and legend reprinted with permission from the Rubin Institute for Advanced Orthopedics, Sinai Hospital of Baltimore

shows a standard FH lengthening with external fixation. In pure external fixation cases, it is typical to use three pins/wires proximally, use two pins/wires in the distal segment, and include foot fixation. A wide space should be left between the proximal and distal rings to allow for better recruitment of soft tissue into the lengthening.

For skeletally mature patients in whom there is no longer a viable proximal tibial growth plate, it is attractive to use IM nails, either in the form of Lengthening over Nail (LON) or in the form of implantable IM telescopic nails. The prototype nail, and the only one FDA approved and marketed in the USA currently, is the PRECICE system (Ellipse Technologies, Irvine, CA). This telescopic nail is magnetically powered, accurately controllable, and can go both in forward and in reverse (in case of over lengthening or in case of poor bone formation). With the LON technique, a standard trauma nail (8.5- to 10-mm diameter) is inserted and over reamed by 2 mm to allow the bone to slide over the nail. The nail is locked proximally. The osteotomy must be proximal enough (and the nail long enough) so that at the end of lengthening, there is at least 5 cm of nail distal to the osteotomy (for stability). An important technical point is to drill holes in the anterior aspect of the tibia prior to reaming to vent the canal and to encourage the reamings to exit anteriorly to stimulate new bone formation at the lengthening site. The external fixator that is applied for the LON technique can be simple, with two points of fixation at either end. The rate of lengthening is adjusted according to bone formation. At the end of lengthening, the patient returns to the operating room for distal locking and frame removal (in that order, to prevent shortening). The bulk of the consolidation then takes place under the protection of the nail, without need for an external fixator. In order to control the foot during IM lengthening or LON, a temporary extra-articular ankle arthrodesis screw may be inserted from the back of the calcaneus into the distal tibia [26]. This screw is removed at the end of lengthening to

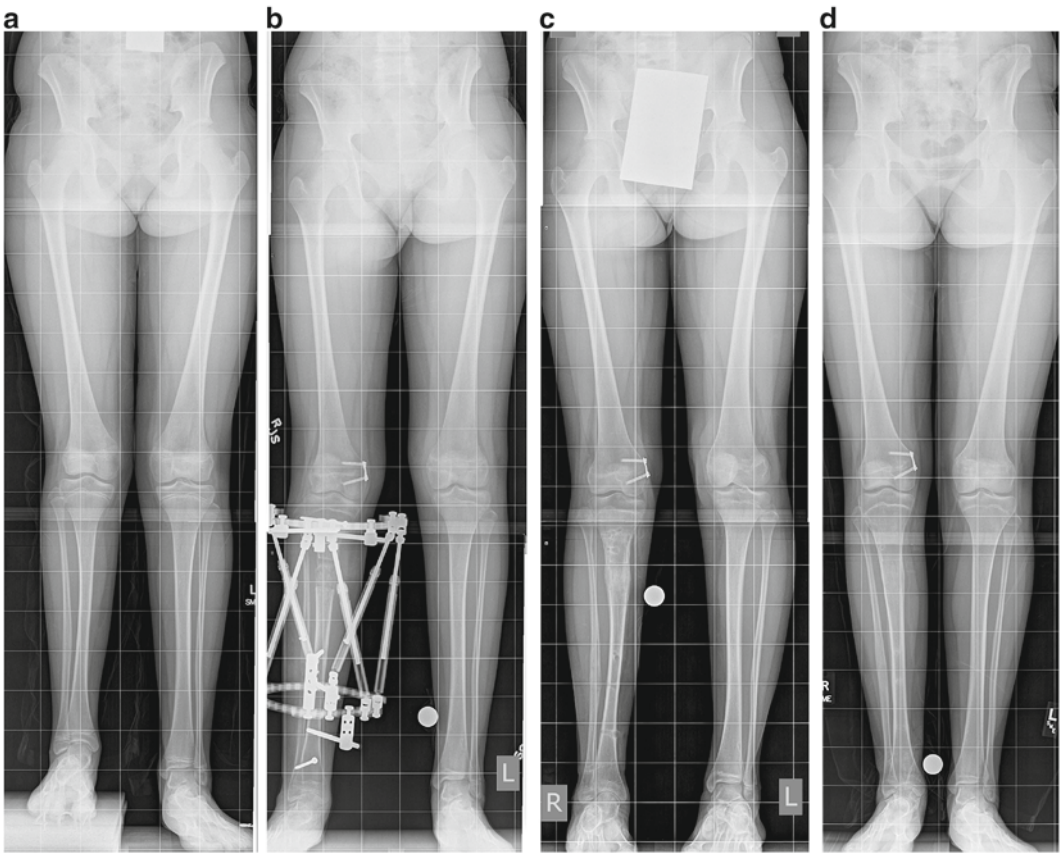


Fig. 23.20 Full-length standing AP view radiographs of a 12-year-old girl with FH before treatment (**a**), after distal femoral hemiepiphysiodesis and application of circular external fixation for lengthening (**b**), after removal of external fixation once lengthening and consolidation were achieved (**c**), and after full correction of distal femoral valgus with a normalized mechanical axis (**d**). Reprinted with permission from the Rubin Institute for Advanced Orthopedics, Sinai Hospital of Baltimore

allow mobilization of the ankle. Figure 23.21 shows an FH lengthening over nail to decrease duration of treatment time with the external fixator (also discussed in Chap. 8).

The technique for self-lengthening telescopic nails is similar to that described above for LON, except that the nail is locked proximally and distally and no external fixator is needed. Blocking screws (Poller screws) can be used to prevent mechanical axis deviations during lengthening. Figure 23.22 shows an FH lengthening with a PRECICE intramedullary lengthening nail, including the extra-articular screw technique to stabilize the ankle (IM lengthening nails are also discussed in Chap. 9).

Box 23.5. Lengthening for Moderate FH

- Lengthen Achilles/Vulpius
- Anterior compartment fasciotomy
- Transfix foot during lengthening
- Options: External fixation, LON, telescopic nails

Lengthening Plus Ankle Realignment for Paley Type 2

In this subset of patients, the recommendations for lengthening are largely identical to those described for Type 1. However, in these cases, the ankle has a flexible valgus deformity. This will worsen with limb lengthening as the peroneal tendons tighten. For such cases, we recommend intramuscular lengthening of the peroneal tendons and bony realignment of the ankle (Fig. 23.23).

Lengthening Plus Super-Ankle Reconstruction for Paley Type 3

For the rigid equinovalgus type of FH, we use a modified Paley Super-ankle approach. This surgery can be performed when a child is as young as 1 or 2 years of age. The goal is to realign the foot relative to the distal tibia and to make it plantigrade. It is an extra-articular reconstruction, as the ankle joint and its ligaments are not opened.

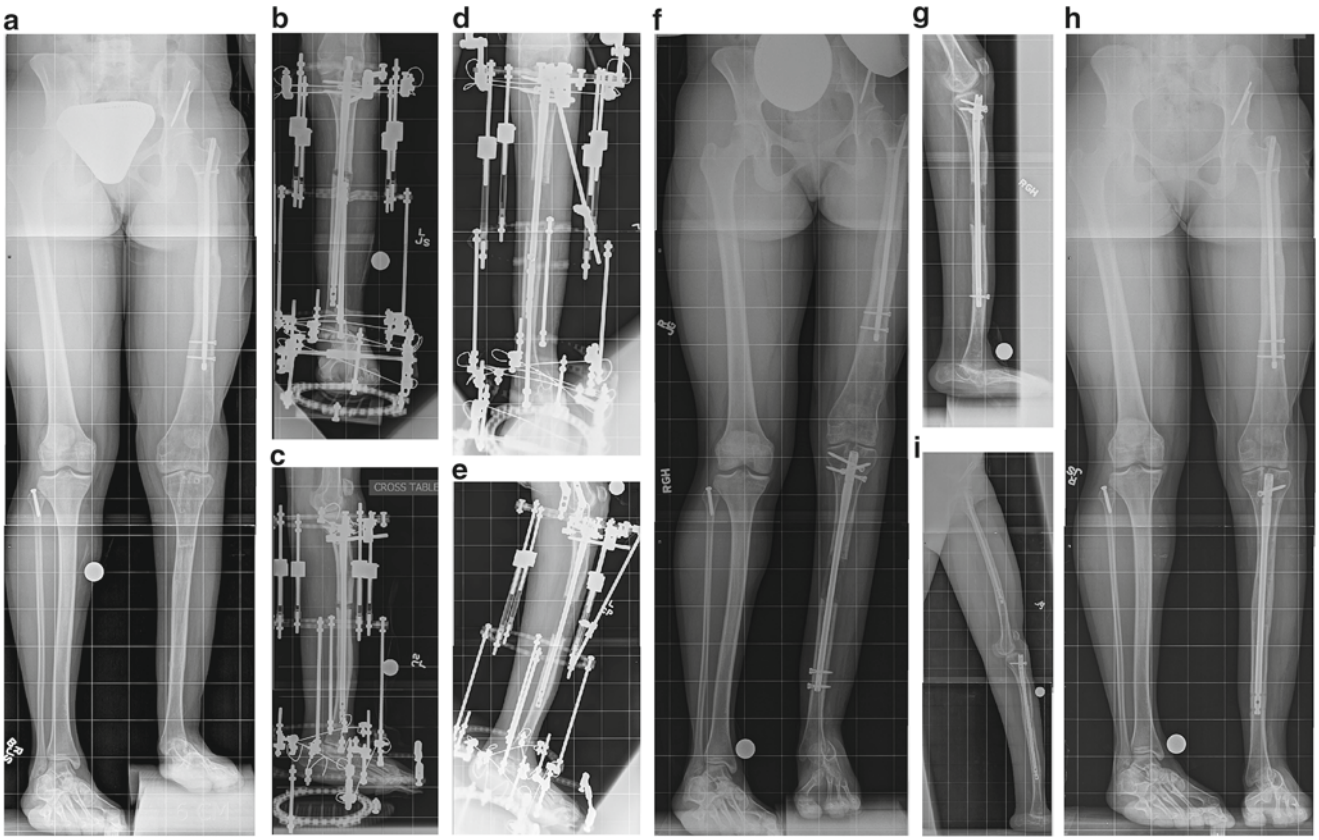


Fig. 23.21 (**a**) Preoperative full-length standing AP view radiograph shows a 16-year-old girl with left congenital femoral deficiency and fibular hemimelia prior to tibial lengthening with lengthening over nail (LON) technique. (**b**, **c**) One week postoperative AP (**b**) and lateral (**c**) view radiographs of the left tibia show the LON technique using an intramedullary tibial nail and concurrent classic Ilizarov circular external fixation device. (**d**, **e**) AP (**d**) and lateral (**e**) view radiographs of the left tibia after completion of the distraction phase and prior to external fixation removal. (**f**, **g**) Full-length standing AP view radiograph (**f**) and lateral view tibial radiograph (**g**) after locking of the intramedullary nail and removal of the external fixation device for the consolidation phase. (**h**, **i**) Final full-length standing AP (**h**) and lateral (**i**) view radiographs show complete healing of the tibial lengthening segment. After three of the four cortices had healed, the distal locking screws were removed for dynamization to improve the anterior cortical thickness. Reprinted with permission from the Rubin Institute for Advanced Orthopedics, Sinai Hospital of Baltimore

Surgical Technique for Type 3a (Figs. 23.24 and 23.25)

Positioning

Supine position, bump under the ipsilateral sacrum, and tourniquet control.

Surgical Approach

Longitudinal (straight) incision starts laterally at the mid-calf and ends just above the sole of the foot. Peroneal tendons (sometimes only one is present) are cut with a Z-technique and later repaired in the lengthened configuration. Distal anlage of the fibula, if present, may be left in place. Anterior compartment fasciotomy is performed, and the superficial peroneal nerve is protected. The anterior compartment muscles are dissected off of the cartilage analage of the fibula and off of the intermuscular septum.

The intermuscular septum is resected from the ankle proximally up to the mid portion of the tibia at the apex of the anteromedial bow. This resected intermuscular septum should be placed in a sterile specimen cup and covered with saline, as it may be recycled for repair and elongation of the Achilles tendon as needed. (Paley's original technique described resection of the anlage up to the proximal tibia and decompression of the common peroneal nerve through a separate, proximal incision. We feel that this step is unnecessary.)

Next, the sural nerve is identified and protected. The Achilles tendon and posterior tibial neurovascular bundle are identified through this posterolateral approach. Be careful not to damage the posterior tibial nerve, which is juxtaposed to the Achilles tendon. The flexor hallucis longus is identified as a guide to the anatomy of the posterolateral corner. The posterior tibial nerve is decompressed as far as can be seen into the tarsal tunnel distally through this posterolateral approach. The Achilles tendon is very short and can be made longer for purposes of Z-lengthening by scraping the distal muscle belly proximally to expose more of the tendon.



Fig. 23.22 Case example of a 14-year-old girl with fibular hemimelia and congenital femoral deficiency who underwent lengthening with the PRECICE. (**a**, **b**) Preoperative full-length standing AP (**a**) and lateral (**b**) view radiographs show that the child had an 8-cm limb length discrepancy and concurrent genu valgus. (**c**, **d**) Full-length standing AP (**c**) and lateral (**d**) view radiographs depict lengthening with the PRECICE magnetic intramedullary telescopic nail. This device contains a small magnet, which will gradually lengthen when activated by an externally applied magnetic field. The PRECICE is also reversible and controllable to a very accurate degree. (**e**, **f**) Full-length standing AP (**e**) and lateral (**f**) view radiographs obtained at the completion of lengthening. (**g**, **h**) Final full-length standing AP (**g**) and lateral (**h**) view radiographs obtained after nail removal. Note the equal limb lengths and the correction of the mechanical axis. Reprinted with permission from the Rubin Institute for Advanced Orthopedics, Sinai Hospital of Baltimore

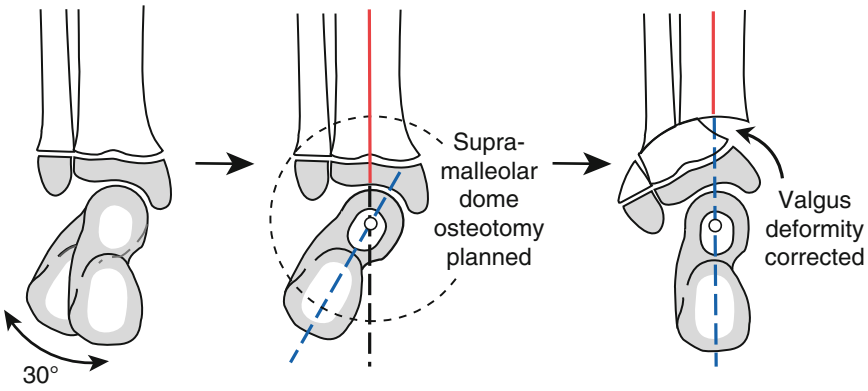


Fig. 23.23 Illustration shows bony realignment of the valgus ankle for cases of Paley Type 2. Reprinted with permission from the Rubin Institute for Advanced Orthopedics, Sinai Hospital of Baltimore

Despite this, it is often necessary to fashion a tendon graft (from the resected intermuscular septum) to obtain sufficient elongation of the Achilles tendon. The foot is then placed into the maximum equinus and valgus position, and two 1.8-mm diameter Kirschner wires are inserted from the plantar surface of the heel, across the calcaneus and talus, and across the ankle joint into the distal tibia about 5–10 mm. This pins the ankle in the maximum valgus/equinus position. At this stage of the operation, these pins do not cross the intended site of the supramalleolar osteotomy.

Osteotomy

Next, a subperiosteal dissection is performed of the distal tibia, being careful not to damage the perichondrial ring of the growth plate. Mini-Hohmann retractors are placed anteriorly and posteriorly. An oscillating saw is used to make an oblique cut from posterolateral to anteromedial, but the anteromedial cortex intact is left intact. The osteotomy is levered open from the posterolateral direction with an osteotome or laminar spreader. This maneuver corrects the equinovalgus position of the hindfoot. The osteotomy is held open with a cortical allograft (small piece of fibular allograft works well) and the two Kirschner wires are passed across the osteotomy and up into the distal half of the tibia, stopping just short of the anteromedial bow in the apex of the diaphysis. The Kirschner wires are bent outside the skin of the heel and can be incorporated into the frame (to be applied shortly) or simply bent and left outside the skin for removal when the frame is removed. The foot is now corrected and plantigrade relative to the distal half of the tibia. The tourniquet can be let down for the closure. The Achilles and peroneal tendons are repaired in their lengthened configuration, and the incision is closed, over a small drain if necessary. At this point, if desired, the operation can conclude and a long leg cast can be applied. The cast and pins are removed in 6 weeks, and then the patient can receive physical therapy for the ankle. If necessary, the tibial diaphyseal bow is corrected through a mid-diaphyseal osteotomy. In severely shortened, angulated cases, the surgeon may elect to perform a closing wedge osteotomy and remove the entire segment between the two osteotomies, thus performing a closing trapezoidal wedge osteotomy. This is only done when the diaphyseal deformity is in very close proximity to the ankle.

Frame Application

Six months after the initial procedure, the tibia can be lengthened with an Ilizarov external fixator through an osteotomy made at the apex of the anteromedial bow. Alternatively, the Ilizarov frame may be applied immediately and used to correct the apex anteromedial bow with minimal lengthening (1–2 cm). This frame is removed after healing of the diaphyseal corticotomy is complete, and a long leg cast is applied for 3–4 weeks followed by mobilization and rehabilitation of the ankle to try to regain as much motion as possible.

Surgical Technique for Type 3b (Figs. 23.26 and 23.27)

Positioning and Surgical Approach

The positioning and approach are identical to what is described previously for Type 3a.

Osteotomy

However, instead of a supramalleolar osteotomy, an oblique, U-shaped osteotomy of the subtalar region is performed. The cartilaginous anlage is discarded, divided, or dissected around. After the osteotomy is complete (need to be able to fully mobilize the hindfoot), the Kirschner wires are initially inserted the same way with the foot in the maximally uncorrected position (equinus and valgus). Then the distal half of the calcaneus and foot is translated medially and angulated to make the Kirschner wires now face directly upwards toward the talus. This reduces the valgus lateral translation of the foot, which is dorsiflexed to create an open wedge posteriorly for bone graft placement to correct the equinus

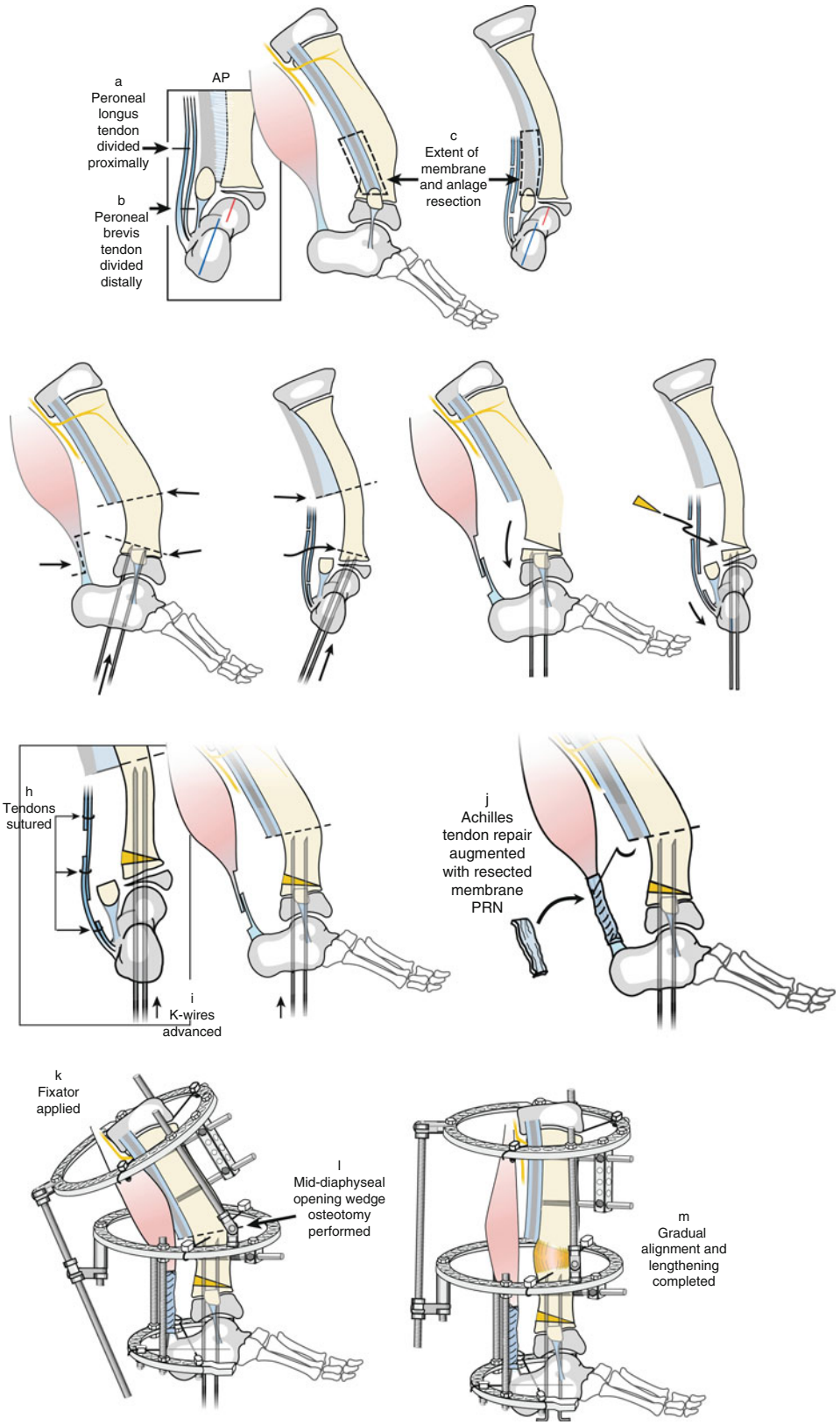


Fig. 23.24 (**a**–**m**) Super-ankle surgical technique used to treat supramalleolar type fibular hemimelia (Paley Type 3A—ankle type). Reprinted with permission from the Rubin Institute for Advanced Orthopedics, Sinai Hospital of Baltimore

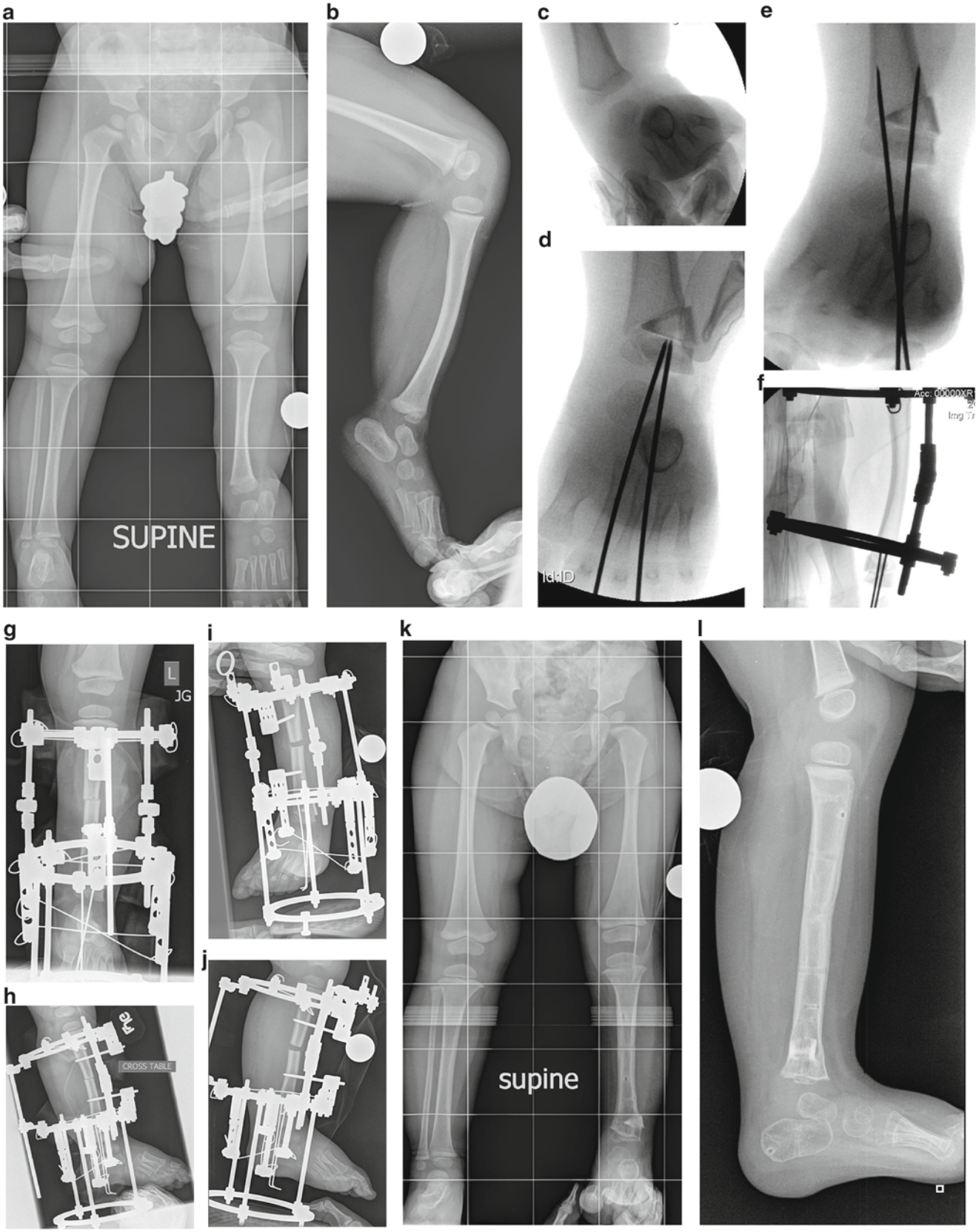


Fig. 23.25 Case example of an 18-month-old boy with Paley Type 3A fibular hemimelia. (**a**, **b**) Preoperative AP (**a**) and lateral (**b**) view radiographs depict the typical equinovalgus foot and ankle deformity along with the anterior tibial bow. (**c**–**e**) Intraoperative fluoroscopic images show the distal tibial osteotomy portion of the Super-ankle procedure (comprehensive reconstruction). (**f**) Intraoperative fluoroscopic image shows the application of the pediatric Ilizarov external fixator with the hinges positioned at the apex of the deformity on the convex side to produce both straightening and lengthening. (**g**, **h**) Immediate postoperative radiographs obtained after the completion of the Super-ankle procedure. (**i**, **j**) Postoperative radiographs obtained 2 weeks after Super-ankle procedure show correction of the midshaft tibial deformity along with limited bone lengthening. (**k**, **l**) AP (**k**) and lateral (**l**) view radiographs obtained at the time of external fixation removal (3 months after Super-ankle procedure). Note the complete correction of the tibia and the ankle. Reprinted with permission from the Rubin Institute for Advanced Orthopedics, Sinai Hospital of Baltimore

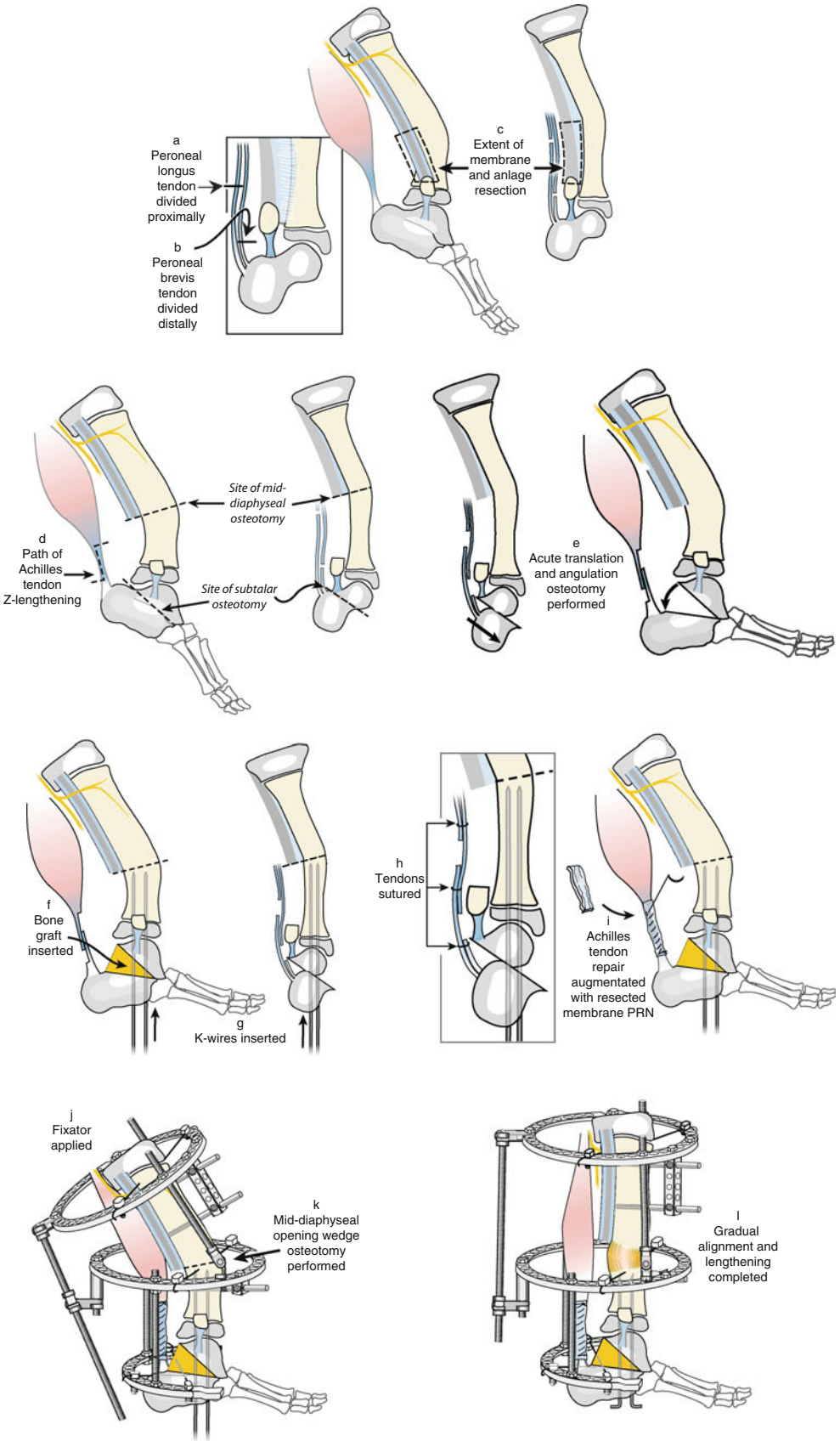


Fig. 23.26 (**a**–**l**) Super-ankle surgical technique used to treat subtalar type fibular hemimelia (Paley Type 3B). Reprinted with permission from the Rubin Institute for Advanced Orthopedics, Sinai Hospital of Baltimore

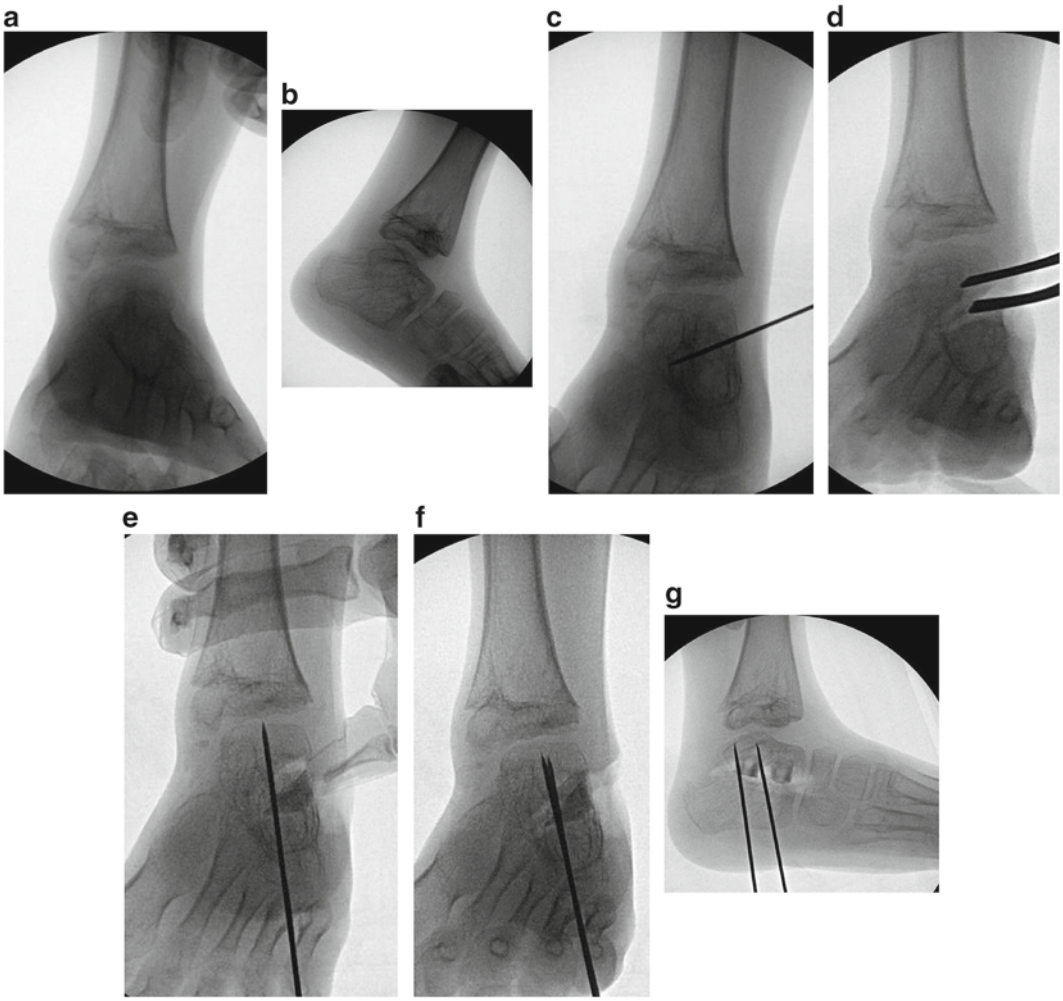


Fig. 23.27 Case example of a 6-year-old boy with left fibular hemimelia and residual hindfoot valgus secondary to a deformity in the subtalar region (Paley Type 3B). The osteotomy is performed through the talocalcaneal coalition. (**a**, **b**) AP (**a**) and lateral (**b**) view fluoroscopic images show residual hindfoot valgus after a previous supramalleolar osteotomy at 2 years of age. (**c**) Fluoroscopic image shows a guidewire inserted into the region of the subtalar coalition demonstrating the oblique osteotomy. (**d**) The subtalar osteotomy is completed with an osteotome and translated medially with concurrent varus angulation with a laminar spreader instrument. (**e**) Fluoroscopic image of fibular allograft wedge position with the surgeon's thumb maintaining medial translation of the calcaneal fragment and initial wire fixation. (**f**, **g**) Final fluoroscopic views of the subtalar osteotomy with medial translation and varus angulation of the calcaneal fragment stabilized with fibular allograft wedges and percutaneous wire fixation. Reprinted with permission from the Rubin Institute for Advanced Orthopedics, Sinai Hospital of Baltimore

position of the calcaneus. The smooth wires are driven across the osteotomy into the talus. In some cases, sufficient purchase is available in the talus, but for very young children, the wires should be driven across the ankle joint into the distal tibia for better purchase. At this point, the tourniquet is let down, and the closure proceeds as described above, including external fixator application and corticotomy of the mid-diaphyseal deformity.

Surgical Technique for Type 3c (Fig. 23.28)

Positioning and Surgical Approach

The positioning and approach are identical to what is described previously for Type 3a.

Osteotomy

Both a supramalleolar osteotomy and a hindfoot subtalar coalition osteotomy are performed, opened (supramalleolar), translated, angulated (subtalar), and held open with bone graft and transfixion Kirschner wires. Intraoperative arthrography of the ankle joint helps to determine the degree of ankle valgus and whether it needs to be corrected or if only a subtalar osteotomy is sufficient. If necessary, the tibial diaphyseal bow is corrected through a mid-diaphyseal osteotomy. In severely shortened, angulated cases, the surgeon may elect to perform a closing wedge osteotomy and remove the entire segment between the two osteotomies, thus performing a closing trapezoidal wedge osteotomy. This is only done when the diaphyseal deformity is in very close proximity to the ankle.

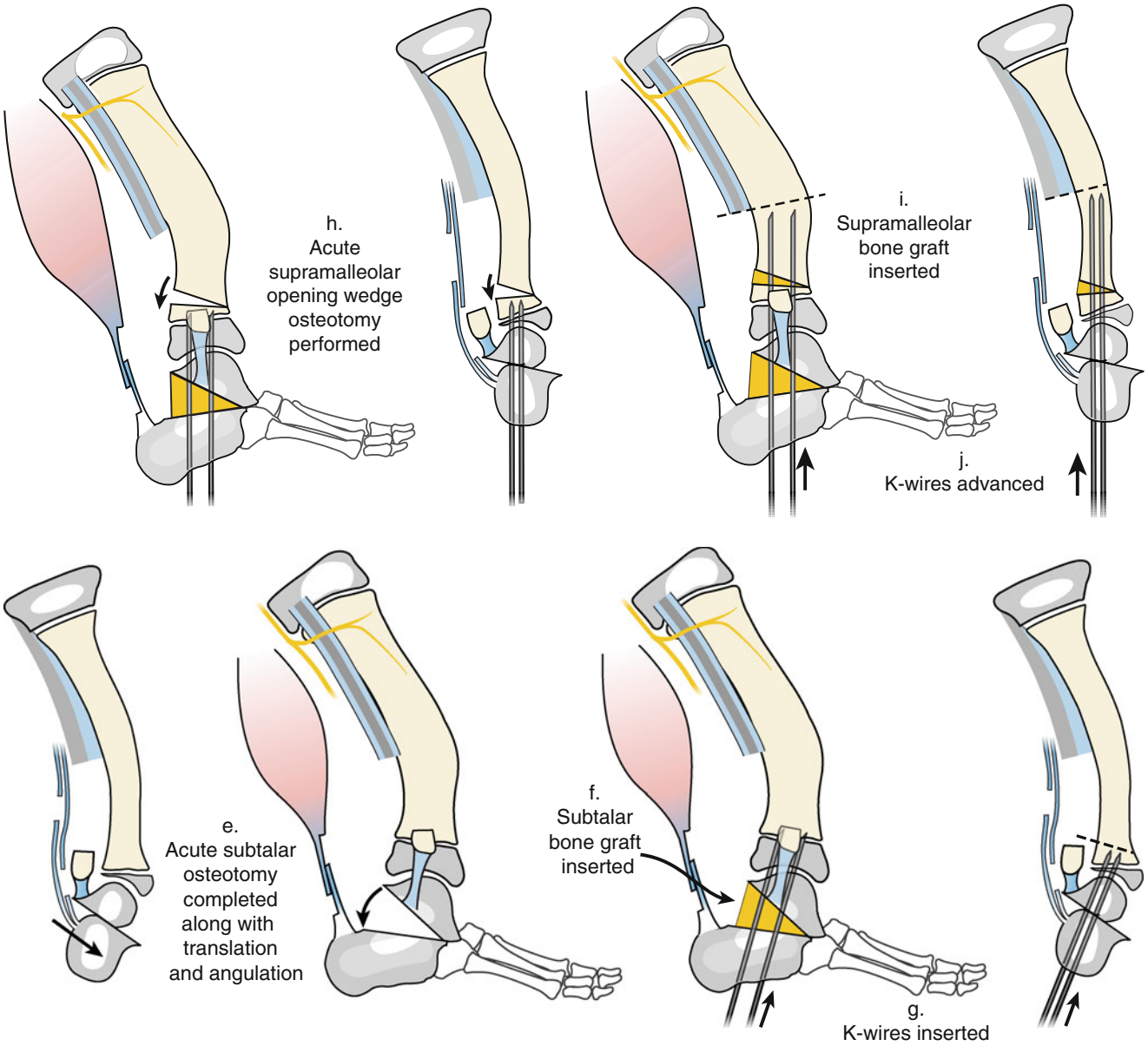


Fig. 23.28 Super-ankle surgical technique used to treat combined ankle/subtalar type fibular hemimelia (Paley Type 3C). Reprinted with permission from the Rubin Institute for Advanced Orthopedics, Sinai Hospital of Baltimore

Box 23.6. Super-Ankle Procedure for Paley Types 3a, b, and c

- Lengthen peroneal tendons
- Lengthen Achilles
- Resect fibular anlage and intermuscular septum
- Osteotomy: supramalleolar or subtalar or both
- Transfixion wires from sole of foot into tibia
- External fixator to correct diaphyseal anteromedial bow

Surgical Technique for Type 3d (Fig. 23.29)

Diagnosing Type 3d

This subtype is extremely unusual and can be recognized in younger children through MRI evaluation or in older children through computed tomography. The hallmark of this deformity is a wedge-shaped talar body.

Positioning and Surgical Approach

The positioning and approach are identical to what is described previously for Type 3a.

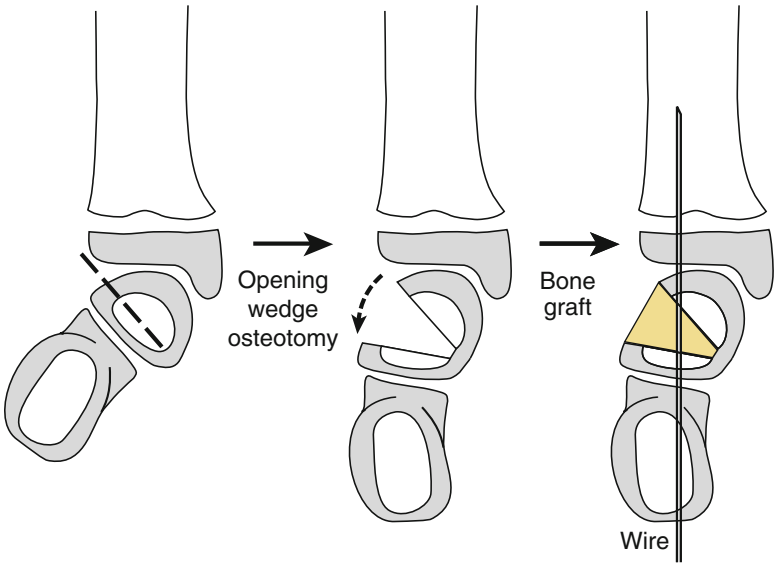


Fig. 23.29 Surgical technique used to treat talar body type fibular hemimelia (Paley Type 3D). Reprinted with permission from the Rubin Institute for Advanced Orthopedics, Sinai Hospital of Baltimore

Osteotomy

The opening wedge procedure is similar to the technique described for the Type 3b (subtalar type), except the osteotomy does not pass completely through the medial side.

Surgical Technique for Type 4

Initial Nonsurgical Treatment

This subtype represents less than approximately 15 % of FH cases; therefore, treatment must be individualized. In the author's experience, the initial treatment is nonsurgical, following Ponseti principles. It is unclear why the Ponseti method is effective in correcting the varus deformity, as the Ponseti technique works through the subtalar joint. In our experience, all of these children have a coalition of the subtalar joint. After casting and tenotomy, the deformity is converted to a more typical equinovalgus type, which can then be corrected using the Super-ankle approaches described above. However, we have seen many children treated elsewhere with preliminary open posteromedial release for Type 4 FH.

Additional Procedures

During tibial lengthening, valgus deformity of the diaphysis, if present, should be corrected. Distal femoral valgus can be corrected with screw-plate devices for growth modulation, which is discussed in greater detail in Chap. 4 (Fig. 23.30). Once corrected, the screw-plate devices are removed, but rebound valgus is commonly observed. After tibial lengthening, the valgus position of the lower extremity can recur due to abnormal growth at the physis of the distal femur and/or the proximal tibia. This progressive valgus deformity in FH may be attributed to hypoplasia of the lateral femoral condyle and/or to the tethering effect of the residual scar-like fibular band called the fibular anlage, if present. After lengthening, the Cozen phenomenon may also account for some of the rebound valgus that can be seen post-tibial lengthening (Fig. 23.31).

We have observed cases of relapse of ankle valgus several years after the Super-ankle procedure. After the initial correction of the foot and ankle with the Super-ankle procedure, the valgus deformity can recur with the appearance that the child is walking on the medial malleolus. This recurrence can be addressed with a revision surgery that includes acute osteotomy of the distal tibia and/or the hindfoot bones. Usually the leg is placed in a cast for 4–6 weeks for healing and then the child is allowed to wear regular shoes.

Post-surgical Care

Lengthening and deformity correction of the tibia begins 7 days after the surgery at a rate of 0.25 mm three times daily. Physical therapy is the most important aspect of the lengthening process. Without proper physical therapy, the lengthening goals will not be achieved and major complications will occur. Therefore, physical therapy requirements are very strict. The patient undergoing tibial lengthening must attend physical therapy sessions 5 days per week while at the same time performing an exercise and stretching program at home. Normally, the foot is included in the frame, so all physical therapy efforts are directed towards the knee and the toes. The knee tends to develop a flexion contracture. The toes also tend to develop flexion contractures. Night splints for the toes can be fabricated by the occupational

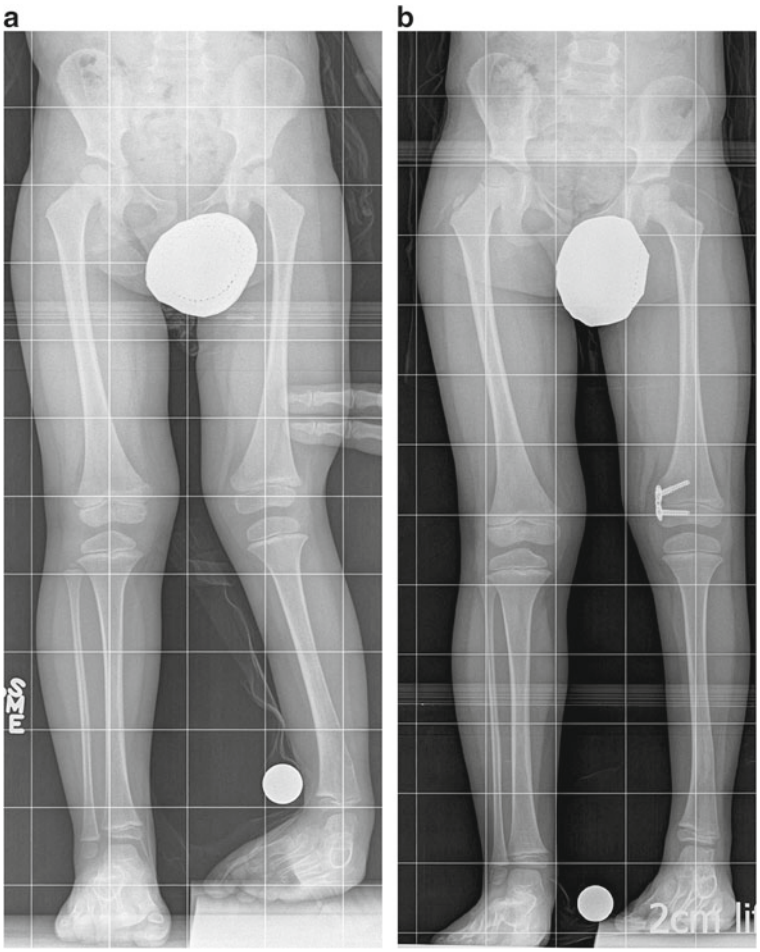


Fig. 23.30 (**a**) Full-length standing AP view radiograph of the same child shown in Fig. 23.25 at 3-year follow-up with genu valgus secondary to hypoplasia of the lateral femoral condyle and supination deformity of the forefoot. (**b**) Full-length standing AP view radiograph of the patient 10 months after distal femoral hemiepiphysiodesis and tibialis anterior tendon transfer to the dorsum of the foot. Reprinted with permission from the Rubin Institute for Advanced Orthopedics, Sinai Hospital of Baltimore

therapists as a forefoot splint to support the toes. The physical therapy continues at this level of intensity for the duration of the distraction phase (2–3 months depending on the planned amount of lengthening). Younger patients can put as much weight as they can tolerate on the operated leg. Older and larger patients are kept at 50 % weight-bearing during the lengthening phase.

Normally, we include the foot in the tibial frame, so there is initially no physical therapy for the ankle. Once the lengthening is completed (provided the knee is not contracted), it is recommended that the foot wires be removed to allow rehabilitation of the ankle. During the lengthening with the foot included in the frame, the therapist may direct all efforts to preventing knee flexion contracture. Nighttime splinting in extension is also helpful. If despite best efforts, the patient develops a knee flexion contracture, then it is possible to extend the tibial frame to the femur in the form of a cast brace and use that as leverage to stretch the knee contracture. This can be done by building a thigh cuff from fiberglass cast material, using waterproof cast padding, and incorporating a ring into the thigh cuff, which is then linked to the proximal tibial ring with two hinges at the knee joint and a posterior distraction rod to stretch out the knee flexion contracture. Please refer to Chap. 12 for more details regarding physical therapy for limb lengthening.

Outcomes

The treatment of FH is still a challenge. The goal is to achieve normal limb alignment and length with a functional plantigrade foot. This undertaking is often accompanied by repeated surgical procedures and a high rate of complications such as pin tract infections, residual LLD, delayed union, ankle and knee stiffness, refractures, knee subluxation, residual foot deformities, and even mild to severe depression.

McCarthy et al. [8] compared the final outcomes after reconstruction and amputation and found that children after early amputation were more active, had less pain, and were more satisfied with the results of treatment. These children also had fewer procedures with fewer complications. The reconstruction group had a total of 21 complications and each patient underwent a mean of 7 procedures.

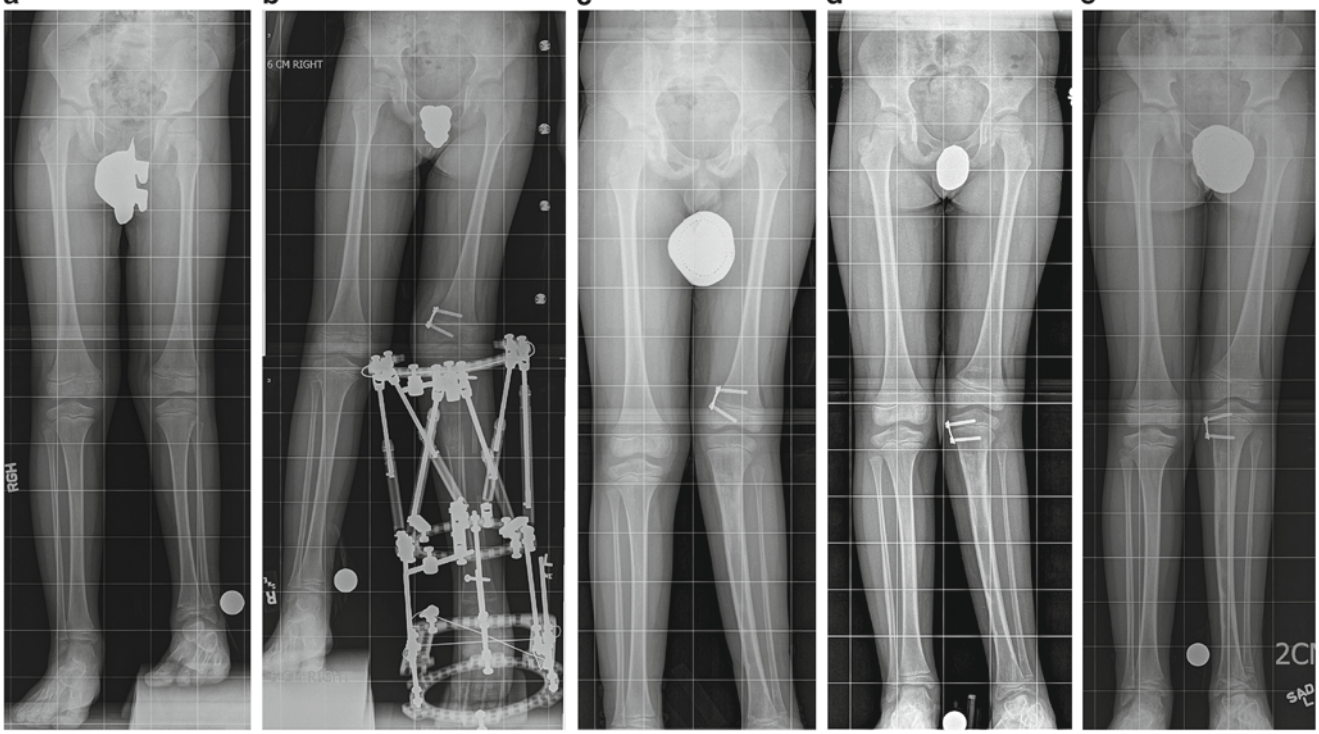


Fig. 23.31 (**a**) Full-length standing AP view radiograph of a 6-year-old boy obtained before he underwent tibial/fibular lengthening and distal femoral hemiepiphysiodesis. (**b**) Full-length standing AP view radiograph after 6 cm of lengthening. Note the mild knee flexion contracture but normalized medial proximal tibial angle. (**c**) Full-length standing AP view radiograph obtained 6 months after external fixation removal. Note the correction of the distal femur and the normalized lateral distal femoral angle. However, progressive recurrent genu valgus from the proximal tibia is also evident, possibly from a Cozen-type phenomenon. (**d**) Full-length standing AP view radiograph obtained after distal femoral guided growth plate removal and hemiepiphysiodesis of the proximal tibia. (**e**) Full-length standing AP view radiograph obtained 8 months after proximal tibial hemiepiphysiodesis shows continued correction of the recurrent valgus. Reprinted with permission from the Rubin Institute for Advanced Orthopedics, Sinai Hospital of Baltimore

In this study, the children in the lengthening group were older and therefore treated later, making treatment more difficult. Also 50 % of the children were treated with the Wagner method, which is associated with more complications [27, 28].

Before 2000, there are several papers with the same poor outcomes in which the authors [12, 27, 29] suggested that amputation is a more effective method of management than limb lengthening in severe fibular hemimelia cases. During the last decade, there was a tendency toward a reconstruction and lengthening treatment even for more severe cases with more favorable results.

Catagni et al. [10, 30] treated 32 patients with type III FH according to the modified Dalmonte classification. In this study, the mean number of surgical procedures was six and the complication rate was 82 % but most of the complications resolved except for two patients who ultimately underwent amputation. The outcomes were satisfactory in 17 of 32 patients.

Changulani et al. [9] reported the results of eight patients, in which each patient underwent an average of three surgical procedures. All patients developed minor complications such as pin-tract infections. At last follow-up (average 15 years), all patients were ambulatory and mobile with acceptable limb lengths and limb alignment.

Oberc and Sulko [13] published a study of 31 patients. Only nine (29 %) were treated with the elongation method. In these 9 patients, there were 12 elongation and 39 surgical procedures in total. The complication rate was 78 %, and there were satisfactory results in five of nine patients.

El-Sayed et al. [14] reported the results of 157 patients with Achterman and Kalamchi type-II FH. All patients underwent elongation, and 12 patients underwent concomitant femoral lengthening (average 13.6 cm). There were 107 minor complications, which were treated without change to the final outcomes and 19 major complications in which the desired lengthening was not achieved.

El-Tayeby and Ahmed [31] performed ankle reconstruction prior to elongation in 13 patients. All patients had a stable ankle without valgus deformity or subluxation after a mean follow-up of 18.6 months and showed good alignment in all cases.

Zarzycki et al. [32] treated ten patients with fibular hemimelia type II according to the Achterman and Kalamchi classification. The mean lengthening was 23 % of the preoperative length. Equal limb length and functional foot positioning were achieved in four patients, and the complication rate was 87.5 %.

Başbozkurt et al. [33] reported his experience treating FH in five patients. At last follow-up, three of five patients had plantigrade feet and two patients had equinus deformity. The other patients had minor complications (pain and pin-tract infection).

There is still a major debate amongst pediatric orthopedic practitioners regarding the relative value of lengthening versus amputation for higher grades of FH. Regardless of the outcome of this debate, there will always be families that refuse amputation; therefore, reconstructive lengthening techniques should be available. For cases with more normal function of the foot and ankle and with at least four rays, the results of reconstruction should be fairly predictable and the need for amputation in these moderately involved patients may be minimized.

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Tibial Hemimelia

24

Dror Paley and David Y. Chong

Introduction

The first case of tibial hemimelia was reported in 1841 by Otto [1] in the German literature. However, he incorrectly described it as fibular hemimelia, and Burckhardt later recognized his mistake in 1880 [2, 3]. Thus, the first correct reported complete case of tibial hemimelia is credited to Billroth in 1861 [4, 5].

Tibial hemimelia often appears as a shortened leg with knee and ankle deformity. Most often there is a knee flexion contracture, though multidirectional instability may also be present due to a lack of collateral ligaments. A patella and quadriceps mechanism may be present or absent and may have limited function if present. There may be cutaneous manifestations such as dimples over the head of the fibula or the free end of the tibia. The tibia may be shortened, dysplastic, or absent; it may also present as an anlage invisible to radiographs. The fibula can be normal or dysplastic, and often subluxated or dislocated proximally and/or distally. The ankle is often in varus and equinus, and the foot may be adducted and supinated. The medial side of the foot may be missing rays.

Genetics

The incidence of tibial hemimelia is reported to be one in one million [6, 7]. In 1941, there were 79 published cases [8]; since then there have been several hundred more reported. Tibial hemimelia may be diagnosed on prenatal ultrasound as early as 16 weeks of gestation [9]. There have been many studies on the genetic inheritance of tibial hemimelia. Multiple reports describe parent to child transmission (Nutt [8], Jones [47]) as well as families with multiple siblings affected (Aitken [10], Emami [11]). Autosomal dominant heritance has been implicated by Clark [12], Cowell [13], and Lenz [14, 15]. Autosomal recessive inheritance has also been described (Fried [16], Mahjlondji [17], and McKay [18]). An interesting breeding trial of Galloway cattle with tibial hemimelia concluded that tibial hemimelia was caused by homozygosity of a single autosomal recessive gene with variable expressivity and pleiotropic effects on various body systems [19].

Tibial hemimelia has been found to be associated with several syndromes. Werner's syndrome, or tibial hemimelia–polysyndactyly–triphalangeal thumb syndrome [20] (THPTTS) is an autosomal dominant disorder which is now thought to be a variant of triphalangeal thumb-polysyndactyly syndrome (TPTPS), which have both been mapped to chromosome 7q [21]. It has also been proposed that deletion on chromosome 8q, contiguous with Langer-Giedion syndrome, or type II tricho–rhino–phalangeal syndrome (TRPS II) may also be responsible for tibial hemimelia [22]. Case reports have also linked tibial hemimelia and other limb anomalies to CHARGE syndrome, a pattern of congenital anomalies including eye, nose, ear, heart, and genital defects [23, 24]. Most cases of CHARGE syndrome are due to a mutation of the CHD7 gene (chromodomain helicase DNA-binding protein 7), located on chromosome 8q, and CHD7 is known to be expressed by the developing limb bud mesenchyme [25, 26]. In addition, tibial hemimelia has been linked to tibial hemimelia-diplopodia syndrome [27], tibial hemimelia–split hand and foot syndrome [28], and tibial hemimelia–micromelia–trigonal brachycephaly syndrome [29].

Tibial hemimelia can be unilateral or bilateral. It is estimated that 30 % of cases are bilateral [30]. It does seem to favor the right side for unknown reasons. Spiegel noted that all 11 of his unilateral cases affected the right side, and in his

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review, about 72 % of unilateral cases in the literature affected the right side [31]. Tibial hemimelia has also been associated with multiple congenital anomalies. In Schoenecker's case series of 57 patients, 34 (60 %) had associated anomalies [32]. Similarly, Launois and Kuss found 24/41 (59 %) cases had associated anomalies [33].

In the ipsilateral limb, additional deformities may be suppressive or duplicated. Reported anomalies include congenital femoral deficiency, bifid femur, missing patella or quadriceps mechanism, knee hyperextension or flexion, clubfeet, missing toes, syndactyly, supernumerary or duplicated great toe, and a "mirror foot" [34–38]. Other reported limb deformities include radial dysplasia, lobster claw deformity, hand syndactyly, polydactyly, triphalangism, missing fingers or toes, hip dysplasia, hip dislocation, coxa valga, hemivertebrae, and myelomeningocele [39–42]. Associated deformities that have been reported include deafness, cleft palate, pseudohermaphroditism, cryptorchidism, and hypospadias [30].

Pathoanatomy

Evans and Smith [43], in an anatomic examination of the leg, found both an absence and duplication of some muscles. He described some muscles as being essentially functionless and attached to only one bone. He postulated a mesoblast origin and not likely a mechanical or traumatic source, confirming what Hovelacque and Noel had found in mouse embryos in 1909 [44].

Turker et al. [45] dissected five Jones type Ia specimens. They found ipsilateral toe anomalies in most cases, ranging from four to eight digits. The affected leg had smaller girth, and a dimple was consistently found where the skin was tethered over fibula. They found intact saphenous and lesser saphenous veins, as well as sural and superficial peroneal nerves in the subcutaneous tissues. The deep peroneal and posterior tibial arteries were found associated with their respective nerves, but the posterior tibial bundle was found to be short and acting as a tether. Lateral and superficial posterior compartment muscles were generally intact with normal insertions. Gastrocnemius and soleus muscles had confluent Achilles tendon, which inserted on the medial side of the calcaneal tubercle. The anterior and deep posterior compartments did not have a discrete boundary. Tendons had anomalous courses and sometimes split. No identifiable posterior tibial or anterior tibial muscle belly was found, but all specimens had a tendon inserting medially on the midfoot that tethered the foot in supination. Three specimens had an anomalous tendon inserting onto the neck of the talus. In addition, all specimens had a flat cable of tendon-like structures on the anterior border of the fibula with minimal muscle associated proximally. This unknown structure wrapped around and inserted on the posterior capsule of the ankle. Abductor hallucis muscles were found in all specimens, even in feet without medial rays. No discrete plantar fascia was found. All specimens had subtalar coalitions, and some had midfoot coalitions. The talus articulated with the distal medial fibula on its posterolateral side with a vertical and sagittal orientation with only one plane of rotation.

Box 24.1. Introduction to Tibial Hemimelia

- Incidence is approximately one in one million.
- Autosomal dominant and recessive inheritance has been reported, with variable expression.
- Defects on chromosome 7q and 8q have been implicated, as well as the CHD7 gene.
- Common associated deformities: congenital femoral deficiency, bifid femur, absent patella, mirrored foot, and polydactyly.
- A dimple is often seen on the proximal fibula.
- Anterior and deep posterior compartments of the leg are most deficient, with tendon remnants still attached to the foot.
- The talus tends to follow the fibula in the deformity.

Classification

Frantz and O'Rahilly described a classification system for congenital skeletal limb deficiencies in 1961 [46]. The authors described amelia, hemimelia, phocomelia, and other limb deficiencies. Hemimelia was further described as being complete, partial, or paraxial. They are then divided into terminal (distal) and intercalary (middle) deficiencies, and further subdivided into transverse and longitudinal deficiencies.

The Jones classification (Fig. 24.1) was proposed in 1978 and was based on radiographic criteria. Type I deficiencies had no visible tibia and were subdivided into two groups: the Ia group had a hypoplastic distal femoral epiphysis and the Ib group had normal ossification of the distal femoral epiphysis, suggesting the presence of a proximal tibial epiphysis. Type II deficiencies had a visible proximal tibia but had a distal tibia deficiency. Type III deficiencies had a visible distal tibia, but had a proximal tibia deficiency. Type IV deficiencies had a shortened tibia with significant distal tibiofibular diastasis [47].

Kalamchi and Dawe, in their study of 21 cases in 1985, proposed a modification to the Jones classification and the results of their treatments for each type [48]. Type I deficiencies were defined as a total absence of the tibia. This group had knee flexion contractures, greater than 45° and no active quad-

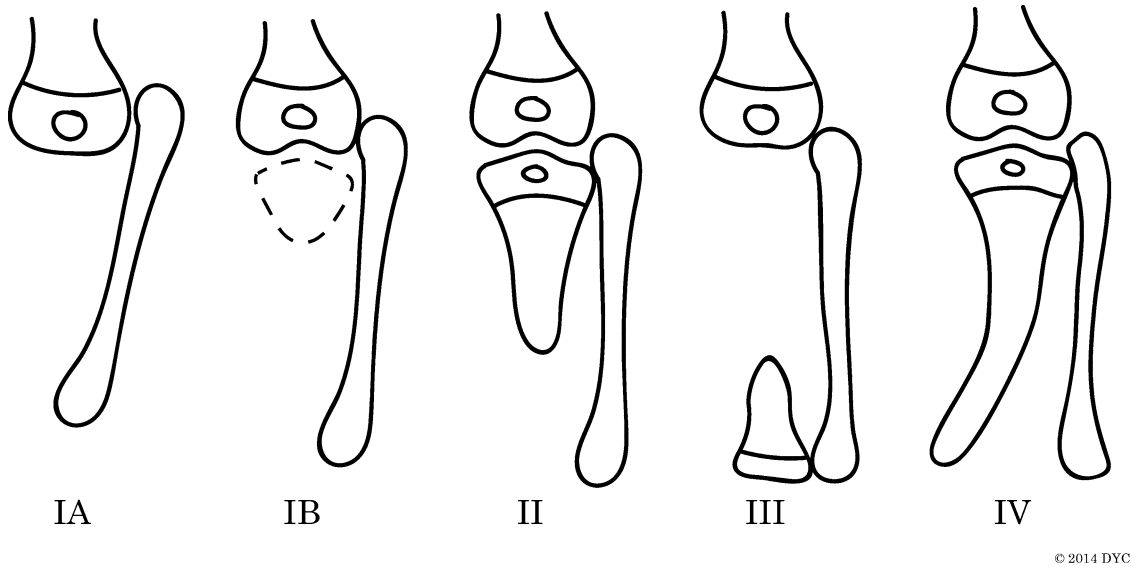


Fig. 24.1 Jones classification of tibial hemimelia

riceps function. These patients also had proximal migration of the fibular head and hypoplasia of the distal femur. Type II deficiencies were defined as distal tibial aplasia, with a proximal tibia present. There was active quadriceps function and had knee flexion contractures between 25° and 45°. They had more normal development of the distal femur, with normal metaphyseal width and normal epiphyseal ossification, indicating at least a proximal tibial anlage was present. There was also less proximal migration of the fibula. Type III deficiencies were defined as distal tibia aplasia with diastasis of the tibiofibular syndesmosis. They had normal knee joints and good quadriceps function. Usually the talus was subluxated proximally with a prominent distal fibula.

Paley proposed a classification in 2003 based on the Jones classification [49]. The Paley classification (Fig. 24.2) is modified in this manuscript and described in more detail below. The classification is oriented around the treatment for each type.

The most recent classification scheme was published by Weber in 2007 [50]. Previous classifications had not taken into account the cartilaginous anlage. He also added a few types of tibial hemimelia which were previously unclassifiable by the previous schemes and previously published by case report [51]. Weber's new classification took into account the entire affected leg, from hip to foot. He proposed seven types of increasing severity, with subgroups based on whether the cartilage anlage was present (a) or not (b). Type 1 was tibial hypoplasia but intact proximally and distally. Type 2 was distal diastasis. Type 3 was distal aplasia. Type 4 was proximal aplasia. Type 5 was bifocal aplasia proximally and distally. Type 6 was complete tibial agenesis with double fibulae. Type 7 was complete agenesis with a single fibula. He then assigned a score to determine the functional ability of the limb, where higher scores indicate less impairment of the limb. The tibia (0–22 points) and presence of an anlage (+10 points) weighed heaviest in importance, and the patella (0–3 points) was given slightly extra weight. The rest of the limb was scored from 0 to 2 points, and included the hip joint, distal femur, fibula, foot, and muscle function over the hip, knee, and ankle joints. Five classes were defined based on the score, indicating the degree of deficiency and difficulty of reconstruction.

The rarity of tibial hemimelia lends itself to other variants, which still may not fit any classification scheme. One reported case which does not fit any current classification is that of the intercalary deficiency, where the central portion of the tibia is missing but the proximal and distal tibia is intact [52]. The authors in this case were able to transfer the fibula into the defect with success.

Box 24.2. Jones Classification

- Type I: No visible tibia
  - Ia: Hypoplastic distal femoral epiphysis
  - Ib: Normal distal femoral epiphysis ossification (possible cartilaginous proximal tibia)
- Type II: Distal tibia deficiency
- Type III: Proximal tibia deficiency
- Type IV: Shortened tibia with distal tibia-fibula diastasis

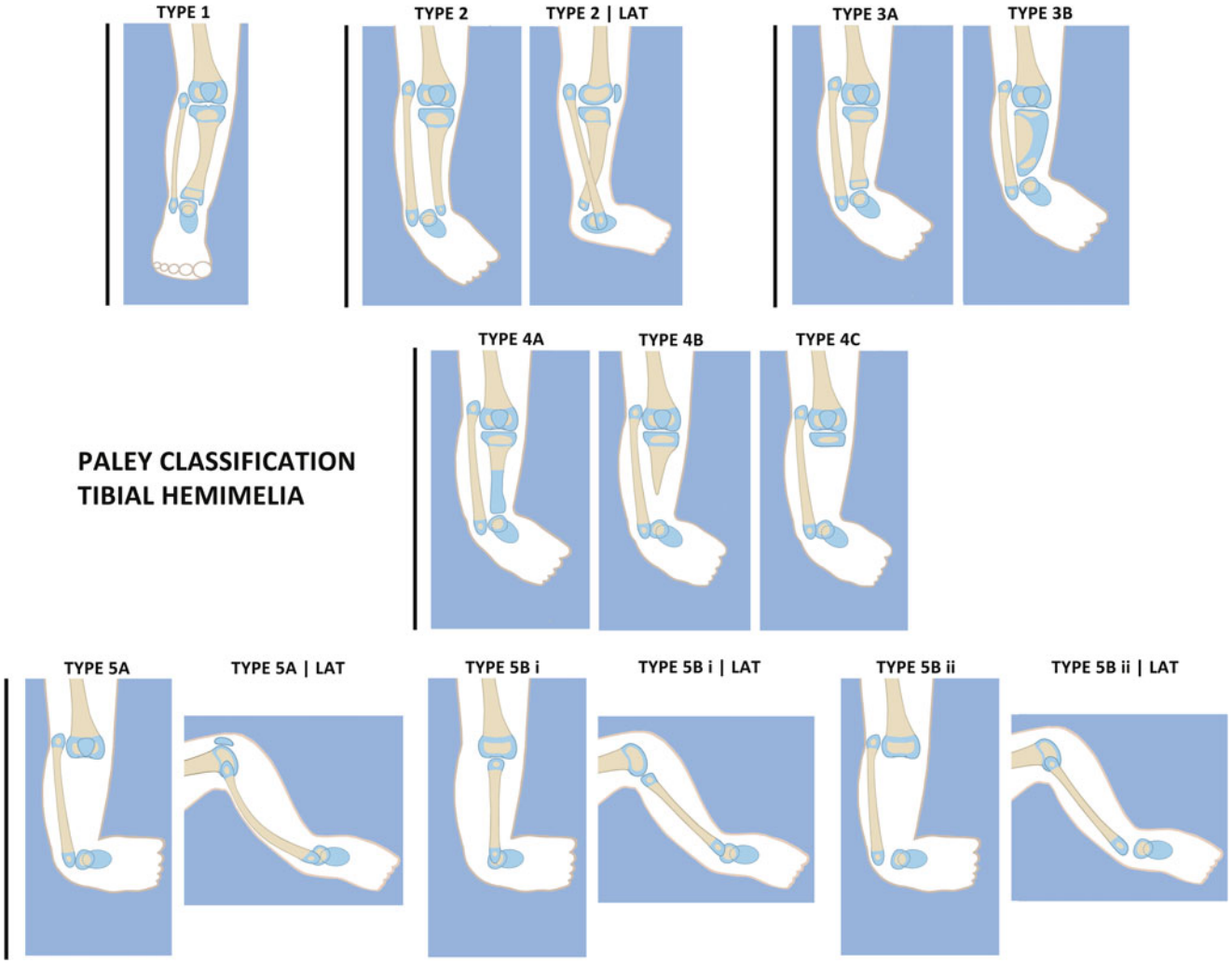


Fig. 24.2 Paley classification of tibial hemimelia

History of Treatment

Early Treatments

In 1877, Albert first published about the centralization of the fibula, fusing the femur and fibula [53]. In 1905, Myers [54] proposed a fibula-femoral arthroplasty, which was further developed by Brown in 1965. In the case of a partial absence of the tibia, there are early published reports of attempted synostosis between the tibial remnant and fibula, either side-to-side or end-to-end (Bade [55], Nove-Josserand [56]). Fraser and Robarts in 1914 reported a case with an intercalary defect in which they transplanted the contralateral fibula with reported success [57]. In 1929, Putti [58] was one of the first to give directives on treatment, describing different approaches to eight different cases: fusing the fibula to the talus in extreme equinus to increase the leg length, transfer of the proximal fibula to the intercondylar notch, and side-to-side synostosis of the fibula to the tibia both proximally and distally. Sulamaa and Ryoeppy [41] performed side-to-side opposition and recommended knee disarticulation for type I deformity.

In the current literature, the treatment of choice for tibial hemimelia has historically been biased towards amputation, though it is dependent upon the classification of the deformity. In addition, in some communities, the option of amputation is not accepted culturally, and patient families wish to pursue limb salvage. With complete absence of the tibia (Jones type Ia), most literature points towards amputation [32, 42, 48, 67, 69], while less severe deformities have several case reports of salvage options [59–61]. The presence of a tibia anlage and an active quadriceps mechanism are important factors for reconstruction, as well as prevention of knee flexion deformities [62]. Their absence may be indicated by the absence of a patella, which can be difficult

to find in young children. Thus, current imaging options such as MRI and ultrasound are a useful method of determining the presence of the patella or tibia anlage [63].

Brown Procedure (Fibular Centralization)

Brown first published his surgical procedure of fibular transfer and centralization under the femur in 1965 [64]. This procedure was done for patients with complete absence of the tibia (Jones type I). He also recommended a Syme-type amputation of the foot. In 1972, Brown published his 15-year follow-up, in which 40 of 56 patients were available for review [65]. Eighteen required secondary surgery due to a knee flexion deformity. Of the remaining 22, 1 was nonambulatory, and all but 2 were wearing braces while ambulatory. He recommended attachment of the patellar ligament to the fibula, preoperative traction, as well as femoral shortening and soft tissue releases as needed to gain extension. He also recommended surgery before age 1 for maximal ambulatory and fibular articulation potential. Inferior results were noted with an absence of quadriceps function.

Jayakumar and Eilert (1979) [66] reported six cases treated with the Brown procedure. All patients received either a Boyd or Syme amputation. Clinical outcomes were graded on several criteria of knee joint function, requiring (1) at least 10–80° of active range of motion, (2) <5° varus/valgus instability, and (3) no flexion contracture. Only two patients achieved these criteria, and they had 3+ quadriceps power. One patient had ipsilateral congenital femoral deficiency and underwent a femoral-fibula arthrodesis. Three patients had "poor" results from knee flexion contractures and two went on to knee disarticulations. The authors concluded that strong active quadriceps function of at least 3+ was necessary for Brown reconstruction to have superior results to an amputation.

Loder and Herring (1979) [67] reported nine cases of Jones type I deficiencies treated with the Brown procedure. All had preoperative knee flexion contractures. Initially, five of nine had good results with active range of motion and no flexion contractures. However, only one patient maintained active quadriceps power, and all progressively developed knee flexion contractures. Despite secondary procedures, outcomes were also poor due to knee instability and poor range of motion. At final follow-up, five had knee disarticulations and the remaining cases had poor range of knee motion (10–45°).

Epps and Schneider (1989) [68] published on three cases of Brown procedures, and noted that their results deteriorated over time due to progressive flexion contractures, and needed secondary procedures. In 1991, Epps et al. [69] reported on 14 patients (20 knees) who underwent centralization of the fibula for Jones type I tibial hemimelia. All patients developed progressive knee flexion deformities and needed multiple secondary procedures, leading the authors to recommend early disarticulation of the knee and prosthetic fitting as the treatment of choice. However, the results were considered to be failures due to the knee flexion deformities.

Simmons et al. (1996) [70] revisited the Brown procedure, publishing his results on five patients and seven limbs. All cases had Jones type Ia deformities, though one case was found to be a 1b during surgery and thus the rudimentary proximal tibia was utilized for fixation. All patients had knee flexion and varus deformities. They also had equinovarus deformities of the foot and underwent Syme amputations. An anterolateral knee incision was used, and the biceps tendon and distal insertion of the patellar tendon released. Femoral or fibular shortening was performed as needed along with peroneal nerve decompression. The fibula was centralized underneath the femoral condyles and the knee stabilized in extension with a Steinmann pin. The patellar tendon was reattached to the fibula and along leg spica cast was applied. Subsequent procedures included split thickness skin grafting, and heel pad release for the Syme amputation, and one quadricepsplasty for lack of knee flexion. All ambulated with a PTB socket with thigh extension. Average range of knee motion was 57° and only two patients achieved 10–80° of knee flexion. However, all patients were satisfied and none went on to knee disarticulation. They recommended narrowing indications for reconstruction to those patients with grade 3+/5 active quadriceps, younger than 1 year of age, no fibular bowing, and ambulatory potential.

Wada et al. (2006) [71] published nine cases of limb reconstruction in Jones type I and II cases. They performed four tibiofibular synostosis and five Brown procedures, with concurrent foot centralization procedures and subsequent lengthening as needed. The tibia-fibular fusion patients had good results with preserved knee function. Of the Brown procedures, four had poor quadriceps function and one had persistent knee instability. All five had unsatisfactory functional results based on Jayakumar's criteria [66]; however, they were all household ambulators with only one patient needing an orthosis.

Hosny (2005) [72] published a case series of 6 patients with tibial hemimelia. Amputation was not accepted in his community and they presented at an older age (3.5–13 yo). Jones type I cases were treated with gradual distraction of the fibula with an external fixator, followed by a Brown procedure 1 month later. Type II cases were treated with gradual distraction followed by fixation of the proximal fibula to the proximal tibia. Fibula lengthening was subsequently performed but had limitations due to knee flexion contractures that resulted. Femoral lengthening was then performed at a later stage. Hosny reported all patient's families were satisfied with the outcome, with all the type II patients ambulating

independently with minimal (5°) to no knee flexion contractures and active ROM greater than 90°. The type I patients could walk with a KAFO and walker and had limited ROM (10–35/40°) but could perform ADLs at home without any pain.

Overall, most authors did not report good outcomes with the Brown procedure and recommended knee disarticulation rather than reconstruction as the best option for total absence of the tibia. Many of the poor outcomes were due to progressive knee flexion contractures, knee instability, and poor range of motion, as previously defined by Jayakumar [66]. However, for some patients in whom amputation is not an option, a limb that is weight bearing though less functional may be considered a success. Certainly the presence of a strong quadriceps, patella, and proximal tibia or anlage seems to favor reconstruction, and tibiofibular synostosis has generally met with good results. In addition, the use of an external fixator prior to reconstruction can help overcome soft tissue contractures.

Amputation

Knee disarticulation has been described for treatment and remains a salvage option for failed Brown procedures, as seen in many of the studies above. Kalamachi (1985) [48] treated three children with the Brown procedure, and all went on to subsequent knee disarticulations. The failure was attributed to knee flexion contractures and no active quadriceps function, leading the authors to recommend early disarticulation of the knee without attempt of reconstruction. Alternatively, if the femur was severely hypoplastic, a femorofibular arthrodesis was performed to effectively lengthen the femur, creating a longer lever arm for improved prosthetic fitting.

Similar results and conclusions were drawn by Schoenecker et al. (1989) [32]. Of a series of 57 patients and 71 limbs, 61 limbs eventually had some type of an amputation. Brown procedures were performed on 14 Jones type Ia limbs. Ten required secondary procedures due to knee instability or flexion contractures. Based on their results, they recommended evaluating for a proximal tibia anlage to differentiate Jones types 1a and 1b, and that a type Ia should be treated by knee disarticulation due to their less-than-satisfactory results with the Brown procedure.

Fernandez et al. (1998) [38] have also been a proponent of early amputation. Twenty-two cases of tibial hemimelia were reported, of which 17 patients (and caretakers) accepted surgery. All ten Jones type I cases underwent knee disarticulation. Three Jones type II and III cases had distal amputations. Only the remaining four Jones type IV cases had conservative treatment. The author recommended early amputation so that the patient would treat it as a "congenital amputation," allowing for better adaptation to their prosthesis and rehabilitation. For many of these patients, their socioeconomic status also required a quicker and more definitive solution.

Spiegel et al. (2003) [31] described some potential complications and sequelae of amputation in patients with tibial hemimelia. They treated nine Jones type I deficiencies with knee disarticulation without any subsequent complications. Five type II deficiencies were initially treated with distal amputation (Chopart or Syme). They developed prosthetic irritation from the proximal fibula from the varus malalignment and prominent fibular head. Three patients subsequently underwent tibiofibular synostosis. One patient had progressive varus deformity and difficulty with prosthetic fitting that improved after hemiepiphysiodesis. Overall, the authors felt unsure about the best course of action for type II deficiencies. Four type III deficiencies were treated with Syme or Chopart amputations and did well.

Tibiofibular Synostosis

In the presence of a tibial anlage (Jones type Ib) or a proximal tibia (Jones type II), most authors have reported good results with tibiofibular synostosis. Kalamachi [48] reported ten patients with Jones type II tibial hemimelia that were treated with tibia and fibula synostosis. Three of these utilized a modified Brown procedure, fusing the fibula with the cartilaginous tibia anlage. They found good stability and adequate range of motion with only mild (20–30°) flexion contractures. Due to leg length discrepancies and foot deformities, half had concurrent Boyd amputations and did well with a prosthesis.

Schoenecker [32] reported 8 of 15 Jones type II limbs successfully treated with tibiofibular synostosis. Twelve were treated with a Syme amputation and were functional below-knee amputees, one had a knee disarticulation, one was nonoperative, and only one patient, who also had a tibiofibular synostosis, retained the foot. They recommended tibiofibular synostosis for Jones Ib and II with concurrent distal Syme or Boyd amputations.

Distal Tibia and Ankle Stabilization

For Jones type II deficiencies, distal tibia aplasia leads to an unstable ankle. These have historically been treated with stabilization of the ankle in the form of arthrodesis and possible amputation. In Kalamachi's [48] series, three cases had essentially normal knees, but the talus was subluxated proximally with a varus foot and prominent distal fibula. These

were treated with calcaneo-fibular fusions and Boyd amputations. Schoenecker [32] treated seven Jones type limbs with Syme or Chopart amputations to function as below-knee amputees. Tibiofibular synostosis was performed in two of these patients.

Jones type IV deficiencies have a shortened tibia with significant distal tibia and fibula diastasis. Tokmakova et al. (2003) [73] felt that the treatment of choice was reconstruction of the ankle mortise, as their patients were independent ambulators with stable ankles and plantigrade feet.

In Schoenecker's series of ten Jones type IV limbs, one limb had a Syme amputation and nine limbs had ankle joint reconstruction and salvage of the foot. Of these nine, five had a Syme amputation within 3 years due to leg length discrepancy. He recommended ankle reconstruction and leg length equalization, similar to Fernandez [38].

Limb Lengthening

Limb lengthening is also an option and adjunct for the treatment of tibial hemimelias [74]. Hootnick et al. (1977) [75] followed the natural history of tibial hemimelia and found that the leg length discrepancy remained proportional over time. Thus, they were able to calculate a final predicted leg length discrepancy to help a family decide on lengthening versus Syme amputation. They recommended amputation for predicted discrepancies greater than 8.7 cm. However, with newer technology and a better understanding of the biology, staged reconstruction and limb lengthening may allow the trained surgeon to address more severe cases of congenital tibial deficiency, preserving more lower limb function than what has been reported previously.

Desanctis et al. (1990) [76] published three cases of Jones type II deficiencies treated with staged reconstruction. They corrected the foot deformity with serial casting and posteromedial soft tissue release, followed by tibiofibular diaphyseal reconstruction, alignment of axis of the foot and leg, and limb lengthening using the Ilizarov technique. Javid (2000) [77] reported on a lengthening after centralization of the fibula in a Jones IIb deformity. They found that the fibula had delayed new bone formation, and compression-distraction technique with an Ilizarov device had to be utilized to promote osteogenesis. Devitt reported good results with differential lengthening of two cases of Weber type I deficiency [51].

Weber Patelloplasty

The Weber patelloplasty was published in 2002 [78] and described a surgical procedure in which the patella was converted into a tibial plateau. The soft tissues were gradually distracted with an external fixator to avoid the need to shorten the femur or fibula. The patella is then brought down with crossing "visor" flaps from the capsule and chondrodesed to a centralized fibula. A Z-plasty of the quadriceps tendon was used to gain length. A hinged external ring fixator was used to stabilize while allowing knee range of motion and weight bearing. A second procedure was done to achieve chondrodesis between the distal fibula and the talus. To our knowledge, there have been no publications of results of this procedure, though Weber did further describe and refine his procedure in 2006 [79].

Box 24.3. Treatment Options

- Outcomes for the Brown procedure (centralization of the fibula) depend on the presence of a quadriceps mechanism and a mobile knee without flexion contractures.
- Progressive knee flexion contractures and knee instability are common reasons for failure of the Brown procedure.
- The presence of a proximal tibia or its anlage has had good results from synostosis with the fibula.
- For some parents, a quicker and more definitive knee disarticulation may be a more attractive option, allowing a child to adapt early to a prosthesis.
- Distal tibia deficiencies can be treated with distal fusion or synostosis or Syme-type amputation.
- Advances in limb lengthening allows for limb length equalization.
- The Weber patelloplasty converts an existing patella into a proximal tibia.

New Horizons and Strategies for Reconstruction in Tibial Hemimelia

The deficiency in tibial hemimelia is a spectrum of pathology, ranging from a congenitally short tibia with relative fibular overgrowth to complete absence of the tibia. It is to be distinguished from fibular hemimelia, where there is a progressive deficiency of the fibula, ranging from a congenitally short tibia with relative fibular hypoplasia to complete absence of the fibula. An important distinction between these two conditions is that the foot and ankle are always in varus with tibial hemimelia and almost always in valgus with fibular hemimelia. The progression of deficiency of the tibia is from distal to proximal, contrary to fibular hemimelia, where

the progression of fibular deficiency is from proximal to distal. Tibial hemimelia should be distinguished from proximal deficiency of the knee, which is most commonly associated with TAR (thrombocytopenia absent radius) syndrome. Proximal deficiency of the tibia ranges from simple genu varum to a ball and socket, complete absence, or congenital fusion of the knee joint. Therefore, in order to prevent confusion, tibial hemimelia should be classified as an incremental deficiency from distal to proximal.

Most classifications in orthopedics classify pathology from the less severe to the more severe (e.g., Garden classification of hip fractures [80], Berndt and Harty classification of talar dome osteochondral fractures [81], Herring and Catterall classifications of Perthes [82, 83], and Kalamchi classification for fibular hemimelia [84]). The Paley classifications [85] for congenital femoral deficiency and fibular hemimelia (FH) follow a similar format. Some well-accepted published classifications follow the opposite format, starting with the most severe or deficient cases (e.g., Pappas classification for CFD [86], Kalamchi classification of tibial hemimelia [48], and Jones classification for tibial hemimelia [47]). The Jones classification for tibial hemimelia has been the most widely used, but it is in reverse order of deficiency. Furthermore, the Jones type III is so rare that the senior author has only seen two cases of it in 27 years of practice and more than 300 cases of tibial hemimelia. The complete absence of the proximal tibia in the Jones III makes its treatment similar to the Jones I where the proximal tibial is missing. One consideration was to add this subtype to the Paley type 5 group. I have chosen not to do this since the treatment is the same whether the distal tibia is present or not. The treatment at the knee still depends on the presence or absence of the patella and whether the fibula is autocentralized or not. The treatment of the foot does not change since the foot is in severe equino-varus as if this remnant of distal tibia was not present (Figure 24.3). Finally, the Jones classification also lacks description of several types of tibial deficiency that involve both the proximal and distal physes, such as the Delta tibia type [87] or types that have a normal appearing tibia beside a longer fibula. The Jones classification also does not consider the presence or absence of a patella.

The Weber Classification [50, 79] changed the order back to increasing severity of deficiency. It also included types with a proximal deficiency (type 3). Similar to the Pappas classification, it splits tibial hemimelia into a large number of types (total of 7). The Weber also subdivides into types that will eventually ossify and change into other types (types 3, 4, 5, 6, 7). The Weber scoring system, combined with the large number of subdivisions, makes this classification challenging for practical use.

Dissatisfied with the existing classifications and prior to Weber's publication, Paley proposed a new classification in 2003 [49]. Each type and subtype has a different surgical treatment. The progression of deficiency is from least to most, and the classification assumes that tibial hemimelia is a progressive spectrum of deficiency from distal to proximal.

Box 24.4. Paley Classification of Tibial Hemimelia (See Fig. 24.2)

- Type 1: Congenital short tibia with relative overgrowth of the fibula, distal tibial plafond intact
- Type 2: Congenital deficiency of the tibial plafond with diastasis of the tibia and fibula and relative fibular overgrowth
- Type 3: Congenital deficiency of tibia with proximal and distal physis of tibia present; distal tibial physis more dysplastic than proximal; relative fibular overgrowth
- Type 4: Absent distal tibia; proximal tibia present; relative fibular overgrowth
- Type 5: Complete absence of tibia, fibula present

Paley Classification with Subtypes and Description of Common Features

Type 1: Valgus proximal tibia (genu valgum), overgrowth of fibula proximally, plafond present and normal.

Type 2: Tibial plafond missing. Medial and lateral malleolus present; distal fibula (lateral malleolus) normally aligned to lateral talus, internally rotated with foot around the tibia. Talus is positioned between the tibia and fibula due to absence of tibial plafond. Relative fibular overgrowth present. Foot follows the fibula. The diastasis can extend as a cleft between the tibia and fibula.

Type 3a: Well-formed distal tibial physis and separate from proximal growth plate of tibia. Plafond present but dysplastic. Major relative overgrowth of fibula.

Type 3b: Delta Tibia: proximal and distal growth plates connected through bracket epiphysis. Malorientation of ankle and knee joints ends of tibia. Major relative overgrowth of fibula.

Type 4a: Delayed ossification of part, or all, of the tibia. Ankle joint present, but distal tibial physis absent. Major relative overgrowth of fibula.

Type 4b: Diaphyseal deficiency of tibia. Complete absence of distal tibia at level of diaphysis; proximal tibia has atrophic, pointed bone end often covered by separate skin pouch. Major relative overgrowth of fibula.

Type 4c: Epiphysis of proximal tibia present but absent proximal physis. Knee joint present. Delayed ossification of epiphysis. Significant relative overgrowth of fibula.

Type 5a: Complete absence of tibia: a remnant of distal tibia may be present in rare cases; patella present; flexion contracture of knee.

Type 5b: Complete absence of tibia; no patella present; flexion contracture of knee; fibula may be (i) auto-centralized or (ii) dislocated.

Limb Reconstruction Surgery

Since Brown introduced centralization of the fibula, many attempts to reconstruct the knee in the most severe types (Jones I, Paley 5) have been made. These have been met with poor results, as previously discussed. Similarly, poor results of reconstruction for Jones types II and IV and Paley types 2, 3, and 4 have led most surgeons to conclude that through-knee amputation for Jones type I, through- or below-knee amputation for Jones type II, and Syme's amputation for Jones type IV are the best treatment for each type of tibial hemimelia. In light of the advances in modern prosthetics, this is definitely a good option and should be considered as the most tried and proven method of treatment. However, advances in the treatment of all types of tibial hemimelia offer new options with excellent functional results as an alternative to amputation. The rest of this manuscript will focus on the reconstructive options for tibial hemimelia according to Paley type and subtype.

Paley Type 1 (Fig. 24.4)

These patients tend to have bilateral involvement. The condition is often familial (autosomal dominant). The tibias are

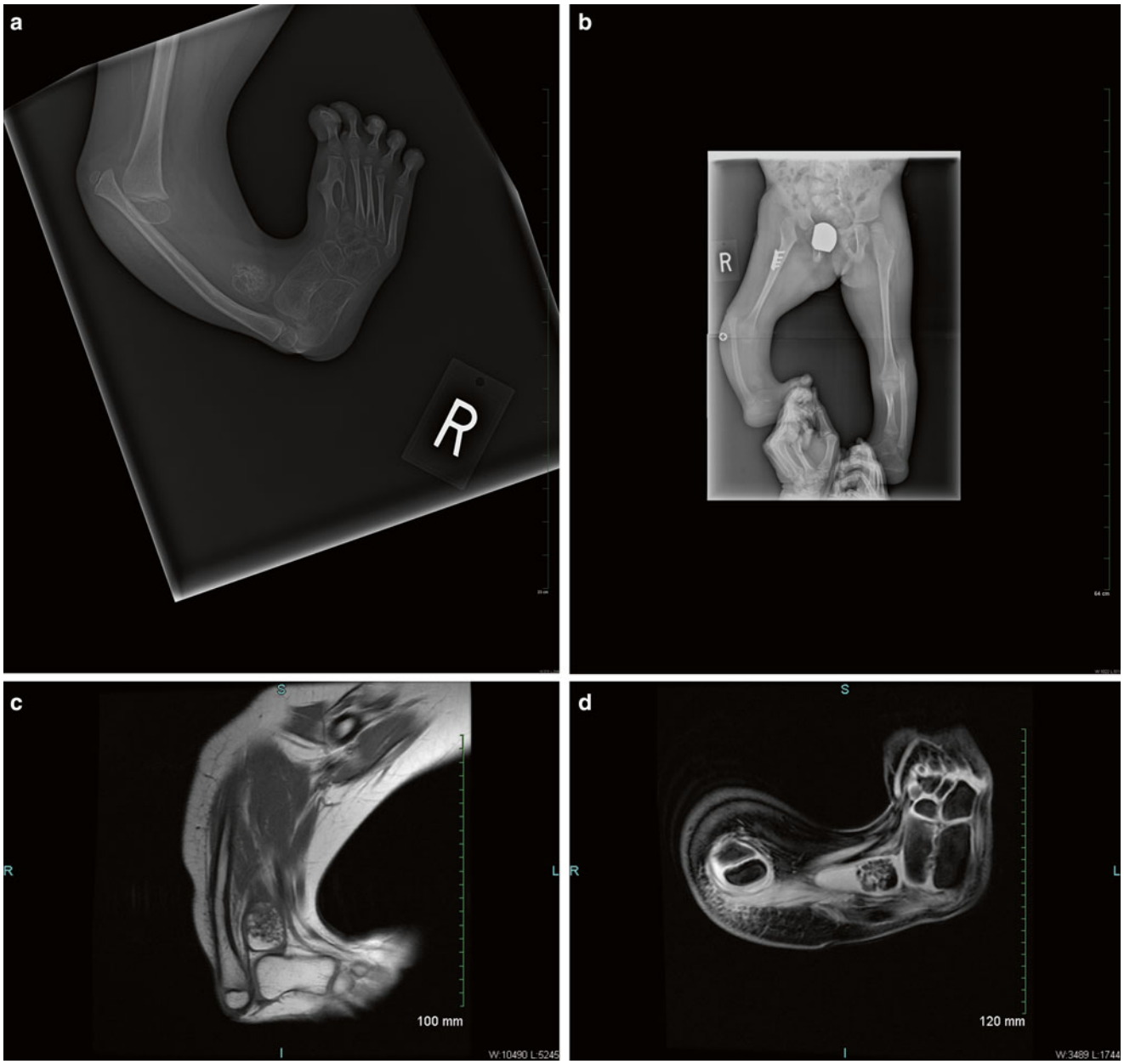


Fig. 24.3 (**a**) Jones type III tibial hemimelia. This would be classified and treated as a Paley type 5 despite the remnant of distal tibia. (**b**) Long x-ray of both legs showing the bilateral nature of this rare type. The left leg is a Paley type 3a and cannot be classified using the Jones classification. (**c**) Magnetic resonance imaging (MRI) confirming that there is no cartilaginous proximal tibia. (**d**) Another MRI cut confirming absence of the proximal tibia

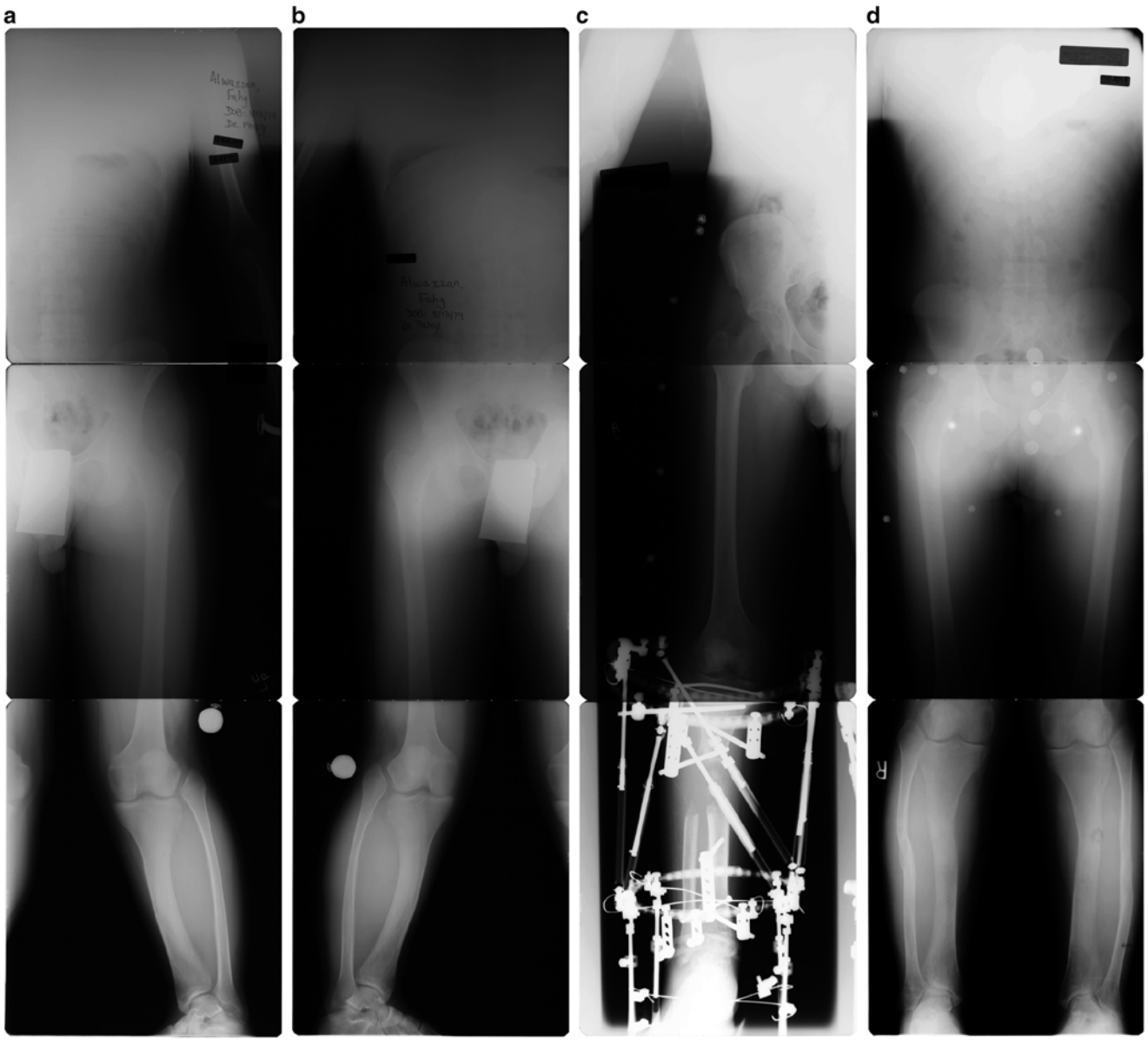


Fig. 24.4 (**a**, **b**) *Right* and *left* AP radiograph of Paley type 1 TH side showing valgus tibial deformity and overgrowth of fibula. (**c**) Lengthening and valgus correction of tibia and fibula with TSF. The fibula is not transported distally. (**d**) Final standing radiograph showing good alignment bilaterally. Proportions of femur to tibia restored closer to normal

short and the knees are in valgus. The valgus is usually from the proximal tibia, but can also be from the distal femur. If treated when the physes are open, the valgus can be corrected using temporary hemiepiphysiodesis. In adulthood, the valgus can only be treated by osteotomy. The biggest complaint of these patients is mesomelic disproportion and short stature. Therefore, the treatment is usually bilateral and includes tibial lengthening for stature in addition to correction of the valgus angulation. In regard to the overgrown proximal end of the fibula, which often articulates with the side of the femur, I prefer to leave it alone and not pull it distally. This avoids the problem of a knee flexion contracture that can result from transporting the fibula distally. If the proximal fibula is producing a noticeable bump that is bothersome to the patient, then the tibia can be lengthened without cutting the fibula, transporting the fibula distally.

Paley Type 2 (Fig. 24.5, 24.6)

The diastasis of the tibia with the fibula starting with the absence of the tibial plafond (Fig 24.4) may extend up the

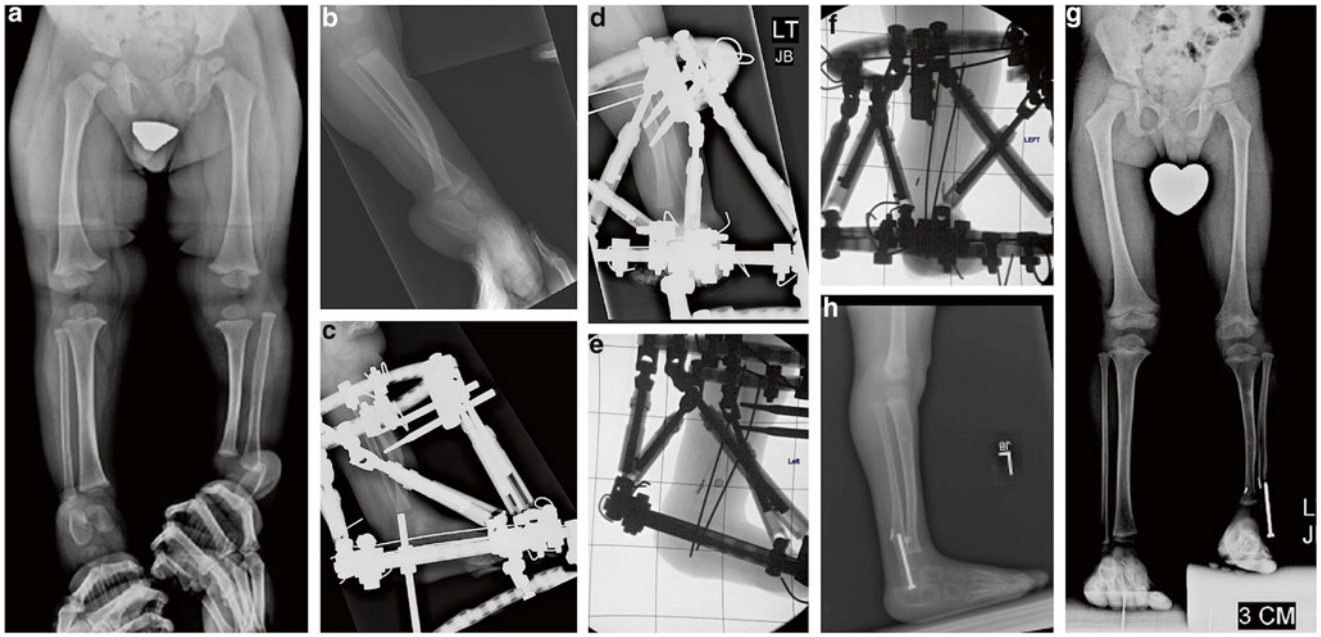


Fig. 24.5 (**a**) AP and (**b**) lateral radiographs of Paley type 2 TH. The distal fibula is more anterior than the tibia on the lateral. The distal fibula goes with the talus and is more hypertrophied than the distal tibia. (**c**, **d**) TSF device applied to foot and tibia for distraction of contracture of foot and to move the talus under the distal tibia. (**e**, **f**) After the talus is under the tibia, the ankle joint is opened and a biologic arthroplasty of the ankle joint is performed. A suture washer system is used to compress the diastasis. An intramedullary wire is placed to hold the foot in place under the tibia and the external fixator is left in place for 3 more months. (**g**, **h**) Epiphysiodesis of the distal fibula is performed to prevent distal overgrowth of the fibula. The final result is a plantigrade stable foot with some motion of the ankle

tibia. The tibia may have a varus bow to its diaphysis and in rare cases there can be a skin cleft between the two bones (Fig. 24.5). The foot always remains with the fibula. The foot is in equinovarus and internally rotated relative to the knee. The talus is proximally migrated relative to the distal tibia. The talus is at the correct level relative to the distal fibula. Since the ankle joint is to be reconstructed, the Achilles tendon is not cut but a gastrosoleus recession may be performed. The foot is repositioned by gradual distraction of the foot from the tibia using a circular ring external fixator. To prevent epiphysiolysis of the proximal and distal fibula, a 1.5-mm wire is drilled retrograde into the fibula and up the fibular diaphysis to exit through the proximal fibular epiphysis. The wire is brought through the skin proximally and then bent backwards on itself to form a hook. A small proximal incision is made and the wire is pulled back into the fibular head to lock into the proximal epiphysis. Distally, the wire is also bent 180°, then shortened and buried under the skin. This creates a temporary epiphysiodesis of the proximal and distal fibula.

One ring is applied to the proximal tibia with one wire and two half pins. The second ring is applied to the foot with three wires. The equinovarus deformity is corrected by gradual distraction of the foot away from the knee, repositioning the talus under the distal tibial epiphysis. Since the fibula is overgrown relative to the tibia, it does not need to be fixed to the distal ring. Its association with the talus and calcaneus causes it to follow the foot distally. This moves the fibula from its relatively overgrown proximal position down to the normal station.

Once the foot is located under the distal tibial epiphysis, a planned second stage surgery can be carried out. Under general anesthesia, the distal ring and wires are removed. The pin sites are covered by Betadine soaked gauzes and the leg is prepped and draped free. A transverse incision is made on the medial side at the level of the tip of the medial malleolus. The tibiotalar joint is opened, and the distal tibia and proximal talus are exposed. The tibialis posterior tendon is often found dislocated between the tibia and fibula, where the plafond should have been. It is moved out of this location and retracted postero-medial to the tibia. The distal end of the tibial cartilage is carved with a knife to the concavity of the tibial plafond, matching the convexity of the dome of the talus, creating a biologic arthroplasty. A wire, perpendicular to the sole of the foot, is passed through the dome of the talus, through the epiphysis of the distal tibia, and continues proximally into the tibial diaphysis. If the tibia has a varus diaphyseal bow to it, a percutaneous osteotomy should be made at the apex of this bow with an acute angular correction, straightening the tibia. The wire is advanced up the tibia to stabilize this osteotomy.

The tibiofibular diastasis is treated next. This is fixed by using a syndesmotic suture system such as the Arthrex-Tightrope™ or the Biomet-Ziptite™. The syndesmotic

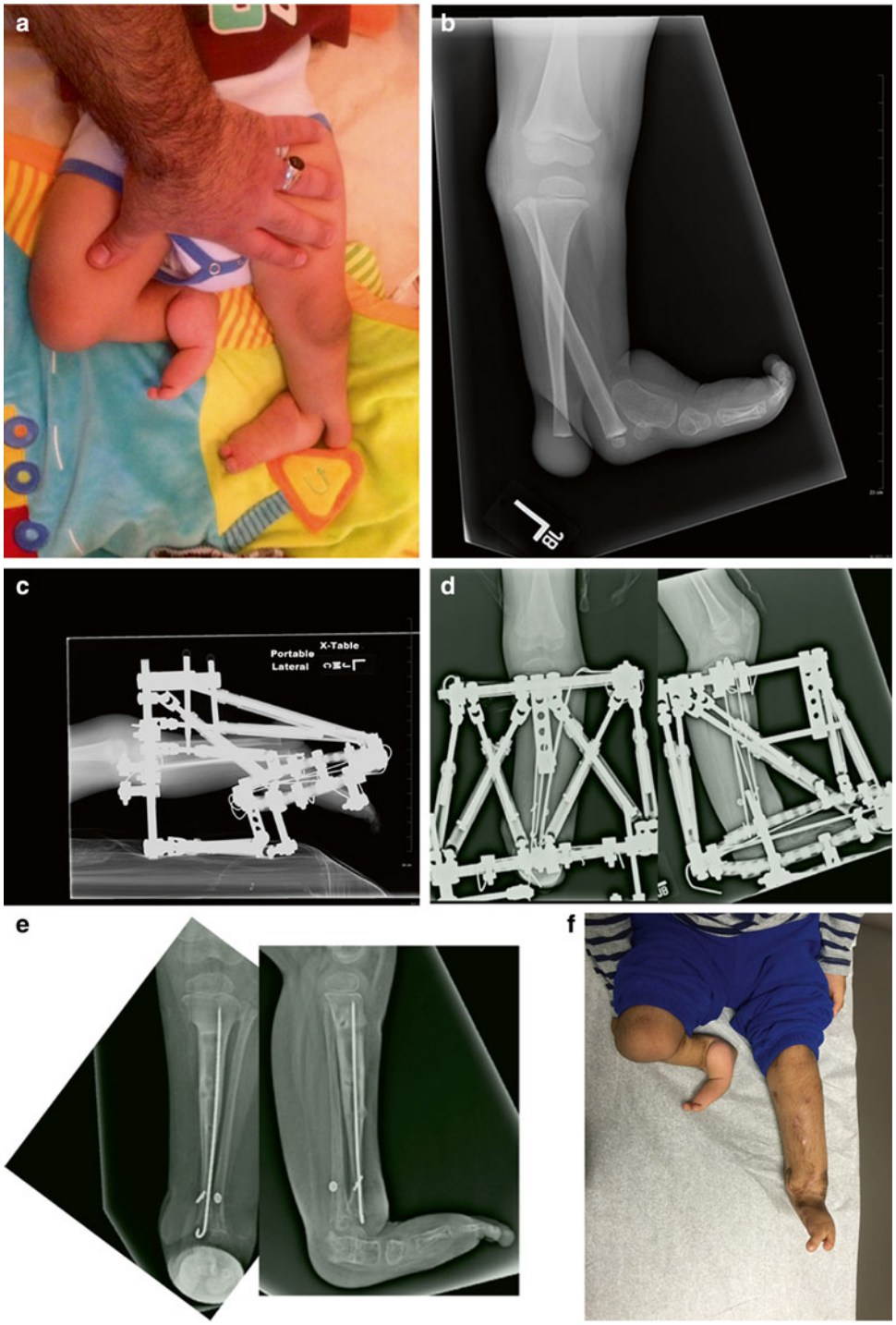


Fig. 24.6 (**a**) Paley type 2 variant: Clinical photo showing there is a cleft associated with the distal tibio-fibular diastasis. Notice that the foot goes with the fibula. There is also tibial hemimelia on the other side Paley type 5. (**b**) Radiograph showing the diastasis and cleft. (**c**) Radiograph of the TSF external fixator applied to the foot and tibia at onset of correction. Notice the 90° equinus contracture requiring the rings to be at 90° to each other. (**d**) AP and lateral radiographs after the correction is completed. The talus was centralized under the distal tibia. The syndesmotic repair was done using the Ziptite suture-washer implant. The two washers can be seen on the x-ray. The cleft was surgically closed. (**e**) Final AP and lateral radiographs showing the foot is plantigrade and the talus is centered under the tibia. The fibula is lateral to the talus. The cleft and diastasis are closed. (**f**) Final clinical photo from the front showing that the cleft is closed

suture with its two washers is used to reduce and compress across the diastasis. The incision is then closed, and the foot ring is reapplied with three new wires. This helps ensure that the foot remains in a plantigrade position.

The external fixator is left in place for 3 more months. The fibular wire as well as the transarticular tibial wire should be left in place even after fixator removal. The transarticular wire can be advanced into the calcaneus to allow for weight bearing. I prefer to leave both of these in place for 6 more months. This serves several purposes: prevention of fracture of the now osteoporotic tibia and fibula, stabilization of the ankle joint to prevent recurrence of equinus, and retardation of the faster-growing fibula to prevent recurrent relative overgrowth. Six months later, both wires should be surgically removed. A solid AFO is used until the wires are removed, after which the patient is placed into an

articulated AFO with a plantarflexion stop. Physical therapy is initiated after the transarticular ankle wire is removed.

Box 24.5. Surgical Tips

- Due to often aberrant anatomy, we recommend a mini-open incision for the Achilles tenotomy.
- A single 1.5-mm intramedullary wire protects the fibular physes at both ends and prevents epiphysiolysis.
- After the ankle arthroplasty, pass the wire antegrade through the center of the dome of the talus, then pass it retrograde through the distal tibia.
- A suture system is used to reconstruct the syndesmosis between the tibia and fibula.

Paley Type 3

The presence of a proximal and distal growth plate defines type 3. As such, the knee is usually mobile albeit often unstable due to absence of cruciate ligaments and depression or deficiency of part of the tibial plateau. The ankle plafond is present but often dysplastic, and thus does not have much motion despite its presence. The presence of the plafond differentiates it from type 2. Ankle diastasis is not typical, but some degree may be present depending on the severity of dysplastic changes of the tibial plafond.

Paley Type 3a (Fig. 24.7)

The foot is in equinovarus, and the tibia is also deformed. The circular external fixator is applied, and gradual correction of the bony deformity is carried out together with the foot, similar to the correction described for Paley type 2. At times, the foot deformity exceeds the bony deformity. After the lengthening and deformity correction of the tibia is completed, the distal tibia pins can be separated from the foot ring and connected to the proximal tibial ring. The foot and ankle are then freed to move independently with the distal ring. Once all of the deformities are corrected by distraction, the bone is allowed to consolidate and eventually the external fixator is removed. In these cases, the fibula can be treated in one of two ways: (1) resection of the mid-diaphysis of the fibula, in an attempt to create a fibular nonunion, or (2) distraction of the tibia without fixation of the fibula, to pull the fibula down to its correct station at the knee and ankle. The external fixator should always be extended to the femur to stabilize the knee joint.

Paley Type 3b (Fig. 24.8)

This type is often associated with syndromes (e.g., Werner's dysplasia). There is usually duplication of toes. This is often a familial condition (autosomal dominant) and bilateral. The bracket epiphysis can be oriented in any direction and does not always correspond to the deformity seen. The fibula is much longer than the tibia. The treatment in these cases is to consider the direction of the bracket in planning the surgery. To interrupt the bracket, the cartilage of the epiphysis and physis is cut, and an osteotomy is performed through the bone at the same level. To allow for acute correction of the deformity, part of the fibula must be resected. The acute correction is accomplished by an opening wedge osteotomy on the side of the bracket, with or without a partial closing wedge on the opposite side. The correction can be done with or without lengthening at the same time. With lengthening, it is done with the external fixator extending to the femur. Without lengthening, fixation is obtained with axial retrograde wires entering through the foot and crossing the knee.

Paley Type 4

In this type, there is a knee joint present. The degree of deficiency of the proximal tibia varies, but the knee is present and functional. The foot is in very severe equinovarus. The lack of a distal tibia makes a mobile ankle joint not possible. Articulating the talus with the distal fibula in a biologic arthroplasty has met with recurrent deformity and failure. Fusion of the talus to the distal fibular epiphysis is the goal of treatment for this type. Transfer of the fibula to the distal end of the tibia is also part of the reconstructive goal if no cartilaginous anlage exists.

Paley Type 4a (Fig. 24.9)

The salient feature in this type is the delayed ossification of part or all of the tibia. When part of the tibia is affected, it is always the distal part. An MRI is useful to image the articulations between the tibia and the femur and the tibia and the ankle joint. Based on these, a decision can be made as to whether the deformity of the tibia needs to be corrected in order to reorient the ankle to the knee. Since the fibula is longer than the tibia, two options can again be considered for managing the fibula: (1) resection to create a pseudarthrosis of the fibula, or (2) lengthening of the tibia relative to the fibula. The ankle joint, while present in these cases, is not functional. The goal is to create a plantigrade foot with a stable ankle. This usually requires distraction of the foot through the ankle joint, and then a secondary arthrotomy as described for type 2 deficiencies, to best match the articula-

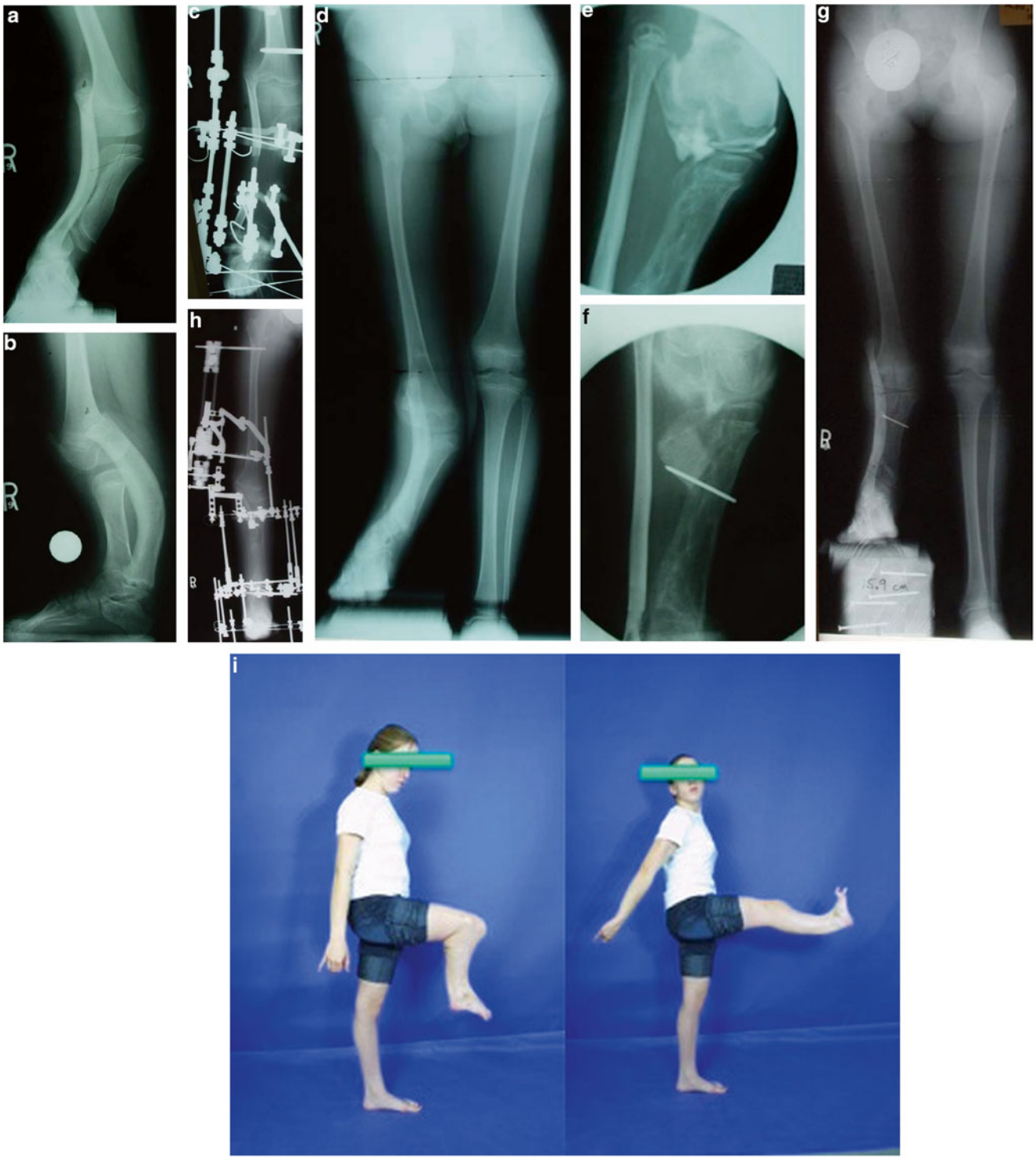


Fig. 24.7 (**a**, **b**) AP and lateral of Paley type 3a TH. (**c**) Lengthening of tibia with distal transport of fibula. (**d**) Recurrent deformity of tibia (valgus). (**e**) Arthrogram of knee showing depressed lateral plateau. (**f**) Hemi-plateau elevation to treat valgus. (**g**) Hemi-plateau elevation of lateral tibial plateau to correct the valgus and stabilize the knee. (**h**) Second lengthening. (**i**) Final result after second lengthening

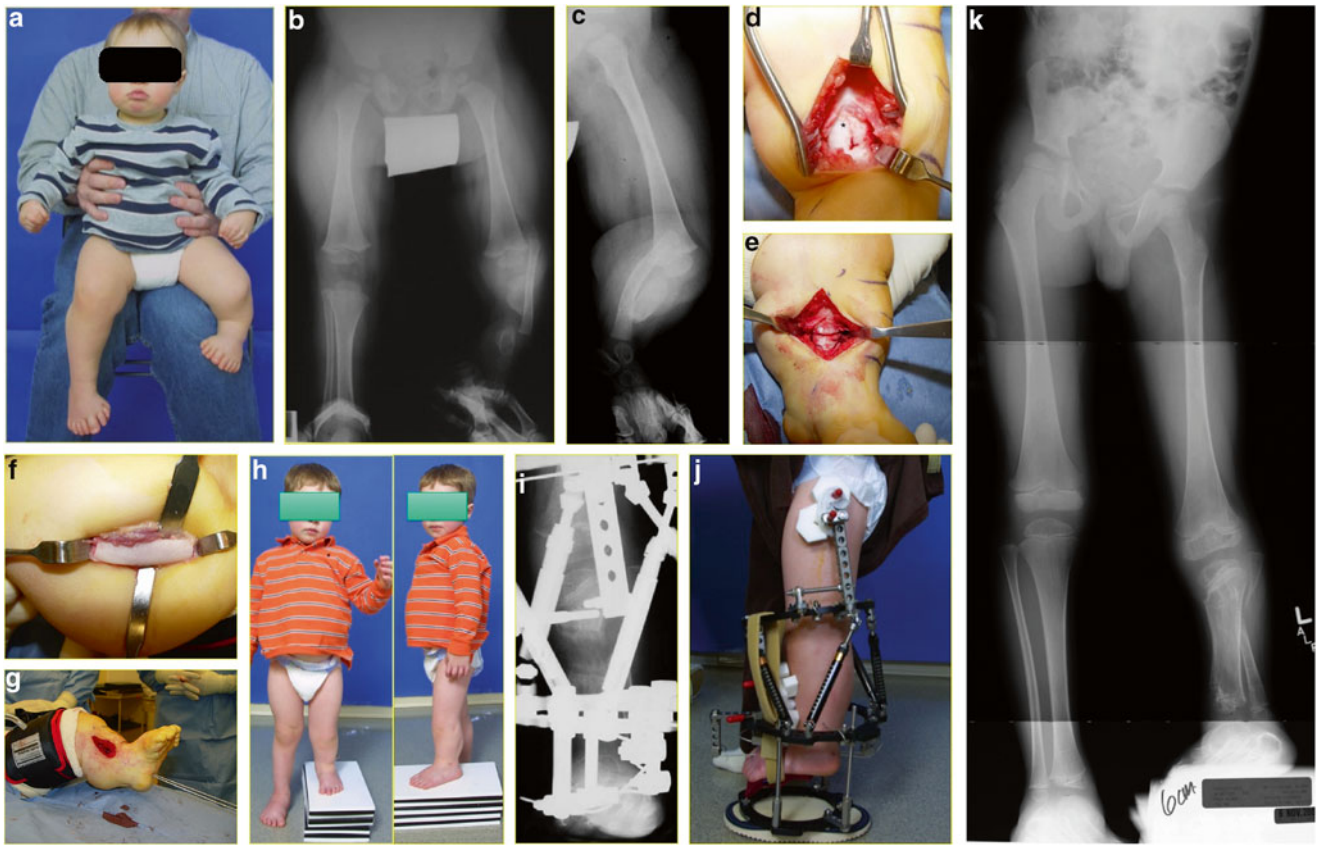


Fig. 24.8 (**a**) A 6-month-old boy with Werner's syndrome. Mother, one sibling, and grandmother have it, too. Note the syndactylized mirror hands and eight toes. (**b**, **c**) Delta left tibia Paley type 3b. The bracket epiphysis is facing anteromedial. (**d**) Cartilage of the bracket epiphysis intact (*asterisk*). (**e**) Cartilage of bracket epiphysis cut with osteotomy. (**f**) Fibulectomy. (**g**) Leg straightened by osteotomy of tibia and pinned retrograde from the foot. (**h**) After correction and amputation of extra toes. (**i**) Lengthening of tibia with external fixator to femur, tibia, and foot. (**j**) TSF device for lengthening. (**k**) Radiograph after lengthening. He will require serial staged lengthening procedures in the future to equalize leg length discrepancy by skeletal maturity

tion between the talus and tibia. The syndesmosis may or may not need to be fixed with the suture technique described for type 2. Biologic arthroplasty may also be necessary.

The unossified portion of the tibia will eventually ossify after many years. To accelerate this process, bone morphogenic protein (BMP) can be inserted into the cartilage. This is an off-label use of INFUSE (Medtronic, Memphis, Tennessee). The basis of its use in tibial hemimelia comes from the senior author's experience with congenital femoral deficiency, using BMP in a region of delayed ossification in the femoral neck [88]. Ossification of the tibia facilitates lengthening and deformity correction of the tibia through bone. If sufficient parts of the tibia are bony, an osteotomy can be made through the bony portion and pins placed in the bony portion. If an insufficient portion of the tibia is ossified to allow for external fixation, then open surgery is performed to acutely realign the foot with a tibial osteotomy, combined with resection of part of the fibular diaphysis. To ossify the tibial anlage (nonossified portion of the tibia), BMP is inserted into drill holes in the cartilage. Stabilization of the osteotomy is achieved with retrograde axial Kirschner wires through the foot and up the tibia. In most cases, ossification of the anlage is already seen by 3 months after BMP implantation surgery. Lengthening is usually done at 1 year after BMP insertion.

Paley Type 4b (Fig. 24.10)

This is the most common type 4 seen. The proximal tibial epiphysis, physis, and metaphysis are well formed, a patella is present, and active and passive knee motion are present through a normal range. The fibula is overgrown and proximally migrated and the foot is in extreme varus and, to a lesser extent, equinus. The goal of treatment is to correct the foot deformity, fuse the talus to the end of the fibular epiphysis distally, and transfer the fibular diaphysis to the distal end of the tibia proximally. This makes a one-bone leg. While this can be accomplished in one stage, it is safer, easier, and more length preserving to do this in two stages.

The first stage involves applying a circular external fixator to the femur, tibia, and foot. The knee is locked in extension by inserting an axial wire through the distal tip of the

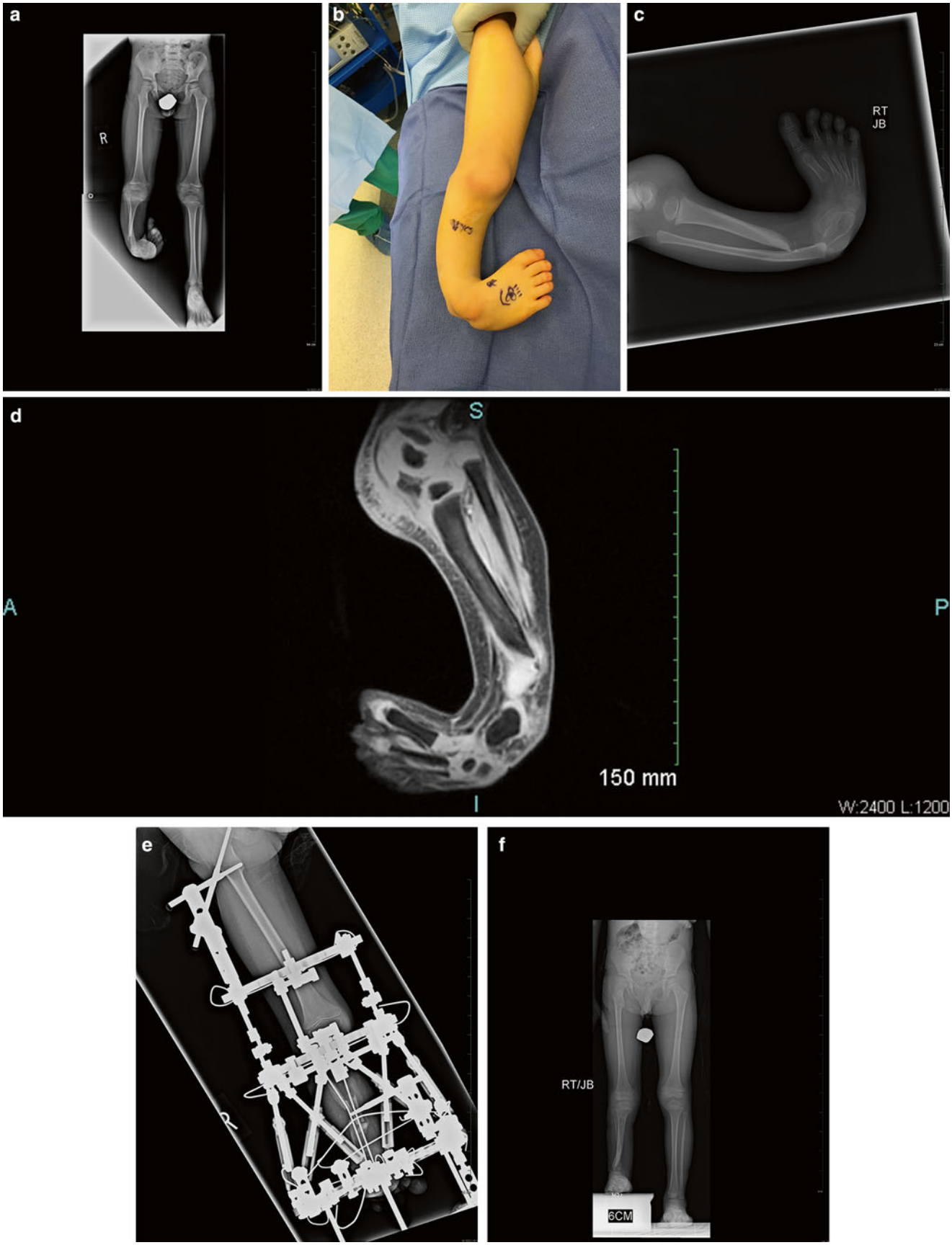


Fig. 24.9 (**a**) Preoperative AP radiograph showing Paley type 4a tibial hemimelia with severe equino-varus. Note the non-ossified region of the distal tibia. (**b**) Preoperative clinical photo showing the severe equino-varus deformity of the foot. (**c**) Preoperative lateral radiograph showing the non-ossified region of the distal tibia. (**d**) MRI showing a cartilage anlage is present articulating with the ankle joint. (**e**) A distal tibial acute angular correction combined with shortening was performed to correct the foot to a plantigrade position. The fibula was also acutely shortened. BMP was inserted into the anlage via drill holes. The tibia and fibula were gradually lengthened through a separate proximal osteotomy. The distraction gap can be seen. A femoral external fixator (to protect the knee) with hinges, is connected to a tibial external fixator for lengthening. (**f**) Final long standing AP radiograph showing the bone is straight and the distal part is ossified. In future additional lengthenings will be required for leg length equalization

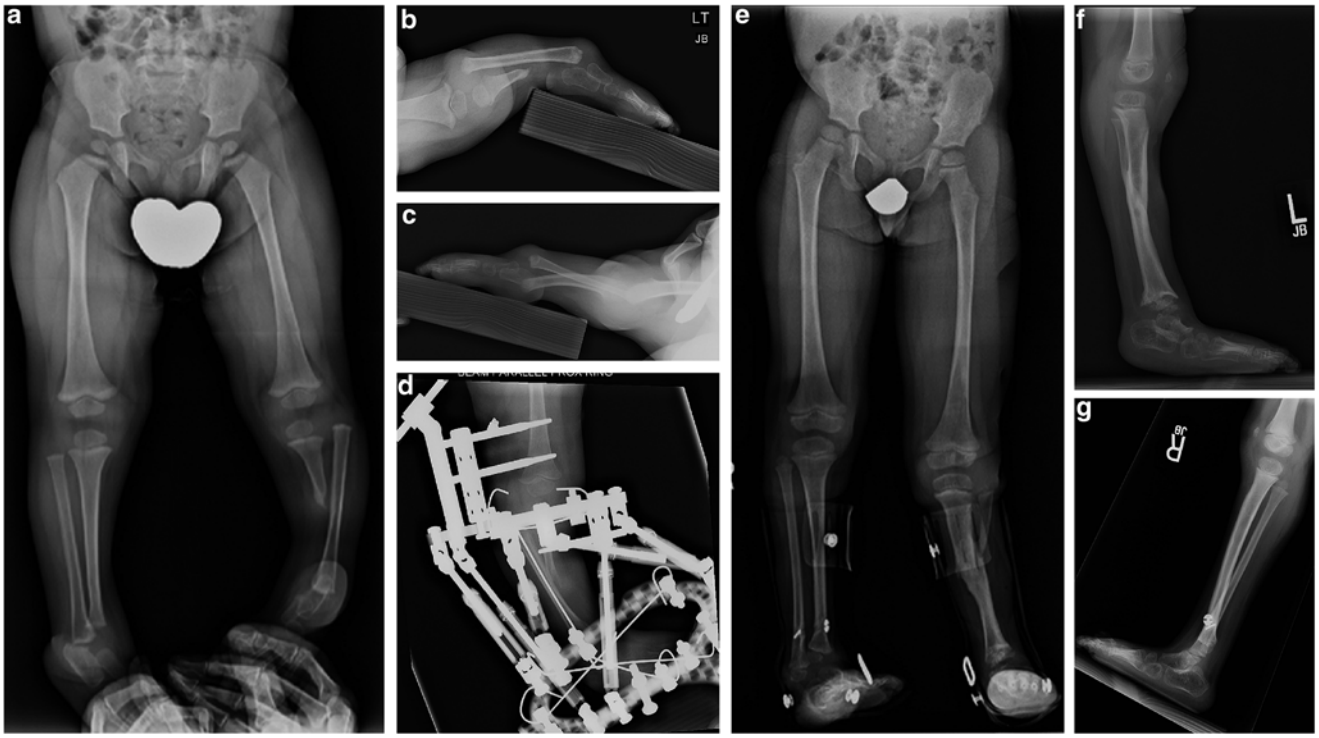


Fig. 24.10 (**a**) AP radiograph of bilateral TH with Paley type 4b on the *left* and 2 on the *right*. (**b**) Lateral radiograph of left leg Paley type 4a with severe equinovarus. (**c**) Lateral radiograph of right leg Paley type 2 with severe equinovarus. (**d**) Application of TSF to left foot and femur to correct the equinovarus deformity and bring the foot under the talus. A temporary epiphysiodesis wire is in place to prevent epiphysiolysis of the fibula during distraction. (**e**) After correction the foot was fused to the distal fibular epiphysis and the fibula was transferred to the tibia to form a one-bone leg. On the *right side* the treatment discussed in Fig. 24.4 was carried out to achieve a stable plantigrade foot. The diastasis on the right was also corrected. (**f**) Lateral of left foot. The fusion at the ankle may not be complete. The foot equinus has partially recurred. (**g**) The right foot is plantigrade and maintains some range of motion after the biologic arthroplasty

tibia, crossing the knee and inserting into the femur. The external fixation pins are in the femur only (two half pins at the femoral neck level and one wire at the ring level) and not in the tibia. This avoids having a contaminated pin site in the operative field at the time of the second stage surgery. Wires are used in the foot (three in the calcaneus and foot and one transverse in the talus). The foot is then distracted away from the tibia to correct its deformity and to gain length. This creates a large space between the talus and the end of the tibia. The goal is placing the talus distal to the lateral malleolus. To prevent epiphysiolysis of the fibula during distraction, a temporary epiphysiodesis wire is inserted into the fibula to protect its two growth plates (as described for type 2).

Once the foot is in the corrected position, the second stage surgery is carried out. The first step is to remove the foot ring and cover the pin sites with an occlusive dressing to minimize contamination during surgery. After the leg is prepped and draped free, a transverse lateral incision is made over the distal tip of the fibula. The distal epiphysis of the fibula and the dome of the talus are exposed. The capsular connections between them are cut to mobilize both bones relative to each other. A small incision is made proximally over the fibular wire. The fibular epiphysiodesis wire is cut either proximally or distally and then removed. A new straight wire is immediately inserted in the same track to protect the fibula from fracture. The fibula is often osteoporotic at this stage and without an intramedullary wire, it can easily break upon manipulation of the foot or leg. The cartilage at the end of the fibula is incised and pared back until the ossific nucleus of the fibula is reached. If there is no ossific nucleus, then BMP can be inserted to get the epiphysis to ossify. The talus is also incised parallel to the sole of the foot to the level of its ossific nucleus. The two ossific nuclei are then aligned, and the wire is advanced from the proximal end through the talus and sole of the foot to hold the foot plantigrade to the fibula.

A longitudinal anterior incision is made over the tibial diaphysis and extended distally. Fasciotomy of the anterior compartment is carried out. The anterior compartment muscles are elevated off of the tibia, and a path is then dissected to the fibula. The fibula is exposed subperiosteally. The wire in the fibula is backed out distally to the level of the planned osteotomy. The fibula is then cut at the level of the distal end of the proximal tibia. The fibula is then shifted over to the tibia. The tibia can either be split longitudinally to accept the end of the fibula inside of it, or it can be cut transversely to allow end-to-end apposition of the two bones. In both cases, the fibular wire is advanced across the proximal tibia up to the joint line of the knee. The wire in the foot is the same wire in

the fibula that is advanced across into the tibia. The external fixation wires in the foot are reinserted and connected to a ring. Struts are now used between the upper femoral ring to the distal foot ring. The external fixator maintains the alignment of the foot and knee to achieve fusion of the tibia and fibula proximally and of the fibula and talus distally. This usually takes 3 months. After that, the external fixator is removed, leaving one wire buried in the foot and fibula to protect the fibula for a few months longer. The knee motion is restored with physical therapy. In the future, lengthening of the one-bone leg can be carried out without crossing the knee joint.

Paley Type 4c (Fig. 24.11)

In this type, there is only a proximal tibial epiphysis present and no proximal tibial physis. The proximal tibial epiphysis is often unossified at an early age. The foot is in severe

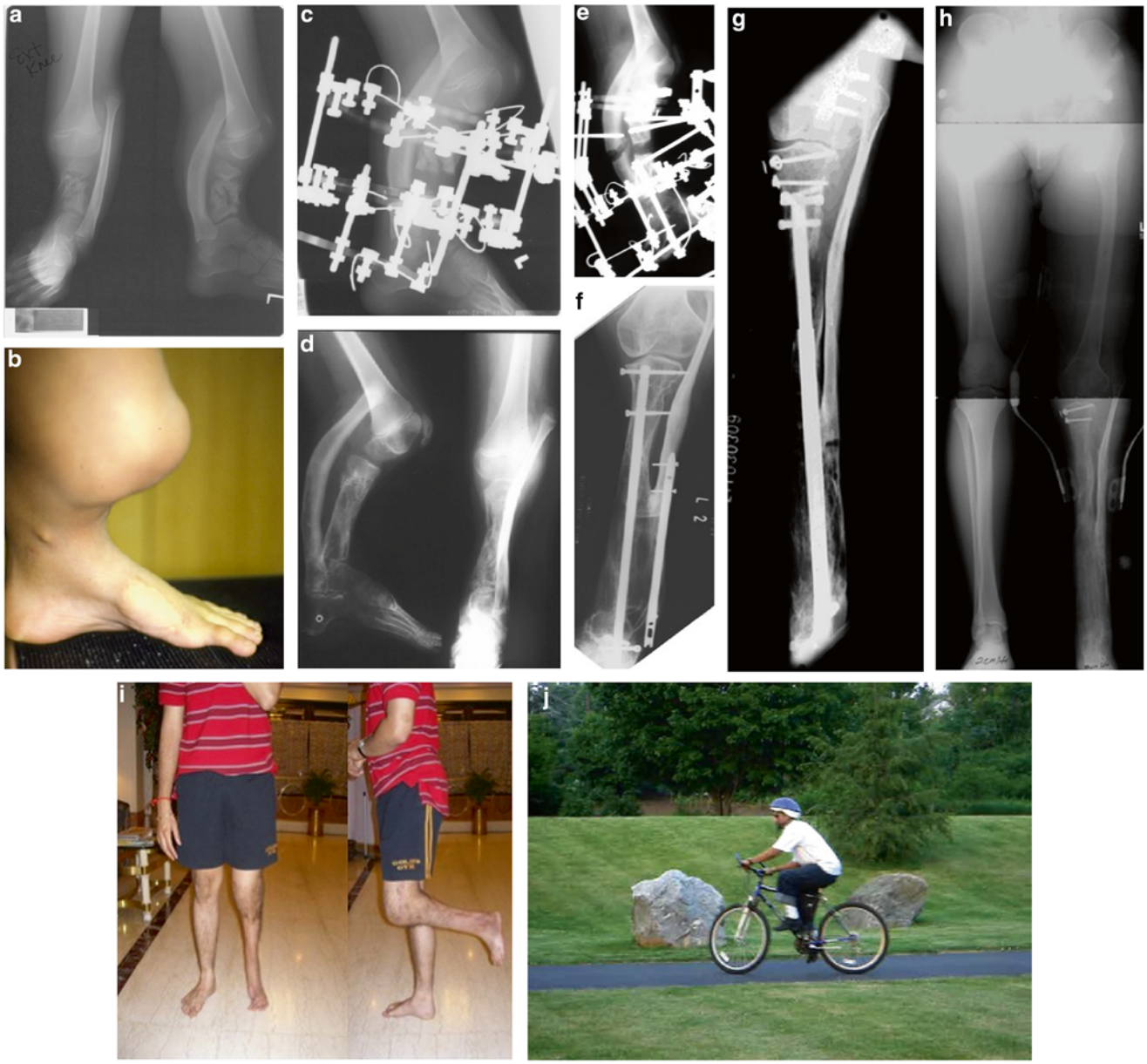


Fig. 24.11 (**a**) Radiograph of Paley type 4c. There were eight metatarsals. They were resected and transferred on a vascular pedicle to the leg to connect with the tibial epiphysis. (**b**) Clinical photo of the same. (**c**) Osteotomy across the three transplanted metatarsals and the fibula for first lengthening (6 cm). (**d**) After first lengthening and bony consolidation. (**e**) Second lengthening of tibia and fibula with Ilizarov device (8 cm). (**f**) Third lengthening over a nail in tibia and fibula (10 cm). (**f**) Fourth lengthening using ISKD internal lengthening device (8 cm). (**g**) Final radiographic result after four lengthenings and 32 cm gained. (**h**) Clinical appearance and knee function 0–90° after 32 cm of lengthening. (**j**) He is sports active, plays soccer, and rides a bicycle

Box 24.6. Synostosis and Ankle Fusion Tips

- The Paley type 4 lacks a distal tibia, and the goal is usually to obtain a fusion of the tibia remnant and the fibula proximally, and a fusion of the distal fibula and talus distally.
- The femoral ring is kept relatively high on the femur to stay out of the surgical field for subsequent surgeries.
- Protect the fibula with an intramedullary temporary epiphysiodesis wire as previously described. This is also important intraoperatively while working on either end of the fibula.
- Once the proximal fibula is brought in line with the proximal tibia, switch the transverse wires in the fibula from the distal ring to the proximal ring. Then focus on correcting the ankle deformity.
- Remove extraneous wires several days prior to surgery to minimize infection risk.
- A longitudinal split in the tibia like a clothespin allows greater surface area and healing potential.
- BMP-2 can be used to promote ossification of cartilage.
- Use the external fixator and intramedullary wires to provide stability and maintain correction until fusion is achieved.
- An arch wire can help provide compression to the talo-fibular fusion.

equinovarus, and the fibula is relatively overgrown and proximally migrated at the knee. The treatment preferred is also a two-stage surgery, with the first stage using a circular fixator to correct the foot deformity. There is often a knee flexion contracture present in these cases. It is also treated by distraction of the knee and foot with the same external fixator. A temporary fibular epiphysiodesis wire is used. A transverse wire can be added to the fibula. Initially this wire is connected to the distal foot ring, and the fibula and foot are both distracted distally, bringing the fibular head under the epiphysis of the tibia. Once this is achieved, the transverse fibular wire is detached from the foot ring and connected to the proximal tibial ring. This allows distraction of the foot deformity, and the foot is pulled distally to the end of the fibula.

In order to minimize the risk of infection, the fibular wire is removed in the office prior to going back to surgery for the second stage surgery. The same treatment is carried out to fuse the talus to the fibula as described in type 4b. At the knee, the fibula is fixed to the tibial epiphysis. If the epiphysis is unossified, then BMP is inserted into drill holes in the cartilage. The proximal fibular epiphysis is pared down as was done for the distal epiphysis. The intramedullary wire used to fix the foot is advanced proximally to transfix the proximal tibial epiphysis. Finally, the foot wire and ring are reapplied and connected with six struts to the proximal ring. It is important to use an external fixator because the axial foot wire cannot maintain the foot in a plantigrade position. The fixator remains in place for 3 months until the fibula fuses to the tibial epiphysis proximally and the talus distally. Knee range-of-motion exercises ensue after that.

An alternative form of treatment in this type is possible when there are duplicate metatarsals, which can be transplanted into the space between the tibial epiphysis and the talus. The metatarsals can be taken as free grafts, but they are preferably taken as vascular pedicle grafts flipped proximally on their pedicle. Iliac crest bone grafting between the metatarsals promotes them to unite together into one-bone mass. Once this occurs, an osteotomy across the new tibia and the fibula can be done for lengthening. The fibula remains proximally dislocated as long as it does not impede knee motion. Repeated lengthenings are needed to equalize limb length discrepancy.

Paley Type 5

Complete absence of the tibia presents the biggest challenge for reconstruction because there is no knee joint. While ankle fusion gives good function with little disability, knee fusion leads to significant disability for sitting and climbing stairs. It is preferable to avoid a knee fusion. Even if active knee motion cannot be achieved, a mobile knee joint supported by a brace is preferable to a knee fusion. This is not dissimilar to a paralytic knee from polio. Therefore, the following two methods have been developed to reconstruct the knee.

Paley Type 5a (Fig. 24.12)

If a patella is present, it can be converted to a tibial plateau. This original idea was first published by Michael Weber [78, 79]. The patella is moved on a vascular pedicle from its normal position, anterior to the femur, to the distal end of the femur. The fibula is centralized to the patella and its epiphysis fused to the patella. This procedure is referred to as the Weber patellar arthroplasty or Weber procedure.

The Weber procedure can be performed acutely (as described by Weber) with gradual correction of a flexion contracture of the knee joint, using a hinged external fixator, or following staged gradual distraction of the fibula to first correct the knee flexion contracture (according to Paley). When performed acutely, the foot deformity is gradually distracted at the same time as the gradual knee contracture correction. When performed after the distraction, the foot is distracted away from the fibula as described above with a temporary epiphysiodesis wire in place. Once the femur,

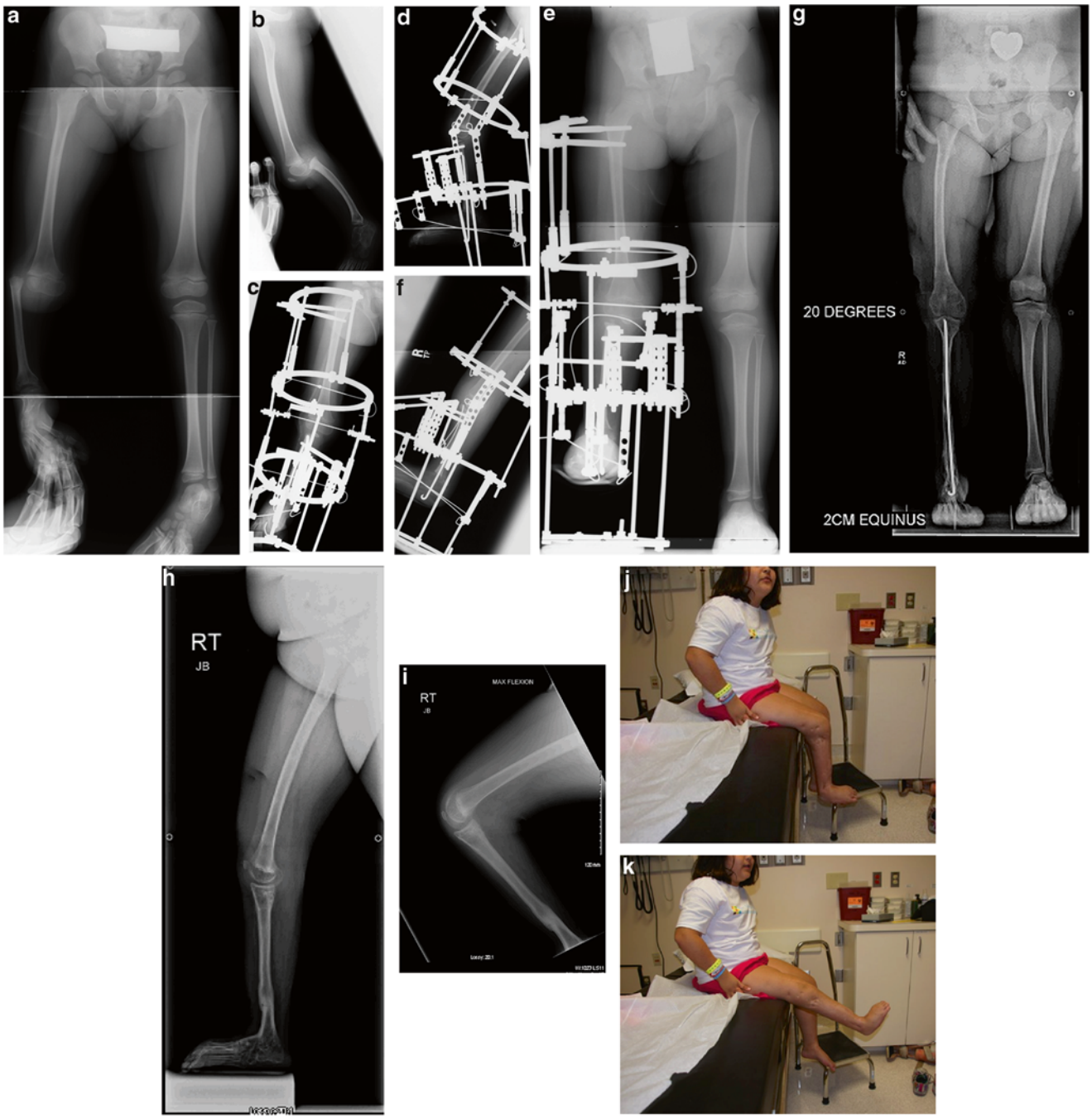


Fig. 24.12 (**a**, **b**) AP and lateral radiographs of Paley type 5a TH. The foot was previously treated and remains in an equinovarus deformity. (**c**, **d**) AP and lateral radiographs after the Weber patellar arthroplasty was performed and the patella jointed to the fibula. The ankle was fused in a plantigrade position for the foot. Initially there was a flexion contracture of the knee. The circular external fixator was used to correct this. (**e**, **f**) After distraction the knee is straight. (**g**) After removal of the fixator an intramedullary wire was used to help protect the fibula. (**h**, **i**) AP and lateral of the knee and tibia after healing from the Weber. Note how well remodeled the knee appears. Due to leaving a small fragment of the patella behind a new patella developed. The knee has excellent active range of motion. (**j**, **k**) Clinical photos of the leg after one lengthening. Note the active knee flexion and extension

fibula, and foot are all aligned, the Weber procedure can be performed through a longitudinal anterior incision over the distal femur and proximal fibula. At the distal end, the ankle fusion can be performed as described above through a transverse lateral incision.

Weber Patellar Arthroplasty (Fig. 24.13)

An anterior midline incision is made over the distal femur and proximal fibula. Medial and lateral skin flaps are elevated to expose the fibula and distal femur. The patella is

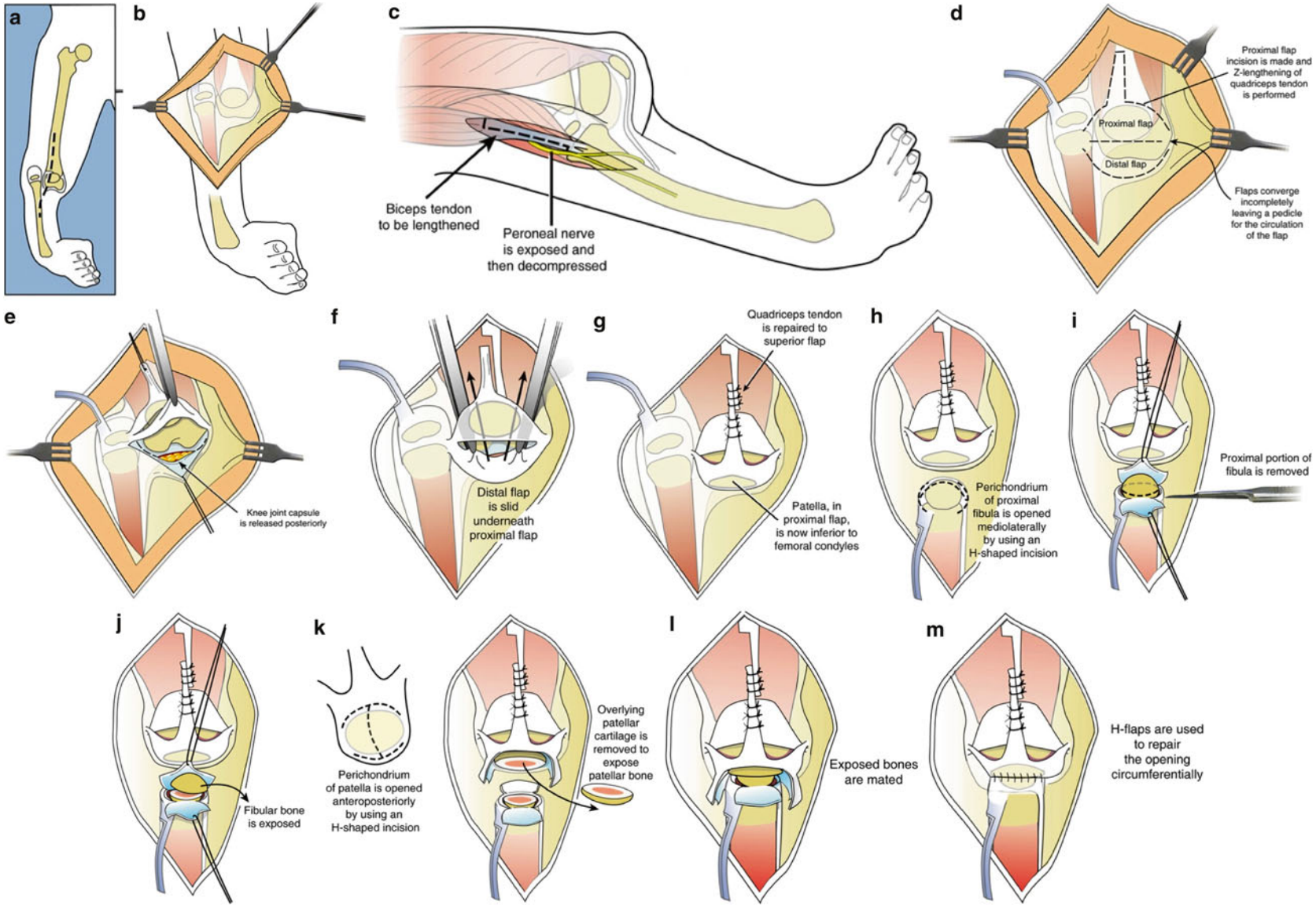


Fig. 24.13 (**a**) Weber patellar arthroplasty incision. (**b**) The underlying fascia is exposed over the patella, femur, and tibia. (**c**) The peroneal nerve should be exposed and decompressed and the biceps tendon lengthened. (**d**) The two visor flaps are marked out. (**e**) The visor flaps are incised and the posterior capsule is cut with great care to identify the vessels posteriorly so as not to injure them. (**f**) The two visor flaps are flipped so that the proximal one goes distal and the distal one goes proximal. (**g**) The quadriceps tendon is repaired and the visor flaps sutured together. (**h–k**) Corresponding H-flaps are incised into the perichondrium of the patella and the fibular head and the ossific nucleus of the fibular head and the patella exposed by resecting some of the overlying cartilage. (**l**) The patella and fibular head are mated to each other. (**m**) The H-flaps are sutured together

Box 24.7. Weber Patelloplasty Tips

- Correct knee flexion contractures as much as possible with frame distraction before surgery. A posterior capsulotomy may be necessary.
- Find and protect the peroneal nerves and the posterior tibial neurovascular bundle.
- If there are remnants of the medial or lateral hamstrings, these can be preserved and reattached to the fibula at the end of the operation.
- Keep the visor flap pedicles as wide as possible. Deepen the cuts just enough to allow the mobility to slide them around each other. Usually the upper patella flap goes underneath.
- Patellas can be present in various shapes. A vertical patella can be easily split, such that a small remnant is translated superiorly with the visor flap and form a new patella.
- The H-flaps should be vertical on one side and horizontal on the other and should be determined by the anatomy and shape of the patella and fibula.
- Cut down into the cartilage of the patella and proximal fibula until reaching the ossific nucleus. If these are not present, then consider placing BMP-2 to promote ossification.
- Suture the posterior H-flap first before fixing the patella and fibula with a wire.

identified. Two "visor" flaps are outlined with a marking pen. At the medial and lateral ends, the pedicle for each "visor" is kept as wide as possible. The proximal visor flap contains the patella. The distal visor flap is all capsular. In the Paley modification, the distal visor flap includes a small part of cartilage of the inferior pole of the patella. This allows a new patella to form anterior to the femur. A Z-lengthening of the quadriceps tendon is performed as part of the proximal visor flap. If the fibula or patella is unossified, Weber recommends using two perichondral H-flaps to get the two cartilage surfaces to heal together. To ensure bone-to-bone healing, we prefer to insert BMP into drill holes in the cartilage of the patella and fibular epiphysis. To avoid injury to the popliteal vessels, it is preferable to find and protect them before completing the posterior capsulotomy.

Since the knee and ankle were predistracted, they can both be fixed by one axial wire across the knee joint. The circular external fixator is used only as a holding device to ensure full knee extension and a plantigrade foot. The medial and lateral collateral ligaments should be reconstructed using local tissues (biceps tendon laterally and semitendinosus medially). After 3 months, the external fixator is removed and the trans-knee wire shortened to span only the fibula and foot. After the external fixator is removed, the patient goes into a cast for a month; when the cast is removed, they are fitted for a KAFO brace. A brace is needed for many years until the knee is stable enough to allow walking without the brace. This is usually after age 10, when adequate hypertrophy of the joint surface and fibula has occurred.

Type 5bi (Fig. 24.14)

If there is no patella, but the fibula is autocentralized, then there is usually a quadriceps muscle in continuity to the fibula. The distal femur is usually less dysplastic in these cases. The knee still presents with a fixed flexion contracture and the foot presents as with type 5a, dislocated and in extreme varus and equinus. The treatment is to distract the knee contracture until the fibula and femur are collinear with each other. The foot should also be distracted relative to the fibula to centralize it under the distal end of the fibula. This is accomplished with an Ilizarov device on the femur using hinges and a circular ring fixator for the foot correction. This permits simultaneous correction at both ends of the bone. This is possible because the fibula is autocentralized. If the fibula is dislocated as in type 5bii, a circular fixator is needed for both knee and ankle and the correction is done sequentially as for type 5a. Once the distraction correction at both knee and ankle is completed, a second stage surgery is performed to reconstruct collateral ligaments at the knee and to advance the quadriceps muscles onto the fibula. An ankle fusion as described above is performed. The distal ring on the foot is removed and then reapplied in this surgery. All wires near the knee are removed in the office at least 1 week prior to the surgery. The external fixator is left in place for another 3 months and then removed. A spica cast is applied for a month and then an HKAFO brace is made with a hinged knee and solid ankle.

Type 5bii (Fig. 24.15)

If there is no patella and the fibula is dislocated, the fibula can be centralized by distraction. First, a temporary epiphysiodesis wire is inserted up the fibula and hooked over each epiphysis at both ends. This is to protect both proximal and distal fibular physes. An Achilles tendon tenotomy is carried out. Then, one proximal and one distal circular fixator ring is applied with two half pins in the proximal femur (one up the femoral neck and one transverse at the level of the lesser trochanter) and one wire on the proximal ring in the mid-femur. A distal ring on the foot is applied with three anterior posterior wires in the foot (one from posterior midcalcaneus to anterior mid-forefoot, one from posteromedial calcaneus to anterolateral forefoot, and one

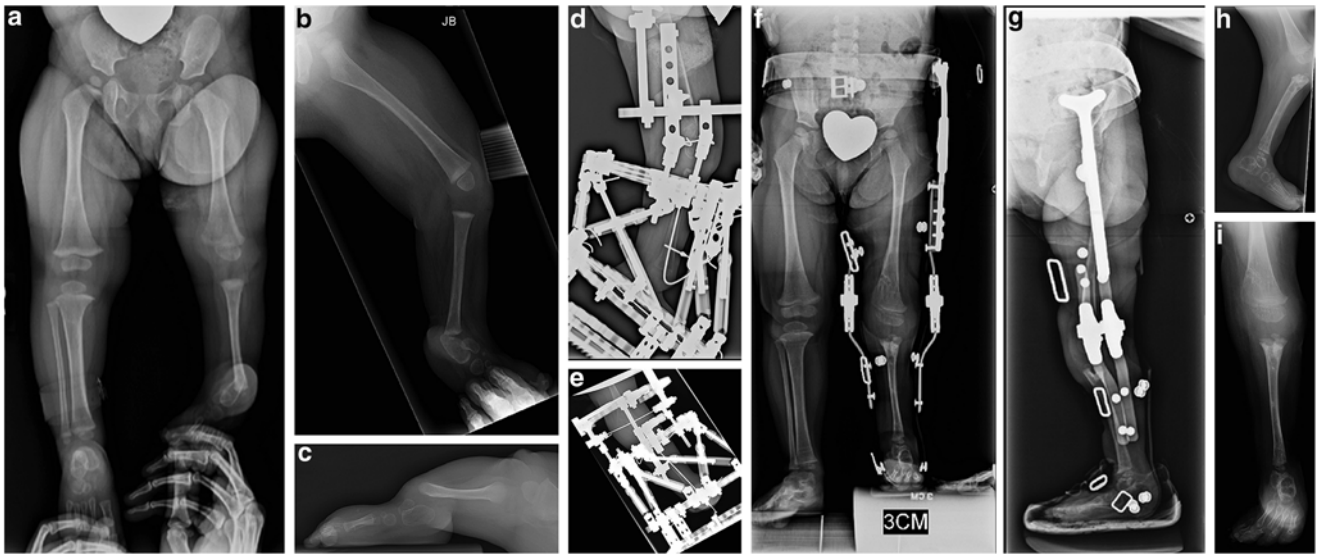


Fig. 24.14 (**a**, **b**) AP and lateral radiographs of Paley type 5b (i) TH. The fibular is autocentralized on the femur. For this reason it has hypertrophied. The knee joint has a fixed flexion deformity. (**c**) The foot is in severe equinovarus relative to the fibula. (**d**, **e**) Hinged construct used to correct knee contracture. A knee hinge wire is located across the distal femur. Taylor Spatial Frame is used to correct foot contracture gradually. A temporary epiphysiodesis wire is in place in the fibula to prevent epiphysiolysis. (**f**, **g**) After the knee and foot are corrected, the knee is reconstructed by open surgery to do a quadricepsplasty and to reconstruct the collateral ligaments. The ankle is fused: talo-fibular fusion. The external fixator is left on for 3 months after this second surgery. It is then removed and a brace is used to support the leg. (**h**, **i**) The distal fibular epiphysis is fused to the talus in a plantigrade position. The knee is well reconstructed

from posterolateral calcaneus to anteromedial forefoot) and one transverse wire across the ossific nucleus of the talus. One wire is inserted into the fibula and connected to the foot ring. The spatial frame planning is done relative to the proximal ring for the knee joint contractures. Once the fibula is reduced at the knee and the knee flexion contracture eliminated, the fibular wire is connected to the proximal ring and the spatial frame planning repeated for the distal ring and foot contractures. When both ends of the fibula are aligned to the knee and ankle, the second stage surgery can be performed.

Paley Centralization of the Fibula on the Femur (Fig. 24.16)

Through a midline anterior incision, the distal femur and the proximal fibula are exposed. The peroneal nerve is liberated and decompressed from the fibula. The biceps tendon is split with half of its tendon left attached to the fibula to be used as a lateral collateral ligament. The other part is transferred to the fibula anteriorly to rebuild a quadriceps muscle. On the medial side, the semitendinosus muscle is also transferred anteriorly. The tensor fascia lata and its iliotibial band are also harvested and transferred. The adductor magnus is also transferred. These four muscles are connected to the quadriceps muscle. The quadriceps muscle usually ends in the mid-thigh and does not continue to the knee. The transferred muscles are all connected together with the lateral and medial muscles balancing each other and centralizing themselves to the iliotibial band, which acts as a central rib for the connection of all of these muscles. It is eventually attached to the front of the fibula to act as the patellar tendon.

Before connecting the transferred muscle, it is important to anchor the fibular head to the femur. This is done by creating an interosseous ligament between the femur and the fibula. One can use allograft or the half of the biceps tendon for this purpose. This single step prevents subluxation of the fibula from the femur. As dislocation was the main reason for failure of the original Brown procedure, the interosseous ligament prevents subluxation from occurring while permitting hinge flexion of the two bones. The ankle is fused as previously described. A wire is inserted from the foot through the fibula and then through the knee joint. The foot ring, which was removed at the start of the procedure, is reapplied with the circular fixator struts. The foot is immobilized in a plantigrade position and the knee in full extension. Three months later, the external fixator is removed, but the medullary wire crossing the ankle and knee is left in place. A cast is used for 1 month and then a KAFO is prepared. The wire is removed 6 months later and knee rehabilitation is started. Other than amputation, the alternative or fall back operation for this type of tibial hemimelia would be a knee fusion. In unilateral cases, this is a reasonable alternative.

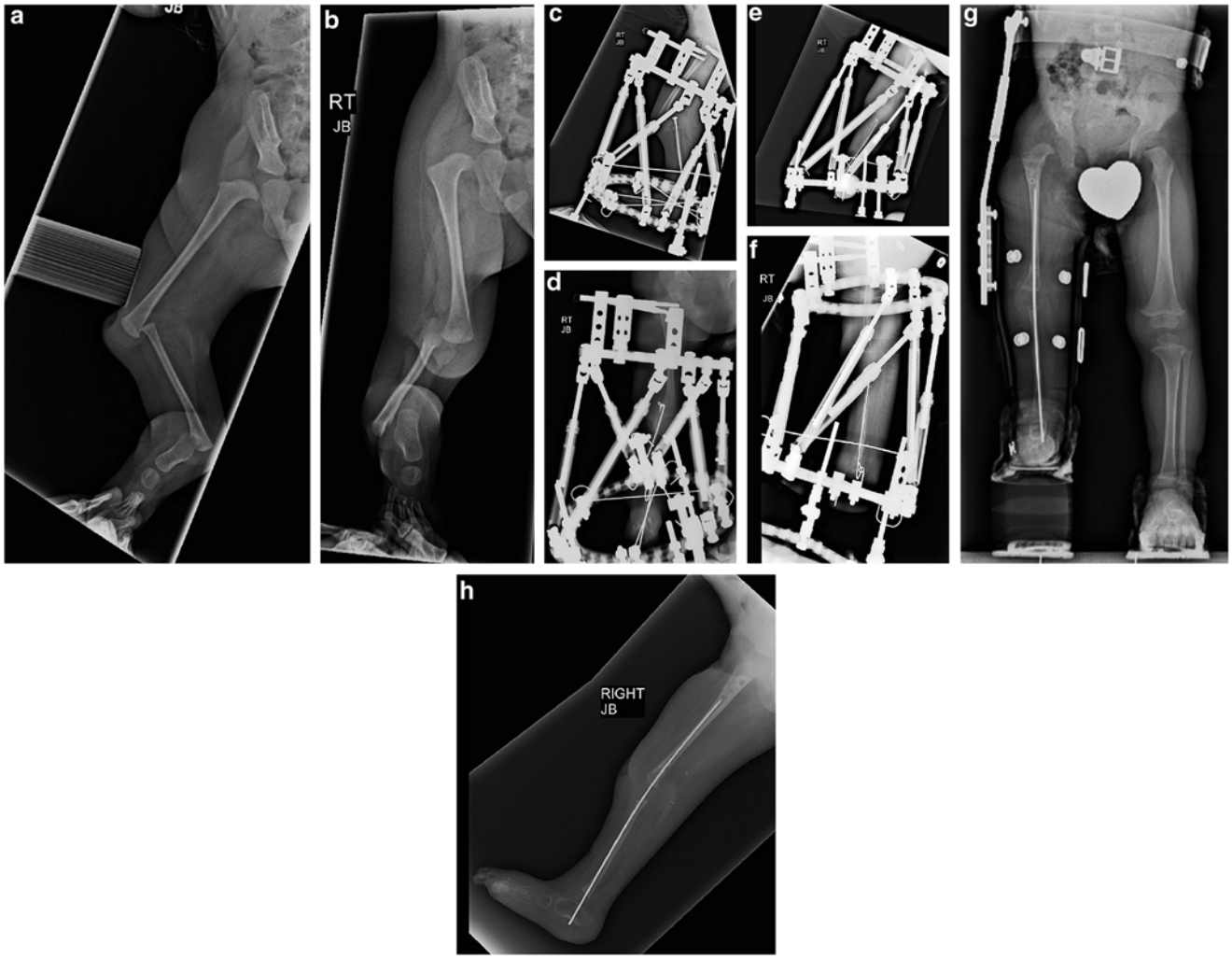


Fig. 24.15 (**a**, **b**) Radiograph of Paley type 5b (ii). There is a dislocation of the fibula on the femur and the foot on the fibula. There is a severe equinovarus of the foot and flexion contracture of the knee. (**c**, **d**) A Taylor Spatial Frame is applied from the femur to the foot with one wire across the tibia. A temporary epiphysiodesis wire is in place in the fibula. (**e**, **f**) After 3 months of distraction the knee contracture is eliminated and the foot is reduced below the end of the distal fibula. (**g**, **h**) The foot is fused to the distal epiphysis of the fibula and the biceps, semitendinosus, tensor fascia lata, and adductor magnus muscles are all transferred to the knee to substitute for the missing quadriceps muscles. A temporary arthrodesis wire is placed across the knee and ankle. The external fixator was removed 3 months after the knee-ankle surgery. The wire across the knee and ankle is left in place for 6 months to allow better loading and minimize the risk of recurrence. It is then removed

Summary

Through-knee amputation remains the most commonly used procedure for complete tibial aplasia. It is certainly a reliable option that can be carried out by most orthopedic surgeons. It should be seriously considered in unilateral cases. In bilateral cases of Paley type 5, complete tibial aplasia reconstruction by one of the techniques described above should be considered, especially on the side that has a patella.

Amputation is probably overused in Paley type 2, 3, and 4 tibial hemimelia. In the senior author's opinion, reconstructive results for these types are reliable and successful in achieving a functional lower extremity. For type 5a, where a patella is present, the results of a Weber patellar arthroplasty in the hands of experienced surgeons are also very satisfactory. The reconstructive options for tibial hemimelia have improved greatly over the past two decades. With continued success and improvements, they may one day overtake the amputation option. Other anatomic variants of tibial hemimelia including duplication of fibula, tarsals, metatarsals and toes, and secondary femoral condyles, have not been discussed. They are not included in the classification either. These associated anatomic variants must be considered in the treatment but are beyond the scope of this chapter.

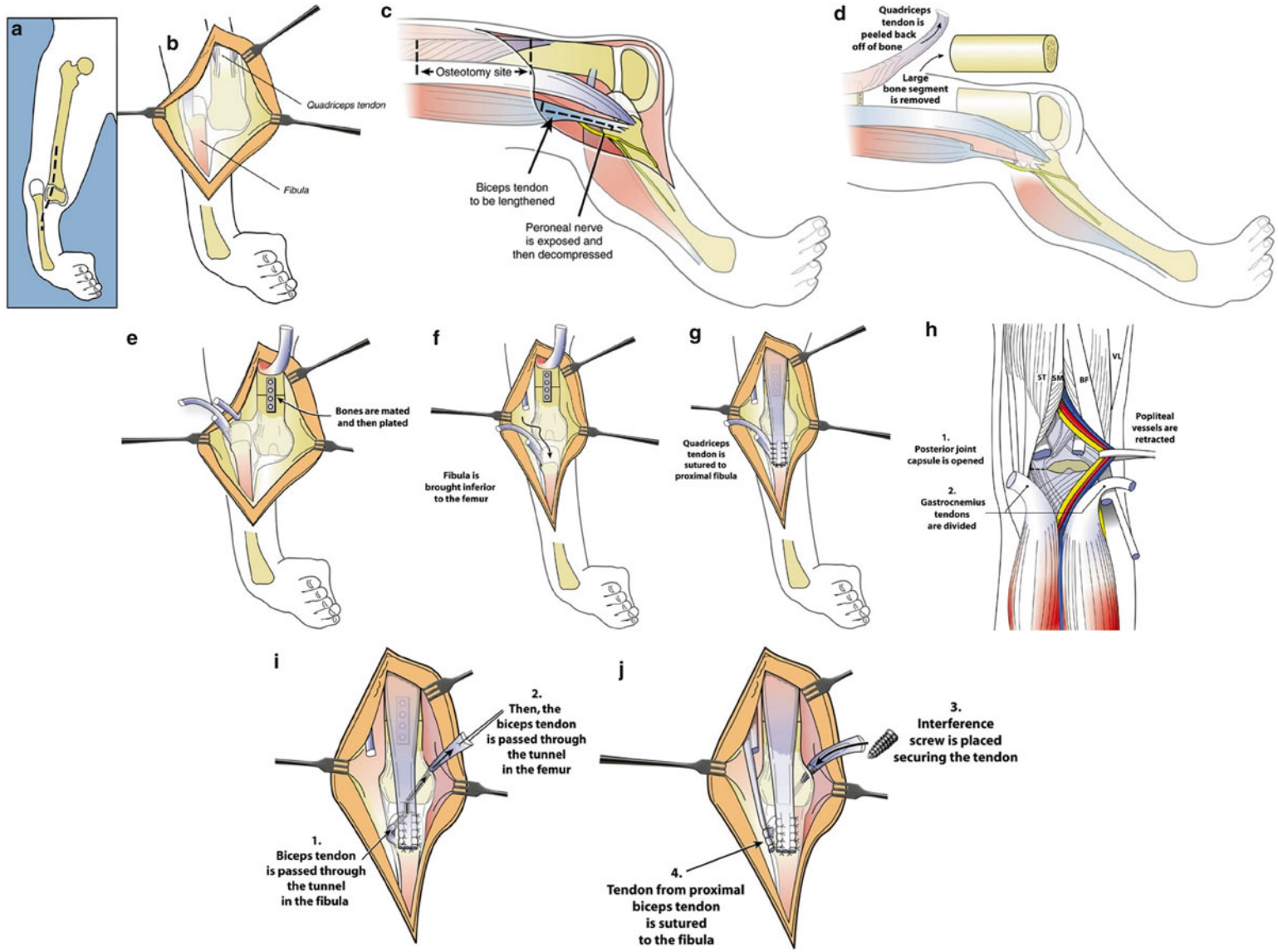


Fig. 24.16 (**a**, **b**) Incision for acute reduction of Paley type 5b (ii). (**c**, **d**) Biceps tendon lengthening, peroneal nerve decompression, and femoral shortening to treat the knee flexion contracture. (**e**) Fixation of the distal femur. (**f**) Reduction of fibula to femur. (**g**) Advancement of the remnant of quadriceps to the fibula. (**h**) To fully correct the knee flexion contracture, the vessels are identified and the posterior capsule is incised. All of the hamstrings and gastrocnemius tendons are released. (**i**) The biceps tendon is used to create a cruciate ligament to tether the fibula to the femur. (**j**) The biceps is advanced and the new cruciate ligament is secured

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Congenital Pseudarthrosis of the Tibia

25

Corinna C. Franklin and Richard S. Davidson

Anatomy and Pathology

Congenital pseudarthrosis of the tibia (CPT) describes a spectrum ranging from anterolateral bowing of the tibia to frank pseudarthrosis and disappearance of bone. It is a rare condition, with an estimated incidence of 1/150,000–250,000 births [1–3]. CPT is typically unilateral and located at the junction of the middle and distal thirds of the tibia; it occurs with equal frequency in boys and girls [1]. Patients may present initially with a fracture or simply with a bowed tibia; the fibula is affected in about one-third of patients [1, 22] (Fig. 25.1).

The pathology of CPT involves fibrous hamartoma and pathological periosteum [1]. Pathologic examination of CPT lesions has revealed highly cellular tissue similar to fibromatosis; the gap contained fibrous tissue as well as both fibrocartilage and hyaline cartilage, and was continuous with the periosteum [5]. This hamartomatous tissue is thought to interfere with bone production and callus formation, perhaps by decreasing the blood supply [2].

Associations

There is a strong association between CPT and neurofibromatosis type 1 (NF-1). Also called von Recklinghausen disease, NF-1 is inherited in an autosomal dominant manner and is relatively common, with an incidence of 1/3,000 [3]. The NF-1 gene is located on chromosome 17 and encodes the product neurofibromin [3]. As many as 75 % of CPT cases are associated with NF-1, though only 5 % of NF-1 patients develop CPT [3]. Histologically, there is no difference between cases with or without NF-1 [4]. Non-orthopedic manifestations of NF-1 include café-au-lait spots, optic gliomas, neurofibromas, malignant peripheral nerve sheath tumors, and increased risk of cardiovascular diseases [3]. Other orthopedic manifestations include scoliosis and abnormalities of the cervical spine; patients often have decreased bone mass due to the loss of neurofibromin function [3].

The method by which NF-1 is related to or causative of CPT is imperfectly understood. A recent study by DY Lee et al. suggested that the fibrous hamartoma of CPT in NF-1 results from periosteal cells that have failed in terminal osteoblastic differentiation, arresting without completing the differentiation process [6]. Another study by SM Lee et al. indicated that double inactivation of the *Nf1* gene could be present in some cases of CPT but is not the essential requirement for development of CPT [7].

Pseudarthrosis of the tibia has also been described in cases of osteofibrous dysplasia or fibrous dysplasia. Boyd described several cases of CPT as having pathology similar to fibrous dysplasia in 1958 [8]; Campbell et al. reported on five cases which they termed a variant of fibrous dysplasia involving anterior bowing of the tibia [9]. Campanacci et al. reported on a series of 35 cases of what they termed osteofibrous dysplasia, one of which had a pseudarthrosis of the tibia, and ten of whom had bowing of the tibia [10].

The tibial bowing in CPT is distinctly *anterolateral*. Bowing in other directions occurs in rickets (typically tibia vara or valga) or in posteromedial bowing of the tibia [11] but is unrelated to pseudarthrosis of the tibia and treated very differently.

Classification Systems

Multiple classifications of CPT have been described; these include the systems of Crawford, Boyd, and Andersen.

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Crawford described a classification system in 1986 based on the radiographic appearance of the tibia in neurofibromatosis; in this assessment type 4 has the worst prognosis (Fig. 25.2) [12]:

Type 1: Anterolateral bow with a dense medullary canal.

Type 2: Anterolateral bow with a widened medullary canal and a tubulation defect.

Type 3: Anterolateral bow and a cystic lesion.

Type 4: Anterolateral bow with evident fracture, cysts, or pseudarthrosis.



Fig. 25.1 Photograph of tibia with anterolateral bow

Boyd described six types of CPT in 1982; in this assessment type 2 has the worst prognosis [13]:

Type 1: Anterior bow and defect of the tibia, possibly associated with other congenital deformities.

Type 2: Anterior bow and hourglass constriction of the tibia with fracture typically before 2 years of age; often associated with neurofibromatosis.

Type 3: CPT developing in a bone cyst, usually at the junction of the middle and lower two-thirds of the tibia.

Type 4: CPT originating in the same location without tibial narrowing but with at least partial, if not complete, narrowing of the canal.

Type 5: A dysplastic fibula with fibular or tibial pseudarthrosis.

Type 6: Intraosseous neurofibroma or Schwannoma.

Andersen described a series of 46 patients with pseudarthrosis of the leg in 1976, which he separated into six types [14]:

Club foot: True congenital fracture present at birth, associated with other congenital anomalies.

Cystic: Cystic lesions of the distal tibia and fibula without angulation or fracture at birth.

Late: Pseudarthrosis developed after fracture through a sclerotic tibia (at 4 and 9 years of age).

Fibular: Pseudarthrosis first developed in the fibula.

Dysplastic: Dysplastic, hourglass tibia with pseudarthrosis; associated w/neurofibromatosis.

Angulated: No radiographic evidence of dysplasia; pseudarthrosis developed after osteotomy; associated with neurofibromatosis.

In practice, the most relevant criterion to guide treatment is whether the tibia is fractured or intact.

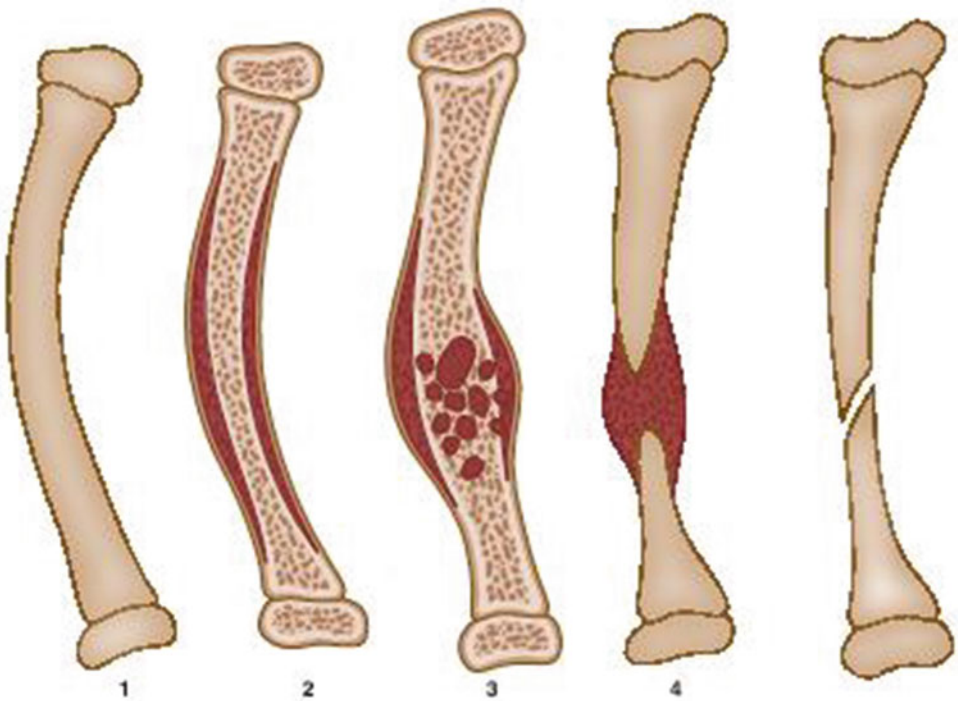


Fig. 25.2 Crawford classification of CPT: Type 1: anterolateral bow with a dense medullary canal; type 2: anterolateral bow with an increased medullary canal and a tubulation defect; type 3: anterolateral bow and a cystic lesion; type 4: anterolateral bow with evident fracture, cysts, or pseudarthrosis

Imaging

The mainstay of imaging for CPT is anterior-posterior and lateral radiographs of the affected leg. Computed tomography (CT scanning) may be used to quantify healing at the CPT site. More recently, magnetic resonance imaging (MRI) has been used to assess CPT; it has been suggested that MRI can provide additional information such as the type, length, and structure of the pseudarthrosis, as well as the extent of the affected periosteum and subtle soft-tissue changes [15]. This may assist in treatment planning and monitoring.

Prognostic Factors

Multiple prognostic factors have been proposed and debated, much of which remains controversial. Association with NF-1 was thought to indicate a poorer prognosis; however, this is no longer thought to be the case, as this has not been borne out in the literature [1]. The relationship between age and prognosis is complex. There has been debate over when to initiate surgical management, with some authors advocating early intervention and others urging waiting until at least age 4 [4]. Boyd asserted that older children would do better after surgical treatment [13]. However, Morrissy et al., reporting on a series of 40 cases of CPT, felt that older children were more likely to have a poor surgical outcome; they found that patients who still required grafting at or after age 13 years did poorly [16].

Tudisco et al. examined 30 patients with CPT who had reached the end of skeletal growth, and found that radiographic classification was correlated with prognosis. Crawford type 2 and 4 had the worst prognosis for union, with type 4 having the worst functional results [17]. Ohnishi et al., reporting on a series of 73 patients with CPT, felt that residual deformity and narrow diameter of the bone at the CPT site would lead to refracture [18].

Finally, a more distal location in the tibia may worsen the prognosis of CPT as this location is more difficult to control and necessitates the involvement of the ankle and the hindfoot in fixation [1].

Non-operative Management

In general, the treatment of CPT is surgical. Early on, patients with anterolateral bowing may be braced in order to attempt to delay progression to fracture and CPT [1]. At first, when a child is younger than 3 or 4 years old, knee-ankle-foot orthoses (KAFOs) are used in order to control rotation of the lower extremity and prevent progression to fracture . Thereafter, a patella-tendon-bearing clamshell orthosis is used with medial and lateral femoral flanges, which will also control rotation. Bracing is also frequently used postoperatively in order to support the surgical procedure.

Operative Management

CPT is a challenging disorder to treat. The underlying condition makes healing difficult, and multiple procedures and revisions may be needed. Ultimately some cases may eventually be treated with an amputation. Many procedures exist for the treatment of CPT. These include bypass grafting, intramedullary fixation, free fibula transfer, Ilizarov external fixation, periosteal grafting, Masquelet's procedure, and amputation.

Modified McFarland Bypass Graft

Ofluoglu et al. describe a method of prophylactically treating an anterolaterally bowed tibia in NF1 with a posteromedially placed allograft fibula combined with bracing until maturity. In their series of ten, no patient had a fracture or a pseudarthrosis at final follow-up (Figs. 25.3a–c and 25.4a–f) [19].

This technique is performed under tourniquet with the use of an image intensifier. The patient is positioned supine and the lower leg is placed on the operating table such that the tibial deformity is in the horizontal plane. The skin and soft tissues are incised anteromedially from the proximal to the distal physis; the periosteum is then resected, taking care not to incise too close to the physes. Notches are then made in the tibial metaphysis proximally and distally about 2 cm from the physes. An allograft fibular strut, several millimeters longer

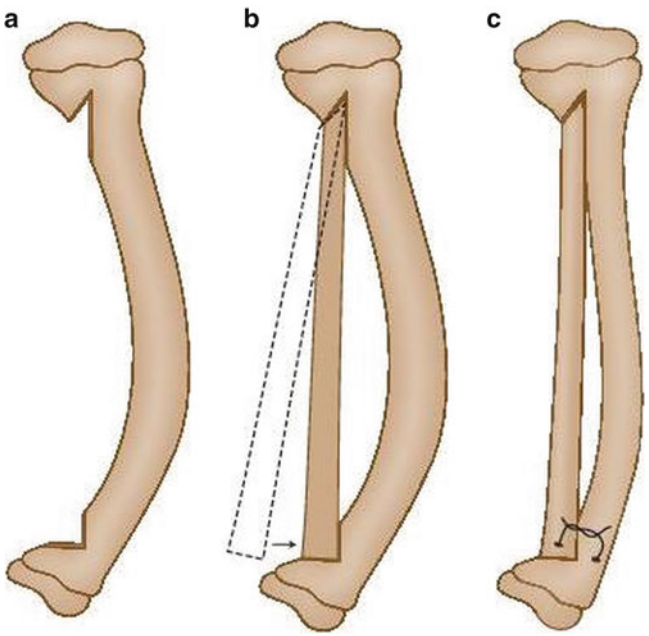


Fig. 25.3 (**a**–**c**) Modified McFarland bypass graft technique

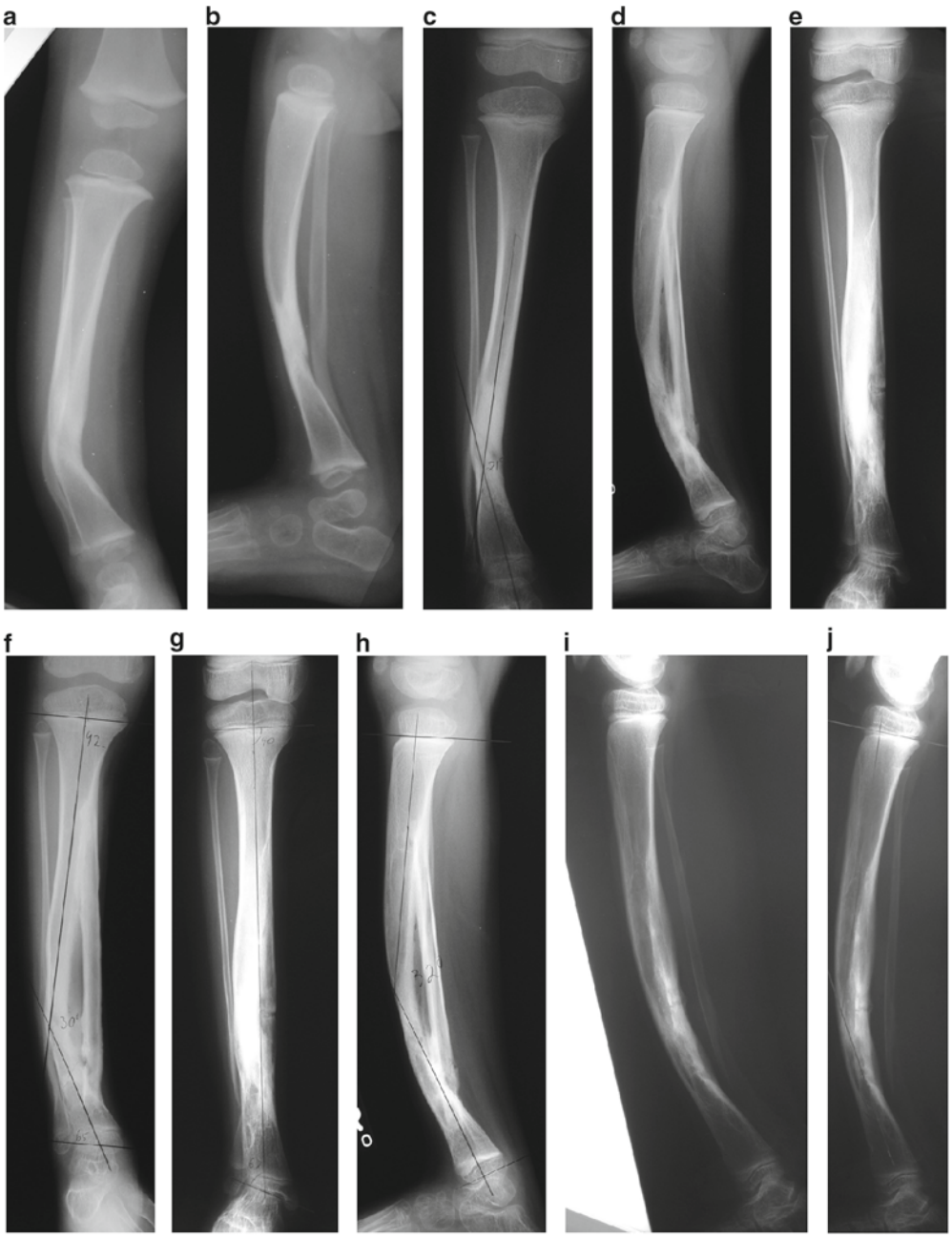


Fig. 25.4 (**a**–**j**) CPT treated by the modified McFarland bypass graft method

than the span of the two notches, is impacted into the two notches under compression, taking care to place the graft on the concavity of the bow of the tibia to accept weight bearing. Thus, this bypasses the bowed tibia. A screw, a wire, or heavy suture is then used to fix the distal end of the graft to the tibia, and the tibia facing the graft is decorticated. Bone graft is then used to facilitate incorporation of the allograft. The patient is then placed in a long leg cast and kept non-weight bearing until radiographic evidence of bone healing is noted (6–8 weeks). Thereafter the patient wears a total contact clamshell patellar tendon-bearing orthosis (flared at the knee to control rotation) or a hinged KAFO until maturity [19]. Remodeling often leads to resorption of the tibia in favor of enlargement and incorporation of the fibular graft. Proximal and distal metaphyseal deformity may need guided growth plates and screws if remodeling is incomplete.

Intramedullary Nailing

In 1956, Charnley described the use of an intramedullary nail to treat CPT; two patients were treated with Steinmann nails in the intramedullary canal [20]. Sofield, in 1971, reported on a series of more than 100 patients with CPT, and



Fig. 25.5 X-rays of CPT treated with an intramedullary nail

asserted that those treated with intramedullary fixation did best; he advocated fragmentation, reversal of the fragments, and fixation with an intramedullary rod (Fig. 25.5) [21].

Dobbs et al. describe a treatment method of CPT using a Peter Williams rod for stabilization after excision of the pseudarthrosis. They report a satisfactory outcome in 16 of 21 patients; complications included reoperation in 5, amputation in 5, ankle valgus in 10, and leg-length discrepancy in 11 [22].

This technique is performed under tourniquet with an image intensifier. Iliac crest bone graft is first acquired. An anterior incision is made longitudinally extending proximally and distally from the pseudarthrosis site. The pseudarthrosis is excised down to normal, bleeding medullary canal, which is then reamed both proximally and distally. A Williams rod is used to stabilize the tibia (implants such as the Fassier-Duval rod may also be considered, although there is concern for threaded screws in the growth plate). The size (length and diameter) of the rod is determined preoperatively, based on anticipated growth. In most cases, the rod traverses the ankle. In children younger than 4, it will go into the calcaneus; in patients who are 5–10 years old, the rod will end in the talus; in those older than 9 or 10 (or if the pseudarthrosis is quite proximal), the rod will not cross the ankle. The assembled rod is passed antegrade into the distal tibial segment, and then out through the talus and calcaneus, finally passing out through the heel pad, taking care to position the foot and ankle correctly in neutral. Then, the rod is passed back up retrograde into the proximal tibial segment, again taking care to align the tibia properly (this may require a second, proximal osteotomy). If the fibula is too long, it should be cut and stabilized with a Kirschner wire. The bone graft is placed around the pseudarthrosis site and secured with heavy suture. Patients are cast for about 4 months—spica initially in young children, and then above knee. Thereafter a clamshell KAFO is used with a free knee and locked ankle. In older children, this may eventually be converted to a total contact ankle-foot orthosis (AFO). Once the distal edge of the rod approaches the talar articular surface, via longitudinal growth of the tibia, a second procedure is done to advance the rod across the ankle. The orthosis is then converted to a hinged ankle and is worn until maturity [22].

Free Vascularized Fibula

A free vascularized fibular (FVF) graft from the contralateral leg may be used as a strut after excision of a tibial pseudarthrosis. This functions as a structural replacement for the excised portion of the tibia. In the ideal case, the vascularity of the graft allows it to heal more quickly, avoid resorption, and add vasculature to a scarred and dysplastic site [23]. FVF transfer requires a specialized team with microsurgery expertise, and careful attention to detail.

Several authors have reported on the results of FVF transfer in the treatment of CPT. Dormans et al. reported on a series of 12 patients in which FVF was used to treat CPT; at final follow-up, one patient had a persistent nonunion [24]. Similarly, Weiland et al. reported on a series of 19 patients with CPT treated with FVF; of these, 18 had healed at final follow-up [25]. Iamaguchi et al. present a cautionary view of this procedure; in their series of 16, 63 % required multiple surgeries to achieve union, but eventually all 16 did [26].

Ilizarov External Fixation

The Ilizarov circular external fixator may be used in CPT. Several different techniques and frame configurations have been described. Paley et al. describe a series of 15 patients with 16 CPT who were treated using the Ilizarov device. The rate of union was 94 % after one treatment and 100 % after two; there were five refractures and two residual deformities [27]. Boero et al. describe a series of 21 patients with CPT treated with the Ilizarov device; at final follow-up, 9 had consolidated without deformity or shortening, 5 consolidated with axial deviations, and 7 did not consolidate [28]. Ghanem et al. reviewed a series of 14 patients with CPT treated using the Ilizarov fixator; in all instances the pseudarthrosis site was not excised but the leg was realigned and lengthened. Union was ultimately achieved in 13 cases, 6 having required additional grafting [29] (Fig. 25.6a, b).

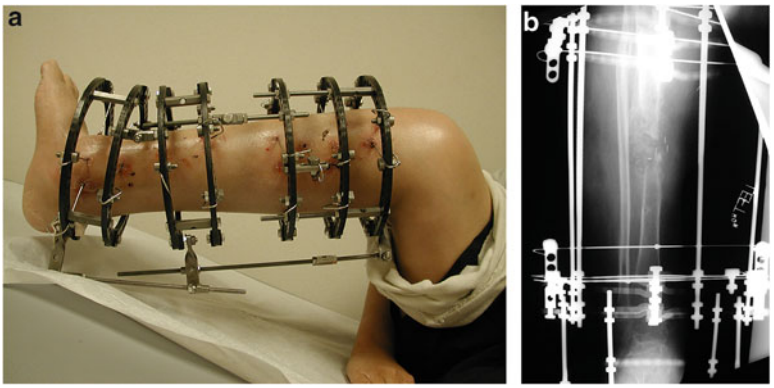


Fig. 25.6 (**a**, **b**) Clinic photo and X-ray demonstrating CPT treated with the Ilizarov device

The pseudarthrosis site may be treated by direct end-to-end compression, by side-to-side compression, or by inserting one part into another; it may also be resected, with lengthening through a proximal corticotomy, through the site itself, or by bone transport [27]. Typically, a four-ring assembly is used, with two rings proximal and two distal to the pseudarthrosis site; the foot may be included if the pseudarthrosis is very distal in the tibia [28].

The versatility of the external fixator may also allow associated deformities in the ipsilateral lower extremity to be treated. Lengthening as well as correction of angular deformities may be performed [27, 28].

Periosteal Grafting

Thabet et al. report on the results of a technique that is a combination of several methods, using intramedullary fixation and Ilizarov circular fixation along with bone and periosteal grafting. Their case series includes 20 patients, all of whom united after the first procedure; 8 patients sustained refractures, reoperation was required in 7 cases, and no amputations were performed [30].

This technique is performed under tourniquet and with an image intensifier. An anterior incision is used; the cuff of abnormal periosteum is incised, circumferentially dissected, and removed. A small longitudinal split is made in the proximal tibia segment, and the distal end is drilled open and then inserted into the proximal end. The same procedure, including periosteal excision, is performed on the fibula. Intramedullary fixation is then placed in both the tibia and fibula (static or expandable rods were both used in the referenced paper). Next, iliac crest bone graft is harvested, as well as the periosteum over the iliacus muscle. These are placed around the pseudarthrosis site, periosteum first. Finally, an Ilizarov fixator is applied, with three wires and a ring proximally and the same distally; the frame is extended to the foot [30] (Figs. 25.7a–f and 25.8a–h).

Masquelet's Procedure

Masquelet has described a procedure, called induced membrane, for reconstruction of long bone defects which he has applied to CPT [31]. This is performed in two stages. Initially, a broad debridement is done, followed by the insertion of a cement spacer (made out of polymethyl methacrylate). In the second stage, performed 6–8 weeks later, the spacer is extracted but the membrane surrounding it is not removed; this is filled with iliac crest bone graft (and supplemented with allograft if needed). An external fixator is used, and the fibula is often incorporated [31]. Further investigation is required on this method as the available results in the literature are limited (Fig. 25.9a–d).

Amputation

Amputation is usually considered an option of last resort. However, some difficult cases of CPT end up requiring many complex surgeries, with long periods in which the patient is unable to fully bear weight, play sports, or engage in many activities shared with their peers. In these cases, amputation may become the most attractive option, as it would mean an end to the frequent procedures and, with current advanced prostheses, allow most sports and activities to be possible. Furthermore, many patients with neurofibromatosis will have other conditions requiring treatment, such as malignancies or spinal deformities, and may wish to curtail the multiple surgeries involved in treating a recalcitrant CPT in order to keep some years of childhood free of hospital visits if possible. In younger children, an ankle disarticulation is appropriate (as the malleoli may be expected to remodel), whereas a Syme's amputation is indicated in older patients. Amputation through the pseudarthrosis itself is indicated if infection is present (Fig. 25.10a, b).

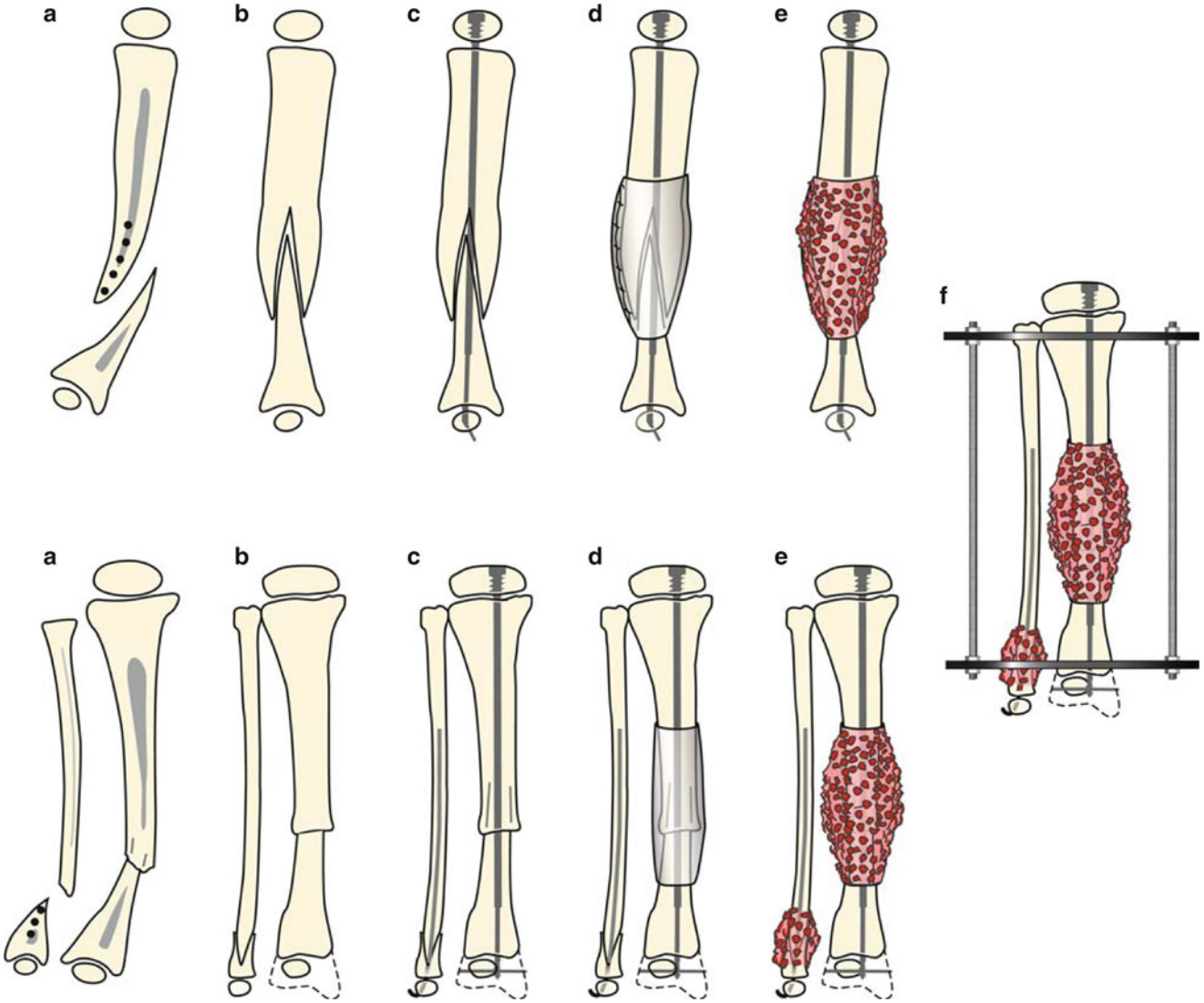


Fig. 25.7 Treatment for type I CPT in the lateral and anteroposterior (AP) views is shown. (**a**) There is longitudinal splitting of the proximal tibial and fibular fragments. (**b**) The bone ends are docked and (**c**) IM rods are inserted (Fassier-Duval telescopic IM nail with Paley modification illustrated) into the fibula and tibia. (**d**) Periosteal graft is wrapped around the pseudarthrosis and (**e**) iliac crest bone graft is applied to the tibial and fibular docking site. (**f**) External fixator is applied. With kind permission from Springer Science+Business Media: Clinical Orthopedics and Related Research, Periosteal grafting for congenital pseudarthrosis of the tibia: A preliminary report, 466(12), 2008, Thabet AM

Medical Management

Several medical adjuncts to surgical treatment have been described in the literature. These include the use of bone morphogenetic proteins (BMPs), bisphosphonates, bone stimulation, and bone marrow grafts. In all cases, further research is needed before firm conclusions can be drawn or recommendations made.

Bone Morphogenetic Protein

BMPs are biologic substances, several of which stimulate bone formation. BMP-2 and BMP-7 are commercially available in the USA, and have been used in procedures such as spinal fusion and repair of open or non-united tibial fractures in order to promote or accelerate fusion [32, 33]. These proteins may be a useful adjunct to CPT procedures; currently this is an off-label use.

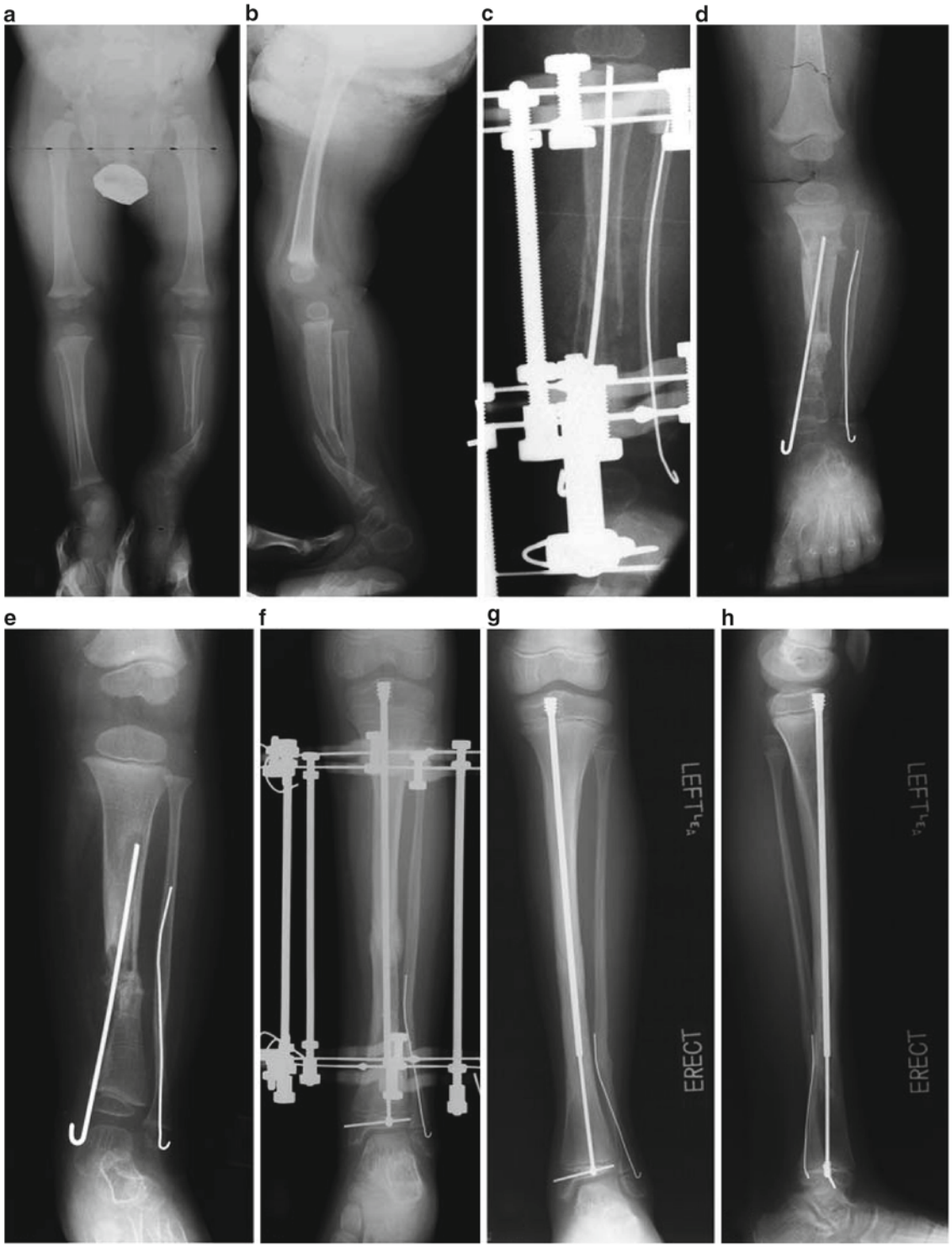


Fig. 25.8 (**a**) Anteroposterior (AP) and (**b**) lateral view radiographs of a 2-year-old boy with neurofibromatosis and CP that had not been previously treated are shown. (**c**) AP view radiograph shows split of the proximal segment with invagination of the distal segment, rodding of tibia from medial malleolus and of fibula through lateral malleolus, and application of Ilizarov external fixator. (**d**) AP view radiograph obtained immediately after external fixator removed shows union of the CPT. (**e**) AP view radiograph shows refracture of the tibia 1 year later. (**f**) AP view radiograph shows retreatment with an Ilizarov device and rerodding with Fassier-Duval telescopic IM nail locked in both proximal and distal epiphysis. (**g**, **h**) Tibia remains healed 2 years later with good remodeling. The telescopic nail has extended with growth. With kind permission from Springer Science+Business Media: Clinical Orthopedics and Related Research, Periosteal grafting for congenital pseudarthrosis of the tibia: A preliminary report, 466(12), 2008, Thabet AM

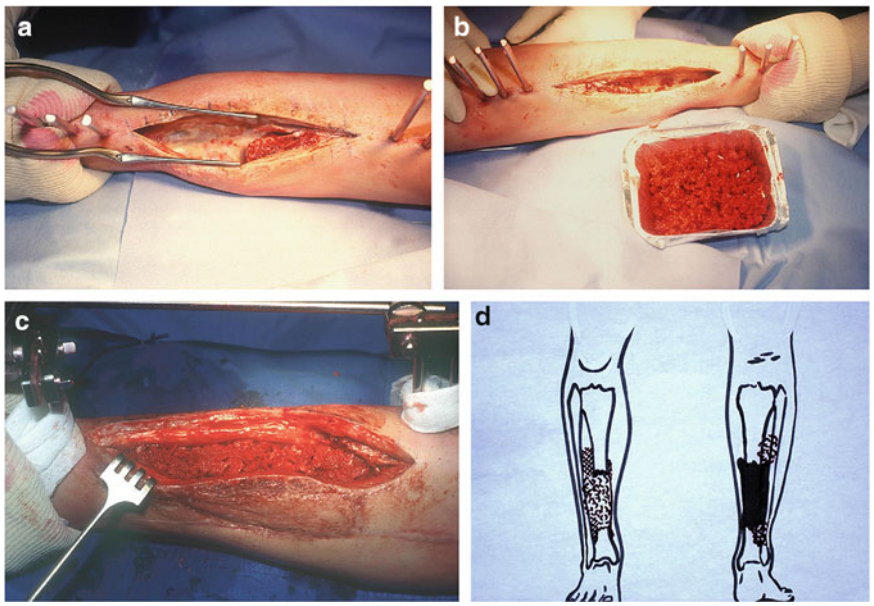


Fig. 25.9 Masquelet's induced membrane technique principles. Elements of technique. (**a**) The preoperative aspect of the membrane just before filling up the cavity. Note the petaling of the bone extremity. (**b**) Morcellized cancellous bone graft; the chips should be as small as possible. The alveolar structure of the cancellous bone permits a fast revascularization by the vascular buds issued from the membrane. (**c**) The cavity is filled up; the suture en bloc of the membrane and subcutaneous tissue results in a containment system. (**d**) Principles of reconstruction of the tibia. At the first stage the PMMA spacer is applied on the fibula. At the time of reconstruction an intertibiofibular graft is performed on both extremities. Reprinted from Orthopedic Clinics of North America, 41 (1), Masquelet AC, Begue T, The concept of induced membrane for reconstruction of long bone defects, 27–37, Copyright 2010, with permission from Elsevier

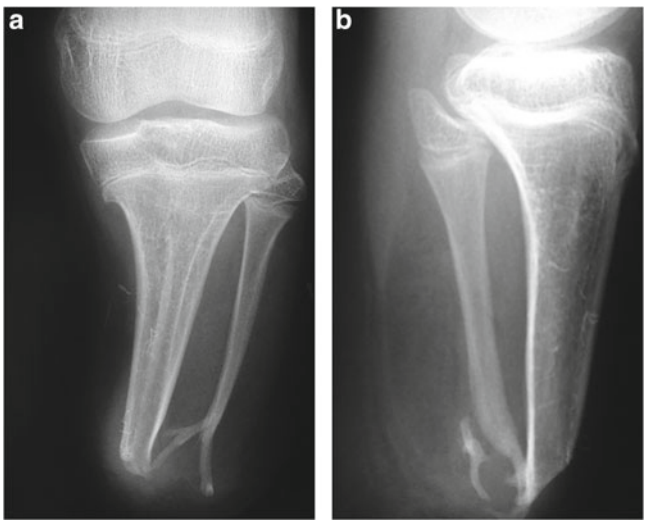


Fig. 25.10 (**a**, **b**) X-rays of a patient who eventually underwent a below-knee amputation following multiple attempts at reconstruction as well as history of three primary malignancies and a large leg-length discrepancy that would have required several more surgeries

Richards et al. reported on a series of seven patients with CPT in whom BMP-2 was used as an adjunct; in all cases a Williams rod was used for fixation [34]. Five patients went on to radiographic union, one of whom later had a fracture that was retreated in the same manner and went on to heal. A further patient required revision but went on to union. One patient had a persistent nonunion and ultimately underwent an amputation. No complications related to BMP were reported. The authors concluded that the use of BMP-2 in this setting improved outcomes [34].

In contradistinction, Lee et al. reported on a series of five patients in which BMP-7 was used as an adjunct to surgery but concluded that this did not promote improved bone healing in these patients [35].

Bisphosphonates

Bisphosphonates are analogues of pyrophosphate that interfere with osteoclast activity, and thus slow bone resorption; they have been used in several diseases in which bone metabolism is disturbed or bone turnover is excessive, such as fibrous dysplasia [36]. Birke et al. proposed that bisphosphonates would be a useful adjunct in treating CPT, particularly as NF-1 involves deficient bone metabolism [37]. They reported on a series of eight cases in seven patients (six of whom had NF-1) in whom BMP-7 and either pamidronate or zoledronic acid were systemically administered as adjuncts to CPT surgery. Primary healing was achieved in six of the eight cases, with the remaining two cases eventually going on to heal after multiple procedures and subsequent refractures.

The authors concluded that bisphosphonate therapy was a useful addition to CPT surgery [37]. It should be noted that there exist limited data on the long-term effects of bisphosphonate use in children or women of child-bearing age.

Electrical Stimulation

In the past, several studies looked at using electrical current to augment surgical treatment of CPT [38, 39]. This then fell out of favor for some time. More recently, however, Cebrian et al. published a study examining the use of pulsed electromagnetic fields (PEMFs) in adult cases of CPT treated with intramedullary nailing. They found improved healing in those cases in which the PEMFs were used compared to those in which the PEMFs were not used, and suggested that this is a useful addition to CPT surgery [40].

Bone Marrow Grafting

In 1995, Garg and Gaur published a report of one patient with CPT in whom a percutaneous injection of harvested bone marrow cells from the iliac crest was used; this patient healed with casting (after prior excision of the pseudarthrosis) [41]. A similar case was reported by Kitoh et al., in which culture-expanded osteoblast-like cells as well as autologous platelet-rich plasma were injected into the distraction site of a CPT being treated with distraction osteogenesis, with a good result [42]. A study of the biological basis for the use of bone marrow cells in the treatment of CPT noted that this was a promising strategy meriting further study [45].

Combined Approach

Recently, Dror Paley has described a combined approach for the treatment of CPT for which he has reported excellent results, though longer follow-up and confirmation of this approach's feasibility in the hands of other surgeons are required [44]. This method combines pharmacologic and surgical management in order to maximize the chances of a successful outcome.

Either 1 week prior to or 1 week following surgery, the patient is given zoledronic acid intravenously (0.2 mg/kg over 30 min), followed 1 hour later by 60 mg/kg of calcium gluconate intravenously over 60 minutes. The patient also is to take 2 g elemental calcium for 7 days and 400 IU of vitamin D for 14 days.

The surgical procedure is performed under tourniquet; the approach is via an anterior longitudinal incision. The diseased periosteum is incised longitudinally and excised circumferentially in its entirety. Posterolaterally to the tibia, the fibular hamartomatous periosteum is also excised. A split is created in the proximal tibia, and the distal tibia canal is opened with a drill. The distal portion is inserted into the proximal split. Similarly, the distal fibula is split and the proximal portion inserted into it. If there has been prior surgery, all dead bone should be resected (the tourniquet may be let down in order to observe which portions of the bone are bleeding and healthy). An intramedullary rod is then inserted into the tibia across the site of the CPT, followed by another rod in the fibula. The ankle is not crossed.

Iliac crest bone graft is then harvested, as well as periosteum from the underside of the iliacus muscle. The periosteum is meshed using a skin graft mesher and then placed around the invaginated CPT site, with the cambium layer facing the bone; this is also done on the fibular site. The harvested cancellous graft is then placed around the CPT site and in the space between the tibia and fibula. BMP-2 is placed as well, including into the space between the tibia and fibula, in order to promote cross-union.

Finally, an Ilizarov fixator is placed, with three proximal wires, three distal wires, and fixation of the foot, followed by a walking ring. After union (about 12 weeks after surgery), the fixator is removed and a long leg walking cast is placed. This is then followed by a KAFO with a free knee and solid ankle. In patients older than 6 years, the ankle is articulated; as the patient grows, the brace is modified, progressing to a total contact articulated AFO or patellar-tendon-bearing brace only. Once the patient is older than 10, a gator brace (with medial/lateral malleolar flanges) is used. The brace is worn at all times (other than bathing and therapy) until skeletal maturity.

The intramedullary rod is changed as needed as the patient grows, with zoledronic acid infusions accompanying rod exchange surgeries. A telescoping rod may be useful in this setting. Hemi-epiphysiodesis may also be performed as needed for valgus at the ankle or knee [44].

Summary

Congenital pseudarthrosis of the tibia is a rare condition that encompasses a wide spectrum from anterolateral bowing of the tibia to true pseudarthrosis with bone loss. The periosteum in CPT is pathologic, with the presence of fibrous hamartoma. There is a strong association between CPT and neurofibromatosis-1 (NF-1); it may also be associated with fibrous dysplasia or osteofibrous dysplasia. Multiple classification systems exist; the key criterion in guiding treatment is whether the tibia is fractured or intact. Treatment is primarily surgical, though medical augmentation with BMPs and/or bisphosphonates may also be useful. A wide variety of surgical procedures exists, including bypass grafting, intramedullary fixation, free fibula transfer, Ilizarov external fixation, periosteal grafting, and Masquelet's procedure; some cases may ultimately end in amputation.

Treatment of CPT has continued to evolve over the past 25 years in a mostly additive fashion based on the principles of removal of pathologic tissues, replacement with viable tissue, stabilization, and, most recently, addition of medical stimulation. While this combined method has produced the best chance of healing and avoidance of refracture, additional study will be needed to distill this extensive complex series of treatment measures to ascertain the minimal number of steps to optimize treatment.

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Congenital Posteromedial Bowing of the Tibia

26

Benjamin Joseph, Hitesh Shah, and N.D. Siddesh

Introduction

Congenital posteromedial bowing of the tibia is a relatively uncommon condition. It is one among a group of congenital bowing deformities of the tibia that include anteromedial, anterolateral, and posteromedial bowing. In the past these three deformities were arbitrarily grouped together and referred to as congenital kyphoscoliotic tibia [1]. It is well recognized that the clinical features, natural history, and long-term prognosis of posteromedial bowing of the tibia are distinctly different from the more sinister anterolateral bowing associated with neurofibromatosis and congenital pseudarthrosis and anteromedial bowing associated with fibular deficiency [2–5]. Though a diagnosis may be made in the antenatal period during routine ultrasound evaluation of pregnancy [6, 7] in the majority of instances the diagnosis is made at birth.

Incidence

The exact incidence of congenital posteromedial bowing of the tibia is unknown though hospital-based reports suggest that it is an uncommon condition. Pappas [5] reported a series of 33 patients gathered over a 36-year period; Hoffman and Wenger [8] reported 13 patients seen over a 14-year period; Johari et al. saw 31 children in an 18-year period [9] and we reported 20 patients seen over a 19-year period [10]. These four reports, all based on retrospective data from tertiary care centers, suggest that most of such centers would probably encounter one or two new cases of congenital posteromedial bowing per year. These hospital-based estimates, however, are prone to several biases and hence are, at best, rough indicators of the magnitude of the problem. In 2008 we put in place a registry for congenital posteromedial bowing of the tibia and have been prospectively following up all new cases. In these last 5 years we documented 34 new cases. This suggests that the condition is much more common than previously estimated from retrospective studies. The greater frequency of cases noted over the last 5 years is likely to be a reflection of more reliable nature of data generated prospectively rather than an increase in the incidence.

Demographic Features

Bilateral involvement is rare; we encountered one bilateral case among 54 cases. There is an unexplained predilection for the left limb to be affected; left-sided involvement is almost twice as frequent as the right [10]. Boys and girls appear to be equally affected.

Etiology

Dawson attributed the deformity to malunion of an intrauterine fracture though he provided no real evidence to support this hypothesis [11]. De Maio et al. [7] had the unique opportunity of having performed an autopsy on a fetus with posteromedial bowing aborted at 24 weeks of gestation. They noted evidence of amniotic rupture and attributed this as the possible underlying cause of the deformity in that fetus.

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Deformities

The Tibia

The tibia is bowed posteriorly and medially (Fig. 26.1a–d); in the newborn the posterior bow is usually more pronounced than the medial bow though the medial bow may exceed the posterior bow in some instances [10]. The degree of posterior angulation in the newborn may be as high as 90° (see Fig. 26.1b). The site of the deformity is characteristically at the junction of the middle and lower thirds of the leg. A skin dimple may be seen at the apex of the bow. The tibia is invariably shorter than the unaffected side and the degree of shortening is proportionate to the severity of bowing (Fig. 26.2a, b).

Though the degree of shortening frequently is in the order of 15 or 20 % up to 40 % of shortening has been recorded [10]. Occasionally there may be an associated torsional deformity of the tibia (see Fig. 26.2b).

The Fibula

The fibula is also bowed and the degree of deformity parallels the tibial deformity. The distal fibula physis is often at a higher level than normal suggesting that there is a relative growth retardation of the fibula as compared to the affected tibia (Fig. 26.3).

The Ankle and Foot

The tibial deformity is invariably associated with a calcaneo-valgus deformity which may be very severe at birth with the dorsum of the foot lying almost in contact with the shin (Fig. 26.4). The distal tibial epiphysis may appear late and when it does appear it may be eccentrically positioned towards the medial side (Fig. 26.5a) [10]. The epiphysis may then develop asymmetrically into a wedge-shaped epiphysis (see Fig. 26.5b) resulting in a valgus tilt of the ankle mortise [10]. The growth delay of the fibula with a high lateral malleolus referred to above also contributes to ankle valgus.

Box 26.1. The Deformity

- Posteromedial bowing at the junction of the middle and lower thirds of the tibia and fibula.
- Posterior bow is usually more severe than medial bow.
- Shortening is proportionate to severity of bowing.
- Always associated with a calcaneo-valgus deformity of the foot.
- Valgus deformity of the ankle may develop later due to growth abnormalities of the distal tibia and fibula.

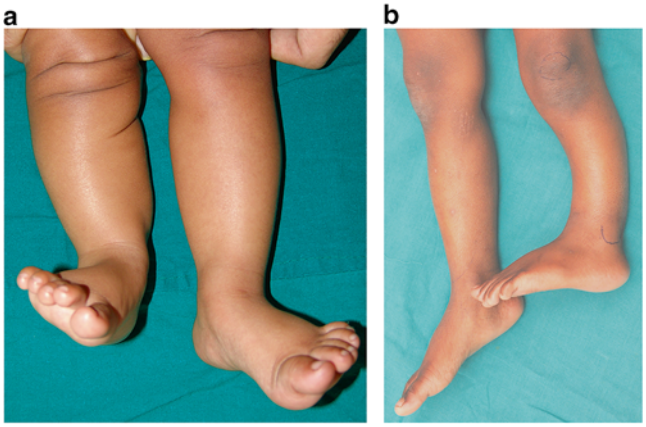


Fig. 26.2 Examples of a short tibia associated with congenital posteromedial bowing. The shortening is mild when the bowing is mild (**a**) and greater when the bowing is more severe. Occasionally, a torsional deformity may also be present (**b**). The patella and lateral malleolus have been marked to demonstrate the torsional deformity

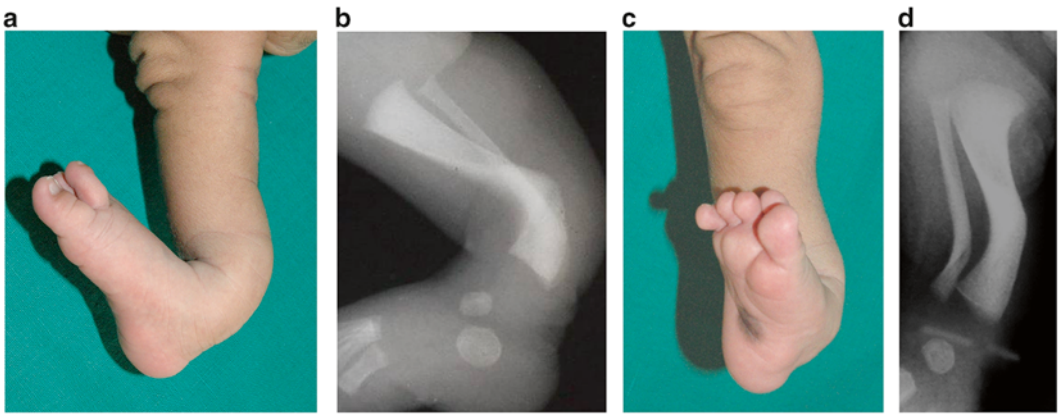


Fig. 26.1 Congenital posteromedial bowing of the tibia in a newborn. The clinical and radiographic appearances of the posterior bow (**a**, **b**) and the medial bow (**c**, **d**) are clearly seen. A delay in the ossification of the proximal tibial epiphysis and the cuboid is evident



Fig. 26.3 The fibular physis is at a higher level than normal (at the level of the ankle joint)



Fig. 26.4 Calcaneo-valgus deformity seen in association with congenital posteromedial bowing of the tibia

The Natural History

Rate and Pattern of Spontaneous Remodeling

The posteromedial bowing deformities of the tibia and fibula tend to improve spontaneously over time. The resolution of the bony deformity occurs by two distinct mechanisms; one entails resorption of bone from the convex surface and new bone deposition on the concavity of the bow in accordance with Wolff's law [10]. De Maio et al. demonstrated that this pattern of remodeling begins in utero [7]. At 24 weeks of gestation an abundance of osteoclasts were present at the apex of the deformity while an osteoblastic response was seen in the concavity. As this process of remodeling progresses the angular deformity decreases and the angulation between the proximal and distal segments of the bone diminishes. The second mechanism of spontaneous correction of the deformity involves realignment of the proximal and distal growth plates. Physeal reorientation occurs at a faster rate than remodeling at the site of the deformity [10]. Remodeling of the deformity by both these mechanisms occurs at a rapid rate during the first year of life and thereafter the rate of remodeling decreases quite substantially dropping to half the initial rate (Fig. 26.6) [10].

Residual Deformities, Shortening, and Functional Problems

Complete resolution of the diaphyseal deformity does not occur in a proportion of children and residual deformity that is clinically discernable may be seen in children older than 4 or 5 years of age (Fig. 26.7a, b) [10].

Shortening of the limb increases with growth [8] and the limb length discrepancy at skeletal maturity may frequently be of a magnitude to warrant treatment. Pappas noted a median shortening of 4.1 cm at skeletal maturity with a range between 3.3 and 6.9 cm [5]. The foot length of the affected limb is often less than the normal foot [5, 8, 12].

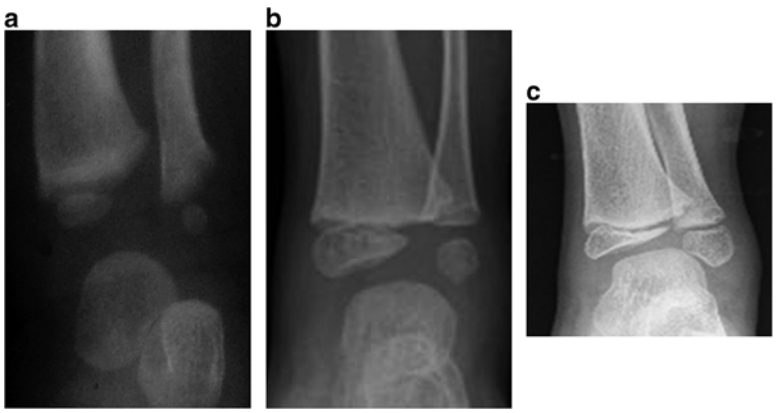


Fig. 26.5 Eccentric ossification of the distal tibial epiphysis in a toddler with congenital posteromedial bowing of the tibia (**a**) and a *wedge-shaped* distal tibial epiphysis in an older child with congenital posteromedial bowing of the tibia (**b**). The valgus tilt of the ankle becomes more evident as the child grows older (**c**)

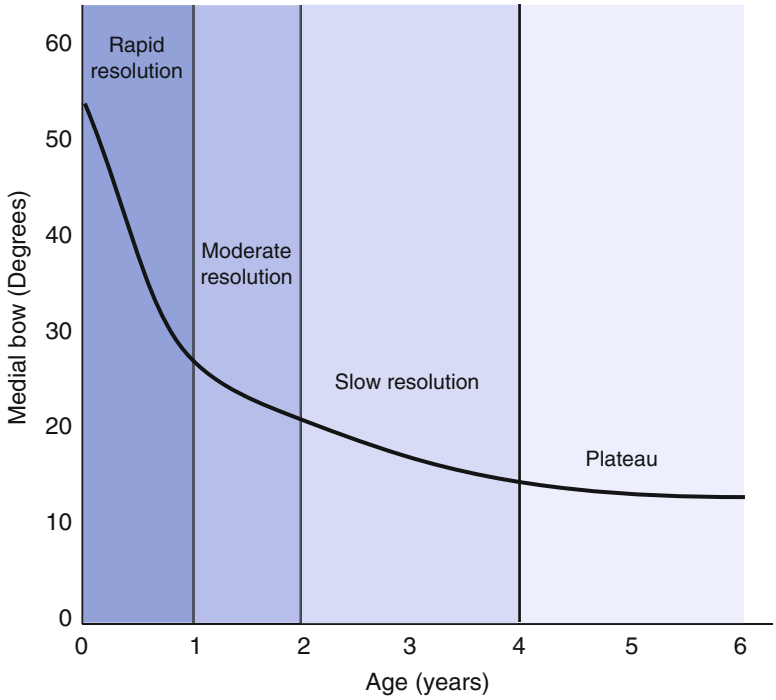


Fig. 26.6 The rate of spontaneous resolution of bowing is rapid in the first year of life, slows down thereafter, and reaches a plateau by 4–5 years of age

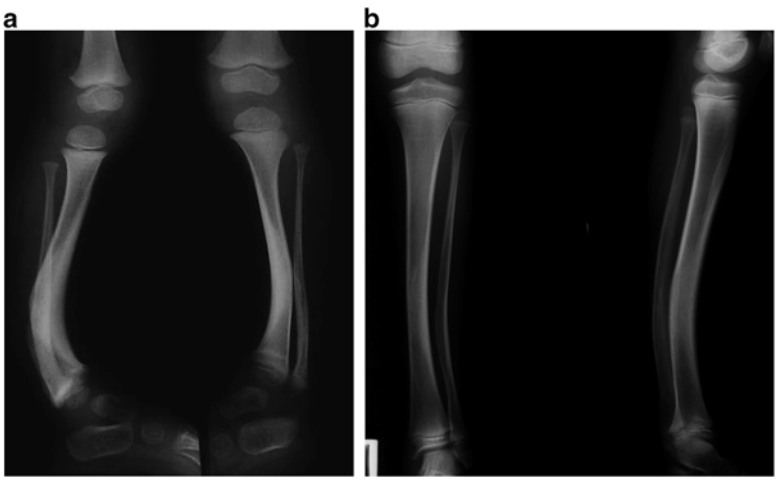


Fig. 26.7 Residual posterior bowing of both tibiae in a child aged 4 years with bilateral posteromedial bowing of the tibia (**a**) and residual bowing at 8 years in another child (**b**). The AP radiograph shows that the distal tibial articular surface is not parallel to the plane of the knee joint even though the medial bow has remodeled to a very large extent

Limitation of ankle motion may persist; limitation of both plantar flexion and dorsiflexion has been reported [8, 10].

Weakness of plantar flexion (Fig. 26.8) has been observed in some children even though the bony deformity may have remodeled almost completely [10]. On occasion the weakness of the gastrocsoleus may be severe enough to result in a calcaneal hitch and a visible gait abnormality requiring surgical intervention [10]; we have encountered two such children.

Box 26.2. Spontaneous Resolution of Bowing

- Occurs by local remodeling and by physeal realignment.
- Occurs rapidly in the first year of life.
- Little remodeling occurs after 4–5 years of age.
- Complete resolution may not occur.

Evaluation

Establishing the diagnosis is not difficult once the plane of the deformity is clear. The evaluation should include a careful documentation of the clinical estimate of the severity of the diaphyseal deformity in the coronal and sagittal planes. The foot should be passively plantar flexed and inverted to assess the rigidity of the calcaneo-valgus deformity. Anteroposterior and lateral full-length plain radiographs of both tibias and a scanogram should be taken to facilitate accurate estimation of the angulation and shortening. The degree of shortening of the tibia should be documented and expressed both as a percent of the normal tibial length and in centimeters. The extent of bowing should be estimated by measuring the diaphyseal angle and the inter-physeal angles



Fig. 26.8 The power of plantar flexion is being tested in a boy with congenital posteromedial bowing of the tibia. Weakness of plantar flexion on the effected side is clearly evident. This weakness that was present before he underwent corrective osteotomy for residual deformity still persists

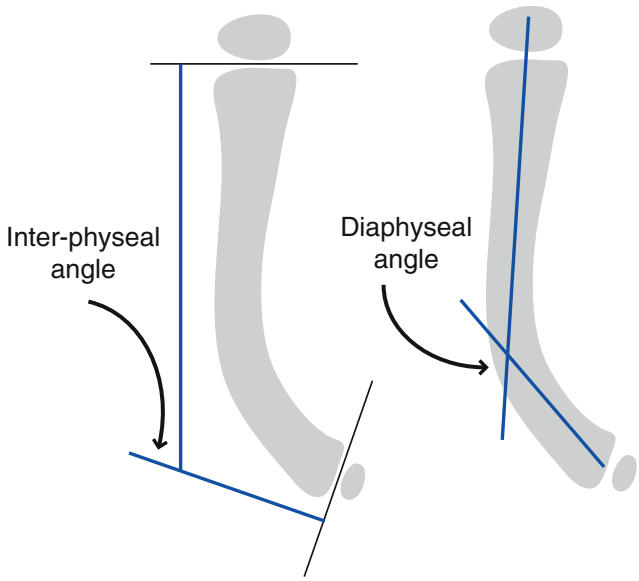


Fig. 26.9 The diaphyseal bow and the inter-physeal angles can be measured on the anteroposterior and lateral radiographs

on the radiographs (Fig. 26.9). The parents should be counselled and reassured that the deformity will reduce spontaneously but they need to be warned that complete resolution may not occur. They should also be instructed in performing passive stretching exercises of the foot into plantar flexion and inversion and encouraged to do it at least twice a day.

The child should be reexamined at 6 months of age to evaluate the extent of correction of the calcaneo-valgus deformity and the tibial bowing. If sufficient resolution of the foot deformity has not occurred casting should be considered. The child should be seen again at 1 year of age to plan the extent of shoe raise that may be required to facilitate more normal walking. The child should then be followed up annually for the next 4 years at which time a decision should be taken regarding intervention for residual deformities and shortening.

The child should be reviewed finally at skeletal maturity to document if limb lengths have been equalized and if there are any residual deformities or functional limitations.

Treatment

Indications

1. Treatment is indicated for any deformity of the leg, ankle, or foot that is cosmetically unacceptable provided adequate time has been given for spontaneous resolution of the deformities.
2. Treatment should be recommended if by the age of 6 years sufficient resolution of the deformity has not occurred to make the articular surface of the distal tibia parallel or almost parallel (<5°) to the knee joint on the anteroposterior radiograph of the tibia.
3. Treatment should be recommended for any functional deficit that may affect normal gait.

Aims of Treatment

- Correct the calcaneo-valgus deformity of the foot in the infant.
- Correct residual coronal, sagittal, and transverse plane (torsional) deformities of the tibia and fibula in the older child.
- Restore the normal alignment of the ankle to restore a horizontal tibial plafond.
- Equalize limb lengths.
- Improve the power of ankle plantar flexion if there is severe weakness of the gastrocsoleus.

Treatment Options

Correction of the Calcaneo-Valgus Foot Deformity

Passive stretching: The mild degree of calcaneo-valgus deformity responds to gentle passive stretching similar to the response seen in idiopathic congenital calcaneo-valgus deformities not associated with posteromedial bowing of the tibia.



Fig. 26.10 A medial screw epiphysiodesis has been performed to correct ankle valgus. The wedge-shaped distal tibial epiphysis and the abnormal proximal location of the distal fibular physis are seen

Serial casting: The more severe degrees usually respond to serial manipulation and cast application; very rarely anterior soft tissue release may be required [13].

Correction of Sagittal, Coronal, or Torsional Deformities of the Tibia and Fibula

Acute correction: Acute correction by performing an osteotomy at the site of the deformity is a simple and attractive option particularly when the residual deformity is in a single plane and when the extent of limb length discrepancy does not warrant formal limb lengthening. Fixation of the osteotomy may be with either internal or external fixation.

Gradual correction: Gradual correction with an external fixator may be the more appropriate option when there are deformities in two or three planes and if concomitant limb lengthening is required.

Correction of the Valgus Ankle

Physeal manipulation (*guided growth*): Valgus deformity of the ankle due to abnormal distal tibial physeal and epiphyseal growth is best addressed by modulating physeal growth such that the growth of the medial part of the physis is restrained using either an extra-periosteal plate and screw device or a single transphyseal screw. The latter option is simpler and the authors' preferred option (Fig. 26.10).

Supra-malleolar osteotomy: If the distal tibial epiphysis is normal in shape and the relative lengths of the tibia and fibula are normal a valgus deformity of the distal tibia should be corrected by a supra-malleolar osteotomy.

Correction of Limb Length Inequality

Contralateral epiphysiodesis: In many instances the projected limb length discrepancy at skeletal maturity is less than 5 cm. An acceptable option is to perform a contralateral proximal tibial and fibular epiphysiodesis at the appropriate time based on the anticipated correction estimated from a growth chart. This option is particularly attractive when the shortening is in the order of 2–3 cm and there are no residual deformities to be addressed.

Limb lengthening: Gradual lengthening of the tibia is typically needed when the limb length discrepancy at skeletal maturity is likely to exceed 5 cm. Lengthening may also be considered for smaller degrees of discrepancy when deformity correction is also required. In such situations both these problems can be addressed by a single procedure [14]. However, the lengthening cannot usually be performed through the osteotomy site used for correcting the tibial deformity as the apex of the tibial diaphyseal deformity tends to be too far distal; a second osteotomy in the proximal tibia is typically required for more reliable new bone formation during limb lengthening. If a circular fixator is used, gradual correction of the deformity at the distal osteotomy site can be undertaken while distraction for limb length equalization proceeds at the proximal osteotomy site. Limb lengthening through an osteotomy in the proximal tibia can also be undertaken with a monolateral frame once acute correction of the biplanar deformity is achieved at the site of bowing in the same sitting (Fig. 26.11a, b).

Correction of Muscle Imbalance Across the Ankle

Tendon transfer: Tendon transfers are seldom effective in restoring powerful push-off if the gastrocsoleus is completely paralyzed [15] but may be effective in posteromedial bowing of the tibia where the gastrocsoleus is weak with some residual muscle function. We prefer to perform a peroneal translocation in which the peroneus longus tendon is re-routed behind the calcaneum under tension (without dividing and re-attaching the tendon as in a tendon classic transfer) to augment the gastrocsoleus in this situation (Fig. 26.12) [16].

Box 26.3. Treatment

- Non-operative correction of calcaneo-valgus foot deformity in infancy.
- Surgical correction of residual deformities of the tibia and fibula after 6 years of age
- Equalization of limb lengths
- Correction of valgus ankle
- Correction of weak plantar flexion

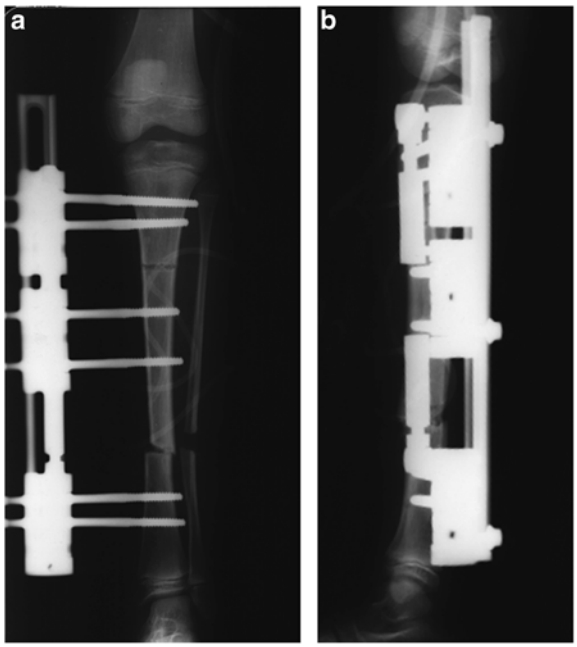


Fig. 26.11 Corrective osteotomy for correction of residual bowing combined with a more proximal osteotomy for limb lengthening has been performed. Acute correction of the posterior (**b**) and medial (**a**) bow was first performed through the distal osteotomy and the fixator was applied taking care to ensure that the normal alignment of the tibia was restored in both planes. The proximal osteotomy for the limb lengthening was then performed

Decision Making

The factors to be taken into consideration while planning treatment are [17]:

- The degree of spontaneous resolution of the tibial deformity in a child over 4 years of age
- The extent of anticipated limb length inequality at skeletal maturity
- The site of ankle deformity, if present

The outline of treatment is shown in Table 26.1.

Pitfalls in Diagnosis and Treatment

Pitfalls in the diagnosis and management of posteromedial bowing can be prevented by careful observation of the site and the plane of the deformities. An erroneous diagnosis of congenital pseudarthrosis of the tibia can be avoided by noting that the direction of tibial bowing is diametrically opposite to anterolateral bowing of pseudarthrosis.

Before embarking on treatment of ankle valgus it is imperative that the site of the deformity is clearly identified. An abnormal wedge-shaped distal tibial epiphysis with an inclined distal tibial articular surface should be treated by medial hemiepiphysiodesis while ankle valgus caused by distal tibial angulation should be treated by a supra-malleolar osteotomy.

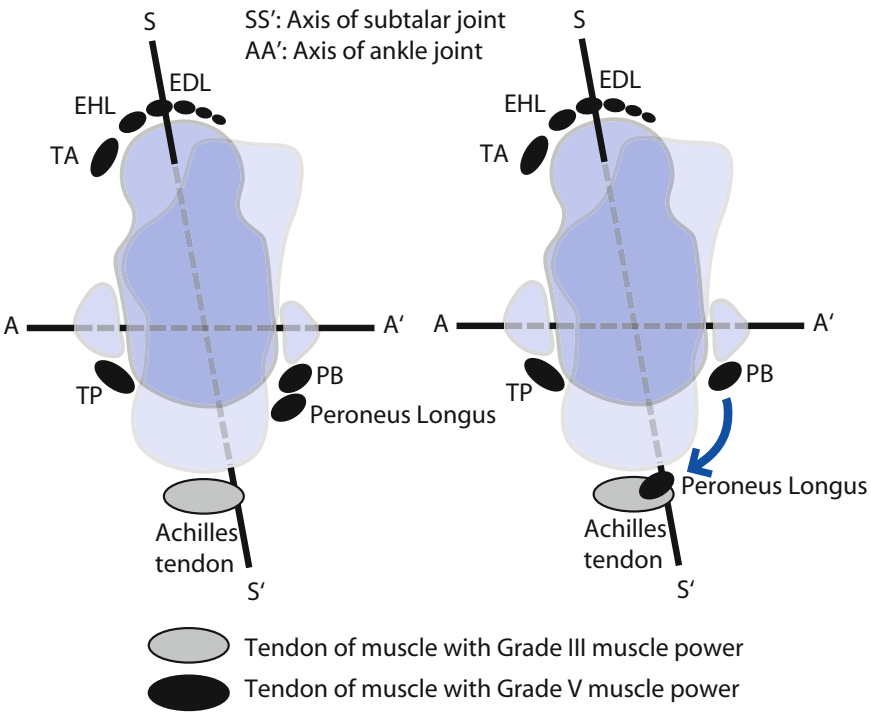


Fig. 26.12 Diagrammatic representation of how the muscle balance is restored across the axis of the ankle joint by performing a peroneal translocation operation

Table 26.1 Outline of management of congenital posteromedial bowing of the tibia[a]

Indication	Treatment recommendation
Anticipated limb length discrepancy at skeletal maturity < 2 cm Near-complete resolution of tibial deformities in all planes No ankle deformity	No intervention
Anticipated limb length discrepancy at skeletal maturity 2–4 cm Near-complete resolution of tibial deformities in all planes No ankle deformity	Contralateral epiphysiodesis of the proximal tibia and fibula at appropriate time so as to equalize limb lengths by skeletal maturity
Anticipated limb length discrepancy at skeletal maturity > 4 cm Near-complete resolution of tibial deformities in all planes No ankle deformity	Proximal tibial lengthening to equalize limb lengths close to skeletal maturity
Anticipated limb length discrepancy at skeletal maturity > 2 cm Unacceptable residual tibial deformity No ankle deformity	Tibial metaphyseal lengthening with diaphyseal osteotomy to correct tibial deformity
Ankle valgus at supra-malleolar level (no wedging of distal tibial epiphysis)	Supra-malleolar osteotomy (in addition to treatment outlined in rows 1, 2, 3, or 4)
Ankle valgus at physeal level with wedging of the distal tibial epiphysis	Medial screw epiphysiodesis of the distal tibia (in addition to treatment outlined in rows 1, 2, 3, or 4)

[a]Modified from [17]

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Controversies in Blount's Disease

27

John G. Birch

Introduction

Much has been written about Blount's disease since its presumed earliest description by Erlacher [1], including early "definitive" (and classic) works of Blount [2] (for whom many have adopted his name to describe the condition) and Langenskiöld [3] (who described the classic radiographic classification of the infantile form of the disease). Blount and Langenskiöld both described two forms of the disorder: occurring in a younger population with generally greater epiphyseal and physeal distortion (infantile Blount's disease) and that occurring in older patients with generally less pronounced epiphyseal and physeal distortion (adolescent Blount's disease). Despite having been described so long ago, this disorder can be aptly described as an arena of controversy: controversy as to name, classification (by age and radiographic type), etiology, and treatment of all types. In this chapter, we review these controversies as well as what is known, and unknown, about Blount's disease

Terminology

Usually progressive proximal tibial varus deformity in an otherwise healthy child or adolescent was termed osteochondrosis deformans tibiae by Blount, and tibia vara by Langenskiöld. Tibia vara (infantile or adolescent) is commonly used in the literature, while some authors prefer to refer to both forms as "Blount's disease."

Classification

Separation of Blount's disease into two distinct forms, infantile and adolescent, is generally accepted. The infantile form typically onsets as an evolution or persistence of physiologic varus, with (usually) progressive clinical and radiographic deformity. The Langenskiöld classification of infantile Blount's is frequently cited (Fig. 27.1). He described six presumably progressive radiographic stages, the mildest form characterized by mild medial epiphyseal and physeal irregularity, with progressively severe distortion, culminating in Stage VI. Stage VI is characterized by complete medial physeal arrest.

Adolescent Blount's disease (Fig. 27.2a) is characterized by later onset, with more subtle physeal and much less epiphyseal distortion. Interestingly, there are neither radiographic classifications nor descriptions of the natural history of adolescent Blount's disease during remaining growth: steady progression of varus deformity once onset during the remainder of growth is assumed.

There is some controversy as to the existence of an intermediate form of Blount's disease (called "juvenile" by Thompson) [4, 5]. This form is characterized as intermediate in age of onset and radiographic severity of epiphyseal and physeal distortion. At best, this form is much less common that either infantile or adolescent Blount's disease. Some cases have been described as having an appearance of a "slipped proximal tibial epiphysis" with diffuse widening and irregularity of the physis, and the impression of medial displacement of a relatively intact epiphysis to produce the varus deformity [5] (Fig. 27.3).

Differential Diagnosis

The most difficult differential diagnosis for infantile Blount's disease is the distinction between persistent physiologic varus and early infantile Blount's disease. Some authors report the efficacy of the metaphyseal-diaphyseal angle in

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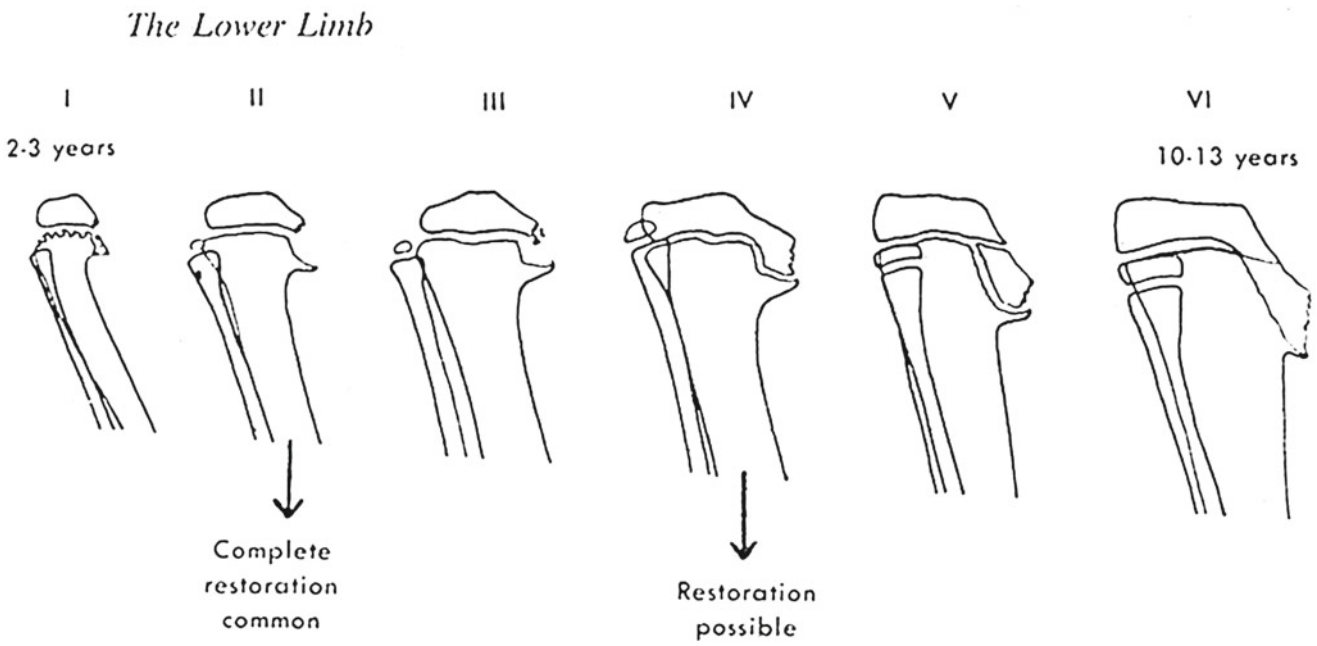


Fig. 27.1 Langenskiöld radiographic classification of infantile Blount's disease. Langenskiöld described six presumably progressive radiographic stages of deformity in infantile Blount's disease. Stage I is often difficult to distinguish from persistent physiologic varus. Langenskiöld noted that spontaneous correction was possible despite radiographic severity as advanced as Stage IV. Stage VI is characterized by complete medial proximal tibial physeal closure (bar). Reprinted from Langenskiöld A. Tibia vara. Acta Chir Scand 1952; 103:9, with permission from John Wiley & Sons

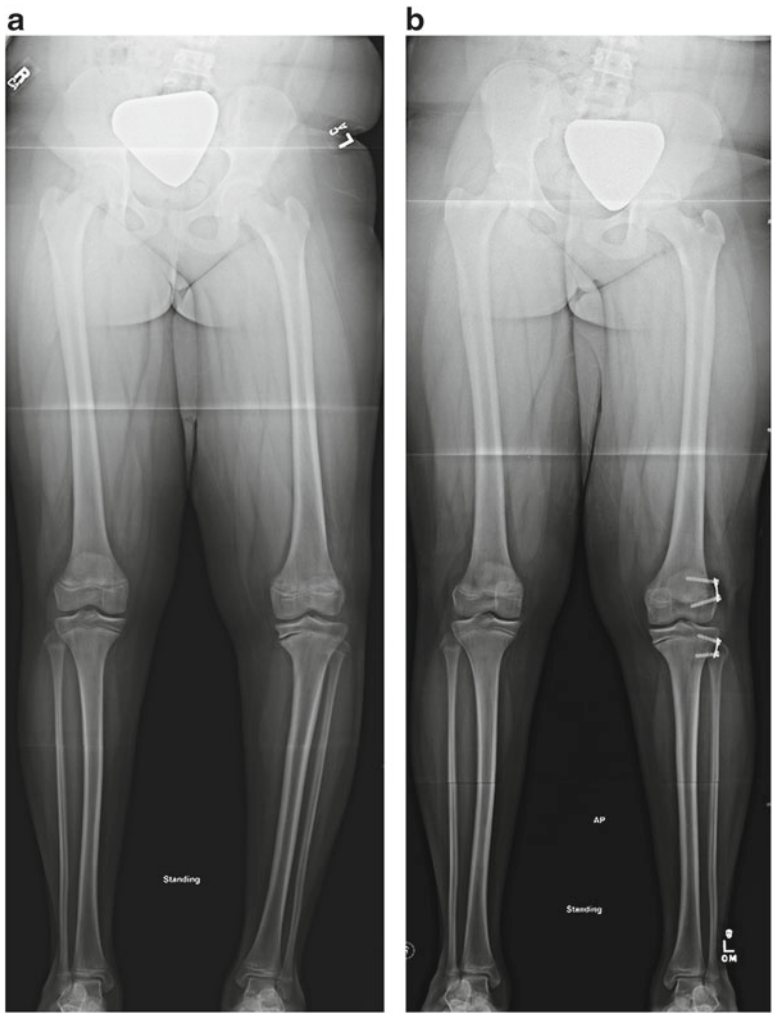


Fig. 27.2 Radiographic appearance of adolescent Blount's disease. (**a**) The characteristic radiographic features of Adolescent Blount Disease include varus deformity of the proximal tibia and medial proximal tibial physeal widening. Distal femoral varus deformity (accentuating the varus deformity) and presumably secondary distal tibial valgus deformity may be present. (**b**) Radiographic appearance after growth modulation of lateral distal femur, lateral proximal tibia, and proximal fibular epiphysiodesis. Note the residual leg length inequality

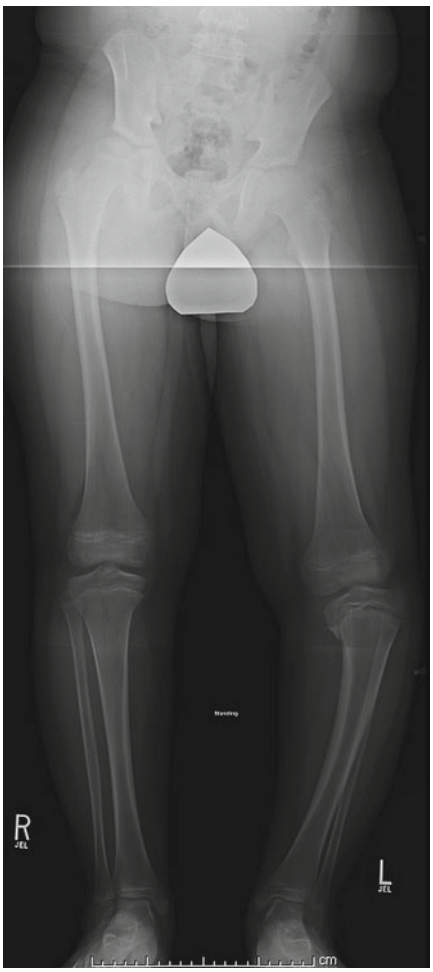


Fig. 27.3 Nine-year-old boy presenting with progressive varus deformity of the left tibia. Note the "intermediate" nature of the patient by presenting age and radiographic epiphyseal/metaphyseal distortion (between "infantile" and "adolescent" Blount's disease). Note the irregularity of the entire proximal tibial physis. Some authors refer to such intermediate cases as "juvenile Blount's disease"

providing this distinction [6–9]. The problem is compounded by the opportunity for true infantile Blount's disease to resolve spontaneously (as reported originally by Langenskiöld [3], and for presumably true persistent physiologic varus to progress to infantile Blount's disease.

Box 27.1. Differential Diagnosis of (Progressive) Varus Deformity in Children

- Persistent physiologic varus
- True infantile Blount's disease (resolving or progressive)
- Renal-metabolic disorders (vitamin D deficiency, renal osteodystrophy)
- Vitamin D-resistant rickets (VDRR)
- Thromocytopenia absent radius (TAR) syndrome
- Focal fibrocartilaginous dysplasia of the proximal tibia
- Epiphyseal dysplasias (multiple; spondylo-epiphyseal; metaphyseal dysostosis)



Fig. 27.4 Anteroposterior radiograph of the left lower extremity of a 13-year-old male with poorly controlled vitamin D-resistent rickets (VDRR). Note significant varus deformity of the distal femur, proximal tibia, and distal tibial as well as widening and irregularity of all physes

Other disorders to consider are epiphyseal dysplasias, dwarfing syndromes, thrombocytopenia-absent-radius (TAR) syndrome, and metabolic bone disease (true vitamin D deficiency, vitamin D resistant (hypophosphatemic) rickets, and the mimicking metaphyseal dysostoses (Schmidt and Jansen types) (Figs. 27.4 and 27.5).

Natural History

Langenskiöld reported that spontaneous correction can occur in infantile Blount's disease, even occasionally in advanced stages [3]. While this certainly seems to be true in milder stages, our experience has been less fortunate in later stages. Thus, the short-term natural history for infantile Blount's disease may be to progress or resolve. The long-term natural history is towards premature intra-articular pathology or early degenerative joint disease, even when angular deformity has been addressed, presumably because of the associated epiphyseal distortion [10, 11].

Interestingly, the short-term natural history of adolescent Blount's disease is not known: no documentation of radiographic changes during growth has been reported. In general, patients appear to have a more benign long-term natural history with respect to degenerative arthritis [10, 12]. However, the association of adolescent Blount's disease with morbid obesity,

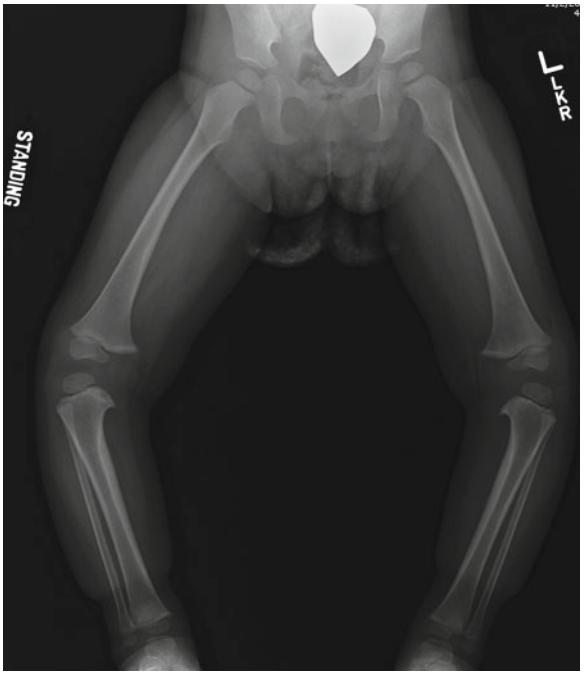


Fig. 27.5 Anteroposterior radiograph of a 3-year-old girl with thrombocytopenia-absent-radius (TAR) syndrome. Note the apparent medial proximal tibial epiphyseal defect and severe resulting varus deformity of the legs. The patient has no thumb or radius

and the recent reports of the failure of deformity correction to impact body mass index portend poorly for the general health and longevity of patients with associated obesity [13–18].

Treatment

Controversies in the Treatment of Infantile Blount's Disease

Does Bracing Work?

There are several studies that address the question of efficacy of bracing in the prevention of progression, or more importantly, the resolution of deformity in infantile Blount's disease [19–21]. Unfortunately, these studies are retrospective, and lack the ability to address fundamental questions such as patient compliance with treatment protocols. Furthermore, wear protocols (day, night, or day-and-night) also vary among published studies. It is not surprising then that authors have arrived at different conclusions, that is, "bracing is effective" (in specific cases) or "bracing is ineffective". We still typically prescribe long-leg, anti-varus braces in patients who are age three or less, and have radiographic Langenskiöld stage II or less infantile Blount's disease, based on our intra-institutional anecdotal and retrospective experience. We further are skeptical about the efficacy of bracing in patients with bilateral infantile Blount's disease, although we may prescribe them even in such cases from time to time, if the clinical picture (e.g., family expectations, resistance to surgery, or similar scenarios) seems to warrant a trial of bracing.

Growth Modulation

Whether identifiable in the published literature or not, it is likely that any experienced pediatric surgeon approaches high tibial osteotomy with trepidation, based on some previous bitter or frightening experience with the major acute complications of that surgery, specifically, compartment syndrome, deep infection, or peroneal nerve injury (never mind skin irritation, challenging fixation/casting scenarios, and recurrent deformity). In that context, the wise surgeon always seeks a more benign treatment, and will generally recommend a trial of the same, if there is any hope of effectiveness. This is the current state of "growth modulation" in infantile Blount's disease [22–26]. The concept is tantalizing: implant one of the commercially available 2-hole (or more) plates on the proximal lateral tibia spanning the physis, for the purpose of temporarily tethering growth in that area, thereby avoiding the significant complication risks associated with high tibial and fibular osteotomy, hope to effect gradual correction of deformity with further growth. Peer-reviewed data confirming the efficacy of such treatment is currently limited, but favorable, Scott et al. [24] reporting 89 % success rate in 18 affected limbs. The likelihood of recurrence of deformity after correction and removal of a growth modulation device is not known. More esoteric questions such as lateral tether due to perichondral ring damage by disruption of appositional epiphyseal/physeal growth by the implant in the very young patient or by direct surgical injury; indications for concomitant epiphysiodesis of the relatively overgrown fibula; or management of the often-associated internal tibial torsion and proximal tibial flexion deformities (Fig. 27.6), remain to be clarified in the era of growth modulation. It is clear that such devices, particularly but not exclusively canulated, titanium screws are at risk for fracture [24]. At a minimum, solid, stainless steel screw devices, with or without more robust (i.e., "H" or similar 4-hole plates) are preferred for growth modulation in Blount's disease. The surgeon and family must contract to regular longitudinal follow up to monitor growth and intervene as needed, and the family should be educated as to the possibility of implant breakage, and failure of this surgical modality, even in the presence of good radiographic indications and apparently technically adequate implant insertion. It is also not clear at what stage of successful correction in the still-growing child a growth modulation plate should be removed, i.e., when the mechanical axis has been corrected, or after allowing the limb to overcorrect into some valgus. Scott et al. [24] reported that three of eight patients with adequate follow up after full correction and implant removal developed mild recurrent varus deformity, although none required treatment.

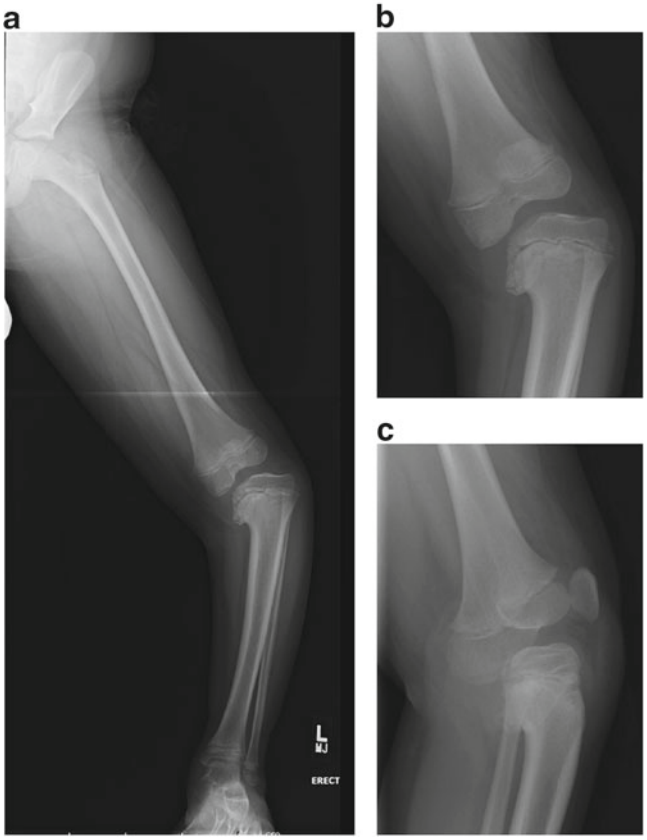


Fig. 27.6 Radiographic appearance of infantile Blount's disease. (**a**) Radiographic appearance of left lower extremity. Note the epiphyseal distortion and varus deformity of the proximal tibia, and mild compensatory valgus deformity of the distal femur and distal tibia. (**b**) Radiographic appearance of the left knee, same patient. (**c**) Lateral radiograph of the knee. Note the procurvatum deformity of the proximal tibia, and the rotational deformity of the limb

Those results and the adage of overcorrecting varus deformity when high tibial osteotomy is performed in similar patients suggest that the surgeon should allow slight overcorrection to occur before removing the growth modulation plate in skeletally immature patients.

Risks of High Tibial Osteotomy and Acute Deformity Correction

High tibial (below the tibial tubercle) osteotomy of the tibia with concomitant osteotomy of the fibula to "over-correct" the varus deformity is the presumptive "gold standard" management of infantile Blount's disease [27–30]. Typically, the distal fragment is externally rotated as well (Fig. 27.7a, b). This operation has serious risks, the most important of which are compartment syndrome and direct or indirect injury to the peroneal nerve from manipulation, traction, or entrapment within fascial planes during manipulation of the fragments (Fig. 27.8). Soft tissue irritation, deep infection, and recurrent deformity are also not inconsequential risks. There are several important considerations the surgeon must assess before proceeding:

1. Is the physis still "open" medially, i.e., is the extent of medial physeal involvement less than Langenskiöld stage VI? If the patient has progressed to Langenskiöld stage VI, recurrent deformity is inevitable, and an alternative or adjunct procedure (e.g., completion of epiphysiodesis effect and management of limb length inequality) selected.
2. What is the current leg length discrepancy, and what's the plan to manage it?
3. What are the concomitant deformities, where are they, and how severe are they? Typically, concomitant deformities include distal femoral valgus (variable), proximal tibial "procurvatum" (flexion deformity), and internal tibial torsion. Typically, all of these should be addressed by at least full correction (some argue for "overcorrection" in the skeletally mature patient) to unload the medial proximal tibial epiphyseal load-bearing [30].

The surgeon has to decide on the geometry and type of osteotomy: opening wedge, closing wedge, oblique osteotomies have all be described [27–30]. In contradistinction to adolescent Blount's, there has been no argument for leaving the fibula intact, presumably because the magnitude of deformity, the desire to overcorrect the varus (coronal plane) deformity, and the need to externally rotate the distal fragment more or less mandate that the fibula be divided to allow full correction. Next to be decided is fixation: cast only, cast and (one or two pins), and external fixation have all been used. Finally, should prophylactic anterior compartment fasciotomy be performed? Some authors support performing a limited anterior compartment fasciotomy in conjunction with tibial osteotomy, while others make no recommendation [27–30]. At a minimum, careful and continuous evaluation of the patient for the presence of compartment syndrome (untoward pain despite adequate release of constrictive splints or casts, or inability to dorsiflex the foot) is essential. In my personal experience, partial prophylactic anterior compartment fasciotomy neither absolutely protects against the development of compartment syndrome, nor is it an entirely innocuous procedure: patients may complain of muscle herniation, discomfort, weakness, or suffer cutaneous nerve injury. The surgeon must carefully weigh his or her strategy in the context of these risks.

Effectiveness of Physeal Arrest Resection

Studies have demonstrated that at least in some cases of infantile Blount's disease (approximately 50 % in our experience), medial proximal tibial physeal growth may be restored by a variation of Langenskiöld's physeal arrest resection surgery [31, 32], usually combined with high (below the physis) osteotomy. This procedure is neither universally accepted nor successful. Patients with Blount's disease typically have

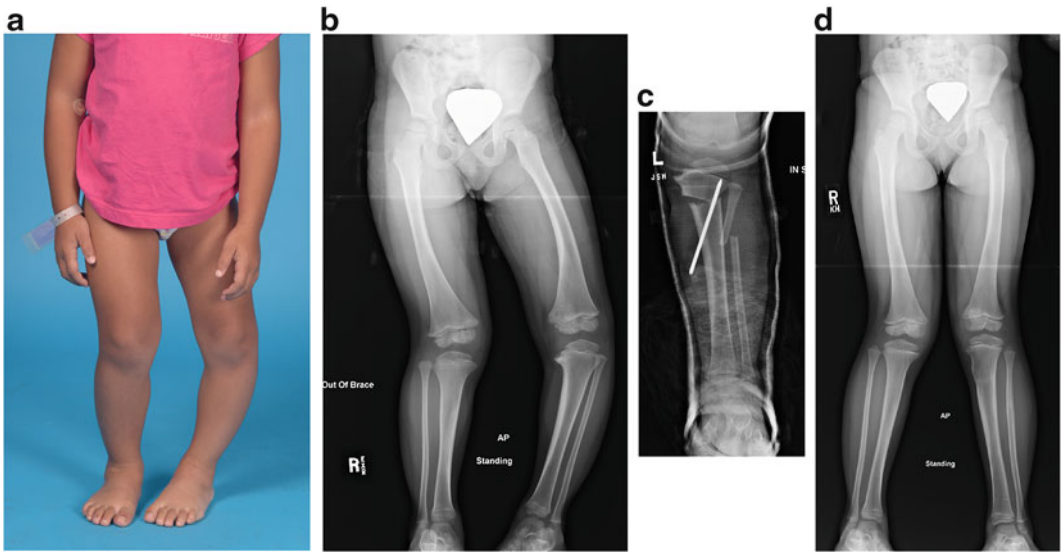


Fig. 27.7 Three-year-old girl with progressive infantile Blount's disease of the left leg, Langenskiöld stage III, who failed conservative treatment with an anti-varus brace. (**a**) Preoperative clinical appearance. (**b**) Preoperative radiographic appearance. (**c**) Postoperative radiographic appearance. Note the lateral translation of the distal fragment to effect restoration of the tibial mechanical axis. The distal fragment is manipulated to correct both varus and internal rotational deformities associated with infantile Blount's disease. (**d**) Radiographic results 2 years postoperatively, age 5 years. Patient has symmetric valgus deformity, with improvement of epiphyseal distortion of the left proximal tibia

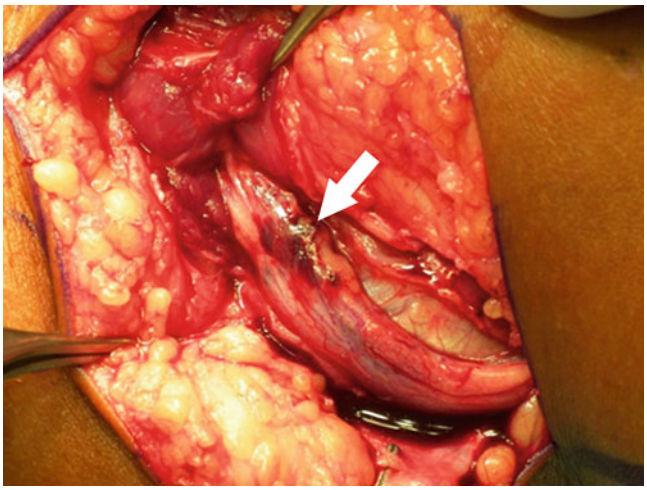


Fig. 27.8 Peroneal nerve compression detected and released in the immediate postoperative period. The patient developed peroneal nerve palsy shortly after surgery. Her compartments were soft. At exploration, the nerve was found to be compressed under a fascial band as it passed into the deep anterior compartment. *Arrow* indicates contused peroneal nerve after release of the fascial band. Same patient as Fig. 27.7

an advanced bone age [33]. The wise surgeon will assess the patient's bone age and the adage of "average" proximal tibial physeal growth of 6 mm per year in the healthy proximal tibial physis to determine if adequate growth remains to warrant an effort at this procedure. A minimum of 4 years of growth remaining would seem to be a reasonable threshold criterion for consideration of physeal bar resection as potentially indicated. Our experience is that even patients who experience resumption of growth will subsequently develop late deceleration/cessation of that growth. Embedding metallic markers or some other method of exact determination of growth persisting is important to allow prompt recognition of secondary cessation of growth (Fig. 27.9a, b). Completion of the epiphysiodesis at that time can save the patient from the development of recurrent deformity requiring an additional osteotomy, and appropriate management of any associated leg length inequality.

Utility of Physeal Arrest Resection Surgery in the Absence of a Bony Physeal Arrest

Andrade and Johnston [34] have recommended "physeal bar resection" surgery in patients with advanced infantile Blount's disease, but at Langenskiöld stage less than VI, i.e., without demonstrable frank physeal bony bar formation on advanced imaging. They report good (>80 % successful in preventing recurrence of varus deformity, in patients less than 7 years of age) results, but there is not a comparable group treated by high tibial osteotomy alone to confirm the relative efficacy of this procedure.

What to Do with the "Failed" Case?

Usually, the surgeon is faced with many things to consider, when the patient has undergone previous high tibial osteotomy with recurrent deformity. These include: the extent of

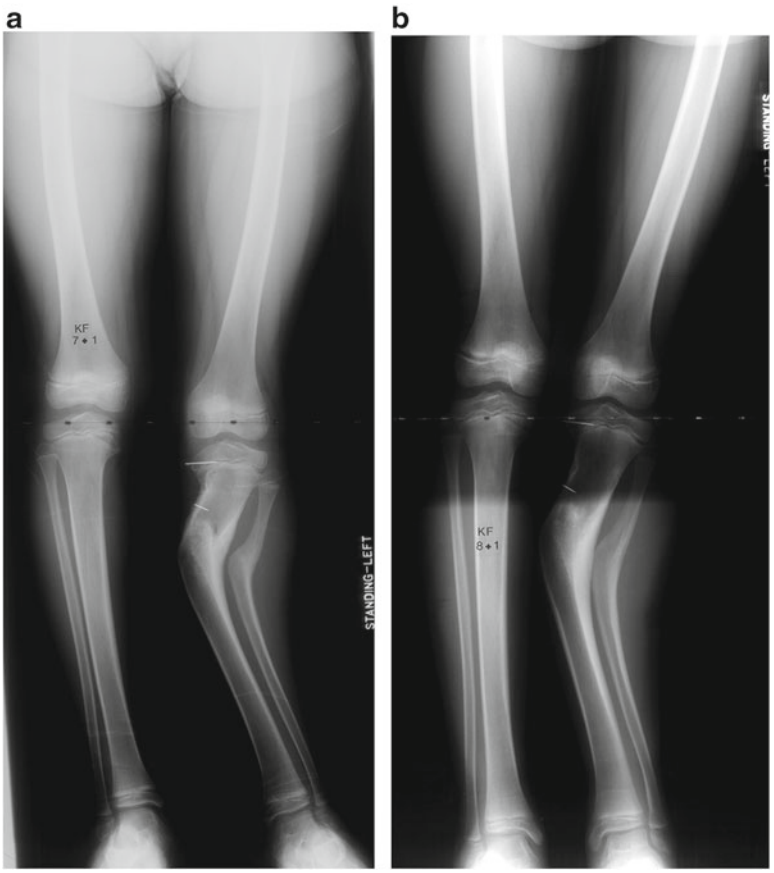


Fig. 27.9 This 6-year, 9-month old girl underwent third-time repeat hight tibial osteotomy, combined with partial physeal bar excision for recurrent infantile Blount's disease of the left tibia. (**a**) Four months postoperatively, the patient has persistent iatrogenic valgus deformity at the osteotomy site. Note the metallic markers in the proximal medial tibial epiphysis and metaphysis. (**b**) Sixteen months postoperatively (1 year after radiograph in part **a**), the patient has persistent valgus deformity. The markers have separated by approximately 20 mm

lower extremity scarring, recurrent (potentially complex) deformity, including variable amount of varus, procurvatum, and internal tibial torsion, leg length inequality in unilateral cases, and potential continued asymmetric growth in the skeletally immature patient (both within the tibia due to the medial arrest, and between the affected and unaffected legs).

There can be no "cookbook" strategy in such cases (Fig. 27.10). The surgeon must individualize the correction plan with careful consideration of all these parameters in the development of a treatment strategy suitable and acceptable to the family and their circumstances. External fixation, with either acute or gradual correction, provides the surgeon with an excellent tool to manage all of these deformities during one treatment stage, if acceptable to the patient and family.

The surgeon must always keep in mind the fact that patients with infantile Blount's disease may have premature or "atraumatic" meniscal tears or other degenerative changes, even in the presence of presumably adequate angular and rotational deformity correction [10, 35]. This risk is presumably the result from the epiphyseal distortion that is unique to infantile Blount's disease and which distinguishes it from adolescent Blount's disease. Thus, if the patient has significant intra-articular complaints or abnormalities on physical examination, appropriate pursuit of the potential for these structural abnormalities (MRI or arthroscopy) should be carried out.

Is There a Role of "Hemi-Plateau Elevation"?

Radiographs in the advanced stage of infantile Blount's leave the observer with the distinct impression of a "sagging" or "depressed" medial tibial plateau. This perception (or reality) has led a number of authors to describe "hemi-plateau elevation" [36–38] (in effect, an intra-articular fracture with restoration of the transverse condylar relationships [unless the surgeon is fortunate enough to effect cartilaginous deformation of the articular surface without fracture]). It should be noted that some other authors dispute at least the universal presence of a true depression of the medial plateau, and confirmed the presence of a cartilaginous medial tibial plateau by knee arthrogram [39], or more recently, MRI [35, 40]. The procedure of hemi-plateau elevation has been described in isolation or in conjunction with second osteotomy of the tibia to correct coexisting varus deformity. The surgeon should recognize the most likely event of intra-articular fracture, and the inevitability of no further (favorable) physeal growth, if performing this surgery; thus, completion of the proximal tibial and fibular epiphysiodesis at the time of surgery would seem appropriate. We have not, in our institutional experience, used this procedure to manage even the most severe cases of infantile Blount's disease.

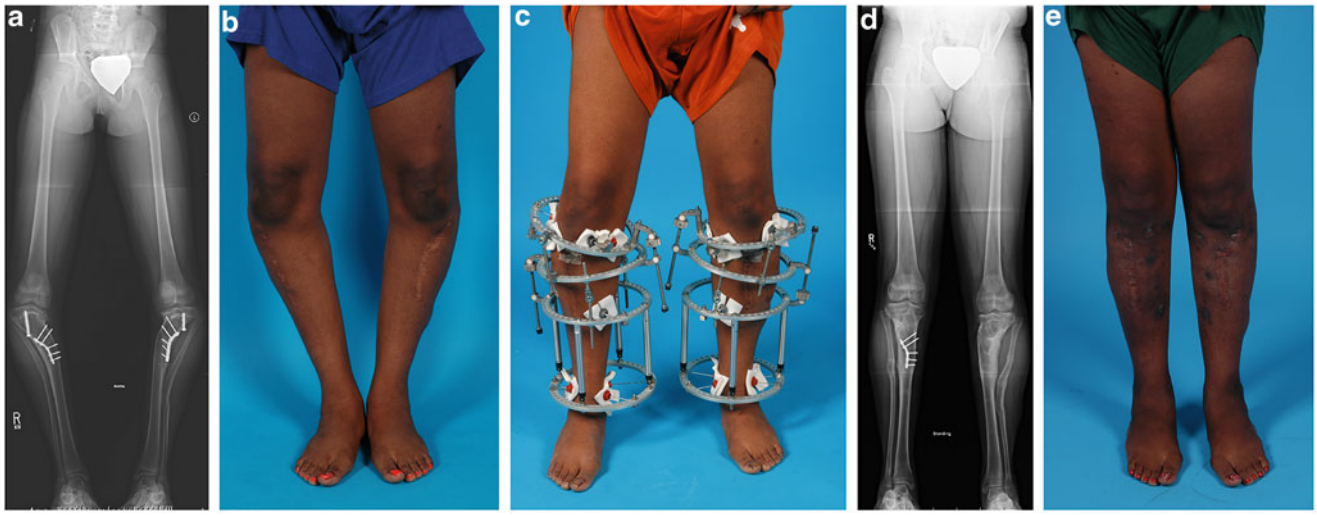


Fig. 27.10 Patient with failed previous treatment of infantile Blount's disease. (**a**) Radiographic appearance. The patient has had several prior surgical procedures, including high tibial osteotomies and attempted lateral proximal tibial physeal arrest with retained implants, recurrent varus deformity, and post-surgical proximal metaphyseal valgus deformity. Note the medial translation of the patient's mechanical axis caused by a failure to lateralize the distal fragment in conjunction with the previous valgus-producing osteotomy. (**b**) Clinical appearance. (**c**) Clinical appearance during staged treatment. The patient was managed by staged surgical procedures, including (partial) implant removal, and two-level osteotomies of the tibiae with gradual correction of the varus and valgus deformities using a circular external fixator. (**d**) Final postoperative radiographic appearance. (**e**) Final clinical appearance

Box 27.2. Controversies in Infantile Blount's Disease

- Etiology, including obesity
- Differentiation between persistent physiologic varus and true infantile Blount's disease
- Efficacy of lower extremity bracing
- Efficacy, timing, and time-to-removal of growth modulation plates for surgical correction

Box 27.3. Controversies in Adolescent Blount's Disease

- Etiology, including obesity
- Role of growth modulation in surgical armamentarium
- Indications for correction of associated deformities (distal femoral varus, distal tibial valgus)
- Gradual versus acute correction by high tibial osteotomy, with or without external fixation

Adolescent Blount's Disease

While adolescent Blount's disease was described by both Langenskiöld and Blount in their original series, side by side with infantile Blount's disease, it is clear that adolescent Blount's disease is a completely distinct disorder. Not only do infantile and adolescent Blount's diseases affect an entirely different age group, the radiographic features, nature and location of concomitant ipsilateral limb deformities, incidence of intra-articular abnormalities, and published long term premature degenerative arthritis prevalence are all distinctly different as well [10–12]. Furthermore, while obesity seems to be a risk factor for both conditions, obesity seems to be more specifically related to adolescent Blount's disease [13–16, 18].

No specific useful radiographic classification exists for adolescent Blount's disease, either for morphological characterization or prognostic purposes. The "classic" deformity consists of proximal tibial varus deformity of the affected extremity with a medial proximal tibial physeal widening or "zone of injury" on radiographs (see Fig. 27.2). Biopsy specimens from this area of radiographic physeal lucency show disorganized microscopic physeal architecture, and/or replacement of normal physis by fibrous tissue. It is assumed that some patient risk factor (familial, persistent mild physiologic varus, subclinical metabolic abnormality) with or without obesity results increased compressive forces across the physis with disruption of normal function and deceleration of growth [4, 41, 42]. There are no short-term natural history reports to confirm the presumption of progressive worsening varus deformity or radiographic abnormality in

untreated patients during the remainder of growth. Setting aside general health issues related to obesity, with or without persistent varus deformity of the leg, published reports suggest a more benign natural history with respect to degenerative arthritis of the knee in patients with adolescent Blount's disease compared to infantile [10–12].

Treatment Considerations in Adolescent Blount's Disease

Role of Obesity and Impact of Treatment on Obesity

It is well recognized that obesity in the general population is increasing and represents a significant health problem. Sadly, that increased incidence is extending into the adolescent and now pediatric population [14–16, 43–45] with significant implications for long-term health consequences. It has been long noted that patients with adolescent Blount's disease are frequently (but not universally) obese or morbidly obese, and obesity, rightly or wrongly, is considered a risk factor for the development of adolescent Blount's disease. Interestingly, there are no reliable population surveys that confirm an increased incidence of adolescent Blount's disease in association with the increased prevalence of adolescent obesity in the general population. Soberingly, Sabharwal et al. [17] have noted that correction of angular deformity (both of infantile and adolescent Blount's disease) is NOT associated with subsequent reduction in BMI in their study population, thus calling into question the myth or circular argument that leg deformity begets pain and exercise intolerance, with the implicit expectation of surgeon and patient that deformity correction will result in increased physical activity and subsequent weight reduction (Fig. 27.11). This calls into question what our responsibility as surgeons should be: while we console ourselves that deformity correction will open the door to a healthier life-style and thus represents a noble pursuit on behalf of the patient, warranting tolerance of myriad increased challenges and complications associated with performing angular deformity correction in patients with morbid obesity (anesthesia risks, sleep apnea, wound complications, deep infection, fixation challenges, and greater difficulty effecting complete deformity correction). Furthermore, there is no clear evidence that correcting angular deformity associated with Blount's disease increases the longevity of the knee in affected extremities. Thus, as patient health advocates, we must seriously question the merit of surgical treatment for adolescent Blount's disease until an effective weight management program has been embraced by the patient.

In the overall picture, probably the most important associated abnormality in patients with adolescent Blount's disease is morbid obesity, because not only is it a threat to the patient's overall health, its presence results in specific potential for sleep apnea, type II diabetes, and increased risks of delayed wound healing, deep infection, and myriad soft tissue challenges in the perioperative period. In addition, patients with obesity will mask significant varus deformity (indeed, the "fat-thigh syndrome" as described by Davids [46] is a potential cause of adolescent Blount's: the need to stand with the legs separated to accommodate fat thighs results in the skeletally immature a secondary adaption of medicalization of the lower leg to bring the feet closer to the midline) (Fig. 27.12). One of the consequences is that "full" correction of the varus deformity will typically make the patient appear to have valgus deformity (Fig. 27.13), which



Fig. 27.11 Recurrent infantile Blount's disease in an obese child. (**a**) At age 8, this patient had progressive, recurrent infantile Blount's disease associated with obesity. (**b**) At age 15, the patient's deformities have recurred after high tibial osteotomy, but through a self-directed program of diet and exercise, the patient lost weight despite the leg deformities, and at this time, weighs less than he did at age 8



Fig. 27.12 Patient with adolescent Blount's disease and morbid obesity. The "fat thighs" require the patient to stand with the legs relatively abducted. Some conjecture that this posture in an adolescent may be the cause of adolescent Blount's disease, at least in some patients

is usually cosmetically displeasing to the family/patient. This puts the surgeon in a vexing situation of either inadvertently or deliberately undercorrecting varus deformity, or justifying the production of deformity in the patient's perception. There is no easy answer here, but we usually prefer to correct the deformity anatomically (as best we can), explaining the outcome and its rationale in advance and in the (perhaps unlikely) hopes that the patient will lose weight in the future. The surgeon should educate the patient and the family regarding the presence of associated deformities, and be prepared to address them, should the need arise.

Associated Deformities

In contradistinction to infantile Blount's disease, ipsilateral deformities of the distal femur and distal tibia are frequent (see Figs. 27.2a and 27.13) [47–50]. Interestingly, the most common associated deformity of the distal femur is an accentuating varus, in contradistinction to some cases of infantile Blount's disease and many traumatic physeal

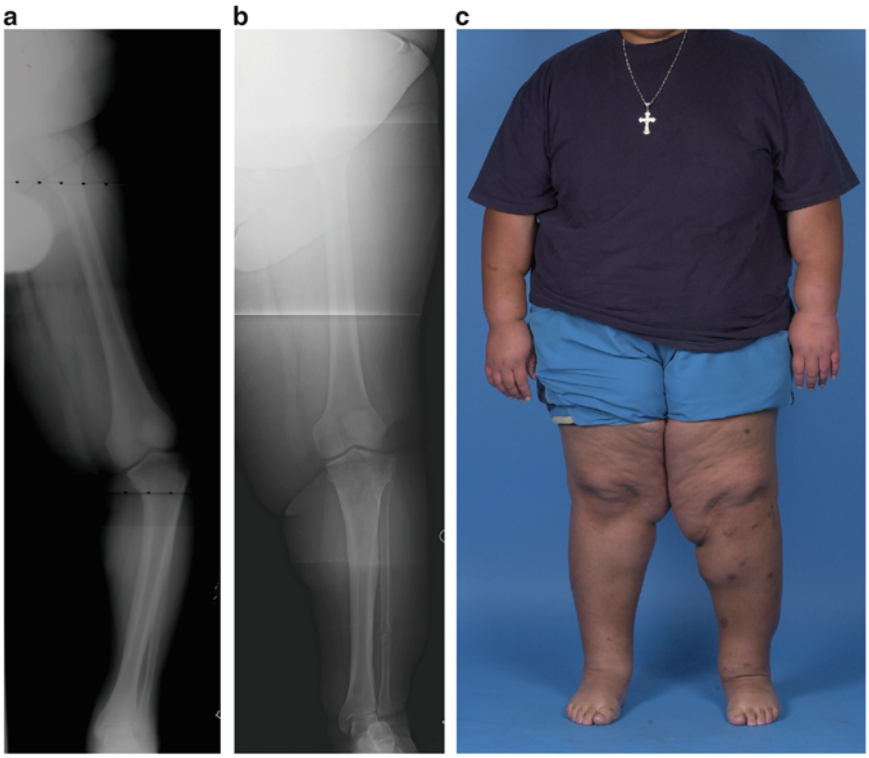


Fig. 27.13 Apparent valgus "deformity" after correction of adolescent Blount's disease varus deformity in the left leg of an obese patient. (**a**) Preoperative radiographic appearance. (**b**) Postoperative radiographic appearance. Note the incomplete correction of the varus deformity. (**c**) Postoperative clinical appearance. Note that despite incomplete correction of the varus deformity radiographically, the patient appears to have valgus deformity clinically



Fig. 27.14 Apparently healthy adolescent with left-sided varus deformity typical of adolescent Blount's disease, developed apparently idiopathic contralateral valgus deformity ("windswept" deformity). There was no identifiable endocrine or skeletal dysplasia abnormality in this patient

growth disturbances which often develop at least a partial compensating reverse deformity at the adjacent physes. This suggests a more global etiology (rather than simple deceleration of growth of the medial proximal tibial physes or unrecognized endocrinopathy as the underlying cause or at least accelerant of adolescent Blount's disease. Along those lines, interestingly, several patients we have treated have developed a contralateral valgus deformity (i.e., "windswept" appearance) during the course of treatment for presumptive adolescent Blount's disease (Fig. 27.14), in the absence of an identifiable metabolic disorder or endocrinopathy. This too, would suggest a systemic component to the etiology of adolescent tibia vara, at least in some patients.

Distal tibial valgus (which would seem a more intuitively logical consequence of proximal varus deformity) is often noted in patients with adolescent Blount's disease. "Comprehensive correction" strategies [47] describe surgical correction of this deformity. However, it has been our experience that patients rarely complain of ankle pain or deformity, and only rarely has primary or secondary correction of this deformity been necessary in our patient population.

Finally, similarly to infantile Blount's disease, patients will usually be noted to have an internal tibial torsional deformity on clinical examination, and on careful radiographic assessment, "procurvatum" (flexion deformity) of the proximal tibia [50]. The severity of these deformities must be taken into consideration in determining the best surgical treatment strategy for individual patients.

Growth Modulation

Open hemi-epiphysiodesis of the lateral proximal tibia, with or without concomitant epiphysiodesis of the proximal fibula and/or lateral femur was the first described method of "growth modulation" of this deformity [51]. It is interesting to note that in at least some cases, this procedure is effective despite the persistence of presumed "causative roles" of deformity and obesity in the disruption/deceleration of normal medial proximal tibial physeal growth. Considering this procedure with acceptance of an uncertain outcome is almost universally motivated by the challenges and risks created by osteotomy, particularly in the obese patient. Open hemi-epiphysiodesis has been replaced by plate (or staple) growth modulation methods [25, 52, 53] (see Fig. 27.2b). Enthusiasm for the original "8" plate (titanium cannulated screws and 2-hole plate) has waned in favor of stronger non-cannulated stainless steel screws, and/or more robust "H" or similar plates (i.e., two screws on either side of the physis) due to reports of screw fracture with the original device [22, 23], an event which seems to be nearly unique to the metaphyseal screws in Blount's patients (particularly adolescent type). Both the surgeon and patient must accept that even apparently technically adequate implant placement and retention do not guarantee effective correction of the angular deformity. For this reason, and for the happy possibility of correction prior to skeletal maturity, both the patient and surgeon must commit to regular longitudinal follow up to prevent overcorrection, and manage failure to correct appropriately.

Acute Correction with Internal Fixation

High tibial osteotomy (either above or below the tibial tubercle, at the surgeon's preference, because the patient is nearly skeletally mature) can be opening wedge, closing wedge, or a combination at surgeon's preference. Due to the average patient's age, size, weight and body habitus, rarely is pin fixation and cast immobilization practical or effective. The surgeon is therefore left with the decision of internal fixation or external fixation, the latter of course then subjecting all to the inconvenience, pin care, and increased soft tissue irritation of such patients. Internal fixation is typically of some plate device. Some authors have noted that fibular osteotomy

is not necessary if modest opening-wedge correction strategy is used [54].

The patient is at risk for wound dehiscence, deep wound infection, delayed union, non-union, implant failure, compartment syndrome, inadequate correction, and the inability to adjust correction postoperatively (without revision surgery); some of these complications are inherent to tibial osteotomy and the patient's body habitus, and some are risks associated with acute correction and internal fixation strategy: the surgeon and patient should be cognizant of these risks and deem them acceptable in context.

Correction with External Fixation (Acute or Gradual) (with 6-Strut Fixators/Others)

Gradual correction of adolecent tibia vara [54–63] offers several theoretical and practical advantages: there is presumably a lower risk of compartment syndrome and deep infection, greater deformities can be addressed more precisely, lengthening can be achieved, it is easier by fragment translation to effect a truly anatomic restoration of limb alignment, and the complex deformity (with procurvatum, internal tibial torsion, with or without shortening) can be addressed, either acutely, gradually, or sequentially. For smaller deformities with minimal lengthening required, the fibula may remain intact and unfixed (at least proximally). Six-strut (hexapod or Taylor Spatial Frame™) are particularly valuable for this deformity, although traditional circular fixators or monolateral fixators are preferred by some authors.

The flexibility of external fixation devices and strategy however, have to be weighed against the generally more prolonged postoperative recovery and management of the device and the patient in the device, particularly with the increased soft tissue irritation in this specific patient population [63].

Adolescent Blount's Disease Summary

Patients with adolescent Blount's represent a significant challenge to the clinician because of the complex nature of the deformity itself, the frequent coexistence of other ipsilateral deformities, and the frequent occurrence of large size and/or obesity. The surgeon must carefully assess the overall health status, commitment and expectations of the patient, the presence and severity of concomitant deformities, and existing and projected leg length inequality before adopting a well-reasoned treatment strategy for the patient. Epiphysiodesis, growth modulation, acute correction with internal or external fixation, and gradual correction with external fixation are options that may in any combination represent excellent strategy in individual cases.

Does Juvenile Blount's Disease Exist?

Some authors [4, 5] note a third, intermediate type of Blount's disease, intermediate in time of onset (between infancy and adolescence) and radiographic deformity (having less epiphyseal distortion typical of infantile Blount's, but more extensive physeal changes than seen in classical adolescent Blount's). We have a few cases which fit this description, and whom do not neatly fit either "traditional" category, and thus believe this is a separate entity. Treatment decisions, however, remain the same as for either condition, except that conservative treatment (observation or bracing) would seem to be unhelpful modalities. There are no reports or personal experience with growth modulation strategies in such patients to our knowledge.

Summary

Infantile and adolescent Blount's diseases must be considered separate and distinct entities, occurring in a dissimilar patient population, at different ages, with entirely different radiographic features, concomitant deformities, and, likely natural history. What they share are a lack of our understanding as to their etiology, and (presumably) progressive, complex varus deformity of the tibia, which challenges the surgeon to create an effective treatment plan. Gradual correction of these deformities by growth modulation or by osteotomy and external fixation, with or without lengthening, are important and valuable surgical correction techniques that should be in the surgeon's armamentarium. Many aspects of infantile and adolescent Blount's disease warrant attention by further research, including identification of their etiologies, favorably influencing childhood obestity, unequivocally determining the effectiveness of bracing in the infantile form, developing an effective growth modulation method of correcting the associated internal tibial torsional deformity, and determining an appropriate threshold for correcting concomitant deformities of the distal femur and distal tibia.

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Part V

Sequelae and Complications

Methods to Enhance Bone Formation in Distraction Osteogenesis

28

Hae-Ryong Song, Dong Hoon Lee, Seung-Ju Kim, and Ashok Kumar Ramanathan

Introduction

Regeneration of bone defects resulting from conditions including trauma and congenital deformity correction by distraction osteogenesis (DO) is one of the most interesting issues in the field of regenerative medicine [1]. DO is an increasingly popular technique used to stimulate new bone formation to treat orthopedic disorders resulting from bone defects and deficits [2]. However, the long treatment period with various complications [3] and the potential for nonunion under some circumstances remain major limitations of current treatment procedures. Regeneration of bone in the presence of rigid fixation and maintenance of the osteogenic tissue (marrow, endosteum, nutrient artery, and periosteum) require many factors for stimulation of bone regeneration [4]. Therefore, the development of procedures to accelerate the formation and maturation of regenerate is clearly desirable.

To regenerate the bone defects, autologous and allogenic bone grafting are the most widely used methods. However, these methods are associated with a number of drawbacks, including donor site morbidity and blood loss. To overcome these drawbacks, tissue engineering strategies have demonstrated a potential role in developing bone graft substitutes. Different mechanical and biophysical stimuli could provide effective augmentation of bone regenerate maturation. These include biologic stimulation such as bone morphogenetic proteins (BMP), platelet-rich plasma (PRP), stem cells, parathyroid hormone, and growth hormone [5–7]. In addition, physical stimulation such as ultrasound, mechanical, and electromagnetic stimulation has been used [8–11]. Even though a bone defect during DO possesses the intrinsic capacity to heal spontaneously following surgery, beyond a certain critical size defect bone may not heal adequately by itself and further intervention is often required.

Box 28.1 Quantitative Methods to Monitor the Regenerate Bone

- Quantitative ultrasound
- Dual-energy X-ray absorptiometry
- Quantitative computed tomography
- Pixel value ratio method

Monitoring of Regenerate Bone

Various methods have been proposed for the monitoring of the regenerate bone [12] quantitatively and qualitatively. Indirect quantitative methods, including ultrasound, dual-energy X-ray absorptiometry (DEXA), and quantitative computed tomography, have already been found to be sensitive [13–15]. However, owing to its high cost, inability to detect soft tissue changes, and poor measurements in the presence of artifacts, the routine use of such modalities in clinical practice is not popular. An image analysis method is also proposed for the monitoring of the regeneration of the bone. Radiographic investigation is one of the most important methods because it is the simplest; it provides continuous information and it can

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be easily interpreted. A recent approach that is being used to determine callus stiffness is to evaluate the pixel value ratio (PVR) using a picture archiving and communication system (PACS). The PVR is the ratio of the proximal segment to that of the regenerate, and it correlates well with the bone mineral density (BMD) [16, 17]. Shim et al. [18] analyzed serial pixel values of different cortices in both femoral and tibial lengthening and demonstrated that pixel values for distraction callus conformed to a sigmoidal curve. Song et al. [19] reported that BMD measurement can thus function as an effective adjunct to measure callus stiffness, along with PVR, using digital radiographs, especially in situations where callus maturation and stiffness are doubtful.

Many studies [20–22] have been published in an attempt to find a relation between the callus pattern and the prognosis of the limb lengthening procedure because such a system or classification can help the surgeon to quantitatively assess the progress of the distraction callus and to predict the chance of callus fracture and can thus impact the clinical outcome of the procedure. Devmurari et al. [23] reported that the Ru Li classification [24] is an effective method for the evaluation of the chance of callus fracture and a lucent pathway was seen in all fracture cases with concave, lateral, and other shapes (Fig. 28.1). Early removal of the fixator may lead to fracture of the regenerate, whereas prolonged use leads to joint stiffness, pin-site infection, and discomfort to the patient, as well as delayed complications like subsidence and angulation [25, 26]. Research is in progress to define callus maturation and corticalization and hence to identify the ideal time for removal of the fixator.

Physical Stimulation

Mechanical Stimulation

Controlled mechanical stimulation is considered to be an important stimulus for the regenerate in distraction osteogenesis. There are preclinical and clinical evidences that physiologic weight bearing stimulates new bone formation and consolidation of new bone in distraction osteogenesis [27–30]. Controlled compressive but not shear force has been shown to affect the biologic nature of the healing response by early expression of BMP 2/4 and the messengers for collagen 1 and osteocalcin [31] and increase the size and strength of the regenerate [30, 32].

"Pumping" of the regenerate (also referred to as the accordion maneuver)—the procedure of alternative compression and distraction—is a well-known method to stimulate new bone formation in the clinical situation of poor regenerate [33, 34]. But, from the literature review, we found that it is still a conflicting issue. Mofid et al. [34], in an animal model of mandibular distraction, performed daily alternative compression and distraction at an amplitude of 1 mm/day for 3 weeks, and observed improved bone healing in terms of cortical thickness, cortical:cancellous bone ratio, and mineral apposition. In contrast, Greenwald et al. [33], in an animal model of mandibular distraction, distracted to a length of 2.5 mm (at a rate of 0.25 mm twice daily) for 5 days, then compressed 1.0 mm for a 2-day period, and redistracted to a length of 5 mm. They demonstrated no significant differences between experimental ("pumping") and control group in radiological and histological parameter. The timing of oscillating motion is also a conflicting issue. Kassis et al. [35] showed that the effect of axial micromotion is effective only after the end of elongation rather than during active distraction. Further investigations are necessary to establish the effect and the optimal protocol for "pumping" including the timing, amplitude of sequential movement, and frequency.

Rhythm of distraction is another conflicting issue for enhancing bone regenerate. Ilizarov in an animal model demonstrated better callus in motorized distraction group (1/60 mm, 60 times/day) compared with the standard rhythm (1/4 mm, 4 times/day) [36, 37]. Mizuta proved that an increase in the distraction frequency (eight steps vs. four steps) in open-wedge osteotomies of the proximal tibia with hemicallotasis provides better bone formation, resulting in a shorter external fixation period [38]. Meanwhile, there have been reports that the rhythm of distraction does not significantly influence bone healing [39, 40]. Lengthening procedure is not confined to the bone and soft tissues are also important. Makarov et al. demonstrated in a goat model that a more fractionated rhythm of distraction was associated with enhanced preservation of muscle fibers and greater regenerative activity of the muscle [41]. Further investigation is necessary to clarify the benefit of the more fractionated distraction, and the optimal rhythm.

Low-Intensity Pulsed Ultrasound

Low-intensity pulsed ultrasound (LIPUS) is a form of mechanical stimulation that is delivered as high-frequency acoustic pressure waves. The physical process through which LIPUS stimulates bone healing remains unclear. It is speculated that ultrasound enhances fracture healing by inducing low-level mechanical forces at the fracture site, reproducing the effect of functional loading [42]. The mechanisms through which the mechanical signal is translated into a biochemical signal have not been fully understood but ultrasound seems to influence certain cellular reactions involved in each phase of the healing process such as inflammatory reaction, angiogenesis, chondrogenesis, intramembranous ossification, endochondral ossification, and bone remodeling [42, 43]. LIPUS is one of the best studied treatment modalities in distraction osteogenesis, but the results from

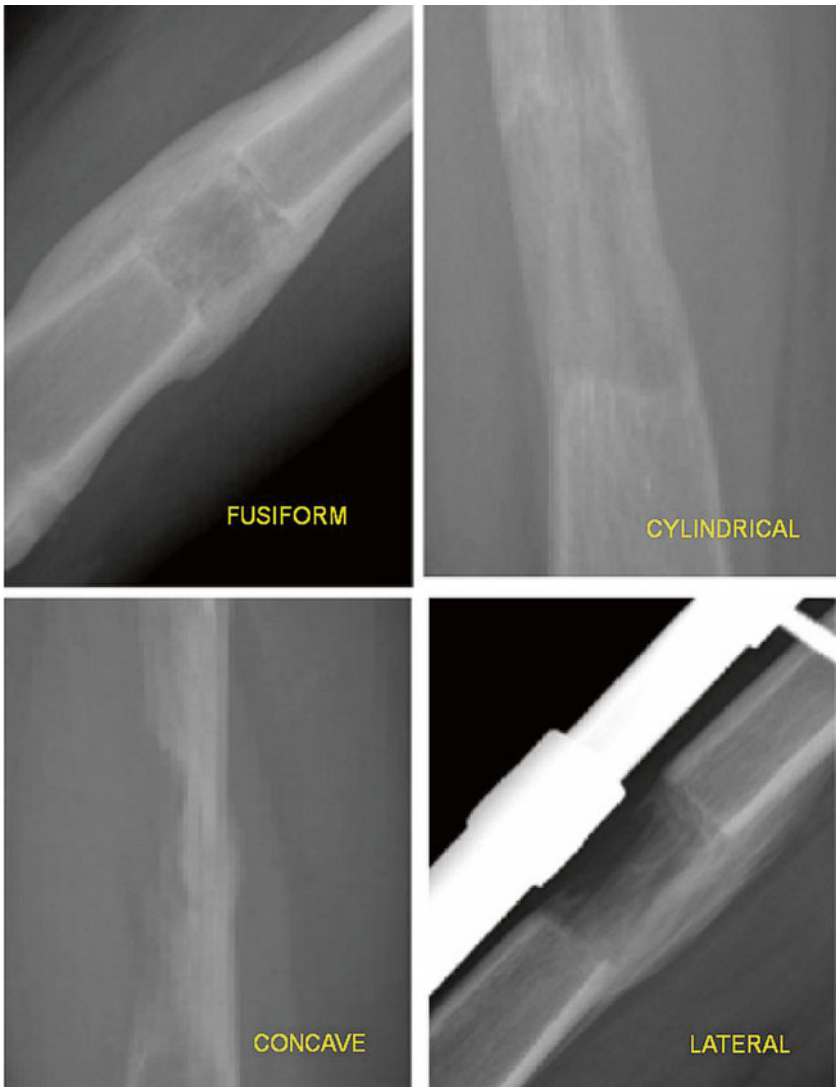


Fig. 28.1 Callus patterns

preclinical experiments are still conflicting regarding its efficacy [44, 45]. There are well-designed clinical studies for the acute fracture healing, suggesting that LIPUS can be beneficial in the treatment of certain non-operatively treated fracture by shortening the time to radiological and clinical union [46–48]. Most successful reports with LIPUS are in the treatment of conservatively managed acute fractures. There are only scanty evidences for surgically treated fracture and most of them reported disappointing results [49–51]. Emami et al. concluded that low-intensity ultrasound treatment did not shorten healing time in acute tibial fractures treated with a reamed and statically locked intramedullary nail [49].

The effect of LIPUS on distraction osteogenesis is not conclusive especially regarding the timing of application [44, 45, 52, 53]. Further well-designed studies are required to clarify the role of LIPUS in humans undergoing DO.

Pulsed Electromagnetic Field

Most of the available studies regarding pulsed electromagnetic field (PEMF) are related to its effect on the treatment of acute fractures [54–56] and suggest that PEMF may be beneficial in the treatment of certain non-operatively treated fractures. However, there are only a few reports about its effect on distraction osteogenesis [11, 56–58]. Though these studies are reporting supportive effect on enhancing callus healing, further high-level studies in human subjects are necessary before we can recommend PEMF as a valid treatment option.

Box 28.2 Biologic Stimulants

- Local application
  - BMP
  - BMAC/PRP
  - Stem cells
  - Experimental—TP508, ED71, FGF-2
- Systemic application
  - Parathyroid hormone
  - Growth hormone
  - Experimental—hyperbaric oxygen, androgens

Biologic Stimulation

Local Application-Bone Morphogenetic Protein

The BMPs are known to have a strong osteoinductive potential [59–61]. There are several preclinical studies on the effect of BMPs on distraction osteogenesis [5, 62–65]. Mizumoto et al. [65] used a single injection of rhBMP-7 into osteotomy gap on the day of surgery in a rat DO model and observed a larger amount of new bone throughout the distraction phase. Hamdy et al. showed that application of OP-1 early during the distraction phase in rabbit tibiae accelerated new bone formation by densitometry and micro CT analysis [62, 64]. Also, Song et al. [5] demonstrated enhanced bone formation by injection of BMP-2 at the end of distraction in rat femora. Several clinical studies have been reported using BMP-2 or BMP-7/OP-1 for open tibial fractures and spinal fusion [66, 67], but not for distraction osteogenesis so far. There are conflicting issues about the risk of cancer by using rhBMP-2 [68–73]. TGF-beta ligand is known to have the ability to alter cell signaling that can lead to tumor promotion, and certain BMPs are expressed during the growth of some types of cancer [68, 69, 71, 73–76]. Further investigations for optimal dose, optimal timing, and potential risk of various BMPs are required before they can be recommended for clinical use.

Bone Marrow Cells/Platelet-Rich Plasma

Osteogenic differentiation from bone marrow-derived mesenchymal stem cells (MSC) has several advantages when compared to the differentiation of other mesenchyme tissues [77]. First, bone marrow cells (BMCs) have abundant mononuclear cells. Second, the differentiation of osteoblasts from BMCs is well described and standardized. Third, due to the autogenous properties the complications such as infection, immunogenic reactions, and disease transmissions can be minimized. Fourth, the harvesting process of bone marrow cells using a Jamshidi vacuum aspiration is minimally invasive and has low donor-site morbidity. Lastly, we are not aware of any reports of malignant transformation of autologous BMCs in the literature [77]. Though a lot of animal studies using bone marrow transplant have shown that bone marrow has enriched osteoblast progenitor cells and it is a valuable bone graft in bone defect, there have been only limited investigations on its application in distraction osteogenesis [78, 79].

The PRP is abundant in various growth factors released from platelets, which have been known to increase vascular ingrowth and mitogenic effects on bone-forming cells [80–83]. Kawasumi et al. [82] observed that a high platelet concentration combined with osteoblastic cells in PRP accelerated new bone formation during distraction osteogenesis in a rat model. In contrast to these findings, Arpornmaeklong et al. [84] noted an inhibitory effect of PRP on osteogenic differentiation of marrow-derived pre-osteoblasts.

There have been few clinical studies using PRP in human subjects [85–87]. Latalski et al. [85] reported a shorter healing index in the PRP group in a retrospective human DO study with a small sample size. The role of PRP in bone regeneration is still a controversial issue and its mechanism is not yet fully understood [84, 85, 88–98]. BMCs and PRP have been used simultaneously showing favorable clinical results. Kitoh et al. [78, 99] studied transplantation of culture-expanded bone marrow cells combined with PRP that accelerated new bone formation during the distraction osteogenesis, especially in the femur.

Lee et al. [86] showed that the combined injection of bone marrow aspirate concentrate (BMAC) and PRP during surgery enhanced bone regeneration in human tibial distraction osteogenesis (Fig. 28.2). The difference in the abovementioned studies conducted by Kitoh et al. and Lee et al. lies in how to process BMC-culture expansion or concentration. Jäger et al. [77] described the strength of the concentration compared with culture-expanded method. An immediate autologous transplantation of bone marrow concentrate can prevent complications related to the reduced quality of the transplanted cells such as pre-aging due to telomere shortening, reduced viability, or dedifferentiation/reprogramming that is associated with in vitro cultivation. The risk of infection is reduced by decreasing the ex vivo time period; and injection can be performed by single-stage surgery. Although there is only limited clinical evidence, BMAC and/or PRP seems to be a promising treatment option. Though all these studies show a favorable prophylactic role of BMAC/PRP in enhancing the regenerate in DO, their role in established delayed/nonunion after DO is uncertain and warrant further studies.

Well-designed preclinical and clinical studies are necessary, especially to evaluate whether BMAC or PRP contributes more to bone formation—or if the effect is synergistic, the optimal timing of injections and the optimal concentrations and cell counts.

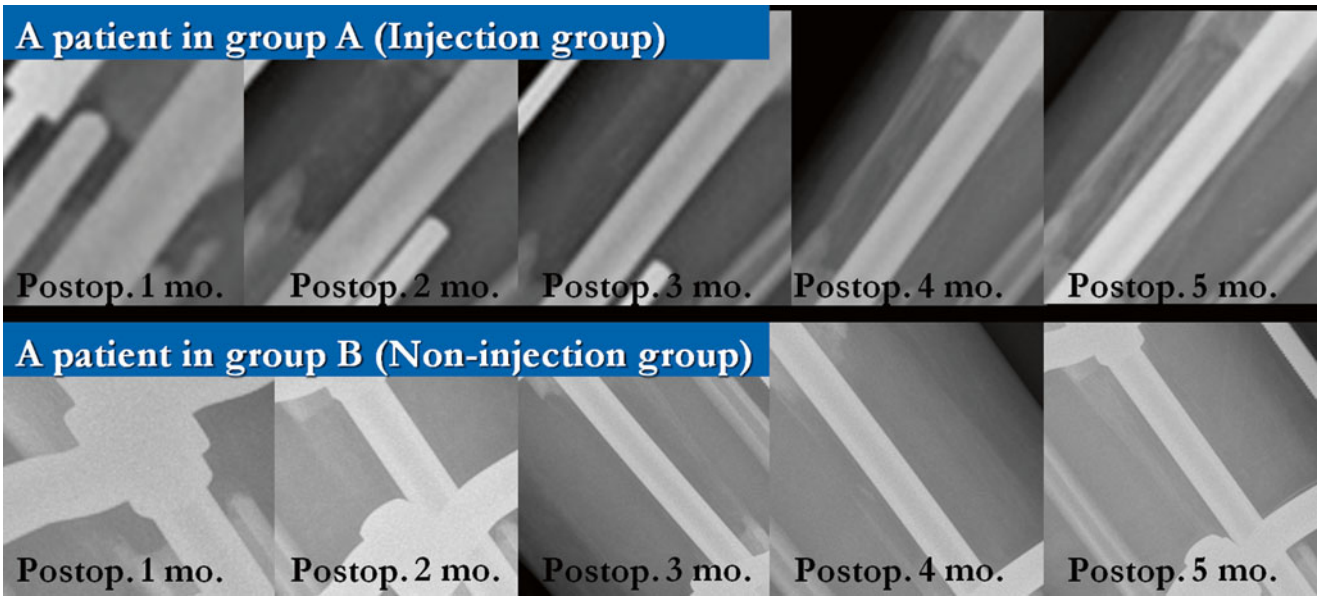


Fig. 28.2 Serial anteroposterior plain radiographs (postoperative 1–5 months, monthly) of the tibia in a patient undergoing lengthening over nail. More callus regeneration was seen in the injection group (group A). Images in group A were obtained from a patient in an injection group, showing postoperative 1- to 5-month states with an increment of 1 month. Images in group B (non-injection group) were from a patient in a control group with the same postoperative months above

Stem Cells

There have been many debates about using biotechnology on fracture healing but there is little scientific evidence about the positive effect of tissue engineering on osteogenesis. Tissue engineering combines osteogenic bone marrow mesenchymal stem cells, synthetic scaffolds, and growth factors in order to form hybrid constructs. Stem cells have been used for other indications including the treatment of long bone defects [100], fracture healing [101], and avascular necrosis [102] apart from enhancing the regenerate in distraction osteogenesis.

Stem Cells

Stem cells can be of two types:

1. Bone marrow-derived mesenchymal stem cells (MSC)
2. Osteogenic differentiated progenitor cells

Bone marrow-derived mesenchymal stem cells (BMC) can be directed towards the osteogenic lineage if cultured with osteogenic supplements like dexamethasone, β-glycerophosphate, and ascorbic acid phosphate [103]. In a study done by Peters et al. [104] in rats using histomorphometric analysis, it has been shown that locally applied osteogenic differentiated progenitor cells are more effective than mesenchymal stem cells in enhancing bone healing.

Growth Factors

Growth factors may be added to improve the results of stem cell therapy. There are several osteoinductive growth factors including platelet-derived growth factor, insulin-like growth factor, and transforming growth factors. PRP contains all these growth factors [105]. PRP can also be a suitable carrier for cell transplantation because it coagulates immediately by an addition of thrombin and calcium. Combination of stem cells and PRP accelerates bone healing and bone remodeling process by stimulating angiogenesis [106].

In a study conducted by Kitoh et al. [99] in 46 patients of skeletal dysplasia and limb length discrepancy, the group that received transplantation of osteogenic differentiated stem cells (approx. 2.3×10^7 cells) and PRP around 3 weeks after the lengthening surgery had a shorter healing index of approx. 31 days/cm compared with the control group which had an average healing index of 55 days/cm and also had reduced associated complications.

Scaffolds

Stem cells may need a scaffold for osteoconduction. The 3-D structure of a scaffold determines the extent of cell migration and differentiation, bone ingrowth, vascularization, and mass transfer between the cells and the environment. All of these processes benefit from increased scaffold porosity, interconnected pore networks, large surface area-to-volume ratio and increased surface roughness [107]. Bioactive scaffolds aid in bone bonding. It is important for scaffolds to interact with surrounding tissues to induce specific cellular responses [108].

Box 28.3 Properties of an Ideal Scaffold

- Increased and interconnected porosity
- Large surface area-to-volume ratio
- Same mechanical strength as bone
- Bioactive
- Biocompatible
- Biodegradable
- Optimal release profile

One method to achieve this goal is to construct the scaffold with biomaterial that can stimulate platelets to release growth factors. For instance, collagen is known to be as effective as thrombin in activating platelets [109]. The activated platelets would then release growth factors, which will enhance tissue regeneration, proliferation, and differentiation. Therefore, a scaffold made with collagen type 1 will likely be effective in bone formation and bonding.

The mechanical strength of a scaffold should approximate that of bone. Weaker scaffolds cannot support the skeleton, whereas stiffer scaffolds lead to stress shielding. The carrier should be biocompatible to minimize interference with bone induction. It must be biodegradable to minimize the effects on the biomechanical properties of the regenerate, and yet it must persist in vivo long enough to maintain bioactive elements at the site of implantation and optimize their release [110].

Scaffolds available for use with stem cells include PRP gel carrier, collagen, hydroxyapatite (HA) ceramic scaffold, resorbable polylactide membrane, and polycaprolactone (PCL). Collagen scaffold seems to be a good delivery vehicle for other graft materials [110]. Maracci et al. [100] have demonstrated that extensive long bone defects in human subjects treated with stem cell-seeded HA scaffolds have healed and integrated well with the host bone. Even after a 7-year follow-up the durability of the regenerated bone was good and there were no long-term complications noted.

Other Uses of Stem Cells

A case of avascular necrosis of femoral head being treated effectively by stem cells has been reported by Kim et al. [102]. Lee et al. [111] have reported successful reconstruction of 15-cm segmental defects by bone marrow stem cells and autogenous bone graft in central hemangioma of mandible. Trials are also being carried out by using stem cells in osteoporotic fractures [112]. "Osteogenic matrix coating," a method to prevent loosening of joint prostheses by coating them with osteogenic cells or their precursors, has been proposed by Ohgushi and Caplan [113].

Osteogenic Progenitor Stem Cell Culture Technique

Our technique for procuring and processing the bone marrow aspirate and culture of osteogenic progenitor stem cells is explained below [101].

Approximately 3–5 ml of bone marrow is collected from the anterosuperior iliac spine and added to a container filled with 30 ml of 10 % FBS-α MEM and 350 units of heparin (Fig. 28.3). The mixture is then taken to the lab and centrifuged at 4 °C for 10 min, after which the supernatant was discarded and 20 ml of culture medium was added to the remaining pellets. The mixture is then filtered, 10 ml of the medium is added per T-75 culture flask, and the culture is initiated. The incubator is maintained at 37 °C with 5 % CO_2. The next day, 50 μg L-ascorbic acid/10 ml and dexamethasone 10^{-7} M are added to facilitate cell differentiation into osteoblasts. The cell culture condition is evaluated using a light microscope, and the culture medium is changed on the fifth day of culture, after which the culture medium is changed every 3 days with the subsequent addition of L-ascorbic acid. On the 14th day of culture, nitro blue tetrazolium chloride 5-bromo-4-chloro-3-indolyl phosphate (NBTBCIP) staining was performed to confirm activation of the alkaline phosphatase. Twenty-four days after beginning the culture, Alizarin red staining is performed to detect newly produced calcium, and thus confirming that most of the cultured cells are osteoblasts. 1 vial (0.4 ml) contains autologous cultured osteoblasts over 12 million. Every patient may need 1–6 vials (12–72 million cells).

Injection Technique (Fig. 28.4)

In the non-sterile area, transfer whole amount of the cell suspension in two vials into the mixing vial using an 18-G needle. In the sterile area, transfer only 0.1–0.2 ml of thrombin into the mixing vial and discard the rest of thrombin. Mix stem cell and thrombin in a mixing vial and transfer 1 ml into red (thrombin) syringe. Mix 1 ml of collagen and 1 ml of fibrinogen with two 3-ml syringes using a three-way connector (Fig. 28.5). Transfer 1 ml of mixed collagen and fibrinogen into blue (fibrinogen) syringe. Install the red syringe and blue syringe into Y-piece and implant stem cells with injector and spinal needle by checking the defect site through C-arm (Fig. 28.6). After injection, this becomes a gel within 3–5 min.

Although the initial results are encouraging, further studies are necessary to assess the safety and efficacy of stem cells in distraction osteogenesis.

Other Experimental Local Stimuli

Osteogenic peptides are cheaper to produce as they are synthetic. They are less likely to lose their bioactivity during storage and delivery because of their short, linear structure.

TP508

One of these osteogenic peptides is TP508, which includes a 23-amino acid peptide that is the non-proteolytic receptor-binding domain of human thrombin. The effects of TP508

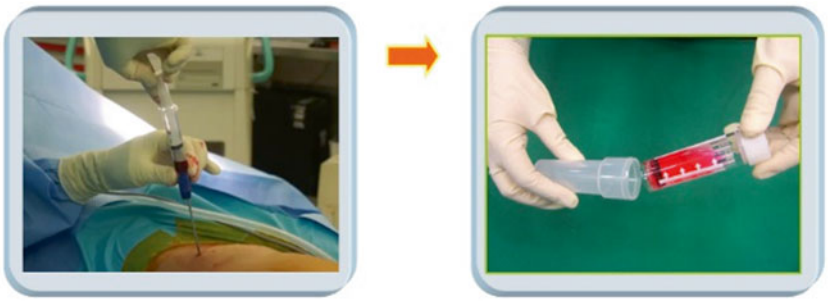


Fig. 28.3 Collection of bone marrow

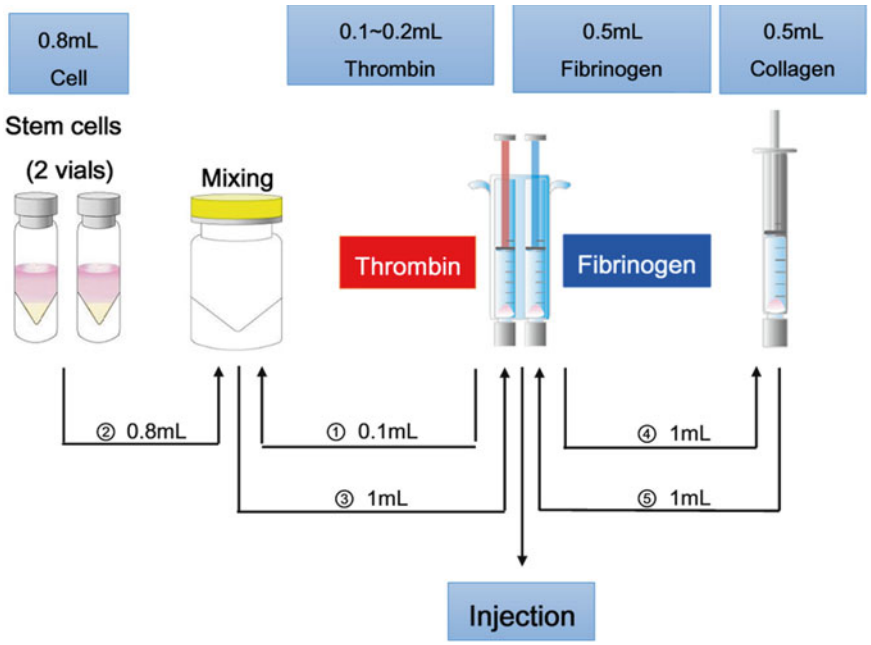


Fig. 28.4 Flowchart depicting the preparation of stem cell mixture

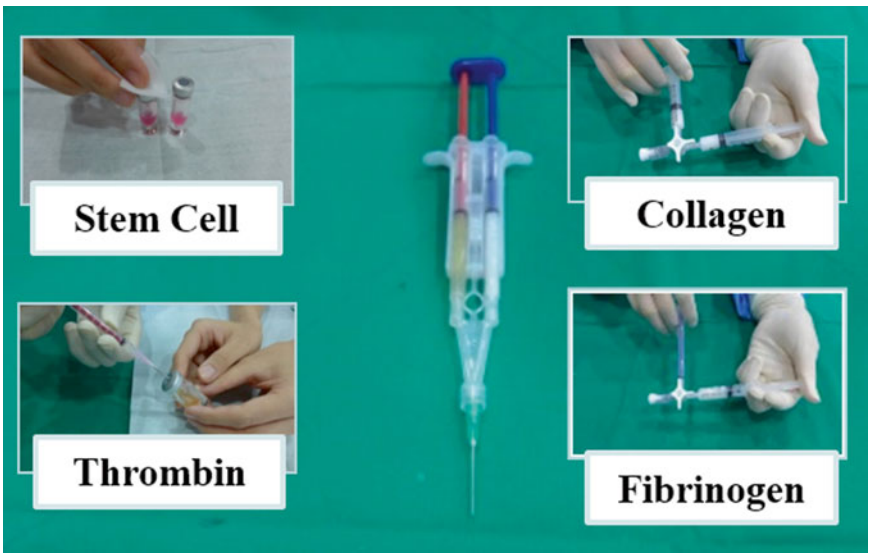


Fig. 28.5 Using a three-way connector, stem cell and thrombin are mixed in a red syringe. Similarly, collagen and fibrinogen are mixed in a blue syringe. Then both of these are attached to the Y-piece

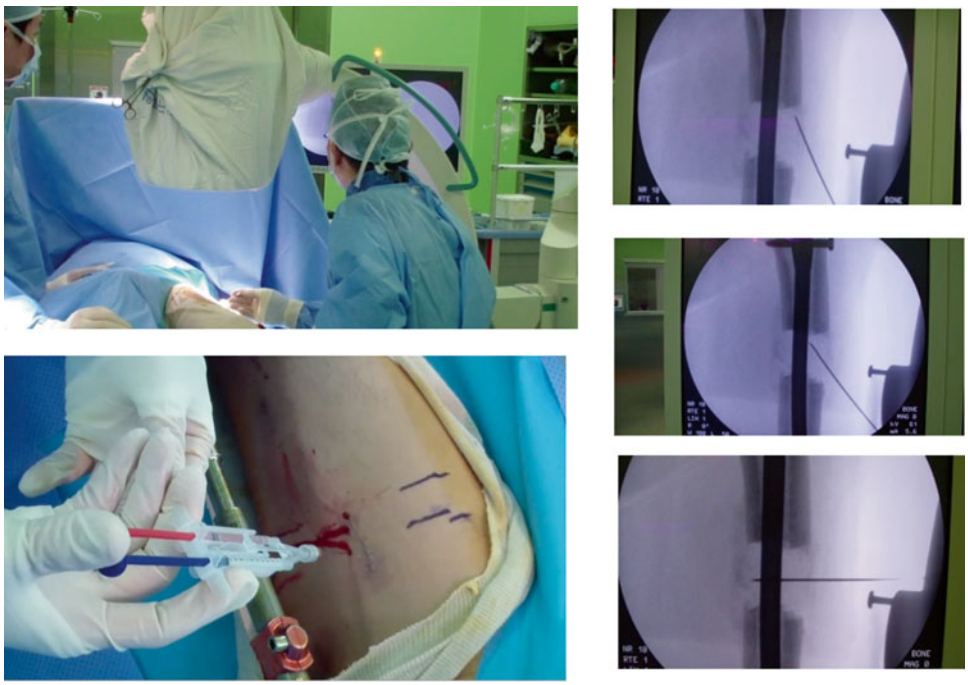


Fig. 28.6 Injection of the stem cell preparation under C-arm control

include changes in the inflammatory response, improvements in cell recruitment, and angiogenesis [114]. The doses of TP508 used in the experimental studies vary from 0.1 μg to 300 μg, and results differ depending on the dose. The animals given the higher dose had a larger area of newly formed bone [115–117].

ED-71

Yamamoto [118] reported that administration of 2-β-(3-hydroxypropoxy)-1α, 25-dihydroxyvitamin D3 (ED-71), an analog of synthetic vitamin D3, increased the bone mineral content (BMC) at the lengthened callus. According to Yamane et al. [119] ED-71 promoted both bone matrix formation and mineralization of the callus at 1 week after completion of lengthening, during which BMC increased markedly and may be effective for shortening the duration of therapy in callus distraction.

FGF-2

Fibroblast growth factor-2 (FGF-2 or basic FGF) is recognized as a potent mitogen for a variety of mesenchymal cells. In skeletal tissues, FGF-2 is produced by cells of osteoblastic lineage, accumulated in bone matrix, and acts as an autocrine/paracrine factor for bone cells [120]. FGF-2 shows variable regulations of proliferation and differentiation of cells of osteoblastic lineage and modulates bone formation. Aronson [121] has reported that the age-related deficits of endosteal bone formation were reversed by administering exogenous rhFGF-2 in an animal study. In another study Okazaki et al. [120] have reported that a single local injection of FGF-2 at the center of distracted callus facilitated the consolidation during bone lengthening in rabbit tibiae even under strenuous conditions of rapid and long distraction. In a study conducted by Jiang et al. [122], osteodistraction was applied in craniofacial bone of rabbits to observe the effects of MSCs with or without bFGF gene transfected on bone regeneration. They noted that excellent bone formation and highest BMD and BMC were achieved in the bFGF gene transfected group.

Optimal Timing of Biologic Stimulants

There is no clear evidence to guide us in terms of the timing of injection to achieve better bone regeneration. Angiogenesis has been shown to be of vital importance and intricately involved in the inflammatory response, soft callus formation, and transition from cartilaginous callus to bone [123, 124]. Proangiogenic factors such as vascular endothelial growth factor (VEGF) are especially important during early phase of distraction, when their expression is significantly higher than in later periods [125]. So, administration of growth factors during the early phases may stimulate neoangiogenesis and/or enhance the formation of regenerate bone. Several preclinical studies showed enhanced new bone formation by applying BMPs on the day of the surgery [65, 126, 127] and immunohistochemical analysis has shown high expression of growth factor receptors during the distraction phase [82, 125, 128–131]. In a randomized controlled trial, Lee et al. [86] clinically demonstrated that injection of BMAC and PRP on the day of surgery enhanced new bone formation (Fig. 28.7).

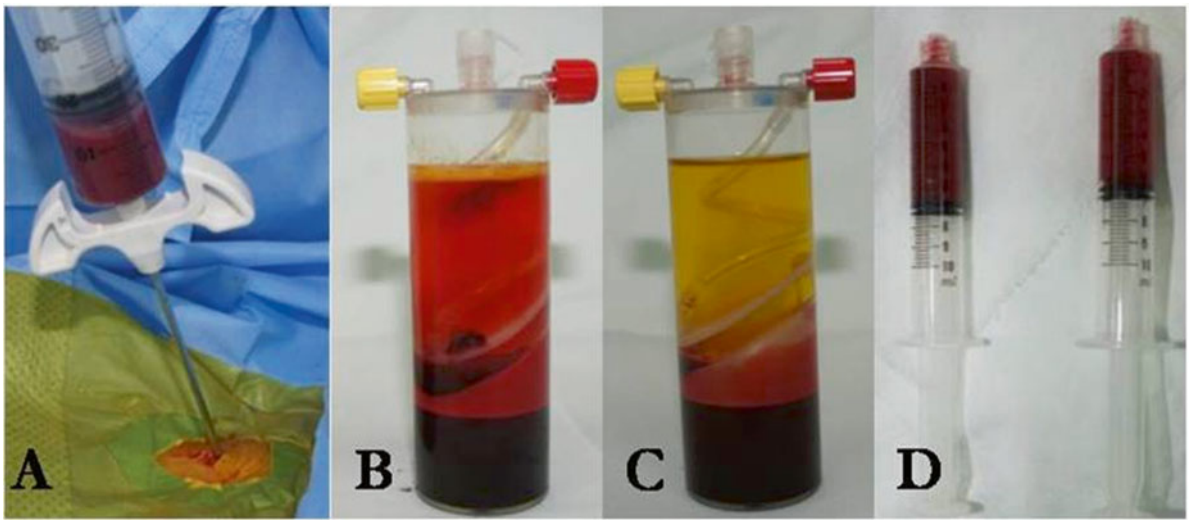


Fig. 28.7 BMAC and PRP are prepared in the operation room at the end of index surgery: 60 ml of bone marrow is aspirated at the iliac crest (**a**) and 60 ml of peripheral blood is obtained. Bone marrow and peripheral blood are centrifuged to make BMAC (**b**) and PRP (**c**). Six milliliters of the aspirate sampled from the mid-layer are prepared for the injection (**d**)

Hamdy and Rauch et al. [62, 132] observed high levels of expression of BMP-2, 4 and OP-1 during the distraction phase, with a rapid decline thereafter. They hypothesized that the best time for exogenous injection of BMPs would be at the end of the distraction phase, when the endogenous expression of BMPs is decreasing. But, they could not observe enhanced bone healing when OP-1 is injected at the end of distraction, but found twofold increase in bone volume when injected during early phase of distraction [62, 64]. There are also several reports of enhanced healing when applied at the end of distraction [85, 133, 134]. The optimal timing of the injection of the biological stimulants is an important but still unresolved issue. Hence further well-designed investigations are required.

Biological Stimulation: Systemic Application

Parathyroid Hormone

Parathyroid hormone (PTH) is one of the most promising therapeutic agents for osteoporosis. Human and animal studies have demonstrated that daily systemic injection of parathyroid hormone increases BMD [135–138]. PTH stimulates bone formation and resorption and can increase or decrease bone mass, depending on the mode of administration. Continuous infusions and daily subcutaneous injections of PTH stimulate bone formation similarly but have different effects on bone resorption and bone mass [139, 140]. It has been reported that the combination of systemic and local parathyroid hormone led to higher BMD, BMC, and bone area, a trend for greater radiographic-detected bone area and higher expression of osteocalcin in osteotomy sites when compared with the individual treatment or control groups [141]. Skripitz et al. [142] conjectured that since PTH seemed to have a greater effect on new bone formation than on normal bone remodeling, it might become useful for improving the incorporation of orthopedic implants and stimulating fracture repair. Neer et al. [143] reported that the clinical benefits of PTH reflect its ability to stimulate bone formation and thereby increase bone mass and strength. Given the enhancement of bone formation by the local stimulator such as BMP and the systemic application of PTH, combination therapies may be beneficial for bone tissue engineering.

The mechanism of this anabolic effect, however, has not been well established. It is widely believed that the anabolic effect of PTH may be due to increased osteoblast differentiation. Other report indicated that prevention of osteoblast apoptosis is a crucial mechanism for the anabolic effects of PTH on bone [144].

Growth Hormone

Systemic administration of recombinant homologous growth hormone (GH) has been reported to greatly accelerate ossification of bone regenerate in distraction osteogenesis [145]. GH administration increases bone turnover by enhancing both bone formation and resorption [146]. Correspondingly, an increase in the mechanical strength of the whole bone occurs, whereas the mechanical quality of the osseous tissue is equivalent in GH-injected animals and controls [7]. Various studies demonstrate that systemic administration of

GH is able to enhance bone regeneration and mechanical strength of healing critical size bone defects in animal models [147–149].

Other Experimental Systemic Stimuli

Hyperbaric oxygen therapy has been used to treat a variety of diseases and has been described as helping patients who have delayed healing or bone defects [150]. This technology consists of intermittently administering 100 % oxygen at pressures greater than one atmosphere absolute entirely enclosed in a pressure chamber [150]. However, one systematic review [151] performed recently failed to locate any relevant clinical evidence to support or refute the effectiveness of hyperbaric oxygen therapy for the management of delayed union or established nonunion of bony fractures. Good-quality clinical trials are needed to define its role in DO.

Androgens such as testosterone are also known to have proliferative effects on osteoblasts and increase fracture healing by systemic and local stimulation of bone formation [152]. In clinical application, androgens may be a possibility to increase bone formation, especially in elderly patients. Furthermore, it may be possible to shorten postoperative rehabilitation because of the effects of androgens on muscles [153].

Anticatabolic Therapy: Systemic Application

Bisphosphonate

The potential anabolic effect of bisphosphonates has been reported [154, 155]. Recent advances in the pharmacological treatment of osteoporosis with bisphosphonates [156–158] have encouraged surgeons to explore their use in limb lengthening. In general, bisphosphonates have been considered antiresorptive agents with limited effect on osteoblasts [159]. The anabolic effect of bisphosphonates has been questioned as they are not internalized by osteoblasts, and therefore, they may not exert a direct effect on osteoblast activity. However, there is evidence to support that there is an effect of bisphosphonates on the osteoblast side not directly on mature osteoblasts but more likely on osteoblast precursors in the bone marrow. First, only nanomolar concentrations of bisphosphonates are needed to stimulate the production of osteoclast inhibitory factor, which is mostly secreted by osteoblast precursors [160]. Second, bisphosphonates have shown to stimulate fibroblastic colony formation by murine and human bone marrow both in in vitro and ex vivo cultures [161].

The use of external fixators or any form of fixation may be associated with stress shielding of the spanned bone interval in this case, the regenerate and the surrounding bone resulting in osteopenia. Eyres et al. [15] noted a mean decrease in BMD of 44 % in the tibia and 61 % in the femur distal to the regenerate in children undergoing limb lengthening. Maffulli et al. [162] noted that the BMC adjacent to the regenerate decreased to less than 40 % of its original value in 6 of 11 patients. There was a reversal of osteopenia, increased regenerate volume, and increased mechanical strength of the bone when bisphosphonates were administered. However, healing index remained very prolonged [163].

Im et al. [164] have shown that both risedronate and alendronate could increase osteoblast and osteoblast progenitor numbers in primary human trabecular cultures with enhanced expression of BMP-2, type I collagen, and osteocalcin. These effects are not limited to osteoblastogenesis but also through the promotion of osteoblast survival as per their efficacy as anti-apoptotic agents [165]. Furthermore, bisphosphonates were reported to enhance bone marrow mesenchymal stem cell osteogenic differentiation [166] and facilitate human adipose-derived stem cell osteogenesis for bone regeneration [167]. Not only bone formation but also bone resorption are highly activated in the regenerated bone, implying high bone turnover. Sufficient nitrogen-containing bisphosphonate (N-BP) causes a notable modulation in morphological properties of the regenerated bone through inhibition of highly activated bone resorption and eventually increased mechanical properties [168]. The main effects of N-BP are at the lumbar spine and proximal femur, where they stop bone loss, reduce fracture risk, and increase BMD [169]. In addition, N-BP are prescribed for the treatment of bone diseases such as osteoporosis, multiple myeloma, cancer metastases, and Paget's disease.

However, the conventional systemic way of administration of bisphosphonates may have some undesirable side effects such as gastrointestinal ulceration and jaw osteonecrosis [169, 170]. Recent studies have indicated that local administration of a low dose of bisphosphonates was able to enhance bone regeneration in vivo [171–173].

Other Experimental Anticatabolic Therapies

The influence of calcitonin on various stages of bone formation has been investigated [174]. Weiss et al. [175] reported that when calcitonin is administered during the initial phases of bone formation, it increases bone formation due to a stimulation of proliferation of cartilage and bone precursor cells. In humans, serum calcitonin rises during pregnancy, growth, and lactation [176]. It is during these periods of calcium stress that a tonic antiresorptive hormone will best exert its effect to limit skeletal loss. This may be the primary role of calcitonin in skeletal conservation [177]. There is considerable support for the thesis that, in addition to its inhibitory effects on bone resorption, calcitonin enhances osteoblastic bone formation both in vitro and in vivo [178]. The putative

anabolic osteoblastic effect of calcitonin may be of clinical importance in sustaining the bone formation rate despite inhibition of bone resorption.

Summary

Insufficient bone regeneration is an unpredictable and unsolved issue in distraction osteogenesis. Avoiding the detrimental factors is also important, but there are unavoidable factors like host-related causes including smoking, diabetes, and systemic illness. Biologic stimulation is more promising because it can be a concept of preemptive stimulation rather than the treatment after poor bone regeneration is observed. Efforts to improve regenerate bone formation are still in progress. The efficacy, safety, optimal dose, timing and route of administration, and cost-effectiveness of each of the agents that can enhance bone formation in distraction osteogenesis should be studied further by well-designed investigations.

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Residual Deformities of the Hip

29

Shawn C. Standard and Daniel K. Ruggles

Introduction

There are many causes of hip deformity in the growing child. Although the etiologies are numerous, the resulting deformities can be categorized anatomically by several basic descriptive terms. Angular deformities of the hip in the coronal plane result in coxa valga or coxa vara. Similarly, angular deformities in the sagittal plane are described as flexion or extension deformities. Axial plane torsional deformities are either external with retroversion of the hip or internal with anteversion of the hip. These six basic deformity terms; varus, valgus, flexion, extension, internal or external torsion can be applied to other joints and long bones. However, alteration of the hip joint's unique configuration creates deformities specific to this joint. These deformities are coxa breva, coxa magna, and acetabular deformities, such as dysplasia, retroversion, and protrusio (Box 29.1).

Coxa breva is defined as shortening and widening of the femoral neck, which alters the relationship of the tip of the greater trochanter to the center of the femoral head (Fig. 29.1). This creates a mechanical disadvantage of the hip joint by decreasing the lever arm of the hip and decreasing the distance between the origin and insertion points of the gluteus medius muscle [1, 2]. This deformity is caused by a growth disturbance of the proximal femoral physis. The most common causes of the abnormal growth are avascular necrosis (AVN) of the proximal femoral epiphysis, osteomyelitis of the proximal femur, and abnormal bone development that occurs in the various skeletal dysplasias.

Coxa magna is defined as the enlargement and deformation of the femoral head. This deformity can cause loss of congruency between the femoral head and acetabulum resulting in femoral-acetabular impingement (FAI) (Fig. 29.2a, b). Severe coxa magna is associated with chronic inflammation, loss of motion, labral pathology, chondromalacia, and eventual degeneration of the hip joint [3–6]. Coxa magna is caused by a combination of the collapse of the proximal femoral epiphysis and altered growth of the articular cartilage. The immature femoral head's articular cartilage is a true growth cartilage, which allows the femoral head to grow in a circumferential fashion [7]. The alteration of this growth due to abnormal joint pressures from joint contractures along with concurrent collapse of the epiphysis can result in a coxa magna deformity. This deformity is caused by avascular necrosis of the proximal femoral epiphysis. The most common cause of proximal femoral epiphyseal AVN is Legg–Calvé–Perthes disease. Other causes of pediatric hip AVN include infection, sickle cell disease, significant steroid use, trauma of the proximal femur, and iatrogenic loss of proximal femoral blood supply [8].

Acetabular dysplasia is defined as an abnormal shape of the acetabulum with a decrease in depth, increased in circumference, and an increased slope in relation to the horizontal pelvic line [9]. This deformity results in an oblong-shaped, or "capacious" acetabulum, that allows lateral and proximal migration of the femoral head with concurrent reduction of joint coverage (Fig. 29.3). In this abnormal position, the contact surface between the articular cartilage of the femoral head and acetabulum is decreased. Consequently, there are increased and concentrated contact forces that ultimately increases wear rate of the articular cartilage.

Concurrently, the surgeon needs to recognize that a normal shaped femoral head must be present for an acetabulum to be defined as truly dysplastic. In other words, an oblong

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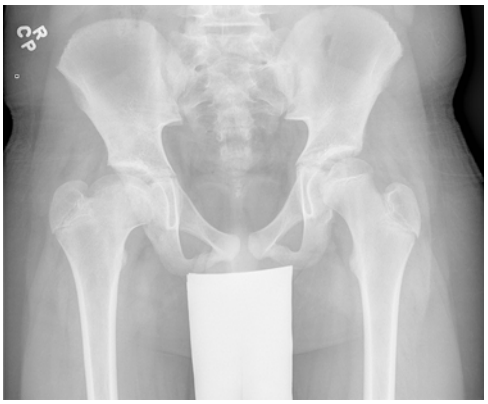


Fig. 29.1 Typical appearance of a coxa breva deformity demonstrated in this patient with a history of Perthes disease of the right hip. Reprinted with permission from the Rubin Institute for Advanced Orthopedics, Sinai Hospital of Baltimore

acetabulum with a congruent enlarged femoral head does not constitute acetabular dysplasia (Fig. 29.4). This situation is better described as coxa magna with a congruent oblong acetabulum, which can produce a functional, mobile hip joint in certain situations. The abnormal acetabulum with a concurrent abnormal femoral head must be carefully evaluated for congruency, impingement, and the area of force transfer of the articular surfaces.

The etiology of acetabular dysplasia is most commonly developmental in nature. However, other conditions resulting in chronic joint instability, such as Down's syndrome, or severe joint contractures, such as cerebral palsy, can result in hip subluxation with concurrent acetabular dysplasia [10, 11].

This chapter will concentrate on the residual hip deformities resulting from various etiologies including; developmental dysplasia of the hip (DDH), slipped capital femoral epiphysis, and Legg–Calvé–Perthes disease. Innovative treatment strategies are presented with detailed surgical techniques of six innovative procedures: triple pelvic osteotomy, modified Dunn procedure (with surgical hip dislocation), modified Southwick intertrochanteric osteotomy, Morscher femoral neck lengthening osteotomy, femoral head reduction osteotomy, and hip distraction with a hinged external fixator.

Box 29.1 Residual Deformities of the Hip

Defining key points:

- True *acetabular dysplasia* includes INCONGRUENCY of the hip joint
- The significance of *coxa breva* is determined by the center of femoral head to tip of greater trochanter distance
- *Coxa magna* is caused by avascular necrosis of the immature femoral head leading to abnormal and asymmetric growth.

Residual Deformities of the Hip Joint Secondary to Developmental Dysplasia

The early postnatal examination and screening for congenital dislocation of the hip has substantially decreased the incidence of patients presenting with late dislocations of the hip joint during infancy and early childhood [12, 13]. However, the stable hip with mild to moderate hip dysplasia that is silent during the early developmental years remains a challenge to detect with clinical exam alone. The primary treatment for hip dysplasia depends on the age of the child at diagnosis. The various treatment options range from simple observation or bracing to complex periacetabular osteotomies. The diagnosis of hip dysplasia for the older infant, toddler, and young child, is consistent with the radiographic Acetabular Index (AI) measurement >30° [14, 15]. Hip dysplasia for the older child, preadolescent, or adolescent is confirmed by the radiographic Center Edge Angle of <20° [16] (Box 29.2). Other radiographic signs of acetabular dysplasia are an abnormal sourcil, decreased femoral head coverage, and lateral/proximal migration of the femoral head, which collectively help to confirm and stratify the severity of the hip dysplasia [16].

Box 29.2. Radiographic Parameters of Acetabular Dysplasia

- Acetabular index >30°
- Center Edge Angle <20°
- Flattened/upsloped sourcil

Treatment of Residual Hip Dysplasia for Patients <6 Months of Age

The treatment for residual hip dysplasia without clinical instability for infants <6 months of age consists of application of a Pavlik harness or abduction bracing [17]. The harness is worn initially 16–18 h per day and then weaned to a nighttime brace within the first month of treatment. The night time bracing continues until the radiographic AI measurement is <30° and the sourcil is normal in appearance. After the hip has normalized based on such radiographic parameters, the bracing is discontinued and the patient is followed annually until the age of 5 years.

Treatment of Residual Hip Dysplasia for Patients >6 Months to 2 Years

The treatment for residual hip dysplasia for patients >6 months to 2 years of age consists of hip abduction bracing (Box 29.3). These older infants and toddlers are often too large and strong for the Pavlik harness, but a nighttime

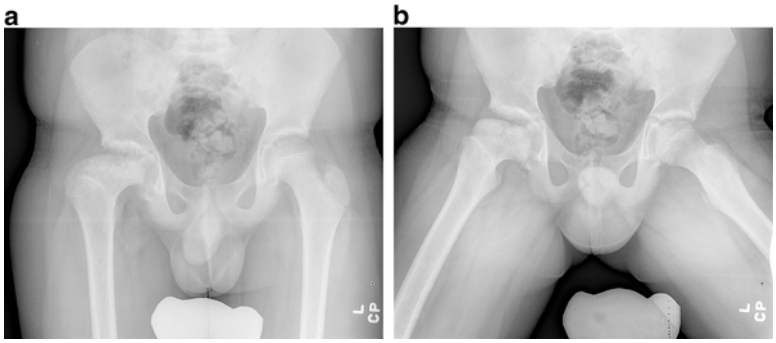


Fig. 29.2 A 12-year-old patient with severe coxa magna resulting from Perthes disease of the right hip as demonstrated on the AP (**a**) and lateral (**b**) views. Reprinted with permission from the Rubin Institute for Advanced Orthopedics, Sinai Hospital of Baltimore



Fig. 29.3 A 14-year-old female with Down's syndrome and bilateral acetabular dysplasia noted by the enlarged, oblong, and shallow acetabulum and concurrent femoral head migration. Reprinted with permission from the Rubin Institute for Advanced Orthopedics, Sinai Hospital of Baltimore

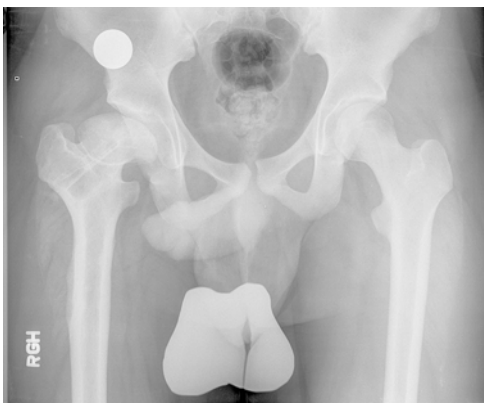


Fig. 29.4 An example of an enlarged and oblong acetabulum secondary to remodeling effects of a large femoral head after Perthes disease. This acetabulum should not be termed dysplastic due to the absence of a normal femoral head. In this situation, the right acetabulum is insufficient due to the decrease in force transfer noted by the reduced sourcil and patient's symptoms. Reprinted with permission from the Rubin Institute for Advanced Orthopedics, Sinai Hospital of Baltimore

abduction brace is very effective. The main difficulty with this treatment in the older toddler is the compliance with the orthosis. However, in the authors' experience, once the potential surgical option is discussed with the parents, the compliance with the bracing often improves. Despite limited published data, this simple orthotic treatment is common among pediatric orthopedists and can yield a high success rate with resolution of the hip dysplasia in the vast majority of patients if the brace is worn consistently. A recent study has shown that part-time abduction bracing significantly improves residual acetabular dysplasia at 6 months of age versus observation alone [18]. After the hip has normalized radiographically, the abduction brace is discontinued and the patient is followed annually at least until the age of 5 years.

Box 29.3. Nonoperative Treatment of Acetabular Dysplasia <2 Years of Age

- Pavlik harness for patients <6 months of age
- Simple hip abduction orthosis for patients >6 months of age
- Night time bracing until radiographic parameters and appearance normalizes

Treatment of Residual Hip Dysplasia for Patients >2 Years Until Closure of the Triradiate Growth Plate

Once the child is older than 2 years of age, the nighttime bracing becomes very difficult due to patient cooperation and compliance. However, in the older toddler, if the nighttime hip abduction brace could be tolerated, then this would be the initial treatment strategy as stated above for the younger child. For the majority of older toddlers and young children, the treatment for clinically significant hip dysplasia consists of a single cut pelvic osteotomy that hinges on the triradiate growth plate or the pubic symphysis (Box 29.4). The direction and severity of acetabular deficiency influences the exact type of pelvic osteotomy that is performed. The type and direction of coverage of the commonly used pelvic osteotomy is as follows: the Pemberton pelvic osteotomy primarily provides anterior coverage, the Dega pelvic osteotomy covers anterolaterally, and the San Diegoplasty covers laterally [10]. These three "volume-reducing" osteotomies are very similar but with distinct differences. All three

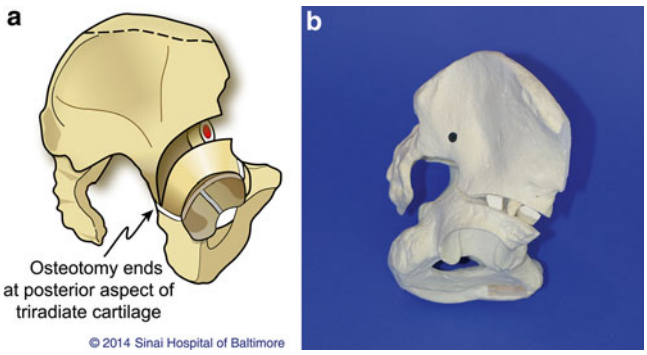


Fig. 29.5 The classic Dega osteotomy curves from the anterior inferior iliac spine to the triradiate cartilage posteriorly hinging on both the inner iliac table and the triradiate cartilage. This is demonstrated in both the schematic drawing (**a**) and bone model (**b**). Reprinted with permission from the Rubin Institute for Advanced Orthopedics, Sinai Hospital of Baltimore

are incomplete osteotomies, hinging on the triradiate cartilage and do not usually require internal fixation for stability.

The Dega osteotomy is the classic pelvic osteotomy that hinges on the triradiate growth cartilage [19, 20]. The Dega osteotomy begins at the anterior inferior iliac spine and curves posteriorly around the acetabulum to end at the triradiate cartilage. Medially, the Dega pelvic osteotomy is directed in a curvilinear fashion over the acetabulum towards the triradiate cartilage, leaving the inner table intact. Wedges of bone graft (autograft harvested from the iliac crest or the proximal femur when a shortening is being performed, versus allograft bone), can strategically be placed in the osteotomy site to provide a combination of increased anterior and/or lateral coverage (Fig. 29.5a, b).

Two modifications of the Dega-type osteotomy are the San Diegoplasty osteotomy and the Pemberton osteotomy (Fig. 29.6a–e). The San Diegoplasty osteotomy does not violate the inner table; however, a notch of bone is removed with a Kerrison rongeur at the anterior inferior iliac spine and the sciatic notch. This allows more coverage directed laterally without sacrificing posterior coverage, and was originally described for hip dysplasia associated with neuromuscular disease, which typically entails posterior acetabular deficiency [21] (see Fig. 29.6a–c).

The Pemberton osteotomy, first described in 1965, also hinges on the triradiate growth cartilage, but the direction of this osteotomy is from an anterior to posterior direction instead of a lateral to medial direction, as with the Dega and San Diegoplasty [22]. The Pemberton osteotomy begins at the anterior inferior iliac spine and curves over the acetabulum and ends at the triradiate growth cartilage posterior to the joint identical to the Dega osteotomy. However, the Pemberton osteotomy divides both the inner and outer tables of the ilium as it heads over the acetabulum to the triradiate cartilage. This creates a pure anterior opening wedge osteotomy with a greater increase of anterior coverage for the acetabulum (see Fig. 29.6d, e).

Another type of pelvic osteotomy that can be used in this age group is the Salter osteotomy that consists of a complete osteotomy from just above the anterior inferior iliac spine to the sciatic notch, including both inner and outer tables of the ilium [23]. The Salter is a redirectional osteotomy that hinges on the flexible symphysis pubis and requires internal fixation due to the amount of inherent instability. However, the Salter osteotomy affords greater movement of the acetabulum and thereby a greater ability to correct a more severe dysplasia. The surgeon must be aware that a dysplastic acetabulum with poor posterior coverage is a relative contraindication for a Salter osteotomy, as while it provides excellent anterolateral coverage, it can further uncover the femoral head posteriorly (Fig. 29.7a–f).

Postoperative care for innominate pelvic osteotomies requires non-weight bearing for approximately 6 weeks. The patients between the ages of 2 and 5 years are placed into a 1-½ hip spica cast for the initial 6 weeks, whereas the older patients (6 years and older) are often placed into a hip abduction pillow. After the initial 6-week period, the cast/pillow is discontinued and physiotherapy is started. The patient gradually returns to full weight bearing. Full activity is allowed after reestablishing full hip strength and range of motion. The patient is then followed annually until skeletal maturity.

Box 29.4. Acetabuloplasties in the Skeletally Immature

Choosing your osteotomy:

- Planning your osteotomy is determined by acetabular deficiencies specific to underlying etiologies (i.e., DDH = anterolateral, congenital femoral deficiency = lateral, neuromuscular = posterolateral).
- The Salter osteotomy can provide significant deformity correction, but requires internal fixation for stability.
- The Dega, San Diegoplasty, and Pemberton osteotomies do not require fixation routinely.
- Bone graft wedges can strategically be placed in a Dega osteotomy to provide more anterior, posterior, or lateral coverage.

Treatment of Residual Hip Dysplasia for Patients After Closure of the Triradiate Growth Plate

As the patient becomes older, the treatment for hip dysplasia becomes more complicated and invasive. However, unlike the asymptomatic younger child who is usually diagnosed incidentally and treated prophylactically, the older patient typically presents with mild to moderate pain associated with activity. Therefore, the treatment goals in this age group

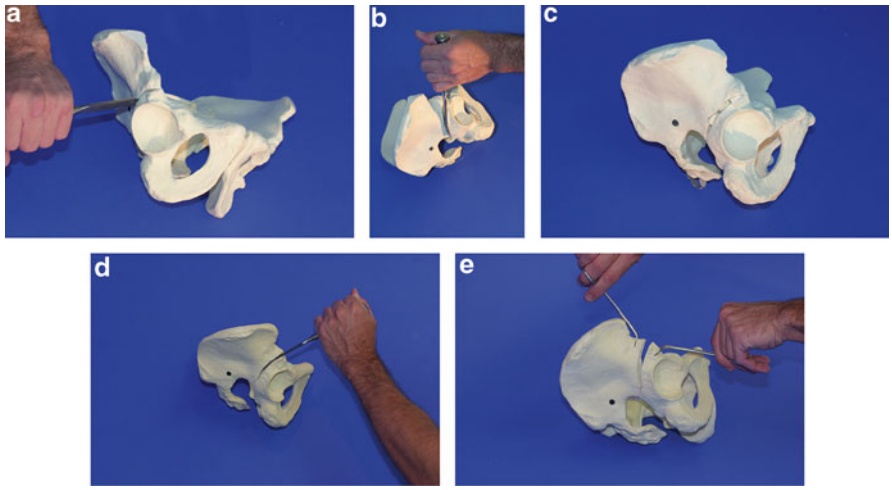


Fig. 29.6 (**a**–**c**) The bone model depicts the San Diegoplasty pelvic osteotomy that begins at the anterior inferior iliac spine (AIIS) and ends at the sciatic notch, hinging on the inner table of the ilium. (**d**, **e**) The bone model depicts the Pemberton osteotomy that is directed from anterior to posterior, beginning at the AIIS and ending at the triradiate cartilage behind the acetabulum. Unlike the Dega osteotomy, the Pemberton osteotomy divides both the inner and outer tables of the ilium. Reprinted with permission from the Rubin Institute for Advanced Orthopedics, Sinai Hospital of Baltimore

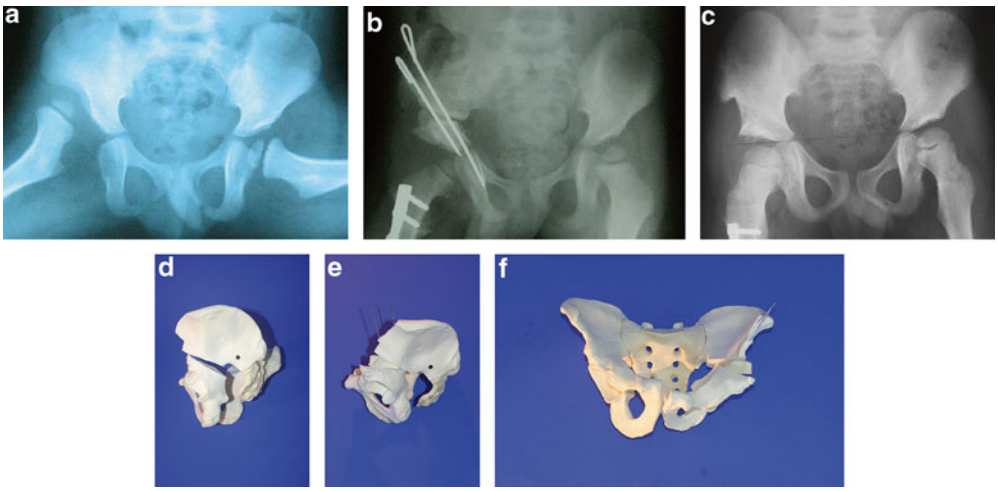


Fig. 29.7 (**a**) A 5-year-old male with R DDH. (**b**) Status-post open reduction, capsulorraphy, femoral shortening derotational osteotomy, and Salter innominate osteotomy. (**c**) Healed osteotomy with complete restoration of normal acetabular index, center-edge angle, and Shinton's line. (**d**–**f**) Bone models representing the Salter osteotomy. Reprinted with permission from Scott Nelson, MD

are the alleviation of pain, improved joint function, and prevention of early degenerative joint disease of the hip.

In the authors' practice, the preferred method of treatment for symptomatic residual hip dysplasia in the young patient after the closure of the triradiate cartilage is the periacetabular triple osteotomy (Box 29.5). The triple is a redirectional osteotomy including ischial, pubis, and supra-acetabular innominate bone cuts. An early technique described by Steel has continued to evolve [24]. Tonnis modified the procedure by performing the ischial cut in closer proximity to the acetabulum, facilitating acetabular fragment mobility and greater correction of severe acetabular dysplasia [25]. Originally described as two-incision procedure, further modifications of the triple osteotomy utilize a one-incision technique [26]. Because this is a complex osteotomy, a brief description of the technique will follow, highlighting certain technical tips.

The Ganz periacetabular osteotomy is another very popular type of pelvic osteotomy [27]. This particular osteotomy maintains the integrity of the posterior column by performing a bone cut connecting the iliac osteotomy to the partial ischial osteotomy. This osteotomy travels through a very narrow column of bone in the young patient. Therefore, the author prefers the periacetabular triple technique for adolescent and smaller adult patients

Periacetabular Triple Osteotomy of the Pelvis: Operative Technique

Positioning and Exposure

The patient is placed supine on the operating table with no bumps placed under the pelvis. The entire lower extremity, to include the groin, gluteal region, and subcostal area are prepared and isolated into the sterile field. A single incision is performed in the flexion crease of the hip joint, approximately 1 cm below the anterior superior iliac spine. This incision starts just medial to the palpable neurovascular bundle and extends 6–8 cm lateral to the anterior superior iliac spine (Fig. 29.8).

The entire operation is performed through this single anterior incision. The interval between the sartorius and tensor fascia lata muscles is defined in the typical fashion protecting the lateral femoral cutaneous nerve. The rectus femoris tendon is identified and released, tagging the insertional portion of tendon. Next, the iliac crest is exposed. Both the inner and the outer tables are subperiosteally dissected to the sciatic notch. Surgical gauze and/or bone wax may be used to control bleeding from the iliac wing. Dissection of the medial structures is subsequently performed. The fascia overlying the sartorius and iliopsoas muscles is divided. The iliopsoas tendon is identified (typically on the posterior aspect of the muscle) and fractionally lengthened within the muscle belly. Gentle retraction of the iliopsoas muscle belly brings the femoral nerve into the operative field. The femoral nerve should be identified prior to the division of the iliopsoas tendon. The femoral nerve, artery, and vein are carefully identified and exposed with individual small vessel loops placed around each structure. Fluoroscopic visualization of the superior pubic ramus is performed to confirm the osteotomy position just medial to the pubic eminence (Fig. 29.9). It is always surprising how medial the dissection must be carried out in order to place the superior pubic ramus osteotomy in the correct position. Typically, the osteotomy site lies between the femoral nerve and artery. However, with smaller patients, the osteotomy position can be medial to the femoral vein. The interval between the femoral nerve and artery is exposed down to the periosteum of the superior pubic ramus. Great care must be used when retracting the neurovascular bundle. If the interval for the exposure of the superior pubic ramus is performed lateral to the femoral nerve, the tension placed on the neurovascular structures will be substantially increased in order to reach the proper osteotomy position.

Osteotomies

After the exposure of the correct interval for the pubic ramus osteotomy is completed, the overlying periosteum is divided. Multiple drill holes are created in the superior pubic ramus using a smooth 1.8- to 2.0-mm wire. The osteotomy is completed with an osteotome. The interval for the ischial osteotomy is then created. This interval lies between the medial aspect of the hip capsule and the iliopsoas muscle. The interval is carefully created with gentle blunt dissection using a large curved Mayo scissor. The interval is opened and widened by gentle spreading of the blunt tipped scissor. A small Cobb elevator is placed into the interval to palpate the superior portion of the ischium, just below the acetabulum (Fig. 29.10). A Cobb elevator is advanced on both the medial and lateral sides of the ischium to push the soft tissues away and prepare a path for the Ganz osteotome (see Fig. 29.11d, e).

The Ganz osteotome is inserted into the prepared interval and positioned just under the acetabulum. It is critical to have adequate fluoroscopic visualization for this osteotomy. The osteotome is aimed approximately 45–55° proximally in order to direct the osteotomy above the ischial spine (Fig. 29.11a–e).

This osteotomy is visualized with an oblique fluoroscopic view, similar to an obturator Judet view (C-arm is rotated

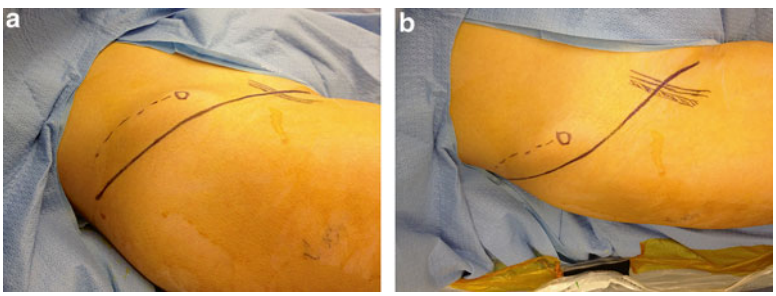


Fig. 29.8 (**a**, **b**) The single incision technique is depicted from an anterior and lateral view. The iliac crest is marked with the *dashed line* ending at the ASIS. The *longitudinal marked structures* are the palpable neurovascular bundle. Reprinted with permission from the Rubin Institute for Advanced Orthopedics, Sinai Hospital of Baltimore

toward the operative hip between 30° and 50° from the AP position). Again, fluoroscopic visualization of the acetabulum, ischium, and ischial spine is essential in this oblique view in order to complete the periacetabular ischial osteotomy successfully. Another critical technical tip is the protection of the femoral nerve while completing the ischial osteotomy. During this osteotomy, the hip must be flexed 15–20° to prevent stretch of the femoral nerve as the Ganz osteotome handle is levered distally in order to aim the osteotomy above the ischial spine. Before the final osteotomy of the ilium, two reference half pins are inserted into the acetabular fragment. These two reference half pins enhance the accuracy of the bony correction and help prevent secondary deformities (Fig. 29.12).

The first half pin is place from the anterior to posterior direction, just lateral to the superior pubic ramus osteotomy. This anterior reference half pin marks the acetabular version. The second half pin is inserted from lateral to medial in the supra-acetabular region, parallel to the roof or sourcil of the acetabulum. This lateral reference half pin marks the dysplastic acetabular roof. After the reference half pins are placed, the final osteotomy of the ilium is performed. A Gigli saw is carefully passed through the sciatic notch. The final osteotomy is completed with the Gigli saw from the sciatic notch to the anterior aspect of the ilium, just above the anterior inferior iliac spine.



Fig. 29.9 A 4-year-old female undergoing a triple osteotomy for right hip dysplasia. Intraoperative fluoroscopic image demonstrating the position for the superior pubic ramus cut. This position is usually directly behind or medial to the neurovascular bundle. Reprinted with permission from the Rubin Institute for Advanced Orthopedics, Sinai Hospital of Baltimore

Deformity Correction and Fixation

The acetabular fragment should become freely mobile after the third cone cut. If the fragment does not easily move, then the pubic ramus osteotomy and ischial osteotomy are revisited with repeat passage of the osteotome. After mobilizing the acetabular fragment, the reference half pins are used to reorient the acetabulum and temporary fixation is performed (Fig. 29.13a). The anterior half pin should still be pointed in a straight anterior to posterior direction if the version of the acetabulum was planned to remain the same. If anteversion is needed, then the anterior half pin should be internally rotated. Similarly, if the anterior half pin is externally rotated, retroversion has been imparted to the new acetabular position. The temporary fixation consists of three separate

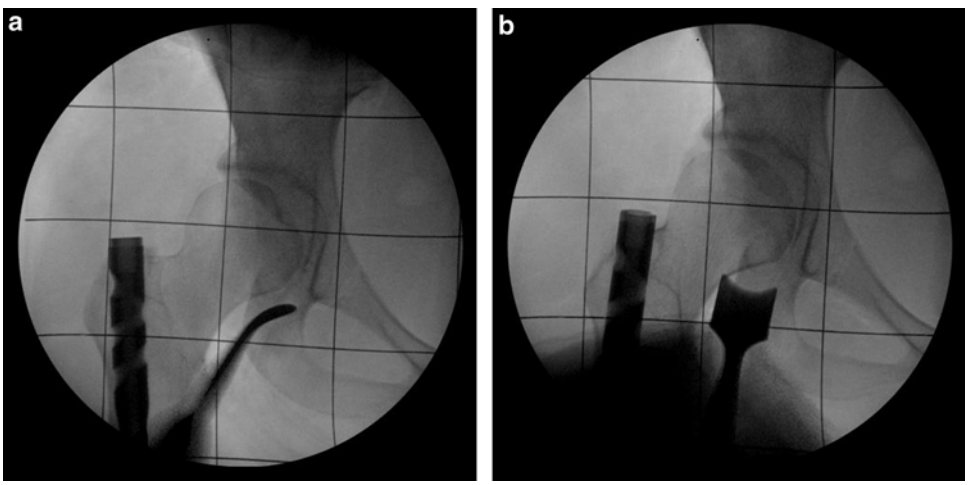


Fig. 29.10 A 16-year-old female undergoing a triple pelvic osteotomy for retroversion of the right hip. (**a**) Intraoperative fluoroscopic image of a Cobb instrument palpating the superior portion of the ischium just beneath the acetabulum. (**b**) Image demonstrating the placement of the Ganz osteotomy at same location as identified by the Cobb instrument. Reprinted with permission from the Rubin Institute for Advanced Orthopedics, Sinai Hospital of Baltimore

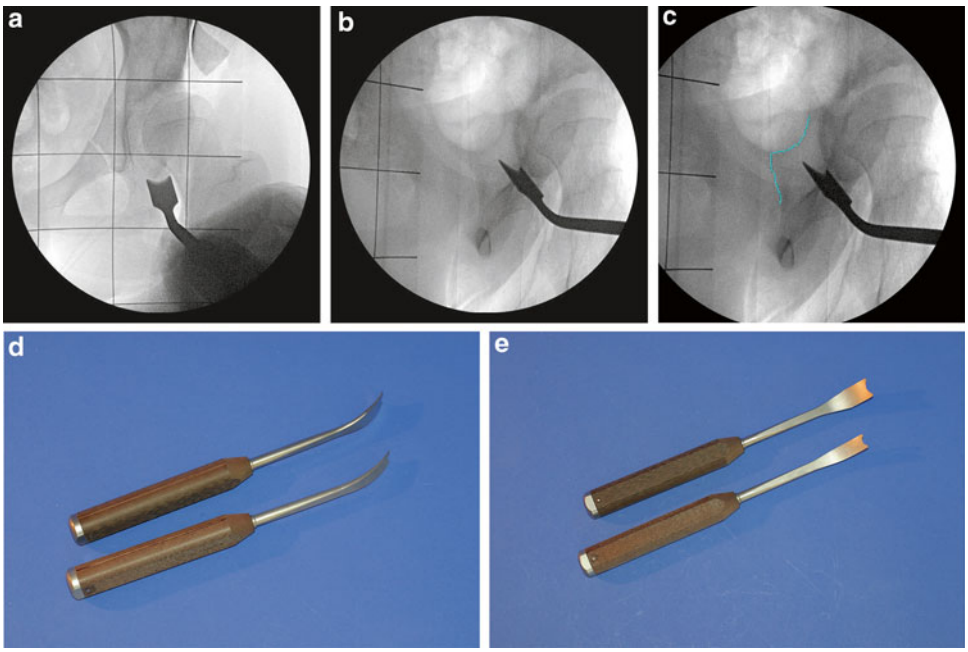


Fig. 29.11 (**a**) Fluoroscopic view denoting the position of the Ganz osteotome in the AP view preparing to perform the lateral cortical cut of the ischium. (**b**) Fluoroscopic view in the oblique position to confirm placement of the Ganz osteotome under the acetabulum and directed above the ischial spine. (**c**) Same fluoroscopic view as Fig. 29.11b with the ischial spine highlighted. (**d**, **e**) Clinical pictures of the specialized Ganz osteotomes. Reprinted with permission from the Rubin Institute for Advanced Orthopedics, Sinai Hospital of Baltimore



Fig. 29.12 Fluoroscopic view demonstrating the placement of the two reference half pins prior to the completion of the iliac osteotomy. The AP half pin marks the acetabular version whereas the lateral to medial half pin marks the slope of the acetabular roof. Reprinted with permission from the Rubin Institute for Advanced Orthopedics, Sinai Hospital of Baltimore

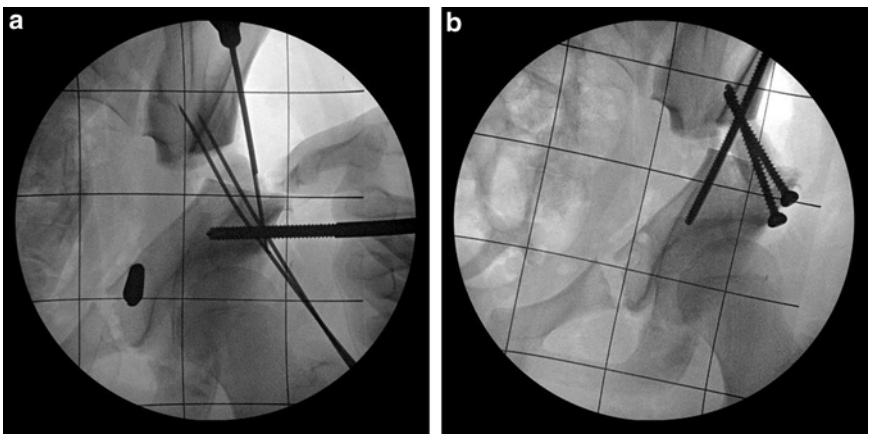


Fig. 29.13 (**a**) After the acetabular fragment is reduced into the corrected position, temporary fixation is achieved with smooth wires. (**b**) The smooth wires are sequentially overdrilled with a 3.2-mm cannulated drill and 4.5-mm solid screws are inserted. This is the same patient as Fig. 29.16. Reprinted with permission from the Rubin Institute for Advanced Orthopedics, Sinai Hospital of Baltimore

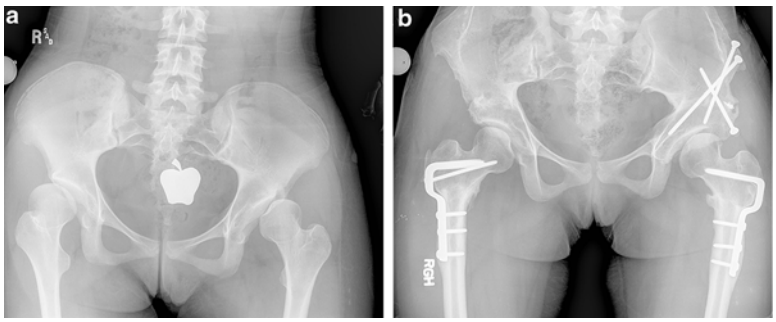


Fig. 29.14 (**a**) A 14-year-old female with Down's syndrome and concurrent severe coxa valga and acetabular dysplasia. (**b**) Left hip corrected with proximal varus osteotomy of the femur with periacetabulr triple osteotomy. Reprinted with permission from the Rubin Institute for Advanced Orthopedics, Sinai Hospital of Baltimore



Fig. 29.15 (**a**) An 18-year-old male with history of Perthes disease of the right hip who had previously undergone aproximal femoral valgus osteotomy with residual impingement pain and gluteus medius weakness. (**b**) Same patient after Ganz femoral head reduction surgery and relative femoral neck lengthening with greater trochanter transfer. (**c**) Subsequent triple pelvic osteotomy in same patient to address residual hip pain due to decrease contact area between newly shaped femoral head and oblong acetabulum. Reprinted with permission from the Rubin Institute for Advanced Orthopedics, Sinai Hospital of Baltimore



Fig. 29.16 A 13-year-old female basketball player presents with new-onset left hip pain with radiographs depicting underlying acetabular dysplasia (**a**), radiographs after triple osteotomy with improvement of femoral head position and coverage and normalization of the center edge angle (**b**). (**c**) Radiographic appearance of pelvis after hardware removal with excellent left hip configuration and no compromise of the pelvic outlet for future childbearing. This is the same patient shown in Figs. 29.11, 29.12, and 29.13. Reprinted with permission from the Rubin Institute for Advanced Orthopedics, Sinai Hospital of Baltimore

1.8-mm Ilizarov or similar sized smooth k-wires. After adequate correction and repositioning of the acetabulum is completed and confirmed under fluoroscopy, the wires are sequentially overdrilled with a cannulated 3.2-mm drill bit and solid 4.5-mm screws are inserted for final stabilization (Fig. 29.13b).

The following figures are case examples of the triple pelvic osteotomy (Figs. 29.14, 29.15, and 29.16).

Closure proceeds with repair of the iliacus fascia to the gluteus medius fascia to cover the exposed iliac crest, repair of the rectus femoris tendon, and a layered closure of the subcutaneous tissues and skin over a drain.

Postoperative Care

The patient is placed into a hip abduction pillow prior to leaving the operative suite and converted to a single leg hip abduction brace before discharge from the hospital. Toe-touch weight bearing is allowed with crutches or a walker for 6 weeks. After radiographic confirmation of healing, the patient is advanced to full weight bearing and physiotherapy.

Box 29.5. Modified Triple Osteotomy

Technical tips:

- Can be performed with a single anterior ilioinguinal incision.
- Identify, isolate with a vessel loop, and protect the femoral nerve, artery and vein prior to iliopsoas tenotomy as well as during the superior ramus and ischial bone cut.
- Adequate medial exposure must be achieved for the superior rami bone cut.
- The superior rami osteotomy site usually lies directly behind the neurovascular bundle.
- A multiple-drill hole technique, Ganz osteotome, and Gigli saw are utilized for the pubic rami, ischial, and ileal bone cuts, respectively.
- Proper oblique fluoroscopic views are critical to performing the ischial osteotomy.
- The Ganz osteotomy must be directed above the ischial spine for the correct ischial osteotomy.
- It is critical to flex the hip 15–20° while levering the Ganz osteotome distally to achieve the proper osteotomy direction; this position protects the femoral nerve from a tension injury.
- Two half-pins are used as anatomical reference points and for controlled mobilization of the fragment.
- Three to four large fragment screws (4.5 mm) are adequate for fixation.

Residual Deformities of the Hip Joint Secondary to Slipped Capital Femoral Epiphysis

The residual deformities that result from a previous slipped capital femoral epiphysis (SCFE) are typically varus, extension, external femoral rotation, leg length discrepancy, and anterolateral femoral acetabular impingement (FAI) [28, 29]. The clinical relevance of each of these deformities depends on the severity of the SCFE and the prior treatment. Some patients will present with a long standing SCFE with severe deformity that has never previously been diagnosed.

The typical treatment of in situ screw fixation for minimal or mild SCFE has proven effective with long-term success [30]. However, the same strategy for moderate or severe magnitude of SCFE often results in unacceptable residual deformities and is associated with pain, gait abnormality and joint dysfunction [31, 32]. There is no absolute criterion for the amount of deformity that requires immediate or eventual reconstruction. However surgeons are increasingly managing even milder deformity associated with FAI in order to prevent poor long term outcomes [33, 34]. The decision to perform a corrective osteotomy should be based on the patient's function and symptoms, as well as the magnitude of deformity. If conservative observation is the treatment strategy, then the physician should follow the patient well into young adulthood to monitor the presence of early degenerative joint disease or impingement that would require a more aggressive treatment plan.

The recommended treatment strategy for residual hip deformities secondary to SCFE consists of either a modified Southwick intertrochanteric osteotomy or a modified Dunn procedure for femoral head reorientation. The osteotomy elected may depend on whether the proximal femoral growth plate is open or closed. If the physis remains open, then the author's treatment of choice is the modified Dunn procedure via a surgical hip dislocation described by Ganz [35]. Once the physis has closed, the proximal femoral periarticular retinaculum becomes very thin resulting in a higher risk of avascular necrosis for the modified Dunn procedure. In the face of a significant proximal femoral deformity with a closed capital femoral physis, a modified Southwick intertrochanteric osteotomy is recommended.

Performing a subcapital osteotomy versus an intertrochanteric osteotomy for a chronic SCFE deformity remains controversial. Several studies of the modified Dunn procedure have reported favorable results with significantly low rates of avascular necrosis [36–38]. However, there exists inconsistent reports on the incidence of avascular necrosis associated with subcapital osteotomies [39–41]. The decision may be surgeon dependent, and related to experience with surgical hip dislocation and the Modified-Dunn procedure. Surgeons may elect to stage treatment of a skeletally immature, chronic stable SCFE by first performing in situ screw fixation, and then following with a planned intertrochanteric osteotomy after physeal closure. An open or arthroscopic osteoplasty, or "bumpectomy," of the femoral neck deformity can also be performed in combination with this approach to avoid femoral-acetabular impingement.

The Modified Dunn Procedure for Residual Deformities of the Proximal Femur Secondary to SCFE: Operative Technique

The modified Dunn procedure has been described multiple times in the current literature. H. Huber et al. provided an excellent description of this procedure in 2009 [39].

Positioning and Exposure

The modified Dunn procedure is performed with the patient in the lateral decubitus position. The incision is centered on the greater trochanter with approximately 5–8 cm distal and 5–8 cm proximal (Fig. 29.17).

The interval between the tensor fascia lata and gluteus maximus muscles is used to gain access to the level of the vastus lateralis and gluteus medius muscles. The posterior border of the vastus lateralis muscle is identified at the level of the vastus ridge. The posterior border of the gluteus medius muscle is identified along with the piriformis tendon and the sciatic nerve. A greater trochanteric osteotomy is performed with an oscillating saw that connects the posterior border of the vastus lateralis muscle to the posterior border of the gluteus medius muscle. The greater trochanteric fragment should be approximately 1–1.5 cm thick for adequate fixation at the end of the procedure. The digastric muscle flap to include the vastus lateralis muscle, the greater trochanteric fragment, and gluteus medius muscle is flipped medially allowing exposure of the gluteus minimus muscle and underlying hip joint capsule (Fig. 29.18a–c).



Fig. 29.17 The incision is centered on the greater trochanter for a total length of 15–20 cm. The clinical photograph depicts the outline of the iliac crest, the ASIS, the greater trochanter, and the longitudinal incision centered on the greater trochanter. Reprinted with permission from the Rubin Institute for Advanced Orthopedics, Sinai Hospital of Baltimore

Surgical Hip Dislocation

The next step in the procedure is the exposure of the hip capsule and the Z capsulotomy. A small Cobb elevator is used to lift the gluteus minimus muscle off of the capsule and provide an interval for a Taylor retractor to be inserted onto the outer table of the ilium. The surgeon will note a prominence in the hip capsule caused by the superior ridge of the femoral neck that was "left" behind as the femoral head progressively slipped in a posterior and medial direction. The Z-capsulotomy is performed with the longitudinal limb in the midline of the capsule. The anterior limb of the capsulotomy is performed at the base of the femoral neck while the posterior limb is extended along the posterior rim of the acetabulum (Fig. 29.19).

The critical step at this point in the procedure is the creation of the retinacular soft tissue flap that will contain the blood vessels to the femoral head. Careful removal of bone from the base of the greater trochanter will create this flap that consists of the proximal femoral periosteum, pericapsular retinaculum, and short external rotator muscles. The creation of this flap is accomplished by removing 5–8 mm of bone from the posterior aspect of the greater trochanteric base by the "inside out technique"(Fig. 29.20). It is critical to minimize soft tissue tension and stretch while excising the bone.

Similarly, the proximal aspect of the greater trochanter base is removed to create a surface that is flush with the superior portion of the femoral neck. This will result in the "relative" femoral neck lengthening after the greater trochanteric fragment is advanced distally during the closure.

If the femoral head has not been previously secured with prior screw fixation, then temporary fixation with a stout single threaded K-wire is performed (Fig. 29.21). The hip is gently dislocated with release of the ligamentum teres from the fovea capitis. After the femoral head is delivered out of the joint, the acetabulum and labrum are inspected for any detrimental changes. Next, the periosteum and retinaculum of the femoral neck is carefully peeled from anterior to the mid-line both medially and laterally. The lateral aspect of this dissection is critical and the continuity of the soft tissue flap to the femoral head must be maintained.

Deformity Correction and Fixation

Following dislocation, the femoral head fixation is removed and a curved osteotome is inserted into the physis. Gentle levering of the curved osteotome is performed to allow the femoral head to gradually detach from the femoral neck. While the femoral head is carefully being levered off the neck, the retinacular soft tissue flap should be visualized to

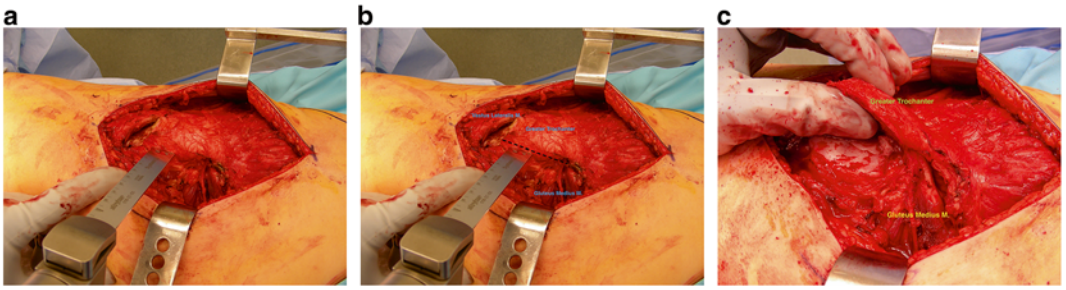


Fig. 29.18 (**a**, **b**) The greater trochanteric osteotomy begins at the posterior border of the vastus lateralis muscle and extends to the posterior border of the gluteus medius muscle. (**c**) The greater trochanteric fragment should be approximately 1.0–1.5 cm in thickness. Reprinted with permission from the Rubin Institute for Advanced Orthopedics, Sinai Hospital of Baltimore

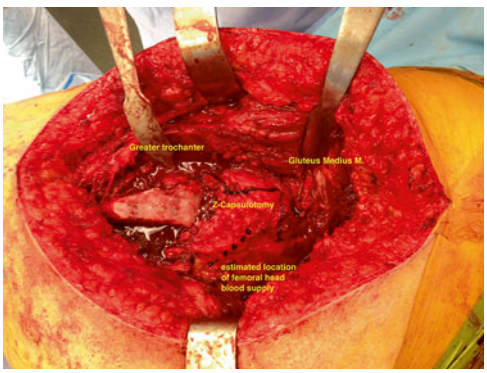


Fig. 29.19 The Z-capsulotomy is created with a longitudinal incision in the capsule in line with the femoral neck. The anterior limb of the capsulotomy is performed anterior at the base of the femoral neck. The posterior limb of the capsulotomy is directed along the posterior border of the acetabulum. Reprinted with permission from the Rubin Institute for Advanced Orthopedics, Sinai Hospital of Baltimore

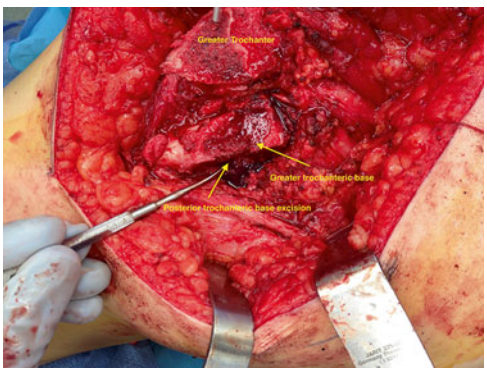


Fig. 29.20 The posterior aspect of the base of the greater trochanter is excised to create the soft tissue vascular sleeve to the femoral head. The excision of 5–8 mm of bone produces soft tissue redundancy that allows greater mobility of the femoral head during the realignment procedure. Reprinted with permission from the Rubin Institute for Advanced Orthopedics, Sinai Hospital of Baltimore

prevent any tearing or stretching. If the final posterior portion of the physis is difficult to detach, the osteotome is carefully directed slightly distally, removing a small portion of the posterolateral femoral neck with the femoral head in order to protect the blood supply. The femoral head along with the soft tissue vascular flap is allowed to fall back into the acetabulum and the femoral neck is lifted into the operative field for inspection (Fig. 29.22).

The femoral neck is trimmed of excess bone and shortened to eliminate any tension of the vascular leash with femoral head reduction. The shortening does not have to be excessive, approximately 1 cm, as long as no tension of the blood supply is evident when the femoral head is reduced. The femoral head is carefully retrieved from the acetabulum and reduced to the "new" femoral neck. The vascular retinacular leash is inspected to insure no tension is present after which temporary K-wire fixation is performed. The hip joint is reduced and its position is confirmed under fluoroscopy. The fixation is finalized via 4.5-mm screws, 3.5-mm screws, or threaded K-wires. The size of the fixation depends on the size of the femoral head and balancing stable fixation while minimizing the amount of area of the femoral head violated to prevent devascularization. The capsule is repaired with care not to over tighten the posterior capsule, which could stretch the blood supply to the femoral head. The greater trochanter is advanced distally and repaired with two 4.5 mm solid screws or two 5.5-mm cannulated screws. The goal of the greater trochanteric transfer is to advance the proximal tip of the greater trochanter below the newly established center of the femoral head (Fig. 29.23). The remainder of the closure is performed in the typical fashion over a drain.

Postoperative Care

The patient is placed into hip abduction pillow prior to leaving the operative room and converted to a single leg hip abduction brace before discharge from the hospital.

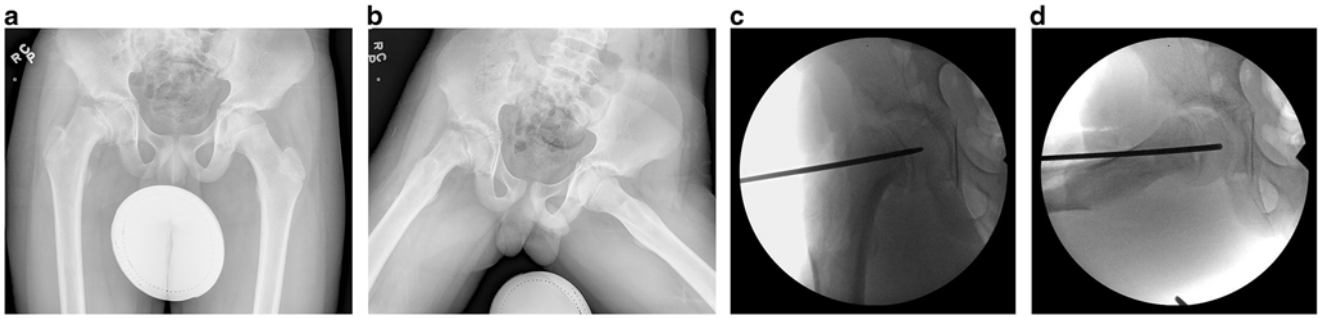


Fig. 29.21 (**a**, **b**) A 14-year-old male who presented with an acute slipped capital femoral epiphysis. (**c**, **d**) Intraoperative fluoroscopic views depicting temporary head fixation to allow for surgical dislocation of the hip without compromising the vascular supply to the femoral head. Reprinted with permission from the Rubin Institute for Advanced Orthopedics, Sinai Hospital of Baltimore

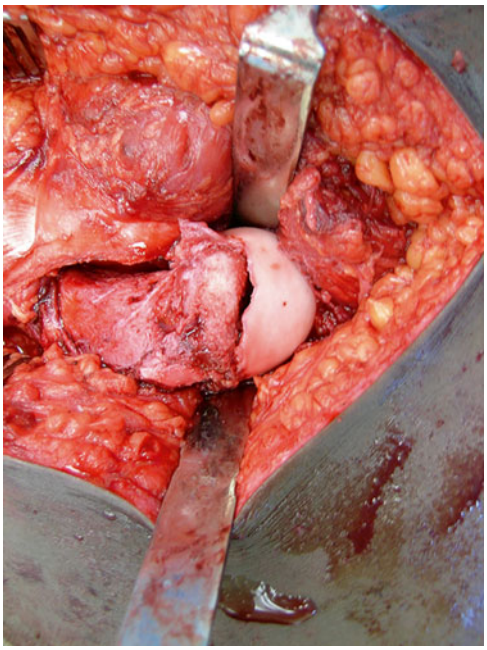


Fig. 29.22 The femoral head is allowed to fall back into the acetabulum after gently levering the femoral head off of the neck through the pathologic physis. Reprinted with permission from the Rubin Institute for Advanced Orthopedics, Sinai Hospital of Baltimore

The patient remains toe touch weight bearing for 12 weeks with gentle and limited passive range of motion of the hip. The initial 6-week postoperative range of motion is performed with a CPM (continuous passive motion) device. The second 6-week period therapy protocol consists of gentle, limited passive hip range of motion and CPM. At the 12-week postoperative mark, the patient begins gradual advance to full weight bearing, hip joint range of motion, and hip muscle strengthening.

Box 29.6. Modified Dunn Procedure

Technical tips:

- Adequate thickness of the greater trochanter osteotomy must be maintained for later fixation (1–1.5 cm width).
- The most critical aspect of the exposure is creating and maintaining the posterior retinacular soft tissue sleeve.
- The femoral head must be temporarily stabilized before the hip is dislocated to insure the vascular leash is not torn.
- Removing 5–8 mm of bone from the posterior margin and the base of the greater trochanter as well as shortening the neck up to 1 cm will aid in relieving tension on the vascular flap.
- Fixation of the femoral head can be done with 4.5 mm screws, 3.5 mm screws, or threaded Steinman pins.
- The hip capsule should be repaired with minimal tension to avoid stretch of the vascular supply to the femoral head.
- The greater trochanter should be advanced until the proximal tip is at or below the center of the femoral head.
- The greater trochanter is repaired with two 4.5-mm or 5.5-mm screws with washers.

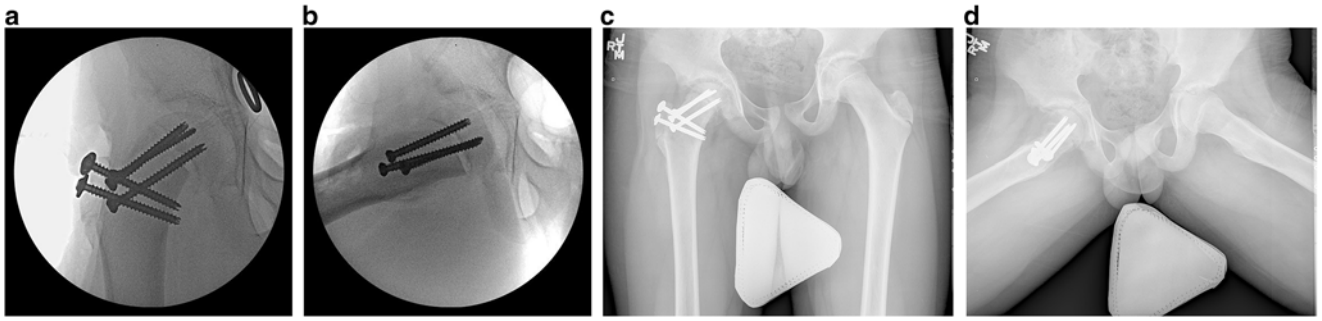


Fig. 29.23 (**a**, **b**) Demonstrating the same patient as in Fig. 29.21 with intraoperative fluoroscopic views of the final femoral head alignment after reduction of the slipped capital femoral epiphysis. (**c**, **d**) AP and frog pelvis radiographs at 8-month postoperative follow-up. Reprinted with permission from the Rubin Institute for Advanced Orthopedics, Sinai Hospital of Baltimore

The Modified Southwick Osteotomy for Residual Deformities of the Proximal Femur Secondary to SCFE: Operative Technique

The Southwick osteotomy has been described in very complicated terms in the past, such as a biplanar or a triplanar osteotomy [42]. In actuality, the osteotomy can be a simple transverse osteotomy stabilized with either an internal blade plate fixation or a simple Ilizarov external fixation. The femoral neck deformity can be described as being in a single oblique plane in the anterolateral direction. This oblique plane deformity is seen as a varus deformity in the coronal view and an extension deformity in the sagittal view. A true second plane of deformity in a residual SCFE exists in the axial plane with retroversion of the hip, or external femoral torsion. However, with proper execution, the Southwick osteotomy can correct both the oblique plane deformity and the external femoral torsion deformity. The one legitimate criticism of the Southwick osteotomy is the inability to resolve the anterolateral prominence at the femoral head-neck junction that causes the femoral acetabular impingement (Fig. 29.24).

The simple modification to the original Southwick procedure is an addition of an anterior approach to the hip with a capsulotomy and excision of the impinging prominence (Fig. 29.25) (Box 29.7). The Southwick osteotomy can be performed with either internal or external fixation. The authors' internal fixation implant of choice is a 130° cannulated locking blade plate. The external fixation of choice can be either a ring fixation device or lower profile external fixator as described by Sabharwal [43].

Positioning and Exposure

The operative technique for the Southwick osteotomy using a 130° blade plate begins with the patient placed on the operating table in the supine position. The entire lower extremity to include the groin, gluteal region, and iliac crest is sterilely prepared and draped. A lateral longitudinal incision is performed starting at the base of the greater trochanter and extended distally for approximately 10–15 cm. A typical lateral approach to the proximal femur is performed. This is accomplished by splitting the tensor fascia lata and retracting the vastus lateralis muscle anteriorly. The lower extremity can be extended, adducted, and internally rotated to visualize the normal profile of the femoral head within the acetabulum. This position can only be obtained by allowing the leg to hang off the side of the operating table. This is accomplished by raising the operative table and allowing the leg to extend off the side of the table on a sterile padded Mayo stand. The position of the leg denotes the deformity that must be corrected.

Deformity Correction and Fixation

A threaded guide pin is inserted into the center of the femoral head under fluoroscopic guidance in both the coronal and sagittal planes. The guide pin is first measured to determine the length of the blade, and then the appropriate-sized chisel is placed over the guide pin. The position of the guide pin determines the correction of the varus deformity, whereas, the rotation of the chisel will determine the deformity correction in the sagittal plane. In order to correct the extension deformity caused by the posteriorly slipped epiphysis, the chisel is rotated the number of degrees towards the ceiling equal to the amount of sagittal plane deformity. After the correct chisel track has been created, a transverse osteotomy in the intertrochanteric zone is performed with an oscillating saw. The chisel is withdrawn and the appropriate sized cannulated 130° blade plate is inserted over the original guide pin. An ideal 130° blade plate design has no medial offset. The distal femoral fragment is first rotated internally to correct the external femoral torsion and then reduced and secured to the plate with cortical screws, correcting the varus and extension deformities. The osteotomy is distal to the center, or apex, of the deformity, which lies at the level of the previous physis, creating an obligatory translation of the distal fragment best visualized in the sagittal plane.

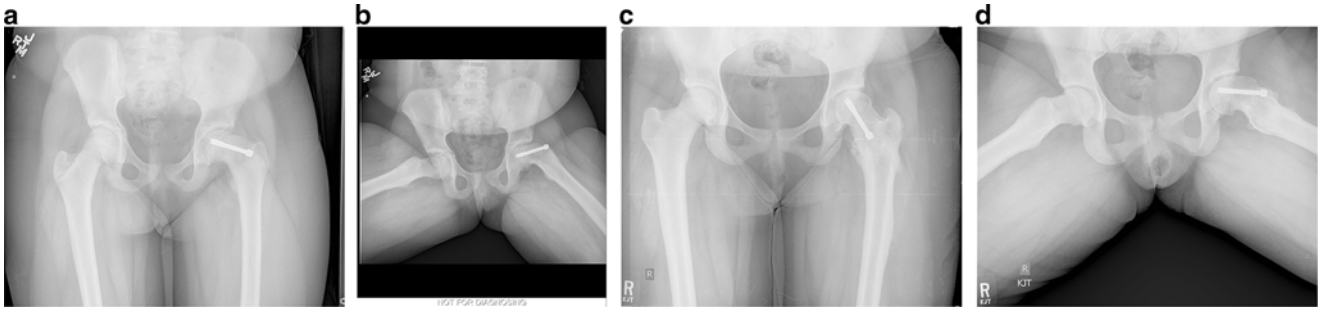


Fig. 29.24 (**a**, **b**) A 13-year-old female AP and frog pelvis radiographs demonstrating residual deformity after slipped capital femoral epiphysis of the left hip treated with in situ screw fixation. (**c**, **d**) AP and frog pelvis radiographs after proximal femoral Southwick realignment osteotomy with residual anterior deformity resulting in femoral acetabular impingement (FAI). Reprinted with permission from the Rubin Institute for Advanced Orthopedics, Sinai Hospital of Baltimore

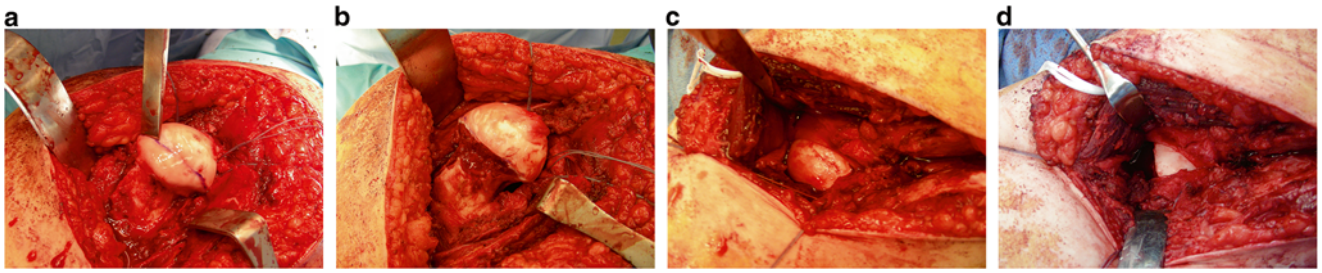


Fig. 29.25 (**a**) An example of anterolateral impingement as seen from a surgical dislocation approach; this impingement is similar to that seen after realigning the residual deformity from a SCFE with an intertrochanteric osteotomy. (**b**) The same femoral head as shown in Fig. 29.25a after bony impingement resection. (**c**, **d**) A similar bony resection accomplished through an anterior approach without the need for complete dislocation of the hip joint. Reprinted with permission from the Rubin Institute for Advanced Orthopedics, Sinai Hospital of Baltimore

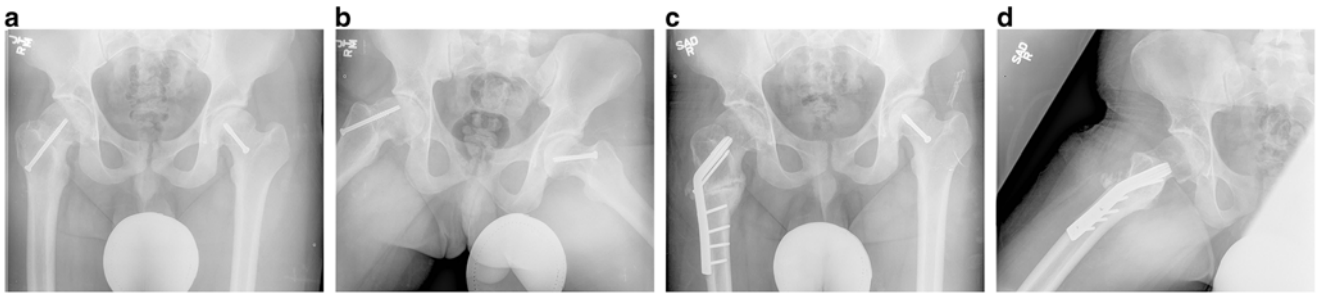


Fig. 29.26 (**a**, **b**) AP and frog pelvis radiographs of 18-year-old male with residual proximal femoral retroversion and external torsion from a previous SCFE treated at an outside facility at age 13. Preoperatively, the patient had hip flexion to 80° with significant impingement and concurrent external foot progression angle during gait. (**c**, **d**) AP pelvis and lateral hip radiographs demonstrating the results after a Southwick osteotomy with concurrent open anterior femoral acetabular impingement resection. Postoperatively, the patient attained hip flexion to 110° with no clinical impingement and a neutral foot progression angle during gait. Reprinted with permission from the Rubin Institute for Advanced Orthopedics, Sinai Hospital of Baltimore

Femoral Neck Osteoplasty

After completing the intertrochanteric osteotomy, all components of the SCFE have been corrected except the anterolateral impingement. An anterior approach is performed through a "bikini" type incision made in the flexion crease of the hip. The incision is approximately 2 cm below the anterior superior iliac spine and measures 6–8 cm centered over the interval between the sartorius muscle and the tensor fascia lata muscle. A typical anterior approach is performed by elevating the sartorius and tensor fascia lata muscles off of the iliac crest and releasing the rectus femoris tendon. The anterior hip capsule is identified and a T or Z capsulotomy is performed. The anterolateral bony prominence is identified and removed with osteotomes and/or ronguers completing the femoral neck osteoplasty

(Fig. 29.26). Anteriorly, the rectus femoris tendon is repaired along with the insertions of the sartorius and tensor fascia lata muscles. Laterally, the vastus lateralis muscle is repaired to the vastus ridge and the fascia lata sutured longitudinally. The superficial tissues and skin are closed in a layered fashion over a drain.

Postoperative Care

Postoperatively, the patient is placed in a hip abduction pillow and converted to a single leg hip abduction brace before being discharged from the hospital. The patient remains with toe touch weight bearing for approximately 6 weeks and then advanced to full weight bearing after radiographic confirmation of healing at the proximal femoral osteotomy site.

Alternative Technique: Percutaneous Osteotomy and External Fixation

The Southwick osteotomy stabilized with an external fixation device is similar in concept, but is performed in a percutaneous fashion. The initial proximal half pin is placed parallel to a proximal reference wire inserted from the tip of the greater trochanter to the center of the femoral head. This half pin is placed directly lateral to medial at the base of the greater trochanter. A femoral arch is attached to the initial half pin and rotated the number of degrees towards the ceiling equal to the amount of sagittal plane deformity. This is the similar technique to the chisel position described above. After the proximal femoral arch is placed in the perfect position mimicking the deformity, a second half pin is inserted from an anterolateral to posteromedial direction stabilizing the proximal construct. A distal femoral arch is mounted via two transverse half pins placed perpendicular to the anatomic axis of the proximal third of the femur. A third half pin is added to both the proximal and distal arches completing and strengthening the final construct. A multiple drill hole percutaneous osteotomy is performed through a lateral 1-cm incision in the intertrochanteric region. The two femoral arches are grasped like handles and the fragments are manipulated correcting the deformity. The sequence of correction is rotation, translation, and then angulation. The two arches should be parallel or nearly parallel once the deformity is corrected. The two arches are now connected via three to four threaded rods with conical washers (Fig. 29.27). The anterior impingement excision is performed as previously described. Postoperatively, the patient with external fixation is allowed 50 % weight bearing immediately with concurrent physiotherapy for range of motion of the hip. Radiographs are performed every 4 weeks to determine complete consolidation of the osteotomy and eventual external fixation removal.

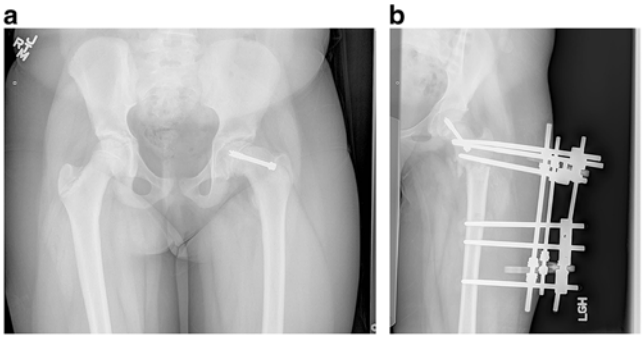


Fig. 29.27 (**a**, **b**) A 13-year-old female as previously depicted in Fig. 29.24 demonstrating the preoperative and postoperative radiographs of a proximal femoral Southwick osteotomy using an Ilizarov external fixation device. Reprinted with permission from the Rubin Institute for Advanced Orthopedics, Sinai Hospital of Baltimore

Box 29.7. Modified Southwick Osteotomy

Technical tips:

- A 130°, zero-offset, cannulated blade plate is an ideal implant for the procedure.
- Directing the blade plate chisel in an appropriate oblique plane is critical for correcting varus and flexion deformities.
- The lower extremity can be extended, adducted, and internally rotated to achieve a correct AP position of the proximal femur—this is achieved by allowing the lower extremity to come off and slightly under the OR table on a sterile padded Mayo stand; this position allows for accurate insertion of the chisel for the blade plate
- To minimize the risk of impingement, the surgeon should consider performing an anterior approach with anterior capsulotomy to remove the anterolateral "bump" at the neck-epiphyseal junction.
- A percutaneous intertrochanteric osteotomy with application of an Ilizarov external frame is a proven, alternative technique.

Residual Deformities of the Hip Joint Secondary to Legg–Calvé–Perthes Disease

Idiopathic osteonecrosis of the epiphysis in the immature hip, better known as Legg–Calvé–Perthes disease or the abbreviated Perthes disease, almost always leaves a mark or "foot print" on the involved hip even after resolution of the process. The evolution of the deformity and final head morphology depends on the age of the patient and degree of vascular insult [44, 45]. The vast majority of younger patients (onset of symptoms <6 years) with Perthes disease progress through the natural healing process with good clinical outcome [46].



Fig. 29.28 A 16-year-old male with a history of left hip Perthes disease with residual coxa magna and coxa breva. Reprinted with permission from the Rubin Institute for Advanced Orthopedics, Sinai Hospital of Baltimore



Fig. 29.29 A 12-year-old male with severe right hip residual deformity after conservative treatment for late onset Perthes disease. Reprinted with permission from the Rubin Institute for Advanced Orthopedics, Sinai Hospital of Baltimore

However, even in the case of a successful outcome with Perthes disease, residual changes or deformities of the hip can still be present. A recent multicenter, long-term study indicates Perthes disease patients treated nonoperatively leads to pain, femoral-acetabular impingement, poor subjective function, and a higher incidence of osteoarthritis than previous reports [47].

The most common residual deformities are coxa breva and coxa magna (Fig. 29.28).

In the older child or in the face of an unsuccessful previous treatment, the deformities can be more severe with substantial effects on patient comfort and function. These types of residual deformities also include coxa breva and coxa magna, but severe femoral head deformity, residual proximal and lateral displacement of the femoral head, and concurrent joint contractures are also present (Fig. 29.29).

Leg length discrepancy is another residual effect of Perthes disease. Paradoxically, the younger patient with Perthes disease has a better prognosis for natural resolution of femoral head deformity, but has a higher incidence of a clinically significant leg length discrepancy (>2.0 cm) [48]. These young patients tend to be <4 years of age at the onset of symptoms. The older patient (onset >7 years) has a worse prognosis for natural resolution, but often does not develop a clinically significant leg length discrepancy. In the authors' opinion, this is due to the fact that the femoral neck has established the majority of its natural length by this age. The more common cause of leg length discrepancy in Perthes disease is iatrogenic in nature. The proximal femur varus osteotomy is a common treatment for Perthes disease. However, an overly aggressive varus osteotomy can result in a significant coxa vara, limb shortening, and a decreased articulotrochanteric distance, which is associated with a mechanically weak hip abductor mechanism causing a long lasting limp [49–52]. Furthermore, the "containment" afforded by the varus osteotomy does not unload the hip or guarantee maintenance of this containment in the postoperative period. Therefore, the author does not recommend the varus osteotomy as the treatment of choice for Perthes disease. Instead, for the Perthes disease that has failed conservative management in the younger age group or in late onset Perthes disease, the author recommends articulated hip distraction via open soft tissue releases and external fixation placement. Details of this technique are described later in the chapter.

Coxa Breva

As previously stated, coxa breva is defined as shortening and widening of the femoral neck, which alters the relationship of the tip of the greater trochanter to the center of the femoral head [2]. This creates a mechanical disadvantage of the hip joint by decreasing the lever arm of the hip and decreasing the distance between the origin and insertion points of the gluteus medius muscle. The radiographic presence of coxa breva is not a definitive indication for surgical correction. In reality, coxa breva is ubiquitous in the post-Perthes patient, but very few actually require surgical reconstruction. The indication for surgical reconstruction is based on the patient's function, gait, and leg length discrepancy. Coxa breva in the presence of a persistent Trendelenburg gait, hip fatigue, and hip pain with prolonged activity requires surgical reconstruction. Surgical treatments described for coxa breva include greater trochanteric epiphysiodesis, and greater trochanter transfer (i.e., relative neck lengthening), but ideal treatment remains ill-defined [1, 53].

The author's preferred method of correction of coxa breva is a femoral neck lengthening osteotomy with concurrent greater trochanteric transfer, originally described by Edwin Morscher in 1980 [54] (Box 29.8). The Morscher osteotomy improves the lever arm length of the hip joint by lengthening the femoral neck, corrects a leg length discrepancy up to 1.5–2.0 cm, and reestablishes the center of femoral head to

tip of greater trochanteric relationship through distal transfer of the greater trochanter. The greater trochanteric transfer not only increases the distance between the origin and insertion of the gluteus medius muscle, but it also adds lateral distance to the mechanical lever arm of the hip joint, which imparts greater mechanical advantage to the gluteus medius muscle. For the Morscher osteotomy to be successful, the hip joint should be congruent with full range of motion with no evidence of femoral acetabular impingement. The following is a brief description of the author's preferred technique for the Morscher osteotomy.

The Morscher Osteotomy for Coxa Breva Secondary to Perthes Disease: Operative Technique

Positioning and Exposure

The operative technique for the Morscher osteotomy begins with the patient placed on the operating table in the supine position. The entire lower extremity to include the groin, gluteal region, and iliac crest is sterilely prepared and draped (Fig. 29.30).

A lateral longitudinal incision is performed starting approximately 5 cm above the tip of the greater trochanter and extending distally for approximately 10 cm. A lateral approach to the proximal femur is performed by splitting the tensor fascia lata and lifting the vastus lateralis muscle anteriorly. Proximally, the gluteus maximus muscle fibers should be dissected away from the tensor fascia allowing the gluteus maximus to fall posteriorly. This allows for better visualization of the gluteus medius muscle and easier dissection of the greater trochanteric fragment-gluteus medius muscle complex later in the procedure.

Deformity Correction and Fixation

The internal fixation of choice is a 130° cannulated blade plate. After exposure of the lateral aspect of the proximal femur, a central threaded guide pin is inserted up the femoral neck and into the femoral head. This guide wire represents the eventual path of the blade plate and the new femoral neck orientation. Three additional guide wires are inserted parallel to the initial guide wire. The second guide wire is inserted at the level of the superior base of the femoral head, which represents the new level of the superior border of the femoral neck. The third wire is inserted approximately 1 cm above the second wire. This represents the new base of the greater trochanter (Fig. 29.31).

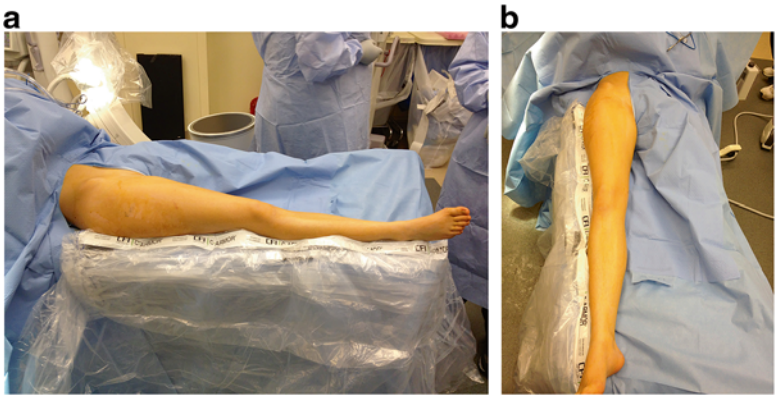


Fig. 29.30 (**a**, **b**) Clinical pictures depicting the typical patient position and draping for the Morscher osteotomy. Reprinted with permission from the Rubin Institute for Advanced Orthopedics, Sinai Hospital of Baltimore

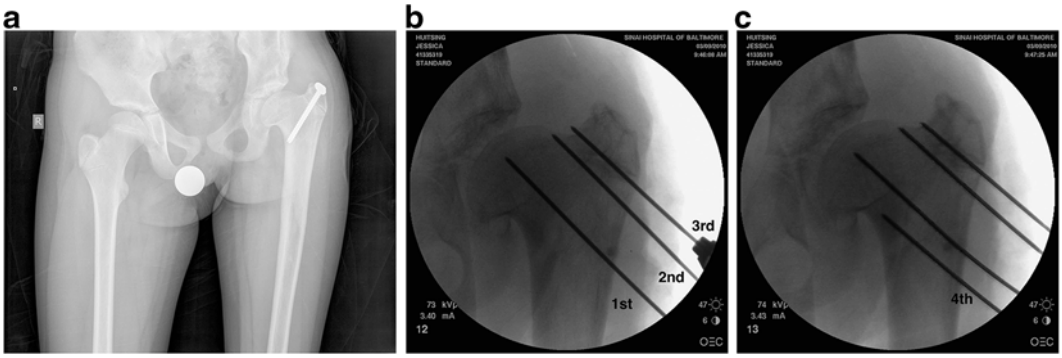


Fig. 29.31 (**a**) AP pelvis radiograph demonstrating severe coxa breva of the left hip. (**b**, **c**) The same patient undergoing left hip Morscher osteotomy with intraoperative fluoroscopic views demonstrating the placement of the guide wires. The first guide wire is inserted at an angle of 130° and marks the path for the blade plate, the second wire marks the superior border of the new femoral neck, the third wire marks the trochanteric osteotomy site, the fourth wire marks the inferior border of the new femoral neck. Reprinted with permission from the Rubin Institute for Advanced Orthopedics, Sinai Hospital of Baltimore



Fig. 29.32 A fluoroscopic image demonstrating the completion of the first two osteotomy cuts allowing mobilization of the greater trochanter, removal of interval bone segment, and the creation of the new superior border of the femoral neck. Reprinted with permission from the Rubin Institute for Advanced Orthopedics, Sinai Hospital of Baltimore



Fig. 29.33 Fluoroscopic image demonstrating the final osteotomy cut that creates the inferior border of the new femoral neck and the insertion of the 130-degree blade plate. Reprinted with permission from the Rubin Institute for Advanced Orthopedics, Sinai Hospital of Baltimore

The interval bone segment between the second and third wire will be removed and used as bone graft. The final guide wire is inserted at the level of the inferior border of the femoral neck. This final guide wire represents the sliding osteotomy surface that will result in femoral neck lengthening as well as lower extremity lengthening. Again, it is important that the guide wires are inserted in a parallel or near parallel fashion. Next, the appropriate sized blade plate chisel is driven over the central guide wire. The chisel should be held in neutral rotation in the sagittal plane to prevent secondary flexion or extension of the osteotomy that would result in minimal contact between the proximal and distal fragments. Once the chisel is fully seated, the appropriate blade length is calculated by adding an additional 1–1.5 cm of length to the chisel length. An oscillating saw is used to cut the bone along the two proximal and the distal most guide wires. The first cut is the most proximal osteotomy is used to mobilize the greater trochanteric fragment along with the gluteus medius muscle. The second cut creates the new superior border of the femoral neck allows for mobilization and removal of the interval bone segment (Fig. 29.32).

This bone should be placed into a moist sponge for later use. The final osteotomy creates the distal border of the femoral neck separates the proximal femoral segment from the distal femoral shaft. After the osteotomies are completed, the chisel is removed and the selected blade plate inserted over the initial threaded guide wire. The blade should be driven to a level that leaves approximately 1–1.5 cm of blade exposed laterally (Fig. 29.33).



Fig. 29.34 Fluoroscopic image showing the reduction of the distal femoral segment reduced to the blade plate that extended out of the proximal segment by 1.0–1.5 cm, which results in a lengthened femoral neck. Reprinted with permission from the Rubin Institute for Advanced Orthopedics, Sinai Hospital of Baltimore

The distal fragment is then reduced to the plate with bone holding forceps. This maneuver causes the distal fragment to slide down the oblique osteotomy, accomplishing femoral neck lengthening, as well as lower extremity lengthening (Fig. 29.34).

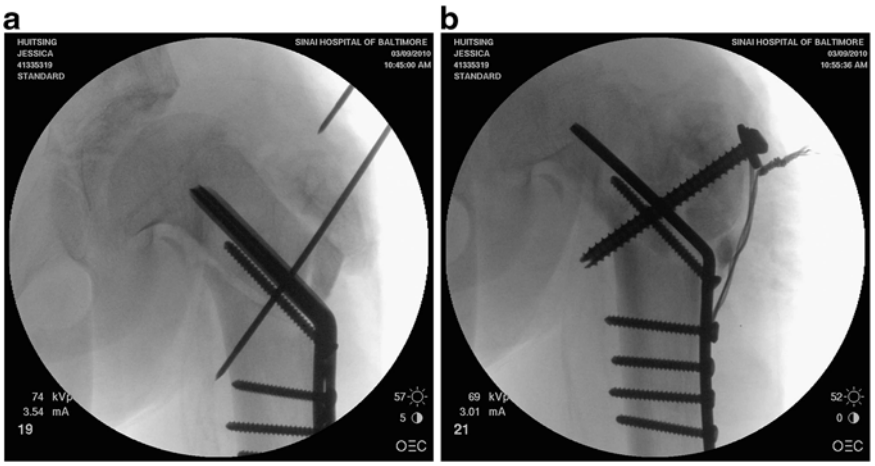


Fig. 29.35 (**a**, **b**) Fluoroscopic images demonstrating the advancement of the greater trochanter with temporary wire fixation and final fixation with a cannulated screw and tension wire. Reprinted with permission from the Rubin Institute for Advanced Orthopedics, Sinai Hospital of Baltimore

The previously removed bone fragment is now notched and placed over the exposed end of the blade plate at the 130° bend.

The final step of the procedure is the mobilization and advancement of the greater trochanter both laterally and distally. The appropriate level of greater trochanteric transfer is accomplished when the tip of the greater trochanter is at or below the level of the center of the femoral head. This position is obtained by carefully mobilizing the gluteus medius muscle and abducting the hip 10–15° before fixation. The greater trochanteric fragment is stabilized with a cannulated 5.5-mm screw and washer, tension band wiring, or a combination of both techniques (Fig. 29.35).

The operative incision is closed as previously described in a layered fashion over a drain.

Postoperative Care

The patient is placed in a hip abduction pillow immediately and converted to a single leg hip abduction brace before discharge from the hospital. The patient is maintained at toe touch weight bearing status and full time brace wear for the initial 6 weeks. After 6 weeks, the brace is used at night for an additional 6 weeks and gradual advancement of weight bearing, hip range of motion, and hip strengthening begins under the supervision of a physiotherapist.

Two key tips for the Morscher osteotomy are the amount of lateral displacement of the distal fragment and the initial position of the chisel in the sagittal plane. First, the distal femoral fragment should not be translated laterally more than 50 % of the original diameter of the proximal femur. If the bone-to-bone contact is less than 50 % of the original diameter, there is a significant increase in implant failure. Second, the chisel position in the sagittal plane should be shifted slightly anterior or posterior to allow enough space for the eventual trochanteric cannulated screw to pass without difficulty (Figs. 29.36 and 29.37).

Box 29.8. Morscher Osteotomy for Coxa Breva

Technical tips:

- The initial guide wire is inserted at an angle of 130°. This is the angulation of the new femoral neck.
- A 130-degree, zero-offset, cannulated blade plate is an ideal implant for the procedure.
- The length of the blade plate is determined by measuring the initial guide wire inserted up the femoral neck and adding 10–15 mm to this measurement.
- When the blade plate is inserted, the plate is left away from the lateral cortex by 10–15 mm; this will allow the distal fragment to shift laterally, lengthening the femoral neck.
- It is critical not to translate the distal segment more than 50 % of the original diameter of the subtrochanteric width of the femur. If greater than a 50 % translation is performed, inadequate bone contact will be present and an increased risk for implant failure will occur.
- The trochanteric fragment must be carefully mobilized without compromising the innervation of the gluteus medius muscle.
- The interval bone segment removed from the superior femoral neck osteotomy should be saved for femoral neck bone grafting around the proximal bend of the 130-degree blade plate.
- The greater trochanter should be advanced until the proximal tip is at or below the center of the femoral head.

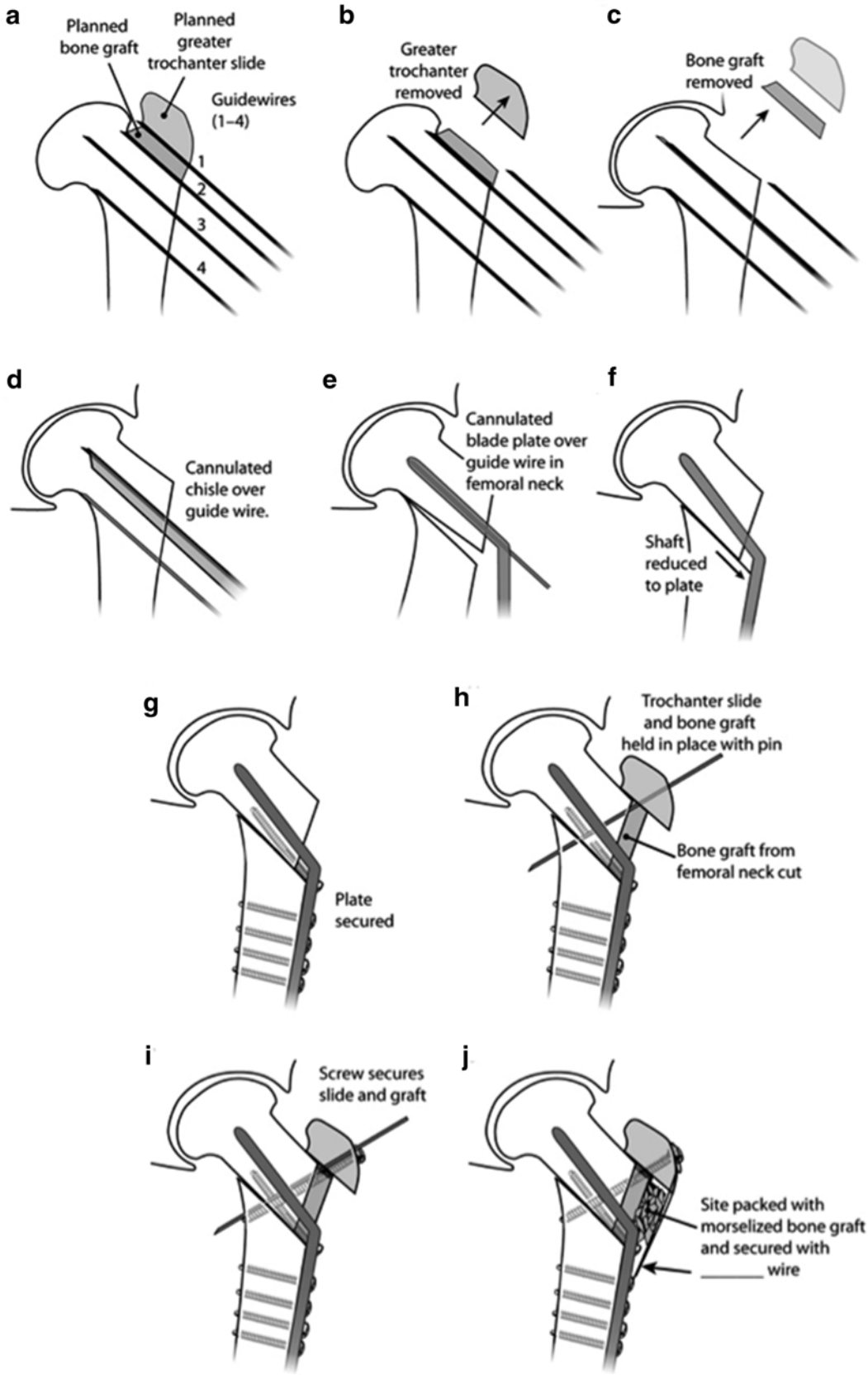


Fig. 29.36 Step-by-step schematic summary of the Morscher osteotomy. Reprinted with permission from the Rubin Institute for Advanced Orthopedics, Sinai Hospital of Baltimore

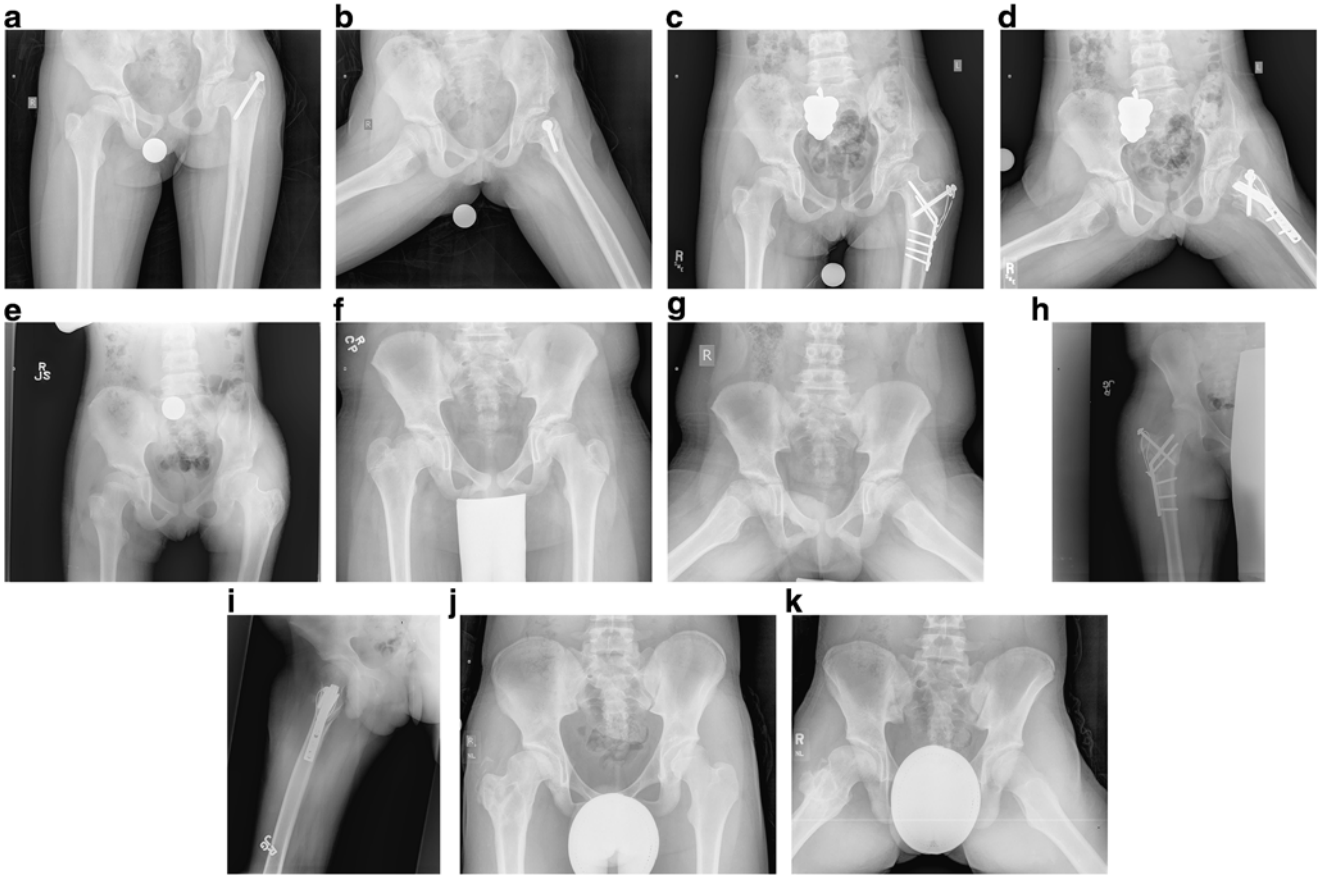


Fig. 29.37 (**a**, **b**) A 12-year-old female with severe left hip coxa breva from previous AVN. This is the same patient depicted in Figs. 29.31, 29.32, 29.33, 29.34, and 29.35. (**c**, **d**) One year post-reconstruction of the left hip using the Morscher osteotomy technique. (**e**) Three years post-reconstruction AP pelvis radiograph after internal implant removal. (**f**, **g**) Another example of coxa breva demonstrated by AP and frog pelvis of a 13-year-old male with residual right hip deformity from Perthes disease. (**h**, **i**) Immediate postoperative AP and lateral radiographs of the right hip after Morscher osteotomy. (**j**, **k**) Five-year follow-up of the same patient after internal implant removal. Reprinted with permission from the Rubin Institute for Advanced Orthopedics, Sinai Hospital of Baltimore

Coxa Magna

As previously stated, coxa magna is a common residual deformity of Perthes disease. Coxa magna is defined as the enlargement and deformation of the femoral head. This deformity can result in the loss of congruency between the femoral head and acetabulum resulting in femoral-acetabular impingement (FAI). Severe coxa magna causes pain, loss of motion, labral and cartilage degeneration, and eventual osteoarthritis [3–6]. Coxa magna is caused by a combination of the collapse of the proximal femoral epiphysis and altered growth of the articular cartilage (Fig. 29.38).

The coxa magna deformity begins as the femoral head settles after the initial subchondral collapse of the epiphysis. This results in a decreased height and an increased width of the femoral head, similar to applying downward pressure on a water balloon. The amount of continued collapse appears to be related to the amount of epiphyseal involvement and the age of the patient. However, there is significant variation to the pattern of collapse between individual patients and an exact method for predicting the amount of eventual collapse has not been developed. The second variable in the development of coxa magna is the abnormal articular cartilage growth. This abnormal growth is caused by asymmetric pressure applied to the growing femoral head. The cause of the abnormal femoral head growth and deformation is directly related to the loss of hip joint motion and the presence of a significant hip joint contracture. In the stiff hip that has lost significant range of motion, the joint pressures are asymmetric over the femoral head. The zone of contact between the femoral head and the superior articular surface of the acetabulum and acetabular rim represents areas of increased pressure, whereas the portions of the femoral head that are anterior and lateral to the acetabular rim and medial within the acetabular fossa are areas of decreased pressure.

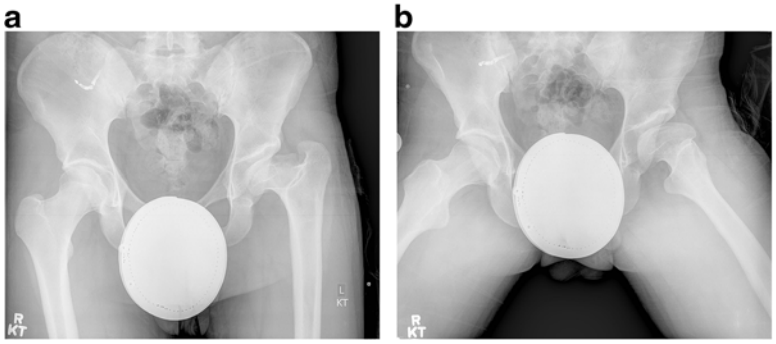


Fig. 29.38 (**a**, **b**) Radiographic example of a coxa magna deformity resulting in loss of congruency of the hip joint and femoral acetabular impingement. Reprinted with permission from the Rubin Institute for Advanced Orthopedics, Sinai Hospital of Baltimore

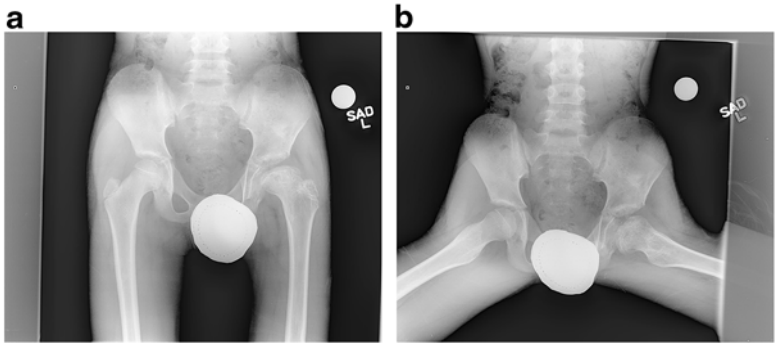


Fig. 29.39 (**a**, **b**) AP and frog pelvis radiographs demonstrating a congruent coxa magna and oblong acetabulum in a patient after late-onset Perthes disease. Reprinted with permission from the Rubin Institute for Advanced Orthopedics, Sinai Hospital of Baltimore

The areas of increased pressure obviously expedite any collapse as the underlying epiphysis loses structural integrity. Furthermore, this pressure also compresses and slows the articular cartilage growth. Conversely, the areas of decreased pressure allow the articular cartilage to grow into the "unweighted" zones, thereby exacerbating the femoral head deformation and coxa magna formation.

Another crucial variable in the evaluation of the coxa magna deformity is the adaptive remodeling changes of the acetabulum. In young patient (<6 years) with Perthes disease, full collapse of the epiphysis can occur with coxa magna formation, but the femoral head remains spherical and the hip joint congruent. It is the author's experience and opinion that this outcome is directly related to the maintenance of adequate hip range of motion during the entire healing process of the femoral head. The only true method to maintain "containment" of the femoral head is to maintain adequate hip range of motion. Adequate hip range of motion is defined by the author as hip motion that is >50 % of the contralateral normal hip. Even in the face of an enlarged and oblong femoral head, adequate range of motion of the hip joint allows the acetabulum to remodel into a larger and oblong congruent joint. This situation can provide a pain-free functional hip joint for several decades with excellent quality of life (Fig. 29.39).

However, in older patients (>10 years for females and >12 years for males), even with successful maintenance of hip range of motion and a relatively spherical femoral head, the amount of acetabular remodeling is minimal in the author's experience. Even mild changes to the overall diameter or shape of the femoral head are not tolerated in this age group. Although these patients have initial good results, the mild to moderate incongruence of the hip joint will eventually lead to FAI and the need for hip reconstruction to avoid early degenerative changes.

The treatment of choice for significant coxa magna deformity of the femoral head is the femoral head reduction osteotomy, also known as the "head splitting" or "head reshaping" osteotomy, initially described by Ganz [55]. This relatively new technique utilizes Ganz's well-described surgical hip dislocation approach, and has shown promising early results [36, 56–58].The following is a brief description of the operative technique for the femoral head reduction osteotomy (Box 29.9).

The Femoral Head Reduction Osteotomy for Coxa Magna Secondary to Perthes Disease: Operative Technique

Positioning and Exposure

The patient positioning and operative approach is identical to the surgical hip dislocation as described above for the modified Dunn procedure. After the capsulotomy and gentle dislocation of the hip, the acetabulum and labrum are inspected for any pathologic changes. With the femoral head

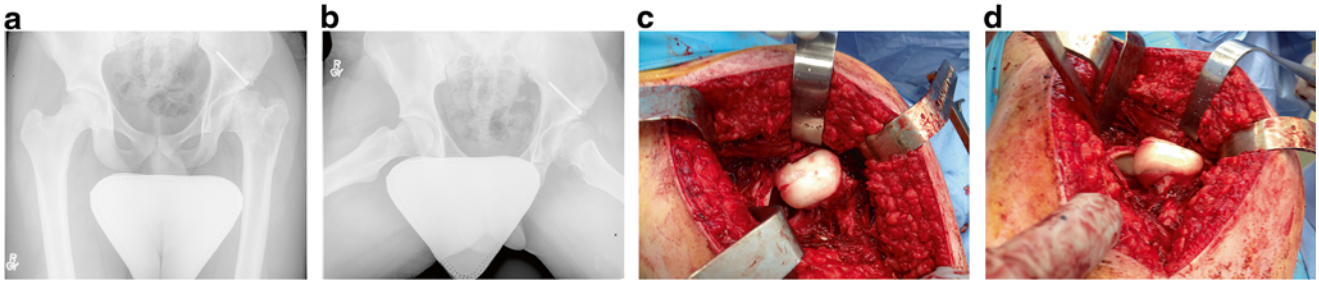


Fig. 29.40 (**a**, **b**) AP and frog pelvis radiographs depicting the same patient in Fig. 29.28, a 16 year old male with coxa magna, coxa breva, and concurrent FAI. (**c**, **d**) Clinical appearance of the femoral head of the same patient after surgical dislocation. It is the author's experience that the true three-dimensional shape of the femoral head does not consistently correlate with various imaging studies; therefore, MRI and CT scans are rarely indicated or performed. Reprinted with permission from the Rubin Institute for Advanced Orthopedics, Sinai Hospital of Baltimore

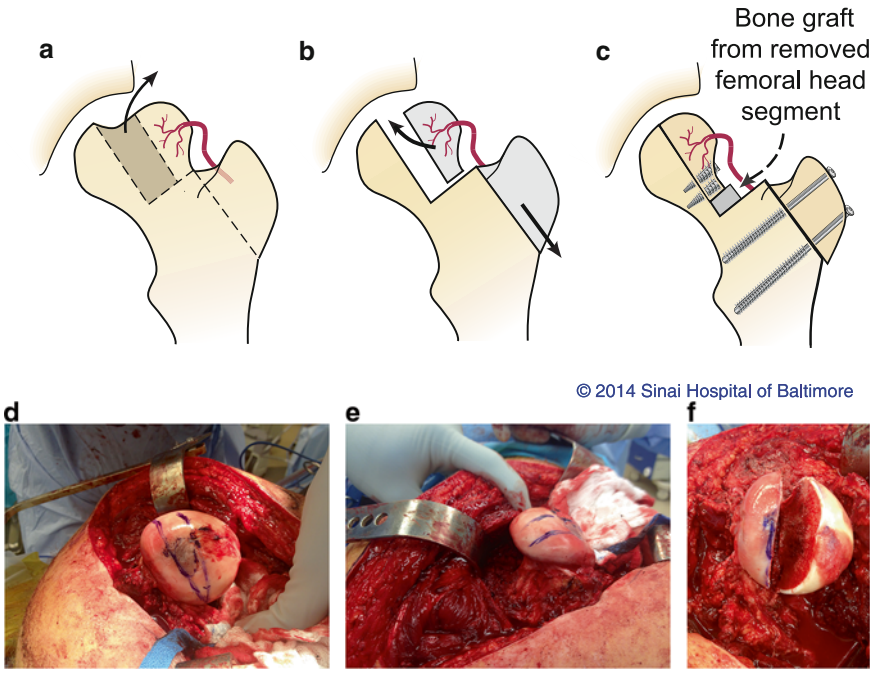


Fig. 29.41 (**a**–**c**) Schematic representation of the femoral head reduction/reshaping osteotomy depicting the creation of a spherical femoral head by excising an interval deformed section of bone. (**d**–**f**) Clinical pictures of another case demonstrating the femoral head osteotomy planning lines that resolves a three-dimensional puzzle into two separate pieces that can be placed together to form a spherical object. Reprinted with permission from the Rubin Institute for Advanced Orthopedics, Sinai Hospital of Baltimore

dislocated, the posterolateral soft tissue leash is inspected for any undue tension and the exact three-dimensional head shape is visualized for planning the osteotomy (Fig. 29.40).

Any cartilage softening and depressed areas are noted as the femoral head is inspected. These areas need to be included in the segmental resection of the mid-portion of the femoral head. Femoral head sizing templates, surgical experience, and three dimensional awareness are used to determine the exact position of the femoral head cuts (Fig. 29.41).

Deformity Correction and Fixation

The femoral head cut is rarely in a pure anterior to posterior direction. Usually the cut is from an anterolateral to a posteromedial direction. The greater the amount of sagittal plane deformity, the more lateral the starting point for the osteotomy needs to be placed. The planned resection of bone can be in a shape of a wafer or a wedge depending on the deformity to be corrected. The greater the sagittal plane deformity present, the more likely a wedge shape is needed

to completely correct the oblique plane deformity. Two K-wires are inserted to represent the two longitudinal cuts (Fig. 29.42).

The most medial cut is performed first with great care to leave greater than a third of the original diameter of the femoral neck intact. Rotating the saw blade in a counterclockwise direction allows a greater segment of the medial neck to remain intact for stability of the medial head fragment. The second osteotomy is now performed. This bone cut is the closest to the posterolateral segment containing the blood supply; so great care is needed to avoid the retinacular vessels entering in the lateral femoral head segment. The final transverse bone cut is performed at the base of the femoral neck, connecting the first two vertical bone cut (Fig. 29.43).

Again, this transverse cut must be performed with great care to avoid the intact medial neck and the posterolateral head-neck junction where the blood supply is entering the lateral femoral head segment. This final cut creates separation of central free fragment from the medial head fragment that is still attached to the intact medial femoral neck, and the lateral mobile femoral head segment attached via the soft tissue vascular leash. The middle segment is carefully removed and the lateral fragment mobilized (Fig. 29.44).

The lateral fragment is reduced to the stable medial head fragment and held with pointed bone reduction forceps. The lateral femoral head fragment usually requires proximal advancement along with the medial translation. The vascular leash is inspected for tension or tenting over the residual base of neck. If a significant medial translation or closing wedge is needed, the posterolateral base of femoral neck must be reduced in size to prevent impingement and tension on the vascular leash. After the femoral head is reduced and vascular supply inspected, three transverse headless screws are inserted to secure the femoral head osteotomy (Fig. 29.45a, b).

The gap between the base of the lateral femoral head fragment and the femoral neck is filled with a bone graft obtained from the removed femoral head fragment. The femoral head is gently reduced into the acetabulum and joint motion is assessed. Any residual impinging bone of the anterior femoral head or neck is excised. The capsule is repaired with absorbable heavy suture and suture anchors at the base of the femoral neck as needed. It is crucial not to over tighten the capsular repair that can place undue tension on the posterior capsule and thereby on the vascular leash.

The hip is placed in mild abduction and the greater trochanter is advanced distally until the tip of the greater trochanter is equal to or below the center of the newly shaped femoral head. The greater trochanter is secured via two 5.5-mm cannulated screws. It is crucial to direct the screws away from the medial femoral neck base to prevent propagation of a fracture form the screw hole to the medial femoral neck which will result in destabilization the medial head fragment (Fig. 29.45c, d).

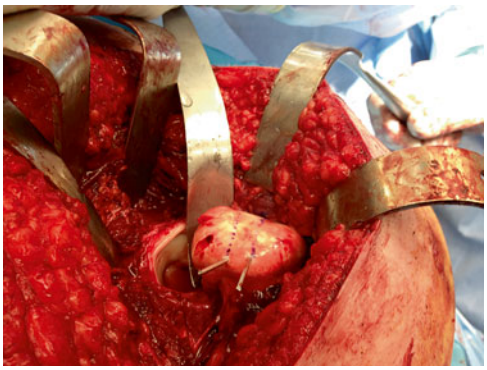


Fig. 29.42 Two guide wires are placed at the planned longitudinal osteotomy sites. Reprinted with permission from the Rubin Institute for Advanced Orthopedics, Sinai Hospital of Baltimore

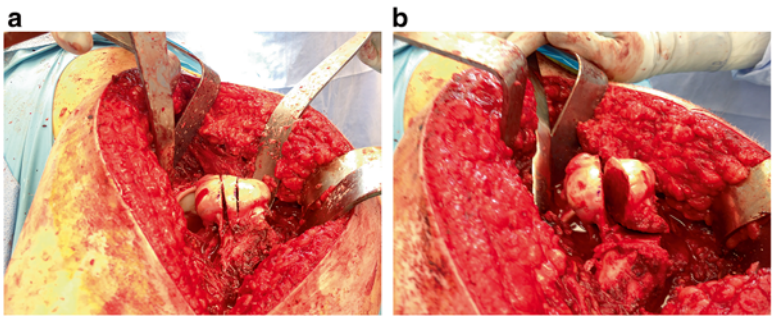


Fig. 29.43 Clinical pictures denoting the two longitudinal cuts (**a**) and the transverse cut (**b**) that create the mobile lateral femoral head fragment attached via the vascular leash. Reprinted with permission from the Rubin Institute for Advanced Orthopedics, Sinai Hospital of Baltimore

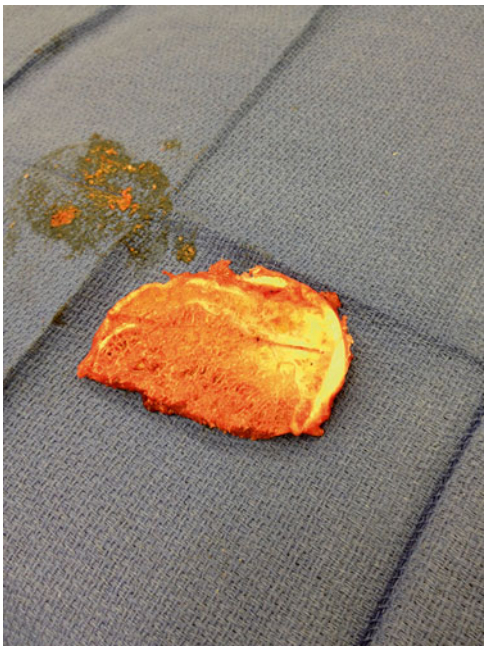


Fig. 29.44 Clinical picture of the excised middle segment. The bone at the base of the cut can be utilized as bone graft in the femoral neck. Reprinted with permission from the Rubin Institute for Advanced Orthopedics, Sinai Hospital of Baltimore

The incision is closed in a routine layered fashion with the tensor fascia lata, subcutaneous tissues, and skin closed over a drain. The patient is gently placed into a supine position with an abduction pillow and a final radiograph is performed (Fig. 29.46).

Postoperative Care

Postoperatively, the patient is placed in a single-leg abduction brace full time except when using a continuous passive motion (CPM) machine for hip flexion and extension. The CPM machine is used 2–4 h during the day and all night for the first 6 weeks. Alternatively, the patient may use the CPM for a greater time during the day and utilize the hip abduction brace at night. The protocol can be adjusted per the patient's preference. Physical therapy consists of simple passive range of motion of the hip not to exceed the following parameters: 75° of hip flexion, 30–40° of hip abduction, full hip extension, zero hip adduction, and 30° of internal rotation and 30° of external rotation. Active sitting and CPM motion can be maximized, increasing the hip flexion to the patient's tolerance. The CPM and daytime bracing are discontinued 6 weeks postoperatively. Toe touch weight bearing is mandated for 3 months to ensure complete healing of the femoral head (Figs. 29.47 and 29.48).

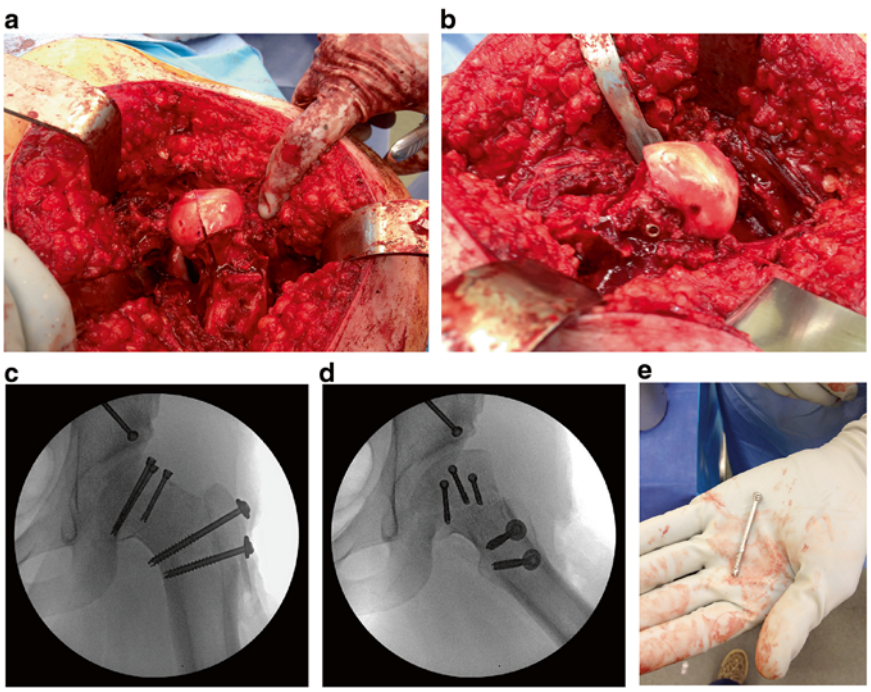


Fig. 29.45 (**a**, **b**) Clinical pictures of the final configuration of the femoral head after fixation of the medial and lateral head fragments with three transverse headless screws. (**c**, **d**) Fluoroscopic images of the final femoral head fixation, hip reduction, and greater trochanter fixation. (**e**) Typical headless screw used to fix the femoral head osteotomy. Reprinted with permission from the Rubin Institute for Advanced Orthopedics, Sinai Hospital of Baltimore

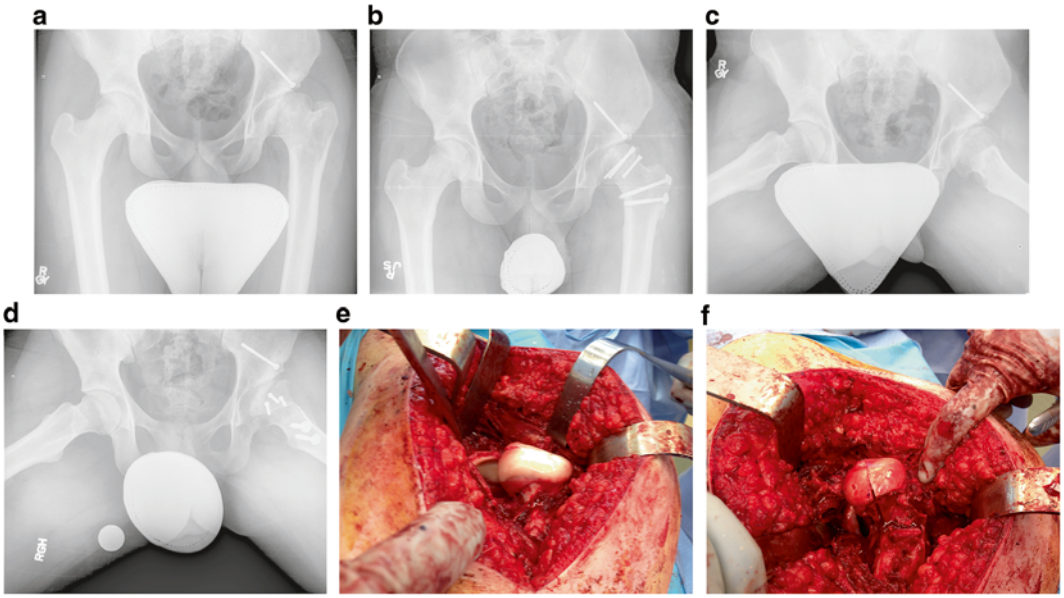


Fig. 29.46 (**a**, **b**) Preoperative and postoperative AP pelvis radiographs after surgical dislocation of the hip with femoral head reduction and transfer of the greater trochanter. (**c**, **d**) Preoperative and postoperative frog pelvis radiographs. (**e**, **f**) Preoperative and postoperative clinical pictures. Reprinted with permission from the Rubin Institute for Advanced Orthopedics, Sinai Hospital of Baltimore

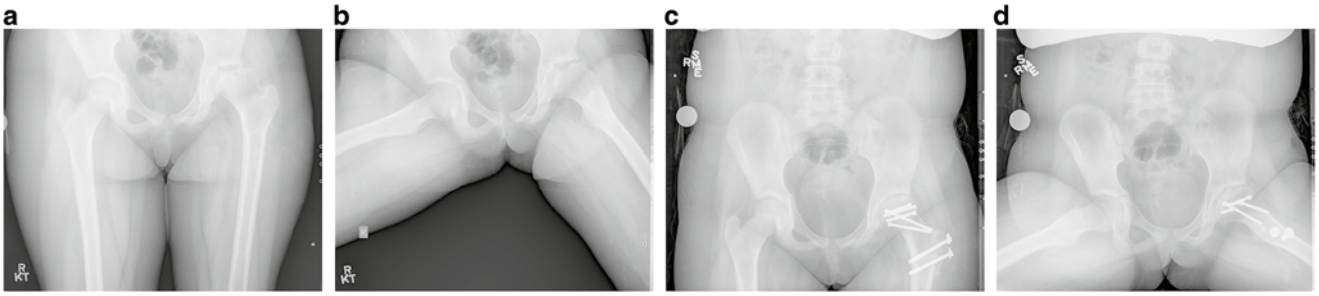


Fig. 29.47 (**a**, **b**) Radiographic example of a 12-year-old female with severe impingement secondary to residual deformity of the femoral head after late-onset Perthes disease of the left hip. (**c**, **d**) Same patient after left hip reconstruction with surgical dislocation, femoral head reduction and reshaping, and greater trochanter transfer. Reprinted with permission from the Rubin Institute for Advanced Orthopedics, Sinai Hospital of Baltimore

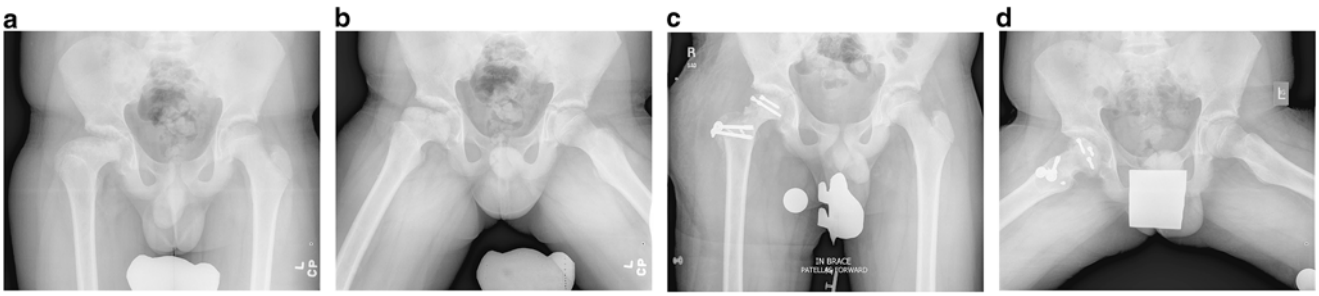


Fig. 29.48 (**a**, **b**) A 12 year-old male with residual right coxa magna resulting in an incongruent hip and FAI secondary to Perthes disease. (**c**, **d**) Radiographs of same patient after right hip reconstruction via a femoral head reduction osteotomy and greater trochanter transfer. Reprinted with permission from the Rubin Institute for Advanced Orthopedics, Sinai Hospital of Baltimore

Box 29.9. Femoral Head Reduction Osteotomy

Technical tips:

- Adequate thickness of the greater trochanter osteotomy must be maintained for later fixation (1–1.5 cm width).
- The most critical aspect of the exposure is creating and maintaining the posterior retinacular soft tissue sleeve to the lateral femoral head segment.
- Removing 5–8 mm of bone from the posterior margin and the base of the greater trochanter will aid in relieving tension on the vascular flap.
- The determination of the medial and lateral head fragments is performed with femoral head sizing templates and intra-operative three-dimensional analysis.
- The medial longitudinal femoral head osteotomy should leave at least 1/3 of the femoral neck intact.
- The lateral longitudinal femoral head osteotomy should be placed to remove all interval damaged/deformed femoral head; the goal is to reduce the femoral head to a size that is completely contained within the acetabulum.
- The transverse femoral neck osteotomy that creates the free interval and lateral femoral head fragments should not violate the medial femoral neck.
- If the residual femoral neck is less than 1/3 the original diameter, then a retrograde 4.5-mm screw should be inserted from the proximal femoral metaphysis into the medial femoral head fragment.
- The femoral head osteotomy is stabilized with three to four 3.0 mm/4.3 mm headless screws.
- The mobile, lateral femoral head fragment is advanced proximally and medially to the stable medial femoral head fragment; the excised bone should be used as bone graft under the lateral femoral head fragment.
- The hip capsule should be repaired with minimal tension to avoid stretch of the vascular supply to the femoral head.
- The greater trochanter should be advanced until the proximal tip is at or below the center of the femoral head.
- The greater trochanter is repaired with two 4.5-mm or 5.5-mm screws with washers.
- It is critical that the fixation screws for the greater trochanter are directed distal away from the residual medial intact femoral neck to prevent postoperative femoral neck fracture.

Residual Femoral Head Subluxation

Residual femoral head subluxation or migration can result from untreated or unsuccessful treatment of Perthes disease. The residual femoral head subluxation is truly a migration or "settling" of the joint as the femoral head deforms from both internal collapse and abnormal femoral head cartilage growth. This migrated joint position is exacerbated by concurrent soft tissue contractures, typically hip flexion and hip adduction contractures. If this abnormal joint position is prolonged, then permanent femoral head deformity will result. The femoral head that demonstrates significant residual deformity after the reconstitution or healing phase can only be corrected with the femoral head reduction osteotomy described above. However, if the femoral head is still within the active phase of Perthes (fragmentation or early reconstitution) and the hip joint is contracted with residual femoral head migration, then a specialized treatment exists that can salvage this situation. This treatment consists of a hip distraction protocol that consists of arthrodiastasis of the hip joint using a hinged external fixation device and selective soft tissue releases. Although there is limited prospective data on hip arthrodiastasis, the concept of using articulating hip distraction as an ideal means to reduce deforming joint pressures, and also maintain range-of-motion, has been described [59, 60]. Adjunctive procedures include a multiple small diameter core decompression and autogenous bone stem cell graft injection. The author has achieved a high rate of success for the most severe late onset Perthes patients after failed previous treatment, and thus utilizes this protocol for primary treatment of Perthes patients in high-risk groups and those that have failed early conservative treatment methods (Box 29.10).

Hip Distraction Treatment for Residual Femoral Head Subluxation Secondary to Perthes Disease: Operative Technique

Patient Positioning

The patient is placed supine on the operating room table after an epidural catheter is inserted. There are no bumps placed under the patient's hip or sacrum. The involved extremity is sterilely prepared to include the entire lower extremity, gluteal region, and iliac crest as proximal as the subcostal margin. A sterile Mayo stand with a pillow is prepared to allow for positioning the leg in adduction during pertinent parts of the procedure.

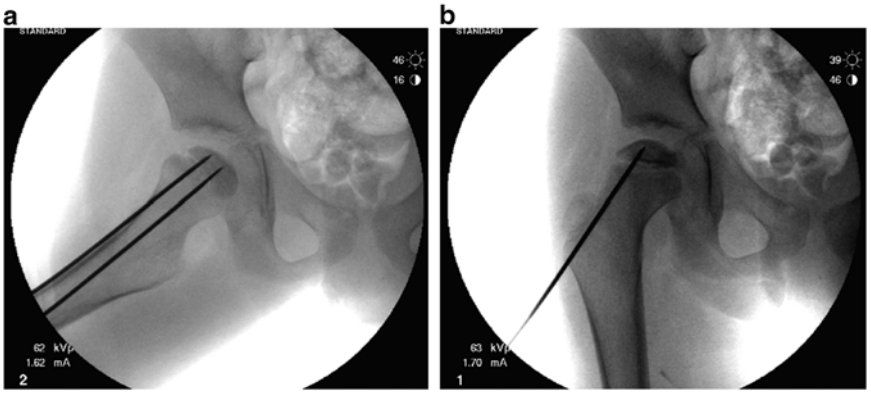


Fig. 29.49 Fluoroscopic views of a small-diameter core decompression. Reprinted with permission from the Rubin Institute for Advanced Orthopedics, Sinai Hospital of Baltimore

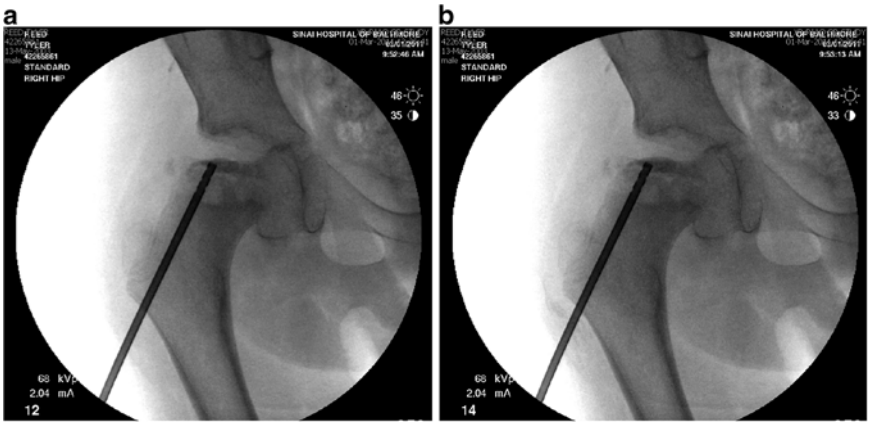


Fig. 29.50 Intraoperative fluoroscopic image demonstrating the insertion of the fenestrated cannula into the femoral epiphysis with injection of bone stem graft. The bone stem cell graft is visualized by adding a small amount of dye, which appears as the darkened epiphysis. Reprinted with permission from the Rubin Institute for Advanced Orthopedics, Sinai Hospital of Baltimore

Core Decompression

The first step of the procedure consists of small diameter core decompression of the femoral head in order to stimulate a healing response within the epiphysis. A 1.8-mm wire is inserted percutaneously from the lateral thigh and driven up the femoral neck under fluoroscopic guidance into the femoral epiphysis (Fig. 29.49).

This is repeated 5–6 times in order to cover all affected zones of the epiphysis.

Stem Cell Injection

The second surgical step begins with harvesting bone marrow aspirate from the iliac crest. A 3-mm trephine needle is used to aspirate approximately 60 cc of bone marrow, followed by centrifuge preparation to separate the bone stem cells. Next, a 1.5-mm wire is driven into the anterolateral portion of the femoral epiphysis. Via a small stab incision, the 1.5-mm wire is overdrilled with a 3.2-mm cannulated drill. A fenestrated cannula is inserted into the bony channel and manually pushed, or carefully tapped with a mallet, into the epiphysis.

The bone stem cell graft is then injected into the femoral epiphysis via the fenestrated cannula. Prior to injection, the bone stem cell graft is diluted with 1 cc of radiopaque dye to allow visualization of the graft distribution under fluoroscopy (Fig. 29.50).

If dye is seen diluting into the joint, the cannula is readjusted as necessary and the injection commences.

Soft Tissue Releases

The leg is placed into a “frog leg” position and an adductor tenotomy is performed through a small stab incision. The adductor longus muscle and gracilis muscle are released during this percutaneous tenotomy. The adductor longus muscle is released with the hip and knee flexed, whereas the gracilis muscle is released with the hip flexed but the knee extended. In a heavier patient, the physician may alternatively make a 2- to 3-cm mini-open approach to perform adductor releases under direct visualization.

Following adductor releases, the leg is placed back into a neutral supine position and a 3-cm anterior oblique incision is performed in the flexion crease of the groin approximately 2 cm below the anterior superior iliac spine. The incision

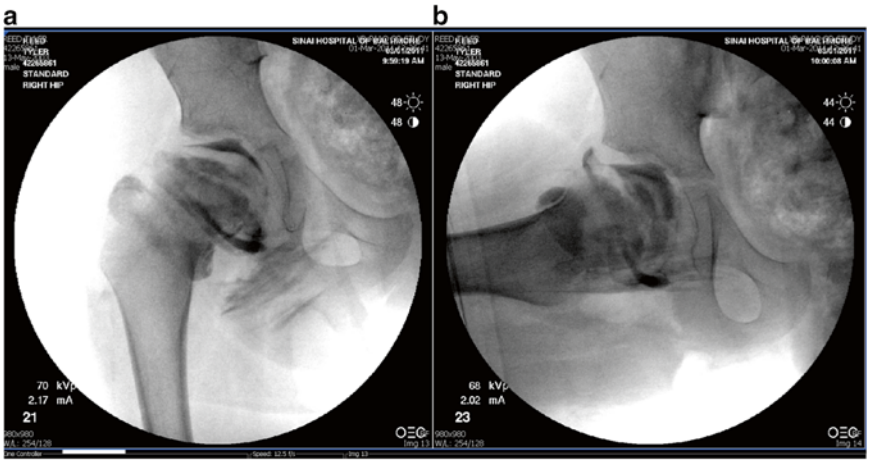


Fig. 29.51 Intraoperative fluoroscopic views of the right hip after intra-articular injection of dye. The actual femoral head deformity can be visualized and documented. Reprinted with permission from the Rubin Institute for Advanced Orthopedics, Sinai Hospital of Baltimore

starts medially at the point of the palpable femoral artery and extends laterally. The interval between the sartorius muscle and tensor fascia lata muscle is palpated and the fascia overlying the sartorius muscle is opened. This fascia is divided from lateral to medial until the septum separating the sartorius muscle and iliopsoas muscle is identified. The fascia overlying the iliopsoas muscle is divided and the muscle is carefully retracted laterally. By gently pulling the iliopsoas muscle laterally, the femoral nerve comes into the operative field and is easily identified. The fascia overlying the femoral nerve is divided and the medial and lateral borders of the nerve are dissected. The overlying fascia proximal and distal along the nerve is released allowing the nerve to mobilize. The femoral nerve is retracted medially and the iliopsoas muscle is retracted laterally and everted exposing the iliopsoas tendon. The tendon is lifted into the operative field with a right angle clamp and sharply released under direct visualization. The incisions are closed in a typical layered fashion and an arthrogram of the hip is performed. The hip is examined under fluoroscopy to determine the exact shape of the femoral head, position of the labrum, and containment of the femoral head with 20° of hip abduction (Fig. 29.51).

Hip Distractor Application

The hinged external fixation device (Orthofix articulated hip distractor) is placed by inserting half pins into the pelvis and into the middle third of the femoral shaft. The leg is positioned on the prepared mayo stand with the hip in full extension, neutral rotation, and 20° of abduction. A 3/32 Steinman pin is inserted perpendicular to the anatomic axis of the femoral shaft 8–10 mm below the center of the femoral head, and serves as the temporary hinge guide pin In the sagittal view, the position of the guide pin is confirmed to be in the center of the femoral head. The hinged external fixation device is placed over the hinge guide pin. The proximal arch is secured to the pelvis by two supra-acetabular half pins. The distal pin clamp is utilized for placement of two half pins into the femoral shaft. Avoid placing the distal half pin anteriorly near the cortex, as this places the patient at risk for femoral shaft fracture. After securing the device to the half pins, a second arch is secured to the distal half pins. The hinge pin may now be removed and fluidity of the articulation checked by passive hip flexion and extension to ensure a properly applied frame. The final femoral fixation point is placed at the level of the lesser trochanter built off the posterolateral distal arch. The final pelvic fixation point is inserted from an oblique anterior to posterior direction under fluoroscopic control down the posterior column of the pelvis (Fig. 29.52).

After completing external fixator application, the hip is acutely distracted 8–10 mm. A modular, removable anterior hip extension bar is attached to the anterior portion of the proximal and distal arches.

Postoperative Care

The postoperative protocol consists of continued gradual distraction at the rate of .5 mm per day for 7–10 days if needed. The goal of distraction is to over reduce Shenton's line by 5–10 mm. In smaller patients, the acute intra-operative distraction is all that is needed. Physical therapy is employed to maintain hip flexion and extension and mobilization with 50 % partial weight bearing. The anterior extension bar is used for 2–3 h during the day and all night for the duration of the external fixation. The external fixation device is maintained for 4 months. Frame removal is performed under general anesthesia with a repeat arthrogram and Botox injection into the adductor longus and gracilis muscles. The dosage of Botox is typically 10 units per kilogram of weight, with a max dose of 200 units. Post-frame removal protocol consists of abduction hip bracing with a Scottish Rite brace and hip mobilization with physiotherapy and home exercises (Fig. 29.53).



Fig. 29.52 (**a**) An AP pelvis radiograph demonstrating a typical articulated hip external fixation device application for arthrodiastasis. (**b**) Another example of articulated hip distraction demonstrating an adequate distraction gap of 8–10 mm. (**c**) Clinical picture of the articulated hip distracting external fixation (Orthofix). Reprinted with permission from the Rubin Institute for Advanced Orthopedics, Sinai Hospital of Baltimore

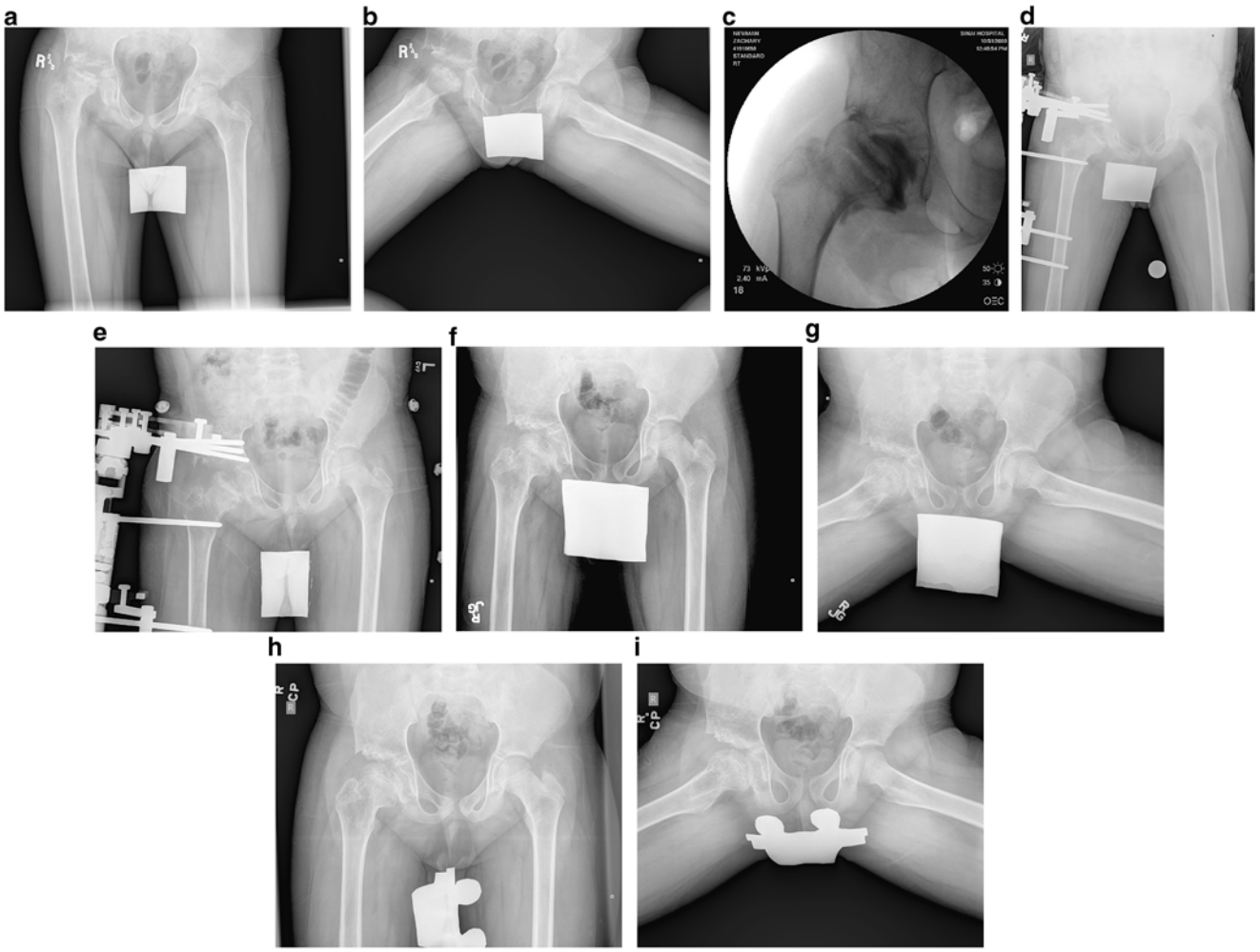


Fig. 29.53 (**a**, **b**) An 11-year-old male with severe residual deformity in late fragmentation stage of Perthes disease; the patient had failed a previous shelf arthroplasty at an outside institution. (**c**) Intraoperative arthrogram demonstrating lateral and proximal migration of the femoral head and true "hinge abduction." (**d**) Radiograph 6 weeks following core decompression, open iliopsoas muscle release, percutaneous hip adductor tenotomy, and articulated hip distraction with full reduction of the hip joint and early reossification. (**e**) AP pelvis radiograph after 4 months of joint distraction prior to removal of external fixation; the radiograph demonstrates significant reossification of the femoral head. (**f**, **g**) One year postoperative AP and frog pelvis radiographs demonstrating excellent healing and sphericity of the femoral head with concurrent excellent congruency of the hip joint. (**h**, **i**) Three-year follow-up with excellent right hip healing and congruency; the patient continues to be pain free and participates in all activities. Reprinted with permission from the Rubin Institute for Advanced Orthopedics, Sinai Hospital of Baltimore

Box 29.10. Hip Distraction with a Hinged External Frame

Technical tips:

- Arthrodiastasis is best performed simultaneously with adjunctive procedures: soft tissue contracture releases, core decompression, autograft bone stem cell injection.
- Six multiplanar 6-mm half-pins are used to stabilize the fixator: two supra-acetabular pins, one oblique posterior column pin, two femoral shaft pins, and one posterolateral pin at the lesser trochanter.
- Acutely distract the hip 8–10 mm, followed for gradual distraction as needed to achieve a distraction gap of 5–10 mm.
- A removable, modular extension bar will help prevent hip flexion contracture; the hip extension bar is inserted for 1–2 h during the day and all night
- Postoperative physiotherapy both prior to and after fame removal is paramount to optimizing outcome.
- After the external fixation is removed, the adductor muscles are injected with Botox, a Scottish Rite hip abduction orthosis is placed, and immediate mobilization with "Perthes exercises" are performed.

Summary

Residual hip deformities may present in any pediatric age group as a result of persistent, recurrent, iatrogenic, or neglected disease. The complex, multiplanar nature of these deformities are challenging for the treating surgeon.

Acetabuloplasty techniques continue to be modified to provide better outcomes for developmental dysplasia of the hip, but emphasis must also be placed on improving screening and early, less invasive treatment with the hope of preventing residual dysplasia. A modernized triple osteotomy is an ideal procedure to reorient a dysplastic acetabulum and regain adequate femoral head coverage in a child approaching or following closure of the triradiate cartilage.

Residual deformity from moderate and severe slipped capital femoral epiphysis leads to both short term and long term sequelae ranging from pain, functional deficit, impingement, and eventual degenerative arthritis. Improvements in surgical approach have led to more aggressive surgical treatment to correct associated femoral neck deformity through the modified Dunn procedure. Long-term studies may emerge in the future to provide surgeons more guidance on whether subcapital or intertrochanteric osteotomies are the ideal treatment of a chronic slipped capital deformity.

Achieving consistent excellent results in treating avascular necrosis of the immature femoral head continues to elude the pediatric orthopedic community. Surgical intervention during the early fragmentation stage of Perthes may be crucial to preserving femoral head morphology and improving long-term outcome. Currently, an appropriately applied hip distraction protocol may be the only treatment method that simultaneously provides containment as well as relief from deforming joint pressures, while allowing maintenance of range-of-motion. Further investigation is needed into hip arthrodiastasis and adjunctive treatments such as core decompression and stem cell injection to optimize the treatment protocol described in this chapter.

Avascular necrosis of the femoral head leads to proximal femur deformities, namely coxa breva and coxa magna. Coxa breva negatively alters proximal femur biomechanics, and is associated with hip abductor weakness, limb length discrepancy, and limping. In contrast to a relative neck lengthening procedure, the Morscher osteotomy is a true neck lengthening osteotomy and directly addresses the femoral neck deficiency. Intracapsular femoral head reshaping osteotomies are technically demanding, but provide the only reconstructive option for a symptomatic coxa magna in the severely deformed and incongruent femoral head.

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Posttraumatic Lower Limb Deformities in Children

30

Ashok N. Johari, Sandeep A. Patwardhan, and Taral Vishanji Nagda

Introduction

Posttraumatic deformities in children are unique. They are different from deformities that are congenital or secondary to other causes since the skeletal and soft-tissue anatomy and physiology are often normal to start with. Hence, reparative mechanisms and many management principles differ in these patients.

Posttraumatic deformities in children are different from adults. Children have the potential to influence future growth due to the possibility of substantial remodeling of a malunited fracture. Such remodeling can occur from different sources including the adjacent physis and differential periosteal growth of the affected diaphysis (Fig. 30.1). On the other hand, injury to a portion of the growth plate can lead to asymmetric growth of the physis with progressive deformities in children.

Deformities following trauma to the extremities are not uncommon and not always dependent on the level of care provided. While some deformities are inevitable due to high-energy trauma and damage to the growing area of long bones, other deformities occur because of suboptimal management allowing the limb to be positioned abnormally or allowing certain muscles to get tight and create a secondary deformity.

Deformities that are not clinically significant or have the potential for substantial remodeling to an acceptable level can be observed. In general, when the residual axial malalignment of a lower extremity long bone exceeds 10°, correction may be warranted.

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Causation and Etiological Factors

Box 30.1. Factors Related to Development of Posttraumatic Deformities

- Injury related
  - Soft tissue
    - Skin and soft-tissue loss
    - Tendon and muscle injury
    - Nerve injury
    - Vascular injury
  - Skeletal
    - Type of fracture, fracture geometry, and comminution
    - Growth plate injury
    - Bone loss
    - Intra-articular injury
- Treatment related
  - Missed diagnosis of a fracture
  - Delayed presentation and management
  - Incorrect treatment choice
  - Suboptimal management
  - Overtreatment
  - Iatrogenic problems
- Complication related
  - Infection
  - Loss of reduction
  - Compartment syndrome
  - Postoperative stiffness
  - Avascular necrosis

(continued)

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Box 30.1. (continued)
- Patient-related factors
  - Preexisting skeletal problems like dysplasia, syndromes
  - Neuromuscular problems like cerebral palsy, poliomyelitis, muscular dystrophy
  - Altered bone physiology due to osteogenesis imperfecta, metabolic bone disease
  - Pathological fracture with underlying tumor, osteomyelitis
  - Immunocompromise
  - Malnutrition



Fig. 30.1 Four-year-old child with femur fracture treated by traction in a Thomas' splint. (**a**) Injury film. (**b**) X-ray in Thomas's splint. (**c**) At 2 months fracture united with 40° angulation and 3 cm shortening. The malunion is due to muscle forces that were not effectively managed

A posttraumatic deformity can be a result of initial soft-tissue and skeletal injury, its management, or complications. In order to accurately identify the cause and anticipate the natural progression of the deformity, it is important to ascertain the mechanism of injury, initial soft-tissue and bony injury, specifics of the treatment given—conservative and operative, postoperative care, and complications and course of the deformity thus far.

Box 30.2. Etiological Mechanisms for Posttraumatic Deformities
- Postural or positional
- Muscle forces
- Neural and vascular injury
- Poor skeletal stabilization
- Damage to the growing area of bone
- Infection

Deformities can be caused by one or more of the following etiological factors:

1. Postural or positional: Lack of immobilization or faulty immobilization may cause a deformity, often related to contracture of the adjacent soft tissues. Examples are an equinus deformity of the foot or a knee flexion deformity because of contracture of the gastro-soleus or the hamstrings, respectively.
2. Muscle forces: Varus deformity of the upper femur following a fracture is a classic example of muscle forces at work. The flexor-adductor predominance over the abductor-extensor forces causes this deformity (e.g., Figs. 30.1 and 30.2).
3. Neural or vascular injury: Neural injury can create deformity because of imbalance of muscle forces or by muscle paralysis causing a postural deformity. Vascular injury may give rise to infarction of muscle followed by its fibrosis and eventually a contracture. Volkmann's ischaemic contracture is a typical example of such an etiology.
4. Poor skeletal stabilization, loss of reduction, and loss of fixation can all give rise to posttraumatic deformities. Factors such as poor bone quality, traumatic bone loss, and fracture comminution may contribute to inadequate skeletal stabilization (e.g., see Fig. 30.9).
5. Damage to the growing area of bone: Classic examples of these are partial physeal injuries with growth arrest giving rise to various angular deformities (e.g., see Fig. 30.7). Symmetric physeal damage would give rise to limb shortening. Damage to the growing area may be a consequence of the mode of injury or may occur because of its poor management. Salter–Harris type 6 injury is a perichondrial ring injury presenting with a normal X-ray. This is an example of the mode of injury giving rise to deformity (e.g., see Fig. 30.10).
6. Infection: Open skeletal injuries or iatrogenic issues can be complicated by infection which works by damaging bone and cartilage, causing bone lysis and loss, growth plate damage and its sequelae, as well as joint ankylosis and deformity.

Classification

Posttraumatic deformities can be classified into different ways to fully understand the current problems as well as potential issues that may arise in the future. Such classification helps in formulating the management plan for the individual patient.

Box 30.3. Classification of Posttraumatic Deformities in Children

Related to the area of bone affected
- Diaphyseal
- Metaphyseal
- Physeal
- Epiphyseal

Related to the anatomical effect
- Angulation
- Rotation
- Limb length
- Nonunion
- Restriction of movement
- Muscle weakness

Related to the tissue affected
- Soft tissue
- Skin, fascia, muscle, capsule, or ligaments
- Skeletal
- Diaphyseal, growth plate, articular

Related to the progression
- Progressive: Improving: Partial or complete
- Progressive: Improving, then progressive

Clinical Manifestations

Fixed deformity of the joint, joint stiffness and ankylosis, shortening, angular and torsional deformities, and axial malalignment are all potential clinical manifestations in a posttraumatic situation. Meticulous clinical assessment may point to the static or progressive nature of the deformity. Presence or absence of infection is to be noted as it is an important consideration in management. Examination for neural injury is important and can be challenging in young children, especially in the presence of fixed contractures.

Investigations

Besides plain X-rays, MRI and CT scans may be useful in some patients.

Plain X-rays are the simplest way of monitoring bony deformity. In addition to standard views one may need X-rays in the plane of the deformity. Full-length standing radiographs and scanograms are useful for monitoring limb length.

MRI can give a lot of information about the growth plate (e.g., see Fig. 30.7f, g) and articular cartilage, periarticular ligamentous injury, osteochondral fractures, and articular alignment.

CT scans help define the fracture geometry, area of growth arrest, and presence of a bony bar. It is also a useful tool to quantify torsional deformities.

Occasionally, specialized scans like the radioisotope scans may be required to detect infection. Hematologic tests including a white cell count, sedimentation rate, and C-reactive protein are helpful in such a situation. A more comprehensive metabolic panel may be required in patients with underlying metabolic bone diseases such as various forms of rickets.

Remodeling of Bony Deformities

Remodeling may impact decision making for correcting any bone deformity. The patient's functional capacity and the surgeon's experience should also be factors in determining whether to depend on the remodeling capacity of the specific fracture or to consider performing a more aggressive, invasive technique to achieve a satisfactory result [1].

In the typical long bone, 75 % of the remodeling occurs by reorientation of the physis while appositional remodeling of the diaphysis can only be expected to contribute 25 % to the remodeling process.

Important factors influencing remodeling are as follows:

1. Age of the patient: The older the child the less is the remodeling potential. In general, remodeling of posttraumatic deformities occurs more effectively in children younger than 10 years.
2. Location of the malunion: Metaphyseal fractures remodel better than diaphyseal fractures. Fractures near the fast-growing physes such as lower femur and upper tibia and lower radius and upper humerus remodel better than their counterparts.
3. The plane of the deformity: Angulation in the plane of the joint axis corrects better. Rotations do not correct for all practical purposes. In some fractures overgrowth phenomenon gives rise to correction of limb shortening.
4. Magnitude of angulation and translation.

Some guidelines on acceptability of fracture reduction and remodeling are given at the end of this chapter (see section Acceptability Criteria for Reduction in Lower Limb Fractures).

Box 30.4. Factors Affecting Remodeling of Fractures in Children
- Age
- Location
- Plane of deformity
- Degree of deformity

Case Study (Fig. 30.2)

Clinical Summary

A 4-year-old child was treated for fracture shaft femur by immediate spica. He had 2-cm shortening and varus deformity 3 months post-fracture. Over a period of 2 years, the deformity corrected and child had equal leg lengths.

Philosophy of Treatment

Fracture healing can stimulate bone growth in certain femoral shaft fractures. The amount of overgrowth varies in different reports from 0.4 to 2.7 cm. Overgrowth in femoral fractures appears to be independent of age, fracture level, and position of the fracture at the time of healing. The effect of growth stimulation may continue for up to 3 years following the fracture.

Case Study (Fig. 30.3)

Clinical Summary

A 6-year-old girl presented 4 months after an open fracture of the tibial tuberosity with patellar tendon tear which was debrided, repaired, and cast. She developed a stiff knee for which the original surgeon did a manipulation under anaesthesia. However, the local swelling and deformity increased. A Salter–Harris type 1 injury was diagnosed at presentation. An MRI revealed a posterior displacement of the distal femoral epiphysis with loculated fluid collection along the posterior border.

Different options of management were considered but it was decided to leave her alone in view of the good possibility of remodeling. For the stiff knee physical therapy with range of motion exercises was started. The sequential X-rays show the excellent remodeling (Table 30.1). She regained nearly full knee movement 2 years after the injury.

Philosophy of Management

The distal femoral epiphysis is one of the fastest growing physes with rapid remodeling in children. Undisplaced Salter–Harris type 1 injuries need not be fixed, as long as they are immobilized with restricted weight bearing and closely followed with serial radiographs. The ones with displacement may need a gentle reduction and fixation. We prefer that pins not be introduced from the knee level upwards but rather in a reverse fashion from the metaphysis into the epiphysis as

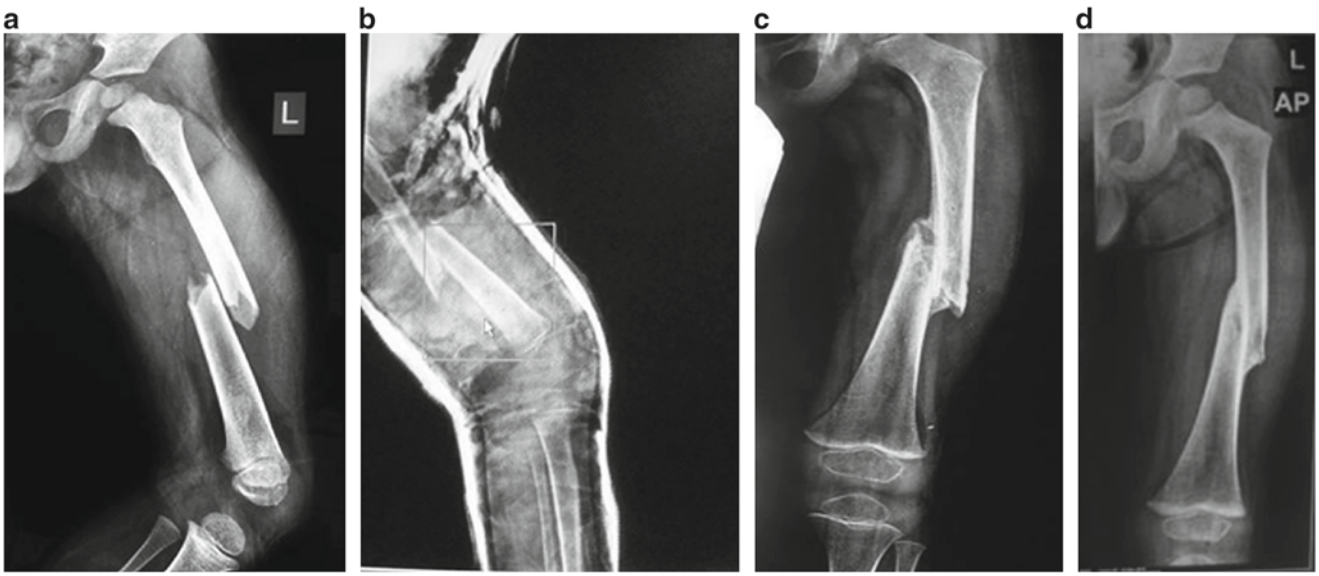


Fig. 30.2 Effect of remodeling and growth: Fracture shaft femur in a 4-year-old child. (**a**) Injury film. (**b**) Position in plaster after immediate spica showing shortening and recurvatum. (**c**) At 6 weeks, uniting with 20 varus and 2.5 cm shortening. (**d**) At 3 months, united with persistent shortening. (**e**) After 2 years, correction of deformity with equal limb lengths



Fig. 30.3 A 6-year-old with malunited type 1 SH fracture. (**a**) Four months from injury, Feb 2011. (**b**) MRI showing posterior displacement of the physis. (**c**, **d**) X-ray and clinical picture 5 months postinjury. (**e**) X-rays 10 months postinjury showing remodeling and correction of angulation and translation. (**f**) Complete remodeling at 1.5 years postinjury in 2012

Table 30.1 Diaphyseal-epiphyseal angle

Feb 2011	38°
Mar 2011	42°
May 2011	54°
Aug 2011	70°
Nov 2011	82°
Apr 2012	90°

recommended by Wall [2]. It is important not to leave pins sticking out of the skin if placed retrogradely. If placed in this fashion they should be buried under the skin for later removal.

Management

Decision making in posttraumatic deformity situations is based on a number of factors, including:

- Age of the patient
- Location of the deformity
- Morphological status of bone, soft tissue, and cartilage
- Functional loss and impairment
- Abnormal mechanical loading with risk of joint degeneration
- Cosmetic aspect

Table 30.2 presents a general schema of the types of interventions.

Important Posttraumatic Deformities: Lower Limb

In this section we briefly discuss some unique posttraumatic lower limb deformities in children and demonstrate a variety of treatment strategies. However, as noted earlier, the decision making should be individualized based on patient, surgeon, and environmental factors.

Posttraumatic Chondrolysis of the Hip and Avascular Necrosis

Acetabular injuries may be complicated by chondrolysis of the hip, and this may give rise to a fixed flexion and adduction deformity. Likewise hip dislocations and fractures of the femoral neck may be complicated by avascular necrosis. Avascular necrosis may follow the commonly described patterns in the classifications of Kalamchi and

Table 30.2 General schema of the types of interventions

Type of deformity	Nature of intervention
Joint contractures	Manipulation
	Splintage
	Serial plasters
	Soft-tissue release
	External fixation
	Osteotomy
	Arthrodesis
Muscle imbalance	Splintage
	Tendon transfer
	Arthrodesis
Angular and rotational bony deformities	Osteotomy
	Growth modulation
	Physeal bar excision
Shortening	Shoe raise
	Limb lengthening
	Contralateral epiphysiodesis
	Limb shortening
Bone loss and nonunion	Bone grafting
	Bone transport

MacEwen [3] or of Bucholz and Ogden [4] and management would follow the lines described for such conditions.

Arthrodiatasis [5, 6] is a useful modality for managing posttraumatic chondrolysis and stiffness of the hip and an example is presented here.

Case Study (Fig. 30.4)

Clinical Summary

A 15-year-old boy sustained a motor cycle accident in 1999. He suffered a fracture of the medial wall of the acetabulum which was managed conservatively by traction. He gradually developed a fixed flexion and adduction deformity of the hip along with stiffness and awkward gait. He presented 2 years later for this problem having tried all conservative measures and physiotherapy. X-rays revealed narrowing of the right hip joint space and MRI was not suggestive of infection or inflammation.

Philosophy of Treatment

He was managed by an adductor release and hip distraction. The flexion adduction deformity improved and he gradually regained his hip motion. The hinged distractor was in place for 3 months and physiotherapy continued. On follow-up 8 years later he still retained excellent motion at the hip and was able to squat and sit cross legged.

Posttraumatic Coxa Vara

Posttraumatic coxa vara may be caused by a malunion or even a nonunion of the proximal femur. Shear forces at the fracture site caused by the body weight and muscle forces may be responsible for the mal- or nonunion.

Restoration of the normal neck–shaft angle is important biomechanically and this would require correction of the malunion by a valgus osteotomy. For a nonunion osteosynthesis may include internal fixation with or without bone grafting and a valgus osteotomy.

Case Study (Fig. 30.5)

Clinical Summary

Case of basicervical fracture of the femur in an 8-year-old child treated conservatively who presented with a limp related to a coxa vara deformity: The deformity was non-progressive and uniplanar with the apex at the neck of femur.

Philosophy of Treatment

Both deformity correction and nonunion of femur neck fracture were treated with a valgus osteotomy performed at the subtrochanteric level and fixation with a plate.

Malunited Shaft Femur or Pseudarthrosis with Deformity

Either situation will demand skeletal realignment and stabilization by some form of internal or external fixation with or without bone grafting depending on the nature of the pseudarthrosis.



Fig. 30.4 Hip deformity due to posttraumatic chondrolysis in a 15-year-old. (**a**) Injury film showing acetabular fracture. (**b**) X-ray 2 months later, showing healed fracture. (**c**) 1.5 years later showing the marked adduction of the hip and loss of joint space. (**d**) Following arthrodiatasis with an adductor release nearly 2 years postaccident. (**e**) X-rays 8 years postinjury. (**f**) Clinical picture at this time

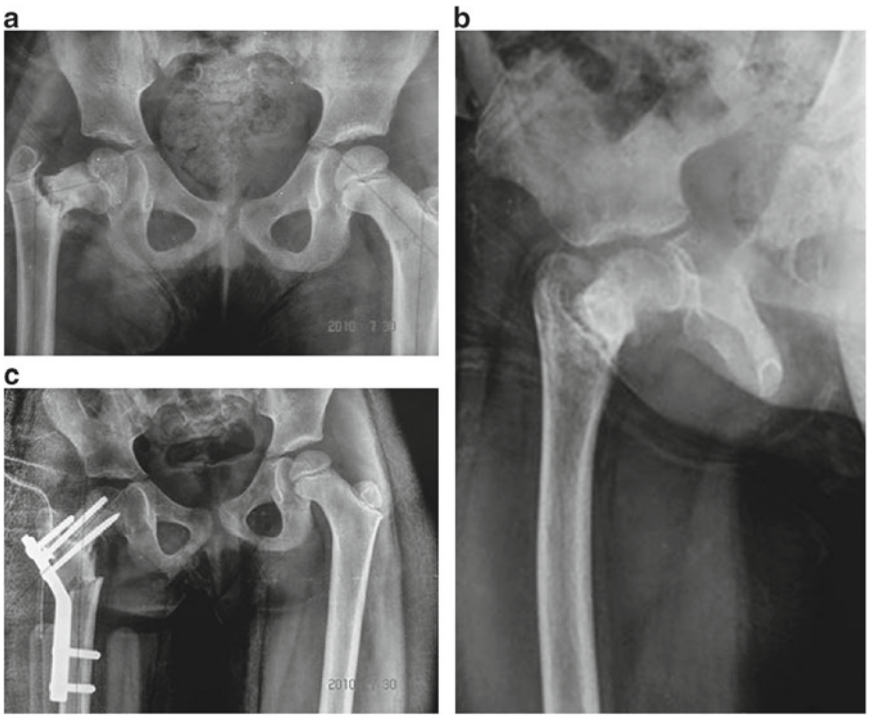


Fig. 30.5 Posttraumatic coxa vara due to fracture neck femur treated in spica. (**a**) Fibrous nonunion in varus with a neck–shaft angle of 100°. (**b**) Lateral X-ray showing ununited fracture line. (**c**) In situ fixation of nonunion with subtrochanteric valgus osteotomy. (**d**) Healing of fracture as well as osteotomy in 3.5 months

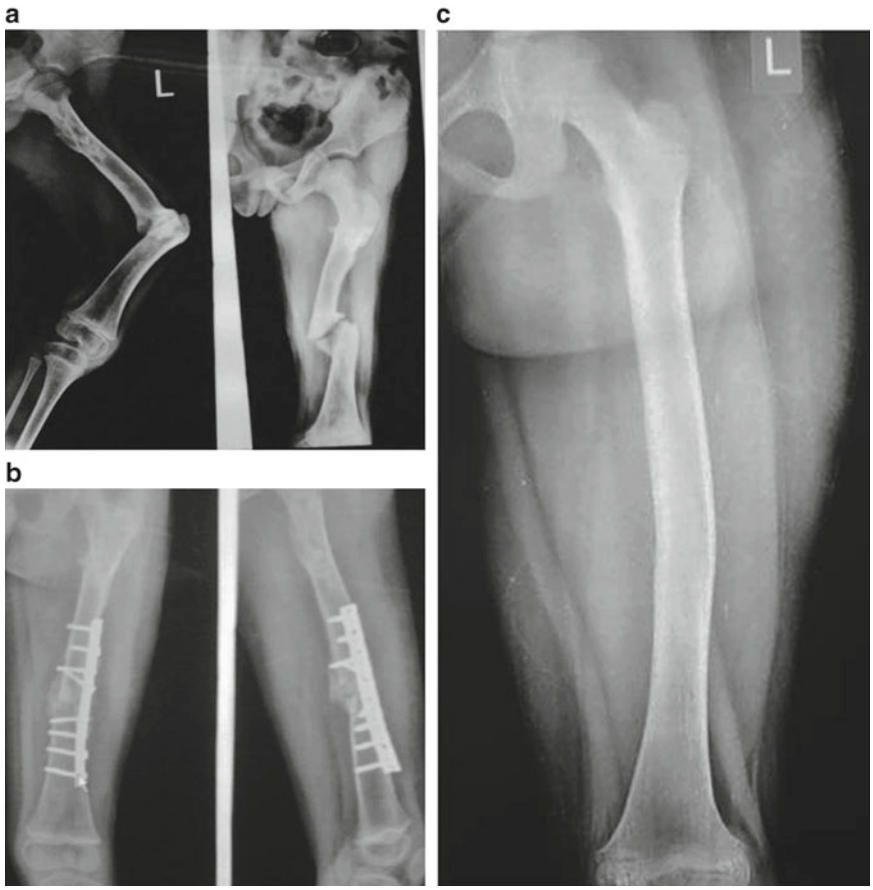


Fig. 30.6 Maluniting fracture shaft femur. (**a**) Fibrous nonunion shaft of femur with triplanar deformation. (**b**) Excision of fibrous tissue, deformity correction at apex, plating without bone grafting. (**c**) Final healing after plate removal at 1 year

Case Study (Fig. 30.6)

Clinical Summary

Fracture femur shaft which was treated conservatively presented with fibrous nonunion with unacceptable deformity at the fracture level.

The deformity was a triplanar deformity with malrotation of the femur, procurvatum of 90° and translation of about 1 cm, and valgus angulation at the fracture site.

Philosophy of Treatment

Fibrous nonunion of shaft of femur was treated with resection and freshening of the edges and fixation along with bone grafting. A corrective osteotomy was performed at the apex of deformity, i.e., at the fracture site, and fixation was done by a plate.

Distal Femoral Deformity Following Iatrogenic Physeal Injury

Growth plate can get injured during insertion of elastic nailing for fracture shaft femur or during implant removal. To avoid this problem it is recommended that the entry point is at least 5 cm away from the growth plate. Damage to the peripheral growth plate and a resultant bar can give rise to progressive angular deformity.

Excision of bar is indicated if there is at least 2-year growth left and area of the bar is less than 30 % of total physeal area. The success of the surgery depends on the age of the patient, etiology, area of the bar, the bone involved, and the location of the bar [7] central or peripheral. The deformities if more than 30° are treated by osteotomy but if less than 30° can often be treated by simultaneous growth modulation surgery.

Case Study (Fig. 30.7)

Clinical Summary

A 11-year-old girl with progressive posttraumatic genu valgum: She had sustained a fracture shaft femur at age 9 years which was treated elsewhere with a retrograde Rush nail.

A progressive deformity was noted after removal of the nail. X-rays revealed a lateral growth arrest. MRI showed peripheral physeal arrest with area less than 20 %.

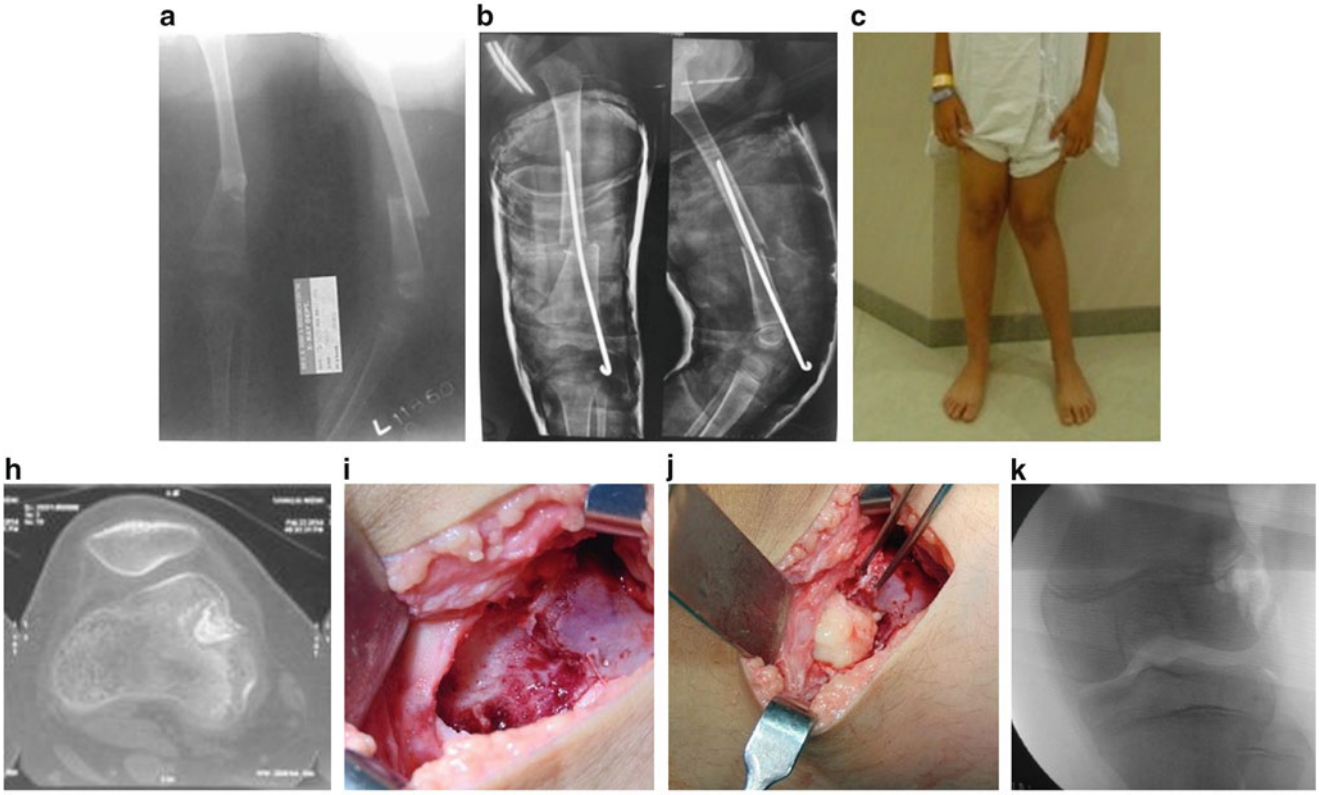


Fig. 30.7 A 11-year-old with fracture shaft femur treated by intramedullary nail with physeal injury of lower femur and genu valgum. (**a**) Injury film. (**b**) X-rays showing fixation with a single intramedullary nail from lateral aspect. (**c**–**e**) Clinical photos and X-ray scanogram showing the deformity. (**f**, **g**) Gradient echo fat suppression MRI image showing lateral growth arrest due to physeal bar. (**h**) Mapping of the physeal arrest using a CT scan. (**i**) Intraoperative photo showing the excision of bony bar using a burr. (**j**) Interposition of the defect with subcutaneous gluteal fat. (**k**) Intraoperative image to confirm adequacy of the bar excision. (**l**) Postoperative X-ray showing hemiepiphysiodesis plate and markers. (**m**, **n**) Nine months postoperative clinical photo and X-rays showing resumption of growth and correction of deformity

Philosophy of Treatment

As she had more than 2 years of growth available and area of the bar was less than 30 % of the underlying physis, she was treated with excision of physeal bar and fat interposition. At the same time she underwent growth modulation by plate hemiepiphysiodesis on medial side.

The medial hemiepiphysiodesis corrected the growth with restoration of lateral growth by physeal bar excision. At 1 year the restoration of growth was evident by diverging screws of hemiepiphysiodesis plate and increase in the distance between markers.

Physeal Injury of the Distal Femur with Angular Deformity

Physeal injuries of the distal femur are notorious for the production of deformity. Serious damage to the physis can occur giving rise to angular deformity and/or shortening. The incidence of angular deformity in such cases is 18–51 % in different series and that of shortening is 36–68 % [7–10]. The risk of growth arrest has been reported to be between 40 and 52 % [11, 12]. Salter–Harris (SH) 1 injuries had the lowest incidence of growth disturbance (36 %), SH2 58 %, and SH 3 49 %, whereas SH 4 injuries had the highest rate of growth disturbance at 64 % [11].

Physeal injuries require very accurate repositioning of the growth plate and restoration of the articular alignment as growth disturbances can result in significant angular deformity. Both SH classification and displacement of the fracture are significant predictors of the final outcome. The treatment method may influence the final outcome [13]. In relation to Salter–Harris type 2 injuries, Ilharreborde and colleagues stated that those with comminution and displacement had a greater risk of growth arrest (75 %) compared with those without these risk factors (38 %) [14].

In a posttrauma situation angular deformities in children are usually caused by a physeal injury. Such deformities can be addressed by a hemiepiphysiodesis procedure utilizing staples, transphyseal screws, or extra-periosteal tension band plates. Appropriate selection of the age for intervention and time for removal of growth modulation devices is important. It is important to avoid repeat corrections via multiple osteotomies and hold the deformity constant in a progressive

situation. Final correction of deformity and limb length discrepancy can be done at skeletal maturity.

Accurate reduction of a physeal injury is mandatory and all efforts must be made to prevent articular step-off. Intra-articular injuries should be open reduced and stabilized and patients with physeal injuries should undergo rapid mobilization of the adjacent joints, generally by the fourth week postinjury, unless the fracture is very comminuted.

Case Study (Fig. 30.8)

Clinical Summary

A 16-year-old boy had an open Salter–Harris type 2 injury of the left distal femur at 1.5 years age. The injury was debrided, open reduced, and fixed with a cancellous screw. Eight months later a valgus deformity of the knee was noted that was gradually progressive. At 10 years from injury, the deformity was progressive and very noticeable. A CT scan revealed the growth disturbance at the distal femur.

Philosophy of Management

An extra-periosteal tension band plate medial hemiepiphysiodesis was done to prevent further angulation. The deformity remained unchanged till skeletal maturity but a residual shortening of 7 cm resulted. Realignment was done by a distal femoral osteotomy. The marked shortening was addressed by a simultaneous gradual lengthening of the femur using external fixation

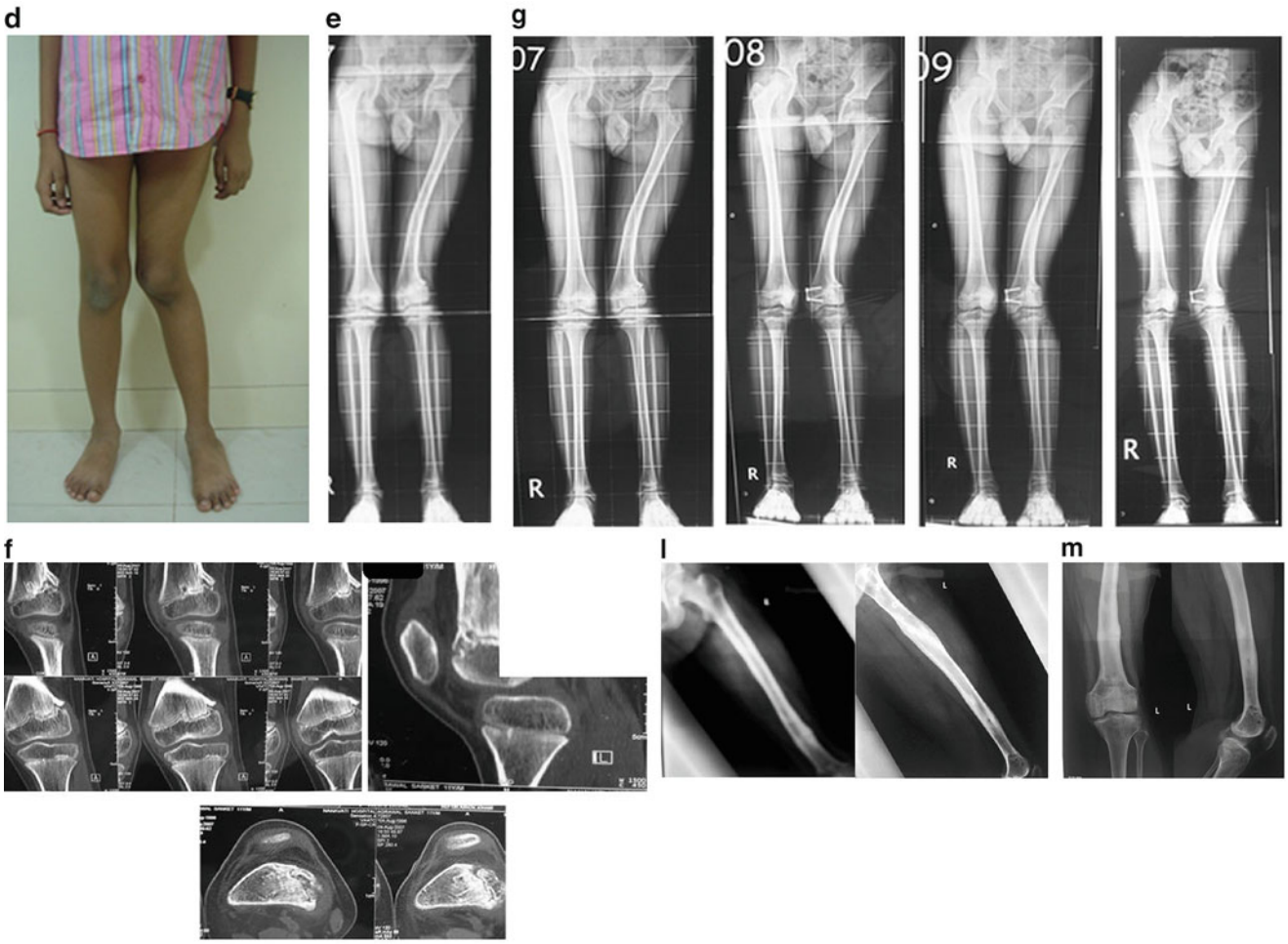


Fig. 30.8 Posttraumatic genu valgum and femoral shortening due to type 2 SH physeal injury of lower femur. (**a**) Injury film 1998. (**b**) Postoperative X-ray showing fixation with screw. (**c**) X-ray taken 6 months post-op showing valgus deformity. (**d**, **e**) Clinical photo and X-ray in 2007, 9 years postinjury showing progressive genu valgum. (**f**) CT shows extensive lateral physeal arrest involving more than 50 % of the growth plate. (**g**) Hemiepiphysiodesis in 2007 using plates. This prevents progression of deformity holding LDFA to 66°. (**h**) Clinical picture, 2010. (**i**) Underwent medial closing-wedge distal femoral corrective osteotomy with limb lengthening proximally using external fixation. (**j**, **k**) X-ray at the end of lengthening after consolidation of the regenerate. (**l**, **m**) 2014 final follow-up showing correction of limb lengths and deformity

Malunited Intra-articular Fractures

If presenting early, maluniting fractures can be addressed with open reduction and the osteochondral fragments repositioned to restore articular and physeal alignment. Late presentations may be fraught with the risks of poor results as articular congruity may be difficult to restore.

Case Study (Fig. 30.9)

A 17-year-old boy sustained a comminuted fracture of the upper tibia. This was treated by internal fixation. The reduction and fixation were suboptimal. The fracture malunited and he presented after implant removal for deformity and loss of motion of the left knee. At this stage management options such as arthrodesis and deformity correction were discussed but the family refused further treatment.

Physeal Injury to Proximal Tibia

Injury to proximal tibial physis is less common than the distal femur but can produce progressive deformities as it is also a fast-growing physis. The management options include excision of the physeal bar coupled with osteotomy or growth modulation. In case where growth cannot be restored, acute angular correction with an osteotomy combined with an epiphysiodesis (to prevent recurrent deformity with future growth) or limb lengthening at a later date can be utilized.

Case Study (Fig. 30.10)

Clinical Summary

A 11-year-old with progressive tibial varus presented with blunt vehicular trauma with normal X-rays 2 years earlier.

He developed progressive tibial varus due to probably a Salter–Harris type 6 medial proximal tibial physeal injury indicating injury to perichondrial ring.

Philosophy of Treatment

The echogradient fat suppression MRI showed that the area of the bar was less than 25 % of the surface area of the underlying physis. He was 11 years and had more than 2 years of active growth remaining. As he fulfilled the criteria for growth restoration, he was treated with physeal bar excision and interposition of bone cement in the defect to prevent reformation of the bar. Growth modulation was added on lateral side with hemiepiphysiodesis plate to correct the deformity.

Case Study (Fig. 30.11)

Clinical Summary

Case of 17-year-old boy who had a fracture of the upper end of tibia at the age of 10 years for which he was treated with above knee cast: He probably had an anterolateral proximal tibial physeal arrest which was undiagnosed. It resulted in progressive recurvatum and valgus deformity.

After 2 years of the injury, the patient presented with a deformity of genu recurvatum which was progressive in nature. There was anteroposterior instability of the knee with partial subluxation of the joint. Radiological examination showed anterior physeal arrest of the proximal tibial physis.

The apex of the deformity was located at the upper end of the tibia.

Philosophy of Treatment

Very high tibial open-wedge osteotomy at apex to correct deformity and filling up of the defect with bone graft: The deformity correction was done acutely with an anterior open-wedge osteotomy at the upper end of the tibia. The osteotomy was stabilized with a plate. Tricortical iliac crest graft which was appropriately shaped was used. Additional tricalcium phosphate interposition material was used.

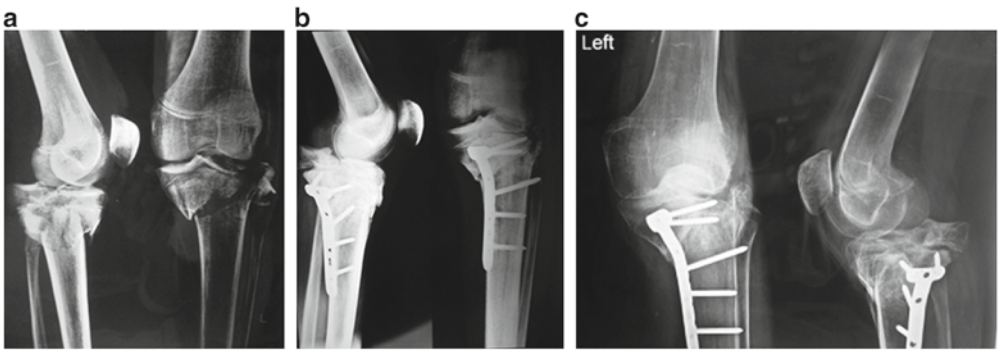


Fig. 30.9 Genu valgum due to malunited upper tibial fracture. (**a**) Injury film. (**b**) Postoperative X-ray. (**c**) Implant failure and malunion. (**d**, **e**) X-rays and clinical picture after implant removal



Fig. 30.10 Type 6 Salter–Harris injury to upper tibial physis perichondrial ring with resultant varus deformity. The child was treated with bar excision and growth modulation. (**a**, **b**) Clinical picture and X-ray 2 years following the injury showing varus deformity upper tibia. (**c**) Gradient echo fat suppression image showing perichondrial ring injury (*white arrow*) with formation of bony bar (*red arrow*). (**d**) Postoperative X-ray showing bone cement used for interposition after excising bony bar, markers, and hemiepiphysiodesis plate. (**e**, **f**) Clinical picture and X-rays 6 months after surgery. (**g**, **h**) Clinical picture and scanogram 13 months postoperatively showing correction of deformity and restoration of growth. (**i**, **j**) Intraoperative images before and after removal of the plate. Note the divergence of the screws

Case Study (Fig. 30.12)

Clinical Summary

Case of posttraumatic tibia vara deformity secondary to fracture of upper end of tibia in a young child: The deformity was progressive and was biplanar with apex at the upper end of tibia. There was severe tibia vara with mild procurvatum of the tibia.

Philosophy of Treatment

Complex deformity in a postphyseal arrest tibia vara with progressive deformity was treated with staged gradual correction with oblique osteotomy through apex and distractors applied for gradual correction. Later a fibular osteotomy was done and the same graft used to fill the osteotomy defect site, which healed and acted like an epiphysiodesis.

Posttraumatic Tibia Valga

An unusual complication of fractures of the upper tibia in children is progressive tibial valgus. There has been a lot of discussion on the mechanism of this valgus and a number of authors have advanced their hypotheses [15, 16].

Operative findings have disclosed soft-tissue interposition in the form of periosteum, pes anserinus, or medial collateral



Fig. 30.11 A 17-year-old with injury to upper tibial growth plate with recurvatum and varus deformity. (**a**, **b**) Anteroposterior X-rays showing the deformity. (**c**) Preoperative picture showing the extent of deformity. (**d**, **e**) Postoperative X-rays showing correction of deformity with osteotomy. (**f–h**) Six-month follow-up showing deformity correction and function with X-ray

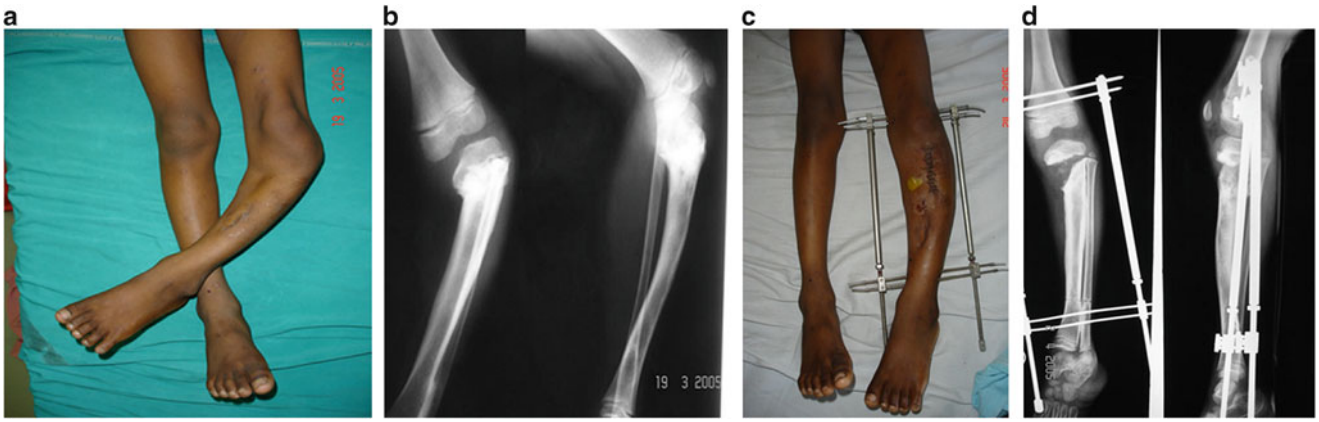


Fig. 30.12 Severe posttraumatic tibia vara secondary to upper tibial physeal arrest. (**a**, **b**) Preoperative clinical picture and X-ray. (**c**, **d**) After differential distraction by a minifixator. (**e**) After osteotomy and grafting. (**f**, **g**) Clinical picture and X-ray on healing

ligament. A consequence of this interposition is loss of periosteal regulation of linear growth on the medial side of the tibia with consequent valgus.

Treatment of this condition is primarily watchful observation. Many authors have shown spontaneous resolution of the valgus with time. If the deformity does not resolve any of the growth modulation techniques can be used. Late presentation may need an osteotomy [16].

Case Study (Fig. 30.13)

A 3-year-old boy was seen in the outpatient clinic with a deformity of the right leg. He had sustained a fracture of the

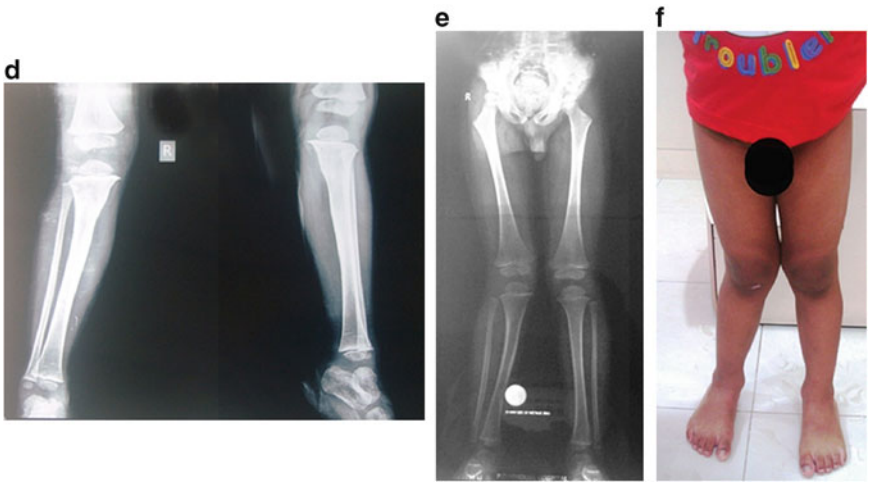


Fig. 30.13 Posttraumatic coxa valga following upper tibial metaphyseal fracture. (**a**) At trauma, May 2013, 1 year and 10 months of age. (**b**) X-ray in plaster cast. (**c**) At 1 year, 11 months of age, June 2013, after cast removal. (**d**) February 2014 at age 2 years, 6 months. (**e**, **f**) X-ray and clinical photo at presentation at 3 years of age, July 2014, MPTA 102°, TFA 18°

right proximal tibial metaphysis at the age of 1 year and 10 months. He had had a cast treatment and there was no history of manipulation at that time. The deformity was apparent a few months after the cast was removed.

Progressive deformity was noted that gradually increased to the current magnitude.

Physeal Injury Ankle with Malunion

The so-called MacFarland fracture is a Salter–Harris type 3 or 4 fracture of the lower tibia and these fractures are associated with a high incidence of growth arrest and deformity. Accurate repositioning of these fractures and accurate restoration of the physeal plate and the joint surface are critical in reducing the incidence of growth arrest. Radiological observation and investigative scans are necessary to determine the progression and location of the physeal arrest.

Case Study (Fig. 30.14)

Clinical Summary

A patient with fracture of distal end of tibia treated conservatively presented with progressive varus deformity at the ankle joint and shortening: The apex of the deformity was present in the distal tibial physis. On radiological examination there was a physeal bar present in the center of the distal tibial physis which was tenting it.

Philosophy of Treatment

An intrafocal osteotomy was done at the distal end of tibia and excision of the bar was done followed by fat graft. A corrective osteotomy was done at the distal end of tibia superior to the previous osteotomy and an external fixator was applied along with an above knee slab.

At present patient walks well with no recurrence of deformity and equal limb length at 5-year follow-up

Case Study (Fig. 30.15)

Clinical Summary

A 11-year-old male with trauma to ankle treated with cast immobilization presenting with progressive valgus deformity within 1 year of the original injury: MRI showed distal fibular growth arrest with damage to lateral part of distal tibial physis.

Philosophy of Treatment

The treatment was planned with very low medial transphyseal osteotomy, and hemiepiphysiodesis of distal tibia to arrest medial growth to achieve correction, with percutaneous physeal drilling for fibular physis.

Patient walks well with full range of ankle motion and no recurrence of deformity at 16 months postsurgery.

Soft-Tissue Loss with Skeletal Injury

It poses a greater challenge for reconstruction as the management involves restoration of soft-tissue cover, muscle length, and skeletal alignment.

This may need soft tissue procedures, use of distraction with external fixation, and osteotomies.

Case Study (Fig. 30.16)

Clinical Summary

A 15-year-old male sustained a runover injury with soft-tissue loss and bony injury (distal physeal arrest) to the lower leg 4 years earlier. He was initially treated with skin grafting

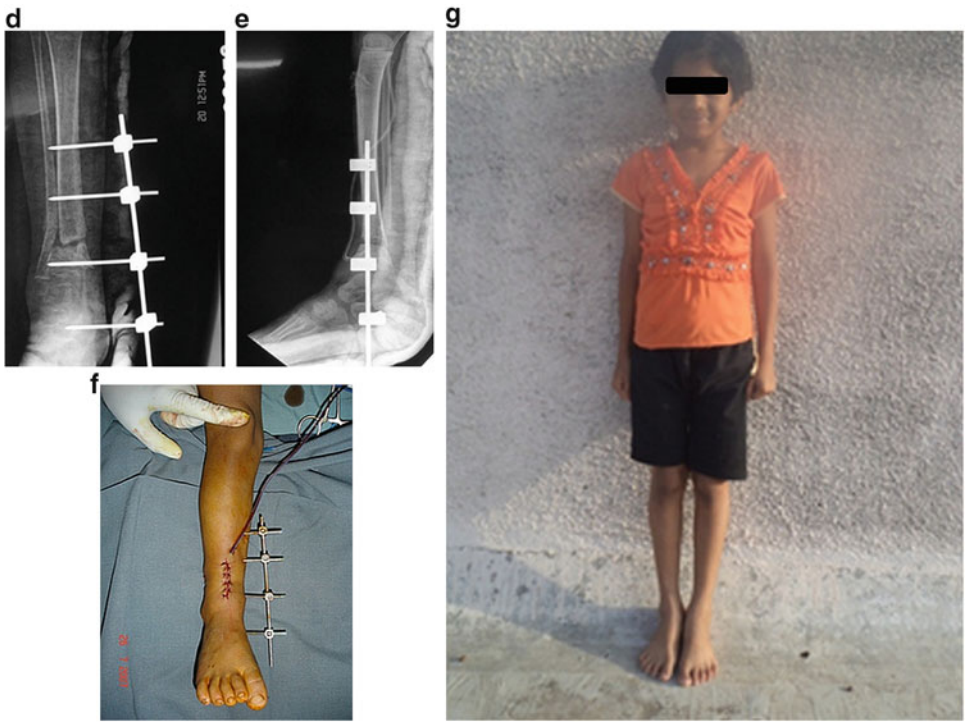


Fig. 30.14 Physeal injury ankle with growth arrest. (**a**, **b**) Postdistal tibial fracture—clinical picture showing deformity and MRI showing physeal central growth arrest with tenting. (**c**) Intraoperative C-arm picture shows osteotomy and central bar excision and fat graft. (**d**–**f**) Postoperative X-ray showing osteotomy and excision of bar with external fixation. (**g**, **h**) Five years postoperative clinical photo and X-rays showing restoration of physeal growth and absence of deformity

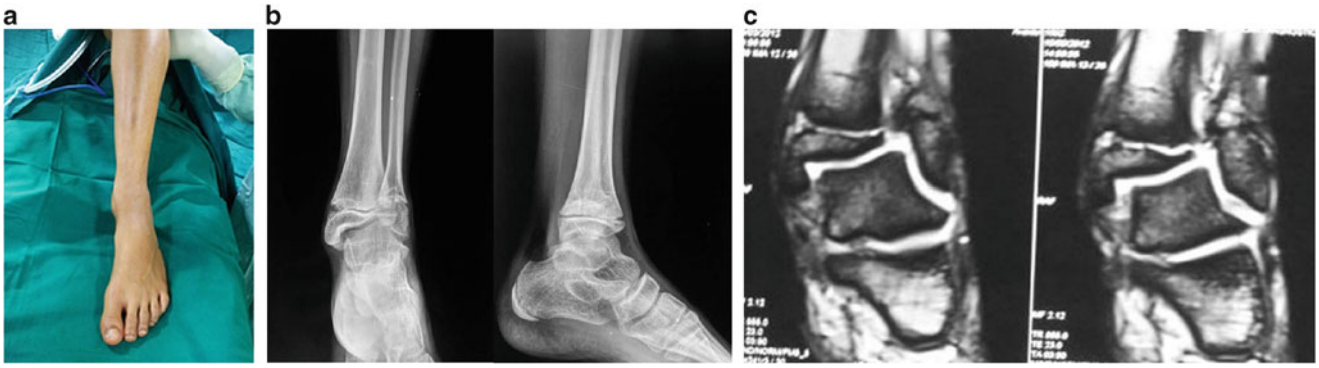


Fig. 30.15 Twelve-year-old child with posttraumatic ankle valgus. (**a**–**c**) Clinical picture, X-rays, and MRI 1 year after injury. (**d**) Intraoperative C-arm image after fibular epiphysiodesis and medial lower tibial hemiepiphysiodesis. (**e**) X-rays at 16-month follow-up showing good correction with physeal closure

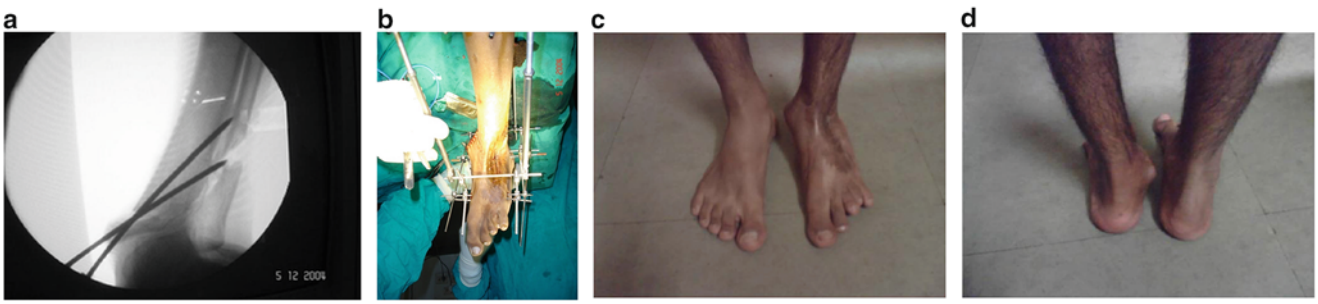


Fig. 30.16 Posttraumatic severe ankle valgus following physeal injury and soft-tissue loss in 15-year-old. (**a**–**c**) Clinical pictures and X-rays at presentation. (**d**) Intraoperative X-ray showing the intraphyseal osteotomy fixed with K wires. (**e**) Intraoperative picture showing mini external fixator. (**f**, **g**) Postoperative pictures on follow-up after fixator removal showing good correction

and external fixation. He presented with a progressive valgus deformity and soft-tissue contracture and was walking on the medial aspect of the foot.

Philosophy of Treatment

We did an oblique closed-wedge intraphyseal osteotomy of distal tibia to correct the deformity and stop further progression of deformity. This was combined with fibular resection and application of a simple distractor system for stretching the lateral scarred soft tissues. We were able to achieve plantigrade foot with this simple strategy.

Summary

Posttraumatic deformity management requires a comprehensive approach in terms of evaluation and management. Such deformities vary in their complexity from a simple extraarticular joint contracture to the more complex and progressive deformities due to physeal injuries or bone loss. In children, the potential for remodeling has to be kept in mind when developing management strategy. Correction may demand combination of techniques and multispecialty involvement. The aim must be to get an acceptable alignment and limb length by skeletal maturity with adequate function of the extremity.

Acceptability Criteria for Reduction in Lower Limb Fractures

Fracture Neck Femur

Only anatomical reduction is acceptable.

Fracture Shaft Femur

0–6 months of age: <1.5 cm of shortening, <30° angulation in varus valgus plane, <30° angulation in AP plane
6 months to 6 years: <2 cm of shortening, <15° angulation in varus valgus plane, <20° anterior angulation
6–10 years: <1.5 cm shortening, <10° varus valgus angulation, <15° AP angulation
>10 years: <1 cm shortening, <5° varus valgus angulation, <10° AP angulation

Fracture: Separation of Distal Physis of Femur

In Salter–Harris Types 1 and 2

<10 years: <20° anterior or posterior angulation
>10 years: Only minimal AP angulation
<5° varus valgus angulation

In Salter–Harris Types 3 and 4

Anatomical reduction and ORIF

Fracture Tibial Tuberosity

Only minimally displaced fractures with possible active extension of knee to 0° can be acceptable. Rest require open reduction and internal fixation.

Fracture Patella

<3 mm articular step-off
<3 mm diastasis on X-ray
Intact extensor mechanism

Fracture of Tibia and Fibula

Proximal Metaphysis

Closed reduction to anatomic position or slight varus is acceptable.

Diaphysis

	<8 years	>8 years
Varus	<10°	<5°
Valgus	<5°	<5°
Ant. angulation	<10°	<5°
Post. angulation	<5°	<0°
Shortening	<10 mm	<5 mm
Rotation	<5°	<5°

Distal Tibial Fractures

Salter–Harris Types 1 and 2

1. In patients with at least 2 years of growth remaining: <15° of posterior angulation, <10° of valgus angulation, 0° of varus angulation
2. In patients with less than 2 years of growth remaining angulation in all planes <5°

Salter–Harris Types 3 and 4

<2 mm displacement

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Postinfectious Deformities of the Lower Limb

31

In Ho Choi

Introduction

There is a diverse range of postinfectious residual deformities of the lower limb depending on the site of infection, patient's age, infecting organism, host resistance, timing and adequacy of initial treatment, and severity of damage to the epiphysis and physis of the affected bone [1–8]. The epiphysis can be damaged extensively, particularly in the neonatal period, by various mechanisms including transphyseal spread of infection from the metaphysis through the physis and into the epiphysis via transphyseal vessels, or following the passage of organisms from the metaphysis to the joint in intracapsular epiphyses such as the hip joint, or from the epiphyses into the adjacent joint following transphyseal spread [9]. As a result, most of the postinfectious juxtaarticular and articular deformities are associated with physeal damage and growth disturbance.

Proper evaluation based on a thorough musculoskeletal examination and radiographic imaging allows the surgeon to determine an effective treatment plan for deformity correction. Meticulous analysis of the complex deformity of the lower limb by determining the location, magnitude, and direction of the deformity would be the first step in characterizing the deformity. Mechanical axis deviation, mechanical and anatomical alignment, and joint orientation should be accurately determined [10]. In addition, LLD and difference in the knee level and the presence of joint laxity and loss of mobility secondary to soft-tissue contractures and/or juxtaarticular skeletal deformity should be evaluated.

The management of severe deformities that occur as a result of fulminant septic arthritis or osteomyelitis is clinically challenging. However, timely and well-performed reconstructive operations can ensure growth and development of the lower limb by restoring the weight-bearing mechanical axis as well as individual joint alignment, and thus providing the best possible joint mechanics at skeletal maturity. This chapter reviews the treatment principles and reconstructive surgical modalities for long-term residual deformities of the lower limb after septic arthritis of the hip, knee, and ankle, and osteomyelitis of the femur and tibia in infancy or early childhood.

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Late Sequelae of Infantile Septic Arthritis of the Hip

Residual deformities of infantile septic arthritis of the hip occur due to necrosis of the articular cartilage, ischemic necrosis of the femoral head (FH), premature closure of the triradiate cartilage, acetabular dysplasia, premature or asymmetrical closure of the proximal femoral physis, subluxation, dislocation, pseudarthrosis of the femoral neck, greater trochanteric overriding, and complete destruction of the FH and neck [1–3, 6–9, 11–14]. The decision to perform either a reconstructive or a salvage procedure should be individualized depending on the type of residual deformity of the hip.

Radiographic Classifications of Late Sequelae of Infantile Septic Arthritis of the Hip

Several radiographic classifications of late sequelae of infantile septic arthritis of the hip have been reported in the literature. In 1982, Hunka et al. [11] first proposed a classification to document the five types of residual deformities of septic arthritis in a logical fashion: Type 1—absent or minimal FH changes; Type II—(A) deformity of the FH, with an intact growth plate; (B) deformity of the FH, with a premature fusion of the growth plate; Type III—pseudarthrosis of the femoral neck; Type IV—(A)

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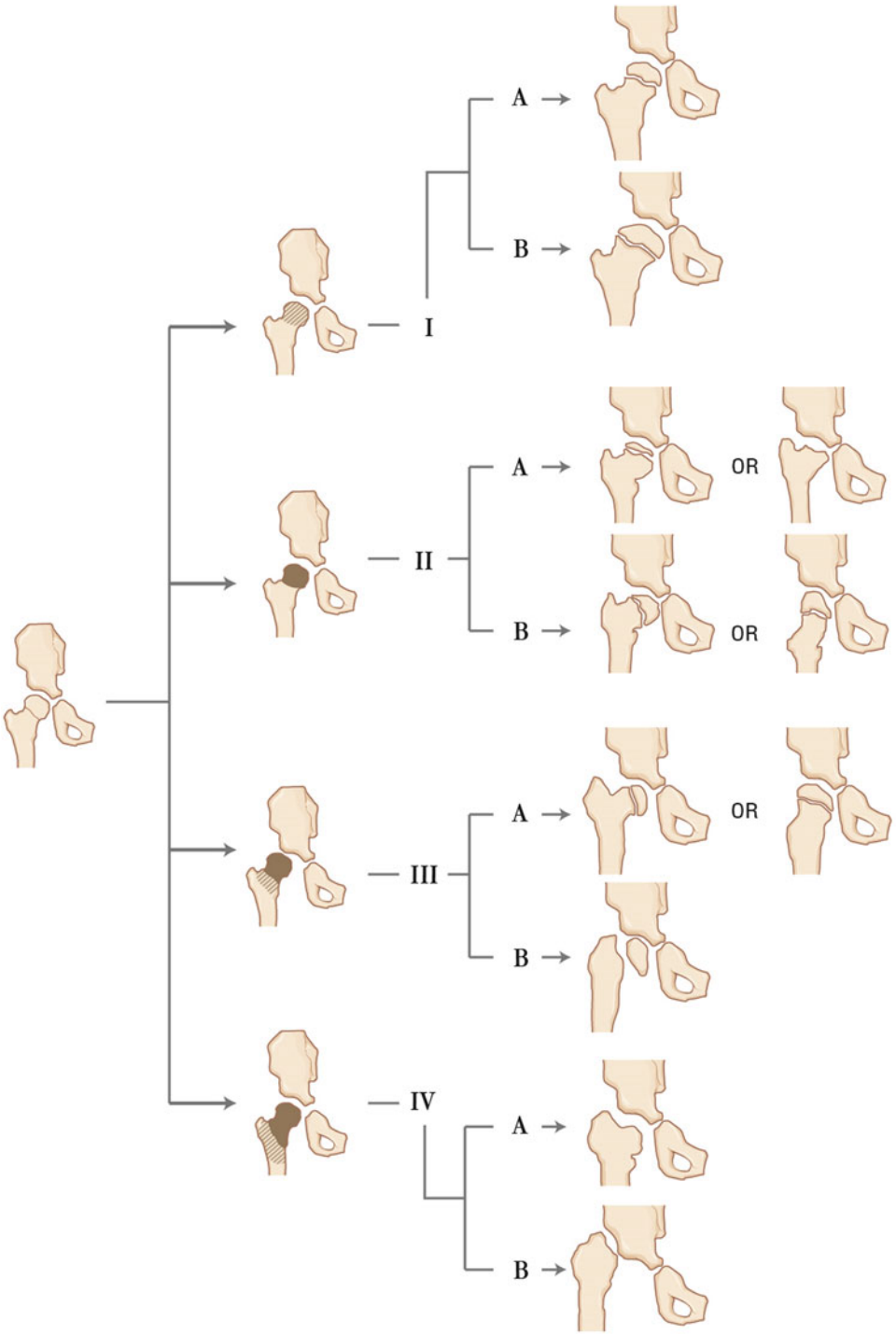


Fig. 31.1 Classification of sequelae of infantile septic arthritis of the hip. Type I: No residual deformity (Type IA) or mild coxa magna (Type IB). Type II: Coxa breva with a deformed head (Type IIA) or progressive coxa vara or valga due to asymmetrical, premature physeal closure (Type IIB). Type III: Slipping at the femoral neck, resulting in coxa vara or valga with severe anteversion or retroversion (Type IIIA) or pseudarthrosis of the femoral neck (Type IIIB). Type IV: Destruction of the femoral head and neck, with a small remnant of the neck (Type IVA) or complete loss of the femoral head and neck and no articulation of the hip (Type IVB)

complete destruction of the proximal femoral epiphysis, with a stable neck segment, (B) complete destruction of the proximal femoral epiphysis, with a small unstable neck segment; Type V—complete destruction of the FH and neck to the intertrochanteric line, with dislocation of the hip.

In 1990 the author devised a radiological classification of residual proximal femoral deformities based on the nature and extent of damage and radiographic appearance at the final follow-up or at maturity, and each of the four major types of residual deformities was further divided into two subtypes (Fig. 31.1) [1]. In 2002, Johari et al. [12, 13]

also reported the classification of the sequelae of septic hips. Group 1: loss of capital femoral epiphysis (CFE)/neck, metaphyseal spike present, stable; Group 2: loss of CFE and neck, unstable; Group 3A: dislocation, CFE present, unstable; Group 3B: subluxation, CFE present, unstable; Group 4, articular incongruity, avascular necrosis, coxa magna, physeal disturbance (coxa breva, coxa vara, coxa valga, and trochanteric overgrowth), stable; Group 5, pseudarthrosis of the femoral neck, stable/unstable. In 2008, Forlin and Milani [14] proposed a relatively simple radiological classification based on the relation between the femur and the acetabulum, and the severity of the lesion of the proximal femur. Hips with sequelae grade 1 were reduced hips, with FH preserved (1A) or absent (1B); hips grade 2 were dislocated hips, with the FH preserved (2A) or absent (2B). The classification systems proposed by other authors [11–14] do not have a subtype equivalent to the Choi Type IIIA (slipping at the femoral neck, resulting in coxa vara or valga with severe anteversion or retroversion) (see Fig. 31.1) [1].

One common drawback of all the aforementioned radiological classifications is that they were mainly devised based on the changes noted on the plain radiographs. Currently, with the help of modern, advanced imaging technologies, e.g., MRI and 3D-CT, deformed osteocartilaginous geometry of the proximal femur can be better visualized in three dimensions, and the dynamics of articular motion between the deformed FH and acetabulum can be better understood. A new, comprehensive three-dimensional radiological classification is anticipated considering the rapid advancement in the imaging technology.

Late Operative Treatment Modalities for Residual Deformities of Septic Arthritis of the Hip

Treatment modalities for the long-term sequelae of infantile septic arthritis of the hip are outlined in Table 31.1. The recommended primary and secondary treatment methods are as varied as the types of deformities. Unfortunately, no definite primary treatment for a severely damaged hip joint makes the affected joint capable of withstanding the forces imposed on it by otherwise healthy children with their long-term physical demands [1]. Thus, any surgical treatment for severe sequelae must be regarded as a measure that temporarily improves the clinical function and delays the more definitive procedures that are reserved for adult patients [1, 2]. The pearls and pitfalls in the management of late sequelae of infantile septic arthritis of the hip are summarized in Table 31.2.

Choi Type II and III Hips Causing Femoroacetabular Impingement and Secondary Hip Dysplasia

Type II and III hips are fundamentally the result of an abnormally shaped, sized, or oriented FH and neck, altered head-neck offset, and acetabular dysplasia. A wide variety of residual deformities of the FH have been observed in Type II and III hips, ranging from deformities similar to those in Perthes disease with coxa vara/valga, coxa plana, coxa breva, coxa magna, coxa irregularis, relative greater trochanteric overriding, sagging rope sign [15], and acetabular dysplasia to the more deformed proximal femur visualized as beard, collared, staghorn, or rugby ball-shaped FH [16, 17].

Children with Type II or III deformities may remain asymptomatic in their younger age, but, in (pre)adolescence, these children often present with clinical problems such as pain, limp, joint instability or contracture, restriction of motion, LLD, spinal disorders, and even emotional disturbances. According to the author's experience, in older children with Type II and III hips, a painful limp is usually associated with femoroacetabular impingement (FAI) with or without damaged acetabular cartilage and/or labrum.

Figure 31.2 depicts a stepwise algorithmic approach to FAI in Perthes-like deformities at the author's institute. First, the extent of asphericity is assessed; if it is mild and the FH is well contained within the acetabulum, osteochondroplasty of the head and neck junction alone is indicated. Next, the presence or absence of a labral tear is determined. If a labral tear is present, labral debridement or refixation is necessary. Similarly, the presence of coexisting acetabular dysplasia is assessed. If the hip becomes congruent in abduction, relative femoral neck lengthening combined with periacetabular osteotomy (PAO) or triple innominate osteotomy (TIO) is indicated. In the author's institute, Bernese PAO is preferred for correction of acetabular dysplasia in adolescents and young adults with closed triradiate cartilage (around 12 years in girls and 14 years in boys) [18], whereas a TIO, in (pre) adolescents with open triradiate cartilage [19]. If the hip becomes congruent in adduction, proximal femoral valgus osteotomy and PAO or TIO are indicated. The next step is to check whether coxa breva and coxa vara associated with relatively greater trochanteric overriding are severe enough to cause FAI, and if so, a relative or true femoral neck lengthening combined with distal advancement of the greater trochanter (GT) is indicated [20–23]. When the FH asphericity is too severe and the FH is "irreducible," the following procedures should be considered. If the hip becomes congruent in extension and adduction, femoral valgus-flexion osteotomy plus/minus PAO or TIO should be considered.

Table 31.1 Late operative treatment modalities for severe sequelae of infantile septic arthritis of the hip

Purpose	Indications	Procedures
To reduce/contain the hip joint	Subluxation or dislocation	Open reduction; soft-tissue release (adductor-iliopsoas tenotomy)
To optimize joint congruity	Incongruity	Proximal femoral realignment osteotomy (varus or valgus osteotomy with a sagittal or rotational component); redirectional acetabular osteotomy (Salter, Pemberton, Dega osteotomy, triple innominate osteotomy, Ganz periacetabular osteotomy)
To improve femoral head coverage	Uncovered femoral head	Redirectional acetabular osteotomy; salvage acetabuloplasty (e.g., Chiari, shelf operation); Ganz femoral head reduction osteotomy
To reduce cam or pincer lesion	Femoroacetabular impingement	Osteochondroplasty via safe surgical hip dislocation or arthroscopy; excision of the femoral head and neck remnant (Choi Type IVA or mobile nonunion type of IIIB)
To address intra-articular pathology of the cartilage or labrum	Torn labrum or articular cartilage lesion	Labral debridement or refixation; articular cartilage repair using microfracture technique or osteochondral grafting or chondrocyte transplantation
To correct acetabular dysplasia	Dysplastic acetabulum	Redirectional acetabular osteotomy (e.g., Salter, Pemberton, Dega, triple innominate osteotomy, Ganz periacetabular osteotomy)
To correct abnormal version of the proximal femur and acetabulum	Anteverted or retroverted femur and acetabulum	Proximal femoral rotational/derotational osteotomy; redirectional acetabular osteotomy (e.g., triple innominate osteotomy, Ganz periacetabular osteotomy)
To improve abductor muscle function; to correct coxa brevis	Greater trochanteric overriding; short femoral neck	Greater trochanteric advancement; true or relative femoral neck lengthening
To repair pseudarthrosis of the femoral neck	Femoral neck nonunion	Bone grafting, often combined with proximal femoral valgus osteotomy
To reconstruct femoro-pelvic articulation	Destruction of the femoral head and neck	Greater trochanteric arthroplasty; pelvic support osteotomy; Ilizarov hip reconstruction (Choi Type IVB or failed type IVA); L'Episcopo, Albee, or Harmon operation, Ilizarov femoral neck lengthening, femoral neck reconstruction using vascularized bone grafting (Choi Type IVA)
To equalize the lower limb length	Limb length shortening	Lengthening by distraction osteogenesis; contralateral epiphysiodesis or shortening of the femur; Ilizarov hip reconstruction
To manage painful unstable hip as a last resort	Unstable hip with advanced osteoarthritis not amenable to the above-mentioned reconstructive procedures	Hip arthrodesis; total hip replacement arthroplasty; Ilizarov hip reconstruction after excision of the femoral head and neck remnant

Table 31.2 Pearls and pitfalls in the management of late sequelae of infantile septic arthritis of the hip

Assessment	The surgeon should be familiar with radiological classifications of femoral residual deformities. Use of advanced imaging technologies (MRI, ultrasonography, arthrography, and 2-D/3D-CT) to visualize unossified cartilaginous structure of the epiphysis, also to determine osteocartilaginous geometry of the proximal femur, and the dynamics of articular motion between the deformed head and acetabulum.
	Full-length standing radiograph (teleradiograph) visualizing the entire lower limbs is helpful not only to identify associated sequelae at other sites, if any, but also to determine axial malalignment, joint malorientation, and limb length discrepancy.
Treatment	Femoroacetabular impingement-related symptoms are often encountered in older children and young adults with severe residual deformities.
	The decision to perform either reconstructive or salvage procedures must be made on an individual basis, after reviewing the clinical findings and imaging studies. Definitive and adjunctive surgical treatment methods can be used alone or in combination (see Table 31.1).
	Author's stepwise algorithmic surgical approach to femoroacetabular impingement would be helpful in decision making of treatment for Choi Type II and III hips (see Fig. 31.2).
	Greater trochanteric arthroplasty for reconstruction of femoro-pelvic articulation is a technically demanding procedure with unpredictable outcomes. If performed at younger age, the transferred greater trochanter can assume a globular shape, but with no or incomplete formation of head-neck offset.
	Ilizarov hip reconstruction is a highly effective treatment modality for the management of Choi Type IVB and IVA hips in which the previous reconstructive surgery has failed, or there is late presentation with a severe abductor lurch and a considerable limb length shortening, particularly in the adolescents and young adults in whom arthrodesis or hip arthroplasty is not suitable.

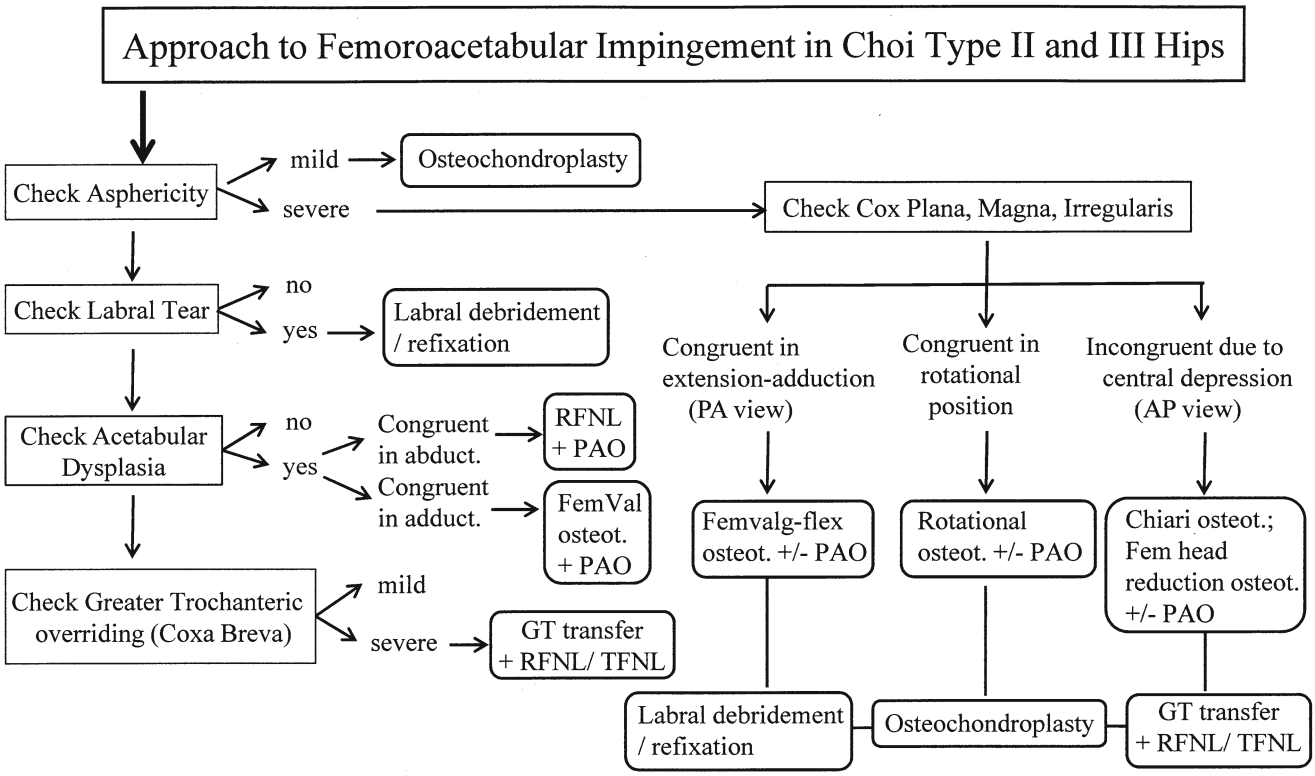


Fig. 31.2 A schema showing the stepwise algorithmic approach to femoroacetabular impingement associated with Choi Type II and III sequelae, which are similar to Perthes(-like) deformities (see text). *RFNL* relative femoral neck lengthening, *TFNL* true femoral neck lengthening, *PAO* periacetabular osteotomy, *PA* posteroanterior, *AP* anteroposterior. Note: A triple innominate osteotomy is performed, instead of PAO, in (pre)adolescents with open triradiate cartilage [18]

If the hip becomes congruent in any rotational position, rotational osteotomy with or without PAO or TIO may be indicated. If the FH with a central depression is too severely deformed such that the FH cannot be contained within the acetabulum in any position, Chiari osteotomy or FH reduction osteotomy [20, 21, 24] plus/minus PAO or TIO can be considered. In addition, combination of osteochondroplasty, labral debridement/refixation, and distal advancement of the greater trochanter should be performed, if necessary.

Late Treatment Options for Choi Type IV Deformity

The available reconstructive procedures for the treatment of Type IV hips are limited, and, in general, have not yielded satisfactory long-term results [1, 2, 7, 11, 25–29]. When only a remnant of the FH and neck is present, it is difficult to maintain a stable reduction; and if instability and dislocation persist, the surgeon can either leave the deformity uncorrected or can attempt reconstruction of femoro-pelvic articulation [25]. In young children in whom the FH and neck have been destroyed, as in Type IVA sequelae, a new femoral neck can be fashioned to articulate with the acetabulum by utilizing the L'Episcopo [30], Harmon [31], or Albee [32] reconstruction methods. When a reasonably sized unossified cartilaginous cap persists as a femoral neck remnant, open reduction can be attempted. The repositioning of the femoral remnant into the acetabular socket can be facilitated using a modification of the Harmon technique [31]. An incomplete, spring osteotomy is made at the base of the femoral remnant adjoining the greater trochanter and the resulting opening wedged gap is filled with a block of cartilage graft taken from the iliac apophysis. This procedure produces a femoral neck-lengthening effect [28]. In older children, gradual lengthening of the remnant femoral neck using the Ilizarov technique may be indicated. Nonetheless, the author believes that open reduction in children more than 6 years of age is not likely to be beneficial because of the high risk of developing stiffness and pain. Therefore, the author recommends that in children older than 6 years of age, Type IVA hips should be treated in the same manner as hips with Type IVB sequelae [2].

Postponing the treatment of Type IVB hips will result in proximal iliac dislocation with a marked abductor lurch, telescoping, limp, and LLD [8, 26]. Although these patients may remain asymptomatic for a number of years [3, 7, 12] they are at a risk of developing degenerative changes in the lumbosacral spine and hips [25]. Therefore, various treatment modalities for

reconstructing a femoro-pelvic articulation in Type IVB hips have been reported: hip reconstruction with vascularized iliac crest grafting [33], arthrodesis, greater trochanteric arthroplasty (GTA) [25, 26, 34–39] with adjunctive or secondary procedures, pelvic support osteotomy (PSO) [40–44], and Ilizarov hip reconstruction (IHR) [44–58]. Of these treatment modalities, the two most commonly performed procedures are GTA and IHR.

Greater Trochanteric Arthroplasty Versus Pelvic Support Osteotomy (Ilizarov Hip Reconstruction)

GTA was originally described by Colonna [34] and was popularized by Westin [35] and others [25, 26, 36, 37]. The underlying concept of GTA is based on the following two aspects: firstly, the GT is viable and therefore retains its growth potential; and secondly, when the hyaline cartilage covering the GT is placed inside the acetabulum, the GT assumes a globular shape, similar to that of an FH [1, 2, 18, 26, 28, 29, 34–39, 44]. The detached abductor muscles must be transferred distally in order to provide hip stability and some degree of abductor function. However, the subsequent progressive subluxation observed in most of the patients necessitates additional procedures, such as femoral varus osteotomy, pelvic osteotomy including Chiari osteotomy, or shelf acetabuloplasty, to improve FH coverage. There are many drawbacks and limitations of GTA for reconstruction of femoro-pelvic articulation due to the uniqueness of the technique. Previous reports have described the potential risk of avascularity of the proximal segment after femoral varus osteotomy, difficulty in osteosynthesis at the varus osteotomy site, gradual straightening of the proximal femur due to remodeling at the osteotomy site, abductor weakness, stiffness, degenerative arthritis, and difficulty during conversion to total hip arthroplasty in adulthood [1, 2, 25, 26, 28, 36, 37, 51]. According to the author's observation, the transferred GT can assume a globular shape with time, but with no or incomplete formation of head-neck offset. For this reason, even those children with satisfactory results at the midterm follow-up often present with FAI-related symptoms (Fig. 31.3).

The two most important factors that determine the success or failure of a procedure after GTA are preoperative abductor weakness and stiffness [2]. Although some authors have reported satisfactory results [25, 26, 34, 35, 37–39], most of the experienced surgeons feel that GTA is a technically demanding procedure with highly unpredictable outcomes. The author has noted that the result of GTA may also be correlated with the patient's age at operation. In a retrospective study performed by the author [28], satisfactory results were obtained only in those children who underwent GTA at less than 6 years of age. This observation may support the opinion that a patient should be old enough to follow the physical therapy instructions, and young enough to allow remodeling of the transferred GT [2].

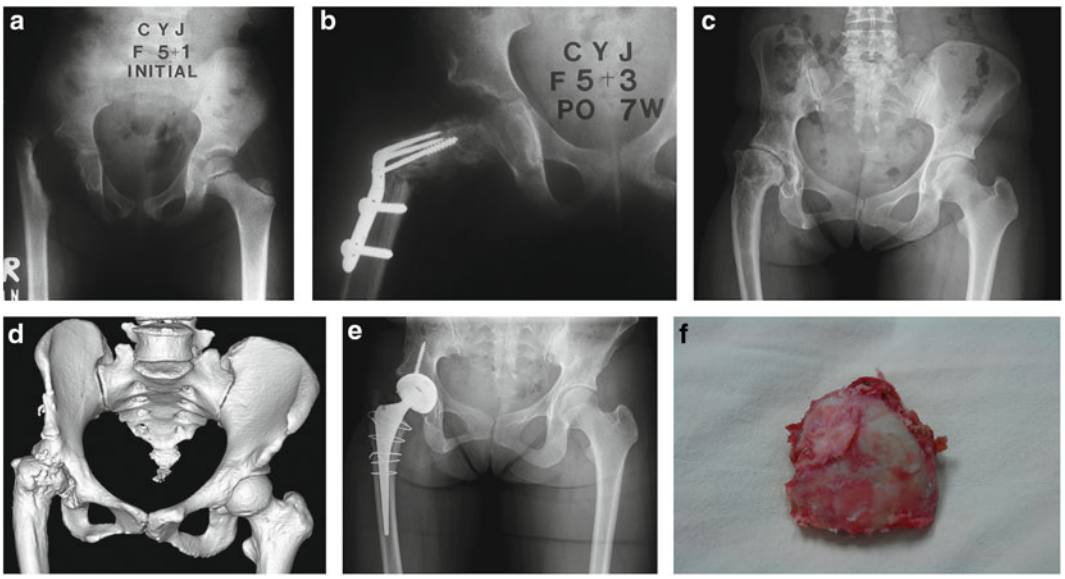


Fig. 31.3 Greater trochanteric arthroplasty in a girl. (**a**) A radiograph in a 5-year- and 1-month-old girl showing Choi Type IVB sequelae in the right hip. (**b**) A radiograph showing trochanteric arthroplasty with adjunctive femoral varus osteotomy. (**c**, **d**) Radiographs taken at 32 years of age showing advanced degenerative arthritis of the right hip joint, despite a well-remodeled greater trochanter which had assumed a globular shape similar to that of a femoral head. 3D-CT image also shows laterally displaced, remodeled greater trochanter with multiple osteochondral prominences on its margin, suggestive of femoroacetabular impingement. (**e**) A radiograph showing total hip replacement arthroplasty which was performed to treat a painful limp. (**f**) A gross photograph of the remodeled greater trochanter; the articular cartilaginous surface had been markedly degenerated, showing some fibrous adhesions

"Pelvic support osteotomy (PSO)" has a long history in orthopedic surgery. The technique was developed in 1910s and quite popular until the advent of total hip arthroplasty [40–44]. The basic concepts and goals of PSO are to enhance femoro-pelvic stability by means of a proximal femoral valgus osteotomy and to improve hip biomechanics by displacing the center of gravity medially, resulting in an improvement in the mechanical efficiency of the abductor muscles [44]. However, the clinical application of traditional PSO is limited due to its indigenous shortcomings. It is difficult to achieve the optimal extent of angulation. If the angular correction is too large, excessive genu valgum, fixed pelvic obliquity, and impingement pain on adduction of the lower limb to neutral position may occur. On the other hand, if the angular correction is too small, the result would lead to an insufficient improvement in hip biomechanics, and most importantly, the issue of persisting LLD cannot be addressed. To overcome the shortcomings of traditional PSO, Ilizarov developed and popularized a modified PSO technique, the so-called IHR, in which a second distal femoral osteotomy is performed in addition to proximal femoral valgus osteotomy for pelvic support to realign the knee joint and to correct the LLD [44–58]. Furthermore, Ilizarov emphasized the importance of extension of the proximal femoral osteotomy to correct the fixed flexion deformity of the hip and to permit locking of the hip joint [44–47, 49–58].

Evidence has accumulated that IHR is highly effective in eliminating or substantially reducing the Trendelenburg gait and in equalizing the lower limb length while improving the stability and range of motion of the hip joint at the same time [44, 47–58]. Hence, the author currently prefers IHR for the management of Type IVB and IVA hips in which the previous reconstructive surgery has failed, or there is late presentation with a severe abductor lurch and a considerable LLD, particularly in the adolescents and young adults in whom arthrodesis or hip arthroplasty is not suitable [44].

The reported complications of IHR in the literature include knee stiffness, pin tract infection, delayed consolidation, refracture, obturator nerve entrapment, straightening of proximal valgus angulation, and persistent Trendelenburg gait. There exists a possibility of ischiofemoral impingement if the apex of the proximal femoral valgus angulation site directly abuts the ischium. The literature demonstrates that an average of 30 % (range, 0–62.5 %) of patients have a persistent positive Trendelenburg sign after IHR, although it is reduced in severity in most of the patients [44].

IHR is not ideal for young children, because in accordance with Wolff's law, gradual straightening of the proximal femur tends to occur at the site of valgus angulation, which may result in loss of pelvic support. Bone remodeling and subsequent gradual loss of valgus angulation remain important considerations in proximal femoral osteotomy [44, 46, 51, 52, 55, 56, 58] (Fig. 31.4). Therefore, when performing IHR at an age before adolescence, as much as 25°–40° of overcorrection should be empirically added to the valgus correction angle based on the single-leg stance drop angle relative to the horizontal line of the pelvis due to the possibility of straightening of the proximal femur after IHR [44, 46, 51]. The author believes that IHR is still indicated even if patients are older than 6 years of age, despite the high probability of repeat PSO, because a second femoral lengthening is usually needed at or near skeletal maturity in patients with hip instability associated with marked LLD due to multiple lower limb growth disturbances secondary to neonatal sepsis. Another alternative is to perform femoral lengthening without a PSO, and insert half pins into the pelvis to prevent proximal migration of the femur at a younger age, and subsequently perform IHR when the patients are near skeletal maturity [44, 55].

Late Sequelae After Septic Arthritis and Osteomyelitis Around the Knee and Ankle Joints

The rate of overall unsatisfactory results in terms of residual deformities after infantile or childhood septic arthritis of the knee and ankle joints is less as compared to that after septic arthritis of the hip [5, 6], because the infection in these joints can be diagnosed sooner than that in the hip [9]. Long-term assessment is essential because radiographic improvement in the involved epiphyseal region can occur several months to years after the infection. In younger children in whom the secondary ossification center has not yet developed, vascular damage can lead to a marked delay in the formation of the ossific center, but the cartilage model continues to grow. Therefore, even when bone deficiency is seen on plain radiography, it is necessary to perform advanced imaging studies such as ultrasonography, arthrography, or MR imaging [9] in order to better visualize the cartilaginous epiphysis (chondroepiphysis) and assess the potential growth of the adjacent physis. When the appropriate treatment is delayed, growth disturbance with a resultant angular deformity, joint incongruity, impingement, and accompanying ligamentous laxity and joint instability may develop gradually. Postinfectious juxta-articular deformity of the knee usually develops as a result of metaphyseal osteomyelitis or primary septic arthritis of the knee, involving the distal femoral physis or the proximal tibial physis, or both. The distal femur is more prone to growth-related sequelae than the proximal tibia. Angular deformities due to partial distal femoral physeal destruction are far more common than complete symmetric growth cessation [9]. The most serious growth sequelae occur following neonatal sepsis and to a lesser extent within the first 2 years of life [6, 9] (Fig. 31.5). In particular, meningococcemia caused by *Neisseria meningitides* or

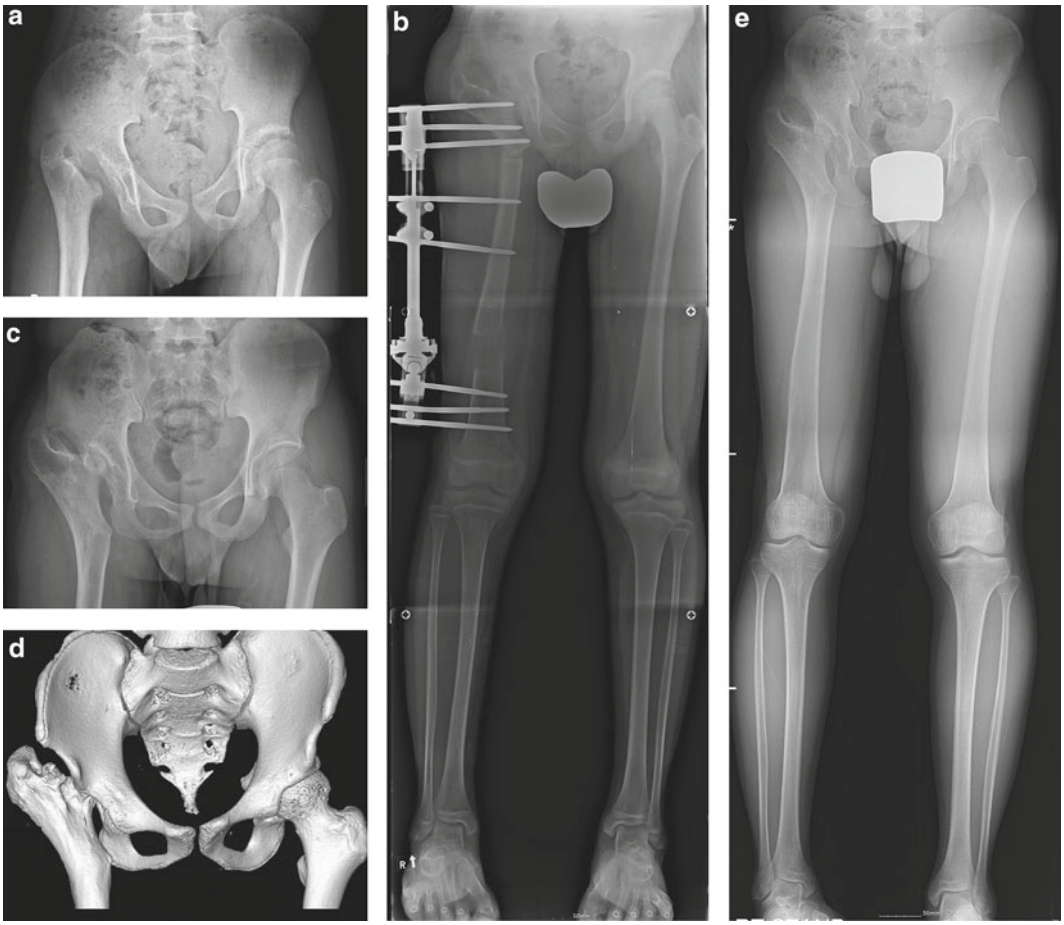


Fig. 31.4 (**a**) A radiograph in a 12-year- and 4-month-old boy with Choi Type IVB sequelae in the right hip. He had a previous history of Ilizarov hip reconstruction with 40° of proximal valgus osteotomy for pelvic support and 4 cm of distal diaphyseal lengthening for leg length equalization performed at an age of 8 years and 1 month. It is noteworthy to observe a completely remodeled, straightened proximal femur showing no evidence of pelvic support. (**b**) A teleradiograph showing a repeated Ilizarov hip reconstruction osteotomy. (**c**, **d**) A plain radiograph and 3D-CT taken at 17 years of age showing displacement of the lesser trochanter into the acetabulum and proximal femur with valgus angulation near the ischium, which apparently enhances the femoropelvic stability and improves abductor mechanics by displacing the center of gravity medially. (**e**) A teleradiograph taken at 17 years of age showing equalization of the lower limb length after surgery. He had a negative Trendelenburg sign

meningococcus, which is found only in human respiratory secretions, is a devastating illness that primarily affects young children. The disseminated intravascular coagulation and focal infections of the acute phase are primarily responsible for the vascular injuries to the growing chondro-osseous tissues and contiguous soft tissues. Ischemic changes also selectively involve the physeal circulation, which adversely affect longitudinal and transverse growth of bone. Therefore, children who survive meningococcal septicemia are prone to develop complex deformities of the lower limb due to multiple areas of growth disturbance [59–61].

The ultimate goal of managing juxta-articular and intra-articular deformity is prevention of osteoarthritis of the affected joint and the adjacent joint by restoring the weight-bearing mechanical axis and by normalizing the joint orientation as early as possible, because both these factors contribute to abnormal loading of the affected joint, leading to compartmental osteoarthritis of the affected joint(s) [6, 62, 63]. Juxta-articular realignment and reorientation osteotomy are preferred because proximity of such an osteotomy to the adjacent joint leads to a more anatomic reorientation correction, without iatrogenic translational deformities. The resultant pain reduction and decreased wear rate of the articular cartilage due to redistribution of joint reaction forces increase the longevity of the major weight-bearing joints [63]. The pearls and pitfalls in the management of late sequelae after septic arthritis and osteomyelitis around the knee and ankle joints are summarized in Table 31.3.

To establish an optimal treatment plan for restoring the weight-bearing axis, for stabilizing the unstable joint, and for equalizing the lower limb length, preoperative three-dimensional assessment of the juxta-articular and articular

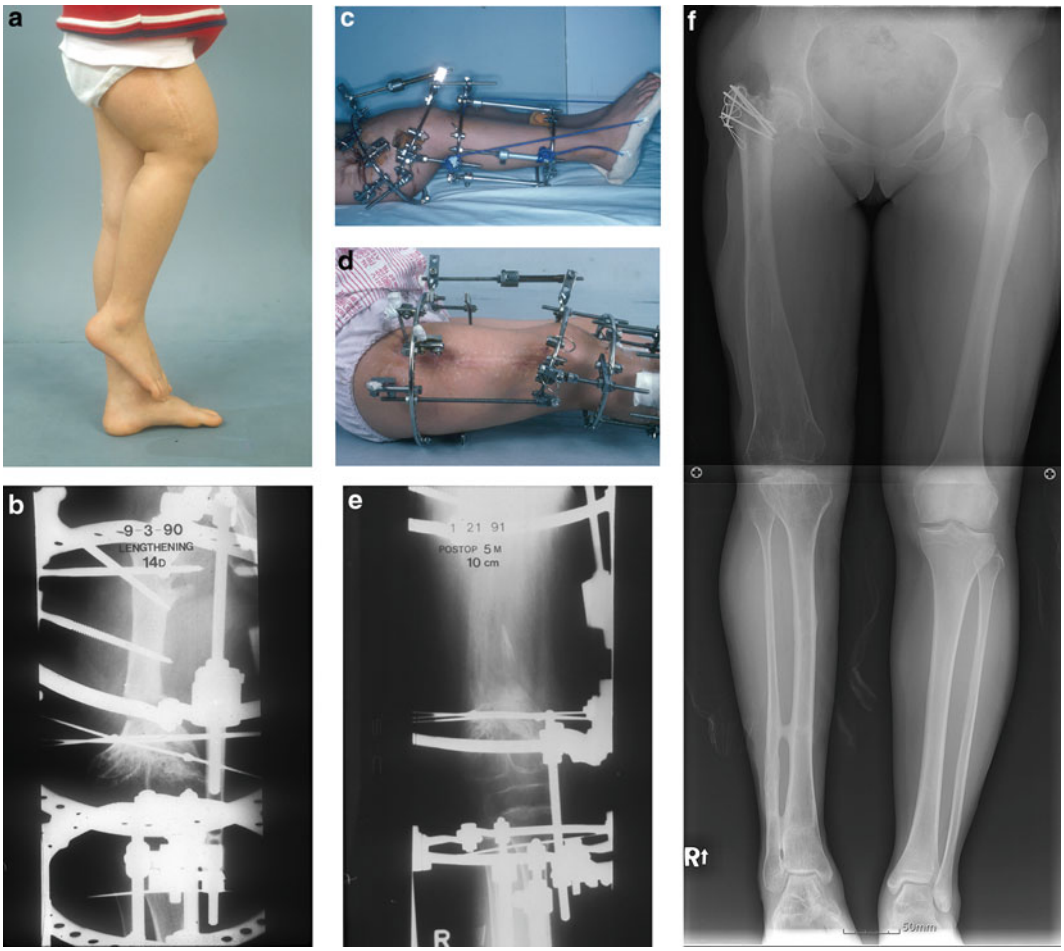


Fig. 31.5 (**a**) A 10-year- and 7-month-old girl presenting with 17.5 cm of shortening of the right femur and a concomitant 30° of flexion contracture of the knee joint after fulminant osteomyelitis of the right femur extending to the hip and knee joints. Further flexion in the knee joint was only marginally limited. (**b**, **c**) A radiograph and photograph showing the femoral and tibial Ilizarov frames constructed with one proximal full ring and a distal two-third ring for concomitant femoral lengthening and deformity correction at the distal femoral corticotomy site. Tibial frame was applied for the purpose of transarticular fixation during lengthening to prevent potential subluxation of the unstable knee joint apparently caused by deformation of the femoral condyle secondary to central physeal arrest in the distal femur. (**d**, **e**) A concomitant 10-cm lengthening and deformity correction could be achieved by differential distraction of the anterior and posterior motor rods. Subsequent procedures included right tibial and fibular lengthening followed by left distal femoral epiphysiodesis, concomitant right femoral and tibial lengthening, and right greater trochanteric advancement. (**f**) At the last follow-up at the age of 26 years, her right lower limb function and gait were excellent with equalization of the lower limb length and no knee flexion contracture despite the difference in the knee level

deformity is essential. The surgeon should recognize that frontal, sagittal, and oblique plane malalignment can result from a variety of sources such as ligamentous laxity, joint subluxation, and altered joint line secondary to physeal growth disturbance, and epiphyseal deficiency or deformation. This may be particularly true in children with severe multifocal, angular, and/or rotational deformities, often associated with joint incongruity, joint instability, LLD, and surrounding soft-tissue scars and contractures. In such children, identification, characterization, and prioritization of each component of the deformity are crucial. The decision and timing of performing multilevel operations either in one stage or in multiple stages must be made on an individual basis.

Patients who have a juxta-articular deformity may also have a compensatory fixed deformity at an adjacent bone and joint, and correction of this deformity must also be included in the treatment plan. Correction of the malunion without addressing the compensatory deformity may result in a straight bone with a maloriented joint, leading to long-term disability due to altered biomechanics. For instance, when distal femoral valgus coexists with compensatory proximal tibia varus or distal femoral varus in the presence of a compensatory proximal tibial valgus deformity, the lower limb may appear aligned with no mechanical axis deviation despite malorientation of the knee joint. However, combinations of femoral valgus and tibial varus would produce shear stress in the knee joint due to increased obliquity of the knee

Table 31.3 Pearls and pitfalls in the management of late sequelae after septic arthritis and osteomyelitis around the knee and ankle joints

Assessment	In young children, it is necessary to perform studies assessing the cartilaginous structure of an epiphyseal region to assess the growth potential by using ultrasonography, arthrography, bone scintigraphy, and MRI.
	Meticulous deformity analysis of the complex deformity by determining the location, magnitude, and direction of the deformity is the first step. Frontal, sagittal, and oblique plane malalignment can also result from ligamentous laxity, joint subluxation, and joint line deformity.
	It is essential to determine whether a juxta-articular deformity has a compensatory fixed deformity at an adjacent bone and joint. When there exists a compensatory fixed deformity, correction of this deformity must also be included in the treatment plan.
	Careful assessment of physeal integrity, intra-articular epiphyseal irregularity, joint instability, surrounding soft-tissue condition, and neurovascular condition is essential.
Treatment	The decision whether to perform acute versus gradual deformity correction, and conventional osteotomy versus external fixator-assisted plating or intramedullary nailing, should be made on an individual basis. When a technique of lengthening over an intramedullary nail using external fixation is used, caution is required to prevent deep infection in patients who have a past history of infection, and open trauma.
	Juxta-articular realignment and reorientation osteotomy are preferred because the closer is each osteotomy to the adjacent joint, the greater is the reorientation with angular correction.
	When correcting a juxta-articular deformity by conventional metaphyseal osteotomy, the use of the Ilizarov technique with an offset hinge system or external fixation which utilizes the six-axis deformity correction principle is preferred.
	When correcting severe juxta-articular deformity, adjunctive procedures (e.g., peroneal nerve decompression and compartment release) may be justified to prevent major complications.
	In the management of juxta-articular ankle deformity, early stabilization of the talocrural joint by leveling the ankle and restoration of the axial alignment and joint orientation are of paramount importance.
	Intra-articular osteotomy is a very technically demanding procedure, and should be selectively performed.



Fig. 31.6 A radiograph showing right distal femoral valgus and compensatory proximal tibia varus in a 16-year-old boy. Although the lower limb may appear aligned with little mechanical axis deviation despite increased obliquity of the knee joint, this deformity can cause lateral subluxation of the proximal tibia associated with progressive degenerative arthritis in late adulthood

joint to the ground (Fig. 31.6). This deformity can cause lateral subluxation of the proximal tibia associated with progressive degenerative arthritis in late adulthood. In contrast, in the case of distal femoral varus and proximal tibial valgus, patients may tolerate well because obliquity of the joint adds to knee joint stability, buttressing against the lateral subluxation forces [64, 65].

Correction of juxta-articular deformity by conventional metaphyseal osteotomy is associated with many problems, including fixation stability, geometrical considerations related to the difference between the osteotomy site and the center of the deformity, multilevel deformities caused by previous osteotomies, and concomitant LLD. In addition, the disadvantages of acute correction using plate and screw fixation include the requirement of extensive soft-tissue dissection, limitation of early weight bearing and function, and inability to correct a significant shortening deformity. It is well known that realignment of the mechanical axis and joint orientation by metaphyseal osteotomy requires a combination of angulation and translation [62, 63]. In this regard, the Ilizarov technique with an offset (juxta-articular) hinge system is preferred for accurately correcting and restoring the mechanical axis and joint orientation. In recent years, the use of external fixators which utilize the six-axis deformity correction principle using a software program has been advocated due to the convenience and accuracy of such adjuncts in correcting complex juxta-articular deformities of the limb. External fixator-assisted acute deformity correction and fixation using a locking compression plate [66–68] is another surgical technique for addressing such deformities. The potential advantages of a fixator-assisted plating include correction of deformities under direct visualization, rigid fixation, and its applicability in various anatomic locations and situations, e.g., open physis, narrow intramedullary canal, and bowing deformity of the femur and tibia. Similarly, external fixator-assisted acute

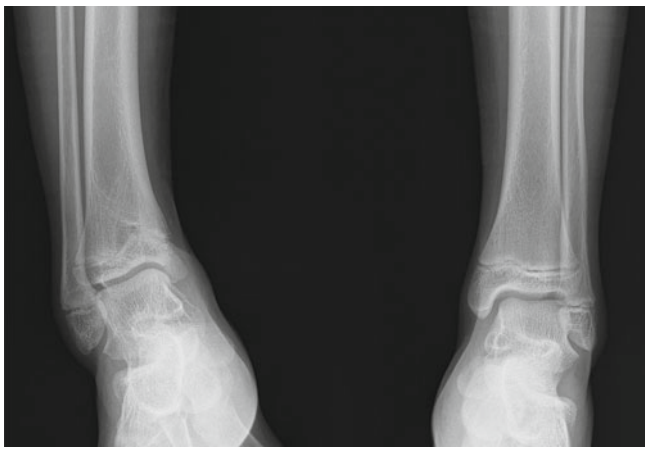


Fig. 31.7 A radiograph showing asymmetrical premature physeal closure of the distal tibia in the presence of normally growing distal fibula in a 13-year- and 8-month-old boy

bone realignment and fixation using an intramedullary locking nail or fixator-assisted deformity correction and consecutive lengthening over an intramedullary locking nail can also be performed in certain instances [69, 70]. However, while the technique of lengthening over an intramedullary nail using external fixation is used to reduce the time of external fixation, caution is required to prevent deep infection in patients who have a past history of bone and joint infection, or an open trauma [71, 72].

Juxta-articular deformity of the ankle may develop as a result of metaphyseal osteomyelitis of the distal tibia or a primary septic arthritis of the ankle joint, involving the physis. Asymmetric or symmetric premature physeal closure of the distal tibia can be seen along with a normally growing distal fibula (Fig. 31.7), or vice versa. When the length discrepancy between the tibia and fibula is severe, secondary distal tibiofibular dissociation can occur. Similar to the juxta-articular deformity of the knee, the ultimate goal of the treatment for juxta-articular deformity of the ankle is to obtain a painless, stable plantigrade foot with a functional, mobile ankle joint. For this purpose, early stabilization of the talocrural joint by leveling the ankle and restoration of the axial alignment and joint orientation are of paramount importance. Treatment of juxta-articular deformity of the ankle should be individualized after considering the patient's age, relative length discrepancy between the tibia and fibula, physeal integrity, presence or absence of lateral talar subluxation and wedge-shaped epiphysis, intra-articular epiphyseal irregularity due to epiphyseal deformation, compensatory foot deformity including subtalar joint subluxation/deformation, range of motion, surrounding soft-tissue condition, neurovascular status, LLD, and other factors, in addition to the deformity itself. For instance, the use of internal fixation may not be feasible in patients with a poor soft-tissue envelope and is contraindicated in the presence of an active infection. Circular external fixation is well suited for the management of patients with complex deformities and/or bone loss with a poor soft-tissue envelope and history or presence of infection. The specific advantage of external fixation is that adjustments to achieve the desired alignment can be performed gradually during the postoperative period. In certain complex scenarios, external fixators are utilized in conjunction with internal fixation to provide additional support in order to avoid hardware failure and development of a residual deformity [73].

Figure 31.8 depicts the author's systematic approach for assessment of the juxta-articular deformity of the ankle. First, the length of the tibia relative to the fibula is assessed, physeal integrity of the distal tibia and fibula is checked, and orientation of the ankle joint in the coronal and sagittal planes is assessed. The location and extent of premature physeal closure, angular or joint surface deformities, and secondary osteoarthritis can be determined by using MRI, bone scintigraphy, and CT, together with plain radiographs [74–76]. Documenting the magnitude of the angular deformity at other site(s) and LLD, if any, is also essential. When a juxta-articular deformity of the ankle, with either a neutral heel or a valgus heel, is associated with a relatively short fibula due to partial deficiency or premature physeal arrest in the presence of intact physis of the distal tibia (Fig. 31.9), it is imperative to determine whether lateral talar subluxation with or without compensatory subtalar varus deformity and wedge-shaped epiphysis of the distal tibia coexist. If possible, acute or gradual reduction of lateral talar subluxation by external fixation should be performed first. Other definitive and adjunctive surgical treatment methods that can be used alone or in combination include fibular lengthening [77, 78], supramalleolar corrective osteotomy [79–81], hemiepiphysiodesis, and occasionally intra-articular osteotomy [82]. Several techniques of supramalleolar osteotomy for acute deformity correction have been reported, including oblique transphyseal supramalleolar osteotomy [79], transverse supramalleolar osteotomy with translation, Wiltse translational step-cut osteotomy [80], and arc osteotomy. The oblique medial closing wedge osteotomy allows for correction of the valgus deformity of the ankle and offers the advantage of improved bone healing as minimal periosteal stripping is necessary and the deformity is corrected by hinging the osteotomy at a point along the bisector of the deformity [81]. Hemiepiphysiodesis of the medial aspect of the distal tibia can be performed by using a transphyseal screw [83, 84], staple, or a tension-band plate [85, 86]. External fixator-assisted acute correction of ankle valgus and fixation using a locking compression plate or multiple screws and gradual correction with external fixation are other viable options.

In contrast, when a juxta-articular deformity of the ankle, with an apparently neutral heel, varus heel, and

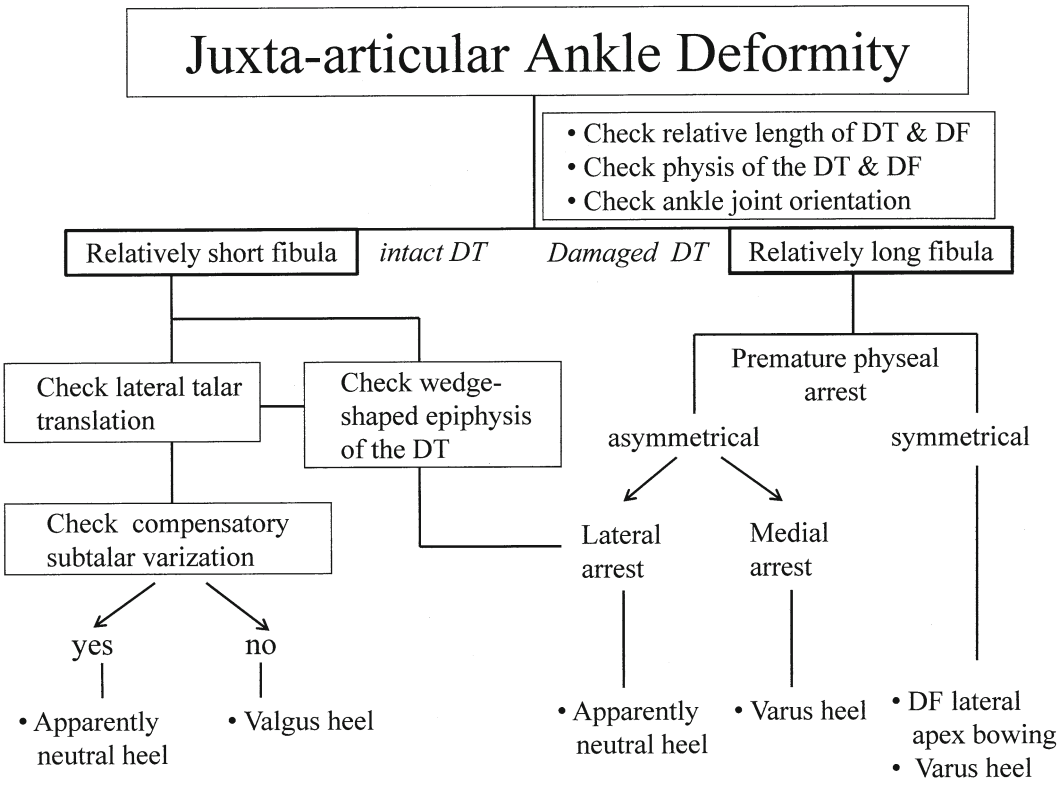


Fig. 31.8 Author's systematic approach for assessment of juxta-articular deformity of the ankle (see text). *DT* distal tibia, *DF* distal fibula

distal fibular lateral apex bowing, is associated with premature physeal arrest of the distal tibia in the presence of intact physis of the distal fibula, the location and extent of physeal damage of the distal tibia should be checked. In this situation, definitive and adjunctive surgical options that can be used alone or in combination include physeal bar resection, supramalleolar corrective osteotomy [79, 81, 87], distal fibular shortening and/or epiphysiodesis, tibial lengthening and subsequent epiphysiodesis, and occasionally an intra-articular osteotomy. When along with a juxta-articular deformity of the ankle there is a compensatory deformity of the subtalar joint and/or calcaneus, the correction of this component must also be included in the treatment plan. When supramalleolar corrective osteotomy is contemplated, the treatment option for acute versus gradual deformity correction should be selected on an individual basis after considering all the aforementioned confounding variables. In patients who have previously undergone surgery for reorientation of the talocrural joint, a second-stage lengthening of the proximal tibia and fibula may be needed, near or at skeletal maturity for equalizing the lower limb lengths.

Postinfectious Articular Deformity Causing Knee and Ankle Joint Incongruity

The residual deformity of septic arthritis of the knee and ankle joints may include partial epiphyseal destruction with a resultant joint incongruity and accompanying joint instability, subluxation, and impingement that may lead to rapid degenerative arthritis. In this situation, juxta-articular osteotomy alone cannot solve the problem of joint incongruity. Although intra-articular osteotomy is not a common treatment option for the hip, knee, or ankle, intra-articular osteotomy can be selectively performed to promote joint congruity and stability [82]. Paley [82] recently demonstrated, through case studies, that intra-articular osteotomy of the FH, the distal femur, or the tibial plafond is technically feasible and can yield successful results when performed for specific indications. When juxta-articular deformity of the ankle is associated with partial epiphyseal deficiency or deformation causing joint incongruity, an intra-articular osteotomy combined with or without juxta-articular osteotomy may be indicated. Theoretically, normalizing the joint articulation between the two adjoining bones is desirable for improving the joint congruency and stability.



Fig. 31.9 (**a**–**c**) A 25-year-old man presenting with a severe equinovalgus deformity of the foot. He had a history of undergoing incision and drainage and sequestrectomy for osteomyelitis of the entire fibula at 5 years of age. Compensatory proximal varus and distal anteromedial bowing deformity of the tibia, and a calcaneal varus deformity, is seen (**c**). (**d**) Tibial osteotomy was made at three levels, for the purpose of acute deformity correction and plate fixation for diaphyseal angulation, and gradual distraction for correction of proximal angular deformity using conventional Ilizarov construct and correction of distal angular deformity at the supramalleolar level using the hexapod system. Lateral calcaneal closing wedge osteotomy was also performed. (**e**–**g**) At 13 months after removal of the external fixator, which was applied for 9 months, he regained full function of the lower limb with a plantigrade foot and ankle joint motion from neutral to 20° of plantar flexion

However, an intra-articular osteotomy is a technically demanding procedure requiring intensive postoperative physiotherapy, and should be selectively performed. Furthermore, long-term follow-up results of such intra-articular osteotomies are currently unavailable.

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Iatrogenic Deformities

32

Austin T. Fragomen and S. Robert Rozbruch

Introduction

As physicians we strive to help our patients improve their pain and function. As orthopedic surgeons we do this through operative or nonoperative intervention. Our treatment choice is based on the standard of care and on our individual training and level of comfort. Despite the best intentions some patients have deformities after treatments that are unacceptable to the patients. *Iatrogenic deformity* refers to the circumstance where medical treatment contributed to the limb deformity. It may be that the management of a tibia fracture with casting resulted in a deformity which the patient now complains about as an adult (Fig. 32.1), or, perhaps, an adolescent with Blount's disease whose deformity was overcorrected resulting in a valgus deformity.

What Is Acceptable Alignment?

There has been an evolution in what constitutes acceptable alignment among our peers. Through the 1990s all focus was on achieving union with the following alignment dogma: less than 10° of angular deformity and less than 2 cm of shortening were well tolerated [1, 2]. Sarmiento stated, "We accept minor losses in length and alignment as small sacrifices in an effort to provide early function and decreased morbidity." Changes in stress patterns at the tibiotalar joint are not significant with angular deformities less than 10° in any plane. In over 25 years of observation small amounts of shortening and angulation have not been associated with osteoarthritis or diminished function at a later date and are cosmetically acceptable to the patient [3]. Many patients were left with large rotation deformities of the femur and tibia, bone shortening, or angular and translational deformities (Fig. 32.2). Surgeons have moved toward more stringent criteria with less than 5° of angular deformity considered ideal [4–6]. The adoption of intramedullary nailing and marginalization of casting as a standard of care for adult tibia fractures has given surgeons the ability to obtain more accurate fracture reductions [7]. Similarly, the management of pediatric femur fractures has moved toward surgery making traction and casting less popular [8] (Fig. 32.3).

What Are the Consequences of Malalignment?

It is not uncommon to see adult patients with childhood deformities present to the office with knee or ankle arthritis secondary to the long-standing malalignment. This poor alignment places abnormal or asymmetric stress across the adjacent joint(s) leading to premature cartilage breakdown [7–10]. This contention is supported by Vallier et al. who found that a tibial malunion of more than 5° was significantly associated with knee and ankle pain [4]. Palmu found that the treatment of femur fractures in children older than 10 years with traction led to a high incidence of deformities greater than 10° that he later correlated with the development of knee arthritis at a young age [8] (Fig. 32.4). We should strive to reestablish these normal parameters for bone alignment to avoid abnormal limb mechanics. This will help to prevent articular cartilage wear and pelvic and low back problems, and facilitate joint replacement, if and when needed. More importantly, if initial treatment results in unacceptable alignment then we are obligated to consider the long-term implications for this patient and suggest further

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Fig. 32.1 This segmental tibia fracture in an adolescent was treated with casting and went onto malunion. The patient subsequently developed knee and ankle osteoarthritis



Fig. 32.2 This distal tibial fracture was sustained during adolescence and was treated with casting. The angulation of 10° was accepted and the fracture healed uneventfully. The patient developed ankle arthritis years later with more joint space narrowing seen in the medial aspect of the tibiotalar joint



Fig. 32.3 This fracture was treated in adolescence with traction and casting resulting in 16° of varus and 3 cm of shortening



Fig. 32.4 This varus femur malunion resulted in malalignment that developed medial compartment osteoarthritis years after the injury

surgical correction for this patient. If the surgeon is not comfortable performing this "residual correction" then the patient should be referred to a specialized center. Many patients will elect no further treatment or may not be candidates for further intervention, but the malalignment needs to be acknowledged.

What Are the Legal Implications of Iatrogenic Deformities?

Nearly 15 % of orthopedic surgeons face a malpractice claim annually [11]. Restricting the criteria for acceptable deformity is analogous to "narrowing the strike zone." This places increased responsibility on the surgeon to have a "perfect" result, thus making our job more difficult. If a malunion is a failure and the criteria for malunion are now broadened then there are legal implications. Patients are glad to have their concerns about their deformity legitimized, but their next question is why the deformity occurred and whether this was malpractice. Based on our experience, in most cases, the best policy is to concede that the result is not ideal, reassure that every effort was made to provide an optimal result and that there was no medical negligence, and counsel that further surgery to deliver a superior outcome is recommended. The greatest predictor of payment to a plaintiff is the severity of the patient's disability [12]. This implies that the best course of action is to do everything possible to improve their condition and decrease disability. At no time should the surgeon abandon or blame the patient. A good doctor-patient relationship and better preoperative discussions of known complications will help to calm the impulse to bring legal action against the physician [13, 14]. Contact the hospital legal department and identify this patient as "at risk" for litigation. The hospital will often have patient services mitigate any feelings of discontent or help finance further treatment. When a patient does seek legal action, an angry patient will be less likely to settle the case. So, one must do as much as possible to avoid a hostile relationship with the patient and his or her caretakers. One must keep in mind that we are judged not only by our successes but also by how we handle adversity. These difficult situations are hurdles along the career path of every orthopedic surgeon.

Common Iatrogenic Deformities

Tibia Malunion

The immature skeleton's ability to remodel a "malunion" is determined by the number of years of growth remaining. If the remodeling potential of an angulated bone is limited, then the fracture is considered a malunion and warrants intervention. If a child of advanced age presents with a displaced long bone fracture of the lower extremity that will likely go on to a malunion (impending malunion) if treated with cast immobilization, the surgeon will often select operative treatment to help ensure acceptable alignment at skeletal maturity. The current criteria for "acceptable" alignment in pediatric fractures fall into two groups. Children 8 years old or less often remodel angular deformities of up to 10°. In children older than eight and in adolescents, angular deformity of up to 5°, 1 cm of shortening, and less than 10° of rotational deformity is acceptable [15]. Most remodeling occurs within 2 years after fracture healing [15]. Due in part to a robust periosteum and rapid healing, most tibia fractures in children can be treated with casting while still maintaining acceptable alignment, length, and rotation [16]. However certain fractures are difficult to manage in a cast or are at high risk for displacement and malunion and are often treated surgically: high-energy unstable fractures, open fractures, fractures with soft tissue injury or compartment syndrome, oblique fractures with an intact fibula, floating knee, and fractures in older children [16–19]. Operative management of tibia factures is most commonly performed with elastic titanium intramedullary (IM) nails [20]. Circular external fixation has been shown to be superior to monolateral fixation for unstable diaphyseal fractures of the tibia in children and adolescents and is gaining popularity [21]. Circular fixation may also be equivalent to elastic IM nailing in adolescents [22]. External fixation may be superior to casting as it allows for ease of monitoring and management of compartment syndrome, which remains one of the most common complications of pediatric tibia fractures [23, 24]. Use of the programmable, six-axis circular fixator in high-energy pediatric tibia fractures has proved successful affording very accurate alignment and early weight bearing [25, 26]. However, the use of external fixation is associated with a small incidence of refracture and fracture through the pin sites [27].

Case 1

A 12-year-old male presented with a noticeable deformity of the lower leg including varus, external rotation, and shortening. He had sustained a Salter–Harris type II fracture of the distal tibia at the age of 8 years. A metaphyseal osteotomy was performed for deformity correction and was stabilized with a plate and screws. The growth plate became tethered medially driving the epiphysis into varus and slowing the growth of the entire physis (Fig. 32.5a, b). This was recognized by his orthopedist who had been following him yearly after the injury. He was referred for deformity correction and limb lengthening. The sum of these deformities led to significant functional problems with walking and running and put him at increased risk for developing arthritis of the ankle and low back pain. He was noted to have limited ankle dorsiflexion of the injured side compared with the unaffected left ankle. His external rotation deformity of the tibia was 20°.

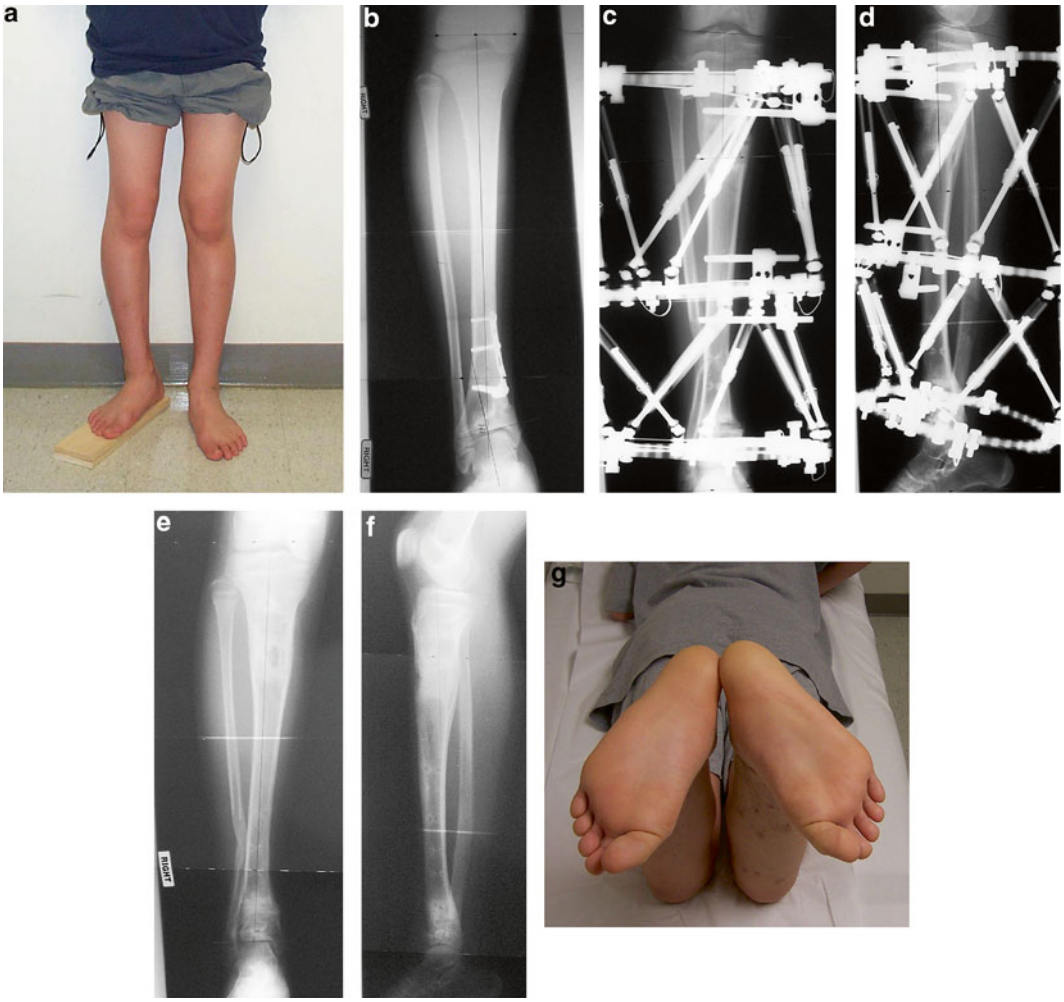


Fig. 32.5 (**a**) This patient has a malunion with shortening, external rotation, and varus. (**b**) AP X-ray shows retained hardware, medial physeal tethering, and distal varus deformity. (**c**, **d**) AP and lateral radiographs during treatment demonstrating the two-level osteotomy and double-level circular fixation for deformity correction and lengthening. (**e**, **f**) These X-rays show healing of both osteotomies with accurate correction of the underlying deformities. (**g**) Restoration of the axial alignment of the tibia is assessed with the prone exam

He had a 2.5-cm shortening of the tibia. Surgical planning included calculation of the future shortening that would result from formal closure of the distal tibial physis. Correction with the six-axis circular fixator was considered a good option. This would allow for deformity correction and lengthening. The entire distal tibial growth plate was closed surgically. A distal metaphyseal osteotomy was created with an osteotome to allow for gradual correction of the varus and rotational deformities. Lengthening through the same osteotomy site has two disadvantages: the patient may develop a significant equinus, and the time to consolidation of the lengthening regenerate would be lengthy. To address these potential problems a second more proximal osteotomy was performed for the lengthening (see Fig. 32.5c, d). The final result was correction of the deformities including length with a full functional recovery (see Fig. 32.5e–g) He had a contracture of the gastrocnemius that fully resolved with physical therapy 6 months after frame removal. Another option would have been to lengthen the gastrocnemius muscle tendon for a more rapid recovery of ankle motion.

Case 2

A high-energy closed tibia fracture in a 13-year-old male was treated with a monolateral pin-to-bar external fixation as the definitive stabilization. The reduction resulted in 2 cm of lateral translation of the distal fragment and 10° of apex anterior angular deformity (Fig. 32.6a, b) The surgeon explained that this was acceptable alignment and that he was unable to improve it with closed reduction; because the deformity was acceptable open reduction was not indicated. The family sought a second opinion. This sagittal plane deformity is greater than 5° and will likely not remodel in this adolescent patient. The clinical concern about residual apex anterior deformity of the tibial shaft is that it may create

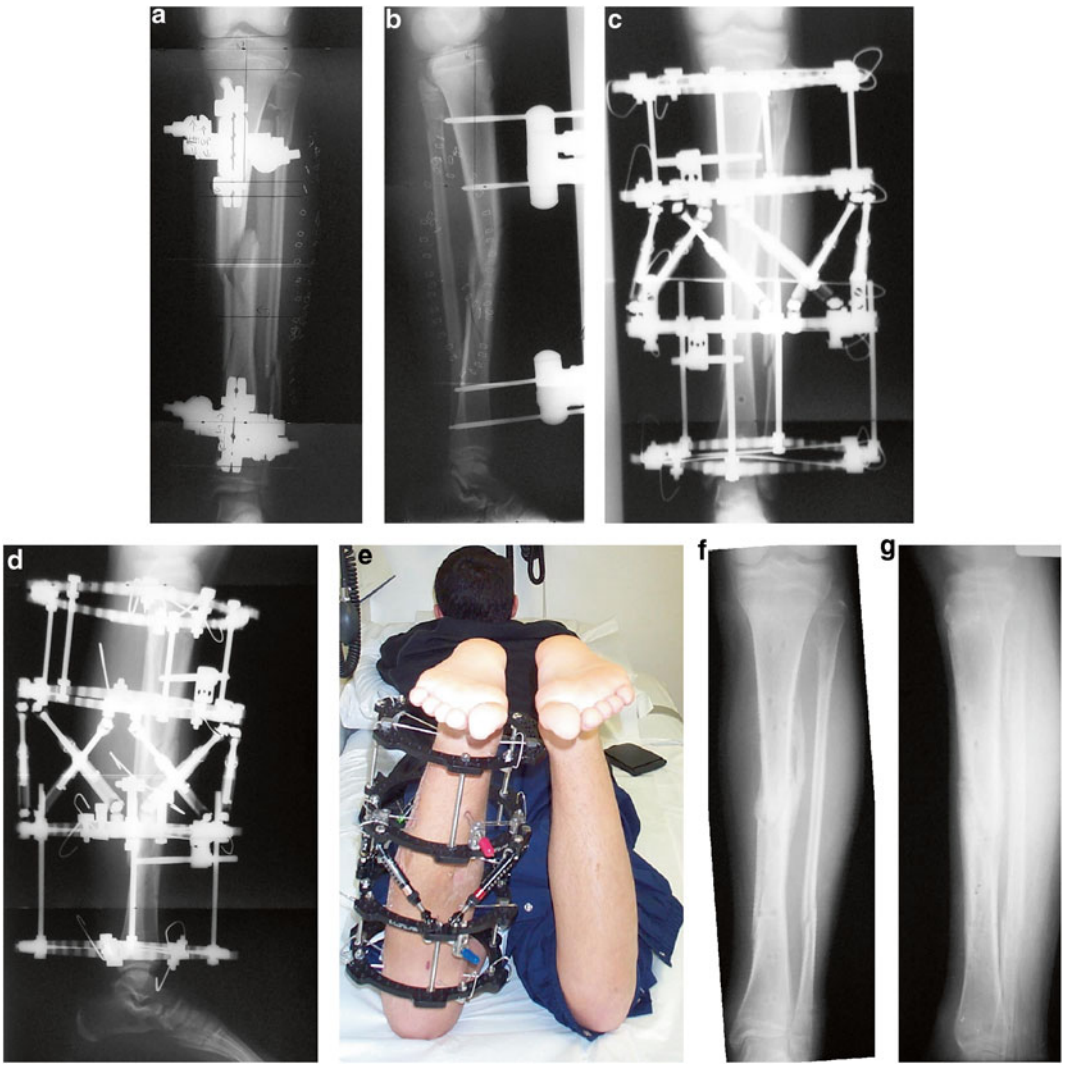


Fig. 32.6 (**a**, **b**) AP and lateral radiographs show a high-energy tibial fracture treated with temporizing external fixation. Soft-tissue swelling and damage made internal fixation a poor option. (**c**, **d**) The circular external fixator provided superior stability and allowed for early weight bearing. (**e**) The prone exam is important to confirm that axial alignment has been restored. (**f**, **g**) The final result is a healed fracture with excellent alignment

abnormal wear on the knee and ankle joints, increases stress on the anterior cruciate ligament (ACL), and prevents full leg extension. Trepidations about the fixation construct included further loss of reduction, impending malunion, and even possible nonunion. Treatment with a six-axis circular fixator using the Taylor Spatial Frame (TSF) (Smith & Nephew, Memphis, TN) was provided. This device allowed for a percutaneous surgery, an accurate and gradual correction of the residual deformity, avoidance of the growth plates, and weight bearing as tolerated ambulation during the entire treatment period (see Fig. 32.6c–e) After 3 months of wearing the external fixator, the final result was a near-anatomic bony alignment and no functional impairment (see Fig. 32.6f, g).

Fibular Malunion

Fibular fractures in children and adolescents commonly heal without incident. In rare circumstances growth arrest of the distal fibular physis can occur causing progressive fibular shortening. This leads to valgus orientation of the hindfoot, widening of the syndesmosis, and early ankle joint degeneration [28–33]. Once recognized, this fibular shortening and syndesmotic laxity can be corrected surgically [34–36]. Manoudis et al. reported a case of fibular shortening from physeal arrest corrected by open osteotomy, acute lengthening, grafting with tricortical iliac crest, and internal fixation [37]. An open release of the syndesmosis was necessary to reduce the fibular tip into its proper location. Other authors

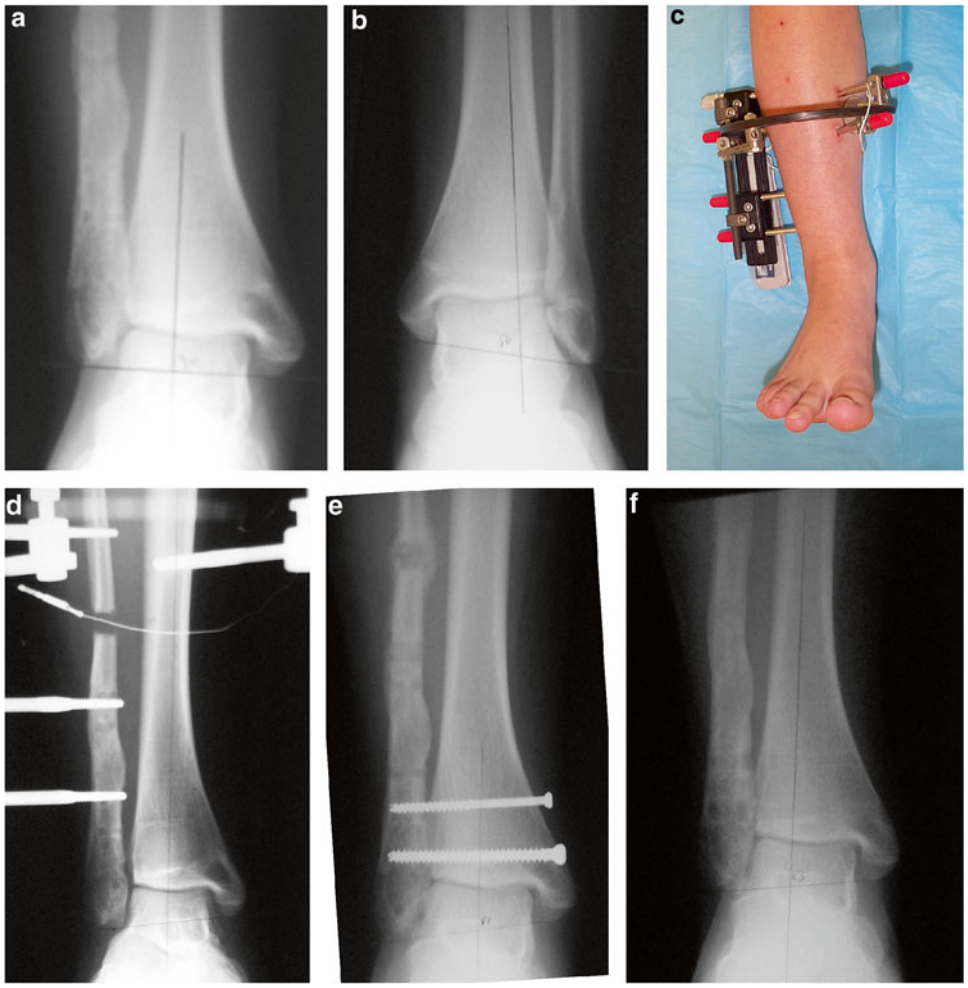


Fig. 32.7 (**a**, **b**) AP radiographs of the shortened right fibula and the normal left fibula. Tibial alignment is within normal. (**c**, **d**) An external fixator was used to lengthen the fibula using half pins. (**e**) At the time of frame removal the syndesmosis was stabilized with two screws. (**f**) Final radiographs show restoration of the length of the fibula and removal of the screws

have reported on the same technique without the need for syndesmotic dissection [29]. Alternatively, a percutaneous technique can be employed to both lengthen the fibula through distraction osteogenesis and utilize distraction histiogenesis to stretch the syndesmotic ligaments and achieve distal migration of the fibular tip.

Case 3

A 13-year-old female presented with a valgus deformity of the hindfoot after premature fibular physeal closure. The patient had previously undergone curettage of a benign distal fibular bone cyst (without apparent physeal involvement) with prophylactic plating to prevent fracture of the distal fibula. The hardware was removed 1 year later without incident. The distal fibular physis closed prematurely postoperatively leading to a shortening deformity of the fibula over time. The etiology of the physeal injury is not clear but an iatrogenic cause could not be excluded. The patient's hindfoot drifted into valgus when compared with the other side. Radiographs revealed a shortened fibula with a distal physeal closure. X-rays of the contralateral ankle were taken to establish the normal fibular length and talocrural angle (Fig. 32.7a, b). This deformity was unacceptable as it would lead to asymmetric wear of the lateral talus and possibly arthritis. Fibular shortening of 1.5 cm was measured. The treatment plan included lengthening of the fibula. The tibia was included in the proximal fibular ring block to protect the proximal tibiofibular joint from subluxation and ensure distal migration and reduction of the fibular tip (see Fig. 32.7c, d). The fibula was lengthened with the external fixator. When the correct length was obtained the patient was brought back to the operating room for insertion of syndesmotic screws and early removal of the frame (see Fig. 32.7e). The patient's hindfoot alignment was corrected with no loss of ankle motion (see Fig. 32.7f).

Transphyseal ACL Reconstruction Causes Premature Physeal Closure

An option for ACL reconstruction in older children (age 11–14 years) is the transphyseal tunnel technique. Most authors report minimal complications with this method including an average growth plate disturbance risk of 3 % [38–44]. Kumar et al. reported on 32 children with an average

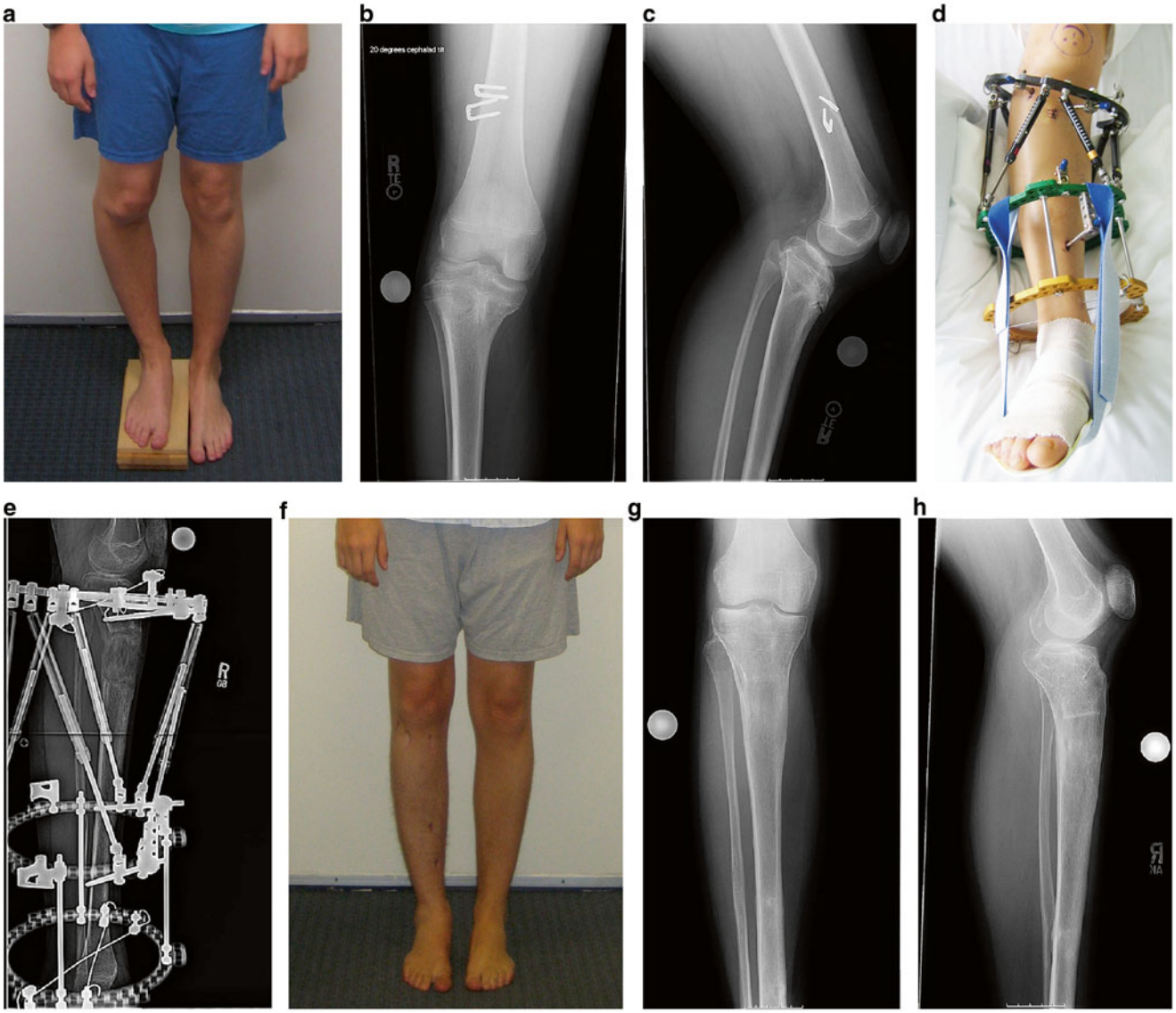


Fig. 32.8 (**a**) This clinical photo shows a proximal tibial deformity with shortening, varus, and malrotation. (**b**, **c**) AP and lateral X-rays show varus and recurvatum angulation with closure of the anteromedial physis. (**d**, **e**) This photo shows the early postoperative external fixator mimicking the deformity. The radiograph was taken near the end of the correction showing the proximal tibial lengthening. (f–h) Final clinical photo and radiographs demonstrate full correction of the deformity

age of 11.25 years where the transphyseal reconstruction was used with one case of mild valgus and no leg length discrepancy [38]. Similarly, transphyseal surgery in a series of 15 patients, average age 12.8 years, resulted in one case of valgus malalignment of the knee [39]. Asymmetric growth of the distal femoral physis is the most commonly reported mechanism for the valgus deformity [41]. Other series report no adverse trauma to the physis with this technique, but authors caution to make sure that the graft bone blocks are not in contact with the physis [42–44]. Physeal sparing reconstruction carries minimal risk to the growth plate but may not be as reliable with regard to maintenance of ligamentous stability of the knee [45]. Although the risk of growth plate disturbance around the knee is small, its occurrence in some cases is undeniable and such iatrogenic angular deformities may require surgical correction.

Case 4

A 15-year-old male presented with a deformity of the proximal tibia including varus, recurvatum, and shortening. At age 12 he sustained an ACL injury. The ACL was reconstructed using the transphyseal technique in the tibia and an over-the-top technique in the femur. The anterior-medial proximal tibial physis was likely damaged. A progressive angular deformity developed with significant retardation of physeal growth (Fig. 32.8a–c). The radiologic deformities included

15° of varus, 28° of apex posterior (recurvatum), and 4.5 cm of shortening. The clinical deformity included 10° of hyperextension of the knee and normal knee ligamentous stability. Growth remaining discrepancy was calculated to be an additional 1 cm. Treatment was closure of the remainder of the proximal tibial physis to prevent further deformity, proximal tibial osteotomy, and TSF application for deformity correction and lengthening (see Fig. 32.8d, e). The latest follow-up at 12 months post-frame removal shows restoration of length and alignment, and a full functional recovery is expected (see Fig. 32.8f–h).

Femur Malunion and Growth Disturbance

Distal femur physeal fractures are afflicted by a high rate of physeal arrest and subsequent deformity [46]. Displaced fractures and Salter–Harris II injuries are predictors of a poor outcome with a 40 % risk of physeal bar formation [47]. Surgical reduction and fixation are recommended, and smooth Steinman pins crossing the physis do not seem to further increase the already high risk of physeal arrest [46, 48].

Distal femur malunion without growth plate arrest is another complication of pediatric femur fractures. Although the skeletally immature femur has a tremendous remodeling potential, angulation and shortening may not be tolerated as well as many surgeons believe. In children over 10 years old varus-valgus angulation of greater than 5° and procurvatum-recurvatum of greater than 10° should not be accepted because such deformities can be associated with premature knee arthritis [8].

Treatment of diaphyseal and distal femoral deformities that occur during childhood and subsequently present in adulthood can be accomplished with various implants including IM nail, plate and screws, and external fixation. The challenge is managing the femoral shortening that is often present in these individuals. External fixation allows for deformity correction and lengthening simultaneously [49]. Integrated fixation techniques provide the best of both devices using external fixation to achieve length and deformity correction and then converting to internal fixation while the immature bone consolidates [50–52]. The newer internal lengthening nails provide an all-internal correction of deformity and bone lengthening solution [53].

Case 5

A 19-year-old male presented with femoral valgus and shortening. He had been struck by a vehicle when he was 11 years old sustaining a distal femur physeal fracture. The fracture was stabilized with internal fixation. It is not known whether the fracture was reduced anatomically or was stabilized with residual valgus angulation. If the initial postoperative alignment was appropriate then this may not qualify as an iatrogenic deformity. He was an avid soccer player but was having increasing difficulty playing due to the lower limb deformity and knee pain. Partial closure of the lateral distal femoral physis caused a valgus deformity and 3 cm of shortening (Fig. 32.9a, b). The patient was at risk for knee arthritis, hip, and low back problems. The proposed treatment would address both the valgus deformity and the femur shortening. A distal osteotomy with lengthening is best done with a circular fixator, but the frame would need to be on the leg for a long time and femoral rings are very uncomfortable. The LAP (lengthening and plating) technique was selected. A femoral osteotomy was created and stabilized with a TSF (see Fig. 32.9c). The correction was performed gradually with lengthening. Once the acceptable length and alignment were reached, the patient was brought back to surgery for insertion of a plate and screws and removal of the frame (see Fig. 32.9d, e). The femur healed and the plate was later removed (see Fig. 32.9f, g). He returned to play soccer without any limitations.

Case 6

A 34-year-old female presented with a deformity of the right femur and tibia. She had a femur fracture when she was 11 years old that was treated with skeletal traction followed a few days later by casting. The traction pin was placed into the proximal tibia at that time. She presented with a deformity of the right femur including valgus and shortening, and deformity of the right tibia including recurvatum and shortening. On examination she had knee hyperextension of 20°, valgus alignment, and lateral knee instability. The radiographs revealed a 20° apex posterior deformity of the proximal tibia with 15-mm shortening and 6° of valgus, relative proximal migration of the proximal fibula, and 5° valgus deformity of the distal femur diaphysis with 20 mm of shortening (Fig. 32.10a, b). The patient has knee instability and pain. She was at a high risk for developing premature knee arthritis and would have likely required living a life of activity modification. The surgical plan included femur surgery to both correct the valgus deformity and lengthen the femur. This was accomplished with a retrograde IM internal lengthening nail. The plan also included a gradual correction of the recurvatum and valgus deformities of the tibia with lengthening. This included translating the intact fibula distally to re-tension the lateral collateral ligament (see Fig. 32.10c, d). After removal of the hardware the patient is recovering well. The deformities have been corrected and the knee is stable with normal extension (Fig. 32.10e–g).

Vascularized Fibula Graft Nonunion and Valgus Ankle Deformity from the Fibular Donor Site

Reconstruction of the tibia after tumor resection with contralateral vascularized fibular autograft is an effective limb-sparing procedure [54]. The rate of nonunion can be as high as 20 % [55]. When the metaphyseal segment is small,

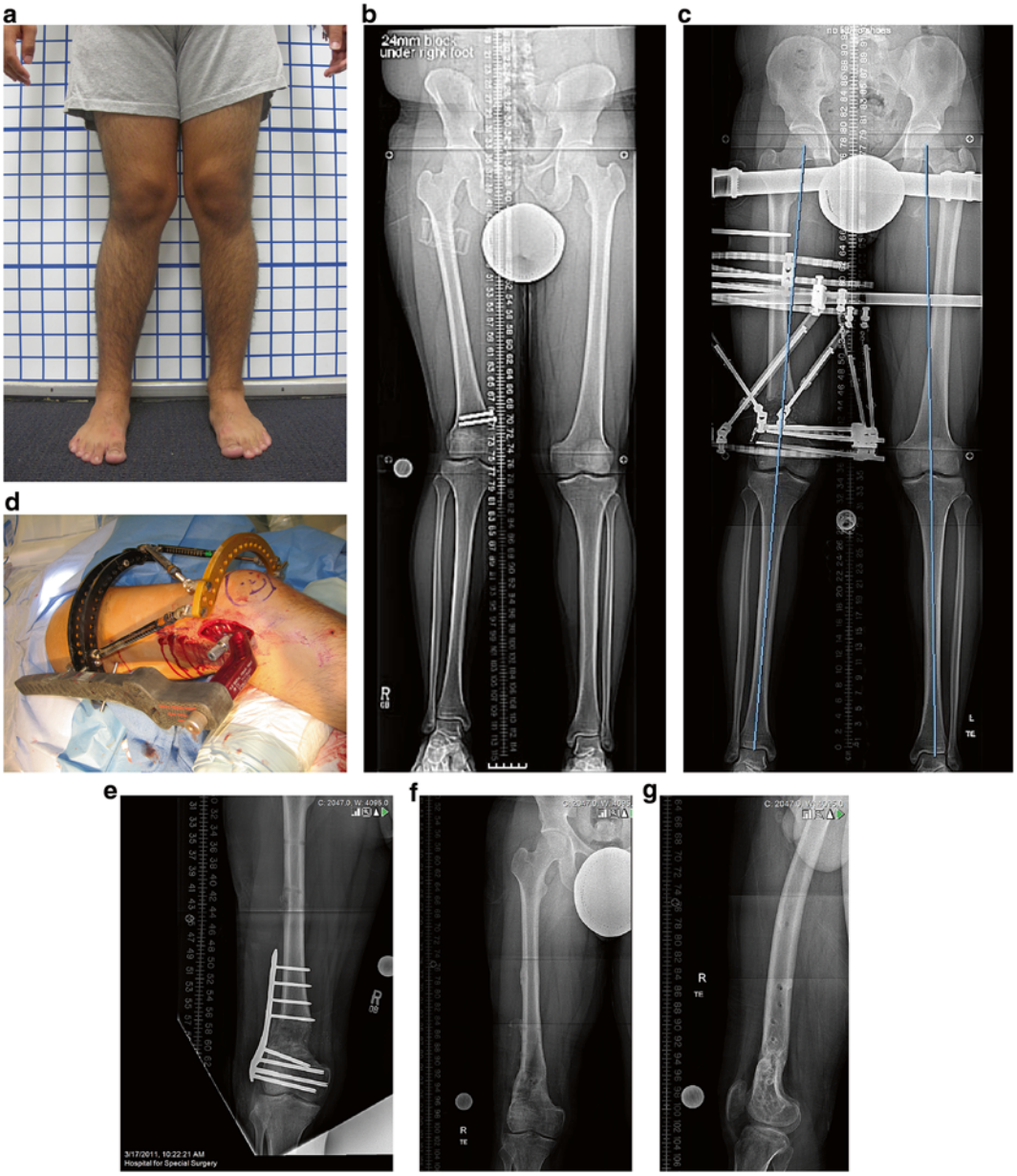


Fig. 32.9 (**a**, **b**) This clinical photo and radiograph show a valgus malunion of the distal femur with shortening. The two medial screws suggest a Salter–Harris 2 fracture with fixation. This X-ray was taken with the patient standing on blocks. (**c**) External fixation was used to distract and angulate the distal femur. (**d**) The external fixator was mounted differently to expose the lateral femur for subsequent plating. Here the plate is being inserted. (**e**) This radiograph shows the plate spanning the new regenerate bone. (**f**, **g**) Final radiographs show full union of the osteotomy and ideal alignment

achieving stability with internal fixation can be challenging. Circular fixation is especially effective in these situations.

Harvesting of the fibula for reconstruction of a bone defect with a vascularized fibular autograft can be associated with valgus deformity of the donor-side ankle [56–58]. Tibia-fibula syndesmosis synostosis has been recommended to prevent this deformity, but valgus can occur even after this measure [59]. Prevention is also possible and has been successful with stabilization of the fibula to the tibia using a distal syndesmotic screw [58]. Correction of the iatrogenic valgus deformity with external fixation through a supramalleolar osteotomy is effective in the short term but can be complicated by recurrence of the deformity [57]. In this case report the guided-growth technique is used to restore the distal tibial alignment resulting in normal ankle joint orientation at maturity.

Case 7

A 10-year-old male was referred for management of a nonunion and deformity at the junction of the proximal tibial metaphysis and a vascularized fibular autograft. The patient had an osteosarcoma excision of the proximal tibia reconstructed with contralateral fibular autograft and flap.

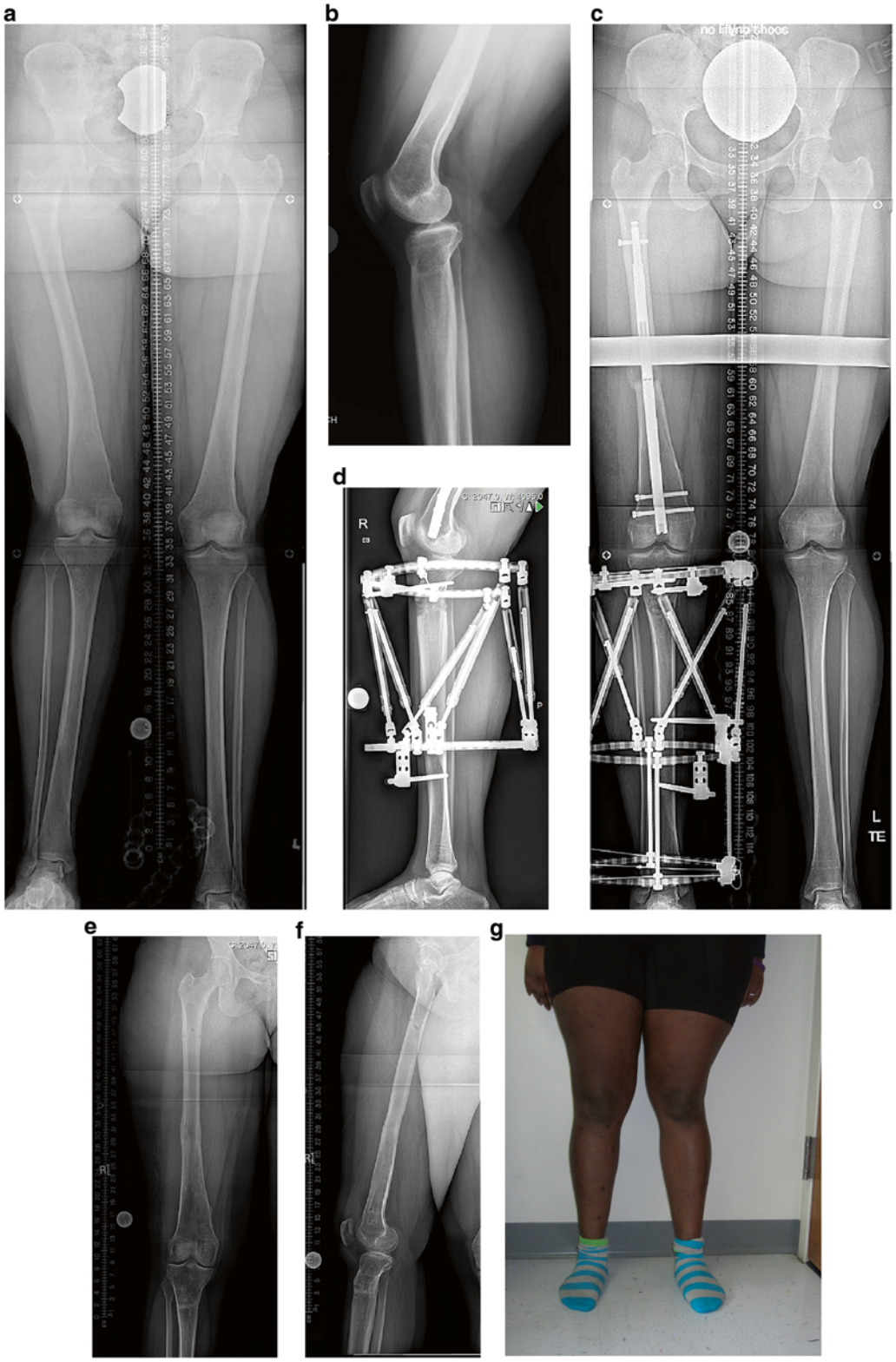


Fig. 32.10 (**a**, **b**) These radiographs show valgus of the distal femur and proximal tibia as well as a 20° recurvatum deformity of the proximal tibia. (**c**, **d**) Correction of the valgus femur malunion and shortening was performed with an internal lengthening nail. The tibia valgus and recurvatum as well as the superior migration of the fibula were all corrected with a tibial osteotomy and external fixation. (**e**–**g**) Final radiographs and a clinical photo show full healing and a successful correction of the deformities

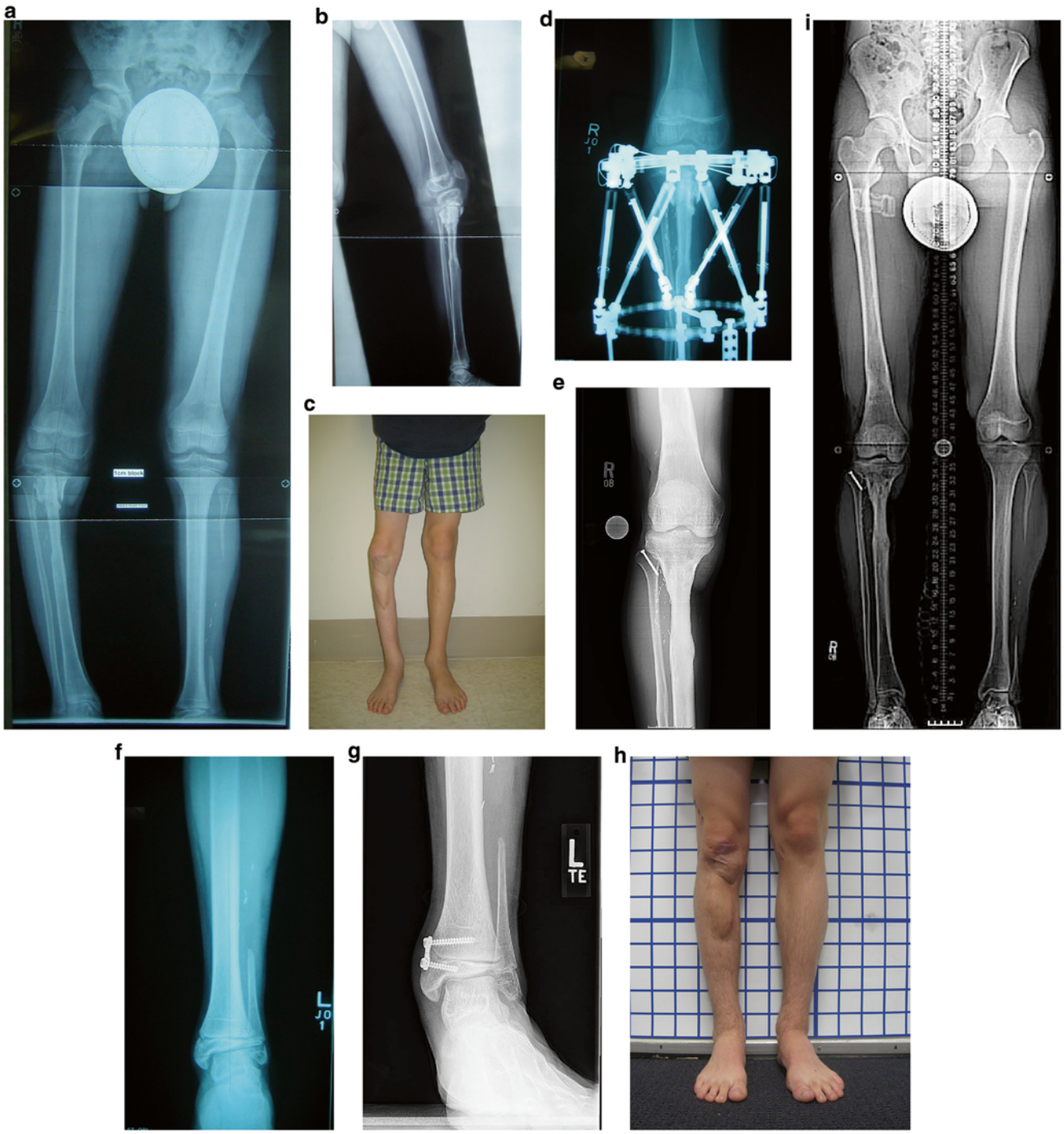


Fig. 32.11 (**a**–**c**) The radiographs show tibial shortening and a nonunion with angulation of the proximal graft-host junction. The vascularized fibular graft has hypertrophied but has failed to unite. The patient had a mobile nonunion with bowing and a lateral thrust. (**d**, **e**) Fixation with fine wires afforded enough stability for bone healing to progress. (**f**, **g**) Valgus deformity was developing at the donor-side ankle, which was treated with guided growth of the physis. (**h**, **i**) The final results were correction of the deformity, bony union, and correction of the donor-side ankle valgus

The patient did not have recurrence of the tumor and the diaphyseal junction of the graft healed satisfactorily. He had pain, a varus and flexion deformity of the proximal tibia, and difficulty walking despite bracing. Radiographs revealed a nonunion of the proximal graft junction (Fig. 32.11a–c). The treatment plan was to repair the nonunion and correct the deformity with fine-wire fixation. The proximal tibial segment was very small making it well suited for this technique.

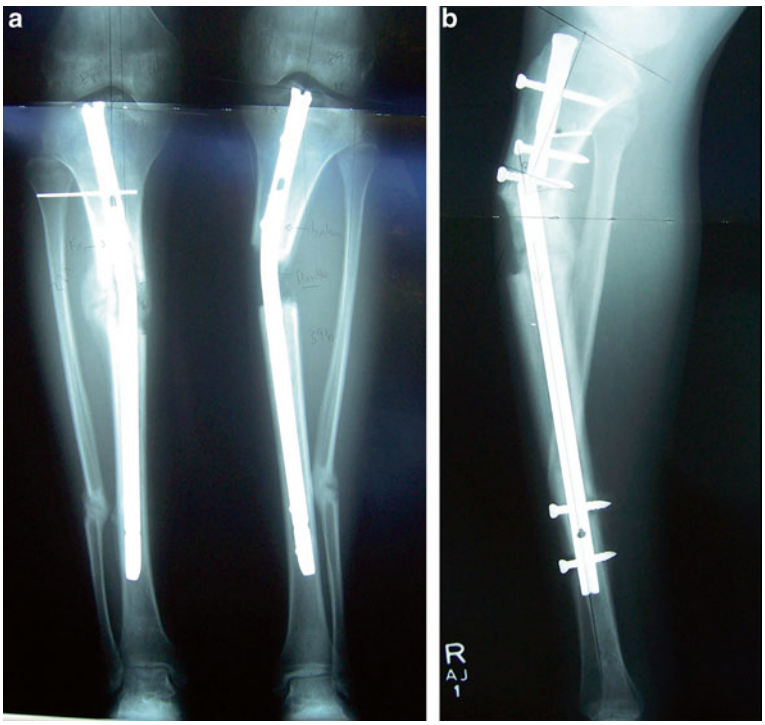


Fig. 32.12 (**a**, **b**) A patient underwent bilateral tibial lengthening surgery by lengthening the tibia with an external fixator over an IM nail. Once the desired length was achieved the fixator was removed and the patient started weight bearing too early. These AP and lateral (respectively) radiographs show collapse and angulation through the tibial lengthening site. The immature bone could not support weight bearing and failed with angulation of the regenerate and fracture of the internal fixation

A percutaneous repair of the nonunion was performed by making a small incision over the nonunion site and passing a drillbit multiple times and in different planes to stimulate the adjacent bone to bleed. A curette was then passed through the drill holes to scrape the bone surfaces and further stimulate bony bleeding. This approach stimulated a healing response while minimizing trauma to the poor soft tissue envelope (see Fig. 32.11d). The minimally invasive wires and pins accomplished the same goals of soft tissue preservation and provided excellent bony fixation. The deformities were corrected with the TSF and the nonunion was compressed (see Fig. 32.11e). Weight-bearing ambulation was allowed postoperatively. The patient was also noted to have an iatrogenic deformity of the contralateral ankle. After the fibula was harvested the ankle had drifted into a valgus deformity. Radiographs showed a valgus deformity of the distal tibia. This was addressed with guided growth using a small extraperiosteal plate and non-locking screws to tether the medial physis and improve the distal tibial valgus (see Fig. 32.11f, g) After frame removal of the side with the nonunion, the graft-host junction of the proximal tibia had healed and the deformities were corrected including the contralateral distal tibial alignment. The patient underwent a distal femoral lengthening of the extremity with the proximal tibial autograft 2 years later to make up for the residual leg length discrepancy. It was felt that the ipsilateral tibia had already suffered enough trauma making a femoral osteotomy a wiser course of action (Fig. 32.11h, i).

Deformity from Tibial Lengthening

Lengthening of the tibia from congenital or acquired shortening can be accomplished using the methods of Ilizarov. However there are unbalanced forces from the posterior and lateral muscle groups acting upon the bone that cause predictable deformity: valgus and procurvatum. The challenge of distraction osteogenesis is to create a straight regenerate, thereby avoiding the creation of a tibial deformity. This iatrogenic deformity needs to be anticipated whether using external fixation or an internal lengthening nail. A further problem is bending of the new regenerate bone when the external fixator is removed prematurely. Even if the lengthening is done correctly, the new bone is still at risk for developing angular deformity (Fig. 32.12a, b). Both deformities that occur during lengthening and after lengthening are considered malunions and are considered iatrogenic. The treatment of these malunions is either casting to prevent further angulation, internal fixation with osteotomy to correct the current deformity, or external fixation with osteotomy. If the immature lengthening regenerate bends and is managed early enough, it can be manually realigned without osteotomy and stabilized with external or internal fixation.

Contracture of the ankle and knee joints is a common complication of tibial lengthening surgery. Tightening of the gastrocnemius muscle and fascia pulls the ankle into equinus and the knee into flexion. A tibial lengthening of greater than

13 % of the length of the tibia bone and congenital etiology were risk factors for an equinus contracture. Further, gastrosoleus recession restored preoperative motion in all cases [60]. Acute correction of these contractures should be avoided to prevent undue stretching on the already tight nerves, especially the tibial nerve at the tarsal tunnel. When these contractures occur before the desired tibial length has been achieved then consideration should be given to stopping or slowing down the lengthening process to allow for improved range of joint motion.

Case 8

A 10-year-old male presented with shortening and deformity of the left tibia. He had no previous trauma, no syndrome, and no femoral involvement. He was felt to have a very mild fibular hemimelia resulting in angular deformity and shortening of the affected tibia and fibula (Fig. 32.13a–c). He had normal range of motion and stability of both ankle and knee joints. There was 8 cm of shortening with additional shortening expected as growth continued. The goal was to obtain at least 5 cm of shortening, which was an 18 % lengthening of the tibia. The patient was treated with double-level tibia osteotomies lengthened at 1.0 mm per day for a total of 2.0 mm per day of limb lengthening (see Fig. 32.13d, e). This rate was chosen to prevent premature consolidation at either lengthening site. The result of this rapid lengthening was equinus contracture of the ankle and knee flexion contracture (see Fig. 32.13f). The patient and his mother were unable to stretch the contracture despite physical therapy. The patient underwent a gastro-soleus recession at the mid-distal-leg level with attachment of a foot ring and hinges. The ankle joint was distracted acutely 3 mm to prevent articular cartilage compression. The correction of the equinus contracture was performed gradually with the foot ring. A spring-loaded hinged brace was attached to the tibial ring to help correct this knee flexion deformity (see Fig. 32.13g). Further lengthening was stopped. After frame removal a total of 5 cm of length had been achieved, and ankle and knee range of motion were eventually normal (see Fig. 32.13h, i). The patient can undergo further lengthening in the future.

Translational Deformity

Iatrogenic translational deformity can occur when an osteotomy is performed at a location away from the apex of the deformity. These osteotomies require a deliberate translation of the bone fragments to ensure that the joints remain in line with the long axis of the tibia and the mechanical axis of the femur. These intentional translations may look like an accidental iatrogenic deformity when in fact they are planned and desirable. Patients will often need reassurance that the translational "deformity" was intended.

Case 9

A 42-year-old female had a correction of Blount's disease at the age of 10 years. A varus and flexion deformity recurred over time and led to medial compartment arthritis (Fig. 32.14a, b). Preoperative radiographs showed a medial mechanical axis deviation (MAD), 17° of varus, and the apex of the deformity at the knee joint line (see Fig. 32.14c, d). The osteotomy performed distal to the tibial tubercle would require a lateral translation of 24 mm (see Fig. 32.14e). This would create an iatrogenic deformity of the tibia in order to restore the mechanical axis and the proper joint orientation angles, the medial proximal tibial angle (MPTA). A circular fixator was used to achieve a gradual correction of the deformity and translate the distal fragment laterally (see Fig. 32.14f). Final radiographs demonstrate an apparent iatrogenic deformity of the proximal tibia (see Fig. 32.14g–j). However, the MAD and MPTA have been restored, and the bone healed without incident. It is likely that this translation, although technically correct, will cause difficulty with conversion to a total knee replacement.

Box 32.1. Highlights

- The ability to correct deformity has improved, making anatomic restoration of bone more attainable.
- Posttraumatic malunions are often seen with displaced pediatric tibial shaft fractures and have directed initial treatment in some adolescents toward surgical fixation rather than casting.
- Transphyseal ACL reconstruction can, in rare cases, result in growth disturbance.
- Limb lengthening often creates predictable deformity that the surgeon must be able to anticipate, recognize, and treat.
- Intentional translation after osteotomy for periarticular deformities maintains proper limb alignment, but the bone may appear deformed to the untrained eye.

Summary

Iatrogenic deformity takes many forms (Box 32.1). The surgeon must consider factors such as the location and magnitude of the deformity and the growth remaining when deciding on treatment options. The tools available to repair



Fig. 32.13 (**a**–**c**) These radiographs show a two-level, congenital deformity of the left tibia with significant shortening. (**d**, **e**) The proximal osteotomy allowed for mild deformity correction and lengthening. The distal osteotomy allowed for deformity correction and mild lengthening. (**f**) Ankle and knee contractures are apparent during the lengthening process. (**g**) The foot ring is seen in an overcorrected position to prevent recurrence. The dynamic, spring-loaded brace is seen bolted onto the proximal tibial ring delivering an extension moment. (**h**, **i**) Final X-rays show full healing, correction of the deformity, and successful attainment of length

malunions and restore alignment have improved, and the outcome of surgical reconstruction has become more predictable. Patients with deformity need not suffer for the remainder of their lives or until they are old enough for a joint replacement. Further research will help determine more accurate parameters for acceptable alignment. Advances in technology will continue to make anatomic restoration of deformities and fractures more easily attainable.

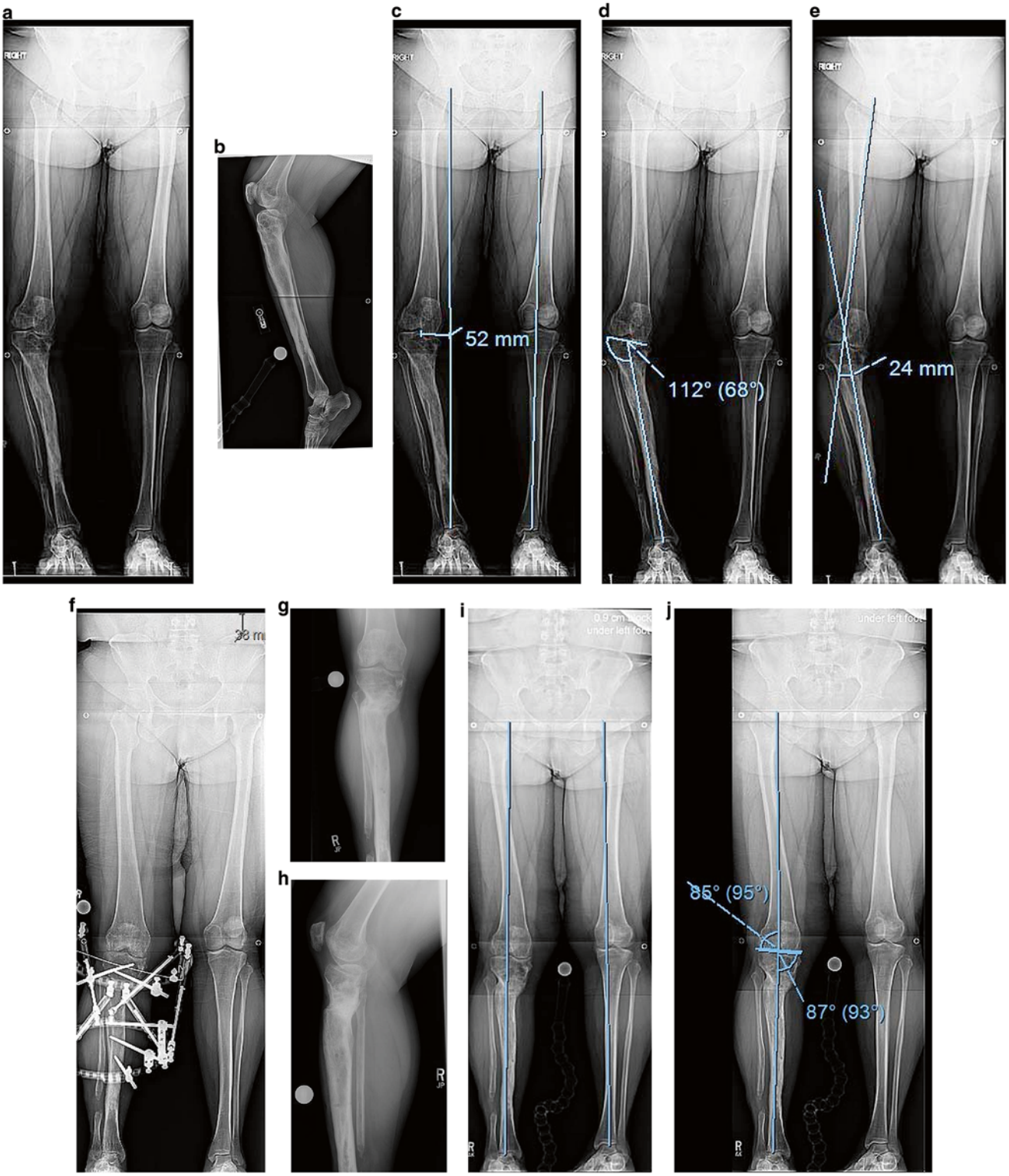


Fig. 32.14 (**a**, **b**) Preoperative radiographs demonstrate varus and flexion deformities of the right proximal tibia. (**c**, **d**) The MAD is 52 mm medial to the midline, and the MPTA is 68°. (**e**) This represents a 17° varus deformity. The CORA is near the knee joint line. The planned osteotomy is below the tibial tubercle. In order to prevent a translational deformity the distal bone fragment must be translated 24 mm laterally in addition to the angular correction of 17°. (**f**) Circular fixation is ideal for executing angular and translational movements of the bone. (**g**, **h**) Final radiographs after frame removal show what looks like an extreme amount of translation at the osteotomy site. (**i**, **j**) Mechanical axis and joint orientation analysis exhibit a slight lateral MAD and an MPTA of 87°; both were ideal for this patient

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Index

A
Acetabular dysplasia, 535
Acetabuloplasties, 538
Achondroplasia, 192–194, 319–321
Achterman and Kalamchi classification, 428–429
ACL. *See* Anterior cruciate ligament (ACL)
Acute complications
  compartment syndrome, 98–99
  iatrogenic fractures, 99
  neurological, 98, 99
  physeal injury, 99
  skin and wound problems, 99
  vascular injury, 99
Acute deformity correction
  acute *vs.* gradual correction, 101
  age considerations, 79, 80
  bone gap creation, 79
  long-standing deformities, 79
  lower limb alignment, 79, 81
  osteotomy
    acetabulum, 88
    acute complications (*see* Acute complications)
    adjuvant fibular, 81, 85
    adjuvant procedures, 88–89
    angular correction and translation, 82–83
    blade plate, 94
    bone void fillers, 95
    closed wedge osteotomy, 81, 82
    Cozen's phenomenon, 84, 85
    deformity memory effect, 84, 85
    diaphysis, 86, 87
    disease-specific indications, 89
    dome osteotomy, 81, 85
    epiphyseal, 85
    external fixation, 94–95
    FAP technique, 95–98
    femoral deformity correction, 91–92
    influence of, 81
    intramedullary devices, 94, 95
    juxta-apophyseal, 85, 87
    Kirschner wires, 93
    knee, 88
    late complications (*see* Late complications)
    LCP, 94
    length discrepancy, 88, 89
    lengthening osteotomy, 83–84
    limb reconstruction systems, 94–95
    mango slice effect, 84
    metaphysis, 85, 86
    multiple bony deformities, 92–93
    oblique osteotomy, 83
    opening wedge osteotomy, 81, 82
    physeal injury, angular deformity secondary, 84
    planning, governing rules, 89–90
    proximal femur, 86–88
    screws, 93
    shortening osteotomy, 83–84, 89
    supracutaneous locked plating, 96, 99
    tension band wiring, 93–94
    tibial deformity correction, 90–91
Adolescent Blount's disease, 510–511
  intermediate types, 514
  treatment considerations
    acute correction with internal fixation, 513–514
    acute/gradual correction with external fixation, 514
    associated deformities, 512–513
    obesity and impact, 511–512
AER. *See* Apical ectodermal ridge (AER)
Aitken classification, 368–369
Amputation management
  cheetah type carbon blades, 199, 200
  congenital femoral deficiency
    Gillespie type A, 201, 202
    Gillespie types B and C, 202–204
    Hamanishi classification, 201, 202
    Paley classification, 201, 203
  congenital fibular deficiency
    Birch classification, 205
    Brown procedure, 205
    Symes amputation, 205–206
  congenital pseudarthrosis, 206, 208, 209
  congenital tibial deficiency
    Jones type 1a classification, 206, 208
    Jones type 2 classification, 206, 208, 209
    Jones type 4 classification, 206, 209
    radiologic description, 206, 207
  fibular hemimelia, 199, 200
  Ilizarov approach, 199
  knee fusion, 199, 200
  marked anterolateral bowing, 199, 201
  patient and family management, 201
  short tibia, 199, 201
  trauma, 210, 211
  treatment concepts, 199–200
  tumor reconstruction, 208, 210–211
  two ray foot, 199, 201
  volume changes, 210
Androgens, 528
Angular correction
  bilateral, 52
  frontal
    anatomic angles, 54
    epiphysiodesis, 53
    follow-up, 54–56

S. Sabharwal (ed.), *Pediatric Lower Limb Deformities*,
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Angular correction (*cont.*)
guided growth, 54, 58–60
Keith needle insertion, 54
longest practical screw, 54
mechanical axis, 53, 54
open/percutaneous techniques, 53
pathologic, 53
physiologic, 52
management, 60, 61
osteotomy, 82–83
patient examination, 52
sagittal/oblique
equinus/posterior plate, 58
knee, 56–58
osteotomy, 58
progressive ankle valgus, 56–58
staples, 51
Ankle and foot
decision making
assessment tools and indications, 160–161
cosmetic improvements, 158
diagnostic matrix, 153, 154
gait cycle (*see* Gait cycle)
intraoperative assessment, foot pusher, 159
level I, II and III deformities, 159
segmental malalignment patterns, 157–158
skeletal surgeries, 158–159
soft tissue surgeries, 158
equinocavovarus deformity, 273, 275
equinoplanovalgus deformity, 272, 274
equinus contracture, 272, 274
myelomeningocele (spina bifida), 290
normal and abducted foot, 273, 274
orthopedic procedure
foot osteotomy, 288–289
gastrocnemius and soleus lengthening, 287–288
SPLATT, 288
SPOTT, 288
Anprolene®, 221
Antegrade femoral lengthening
patient indications, 138–139
planning for, 139
surgical technique, 139–141
Anterior cruciate ligament (ACL), 363
Anterolateral tibial bowing, 9
Anteromedial tibial bowing, 9
Apical ectodermal ridge (AER), 361
Arthrogryposis
ambulation, 309
clinical picture, 300–301
congenital contracture, 297, 298
etiology, 297
genetic aspects, 300
Hall classification system
group 1, 298
group 2, 298–299
group 3, 299–300
intellectual skills, 300
lower limb (*see* Lower limb management)
physical examination, 300
prevalence, 297
prognosis, 301
rehabilitation, 309, 310
timing, 308–309
upper limb, 308
Arthrogryposis multiplex congenita, 4, 6
ASTYMT, 184, 188, 194
Autogenous cancellous bone grafting, 408
Avascular necrosis (AVN), 11, 573–575

B
Beckwith–Wiedemann syndrome, 7–8
Benign bone lesions
dysplasia epiphysealis hemimelica, 343–344
enchondromatosis, 338–340
fibrous dysplasia
anteroposterior, 334, 335
bisphosphonate agents, 336
definition, 333
diagnosis, 333
differential diagnosis, 335
flame-shaped appearance, 334
ground glass appearance, 334
limb length discrepancy, 336
malignant degeneration, 337
Mazabraud's syndrome, 334
McCune Albright syndrome, 334
monostotic and polyostotic fibrous dysplasia, 334
orthopedic management, 335
osteotomy, 336, 337
pathological fractures, 335, 336
RANKL, 336
recurrence, 335
scoliosis, 336
shepherd's crook deformity, 335–336
MHE, 341–343
nonossifying fibroma, 333, 334
osteochondromas, 341
osteofibrous dysplasia, 337–338
Benign tumors, 4, 6
2-β-(3-hydroxypropoxy)- 1α, 25-dihydroxyvitamin D3 (ED-71), 526
Birch classification, 205, 429–431
Bisphosphonates, 256, 260, 264, 491–492, 528
Blocking screws, 113, 138
Blount's disease, 503
adolescent (*see* Adolescent Blount's disease)
classification, 503, 504
differential diagnosis, 503, 505
gradual deformity correction, 108
late-onset/adolescent, 10–11
natural history, 505–506
physeal growth disturbance, 67
physeal injuries, 79, 80
proximal tibia intra-articular deformity, 46
quad plates, 51
treatment
acute deformity correction, 507
atraumatic meniscal tears, 509
bracing, 506
growth modulation, 506–507
hemi-plateau elevation, 509
high tibial osteotomy risks, 507
physeal arrest resection, 507–508
physeal bar resection, 508
Body mass index (BMI), 16
Bone grafting methods, 408
Bone marrow aspirate concentrate (BMAC), 522
Bone marrow cells (BMCs), 522
Bone morphogenetic protein (BMP), 489, 491
Bone transport, 125–127
Bovie cord technique, 96

C
Calcitonin, 528–529
Campanacci's disease. *See* Osteofibrous dysplasia
Center of rotation of angulation (CORA)
  femoral deformity correction, 91–92
  osteotomy, influence of, 81
  sub-capital realignment osteotomy, 87
  tibial deformity correction, 90–91
Cerebral palsy (CP)
  ankle and foot (*see* Ankle and foot)
  causes, 267
  DMD, 293
  GMFCS, 267–268
  hereditary motor sensory neuropathy
    diagnosis, 291
    foot deformity, 292
    hip deformity, 292–293
    HSMN type 2/CMT type 2, 291
    HSMN type 1/CMT type 1A, 291
  knee and lower leg segment (*see* Knee and lower leg segment)
  lower limb assessment (*see* Lower limb assessment)
  musculoskeletal pathology, 268–269
  myelomeningocele (spina bifida), 289
    ankle and foot deformity, 290
    hip dysplasia, 290
    knee flexion deformity and tibial torsion, 290
  optimal timing, 282
  pelvis and hip (*see* Hip)
  poliomyelitis, 290–291
  prevalence, 267
  risk factors, 267
  treatment principles, 282
CFD. *See* Congenital femoral deficiency (CFD)
Charcot–Marie–Tooth disease (CMT), 5–6, 16, 291, 292
Cheetah type carbon blades, 199, 200
Chondroectodermal dysplasia, 329–330
Cidex®, 221
Clinical evaluation
  coronal plane angulation, 27–28
  deformity, definition of, 15
  history, 15–16
  limb length assessment, 23
    AP and lateral alignment radiographs, 23, 24
    CT scanograms, 24
    orthoroentgenography, 25
    single-exposure standing teleroentgenogram, 22, 24
    standardized radiographic technique, 22, 24
    three-view scanogram, 24–25
    X-ray, 24
  pelvic radiograph, 22
  physical examination
    equinus, 18
    foot progression angle, 17
    neurologic, 17
    prone exam (*see* Prone exam)
    short leg gait, 18
    spine and trunk, 17
    supine exam, 18–19
    trendelenburg sign, 18
    upper extremity, 17
    vital signs, 16–17
  quantitative deformity analysis
    anatomic axis, 29, 30
    deformity components, 28
    deformity detection, 29, 31
    joint orientation angles (*see* Joint orientation angles)
    mechanical axis, 28–29
    oblique plane deformities, 32–34
    single-exposure weight-bearing teleroentgenogram, 28
  rickets, chemical findings, 34, 35
  routine radiographic screening, 22
  version diagnostic imaging
    acetabular version, 25–26
    femoral version, 26
    tibial torsion, 26
Closed wedge osteotomy, 81, 82
Clubfoot, 225–226
Computerized tomography (CT)
  acetabular version, 25–26
  CFT, 367
  femoral version, 26
  limb length assessment, 24
  physeal arrest, extent of, 71–72
  tibial torsion, 26
Congenital coxa vara, 9
Congenital dislocation, 9
Congenital disorders
  congenital coxa vara, 9
  congenital dislocation, 9
  congenital femoral deficiency, 8
  congenital fibular deficiency, 8
  hemihypertrophy and hemiatrophy, 7–8
  longitudinal tibial deficiency, 8
  tibia bowing, 9, 10
Congenital femoral deficiency (CFD), 8, 191, 192
  classification systems
    Aitken, 368–369
    Gillespie and Torode, 369–371
    Hamanishi, 371
    Kalamachi, 371–372
    Paley, 372
    Pappas, 369
  complications and treatment
    fractures, 409, 411
    hip subluxation/dislocation, 408–410
    incomplete osteotomy and premature consolidation, 408
    joint stiffness and contracture, 413
    knee subluxation/dislocation, 409
    limb malalignment, 409
    limb reconstructive surgery, 419–420
    nerve injury, 407
    pelvic support osteotomy, 415, 418–419
    poor bone formation, 407–408
    prosthetic reconstructive surgery, 420–422
    superhip 1.5 technique, 413–415
    superhip 2 technique, 414–416
    type 2 treatment, 413, 414
    type 3 treatment, 419
  deformity and pathoanatomy
    ligamentous structures, 363–364
    muscle, 364–365
    osseous deformities, 363
    vascular, 365–366
  embryology, 361–362
  evaluation
    CT, 367
    history, 366
    MRI, 367
    physical exam, 366
    radiographic examination, 366–367
  Gillespie type A, 201, 202
  Gillespie types B and C, 202–204

Congenital femoral deficiency (CFD) (*cont.*)
Hamanishi classification, 201, 202
knee considerations in, 391
knee flexion contracture release, 394–396
Langenskiöld patellar realignment, 394
often-cited incidence, 361
Paley classification, 201, 203
Paley type 1
botulinum toxin injection, 401
femur/tibia, serial lengthenings, 375–376
hemiepiphysiodesis and epiphysiodesis, 376
hip and knee, preparatory surgery, 375
knee instability, 401
limb lengthening, soft tissue releases, 399–401
osteotomy level, 398
superhip procedure, 377
Paley type 2 treatment, 413, 414
pathophysiology and genetics, 362–363
pelvic osteotomies
Paley-Dega osteotomy, 388–389
Paley PATO, 389–390
PAO, 390–391
peroneal nerve decompression, 396–397
preparatory surgery, 391
superhip surgical technique
abductor muscle slide, 385
arthrogram, 386
blade plate insertion, 386
BMP insertion, 387
closure, 387–388
distal femur fixation, 387
external rotation contracture release, 385
fascia lata release, 384–385
femoral shortening, 386–387
flap elevation, 384
guide wire insertion, 386
hip flexion contracture releases, 385
iliac wing osteotomy, 387
incision, 384
muscle repairs and transfers, 387
pelvic osteotomy, 387
periosteum release, 386
positioning, prepping, and draping, 378–384
postoperative course, 388
quadriceps elevation, 386
subtrochanteric osteotomy, 386
superknee surgical technique, 392–394
surgical technique, 401–402
fixator removal and rodding, 406
Ilizarov™ fixator technique, 402–403
modular rail system technique, 404–405
Orthofix™ fixator technique, 403–404
prophylactic rodding, 406–407
rehabilitation and follow-up during lengthening, 405–406
valgus ± flexion deformity, 405
treatment options
limb lengthening, 373–375
rotationplasty, 372–373
syme amputation, 373
type 3
limb reconstructive surgery, 419–420
prosthetic reconstructive surgery, 420–422
treatment, 419
Congenital fibular deficiency, 8
Birch classification, 205
Brown procedure, 205
Symes amputation, 205–206
Congenital posteromedial bowing
deformity
ankle and foot, 496, 497
fibula, 496, 497
tibia, 496
demographic features, 495
diagnosis and management, 501, 502
etiology, 495
evaluation, 498–499
history
residual deformity, 497, 498
shortening, 497
spontaneous remodeling, 497, 498
weak plantar flexion, 498, 499
incidence, 495
treatment
calcaneo-valgus foot deformity, 499–500
coronal deformity, 500
indications, 499
limb length inequality, 500, 501
motivation, 499
muscle imbalance, 500, 501
sagittal deformity, 500
torsional deformity, 500
treatment planning, 501
valgus ankle, 500
Congenital pseudarthrosis of the tibia (CPT)
amputation, 488, 491
anatomy and pathology, 483, 484
associations, 483
bisphosphonates, 491–492
BMP, 489, 491
bone marrow grafting, 492
combined approach, 492
Crawford classification system, 484
electrical stimulation, 492
FVF, 487
Ilizarov circular external fixator, 487–488
imaging, 485
intramedullary nailing, 486–487
Masquelet's procedure, 488, 491
medical management, 489
modified McFarland bypass graft technique, 485–486
non-operative management, 485
operative management, 485
periosteal grafting, 488–490
prognostic factors, 485
Congenital tibial deficiency
Jones type 1a classification, 206, 208
Jones type 2 classification, 206, 208, 209
Jones type 4 classification, 206, 209
radiologic description, 206, 207
Continuous passive motion (CPM) machine, 560
CORA. *See* Center of rotation of angulation (CORA)
Cortical allograft, 408
Coventry and Johnson classification, 428, 429
Cover-up test, 18–19
Coxa breva, 535, 551–552
Morscher osteotomy
deformity correction and fixation, 552–554
positioning and exposure, 552
postoperative care, 554–556
Coxa magna, 535, 556–557
femoral head reduction osteotomy
deformity correction and fixation, 558–560
positioning and exposure, 557–558
postoperative care, 560, 561

Cozen's fracture, 42
Cozen's phenomenon, 52, 84, 85
CP. *See* Cerebral palsy (CP)
CPT. *See* Congenital pseudarthrosis of the tibia (CPT)
Crawford classification system, 484
Custom knee device (CKD), 185–187

D
Decision making
  clinical evaluation, 37–38
  contraindications, 39–41
  equinus contracture
    external fixation, 167–169
    muscles and tendons, lengthening of, 165–167
    nonoperative, 164–165
  knee extension contractures
    nonoperative technique, 176
    quadricepsplasty, 176–178
  knee flexion contracture, 171
    bony correction options, 173–174
    gradual soft tissue correction, 174–175
    nonoperative methods, 172
    soft tissue releases, 173
  osteotomy (*see* Osteotomy)
    in pediatric foot and ankle surgery
    assessment tools and indications, 160–161
    cosmetic improvements, 158
    diagnostic matrix, 153, 154
    gait cycle, ankle and foot function (*see* Gait cycle)
    intraoperative assessment, foot pusher, 159
    level I, II and III deformities, 159
    segmental malalignment patterns, 157–158
    skeletal surgeries, 158–159
    soft tissue surgeries, 158
  physeal bar resection, 42
    advantages, 40
    complete epiphysiodesis, 40
    disadvantages, 40
    hemiepiphysiodesis, 40, 42
    oblique plane deformity, 42, 44
    proximal tibial valgus deformity, 42, 45
    in right distal femur, 40, 43
  problem lists, 38
  soft-tissue modification, 40, 42
  surgeon skills and patient/caretaker preferences, 37
  surgical indications, 38–39
    ankle, 39, 40
    knee, 39
Dega osteotomy, 538
Developmental dysplasia of the hip (DDH), 11, 20
Diastrophic dysplasia, 299
  with hitch hiker thumbs, 323, 325
  laterally dislocated patellae, 323, 325
  operative correction, 324, 326
  rigid clubfeet and hallux varus, 323, 325
  rigid equinovarus foot, 324, 326
  supramalleolar osteotomies, 324, 326
Distraction osteogenesis (DO), 106, 121, 132, 519
  advantages, 357
  anticatabolic therapy
    bisphosphonate, 528
    calcitonin, 528–529
  biological stimulation, 522
    androgens, 528
    bone marrow cells/platelet-rich plasma, 522, 523
    ED-71, 526
    FGF-2, 526
    GH, 527–528
    growth factors, 523
    hyperbaric oxygen therapy, 528
    injection technique, 524–526
    local application-BMP, 522
    optimal timing, 526–527
    osteogenic progenitor stem cell culture technique, 524
    PTH, 527
    scaffolds, 523–524
    stem cells, 523
    TP510, 524, 526
  disadvantages, 357
  indication, 349, 350
  physical stimulation
    LIPUS, 520–521
    mechanical stimulation, 520
    PEMF, 521
  regenerate bone monitoring, 519–520
  type I, 349, 350, 355, 356
  type II, 351
  type III, 352
  type IV, 353
DO. *See* Distraction osteogenesis (DO)
Dome osteotomy, 81, 85
Drill hole technique, 236, 239
Duchenne muscular dystrophy (DMD), 293
Duncan–Ely (DE) sign, 280
Dunn procedure
  deformity correction and fixation, 545–546
  positioning and exposure, 545
  postoperative care, 546–547
Dysplasia epiphysealis hemimelica, 343–344

E
Ellis-van Creveld (EvC) syndrome, 329–330
Endocrine disorders, 3
Equinocavovarus, 157, 158
Equinoplanovalgus, 157
Equinus contracture, 157, 163
  clinical evaluation, 163
  imaging, 164, 165
  management strategy and decision making
    external fixation, 167–169
    muscles and tendons, lengthening of, 165–167
    nonoperative, 164–165
  pathoanatomy, 163–164
Estimated blood volume (EBV), 220
Extensor hallucis longus (EHL), 189

F
Familial hypophosphatemic rickets. *See* Vitamin D-resistant rickets
FAP technique. *See* Fixator-assisted plating (FAP) technique
Femoral deformity correction, 91–92
Femoral head reduction osteotomy
  deformity correction and fixation, 558–560
  positioning and exposure, 557–558
  postoperative care, 560, 561
Femoral metaphyseal-diaphyseal angle (FMDA), 27
Femoral neck osteoplasty, 549–550
  percutaneous osteotomy and external fixation, 550
  postoperative care, 550
Femoral-tibial ratio (FTR), 27
Femur malunion, 608

FH. *See* Fibular hemimelia (FH)
Fibroblast growth factor-2 (FGF-2), 526
Fibrous dysplasia
  benign bone lesions
    anteroposterior, 334, 335
    bisphosphonate agents, 336
    definition, 333
    diagnosis, 333
    differential diagnosis, 335
    flame-shaped appearance, 334
    ground glass appearance, 334
    limb length discrepancy, 336
    malignant degeneration, 337
    Mazabraud's syndrome, 334
    McCune Albright syndrome, 334
    monostotic and polyostotic fibrous dysplasia, 334
    orthopedic management, 335
    osteotomy, 336, 337
    pathological fractures, 335, 336
    RANKL, 336
    recurrence, 335
    scoliosis, 336
    shepherd's crook deformity, 335–336
  genu valgum, 11
Fibular hemimelia (FH), 191–192, 199, 200. *See also* Congenital fibular deficiency
  classification
    Achterman and Kalamchi, 428–429
    Birch, 429–431
    Coventry and Johnson, 428, 429
    Paley, 431–433
    Stanitski, 429, 430
  clinical assessment, 433, 434
  diagnosis, 449
  incidence, 427
  lower extremity anomalies, 427, 428
  outcomes, 451–453
  Paley type 2 lengthening plus ankle realignment, 441, 444
  Paley type 3 lengthening plus super-ankle reconstruction, 441
  Paley type 1 lengthening procedure
    full-length standing, 438, 440, 441
    LON technique, 440–442
    open Vulpius procedure, 438, 439
    PRECICE, 441, 443
    prophylactic anterior compartment fasciotomy, 438, 440
  post-surgical care, 450–451
  prevalence, 427
  radiographic assessment
    arthrogram, 434, 438
    classic "C" sign, 434, 436
    full-length standing AP view, 433, 434
    full-length standing lateral view, 433, 434
    with moderate deformity, 434, 435
    Paley type 3, 434, 437
    with severe deformity, 434, 435
    talo-calcaneal coalition, 434, 436
    talus and calcaneus overlap, 434, 435
    talus and calcaneus side by side, 434, 436
    talus and calcaneus stack, 434, 435
  treatment, 427, 436–438
  type 4, 450–452
  type 3a, 445–446
    frame application, 444
    osteotomy, 444
    positioning, 442
    surgical approach, 442, 444
  type 3b, 447, 448
    osteotomy, 444, 448
    positioning, 444
    surgical approach, 444
  type 3c, 448, 449
    osteotomy, 448
    positioning, 448
    surgical approach, 448
  type 3d, 449–450
  upper extremity manifestations, 427
Fibular malunion, 605–606
Fibular osteotomy, 81, 85
FITBONE device, 133, 145
Fixator-assisted plating (FAP) technique, 95–96
  advantage, 96
  CORA, 96
  disadvantage, 96
  external fixator pins, 96
  femoral shaft, 96, 97
  Ilizarov frame, 96, 98
  indications, 95
  LCP/DCP, 95
  locked plates, 95, 96
  metaphyseal plates, 95
  monoplanar external fixator, 95
  proximal and distal pins, 96
  straight narrow plates, 96
  Taylor spatial frame/multiaxial correction frame, 96
Fixed knee flexion deformity (FKFD), 56–58
Free vascularized fibular (FVF) graft, 487

G
Gabapentin (Neurontin™), 189
Gait cycle
  adduction/abduction, 153
  external rotation, 153
  foot columns, 153, 154
  foot segment, 153, 154
  internal rotation, 153
  pronation, 153
  skeletal alignment, 155
  stance phase, 153
    first/heel rocker, 155
    second/ankle rocker, 155, 156
    terminal stance, 155
    third/forefoot rocker, 155
  supination, 153
  swing phase, 153, 156
Ganz periacetabular osteotomy (PAO), 390–391, 539, 540
Gastrocnemius tightness, 196
Genetic disorders, 4
  NF, 4–5
  osteogenesis imperfecta, 3–4
Genu valgum, 11
Genu varum, 4
  Blount's disease, 10–11
  physiologic genu varum, 9
  tibia vara, 10, 11
Gigli saw osteotomy technique, 236, 239
Gillespie type A classification, 201, 202, 369–371
Gillespie types B and C classification, 202–204, 369–371
Gradual deformity correction
  analysis of
    alignment, 109
    AP and lateral radiographs, 109

bone density, 109
degree and location, 109
delayed healing secondary, 109, 112
hemangiomatosis and tibial bowing, 109, 111
length measurement, 109
planning, 108–109
unossified epiphyses, 110
biology, 106
Blount disease, 108
complications, 108
Ellis-van Creveld disease, 106, 108
history, 105
hypophosphatemic rickets, 106, 107
internal lengthening devices, 113
intramedullary rod, blocking screws, 113, 116
joint subluxation and dislocation, 112, 115
limb length inequality, 106, 109
locking plate, 113
MACT, 112–113
monolateral fixation device, 112, 115
osteotomy, 110, 112, 113
postoperative care, 114, 116
potential complications, 116–119
recurvatum, 114
severe deformity, 106
translation and angular deformity, 106, 107
untreated clubfoot, 114
Graston™, 184
Greater trochanteric arthroplasty (GTA), 592–593
Gross motor function classification system (GMFCS), 267–268
Growth hormone (GH), 527–528

H
Hamanishi classification, 201, 202, 371
Health Inter Network Access to Research Initiative (HINARI), 228
HemoCue device, 219
Hereditary motor sensory neuropathy (HSMN)
diagnosis, 291
foot deformity, 292
hip deformity, 292–293
HSMN type 2/CMT type 2, 291
HSMN type 1/CMT type 1A, 291
Hip
coxa valga, 270, 271
distraction treatment
application, 564
core decompression, 563
patient positioning, 562
postoperative care, 564, 565
soft tissue releases, 563–564
stem cell injection, 563
dysplasia, 290
femoral anteversion, 270, 271
femoral head dislocation, 270, 272
hip at risk progress, 270, 272
orthopedic procedures
adductor releases, 285
femoral derotation, 285
femoral head resection, 286
iliopsoas lengthening, 285
periacetabular pelvic osteotomy, 285–286
varus derotational osteotomy, 285
quantifying hip displacement and acetabular dysplasia, 270, 271
windswept hips, 269, 270
HSMN. *See* Hereditary motor sensory neuropathy (HSMN)
Hydrotherapy, 187–188
Hyperbaric oxygen therapy, 528
Hypertonia, 268–269
Hypophosphatasia, 232
ALP, 244
with bilateral femoral varus deformities, 244–246
with genu valgum deformity, 244, 246–248
with genu varum deformity, 244, 248–250
TNSALP, 244
X-linked rickets, 244, 251–252

I
Iatrogenic deformities
acceptable alignment, 601, 602
femur malunion and growth disturbance, 608
fibular malunion, 605–606
legal implications, 603
malalignment, consequences, 601–603
premature physeal closure, 606–608
tibial lengthening, 612–613
tibia malunion, 603–605
translational deformity, 613–615
vascularized fibula graft nonunion and valgus ankle deformity, 608–612
Iatrogenic injury, 4, 12, 576–577
Idiopathic hemihypertrophy (IH), 7
Ilizarov approach, 199
Ilizarov™ fixator technique, 402–403
Ilizarov hip reconstruction, 415
Ilizarov method, 121, 167
Inflammatory disorders, 4, 6
Informed consent, 219
Infratrochanteric torsion (ITT), 26
Internal lengthening devices, 194–195
Internal limb lengtheners, 132–134
Intramedullary skeletal kinetic distractor (ISKD), 132
Intraoperative nail testing, 144–145
I-STAT, 219

J
Joint line congruency angle (JLCA), 39
Joint orientation angles
in adults, 30
anatomic/mechanical, 30, 32
deformity magnitude, 30
femur and tibia, 30, 31
long segment method, 30, 32
short segment method, 30, 32, 33
Jones classification, 206, 208, 209
Juxta-articular deformity
ankle development, 597
correction of, 596
joint incongruity, 598
knee development, 593
systematic approach, assessment, 598
ultimate goal of, 594

K
Kalamachi classification, 371–372
Knee and lower leg segment
bilateral patella alta, 272, 273
functional contracture, 271

Knee and lower leg segment (*cont.*)
gastrocnemius contracture, 271
orthopedic procedures
anterior hemiepiphysiodesis, 286
distal femoral extension osteotomy, 286
distal tibial (supramalleolar) derotational osteotomies, 287, 288
hamstring lengthening, 286
patellar tendon shortening/advancement, 286, 287
rectus femoris transfer, 287
posterior capsular contracture, 271
torsional malalignment, 272, 273
true coronal plane deformity, 272
weak plantar flexion-knee extension couple, 272
Knee flexion contracture, 172
anterior cortical lines, 171, 172
biomechanics, 170
bony correction options, 173–174
clinical evaluation, 170
decision making, 171
etiology, 169
gradual soft tissue correction, external fixation, 174–175
nonoperative methods, 172
radiographic evaluation, 171
soft tissue releases, 173

L
Larsen syndrome, 299, 308
Late complications
cast-related complications, 100–101
hardware-related complications, 100, 101
joint stiffness, 100
muscle weakness, 101
non-union and delayed union, 100
recurrent deformity, 100
vascular, 100
Lateral distal femur angle (LDFA), 39
Lateral distal tibial angle (LDTA), 56, 58
Legg–Calve–Perthes disease, 190–191, 550–551
Leg length discrepancy (LLD), 195
Lengthening and Then Nailing (LATN), 127, 128
Lengthening and Then Plating (LAP), 125, 126
Lengthening osteotomy, 83–84
Lengthening over Nail (LON) technique, 121–123, 236, 238, 440–442
Limb lengthening
anisomelia
complications, 61, 65
pathologic, 60
physiologic, 60
screw placement, 61, 65
timing, 60–64
complication, 121
hybrid techniques
bone transport, 125–127
LAP, 125, 126
LATN, 127, 128
lengthening and screw fixation, 127, 128
LON, 121–123
PAL, 123–124
physical therapy (*see* Physical therapy)
prolonged treatment, 121
staples, 51
Limb reconstructive surgery, 419–420
Liquid nitrogen (LN) method
advantages, 357
disadvantages, 357
type I, 349, 351
type II, 352
type III, 353
type IV, 354, 355–357
type V, 354–355
type VI, 355
Locked plates (LCP), 94
Longitudinal tibial deficiency, 8
Lower limb assessment
ambulant child (GMFCS levels I-III)
apparent equinus pattern, 276, 277
bony alignment, 280
crouch gait, 277
gait observation, 275
improve independence and physical function, 281
internal rotation gait pattern, 278
jump gait pattern, 276
muscle strength and selective control, 280–281
muscle tone and length, 278–280
optimize gait efficiency, 281
stiff knee pattern, 277
true equinus pattern, 276
unilateral CP (hemiplegia), 278, 279
non-ambulant child (GMFCS levels IV-V), 281
Lower limb management
foot, 301–304
hip, 307–308
knee
with external fixator, 305, 306
knee extension contractures, 305–306
management, 304
quadricepsplasty, 306
supracondylar osteotomy, 304, 305
Low-intensity pulsed ultrasound (LIPUS), 520–521
Lyrica™, 189

M
Maffucci's syndrome, 338
Magnetic resonance imaging (MRI)
CFT, 367
CPT, 485
physeal arrest, extent of, 72
posttraumatic deformities, 571
Malignant tumors, 4, 6
Malunited intra-articular fractures, 579
Malunited shaft femur, 574, 576
Masquelet's procedure, 488, 491
Maximum allowable blood loss (MABL), 219
Mechanical axis deviation (MAD), 28, 29, 109
Medial proximal tibial angle (MPTA), 39
Metabolic disorders, 4
clinical manifestations, 231
definition, 231
deformity analysis, 236, 240
Drill hole technique, 236, 239
drug-induced rickets, 234
Gigli saw osteotomy technique, 236, 239
interference screws, 239, 242
intramedullary static fixation, 236, 239, 242
long sagittal bowing deformity, 236, 238
medulla, 239, 241
monolateral external fixator, 236, 241
nutritional rickets, 233, 234
orthopedic treatment
anterolateral bowing, 234, 235

intramedullary and plate fixation, 234, 236, 237
LON, 236, 238
monolateral/circular external fixator, 236, 238
recurrent deformity, 234, 238
tibial deformity, 234, 235
pathophysiology, 231–232
pediatric metabolic conditions, 231
prematurity, 234
renal osteodystrophy, 3
angular deformity, 243
avascular necrosis, 244
clinical manifestations, 241
features, 242
medical treatment, 243
prevalence of, 241
rachitic changes, 242–243
SCFE, 243
serum vitamin D and calcium levels, 234, 243
rickets, 3, 4
transverse skin incision, 236, 241
tumor-related hypophosphatemic rickets, 241
two parallel Schanz screws, 236, 240, 241
unilateral external fixator, 236, 239
varus bowing deformity, 236, 238
vitamin D-resistant rickets
biochemical abnormality, 233, 234
distal tibial varus deformity, 233, 235
medical treatment, 233–234
severe genu valgum deformity, 233, 234
severe genu varum deformity, 233, 234
Metaphyseal chondrodysplasia, 328–329
Metaphyseal-diaphyseal angle (MDA), 27, 28
Modified McFarland bypass graft technique, 485–486
Modular rail system technique, 404–405
Morquio syndrome, 324–325, 327, 328
Morscher osteotomy
deformity correction and fixation, 552–554
positioning and exposure, 552
postoperative care, 554–556
Motorized intramedullary limb lengthening
complications of, 145–148
reverse planning method, 134–135
Shanz pins, 136, 137
trochanteric entry, 136, 137
venting, canal, 135–136
visual aids, 135, 136
Multi-axial correcting (MACT) system, 112–113
Multiple bony deformities, 92–93
Multiple epiphyseal dysplasia (MED), 323, 324
Multiple hereditary osteochondromatosis/exostoses (MHE), 341–343
Multiple pterygium syndromes, 299
Muscle contractures
analgesic medications, 185
CKD, 185–187
femoral lengthening, 184
myofascial tissues, 184
passive stretching, 185
positioning and splinting strategies, 185, 186
prolonged stretch, 184
removable thigh cuff, 186, 187
soft tissue mobilization, 184
tibial lengthening, 184
tight pin wraps, 185
walking shoe modification, 185, 186
Muscular dystrophy, 293
Musculoskeletal infections, 4, 12
Myelomeningocele (spina bifida), 289
ankle and foot deformity, 290
CP, 289
ankle and foot deformity, 290
hip dysplasia, 290
knee flexion deformity and tibial torsion, 290
hip dysplasia, 290
knee flexion deformity and tibial torsion, 290

N

Neurofibromatoses (NF), 4–5
Neuromuscular disorders, 4
CMT, 4–5
CP (*see* cerebral palsy (CP))
Non-physeal fractures, 4, 12

O

Oblique osteotomy, 83
Ollier's disease, 338
Opening wedge osteotomy, 81, 82
Orthofix™ fixator technique, 403–404
Osseous deformities, 363
Osteofibrous dysplasia, 337–338
Osteogenesis imperfecta (OI), 109
anesthesia, 256
bone deformity(ies), 255, 256
braces
AFOs, 262
HKAFOs, 261
KAFOs, 261–262
molding of, 262
SMOs, 262
coxa vara correction, 261–263
difficult intubations, 256
femoral rodding technique, 258–260
fracture, 256
hemorrhage, 256
long-term immobilization, 260
neurologic complications, 256
open/percutaneous osteotomy, 257–258
patient positioning, 260–262
postoperative rehabilitation, 261
rod types
regular rods, 257
sliding rods, 257, 258
telescopic rods, 257
soft tissue problems, 255–256
surgical correction, 256–257
tibial rodding technique, 259–261
tight muscles stretching, 261
Osteogenic progenitor stem cell culture technique, 524
Osteoporosis, 231
Osteotomy
acute correction
blade and locking plates, 44, 47
cast immobilization, 42
conventional plates, 44–45
distal femoral valgus deformity, 44, 47
implants determination, 42
intramedullary fixation, 42, 44
intramedullary stabilization, 42, 44, 46
Kirschner wire fixation, 42
screw fixation, 44
structural graft, 44, 46

Osteotomy (*cont.*)
acute deformity correction
acetabulum, 88
acute complications (*see* Acute complications)
adjuvant procedures, 88–89
angular correction and translation, 82–83
angular deformity secondary, 84
blade plate, 94
bone void fillers, 95
closed wedge osteotomy, 81, 82
Cozen's phenomenon, 84, 85
deformity memory effect, 84, 85
diaphysis, 86, 87
disease-specific indications, 89
dome osteotomy, 81, 85
epiphyseal, 85
external fixation, 94–95
FAP technique, 95–96
femoral deformity correction, 91–92
influence of, 81
intramedullary devices, 94, 95
juxta-apophyseal, 85, 87
Kirschner wires, 93
knee, 88
late complications (*see* Late complications)
LCP, 94
length discrepancy, 88, 89
lengthening osteotomy, 83–84
limb reconstruction systems, 94–95
mango slice effect, 84
metaphysis, 85, 86
multiple bony deformities, 92–93
oblique osteotomy, 83
opening wedge osteotomy, 81, 82
planning, governing rules, 89–90
proximal femur, 86–88
screws, 93
shortening osteotomy, 83–84, 89
supracutaneous locked plating, 96, 99
tension band wiring, 93–94
tibial deformity correction, 90–91
disadvantage, 51
fibrous dysplasia, 336, 337
gradual deformity correction, 45, 48–49, 110, 112, 113
physeal bar excision, 73

P
Paley classification, 201, 203, 372, 431–433
Paley-Dega osteotomy, 388–389
Pappas classification, 369
Parathyroid hormone (PTH), 527
Pediatric joint contractures
equinus contracture (*see* Equinus contracture)
knee extension contractures
clinical evaluation, 176
etiology, 176
knee stiffness, 176
nonoperative technique, 176
quadricepsplasty, 176–178
radiographs, 176
knee flexion contracture (*see* Knee flexion contracture)
Pelvic osteotomies
Paley-Dega osteotomy, 388–389
Paley PATO, 389–390
PAO, 390–391
Pelvic support osteotomy (PSO), 592–593
Pemberton osteotomy, 538
Percutaneous epiphysiodesis using transphyseal screws (PETS), 52
Periacetabular triple osteotomy (Paley PATO), 389–390
deformity correction and fixation, 541–543
osteotomies, 540–542
positioning and exposure, 540, 541
postoperative care, 544
Peroneal nerve decompression, 396–397
Perthes disease, 190–191
Physeal bar excision
abnormal physis, assessment of
growth remaining, 72
partial *vs.* complete physeal arrest, 70
physeal arrest location, 70–71
timely identification, 70
anterior physeal bar, 75
cannulated ACL reamer, 75
clinical outcome, 76
etiology and prognosis
altered proximal humeral physeal growth, 69
Harris growth arrest lines, 67, 68
infection, 68, 69
neonatal sepsis, 67, 68
neoplasm and tumorlike conditions, 68–69
physeal arrest, 67
repetitive stress, 69
trauma, 67–69
vascular insult, 69
fiber-optic lighted suction, dental mirrors and fluoroscopy, 75
interposition material, 74, 76
local fat harvest, 75
osteotomy role, 73, 75
physeal arrest, extent of, 71–72
radiographic markers, 76
radio-opaque dye, 75
recurvatum deformity, 75
surgical approaches
bony bar, 73
central bars, 73, 74
peripheral bars, 73
type C bars, 73, 74
surgical indications, 72–73
Physeal bar resection, 42
advantages, 40
complete epiphysiodesis, 40
disadvantages, 40
hemiepiphysiodesis, 40, 42
oblique plane deformity, 42, 44
proximal tibial valgus deformity, 42, 45
in right distal femur, 40, 43
Physeal injury, 4, 11–12, 84, 577–578
ankle with malunion, 582
distal femur with angular deformity, 577–578
proximal tibia, 579–580
Physical therapy
equinus contracture, 164
knee extension contractures, 176
knee flexion contracture, 172
limb lengthening and reconstruction surgery
achondroplasia, 192–194
CFD, 191, 192
consolidation phase, 182, 183
fibular hemimelia, 191–192
gait considerations, 195–196
handling techniques, 184

inpatient phase, 181, 182
internal lengthening devices, 194–195
joint stiffness, 186
joint subluxation, 188
Legg–Calve Perthes disease, 190–191
lengthening/correction phase, 181–182
muscle contractures (*see* Muscle contractures)
muscle weakness, 186–188
nerve injury, 189
post frame removal, 182–183
temporary soft tissue dysfunction, 183
weight-bearing considerations, 189–190
Physiologic external rotation contracture of infancy (PERCI), 20
Pixel value ratio (PVR), 520
Plate-assisted lengthening (PAL), 123–124
Platelet-rich plasma (PRP), 522
Poliomyelitis, 290–291
Popliteal pterygium syndrome, 299, 308
Posteromedial tibial bowing, 9, 10
Postinfectious deformities, 587
Choi type II and III hips, 589, 591
Choi type IV deformity, treatment options for, 591–592
GTA *vs.* PSO, 592–593
infantile septic arthritis, 587
knee and ankle joints
incongruity, 598–599
septic arthritis and osteomyelitis, 593–598
radiographic classifications, 587–589
treatment modalities, 589, 590
Posttraumatic chondrolysis, 573–575
Posttraumatic deformities, 569
bony deformities remodeling, 571–573
causation and etiological factors, 569–570
classification, 571
clinical manifestations, 571
distal femoral deformity, 576–577
investigations, 571
lower limb, 573
malunited intra-articular fractures, 579
malunited shaft femur/pseudarthrosis with deformity, 574, 576
physeal injury (*see* Physeal injury)
posttraumatic chondrolysis and avascular necrosis, 573–575
posttraumatic tibia valga, 580–582
soft-tissue loss with skeletal injury, 582–584
Posttraumatic tibia valga, 580–582
PRECICE, 133, 134, 148, 440
Pressure specified sensory device (PSSD), 189
Prone exam
axial foot deformities, 21–22
femoral version and antetorsion, 19–21
tibial torsion, 21
Prosthetic management
children, 210–212
end weight-bearing, 213
first prosthesis, 210
future developments, 213
residual limb length, 211–212
residual limb overgrowth, 212–213
Prosthetic reconstructive surgery, 420–422
Proximal focal femoral deficiency (PFFD), 20
Pulsed electromagnetic field (PEMF), 521

Q
Quadricepsplasty, 176–178

R
Reaming techniques
conventional reaming, 136
rigid reaming, 137–138
Receptor activator of nuclear factor kappa-B ligand (RANKL) inhibitors, 336
Reimer's migration percentage (MP), 270, 271
Renal osteodystrophy, 3
clinical manifestations, 241
features, 242
medical treatment, 243
orthopedic treatment
angular deformity, 243
avascular necrosis, 244
SCFE, 243
prevalence of, 241
rachitic changes, 242–243
serum vitamin D and calcium levels, 234, 243
Residual femoral head subluxation
application, 564
core decompression, 563
patient positioning, 562
postoperative care, 564, 565
soft tissue releases, 563–564
stem cell injection, 563
Residual hip deformities, 4, 11
coxa breva, 551–552
coxa magna, 556–557
developmental dysplasia, 536
femoral head reduction osteotomy
deformity correction and fixation, 558–560
positioning and exposure, 557–558
postoperative care, 560, 561
femoral neck osteoplasty, 549–550
percutaneous osteotomy and external fixation, 550
postoperative care, 550
hip distraction treatment
application, 564
core decompression, 563
patient positioning, 562
postoperative care, 564, 565
soft tissue releases, 563–564
stem cell injection, 563
Legg–Calve–Perthes disease, 550–551
modified Dunn procedure
deformity correction and fixation, 545–546
positioning and exposure, 545
postoperative care, 546–547
surgical hip dislocation, 545
modified Southwick osteotomy, 548, 549
deformity correction and fixation, 548
positioning and exposure, 548
Morscher osteotomy
deformity correction and fixation, 552–554
positioning and exposure, 552
postoperative care, 554–556
periacetabular triple osteotomy
deformity correction and fixation, 541–543
osteotomies, 540–542
positioning and exposure, 540, 541
postoperative care, 544
residual femoral head subluxation, 562
SCFE, 544
treatment
patients after closure, 538–539
patients <6 months of age, 536

Residual hip deformities (*cont.*)
patients >2 years until closure, 537–538
patients >6 months to 2 years, 536–537
Resource-challenged environments
Clubfoot, 225–226
educational resources, 227–228
international collaboration
credentialing rules, 217–218
good working relationship, 217
hosting, 216, 217
medicolegal issues, 218
motivation, 217
program commitment, 216
volunteers, 216–217
operative challenges
Cidex®, 221
cordless power, 223–224
ethylene oxide, 221
indications, 218–219
internal *vs.* external fixation, 222
operating room set up, 221–222
orthopedics, 224–225
perioperative and anesthetic considerations, 219–221
postoperative care, 224
reusing implants and external fixation components, 222–223
sterility, 221
surgical modus operandi, 222
population pyramids, 215, 216
prevalence, 215
severely progressive disparity, 215, 216
SIGN nail, 226–227
Spica Cast, 227
Retrograde femoral lengthening
patient indications, 141–142
planning, 142
surgical technique, 142–143
Reverse planning method, 134–135, 142
Rickets, 3, 4
Ru Li classification, 520

S
Salter-Harris type II, III, and IV injuries, 67, 69
San Diegoplasty osteotomy, 538
Scoliosis, 308
SEDC. *See* Spondyloepiphyseal dysplasia congenital (SEDC)
Shanz pins, 136, 137
Shortening osteotomy, 83–84, 89
SIGN. *See* Surgical Implant Generation Network (SIGN) nail
Single-event multilevel orthopedic surgery (SEMLS), 282
Skeletal dysplasia, 4, 6
achondroplasia, 319–321
characteristic, 313, 314
deformity correction
acute methods, 316–317
with external fixation, 317
with guided growth, 317
diastrophic dysplasia
with hitch hiker thumbs, 323, 325
laterally dislocated patellae, 323, 325
operative correction, 324, 326
rigid clubfeet and hallux varus, 323, 325
rigid equinovarus foot, 324, 326
supramalleolar osteotomies, 324, 326
EvC syndrome, 329–330
genetic mutation and mode of inheritance, 313, 314
limb lengthening, 317–318
MED, 323, 324
metaphyseal chondrodysplasia, 328–329
Morquio syndrome, 324–325, 327, 328
postoperative considerations, 318
SEDC, 319, 321–323
surgical considerations
anesthesia, 315–316
imaging, 315
implant size and design, 316
patient positioning, 316
preoperative evaluation, 315
systemic abnormalities, 313, 314
Skeletal injury, 582–584
Slipped capital femoral epiphysis (SCFE), 3, 20, 22, 243, 544
Soft tissue surgery, 158, 173
Southwick osteotomy, 548, 549
deformity correction and fixation, 548
positioning and exposure, 548
Spica Cast, 227
Split tibialis anterior tendon transfer (SPLATT), 288
Split tibialis posterior tendon transfer (SPOTT), 288
Spondyloepiphyseal dysplasia congenital (SEDC), 319, 321–323
Stanitski classification, 429, 430
Superhip surgical technique
abductor muscle slide, 385
arthrogram, 386
blade plate insertion, 386
BMP insertion, 387
closure, 387–388
distal femur fixation, 387
external rotation contracture release, 385
fascia lata release, 384–385
femoral shortening, 386–387
flap elevation, 384
guide wire insertion, 386
hip flexion contracture releases, 385
iliac wing osteotomy, 387
incision, 384
muscle repairs and transfers, 387
pelvic osteotomy, 387
periosteum release, 386
positioning, prepping, and draping, 378–384
postoperative course, 388
quadriceps elevation, 386
subtrochanteric osteotomy, 386
Superknee surgical technique, 392–394
Supracutaneous locked plating, 96, 99
Supratrochanteric torsion (STT), 26
Surgical Implant Generation Network (SIGN) nail, 226–227

T
Taylor Spatial Frame™, 169, 174, 175
Taylor spatial frame (TSF), 605
Tibia bowing, 9, 10
Tibial deformity correction, 90–91
Tibial hemimelia, 455
genetics, 455–456
horizons and strategy, 461–462
Jones classification, 456, 457
limb reconstruction surgery, 463
Paley classification, 457, 458, 462
centralization, fibula, 477, 479
synostosis and ankle fusion, 473
type 1, 463–464

type 2, 464–467
type 3a, 467
type 4a, 467–469
type 5a, 473, 474
type 3b, 467
type 4b, 469–472
type 5bi, 476
type 5bii, 476–477, 478
type 4c, 472–473
pathoanatomy, 456
treatment history
amputation, 460
Brown procedure, 459–460
distal tibia and ankle stabilization, 460–461
early treatments, 458–459
limb lengthening, 461
options, 461
tibiofibular synostosis, 460
Weber patelloplasty, 461
Weber patellar arthroplasty, 473–476
Tibial hemimelia-polysyndactyly-triphalangeal thumb syndrome, 455
Tibialis posterior tendon lengthening, 288
Tibial lengthening
patient indications, 143
planning, 143
surgical technique, 144
Tibial metaphyseal-diaphyseal angle (TMDA), 27
Tibia malunion, 603–605
Tibia vara, 10, 11
TMA. *See* Transmalleolar axis (TMA)
TMDA. *See* Tibial metaphyseal-diaphyseal angle (TMDA)
Torode classification, 369–371
TP510, 524, 526
TPTPS. *See* Triphalangeal thumb-polysyndactyly syndrome (TPTPS)
Trans-iliac osteotomy, 88
Transmalleolar axis (TMA), 21
Transphyseal ACL reconstruction, 606–608
Trevor's disease. *See* Dysplasia epiphysealis hemimelica
Triphalangeal thumb-polysyndactyly syndrome (TPTPS), 455

V
Valgus ankle deformity, 608–612
Vascularized fibula graft nonunion, 608–612
Vascular malformations, 4, 7
Visual aids, 135
Vitamin D-resistant rickets
biochemical abnormality, 233, 234
distal tibial varus deformity, 233, 235
medical treatment, 233–234
severe genu valgum deformity, 233, 234
severe genu varum deformity, 233, 234

W
Weber patelloplasty, 461
Wingless-type (Wnt) signaling pathway, 362
Wolff's law, 79, 497
World Health Organization has created a Surgical Safety Checklist, 220

X
X-rays, 571

Z
Zone of polarizing activity (ZPA), 361