CHAPTER 28

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Limb-Length Discrepancy

SURGICAL PROCEDURE INDEX

INTRODUCTION

The child with a leg-length discrepancy presents a variety of challenges to the treating orthopaedist who must understand the natural history of the disease process and be able to predict the discrepancy as it will exist at maturity. In order to do so, it is required to be familiar with the methods used to analyze growth and to predict future growth. Implicit to this is an understanding of methodology, risk, and the effect of the wide variety of treatment options on the growing child. Limb-lengthening techniques have evolved rapidly, and the orthopaedic surgeon must consider the ability of these techniques to equalize length discrepancy in light of physical and mental morbidity to the patient. Although enamored with the potential to correct large discrepancies, surgeons and parents need to consider the long-term effects of these treatments on the child.

In addition to understanding the assessment and methodology for treatment of length discrepancies, the surgeon is challenged by the sometimes difficult task of educating the patient and parents. In the case of epiphysiodesis, it can be difficult to explain why a problem in one leg requires an operation on the normal leg; furthermore, patients are not pleased at the thought that it will make them shorter. In the case of leg lengthening, the parents and the patients must understand why the child may wear an external device for many months even after the length is gained. In addition, the family must understand that a fairly high morbidity is associated with this process and the risk of complications can occasionally compromise the final result.

DEFINITIONS

First, we must define what we mean when we say someone has a leg-length discrepancy. Do they have an angular deformity, dislocation, or contracture at the hip, knee, ankle, or foot causing one limb to be apparently shorter or longer? Or is there a true anatomic difference in lengths/size of one of the segments of the lower extremity (femur, tibia, foot)? To avoid confusion, we define *structural* or *true* leg-length discrepancy as a difference in the length of a given anatomic segment (femur, tibia, foot). A leg-length discrepancy that refers to discrepancies that are not true differences in anatomic segment lengths are termed *apparent* or a *postural* discrepancy. As an example, a knee flexion contracture or a dislocated hip may cause an *apparent* shortening of a limb. *Functional* leg-length discrepancy (the sum of the true and apparent leg-length discrepancy) is the most important in treatment decisions (Fig. 28-1). Just as important in the future outcome of leg-length differences is the understanding of age, maturity, and growth potential. *Chronologic age* refers to the actual years of life. *Skeletal (bone) age* is a measure of maturity based on a set of “norms” from which we can make predictions on future growth. From clinical and radiographic assessment, one arrives at a functional length discrepancy and the overall maturity of the patient. Treatment can be considered based on a prediction of the final discrepancy at skeletal maturity and an understanding of the natural history.

EPIDEMIOLOGY, GROWTH, AND ETIOLOGY OF LEG-LENGTH DISCREPANCY

Leg-length differences are common at skeletal maturity. In one study, 77% of 1000 military recruits were found to have differences in leg lengths (1), in another group of military recruits, 36% had differences >0.5 cm (2). In the pediatric population, 2.6% of asymptomatic adolescents were found to

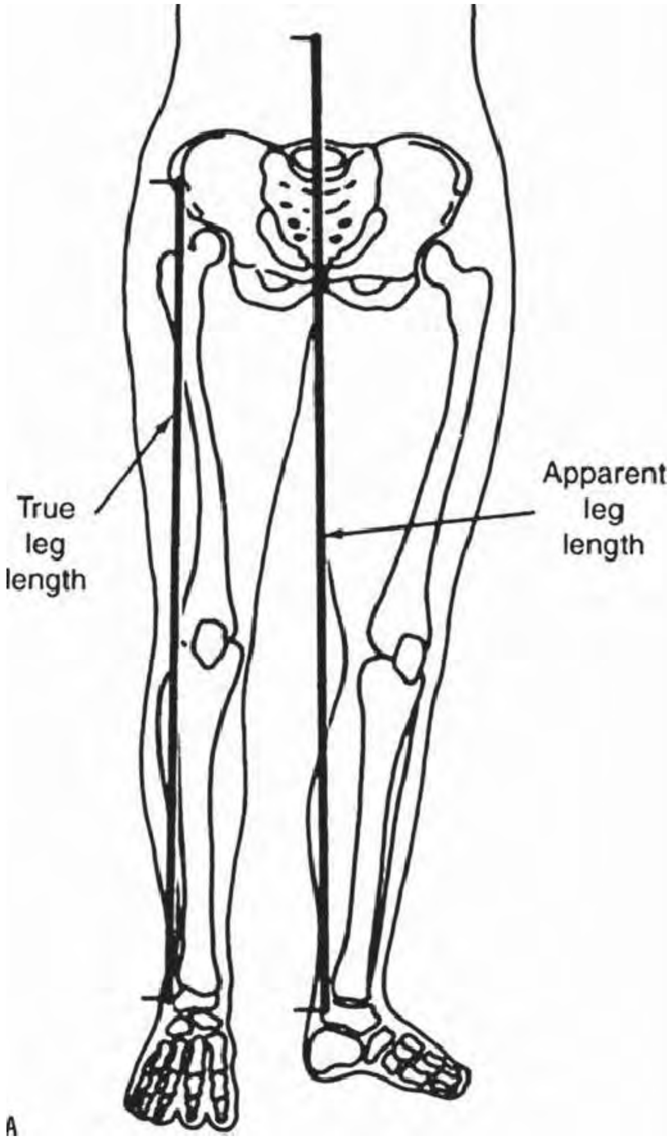


FIGURE 28-1. **A:** *True leg length* is a measure from a point on the pelvis (ASIS) to the ankle and *apparent leg length* is a measure from the umbilicus to the ankle. The latter is affected by hip abduction or adduction as well as knee and hip flexion. **B:** *Functional leg-length* discrepancy takes into account the combined effect of the true leg discrepancy and the hip and knee pathology seen in this child with congenital short femur; even with orthotic shoe modification his lower extremity is still slightly short.

have leg-length differences large enough to cause a clinically noticeable pelvic obliquity during scoliosis screening (3).

In any attempt to predict what will occur in two lower limbs in which unequal growth has been found, one must first understand the normal growth of the lower limb. At birth, the lower limbs are 20% of their final length. The difference in the length of the femur and tibia at birth is 1.2 cm compared with the 10 cm at skeletal maturity (4). The femur and the tibia respectively contribute 54% and 46% of the length of the lower extremity at skeletal maturity; these percentages change throughout growth (5–7). The growth of the lower limb occurs at four growth plates and the foot. The majority of growth of the lower limb occurs about the knee. Anderson found that 71% of femoral growth occurred distally and 57% of the tibia growth occurred proximally (6). This can be shown diagrammatically in terms of percentage of bone growth, limb growth, and actual average growth per year (Fig. 28-2).

In general, overall growth rate and lower extremity growth rate decrease from birth until adolescence when the adolescent growth spurt occurs (6) (Fig. 28-3). More specifically, growth can be considered to happen at different rates throughout development. Dimeglio describes four periods consisting of the antenatal period (exponential growth), birth until 5 years of age (rapid growth), 5 years until puberty (stable growth), and finally puberty (acceleration/deceleration). Using skeletal ages, Dimeglio has demonstrated an early increased growth rate (3.2 cm/yr to 5 cm/yr) at puberty termed the *acceleration phase* followed by a decreased growth rate during the remainder of puberty (4). This *limb* peak height velocity occurred 6 months before that of the peak height velocity of the spine.

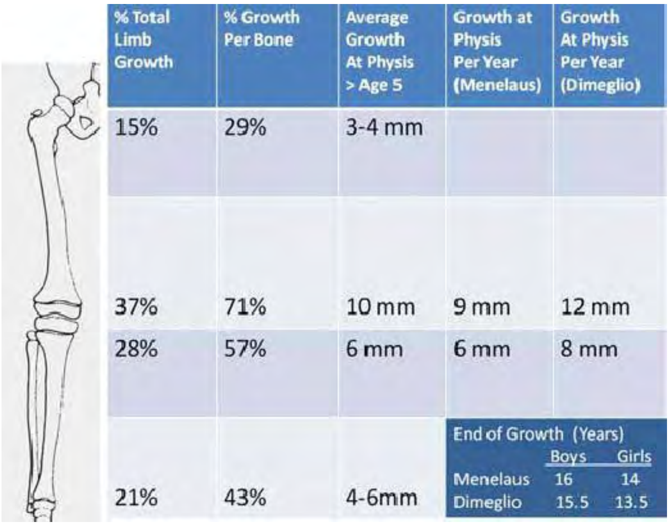


FIGURE 28-2. Cumulative data representing the growth of the lower extremity.

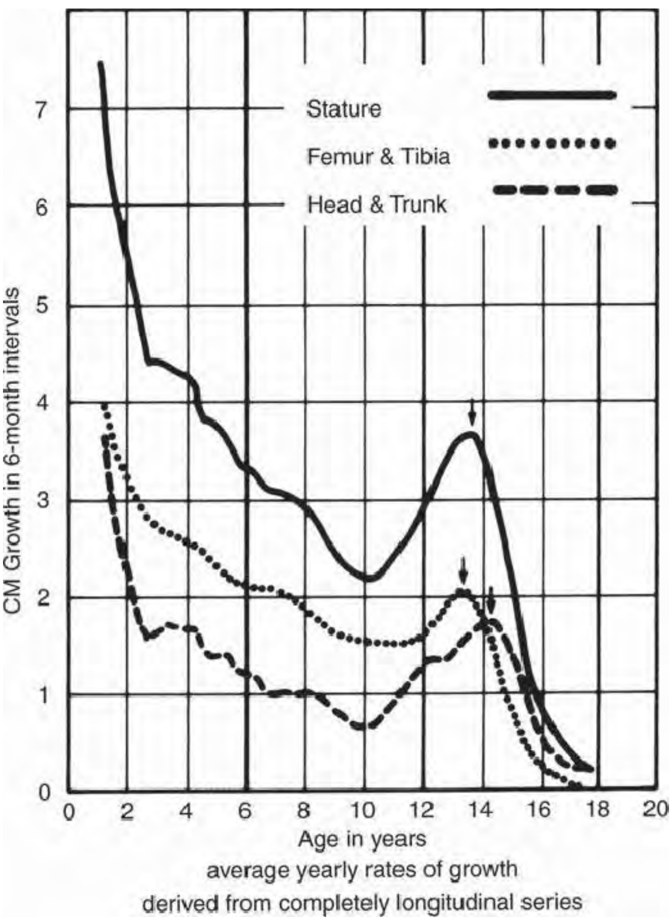


FIGURE 28-3. Green and Anderson growth curve. The examination of growth rate as a function of chronologic age shows a major growth spurt during adolescence. Interestingly, no such spurt appears in the growth curve of Figures 29-6 and 29-7. (From Green W, Anderson M. Skeletal age and the control of bone growth. *Instr Lect Am Acad Orthop Surg* 1960;17:199–217.)

Thus, at the onset of puberty (Tanner stage 2 and skeletal age of 13 for boys and 11 for girls), the average growth remaining in the lower extremities was 10 cm for boys and 9 cm for girls before reaching skeletal maturity. Growth patterns can also be described in the Green and Anderson growth data whereby the lower extremities grow after the age of 5 years an average of 3.5 cm per year (2 cm/yr from the femur and 1.5 cm/yr from the tibia) until puberty.

Etiology Leading to Abnormally Shortened Limb. Several congenital conditions of limb-length discrepancy (congenital short femur, fibular hemimelia, tibia hemimelia) may be apparent at birth and continue to inhibit growth of the short limb as the child ages. Thus in mild cases, the difference in length may only be noticed as the child gets older. In initially apparent cases, a structural deformity may exist within the bones themselves including the physis. For instance, congenitally short femurs are often associated with coxa vara, bowing, hypoplasia of the lateral femoral condyle, and external torsion (8–10). In addition the congenitally short femur may be associated with other clinically noticeable limb abnormalities including fibular hemimelia (11, 12), anterior cruciate ligament (ACL) deficiency (13–17), ball-in-socket ankle, tarsal coalition, and absent metatarsals and toes (18) (Fig. 28-4). Posterior medial bowing of the tibia is another congenital condition that has also been shown to accompany a leg-length discrepancy as well as calcaneal-valgus feet (19–21). Other conditions that lead to limb-length discrepancy include congenital pseudarthrosis of the tibia (22); and patients born with a clubfoot have an increased risk of having a leg-length discrepancy as a result of smaller foot size and also decreased length in the tibia (23) (Fig. 28-5).

An acute change in bone length usually follows trauma and fracture malunion. When a fracture heals in a shortened position, an immediate difference in limb length is observed. Some regrowth is likely to occur in younger patients (24, 25); this is especially seen in the femur (see below). Unfortunately regrowth is unlikely to occur in older children and adolescents with shortening >2.5 cm in the femur, thus leaving a permanent leg-length discrepancy. Similarly, avascular necrosis secondary to Perthes disease, idiopathic, or iatrogenic causes can result in an acute loss of height in addition to damaging the physis of the proximal femur (26, 27).

More commonly, limb-length discrepancies result from gradual changes in leg lengths associated with growth arrests from various causes. Growth plate fractures resulting in partial or total growth arrest may result in growth arrests with or without angular deformities. Similarly neonatal sepsis with multifocal osteomyelitis and septic arthritis with an associated intra-articular growth plate (i.e., proximal femur) lead to physeal destruction and growth arrest (28) (Fig. 28-6). Other causes of premature growth arrest include radiation exposure (29) and neoplastic processes. The latter can be malignant or benign tumors–enchondroma, osteochondroma, and unicameral bone cysts. In these cases, growth arrests result from alterations in local growth or by iatrogenic means in treating them. Mechanical forces can lead to growth retardation and include infantile and adolescent Blount disease, which cause varus deformities of the proximal tibia through medial growth inhibition at the proximal tibia (30). The angular deformities seen in Blount disease and the anterolateral bowing seen in congenital pseudoarthrosis of the tibia, both cause an apparent discrepancy due to the deformity and a true discrepancy in that the affected bones are shorter than their normal counterparts.

Vascular impairment can also result in altered growth plate function and can be seen as a complication of regional disruption of blood supply [e.g., umbilical or femoral catheterization (31)] or local disruption [e.g., hip surgery in the infant (32)]. In these vascular causes, the likelihood of discrepancy can be correlated to the pattern of ischemic damage and increases with increasing involvement (33). Septic and vascular insults to the growth plate tends to lead toward diffuse growth plate dysfunction while trauma or neoplastic process may result in more discrete formation of bony bars between the metaphysis and the epiphysis preventing further growth.

Localized neurological syndromes can cause anatomic limb-length discrepancies. Hemiplegic cerebral palsy (34, 35), polio (36, 37), and other neurologic abnormalities (38) can cause decreased growth of affected limbs (Fig. 28-7). These patients can also have apparent shortening due to concomitant knee and hip contractures and need to be factored in when considering the functional discrepancy. Other nonneurologic causes of apparent shortening include the child with dislocated hip, who may present with apparent shortening without a true leg-length discrepancy.

Etiologies Leading to Abnormally Lengthened Limb. Some of the same factors that can cause shortening of a limb may also cause overgrowth of a limb. In general, it is thought that situations causing increased blood flow to a limb

A B C

FIGURE 28-4. **A:** Patient with congenital short femur. Standing alignment film demonstrates genu valgum and shortened femur. **B:** AP radiograph of the knee demonstrates lateral femoral condyle hypoplasia, lateral subluxation of the patella, and hypoplasia femoral intercondylar notch and of the tibial spine consistent with cruciate deficiency. **C:** AP radiograph of the ankles demonstrates a ball-and-socket ankle.

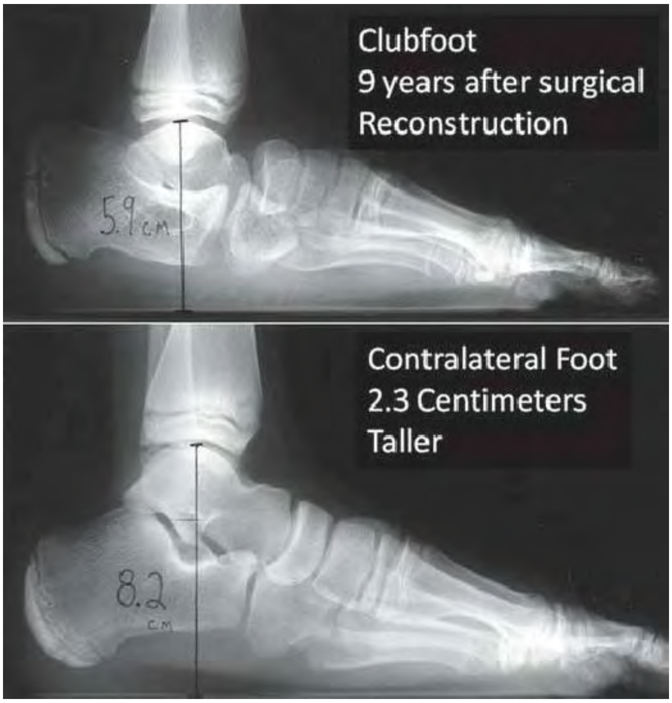


FIGURE 28-5. A 10-year-old boy has a 2.3 cm discrepancy as a result of decreased foot height from the surgical management of idiopathic clubfoot.

FIGURE 28-6. A 2-year-old girl with a history of multiple joint neonatal sepsis and limb-length discrepancy as a result of physeal destruction and joint dislocations.

either transiently or permanently may cause overgrowth of the extremity. Examples of transient increased circulation would be that of posttraumatic or postinfectious overgrowth of the femur and or tibia. Examples of more permanent increases in blood flow would be inflammatory arthritis and arteriovenous malformations (39, 40) that can be seen in Klippel-Trenaunay syndrome (41, 42).

Posttraumatic overgrowth typically occurs following a femur fracture but may also occur after an isolated tibia fracture or a combination of the two (43, 44) (Fig. 28-8). This is more likely to occur in proximal third and middle third femur fractures (45). The most common ages at which this occurs seem to be between 4 and 7 years of age (46, 47), and average overgrowth of the femur has been found to range from 7 to 10 mm (48). While the majority of overgrowth is felt to occur within the first 2 years (44), the clinician should follow these patients periodically until skeletal maturity to ensure similar leg lengths at skeletal maturity. In addition to trauma, increased blood flow from inflammatory conditions can stimulate growth. Examples where growth is stimulated via inflammation near the physis include osteomyelitis or chronic inflammatory arthritis (rheumatoid, psoriatic, or lupus arthrosis etc.) (49–51).

Several syndromes such as Proteus syndrome (53), Macrodactyly, Parke Webber (54), and Klippel-Trenaunay-Weber syndromes (41, 42, 55) have limb-length discrepancy due to generalized overgrowth of one of the limbs and may be accompanied by vascular malformationst. In general, these patients fit within the category of hemihypertrophy, and the discrepancy may be a result of various neurocutaneous disorders that are known to be associated with overgrowth. For instance, neurofibromatosis type 1 (NF-1) (which can also be associated with shortening in congenital pseudarthrosis of the tibia) can present with hemihypertrophy (52). Cutaneous signs of NF-1 include café au lait spots, axillary and inguinal freckling, and cutaneous neurofibroma.

When no apparent systemic disorder is present and the child has apparent idiopathic hemihypertrophy, the treating physician must recognize the possibility of Beckwith-Wiedemann syndrome (56, 57). This disorder is characterized by major criteria (macroglossia, overgrowth abnormalities, and anterior chest wall defects) and minor criteria (ear creases, flame-shaped facial nevi, kidney enlargement, hypoglycemia, and hemihypertrophy) (58). These children are at increased risk for developing intraabdominal tumors such as Wilms tumors, adrenal carcinomas, and hepatoblastomas (59). Because of this risk, regular screening with abdominal ultrasounds is

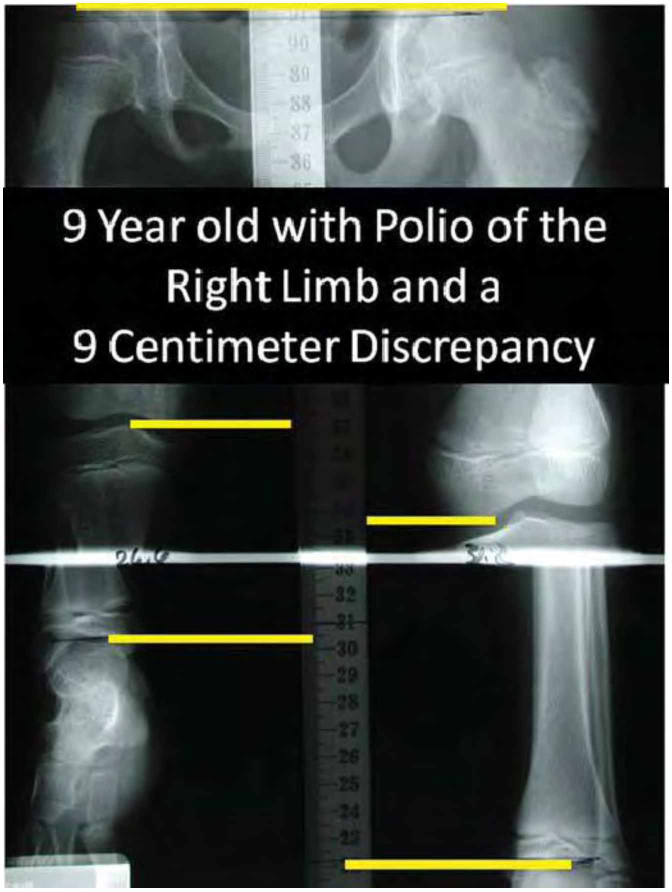


FIGURE 28-7. A 9-year-old girl with infantile polio and a completely flaccid right lower extremity has a 9-cm limb-length discrepancy.

recommended every 6 months for the first 8 years of life. Input from pediatric genetic specialists can be invaluable in evaluating all these hemihypertrophy patients when a diagnosis is not clear.

PATIENT EVALUATION

The parents of a child with limb-length discrepancy may present with concerns referable to a painless limp, pelvic obliquity, and differences in knee height, limb size and shoe sizes. It is important to study the past history of trauma, infection, neurologic conditions, abnormal skin pigmentations, or cutaneous vascular abnormalities. The orthopaedic physical exam is paramount in understanding a limb-length discrepancy. Each patient's height and weight are recorded on the growth chart and parental heights are noted.

The general physical examination is important and will become more focused based on findings and clues toward the etiology are noted. Patients with questionable syndromes should be examined in shorts and with appropriate covering to evaluate for spinal deformity, pelvic obliquity, signs of spinal dysraphism, and hemiatrophy. In the seated position, the clinician should evaluate for any abnormal skin markings such as hemangiomas, axillary freckling, or café au lait spots. Hands can be inspected for differences in size and lower limb hemihypertrophy (Fig. 28-9) can be documented with the "thigh-leg" technique by measuring the foot length, calf, and thigh circumference at set distances above and below the knee in the supine and prone positions (60). In congenital limb-length discrepancies, deficient thigh and gluteal musculature can lead to spuriously larger appearing discrepancies. With the patient in the supine position, the abdomen should be palpated to feel for any intra-abdominal mass such as a Wilms tumor that can be related to Beckwith-Wiedemann syndrome. Examination of gait may suggest underlying neurologic conditions; spasticity (noted by a crouched gait, decreased knee extension, equinus, and raising of one arm) or weakness (Trendelenburg gait) may be uncovered. Functional compensation can be detected in gait and include hip and knee flexion and circumduction of the long limb. On the short side, the patient may exhibit an equinus contracture at the ankle and vaulting over the long limb. Vaulting is typically recognized as the patient's center of gravity is thrust up and down. Upper extremity range of motion and muscle tone should be assessed in order to detect an underlying neurologic disorder.

In the standing position, the level of the popliteal crease, iliac crest, and shoulders is noted and overall coronal and sagittal alignment of the spine and lower extremities is assessed. The clinician must be sure the patient has his or her knees in extension and feet flat on the ground. An Adams forward bend test should be performed to look for occult scoliosis. A fixed pelvic obliquity due to a spine deformity may be the underlying cause of an apparent leg-length discrepancy. Placing their hands on the patient's iliac crest will allow the examiner to observe any difference in height of the posterior superior iliac spines and then by using graduated blocks one can equalize the pelvis estimating the discrepancy. (Fig. 28-10). This indirect method of measuring leg lengths has been shown to be accurate within ~1.5 cm of actual lengths (61–64). Pelvic asymmetry may occur in up to 5% of the normal population and give rise to some inaccuracy (65). In addition, this method may be more unreliable in the overweight child.

Patients with nonsyndrome causes will undergo exam of the affected limb in comparison to the contralateral limb. Examination of the foot for any signs of deformity should be mandatory; the deformity should be evaluated and may be a sign of fibular hemimelia. Hip, knee, and ankle instability should be documented and may suggest congenital short femur (hip and knee instability) which is crucial to consider when planning limb lengthening. Care is needed to document joint motion, muscle girth, and neurovascular function. While supine, a detailed range of motion of the hip, knee, and ankle should be assessed for any contractures. Hip adduction contractures produce a functionally shortened limb while abduction contractures produce a functionally long limb; these can thus produce an infrapelvic obliquity (66–68).

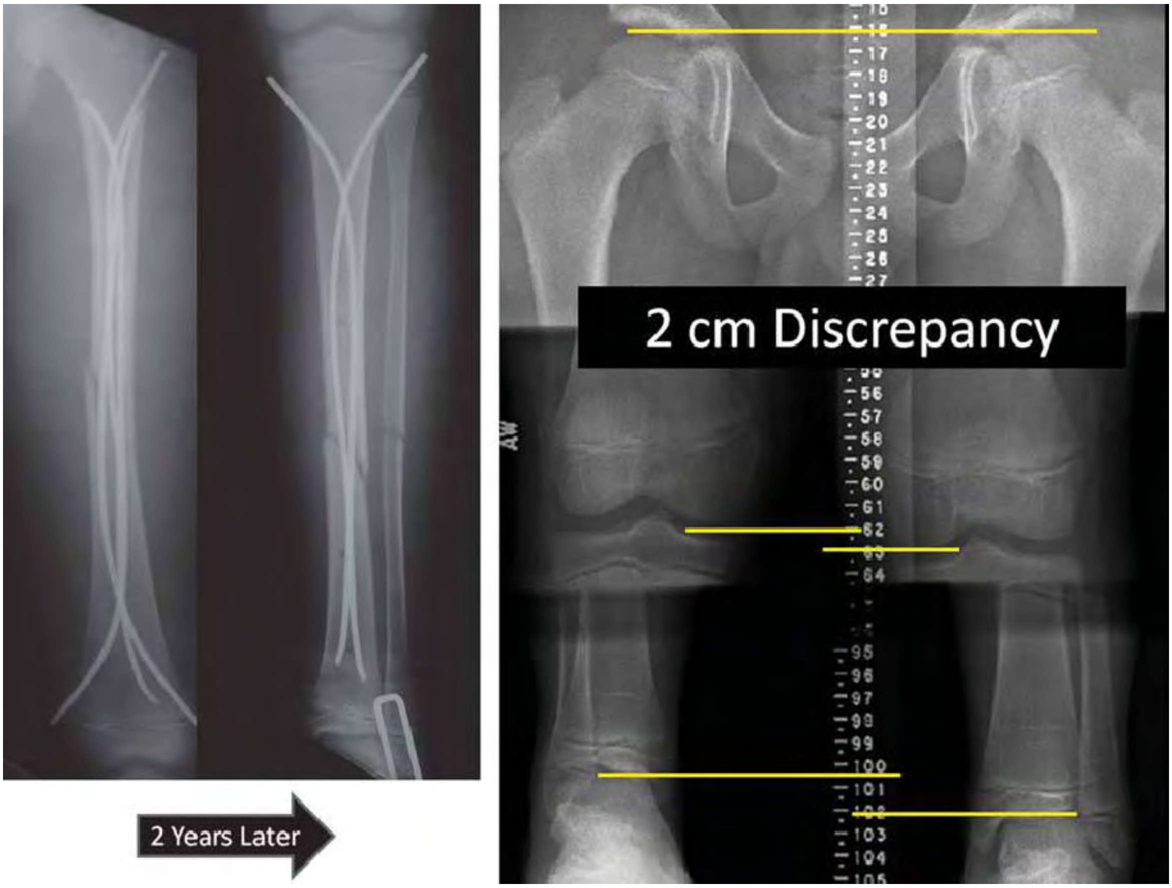


FIGURE 28-8. Two centimeters of limb overgrowth is present 2 years after surgical management of an ipsilateral femoral shaft and tibia fracture.

In the supine position, the true and apparent leg lengths can be measured using a tape measure. The apparent leg length is measured from umbilicus to medial malleolus. The true leg length is measured from the anterior superior iliac spine to the medial or lateral malleolus (60, 61, 64, 69–72) (Fig. 28-1). The knees and hips can be flexed 90 degrees and any difference in knee heights recorded (Galeazzi sign) will suggest pathology in the femoral segment (hip joint to knee joint). Similarly, with the patient prone, differences in the height of the heel pad can be recorded demonstrating likely pathology between the distal femur and the foot. As tape measurements and block measurements are prone to error, the clinician should use them together to screen for a true leg-length discrepancy and should confirm their findings with radiography. While the imaging modalities have been shown to be more accurate than clinical screening methods, clinically measured leg-length differences correlated better with patient's perceived inequality than did imaging (73).

In addition to an orthopaedic examination, an accurate assessment of maturity should be made. While seldom done in the orthopaedic clinic, a full Tanner staging can be useful to determine an adolescent's maturity. For review, the Tanner stage relies on the development of secondary sex characteristics to determine maturity.

RADIOGRAPHIC ASSESSMENT

Several different radiographic methods are available to quantitate a leg-length discrepancy, and each has its benefits and weaknesses. Factors that affect which study to order include the estimated discrepancy, the location of the discrepancy, and the age of the patient. For instance, in children under 2 years of age who are initially presenting with a leg discrepancy, the authors prefer to obtain a standing radiograph of the entire lower extremity. Although this radiograph does not allow the highest accuracy in measuring the discrepancy, it is a simple method that requires only one exposure of a potentially fidgety child. In addition, it provides an evaluation of alignment and lets the clinician see all the bones in the legs.

After the patient grows to the age whereby a more sophisticated study can be reliably performed, this same technique should be employed throughout the child's course so that differences are consistent. Similarly, the clinician should personally measure all his or her own films using consistent landmarks (top of femoral head, medial femoral condyle, center of the ankle joint) to ensure as much consistency as possible. Radiographic measurements should correlate with the clinical exam findings and when discordance is present, the cause of an apparent leg-length discrepancy should be sought. A classic

FIGURE 28-9. Clinical picture is presented of a 10-month-old infant with apparent hemihypertrophy. Although he does not have other signs of Beckwith-Wiedemann syndrome, routine abdominal ultrasounds are needed to detect abdominal tumors.

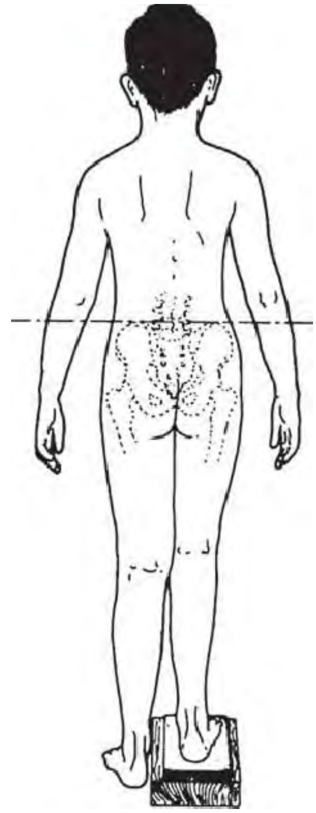


FIGURE 28-10. Placing blocks beneath the heel of the short leg allows assessment of the combined effect of all factors that produce functional leg-length discrepancy.

example is the patient with a concurrent hip or knee flexion contracture; thus the length of the bones may be best assessed with separate lateral films of the tibia and femur or advanced measurement techniques such as CT scanogram (Fig. 28-11). Available in many centers, *Computer Scanogram* has several benefits; it is quick, associated with decreased radiation (74–76) and easily accommodates contractures (77) and external fixators. Even though many clinicians do not have a CT scanner immediately at their disposal in the clinical setting and other reliable methods exist, CT scanograms have been shown in some of these cases to be more accurate than standard radiographic measurements below.

Teleoroentgenography consists of standing alignment film (35 cm × 90 cm) taken with a single exposure at a distance of 2 m centered on the knee joint; a radiopaque ruler will allow one to measure the differences in length (Fig. 28-12). It is perhaps the best radiographic assessment in the young where scanograms are difficult to obtain and where full visualization of the skeleton can assist in diagnosis. Its benefit is that it is a quick single x-ray and it can give information about limb alignment and limb length. Unfortunately, one must account for magnification as parallax becomes an issue. In the past, the storage of these large films was more cumbersome, but with widespread use of digital radiography, this is less of an issue and some programs can assess length without the radiographic ruler. For individuals who cannot stand well, there is no significant difference in length measurements when this technique is performed in the supine position (78).

Recently, several authors have recommended these radiographs (especially the computer variants of this technique) to be the primary imaging tool to be used to diagnose and monitor leg-length discrepancies as the technique is relatively inexpensive, involves less radiation, is widely available, shows angular deformities, and can show asymmetries in the foot and pelvis (79–82) Recently, Sabharwal compared the use of the scanogram and digital teleoroentgenogram (with a 5% magnification factor). The mean differences between the two techniques were 0.5 cm and they had a correlation coefficient of 0.96. The authors endorsed the use of the single x-ray as this gave important information as to mechanical axis deviations not seen on a scanogram, and this limited the patient to a single radiation exposure. This is in agreement with the findings of others (78).

Orthoroentgenography consists of three separate exposures at the hip, knee, and ankle all placed on the same long-standing film with ruler centered over each joint (Fig. 28-13). This technique eliminates the magnification error as the x-ray source moves to the center over each joint (83). It can be adapted to show angular deformities, but throughout all exposures the patient must remain still and thus makes it a challenge for smaller children.

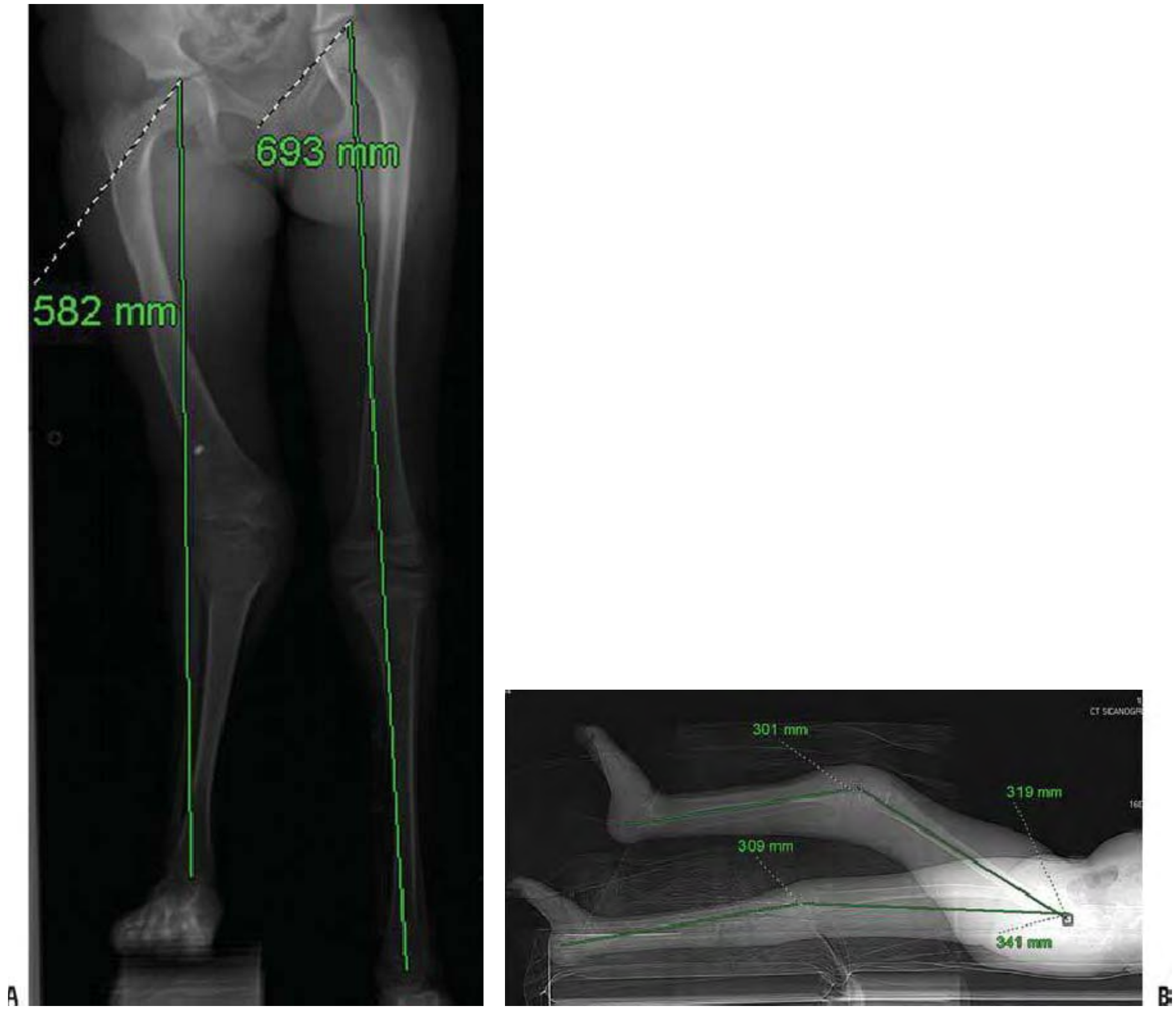


FIGURE 28-11. **A:** Standing alignment film of an 11-year-old girl with arthrogryposis demonstrates a functional discrepancy in length approximating 9 cm. Unfortunately, her severe knee flexion contracture precludes accurate assessment of her anatomic bone lengths. **B:** Lateral CT scanogram demonstrates that her femur is not significantly shortened (2 cm) and her tibia are essentially equal.

*Slit scanography or scanogram*s are performed so that the x-ray source and the film are both adjusted to reduce parallax error and all three joints are placed on one smaller film. Similar to the orthoroentgenography, the patient must remain still as the film and x-ray source move (84) (Fig. 28-14). Both indirect and direct slit scanograms have been described. Indirect scanograms utilize a midline ruler between the extremities from which measurements are made; a direct scanogram places the ruler along the mechanical axis of the limb. The direct method has been shown to be more reproducible than the indirect (63). With today's computer imaging software, one can rapidly determine the differences in the absolute length between certain bones, thus improving clinical efficiency by avoiding marking, measuring, and calculating differences.

Other imaging technologies have been described to decrease radiation exposure; however, most of these are not readily available to the practicing orthopaedic surgeon. Microdose radiography involves an x-ray source and computer detection system which yields data in about 20 seconds. The advantage of this system is its accuracy and the significantly less radiation exposure than in conventional radiographs. However, the widespread availability of the technology is limited (74). As another method, ultrasound has been used to measure leg-length discrepancies and has found to be slightly less accurate than x-rays (72) but eliminates all radiation. Finally, MRI has been used to assess lengths and despite the elimination of radiation, the cost, the length of procedure, and less accuracy have prevented its widespread use (85).

MATURITY ASSESSMENT

Assessment of maturity is another important factor in treating a patient with a leg-length discrepancy. Unfortunately, chronologic age is only a rough estimate of maturity, and as orthopaedists, we are interested in skeletal or physiologic maturity as it relates to growth. While a detailed developmental history (menarche, shaving, and secondary sexual characteristics) can give a general idea of maturity, several imaging techniques

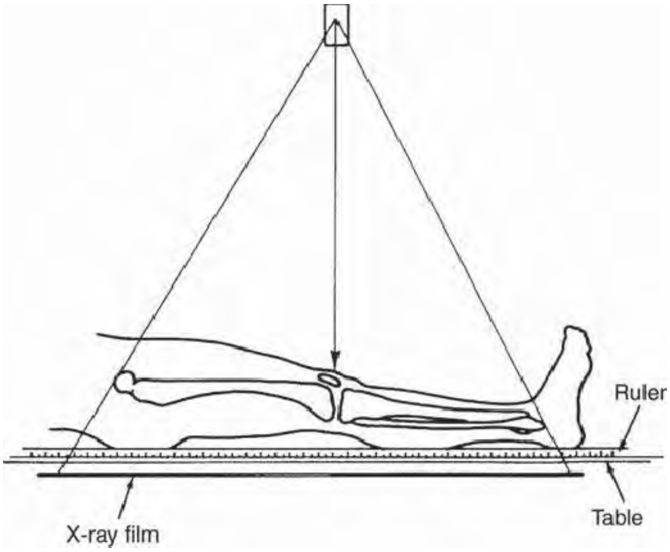


FIGURE 28-12. The teleoroentgenogram reveals angular deformities but is subject to errors of magnification. It is best for children who cannot remain still or as an initial test to locate deformity as well as length discrepancy.

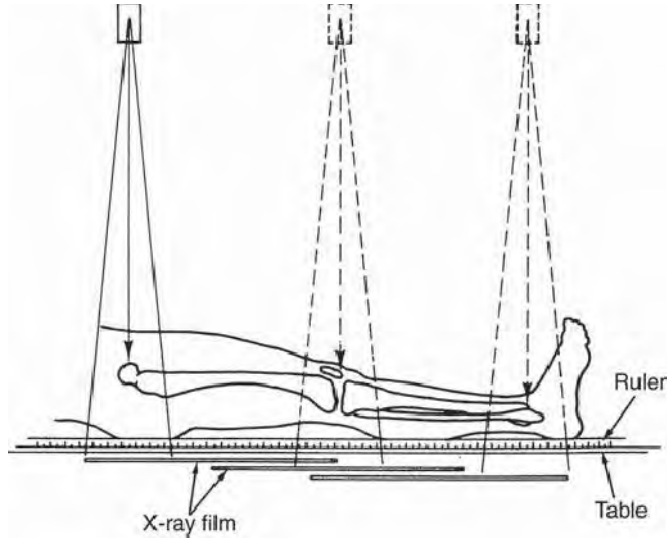


FIGURE 28-14. The scanogram technique avoids magnification error in the same manner as the orthoroentgenogram does and has the advantage of being on a smaller image. It is also useful for children who can be counted on to remain still.

are often used to more accurately evaluate a child's skeletal maturity. Up to 50% of children were found to have a skeletal age that differed from their chronologic age by >6 months (86).

Methods for measuring maturity based on the appearance of various ossification centers in various populations of "normal" pediatric patients have been published. Findings at the foot (Hoerr), knee (Pyle and Hoerr), pelvis (Acheson), and upper extremity have all been used. The most widely used method of assessing skeletal maturity is using the bone age, as described by Greulich and Pyle (87). These authors studied the ossification centers of the left hand in a number of

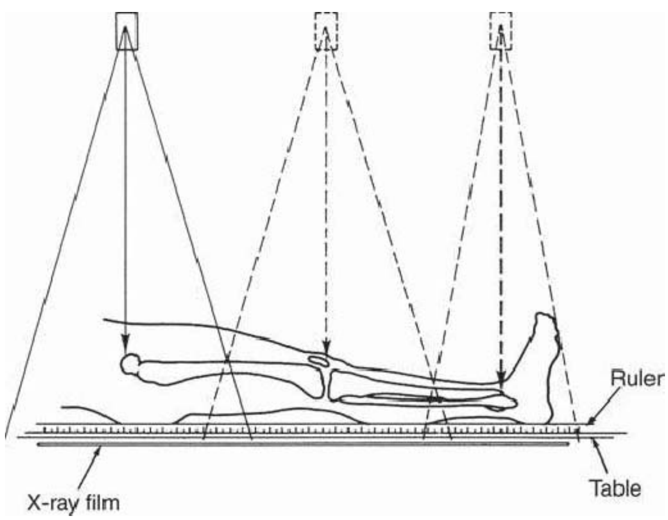


FIGURE 28-13. The orthoradiograph technique exposes each joint individually, thereby ensuring that the x-ray beam through each joint is perpendicular to the x-ray film and thereby avoiding errors of magnification.

healthy normal children at different ages. As the ossification centers appear and coalesce in a reasonably predictable fashion, they were able to develop a norm for each age. When a child has a bone age x-ray, the clinician or radiologist can compare this child's x-ray with those in the atlas and develop a bone age with a given standard deviation. While helpful, it does have significant deficiencies including gaps as far as 14 months, thus giving a rather large standard error. While the average or mean radiograph would likely be chosen for a given skeletal age (placing half the children as more mature and half the children as less mature), the developers chose some of the standards based on what they subjectively felt were most representative of that age. Furthermore, while a general order of ossification occurs in the bones of the hand and wrist, variations do occur; therefore arbitrary choices in age must be made.

Despite the above limitations, the most common technique used in determining skeletal maturity is the Greulich and Pyle technique that is based on standard radiographs, now over 50 years old, from affluent white children. The applicability of these "standards" to other ethnicities and to modern-day children has been questioned. Recent studies have shown children to achieve an older bone age for a given chronological age today versus 25 years ago (88). Similarly, bone ages of Asian and Hispanic children tend to be overestimated using the Greulich and Pyle technique. They appear to mature sooner than the Caucasian and African American children (89). Recent advances have used computer-based systems to score the x-rays and determine the skeletal age (90–92). One of these systems has involved the recent acquisition of an atlas of 1,400 children from four different ethnicities to attempt to limit any ethnic bias from the standards (89). Preliminary results appear promising

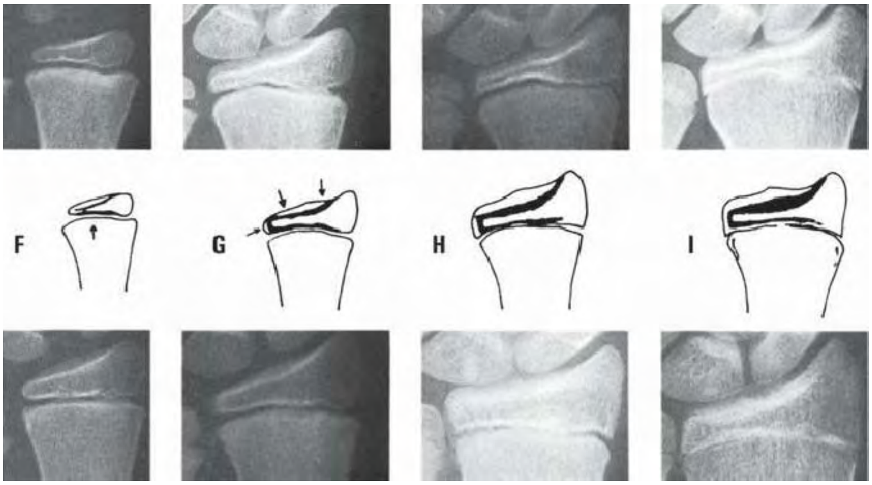


FIGURE 28-15. The Tanner-Whitehouse atlas provides standards for 20 different landmarks of the hand and wrist and allows determination of the skeletal age. (From Tanner J, Whitehouse R, Marshall W, et al. *Assessment of skeletal maturity and prediction of adult height (TW2 method)*, London, UK: Academic Press, 1975.)

using these techniques as they are able to eliminate reader variability and bias.

Similar to the Greulich and Pyle method, the Tanner-Whitehouse method (93) relies on the ossification centers of the hand and wrist (RUS–radius ulna and small bones) to determine maturity, but relies on computerized mathematical procedures to evaluate the successive stages of 20 bony landmarks of the bone and wrist. The radiograph is compared to standard radiographs, and each of the 20 landmarks is designated a letter score. A numeric score is derived from this letter score after gender is taken into account. The result is a skeletal or bone age with a much smaller standard error. This method relies on the cuboid and long bones of the hand. If a discrepancy in age between these two types of bones exists, it may be reasonable to assume those measures of the long bones to be more directly relevant to the maturity and growth of the long bones of the lower extremity, but this is only speculative (Fig. 28-15).

Dimeglio (94) has shown accuracy in using a modification of the Sauvegrain method of bone age especially in the prepubertal and pubertal children. This is beneficial as it is in this age group (9 to 15 years old) that the Greulich and Pyle atlas lacks norms at regular intervals. This French method looks at the four different ossification centers about the elbow and develops a maximum 27-point score for males and females. The score is then plotted on a graph and the appropriate bone age (at 6-month intervals) can be determined. This method has been shown to be very reproducible and is ideal for children in this age group. Before the age of 9 in females and 11 in males, this method cannot be used.

OTHER DIAGNOSTIC STUDIES

When evaluating the limb-length discrepancy and based on the likely etiology, other imaging studies may be required. As previously stated, patients with apparent idiopathic hemihypertrophy require interval screening (every 6 months of age up until age 8 years) via abdominal ultrasound to rule out a Wilms tumor or other abdominal masses that would be consistent with Beckwith-Wiedemann syndrome. When evaluating a growth plate injury from a traumatic, infectious, or neoplastic cause, a CT scan or MRI of the affected growth plate may be essential to evaluate for any potential for bar excision (95, 96). Similarly, an MRI may be advantageous in evaluating the presence of anatomic structures (ACL) in a patient with a congenital short femur or in evaluating the location of an arteriovenous malformation prior to surgical intervention.

GROWTH PATTERNS

In order to predict the ultimate discrepancy, one needs to consider the patient's growth potential and whether future growth could be retarded or accelerated. For instance, one would expect a 3-cm discrepancy from a femur shaft malunion in a 12-year-old boy to remain stable as the growth plate is unaffected and the patient is unlikely to recoup the distance with regrowth. In contrast, a Salter Harris type I distal femur fracture with complete growth arrest will continue to worsen until skeletal maturity. In congenital limb differences, the ratio of the short limb to the long limb has been shown to be constant (97). Clinically, these limbs will stay proportionately the same, but the absolute difference in length will increase (98). Some generalities can be made about the existing congenital deformity according to the patient age. For instance at birth, the ultimate discrepancy will be 5 times the difference at birth, 3 times the difference at 1 year of age, and 1.5 times the difference at 7 years of age (4). Some developmental discrepancies (polio, Ollier disease, growth arrest) have been shown to have a rate of inhibition that is also fixed.

Shapiro (99) described five patterns of growth inhibition (Fig. 28-16). Constant inhibition was only one pattern of growth inhibition recognized. While five patterns were recognized, a given diagnosis may exhibit more than one pattern of inhibition. Unfortunately, these patterns of inhibition are difficult to use clinically. Even so, one may wish to keep these patterns in mind

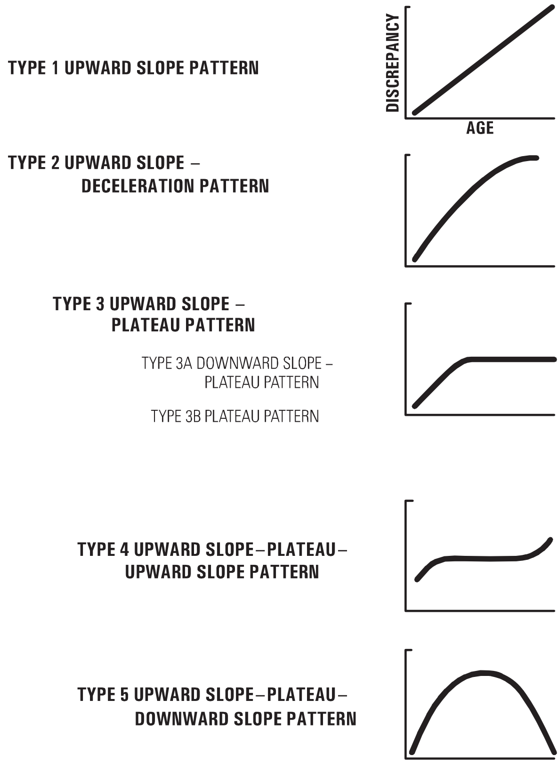


FIGURE 28-16. Different patterns of growth retardation are demonstrated here. (From Shapiro F. Developmental patterns in lower extremity length discrepancies. *J Bone Joint Surg Am* 1982;64(5):639–651.)

when planning an equalization procedure as this may cause a deviation from the prediction models described below.

PREDICTIONS OF GROWTH AND ULTIMATE DISCREPANCY

The most important question that drives prognosis and treatment is "what will be the final limb-length discrepancy, and how and when are we going to equalize them?" Multiple methods have been developed to try to predict final discrepancies and thus guide treatment to allow equal leg lengths at skeletal maturity. From the previous sections, it becomes obvious that accurate knowledge of both the discrepancy and maturity is essential in answering these questions. Essentially, most methods to predict final discrepancies and time treatment rely heavily on the ground-breaking work of Green and Anderson, and each attempts to use this data in different ways (mathematically and graphically). Several generalizations can be made regardless of the technique used. Multiple data points (of discrepancy and skeletal age) over time help make more accurate predictions, and greater accuracy exists in predicting final limb difference as the child gets older (children >10 years of age).

Green and Anderson Growth Remaining Model. Green and Anderson used longitudinal data on the growth of the lower extremities to predict the amount of growth remaining in the distal femur and proximal knee. Initially, semilongitudinal data on over 800 individuals were used to construct a growth remaining chart in 1947 (7). In 1963, a pure longitudinal cohort consisting of 50 males and 50 females was followed yearly to refine the growth remaining chart and a nomogram of femur and tibia lengths. The later prospective cohort provided more accurate standard deviations over time and used skeletal age using Greulich and Pyle bone age. By plotting the skeletal age of the child, the amount of growth remaining in each bone could be read from the chart and allowed the prediction of the outcome of epiphysiodesis within two standard deviations. The growth remaining graph did not take into account the differences in the size of stature or inhibition which might lead to differences from those predicted.

Green and Anderson constructed another graph looking at the yearly growth of the tibia and femur in 67 males and 67 females from ages 1 to 18 years of age. This again was a completely longitudinal study based on *chronologic age* and average tibia and femur lengths (5). From this data, a nomogram was again constructed in which a patient's leg lengths could be plotted. From multiple measurements, percentile growth of the individual could be seen on the "normal leg" and inhibition of the short leg could be observed. They felt this helped with the growth remaining curve to determine whether a patient might be on the high or low side of the average growth remaining (i.e., a patient whose tibia and femur were two standard deviations from the norm might have a greater growth remaining and thus a greater inhibition after epiphysiodesis than someone two standard deviations below the mean). They stressed the importance of obtaining several measurements to get a sense on the pattern of the growth rate abnormality, especially in the 2 to 3 years before a planned procedure as this rate may change over time. When using the chronologic graphs, they stressed the importance of taking maturity into account. Multiple measurements and the patient's overall growth chart percentiles can be useful in identifying discrepancies in maturity between skeletal and chronologic ages on these graphs. A patient consistently in the 80% percentile for height for age should similarly fall in near the second standard deviation for tibia and femoral length; if they suddenly drop in percentile this may mean they are either delayed in their maturation or were previously precocious. Knowledge of this will be useful in altering final predictions and might lead a clinician to use skeletal age rather than chronologic age. This method has been used for years and has been found to be accurate (100) at predicting the timing of epiphysiodesis (Figs. 28-17 to 28-20).

Moseley Straight-Line Graph Method. In an effort to simplify and improve the accuracy of the Green and Anderson method, Moseley developed a nomogram for skeletal age derived from Green and Anderson data to correct for percentile growth (variations in maturity and relative size) (101, 102). On this graph, the growth of each limb is

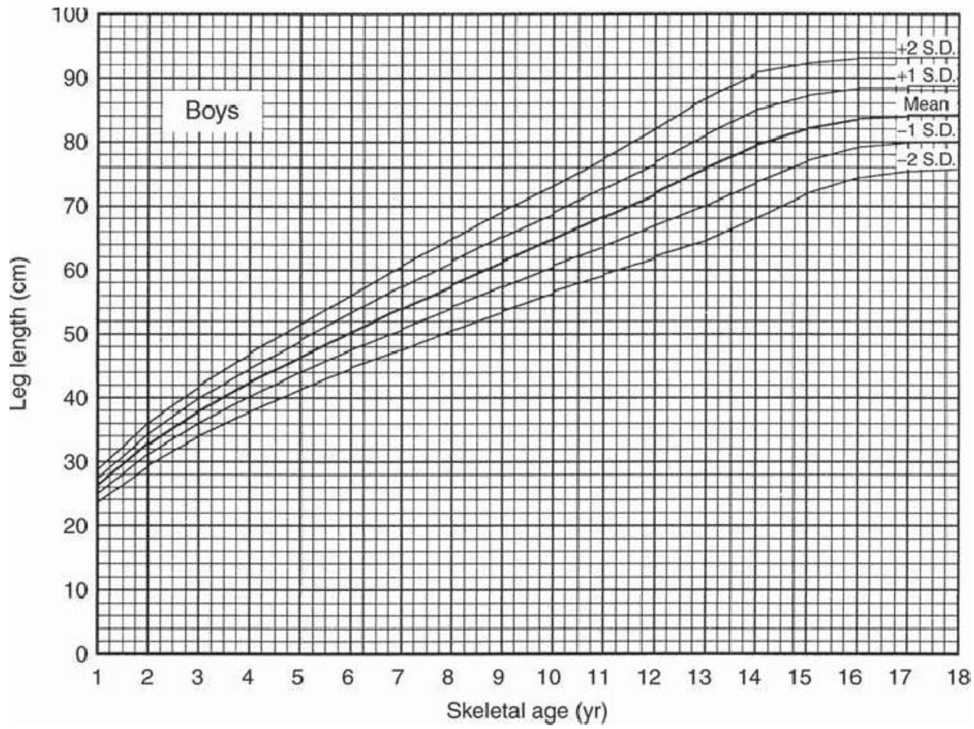


FIGURE 28-17. Graph showing total leg length versus skeletal age for boys allows a specific boy to be related to the population by plotting his leg length as a function of his skeletal age. It is useful in the analysis of leg-length data because it allows a projection into the future on the basis of the present situation. (From Anderson M, Green WT. Lengths of the femur and tibia; norms derived from orthoroentgenograms of children from five years of age until epiphyseal closure. *Am J Dis Child* 1948;75:279–290.)

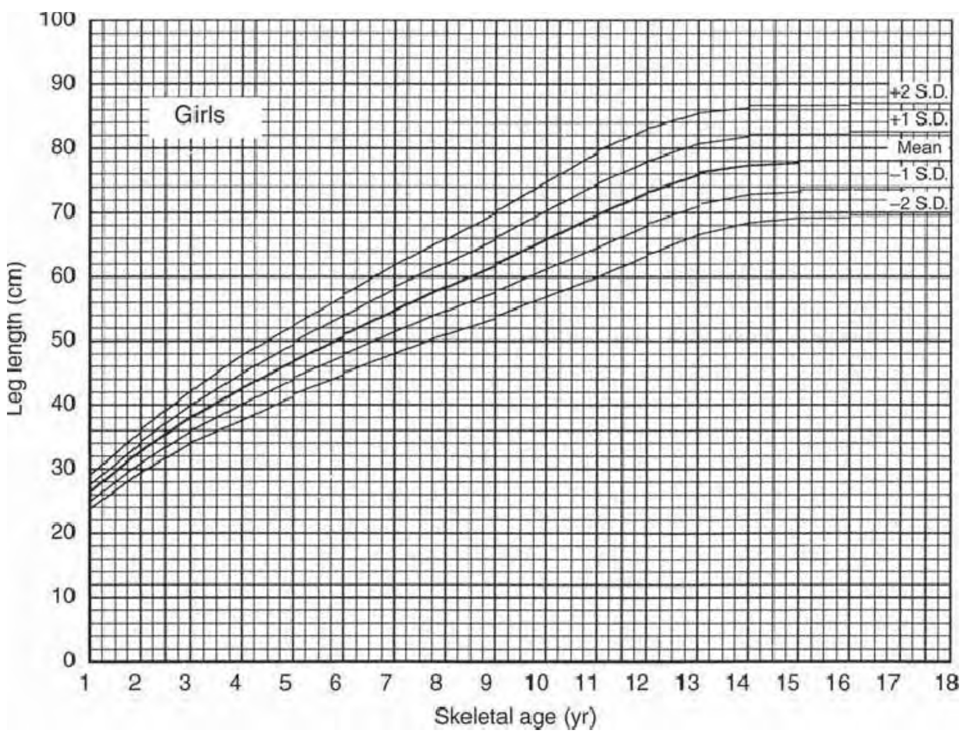


FIGURE 28-18. Graph showing total leg length versus skeletal age for girls serves the same purpose for girls that Figure 28-17 serves for boys. (From Anderson M, Green WT. Lengths of the femur and tibia; norms derived from orthoroentgenograms of children from five years of age until epiphyseal closure. *Am J Dis Child* 1948;75:279–290.)

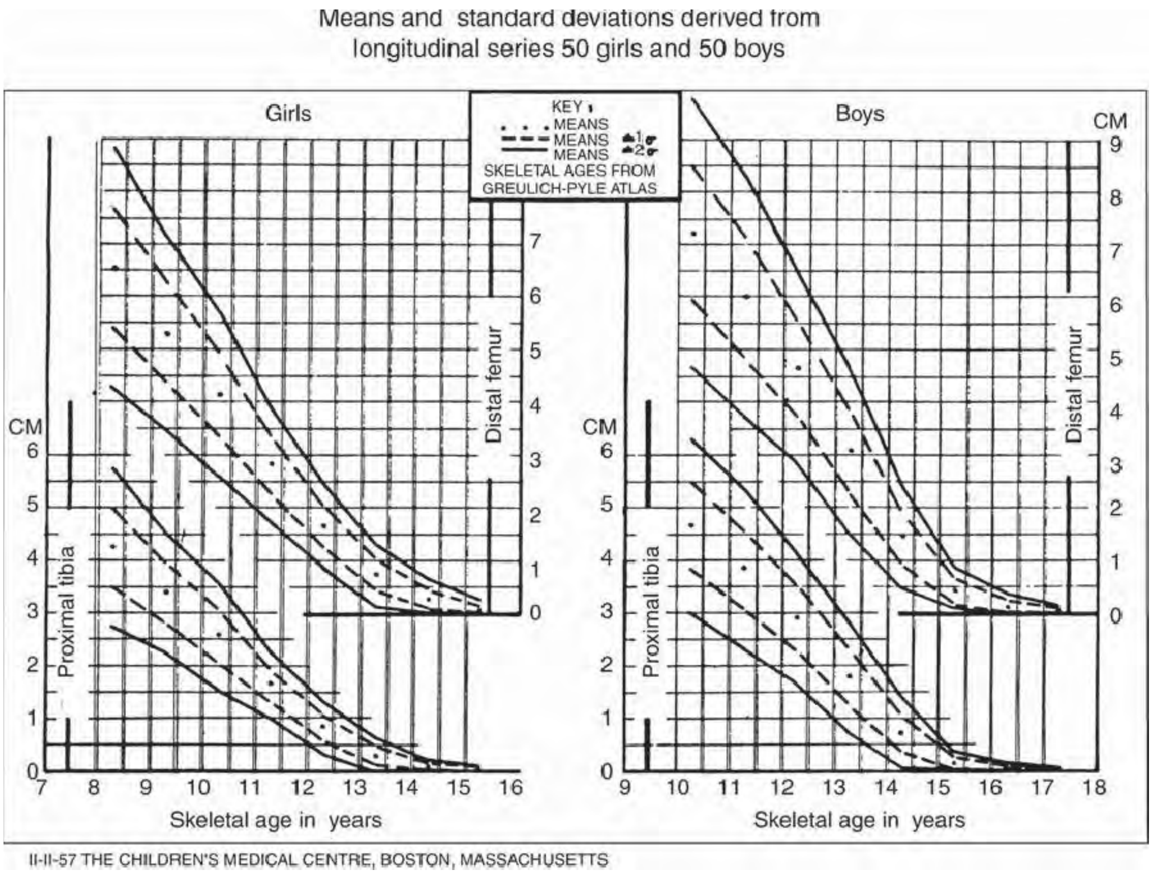


FIGURE 28-19. Green and Anderson growth-remaining graph. This graph shows the amount of growth potential remaining in the growth plates of the distal femur and the proximal tibia of boys and girls as functions of skeletal age. The graph is useful in determining the amount of shortening that will result from epiphysiodesis. (From Anderson M, Messner M, Green W. Distribution of lengths of the normal femur and tibia in children from one to eighteen years of age. *J Bone Joint Surg Am* 1964; 46-A(6):1197–1202.)

recorded as a straight line. The effects of epiphysiodesis can be determined by using any one of three (proximal tibia, distal femur, both) reference lines so that equivalent leg lengths are achieved at maturity. To utilize this chart, the length of the NORMAL leg is plotted on the given normative line. A vertical line is then drawn and the intersection of the reference skeletal age (determined from Greulich and Pyle measures) is recorded. The length of the ABNORMAL leg is then plotted on this same vertical line. The process is then repeated for at least three measurements. Best fit lines are then drawn on the skeletal nomogram and the ABNORMAL limb. A vertical line is drawn from the intersection of the skeletal age nomogram at maturity. The distance between these lines at maturity is the predicted discrepancy. Using the reference slopes for epiphysiodesis, one can determine when different combinations of epiphysiodesis could be performed to allow ultimate correction (Figs. 28-21 and 28-22).

This method remains very useful in predicting final limb length with a mean error of 0.6 cm using this technique (76, 101). After surgical intervention, typically epiphysiodesis or lengthening, the leg-length discrepancy can continue to be monitored on the same graph to see if further interventions will be needed. Recently, this graph has been updated to include more modern growth data, and the originators claim it is more accurate than the original Moseley straight-line graph (103). The authors do not have experience with this graph.

Menelaus/White—Arithmetic Model. The arithmetic model was first proposed by White (104) and is useful when only one data point exists for the prediction of ultimate discrepancy. It was developed to help predict the timing of epiphysiodesis and not to describe growth. He suggested the distal femur grew 3/8 in. (10 mm) per year and proximal tibia grew 1/4 in. (6 mm) per year and the discrepancy increases by 1/8 in. (3 mm) per year. This equated to 37% of total limb growth at the distal femur and 28% of total limb growth at the proximal tibia. White assumed boys stopped growing at 17 years and girls at 16 years. Menelaus later adjusted the age of growth cessation to be 16 years for boys and 14 years for girls. While calendar age was used in developing this method, Menelaus suggested it only be used when skeletal and chronologic age are within 1 year off each other and clinical used leg-length differences as determined by blocks and not radiographic length

Determing leg-length discrepancy: The growth-remaining method

A Assessment of past growth

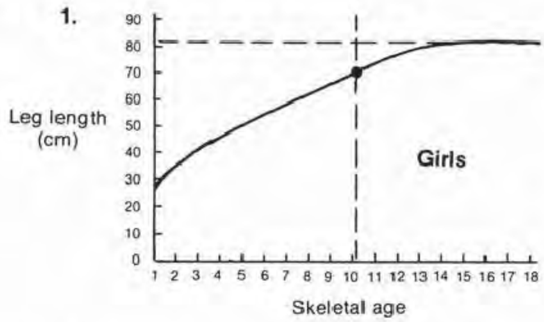
1. Growth of both legs
   = present length − first length
2. Present discrepancy
   = length of long leg − length of short leg
3. Growth inhibition
   $= \frac{\text{(growth of long leg − growth of short leg)}}{\text{growth of long leg}}$

1. Growth of long leg
   = 70.0 − 60.0 = 10.0 cm
1. Growth of short leg
   = 66.2 − 58.2 = 8.0 cm
2. Present discrepancy
   = 70.0 − 66.2 = 3.8 cm
3. Growth inhibition
   $= \frac{(10.0 - 8.0)}{10.0} = 0.2$ cm

B Prediction of future growth

1. Plot present length of long leg on Green-Anderson leg length graph for appropriate sex

1.



2. Project to right parallel to standard deviation lines until maturity to determine mature length of long leg
3. Future growth of long leg
   = mature length − present length
4. Future increase in discrepancy
   = future growth long × inhibition
5. Predicted discrepancy at maturity
   = present discrepancy + future increase

2. Length of long leg at maturity = 81.1 cm
3. Future growth of long leg
   = 81.1 − 70.0 = 11.1 cm
4. Future increase in discrepancy
   = 11.1 × 0.2 = 2.2 cm
5. Discrepancy at maturity
   = 3.8 + 2.2 = 6.0 cm

C Prediction of effect of surgery

1. The effect of epiphysiodesis of the distal femoral and proximal tibial plates for a given sex and skeletal age can be determined by the Green-Anderson growth = remaining graph.
2. The effect of lengthening is not affected by growth.

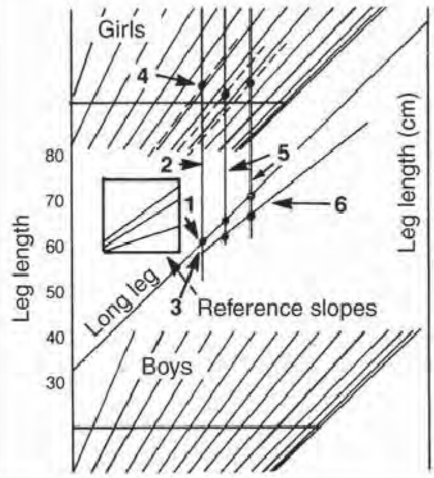
1. Correction from proximal tibial arrest
   = 2.7 cm

   Correction from distal femoral arrest
   = 4.1 cm

   Correction from combined arrest
   = 2.7 + 4.1 = 6.8 cm

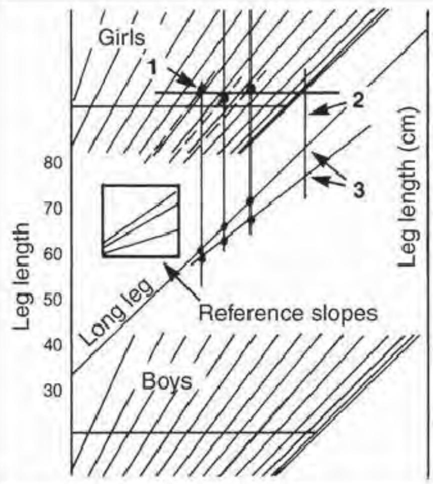
FIGURE 28-20. Step-by-step instructions for the use of the growth-remaining method.

Determining leg-length discrepancy: The straight-line graph method



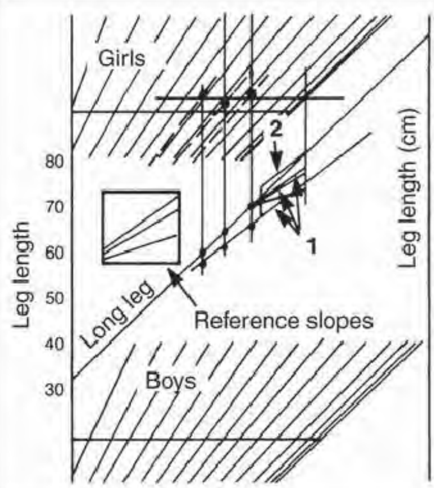
A Assessment of past growth

1. Plot the point for the long leg on the sloping line labeled "LONG LEG" at the appropriate length.
2. Draw a vertical line through that point representing the current assessment.
3. Plot the point for the short leg on the vertical line.
4. Plot the point for skeletal age with reference to the sloping lines in the nomogram.
5. Plot successive visits in the same fashion.
6. Draw a straight line through the short leg points to represent the growth of the short leg.



B Prediction of future growth

1. Draw the horizontal straight line that best fits the points previously plotted for skeletal age. The fit to later points is more important than to earlier points. This is the growth percentile line.
2. From the intersection of the growth percentile line with the maturity skeletal age line, draw a vertical line to intersect the growth lines of the two legs. This line represents the end of growth.
3. The points of intersection of the vertical line with the two growth lines indicate the predicted lengths of the legs at maturity.



C Prediction of effect of surgery

1. To predict the outcome after epiphysiodesis, draw three lines to the right from the last point for the long leg parallel to the three reference slopes. The intersections of these lines with the vertical line representing the end of growth indicates the predicted lengths of the long leg after the three possible types of epiphysiodesis.
2. To predict the outcome after leg lengthening, draw a line parallel to the growth line of the short leg but elevated above it by the amount of length gained.

FIGURE 28-21. Step-by-step instructions for use of the straight-line graph method.

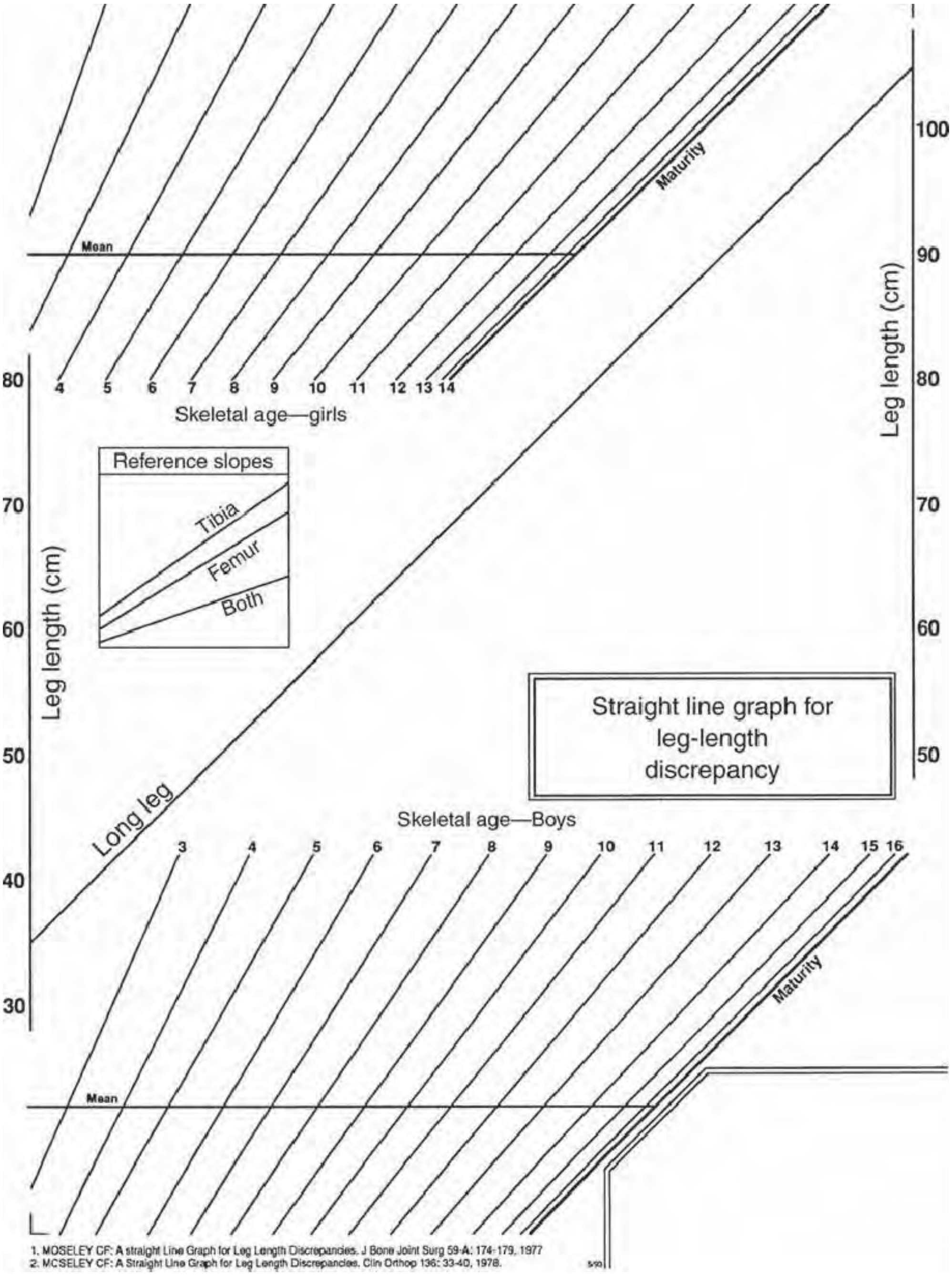


FIGURE 28-22. The straight-line graph comprises three parts: the leg-length area with the predefined line for the growth of the long leg, the areas of sloping lines for plotting skeletal ages, and reference slopes to predict growth following epiphysiodesis.

measurements. This method is best suited for those patients during the last few years of growth whose skeletal age correlates well with their chronologic age. The results of this technique for timing epiphysiodesis have found that 80% of patients were within a ½ in. when compared with the 90% obtained by using the Green and Anderson technique (105, 106) (Fig. 28-23).

Dimeglio Method. This method is similar to the White and Menelaus method; however, different assumptions are made regarding the growth and skeletal age. Dimeglio calculates the growth at the knee to be 2 cm per year (1.1 cm at the femur and 0.9 cm at the tibia) at the onset of puberty (bone age of 11 years in girls and 13 years in boys); in this model, growth ceases at bone age of 13.5 years in girls and 15.5 years in boys. Based upon this, four common scenarios are generated for each discrepancy and timing of epiphysiodesis. To treat a 5-cm discrepancy, epiphysiodesis of the distal femur and proximal tibia should be performed at the onset of puberty. For a 4-cm discrepancy, epiphysiodesis of the femur and tibia is performed 6 months after the onset

Determining leg-length discrepancy: The arithmetic method

Leg-length data

(for examples for all three methods):

Sex: Female

Age (yr)	Skeletal age (yr)	Right leg length (cm)	Left leg length (cm)
7 + 10	8 + 10	60.0	58.2
8 + 4	9 + 4	64.4	61.9
9 + 3	10 + 3	70.0	66.2

Prerequisite growth information

Distal femoral plate grows 10 mm/yr.
Proximal tibial plate grows 6 mm/yr.

Girls stop growing at 14 years of age.
Boys stop growing at 16 years of age.

A Assessment of past growth

1. Longest time interval for data
   = age at last visit – age at first
2. Years of growth remaining
   = 14 (16 for boys) – age at last visit
3. Past growth of legs
   = present length – first measured length
4. Growth rate of long leg
   $= \dfrac{\text{past growth}}{\text{time interval}}$
5. Growth inhibition
   $= \dfrac{(\text{growth of long leg} - \text{growth of short leg})}{\text{growth of long leg}}$

1. Longest time interval for data
   = 9 yr 3 mo – 7 yr 10 mo = 1 yr 5 mo
   = 1.42 yr
2. Years of growth remaining
   = 14 yr – 9 yr 3 mo = 4 yr 9 mo = 4.75 yr
3. Past growth of:
   long leg = 70.0 – 60.0 = 10.0 cm
   short leg = 66.2 – 58.2 = 8.0 cm
4. Growth rate of long leg
   $= \dfrac{10.0}{1.42} = 7.04$ cm/yr
5. Inhibition
   $= \dfrac{(10.0 - 8.0)}{10.0} = 0.2$ cm

B Prediction of future growth

1. Future growth of long leg
   = years remaining × growth rate
2. Future increase in discrepancy
   = future growth of long leg × inhibition
3. Discrepancy at maturity
   = present discrepancy + future increase

1. Future growth of long leg
   = 4.75 × 7.04 = 33.4 cm
2. Future increase in discrepancy
   = 33.4 × 0.2 = 6.7 cm
3. Discrepancy at maturity
   = (70.0 – 66.2) + 6.7 = 10.5 cm

C Prediction of effect of surgery

Effect of epiphysiodesis
= growth rate × years remaining

Effect of epiphysiodesis

Femoral	= 1.0 × 4.75 = 4.75 cm
Tibial	= 0.6 × 4.75 = 2.85 cm
Both	= 1.6 × 4.75 = 7.6 cm

FIGURE 28-23. Step-by-step instructions for use of the arithmetic method. The method presented here is modified from that presented by Menelaus and Westh, in that the future increase in discrepancy is calculated from growth acquired in the past instead of being assumed to be 0.3175 cm per year of growth remaining. An example is shown in the panels in the right column.

of puberty. For a 3-cm discrepancy, epiphysiodesis of the femur alone is recommended at the onset of puberty. For a 2-cm discrepancy, epiphysiodesis of the femur only is recommended 1 year after the onset of puberty. This strategy can be adapted to individual cases but stresses the importance of making treatment decisions at the onset of puberty and detecting this through Tanner staging (stage 2) and skeletal age (11 years in girls and 13 in boys). In this age group, hand and elbow radiographs were found to be effective in determining bone age (4, 107).

Multiplier Method. In order to calculate the ultimate discrepancy, defined multipliers have been determined from previously published growth data. Tables of multipliers (for each age and gender) decrease with age and when multiplied by the existing deformity, an ultimate discrepancy can be predicted. Thus by using the multiplier, the current leg lengths, and knowledge of whether the discrepancy is congenital or developmental, the clinician can estimate leg-length discrepancies at maturity. For congenital discrepancies, the discrepancy at skeletal maturity is easy to calculate.
Discrepancy at maturity = (L – S) × M, where L and S are the long- and short-limb measurements and M is the age appropriate multiplier. As developmental discrepancies have a constant rate of inhibition, the clinician must be able to calculate the rate of inhibition and the amount of growth remaining in the long limb. Thus, **Discrepancy at maturity = (L – S) + [1 – (S – S′) / (L – L′)] × L (M – 1),** where S, L are the current lengths and S′, L′ are the lengths from 6 to 12 months ago. From these two calculations, affects and timing of epiphysiodesis can be estimated using similar appropriate formulae. This method has been found accurate (108–110), and the originators state that chronologic age is as accurate as bone age using this method (Table 28-1).

In summary and as stated previously, all of the above methods assume constant growth and constant inhibition. Despite the knowledge that several inhibition patterns do exist (99), these do not appear to be of real clinical importance in estimating ultimate leg lengths. Studies have not consistently shown that one method is superior over the others and that skeletal age does not necessarily improve estimation in final discrepancies. For example, Kasser et al. (111) found a mean error of 2.4 cm using Anderson and Green data with chronologic age versus 2.6 cm using the straight-line graph with skeletal ages in children <10 years of age. The accuracy of the skeletal age determination has been brought into question. While no one technique is fail safe, the authors would recommend always utilizing at least two techniques when determining treatment. If there is a sizable discordance between these techniques, a third should be employed. Of course, this is not always possible; both the Moseley and Green and Anderson methods require using multiple data points. When a clinician encounters a patient for the first time near the epiphysiodesis date (10 to 14 years of age), a treatment decision based on elbow and hand radiographs may better help determine the true skeletal age.

TABLE 28-1 Multipliers

Age (yr)	Boys		Girls	
	Femur	Tibia	Femur	Tibia
0	5.90	5.40	4.64	4.76
1	3.26	3.21	2.94	2.99
2	2.60	2.56	2.39	2.39
3	2.24	2.22	2.05	2.06
4	2.00	2.00	1.82	1.84
5	1.82	1.82	1.66	1.67
6	1.68	1.69	1.53	1.54
7	1.56	1.57	1.42	1.43
8	1.46	1.47	1.33	1.34
9	1.37	1.38	1.26	1.26
10	1.30	1.31	1.19	1.18
11	1.24	1.24	1.12	1.12
12	1.18	1.17	1.07	1.06
13	1.12	1.11	1.03	1.02
14	1.07	1.06	1.00	1.00
15	1.03	1.03	—	—
16	1.01	1.01	—	—
17	1.00	1.00	—	—

From Paley D, Bhave A, Herzenberg J, et al. Multiplier method for predicting limb-length discrepancies. *J Bone Joint Surg Am* 2000;59A:1432–1446.

NATURAL HISTORY OF LIMB-LENGTH DISCREPANCY

The *natural history* of limb-length discrepancy in adults is of major concern, yet unfortunately the data on this are lacking. To fully test the effect of limb-length discrepancy on a patient's long-term function, one would need to study a cohort of affected individuals which could be compared to normal subjects. These groups would have to be large in order to control for genetic predisposition for arthritis, traumatic and lifestyle (obesity, smoking, exercise habits) factors, and other comorbid conditions that can affect the rates of limb arthrosis and back pain. In the growing years, the long-term effect of discrepancy is also not known. Therefore, it is helpful to consider the effects of limb-length discrepancy on the growing child in terms of *mechanisms of compensation*.

Mechanisms of Compensation. The parents of patients with leg-length discrepancy worry about developing problems of the hip, knee, and the spine. It is important to counsel families that the data on long-term effects of discrepancy are unknown in adults, and no data demonstrate damage to the growing skeleton as a result of discrepancy. The child with a congenital leg-length discrepancy usually compensates better than adults who may have an acquired leg-length discrepancy. The movements about the lower limb joints with simulated and real leg-length discrepancies have been found to be essentially unchanged with small discrepancies (112).

Often times, a young child with a congenital discrepancy of 3 cm may not feel right with a shoe lift and prefer to go without it. Song et al. found that discrepancies >5.5% of the long extremity increased the mechanical work performed by the long limb and increased the vertical displacement of the center of body mass, with consequent energy penalty. Children with lesser discrepancies were able to normalize the work performed by the two extremities. These children compensate for minor degrees of leg-length discrepancy by walking on the toes of the short leg, with the heel rarely touching the ground. This can result in a smooth, symmetrical gait that shows no abnormality except for the lack of heel strike on the short side. Children who are older or who have a discrepancy of 4 to 5 cm may also compensate for the discrepancy by flexing the knee on the long side or more commonly vaulting over the long leg. This action produces excessive up-and-down motion of the pelvis and trunk.

Natural History in Adults. It has been shown that discrepancies of <2 cm are of no functional or clinical consequence in adults and that these discrepancies do not require treatment (113). Despite evidence to suggest that discrepancies of <2.5 cm are not significant in the adult (114), postural sway has been shown to increase when simulated discrepancies are as small as 1 cm (115). Liu et al. (116) proposed the "symmetry index" (SI) as a measure of the quality of gait and found that correction of discrepancy by a heel lift considerably improves the SI.

It has been hypothesized that *idiopathic* arthritis of the hip in the elderly patient may actually be the result of some previously unrecognized mild dysplasia, slipping of the capital femoral epiphysis, or leg-length discrepancy. It is conceivable that pelvic obliquity from limb-length discrepancy would decrease the coverage of the hip of the long leg in two-legged stance and with greater discrepancy; a more significant decrease in the center-edge (CE) angle could be expected to result in arthritis (Fig. 28-24). Despite this theory, there is no documentation to prove this hypothesis, and such a study would be difficult to conduct as mentioned above. Leg-length discrepancy may increase the incidence of knee pain in athletes, although the nature of the relation has not been elucidated (117).

The effects of leg-length discrepancy on the spine are also not clearly established. Contradictory evidence exists about the possibility that leg-length discrepancy causes low back pain in the long term (118–120). Low back pain is unusual in the younger child and is more common in the adolescent, but there is no evidence that low back pain and leg-length discrepancy are related in this age group. Froh et al. (121) studied whether leg-length discrepancy had any effect on the orientation of the facet joints in adults and found none, whereas Giles and Taylor (122) found changes in the facet joints of cadavers with leg-length discrepancy. It is not clear whether the incidence of back pain is higher in patients with leg-length discrepancy than it is in the general population.

It is controversial whether leg-length discrepancy leads to structural scoliosis. Gibson et al. (123) assessed 15 patients with leg-length discrepancy following femoral fractures and found that after 10 years none had structural scoliosis. An increased incidence of structural scoliosis in patients with leg-length discrepancy has been noted when compared with the general population (124, 125); but it is hard to attribute leg-length discrepancy as the cause of scoliosis. If this were the case, the convexity of the scoliosis would be in the direction of the shorter limb; but in up to one-third of the cases, the scoliosis was in the opposite direction (Fig. 29-4). Because the leg-length discrepancy affects the spine only during two-legged stance, the skepticism toward the cause-and-effect hypothesis of length discrepancy and scoliosis seems justified.

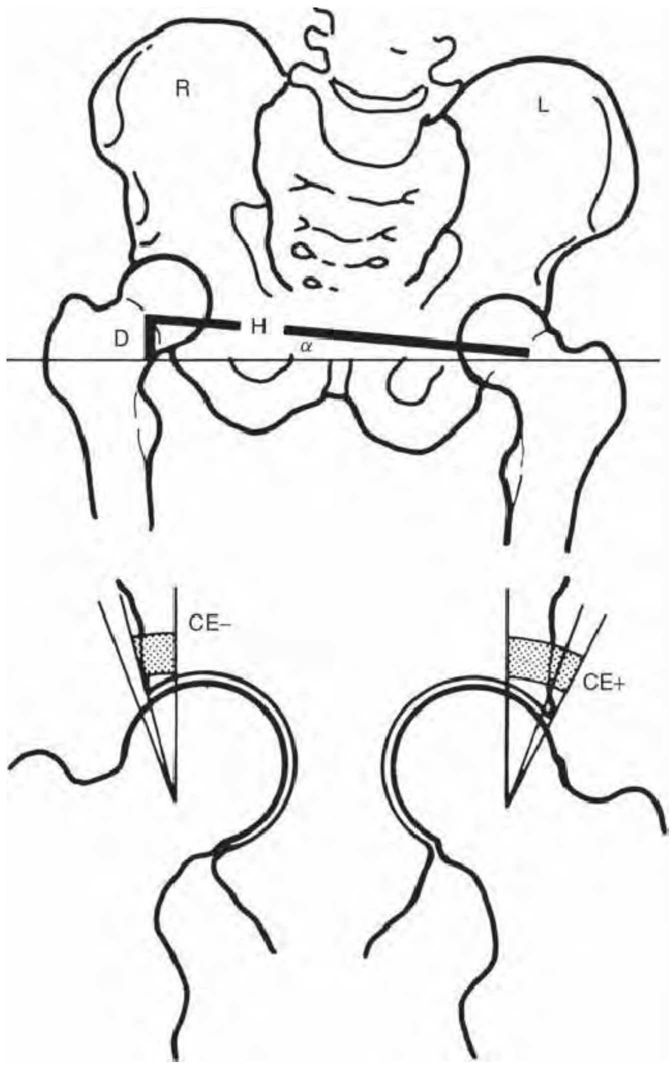


FIGURE 28-24. Decrease in CE angle with pelvic obliquity. The CE angle is decreased on the side of the long leg. Coverage is decreased and the resulting decrease in the load-bearing area causes an increase in pressure. Such a hip may be susceptible to late degenerative arthritis. (*L*, left; *R*, right; *H*, Hilgenreiner line; *D*, leg-length discrepancy.) (From Morscher E. Etiology and pathophysiology of leg-length discrepancies. *Prog Orthop Surg* 1977;1:9.)

TREATMENT OF LIMB-LENGTH DISCREPANCY

General Treatment Principles. Limb-length discrepancy is a condition that the lay public can conceptually understand; unfortunately, the public is also subject to misinformation on the implications of the discrepancy and treatment. As such, the physician is required to discriminate real and apparent (or positional) discrepancies in length, explain the facts and discount the myths which may be well entrenched. Patients referred for apparent discrepancies <1 to 1.5 cm may be due to positioning during assessment or may result from joint contractures. For the overly concerned parent with a child with a positional discrepancy, a standing or supine alignment radiograph can go a long way in allaying fears.

When infants or young children present with a significant limb-length discrepancy, several clinic visits may be required to develop a good relationship with the family. A gradual approach allows the family to understand why the limb is affected and to psychologically come to grips with the implications for their child. For instance, it may take several meetings for the family to appreciate why the disorder exists, quantitate the current discrepancy, predict the discrepancy at maturity, and most importantly, to understand the effect on their child's health. Naturally, families will want to know as soon as possible what treatment is likely; yet caution is needed to prevent informational overload at the early visits. Even in instances of significant shortening in which an infant will require reconstruction (usually several years to a decade later), an in-depth discussion on the risks of limb lengthening is not warranted at the initial visits. With time, a gradual introduction and education for future treatment is critical to develop a good parent–patient–surgeon relationship.

Treatment Goals. Treatment varies according to patient age, the current discrepancy, and the projected discrepancy at maturity. While the child is growing, the family and the surgeon may temporize the situation with shoe lifts or other prosthetic options. The ultimate choice of treatment depends on the predicted discrepancy at maturity. There are three ways to ultimately treat a limb-length discrepancy. The simplest method is to plan to continue the prosthetic or orthotic resources used in the childhood years. This is usually the best option in the two extreme situations—patients with slight discrepancies or in those whose projected discrepancy is so large it precludes limb reconstruction. In some of the later patients, surgical procedures may be needed to optimize the use of these appliances.

Should limb equalization be the ultimate goal, the two options are to either shorten the long limb or lengthen the short limb. In the former scenario, one can consider *procedures* such as epiphysiodesis to slow the growth of the long leg or in mature patients to consider shortening of the bone via removal of a segment of bone. If the limb is to be lengthened, it may require just a simple lengthening which involves an initial procedure and a subsequent distraction of the limb. In some patients, a limb may require a series of surgical procedures to optimize the limb prior to limb lengthening. In either scenario of simple limb lengthening or more complicated limb reconstruction, it is more apropos to consider limb lengthening to be a *process*, more than a procedure.

There is some flexibility in the following guidelines to account for factors such as environment, motivation, intelligence, compliance, emotional stability, patient's and parents' wishes, and associated pathology in the limb. Fairly straightforward guidelines expressed in terms of the magnitude of the predicted discrepancy can be used to choose from among the major treatment categories:

0–2 cm	No treatment
2–6 cm	Orthotic use, epiphysiodesis, skeletal shortening
6–20 cm	Limb reconstruction (limb lengthening with or without adjunctive procedures)
>20 cm	Prosthetic fitting (with or without surgical optimization)

Because there is some advantage to being tall (126–129), lengthening procedures are often preferred by parents and patients. Despite this, lengthening is not generally done for discrepancies <6 cm because of the high morbidity and complication rate of lengthening. This procedure should be avoided in favor of epiphysiodesis or shortening whenever possible. The exception is in those patients who require a concomitant osteotomy to correct deformity, and therefore distraction through the osteotomy site can be performed for residual length discrepancies <6 cm.

Shortening procedures are usually not appropriate for correction of >6 cm, because a disproportionate appearance may not be pleasing to the patient. The exception exists when epiphysiodesis is performed to correct a discrepancy of any magnitude when the long leg is clearly the abnormal one. In this case, the procedure corrects the abnormally long leg and does not result in abnormal proportions. Tibia skeletal shortening is rarely done; in the femur, shortening >10% of the bone length affects the bone–muscle length relationship resulting in muscle weakness. In addition, acute femoral shortening of 5 to 6 cm in mature patients is also not recommended because the bulbous thigh appearance is usually objectionable.

The rough goal of limb shortening or limb reconstruction is to equalize both limbs, provided no other comorbidities need to be accommodated. For instance, undercorrection of 1 or 2 cm would be considered *functionally equal* and best for patients with paralysis of the short leg. The residual discrepancy facilitates clearing of the floor by the weak short leg during the swing phase of gait, and this is even more important in patients who wear braces or have diminished swing phase knee flexion in gait. Whether skeletal shortening, epiphysiodesis, or bone lengthening, the method of correction is chosen with the ultimate goal of making the limb lengths functionally equal; a secondary goal is to consider treatment that will allow a patient's body to be as symmetrical as possible. In order to maintain symmetrical knee height, one should consider lengthening the shortest bone or shortening the bone corresponding to the shortest bone on the long leg side. This only applies to instances where morbidity of treatment is equal. For instance, a mature adolescent with a 4-cm short femur would be a good candidate for acute femoral lengthening on the long side. A similar patient with a 4-cm short tibia could be similarly managed even though this would result in abnormal knee heights. In this instance, femoral shortening is chosen because it is technically easier with fewer complications than tibial shortening.

Orthotic Treatment for LLD. Not every patient with a discrepancy will benefit or want a lift; for instance in mature patients, no lift is required for discrepancies <2 cm. In young children with congenital discrepancies in length, parents may not opt to consider a lift until the child has a discrepancy of 4 cm. These patients are used to a short leg and have adapted to it; practically speaking, these children are often barefoot and would not want to wear a shoe all of the time. Rapidly growing and active children would likely need more than one lift per year and some insurance companies may only pay for per year.

A lift is a potential treatment for discrepancies up to 6 cm and is a satisfactory adjunct for those patients with greater discrepancy who are not appropriate for surgery. It is important to realize that the discrepancy to be treated may be different from the radiographic measurement, perhaps indicating that there is a discrepancy in the foot height. Blocks can be used in clinic to estimate the extent of correction that feels best to the patient; the final height of the lift can be determined by clinical trials in which the lift height is temporarily modified to suit the patient.

Depending on the patient, physicians may opt to recoup discrepancy with lifts inside the shoe (usually 1.5 to 2 cm is the maximum), with the remainder, if necessary, applied to the bottom of the shoe. For larger discrepancies, the height of the lift should be less than the discrepancy. Lifts higher than 5 cm are poorly tolerated because they may be heavy for a patient with congenital limb deformity. In addition, the leg muscles are not strong enough to resist inversion stress, and frequent ankle strains may result. If a higher lift is required, an orthotic extension up the posterior calf or above the malleoli can be added for stability.

In patients who have a congenital amputation in addition to a short leg, the leg-length discrepancy usually can be made up in the prosthesis and thus obviate limb-equalization surgery. Prosthetic fitting after limb ablation surgery is a treatment of last resort but may be eventually needed in infants with a femur that is less than half of the length of the contralateral femur (130, 131). Amputation (Syme or Boyd procedures) and prosthetic fitting is chosen if the discrepancy is anticipated to become >15 to 20 cm and especially if the patient has a functionally useless foot (14). In order to maximize prosthetic function, patients may benefit from adjunctive procedures which may include iliofemoral fusion, knee fusion, and Van Ness Rotationplasty. Foot ablation and prosthetic fitting is the preferred treatment when multiple procedures and lengthenings are needed to correct limb deformity and length discrepancy in extreme cases. Depending on how one defines complications from limb lengthening, families should expect one problem/complication that will require an additional treatment/surgery for each lengthening procedure. Furthermore, families should expect an average treatment period (from surgery to recovery) of at least 1 year for each limb lengthening.

An enormous physical and psychological risk to a child would be expected if heroic attempts are taken to salvage a marginal limb with an 18- to 20-cm discrepancy. Some patients with fibular hemimelia and an unstable ankle do better with amputation and prosthetic fitting than with multiple hospitalizations and surgical procedures to conserve the foot and lengthen the leg. This approach has the advantage of involving only one hospitalization and one definitive operation. They have an almost normal walking gait and can participate in recreational and sporting activities. Children who are treated for proximal focal femoral deficiency require above-knee prostheses and they function well, although not as well as the children who have undergone below-knee amputations. Some of the above-the-knee prostheses can function as below-the-knee prostheses following a Van Ness rotationplasty, in which the reversed ankle functions as a knee, providing active control and motor power to the prosthetic knee (132).

Although it is difficult for the parents to decide on an amputation in young children, those who undergo surgery and prosthetic fitting early in life show very good results. The optimal time for performing the Syme amputation is toward the end of the first year of life and for performing the rotationplasty is at approximately 3 years of age. It is helpful for parents of children who are candidates for these procedures to meet older children who have undergone the same procedure and to talk with their parents.

Epiphysiodesis. Epiphysiodesis is an excellent way to treat mild-to-moderate discrepancy in length; as a requirement, appropriate candidates must have enough growth remaining to recoup differences in length. The advantage to this method is the low morbidity and complication rate, thus making it the treatment of choice for surgical correction of moderate length discrepancy (104, 133–135). It is particularly useful in cases of limb overgrowth from fracture, inflammation, or overgrowth syndromes such as in hemihypertrophy. It is an excellent way to prevent limb-length discrepancy from occurring in the case of growth arrest from trauma or infection or tumor. For instance, in the case of a 12-year-old boy with an established growth arrest of his distal femur from trauma, an epiphysiodesis of the contralateral distal femur would be immediately indicated to avoid the expected 4-cm discrepancy at maturity.

The operation is effective by slowing the growth rate of the long leg and by allowing the short leg to catch up. It is necessary to take into account the ability of the short leg to catch up; this is done by predicting the growth inhibition to correct the discrepancy at maturity. Epiphysiodesis is a highly acceptable procedure because it is straightforward, does not require postoperative immobilization due to instability, and disables the child minimally. It is most suitable for those children who have sufficient leg-length data to enable a confident prediction of the discrepancy at maturity of 2 to 6 cm (136).

After epiphysiodesis, the leg grows at a slower rate, having lost the contribution of the operated physis toward the process of growth. The loss is 27% for the proximal tibial, 38% for the distal femoral, and 65% for combined epiphysiodesis of both plates. The surgeon therefore induces a known degree of growth inhibition and has three discrete choices for shortening strategies. The amount of desired shortening can be achieved only by performing the surgery at the correct time. Performing the operation too late results in undercorrection, and performing it too early results in overcorrection.

The procedure itself is relatively easy to perform; a certain difficulty arises in explaining it to the family and performing it at the proper time. Most families have an initial reticence about performing the operation on the good leg. In addition, the families can be concerned that the surgery will halt all of the child's growth or that they will actually get shorter. Furthermore, it can be difficult to explain why different growth plates grow at different rates. In order to improve understanding, it may be helpful to describe growth plates as automobile engines; some are more powerful than others, and in cases of discrepancy, one leg has a more powerful group of (growth) engines than the other. In order to make the legs equal, the epiphysiodesis is tantamount to placing a governor on the more powerful engines so that the other leg can catch up at the end of growth (finish line).

Determining the appropriate time to perform the procedure is the most challenging part of the treatment. Performing the procedure too early will result in the short leg being longer than the leg undergoing the growth arrest; conversely, if it is done too late, then there will be incomplete correction. The Multiplier method and the Moseley growth chart are appropriate mechanisms to gauge the appropriate time to perform the growth arrest (109). Use of the arithmetic method is most appropriate for patients who present close to the optimal time of epiphysiodesis and for whom there is no information on past growth. In order to do so, we assume that boys stop growth at 16 years of age, girls stop at 14 years of age, and the distal femur provides 10 mm of growth and the proximal tibia 6 mm of growth per year. Therefore a pan-genu epiphysiodesis could be expected to recoup 1.6 cm of growth per year. This works well when the chronologic age and skeletal age are closely related; when discordant it is controversial whether the bone age or the chronologic age should be used. For instance, how does one predict the effect of a pan-genu growth arrest in a 13-year-old boy when his skeletal age is 11.5 years. Based on chronologic age, a growth arrest at this age would lose 4.8 cm in the long leg. On the other hand and based on skeletal age, a growth arrest would lead to a loss of 7.2 cm. Blair et al. (137) reviewed 67 epiphysiodeses and found that correction to within 1 cm had been achieved in only 22 cases, and 35 failures had occurred because of the incorrect use of the Green and Anderson data. It is better to aim for 0.5 to 1.0 cm of undercorrection by doing the epiphysiodesis slightly later than the time for perfect correction. To improve symmetry, the procedure is best done in the bone that is opposite the shortest one on the other-side leg, although this principle may have to be compromised if future growth is insufficient for such an epiphysiodesis to be effective.

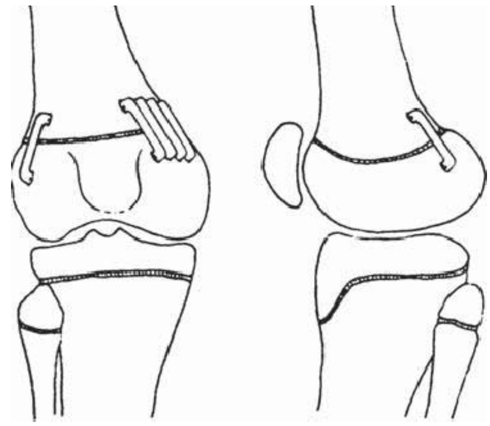


FIGURE 28-25. Epiphysiodesis by stapling. Growth arrest can be accomplished by careful placement of three extraperiosteal staples over the medial and lateral aspects of the plate.

Blount produced physeal arrest by placing three staples across the medial and lateral physis (138–140) (Fig. 28-25). He proposed temporary arrest and that growth would resume after removal of the staples (141, 142). This concept is attractive because it obviated the need to make predictions of future growth. Unfortunately, this could not be predicted with certainty, and cases of undesired arrest, angular deformity, or rebound growth upon removal occurred (143–145). In addition, some staples lost fixation, entering the adjacent joint, or caused overlying bursitis: these implants therefore lost proponents and was considered to be a permanent form of growth arrest. Temporary instrumented epiphysiodesis with medial and lateral rigid staples may be used for the rare patient with overgrowth that is significant at an early age and would be >6 cm at maturity (e.g., limb gigantism associated with Macrodactyly or Klippel-Trenaunay-Weber syndrome). In these cases, the medial and lateral rigid stapling can slow the growth for 1 to 2 years and then be removed to be reinserted some time later or followed with definitive epiphysiodesis. It is possible to perform stapling in such a way that normal growth resumes when the staples are removed (146–148). Technical details are important such as remaining extraperiosteal and not directly exposing the growth plate. Some surgeons have recently considered growth arrest with the use of modular plate and screw devices placed on both sides of the growth plate. It is the author's opinion that these devices, designed for hemiepiphyseal stapling, should not be used to arrest growth because of the risk of implant failure and asymmetrical growth arrest leading to iatrogenic deformity. Other methods of growth arrest include the use of transphyseal screws as originally described by Metaizeau in 1998 (149–151). Khoury et al. (149) recently reported good results of growth arrest with transphyseal screw epiphysiodesis (Fig. 28-26).

The principle of uninstrumented growth arrest is to produce a symmetrical bony bridge that tethers the physis and prevents future growth. Traditional epiphysiodesis requires incisions on both the medial and lateral aspects of the physis, a total

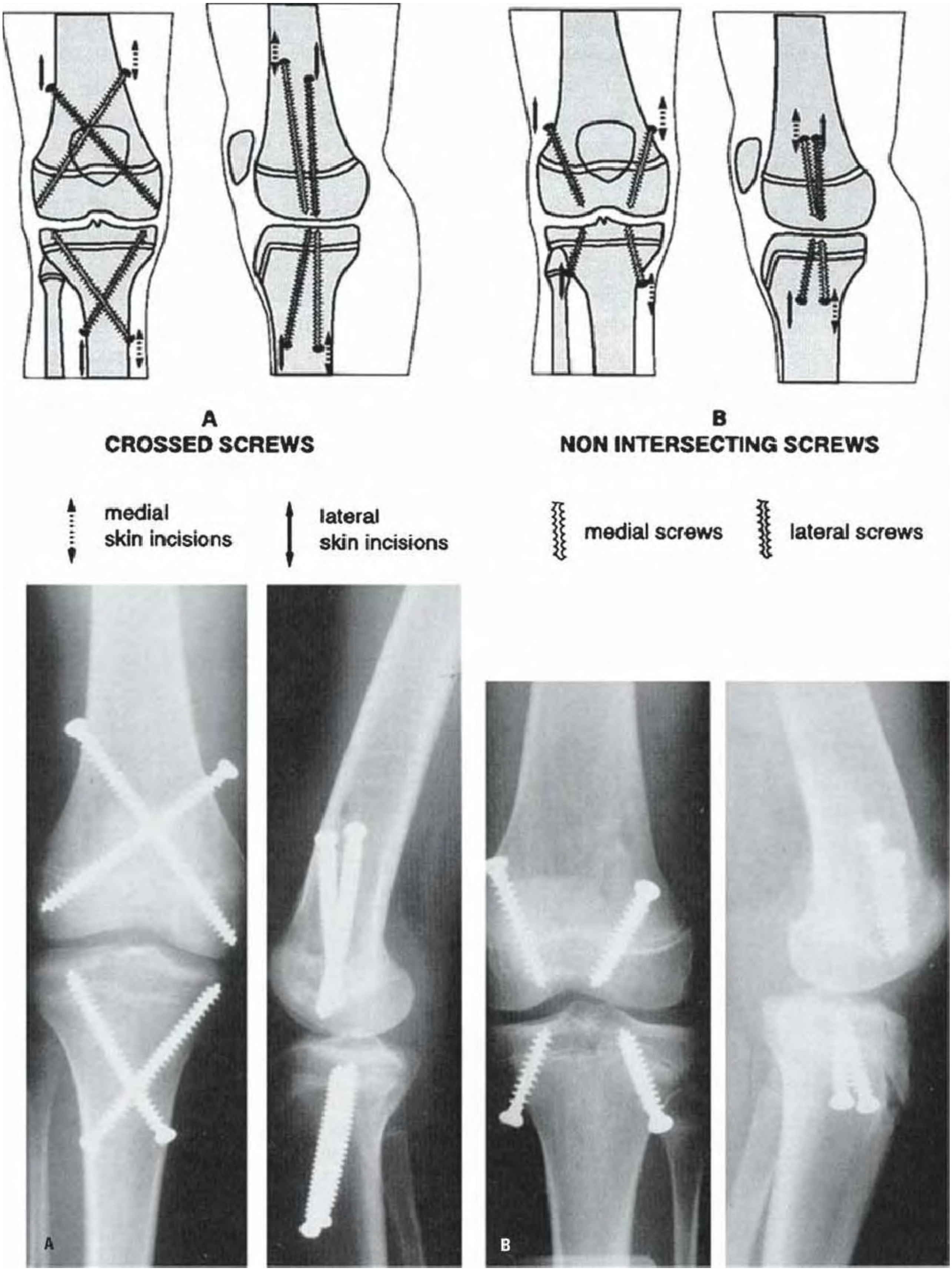


FIGURE 28-26. Epiphysiodesis can be accomplished with percutaneous placement of transphyseal screws as described by Metaizeau et al. (From Metaizeau JP, Wong-Chung J, Bertrand H, et al. Percutaneous epiphysiodesis using transphyseal screws (PETS). *J Pediatr Orthop* 1998;18(3):363–369.)

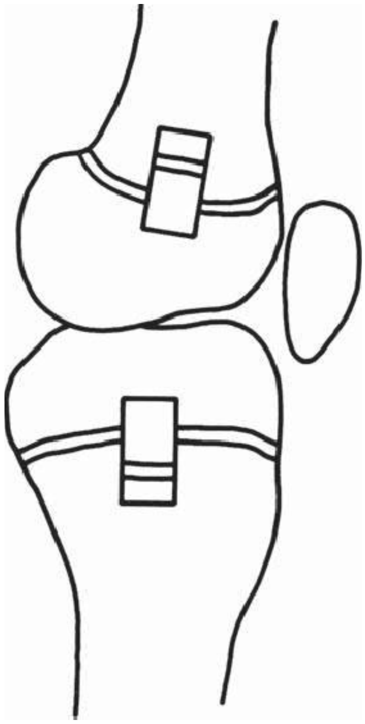


FIGURE 28-27. Epiphysiodesis by the Phemister technique. A rectangular bone block is replaced in reverse position to produce a bar across the growth plate.

of four incisions if pan-genu arrest is performed. Traditional open techniques involve removing a block of bone from both sides of the plate and reorienting the block of bone to produce a bony bridge. Phemister removed a rectangular block that was two-thirds on the metaphyseal side and one-third on the epiphyseal side of the plate, and then put it back in a reversed position (152) (Fig. 28-27). Others have designed a box chisel or circular trephine to remove a square block or cylinder of bone that can be rotated 90 degrees before replacement (104). Macnicol and Gupta have reported a percutaneous version of the Blount technique (153). All these procedures serve to bridge the physis medially and laterally with the solid bone.

The current authors advocate percutaneous epiphysiodesis under fluoroscopic guidance as the current standard of care. Epiphysiodesis has considerable advantages over other approaches because of its low morbidity and low complication rate, but there are minor disadvantages (154, 155). The advantages include small scars, thus avoiding unsightly scarring (153, 156, 157). Disadvantages to the method include potential for incomplete growth arrest resulting in angular deformity or continued growth. Scott, for example, reported a rate of continued growth of the physis of 12% (158). Additionally, we have seen several children develop inflammatory reactions within the soft issues that can be mistaken for cellulitis. Although most percutaneous epiphysiodesis are performed around the knee for length discrepancy, similar techniques with smaller drills can be used for the upper extremity or distal tibia.

In the preoperative area, the patient's surgical limb is identified by the patient and family; the site is marked by the surgeon or the patient. Intraoperatively, a "time-out" by the surgical team confirms patient name, birth date, antibiotic prophylaxis, site of surgery, and planned procedure. After appropriate anesthetic induction, patients are positioned supine on a radiolucent table and fluoroscopic images confirm appropriate visualization of the growth plates by lining up the image intensifier beam perfectly parallel to the growth plates. This can be challenging for the distal femur where the growth plate is biconcave in nature; the tibia growth plate is easier to visualize because it is flat and parallels the joint line which is posteriorly sloped 10 degrees. A sterile tourniquet is applied and inflated to 250 mm of Hg of pressure prior to incision.

Percutaneous epiphysiodesis is usually performed through one medial and one lateral incision. Edmonds et al. (159) found fourfold lower rates of complications when a two-incision approach is chosen over a single portal approach. After incising the skin and fat, the fascia is divided which allows the surgeon to place and drill a k-wire 1 cm into the periphery of the growth plate. Once confirmed radiographically, a 7 mm cannulated drill is placed over the wire and advanced horizontally, two-thirds the way across the growth plate. The drill bit is then backed up to the entry site and is directed anteriorly and advanced along the path of the growth plate and then redirected and passed posteriorly along the path of the growth plate. Between passes, the surgeon must adjust the trajectory of the drill bit anteriorly and posteriorly in order to remove the intervening growth cartilage. Care is needed to prevent inadvertent cortical breech and damage to the surrounding joint or soft tissue when the drill is directed anteriorly and posteriorly (Figs. 28-28 to 28-30). Because the distal femoral plate is biconcave and not perfectly flat, there is a technical challenge in making sure that the tip of drill is in the plate and that it stays there. In the distal femur, the intercondylar notch intrudes into the posterior aspect of the plate; anteriorly, the surgeon must be careful not to inadvertently enter the patella groove by being sensitive to the feel of the drill touching the cortical bone. The opposite incision is then made and the same technique is used to remove the growth plate anteriorly and posteriorly under fluoroscopic control. From the opposite entry site, the surgeon will be able to pass the drill bit all the way across the growth plate connecting the two areas and in an attempt to remove the cartilage adjacent to the opposite entry site. Once a connection between the two sites has been made, a curette can be used to remove the growth plate in the hard-to-reach corners. We flush the growth plate with sterile saline (without bubbles) to remove the debris and more clearly visualize the resected area via fluoroscopy (Fig. 28-31).

Approximately 50% of the area of the plate should be removed in the pattern shown in Figure 28-32. This extent of removal is sufficient to ensure arrest of the physis and maintains enough bone strength through residual plate and surrounding periosteum and perichondral ring to prevent fracture. Tibial

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Technique of Percutaneous Epiphysiodesis (Figs. 28-28 to 28-30)

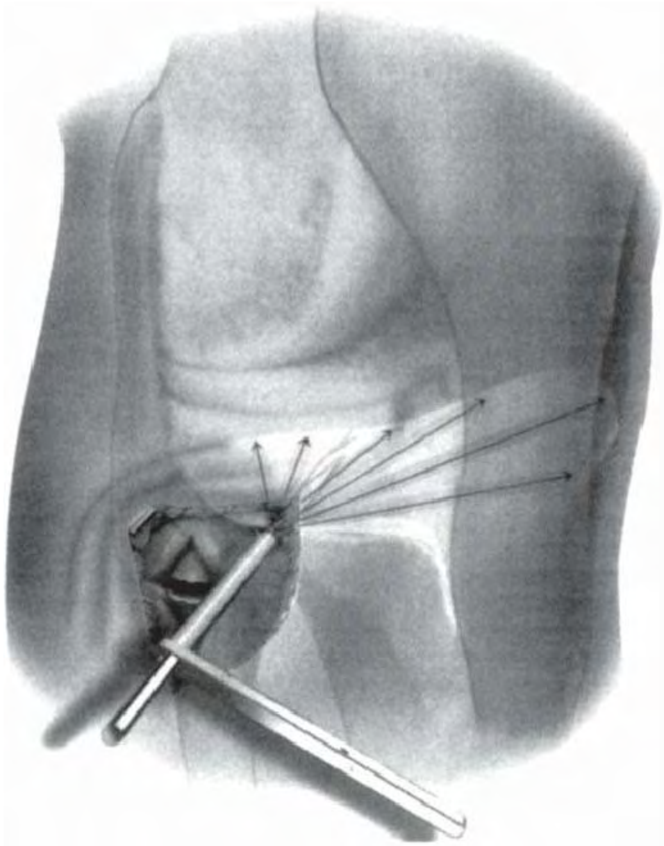
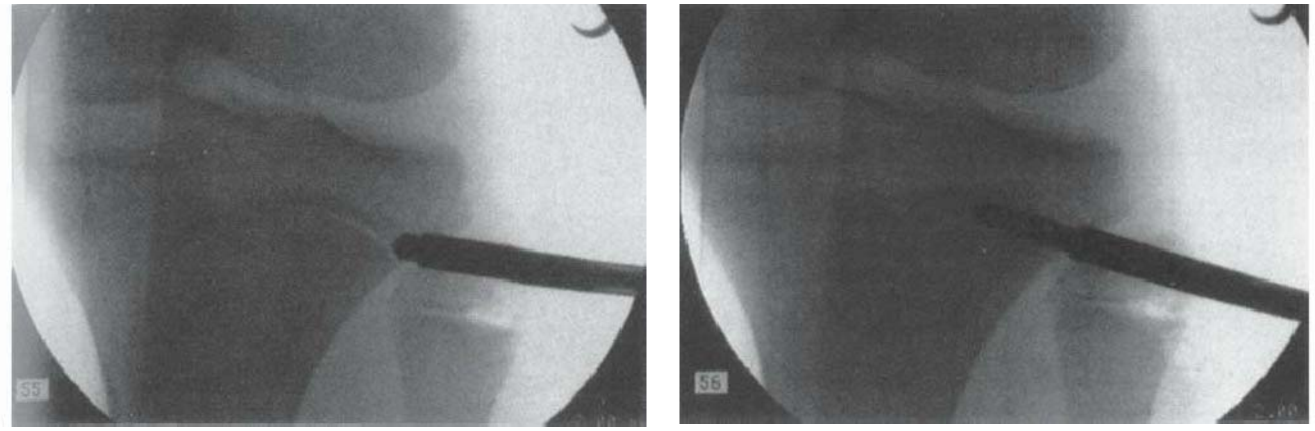


FIGURE 28-28. Technique of Percutaneous Epiphysiodesis. The technique for destruction of the tibial physis is the same as that described for percutaneous epiphysiodesis of the distal femur. A drill is advanced across the physis under image intensifier control. Several passes are made going anteriorly and posteriorly, with care taken that each time the drill is advanced, it remains in the physis and not above or below it. Care need also be taken so that the drill remains within the bone. The closer together these drill tracks are made, the easier the next step will be.



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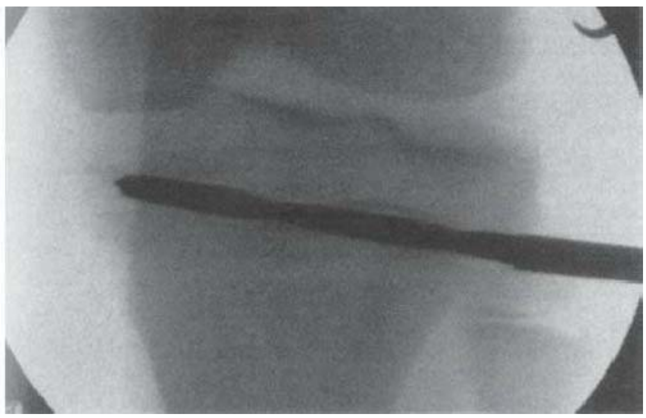

C

FIGURE 28-29. If the fibula was arrested with the open technique, it is a simple matter to start the drill into the bone through the wound. If not, a small stab wound is made opposite the physis. It is possible to do this from only the lateral (or medial) side in a normal-shaped physis. **A–C:** The drill is slowly advanced, checking with the image intensifier to be sure that the drill remains in the physis. This is done with each pass, as the surgeon moves the drill more anteriorly and posteriorly with each successive pass. In some cases, the physis follows an undulating course, which requires the epiphysiodesis to be performed from both the medial and lateral sides. Besides, some surgeons feel uncomfortable passing the drill completely across the physis, especially posteriorly, for fear of going outside of the bone and injuring an important structure.

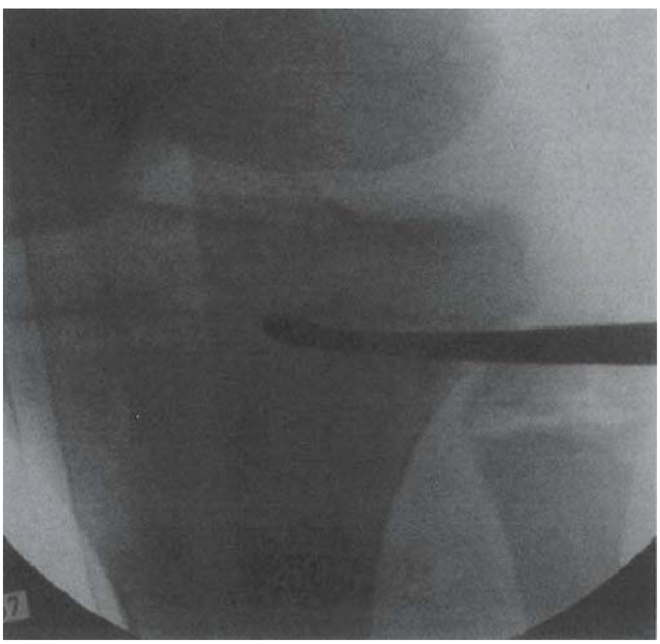

FIGURE 28-30. After multiple passes with the drill, a curette is used to connect the drill holes and further remove additional physis. The curette should be checked with the image intensifier to be certain that it is removing physis and not unnecessary metaphyseal bone. Also, an effort should be made to keep the hole in the cortex as small as possible to minimize the bleeding into the soft tissues. The wound is closed with absorbable sutures. A drain is not helpful because it blocks readily with the material from the bone. If the limb has 2 years or less of growth remaining, it is usually not necessary to arrest the proximal fibula. In this case, it is usually easier to perform the epiphysiodesis from the medial side because the border of the tibia is subcutaneous. This is the only time the operation is truly percutaneous.

FIGURE 28-31. Radiographic images after percutaneous growth arrest demonstrates ablation of both the distal femur and the proximal tibial growth plates.

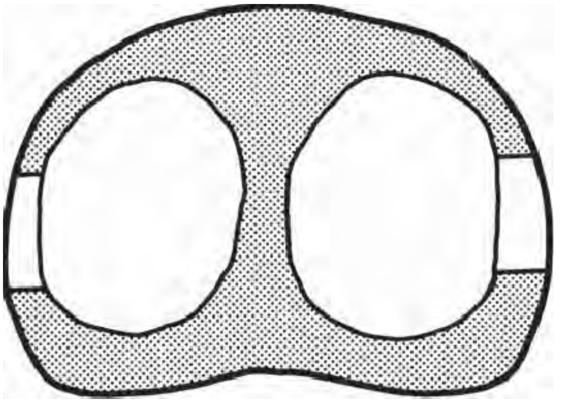

FIGURE 28-32. Area of plate to be removed in epiphysiodesis. Obliteration of medial and lateral circular segments of the plate, leaving the central part and the strong periphery, successfully stops growth, yet the bone retains sufficient strength to forego immobilization.

epiphysiodesis should be accompanied by arrest of the proximal fibular physis if the tibial shortening is >2.5 cm (160). The fibular epiphysiodesis can be performed through the same skin incision as the lateral aspect of the tibia, but the surgeon must approach the fibula from the anterior aspect to avoid the peroneal nerve. Because the tibia has a flat physis, one may be tempted to try and arrest the growth plate from only a single incision; this can lead to complications such as incomplete growth arrest and angular deformity (159).

Postoperatively, we place a compressive dressing for 3 days to reduce swelling and we routinely perform a femoral nerve block with 1/4% marcaine for postoperative pain control. Even though the bones are considered strong enough for routine ambulation, patients receive a knee immobilizer and crutches for ambulation in order to decrease pain. Once the pain resolves, these modalities are not needed as the child is weightbearing as tolerated. Three days after surgery, the dressing can be removed and motion and therapy started to facilitate recovery. We typically restrict all sports activities until 6 weeks and ensure that the patient returns to activities when they have full motion and strength. Patients are seen at 6 months with a standing alignment film to detect any iatrogenic deformity from incomplete growth arrest that may occur. Once complete growth arrest is confirmed, they are followed with scanograms until growth cessation in order to document outcome and prevent possible overcorrection as a result of unexpected growth potential from the short leg. Should the leg length equalize prior to growth cessation, the family and patient may need to consider growth arrest of the previously short leg to ensure they remain equal. Thankfully, this is an extremely rare occurrence as most surgeons tend to time the surgery in order to undershoot correction.

Skeletal Shortening. Skeletal shortening has similar indications as epiphysiodesis (predicted discrepancy from 2 to 6 cm); it is offered to patients who do not meet the prerequisites for epiphysiodesis either because they are too old to gain correction through growth arrest or because their conditions are such that the extent of the discrepancy at maturity cannot be confidently predicted. In patients where a deformed bone needs to be osteotomized and is slightly longer (≈2 to 3 cm), one can simultaneously shorten the bone. Skeletal shortening can also be considered to gain partial correction in patients with larger discrepancy (>6 cm) because they are not candidates for limb lengthening as a result of clinical or psychological concerns. More extensive shortening of 7.5 cm in the femur and 5 cm in the tibia have been reported with no loss of function (161).

The overall advantage of skeletal shortening is that it can be done in the mature patient, when the discrepancy is certain and length correction (and occasionally simultaneous angular or rotational deformity correction) can be precisely obtained. The disadvantage of a major skeletal shortening is the potential for relative overlengthening of the muscle and weakness (162); as such it is unusual to perform more than 3 cm of shortening in the tibia and more than 5 cm in the femur. A good rule of thumb is to consider that a shortening of around 10% of the bone length is usually well tolerated. Furthermore, it is desirable to plan and execute an osteotomy level that leaves as much muscle at *in situ* length; femoral shortenings done proximally will have minimal effect on the quadriceps that originate distally to the osteotomy.

Skeletal shortening is usually performed in the femur; it is rarely done in the tibia except in cases in which the femur does not lend itself to shortening. Tibia shortenings have greater risk of neurovascular complications because of the proximity and the tethering of neurovascular structures. In addition, tibia osteotomies have a higher rate of delayed union, nonunion, and compartment syndrome. Should tibia shortening be performed, prophylactic anterior compartment fasciotomy is advisable to reduce the risk of compartment syndrome. Internal fixation is more difficult in the tibia; closed techniques cannot be used because the bone is subcutaneous, and the muscles of the leg are slower to recover strength than those of the thigh.

The early techniques involved making step cuts or other complex cuts in the diaphysis of the bone, using interfragmentary screws or intramedullary rods for fixation (163, 164). These techniques are only of historical interest, because better techniques with more secure fixation are now available (165). The two principal techniques in use today are diaphyseal shortening, open or closed, with intramedullary rod fixation and proximal shortening with plate fixation. Both approaches provide secure fixation and neither requires postoperative immobilization.

Prior investigators had pioneered a technique of closed femoral shortening with an intramedullary saw set which allows the procedure to be performed entirely within the medullary cavity and without direct approach to the shaft of the femur (166, 167). The femur diaphysis is cut at two levels; the intervening bone segment is fragmented, shortened, and stabilized with a locked intramedullary nail; advantages are a minimal approach and less soft-tissue dissection. The nail provides outstanding fixation and the patient may weight bear as tolerated. This procedure is limited to patients with bones that can accommodate the intramedullary saw and standard locked nails (usually 9 to 10 mm of medullary space is required). This technique is usually precluded in patients with open proximal femoral growth plates due to risk of growth arrest or avascular necrosis; the latter results from damage to the lateral ascending femoral circumflex vessels along the femoral neck as the saw and the nail may enter through the piriformis fossa. Other risks to this surgery include potential fat embolism syndrome as a result of closed femoral shaft reaming (168). Because the fragmented bone stays *in situ*, patients with closed femoral shortening may temporarily complain of the large callus of bone that develops from the incorporation of the comminuted bone fragments that remain.

Alternatively, an open approach can be used to remove a segment of bone which is stabilized by a plate or possibly an intramedullary nail. A significant advantage of proximal femoral shortening (at the level of the lesser trochanter) with plate fixation is that the resection level is proximal to most of the quadriceps origin. As such, the muscle and bone length relationship is preserved and patients recover strength quickly. The open approach allows exact measure of the amount of bone to be removed from the patient. This is an advantage because closed femoral shortening does not allow precise measurement of shortening. On the other hand, the open approach requires a large incision on the lateral thigh and requires elevation of the vastus lateralis muscle. The plate is placed on the lateral aspect of the femur, thus making it potentially prominent and painful. In these cases, a second later operation of moderate magnitude is needed to remove the plate.

Preoperatively, the amount of bone to be shortened is determined from scanogram measurements; anteroposterior and lateral radiographs of the femur are required to measure the diameter of the diaphysis and to assess for anatomic deformity that would preclude intramedullary fixation. Intraoperatively, the patient is placed supine with a bump that allows exposure to the buttock. A guide pin is placed percutaneously to the appropriate starting point as dictated by the proximal nail geometry [either the piriformis fossa (straight nail) or the trochanteric tip (trochanteric entry nail)]. The bony entry portal is enlarged and the femur is prepared for reaming. Acute respiratory distress syndrome has been reported during or following closed intramedullary shortening and may be the result of fat embolization caused by reaming an intact femur (168). In order to prevent this rare but significant complication, we routinely vent the femur with a large cannulated drill bit placed into the junction of the distal metaphysis and diaphysis. If the femur is of sufficient diameter (12 mm minimum), one can use newer reamers (RIA Reamers, Synthes, Paoli, Pennsylvania) that simultaneously ream, irrigate, and aspirate the femur. We further advise the anesthesia team of the risk so that they can adequately hydrate the patient and keep the PaO2 high during the reaming process. The femur is reamed 1.5 to 2 mm larger than the diameter of the planned nail and sufficiently large enough to accept the saw.

A proximal and distal k-wires are placed to detect inadvertent malrotation after osteotomy, and then the osteotomy levels are identified. We prefer to plan the levels in the proximal one-third of the femur in order to minimize the amount of quadriceps muscle that would be functionally lengthened. Alternatively for shorter lengthenings, cuts could be placed so that the shortening is from the isthmus of the femur where the internal diameter is least; this has the advantage of more cylindrical reaming and eventual easier cutting in the cylindrical diaphysis. The bone is cut by a special eccentric cam saw (OEC Intramedullary Saw, Warsaw IN) that is passed down the shaft and cuts through the cortex from within (Figs. 28-33 and 28-34). The size of the saw required is determined by the outside diameter of the bone; the distal cut is made first, and a second cut is then made more proximally at the precise location to give the desired amount of correction (Fig. 28-35). Occasionally, the cuts cannot be completed because the femur is not perfectly cylindrical and the saw will not fully cut a portion of the bone. In these cases, a ¼ in. osteotome can be placed percutaneously to complete the cuts (Fig. 28-36). Once the two cuts have been completed, the intervening cylindrical piece of bone is split into two sections using a special hook-shaped reverse-cutting osteotome, and the pieces are pushed aside (Fig. 28-37). The surgeon then passes a guide rod down the femoral shaft which facilitates the placement of the intramedullary rod. The previously placed k-wires ensure rotational integrity, and the femur is locked distally and the rod is back-slapped to

Text continued on page 1373

Technique of Closed Femoral Shortening (Figs. 28-33 to 28-37)

FIGURE 28-33. Technique of Closed Femoral Shortening. The intramedullary bone saw set is a required equipment in addition to all the equipment necessary for closed intramedullary femoral rod placement. The saw consists of two shafts: an inner one with a saw blade on the end and an outer one with a bushing set eccentrically on it just proximal to the saw blade. This bushing fits tightly inside the medullary canal. The size of the bushing on the saw defines the size of the saw, which ranges from 12 to 17 mm. In reality, the size of the bushing is 0.5 mm smaller than the stated size; therefore, it will fit into a canal reamed to the stated size. On the shaft opposite the saw blade is a T handle, an indexing scale marked 1 through 20, and a spring-loaded index scale locking nut. This T handle rotates the saw blade out from behind the bushing. On a threaded portion of the outer shaft is a measuring device that consists of a trochanteric rest distally and a locking nut proximally.

The intramedullary chisel is a sharp, thick hook on the end of a strong rod. This is hooked on the distal end of the piece of bone to be split and is driven out of the bone with the slotted hammer. Because the piece of bone to be split is a rigid ring, splitting one side of it results in the other side of it splitting (see Fig. 28-37).

FIGURE 28-34. The saw is now advanced down the medullary canal. The distal cut is made first. The measuring device is set to allow the saw to pass down the shaft to the distance that was determined on the preoperative radiographs. This can be checked on the image intensifier, although it may not be as precise. It is of critical importance that the trochanteric rest be held firmly against the tip of the greater trochanter at all times. Not only is this the reference point for all measurements, but it also ensures that the saw remains in the same cut in the bone.

To begin the osteotomy, the index scale locking nut is pulled back and the indexing scale is advanced one hole past the zero mark. The T handle is then used to turn the saw through one or two complete revolutions. The osteotomy proceeds by advancing the indexing scale one hole at a time. Each time after it is advanced, the saw is turned 360 degrees, cutting a small thickness of the bone. Each time the indexing scale is advanced, the saw protrudes slightly more. When the saw has been advanced to the number 20 on the indexing device, it has reached its maximal penetration. The indexing device is returned to the zero mark, and the saw is withdrawn.

Occasionally, after the anterior cortex has been cut through, the saw catches when it is rotated. It is important to make certain that the measuring device is held firmly against the tip of the trochanter. If this fails to correct the problem, the saw should be retracted three or four stops and then advanced one stop at a time. Care should be exercised in the procedure because it is possible to break the saw.

FIGURE 28-35. The saw is now pushed down the shaft until the trochanteric rest sits firmly against the tip of the trochanter, and the entire procedure of cutting the bone is repeated at the proximal osteotomy site. It is necessary to complete this osteotomy. This can be done by using the distal fragment of the femoral shaft as a lever to break this intercalary piece of bone off the proximal fragment. If this is not possible, either of the two methods used to complete the first osteotomy can be used.

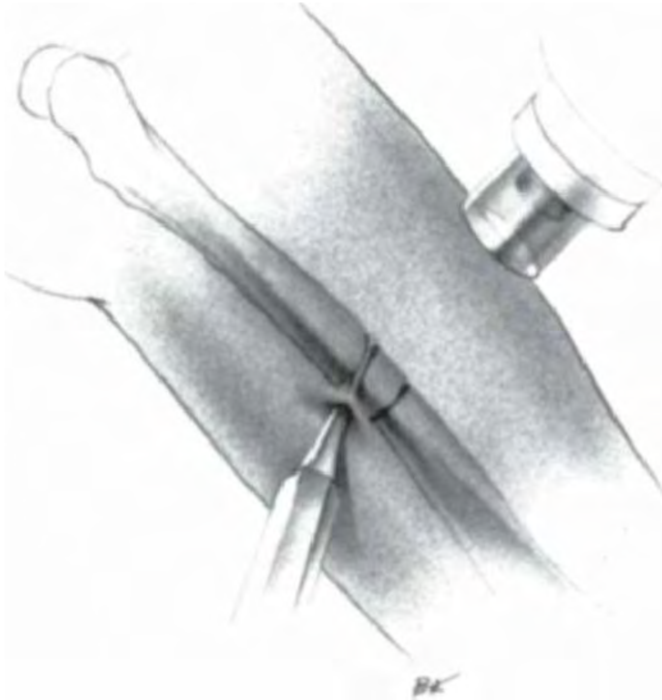

FIGURE 28-36. In some circumstances, the femur may not break. This is usually because the saw has failed to cut through the linea aspera. There are two ways to deal with this problem. The canal can be reamed larger and a larger saw inserted. If this is not possible, the osteotomy can be completed by inserting a 1/4-in. osteotome through a stab wound in the lateral thigh over the osteotomy site. The osteotome is passed into the osteotomy site under image intensifier guidance. The osteotome is then maneuvered posteriorly toward the linea aspera. With a firm grasp on the osteotome and with the hand held firmly against the thigh, the osteotome is struck sharply with a mallet to complete the osteotomy.

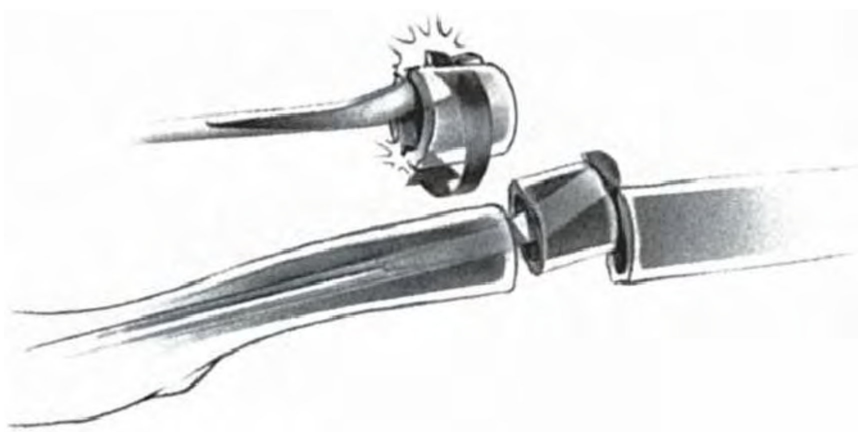

FIGURE 28-37. The intercalary fragment of femoral shaft created by the two osteotomies must now be split and moved out of the way to allow the femur to shorten. If the entire piece of bone or one large piece of bone is displaced, it may create a symptomatic enlargement that interferes with muscle movement.

The intramedullary chisel is inserted through the proximal and intercalary fragments, and the hook is directed posterolaterally to catch on a thick part of the intercalary fragment. It is usually not possible to split the linea aspera, and if the thin anterior cortex is split, one large and one small fragment will result. With the hook in the proper location, the slotted hammer is used to drive the hook out of the canal. The image intensifier is used to verify that the intercalary fragment is split and to avoid splitting the proximal femoral shaft.

After the fragment is split, the hook is used to displace the pieces to each side. Additional manipulation of the fragments can be accomplished by pushing on them with the distal fragment of the femoral shaft as it is brought into apposition with the proximal fragment. It must be possible to displace the split fragments and bring the distal and proximal shaft into apposition.

compress the osteotomy gap; final proximal locking maintains shortening and normal femoral rotation. In the past, complications resulted from inadequate fixation with unlocked nails. Less than rigid fixation can lead to loss of rotational control and opening of the shortening gap, two problems that are difficult to control without locking.

Although closed femoral shortening is an appealing technique, it requires familiarity with the instruments, is technically demanding, and has best results in experienced hands (Fig. 28-38). It leaves a small, cosmetically acceptable scar, and the later procedure to remove the rod is of lesser magnitude than that required to remove a blade plate. The major

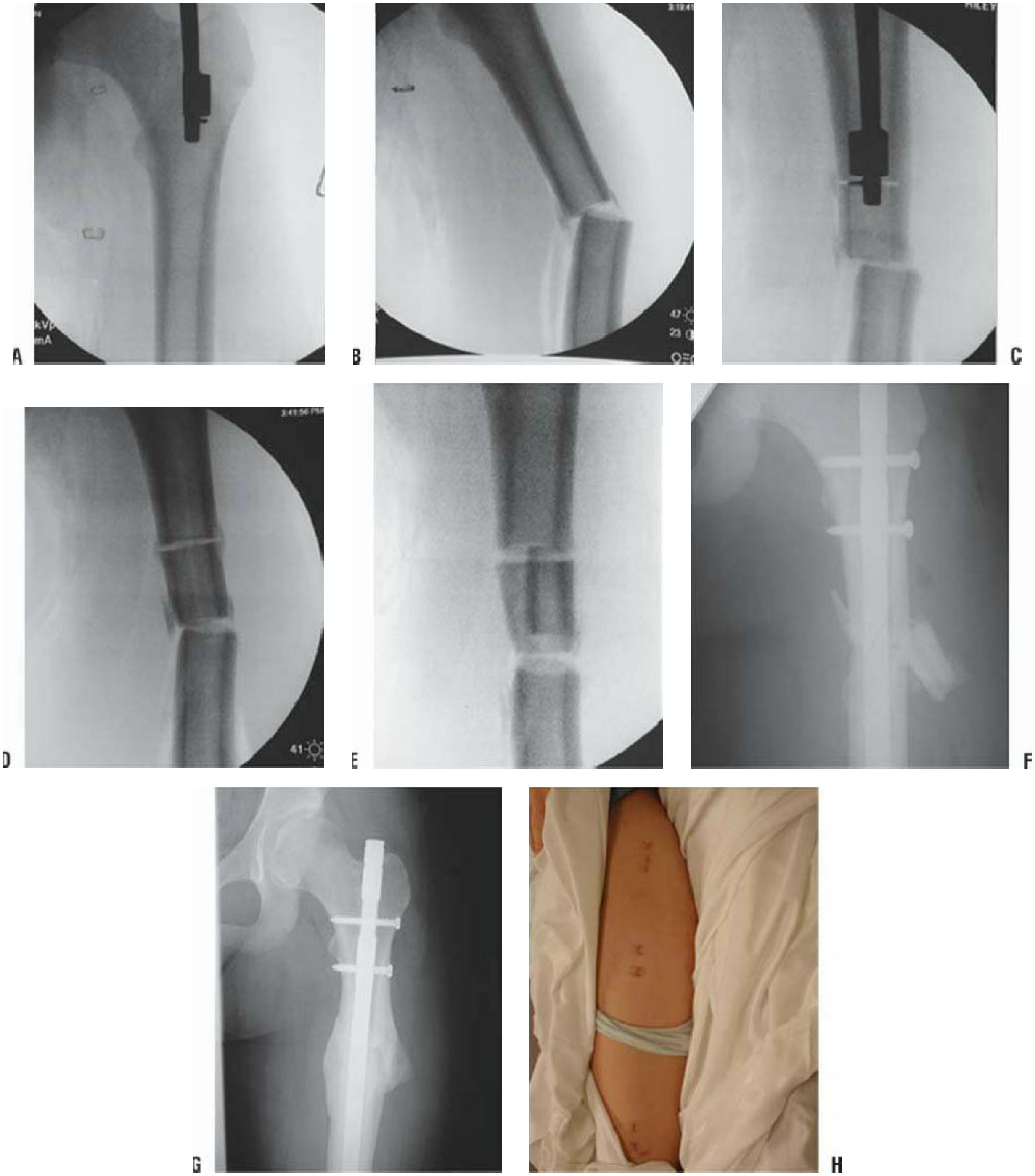

FIGURE 28-38. Technique of closed femoral shortening. **A:** After reaming the canal, the saw is placed into the shaft of the femur. **B:** The femur is osteotomized. **C:** The second cut is begun at a length equal to the distance needed to shorten. **D:** The second cut is completed leaving a cylinder of bone. **E:** The cylinder of bone has been split with the osteotomes, **F:** Femoral nail fixation is performed after the femur is shortened. **G:** Solid healing is present 4 to 6 months after surgery. **H:** Little clinical scarring is noted on her lateral thigh.

disadvantages are the technical complications and the risk of respiratory distress syndrome, as noted in the preceding text, and the significant quadriceps weakness that results. Patients with greater shortening require 6 to 12 months to regain normal knee control and function (169).

Proximal Shortening with Plate Fixation. In certain patients, acute shortening is best accomplished with an open approach that is stabilized with a variety of plate fixation devices. This method is appropriate for patients with concomitant proximal femoral deformity that needs to be simultaneously corrected. This method may be best for patients who may not be great candidates for intramedullary fixation. Such patients have open growth plates (risk of avascular necrosis), very narrow diameter (poor reaming and nail candidates), or with femoral shaft deformity (bowing) that will not easily accommodate the nail geometry. In addition, open osteotomy techniques will ensure exact shortening where fluoroscopy is not readily available or in patients with brittle bones (osteogenesis imperfecta) that may not be cleanly cut with an intramedullary saw. In addition, patients with questionable pulmonary history may be at increased risk of ARDS and should not have closed femoral shortening.

We prefer to plan our shortening in a proximal location when at all possible. This has the advantage of minimizing the degree of quadriceps functional lengthening and therefore minimizing postoperative weakness. Mid diaphyseal shortenings have the disadvantage of weakness as well as potentially decreased healing seen in nonmetaphyseal bone. Today, a host of different fixation options can be used and include devices with proximal fixation into the femoral neck (pediatric sized hip-screw devices or blade plates). Alternatively plates with locked screws (Synthes, Paoili, PA) into the femoral neck can be used. We prefer to use a 110- or 130-degree plates in order to stabilize the femur; these devices have a lower profile than hip-screw devices or 90-degree blade plates which are more suitable for varus osteotomy (varus plates are designed for medial displacement of the femoral shaft). With these implants, we can perform an osteotomy at the junction of the intertrochanteric–subtrochanteric level. This approach combines the advantages of metaphyseal healing without greatly affecting the Blix curve of the quadriceps function. On occasion, distal femoral deformity requires a more distal osteotomy to correct deformity (Fig. 28-39) in addition to shortening. We do not plan shortening >2 cm at this level for risk of long-standing quadriceps weakness.

A lateral approach to the proximal femur is made, the fascia lata is incised, and the vastus lateralis is detached from the vastus ridge and elevated posteriorly to maintain an anteriorly based flap of muscle. The blade plate chisel is placed into the proximal femur over a guide pin; a distally placed k-wire that is placed parallel with the chisel will ensure that rotation is preserved. The chisel is inserted by hammering

FIGURE 28-39. **A:** This 15-year-old girl with NOMID syndrome has a 4-cm discrepancy in length with genu varum from the distal femoral varus. She is a poor candidate for external fixation due to risks of infection and her severe immunodeficiency. **B:** Distal osteotomy is required to correct deformity, but proximal femoral shortening osteotomy is additionally chosen due to concerns for weakness from shortening distally. **C:** Six months postoperatively, the length is equalized and varus is corrected. **D:** Good knee extension is possible secondary to preservation of her *in situ* quadriceps length.

it a short distance, backing it out until loose and advancing further; then it is backed out and readvanced in this manner to the appropriate depth. Failing to advance the chisel in this cyclical way can lead to great difficulty in removing the chisel once the bone has been sectioned. After the chisel site has been appropriately prepared and the chisel seated, the osteotomy level is planned; the most proximal cut is placed at or above the level of the lesser trochanter. The femur is cut with an oscillating saw that is frequently cooled with saline to minimize bone necrosis. The distal level is measured and similarly cut and the intermediary bone segment is removed. The chisel is removed and the blade plate is placed proximally along the chisel path; the plate is then fixed to the distal shaft after simultaneously shortening the bone while maintaining *in situ* rotation. The bone that was removed can be ground up and used as bone graft at the osteotomy level prior to wound closure.

Growth Arrest (Physeal Bar) Excision. Partial growth arrests (bar or bridge) are a consequence of localized growth plate injury resulting from a multitude of etiologies including trauma, infection, or neoplasm. Growth arrests can vary in size and location and lead to variable combinations of angular deformity or limb-length discrepancy. For instance and in comparison to traumatic growth arrests, the bridge from infections tends to be less discrete, larger and more central, and can even consist of multiple small bridges. Diagnosis of growth arrest can be made by identifying deformity (angulation or shortening) and growth plate abnormalities such as localized growth plate narrowing and asymmetric growth arrest lines (Fig. 28-40). Advanced imaging is available to define the extent and location of the bridge and include computerized tomography (CT) and MRI scans. The latter is extremely sensitive and in cases where no obvious deformity (length discrepancy or angulation) has resulted, the surgeon would be wise to document irrefutable growth alteration before considering treatment. CT scanning is very effective for mapping out the location of the bony bar and calculating the size of the bridge in relation to the growth plate bridge.

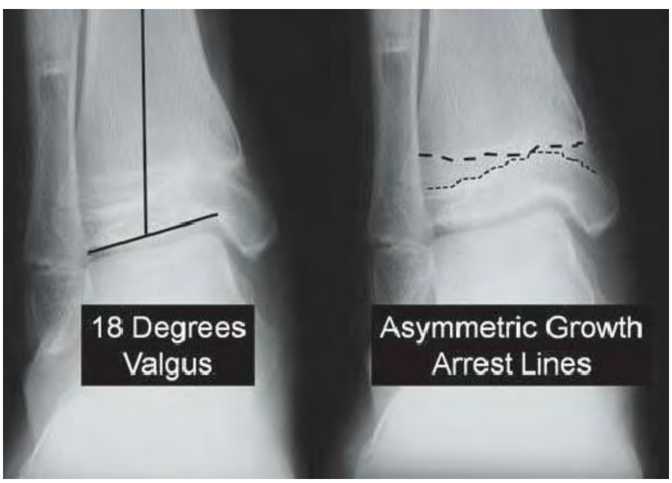

FIGURE 28-40. A 15-year-old boy has ankle deformity 4 years after an ankle fracture. Asymmetric growth arrest lines are present and are indicative of growth arrest.

Bridge resections are usually limited to those that involve <50% of the surface area of the growth plate and who would have at least a 2-cm discrepancy at maturity. In patients who would have less than a 2-cm discrepancy at maturity, completion of the growth arrest is preferable to the unpredictable results inherent to bridge excision. Attempted excision of bars >50% may be considered in selected cases where the predicted growth loss is >5 cm. In these young patients, an attempt at excision may be preferred to the difficult treatment of limb lengthening. Most authors do not consider an angular deformity to be a contraindication to attempted bridge excision; however, concomitant osteotomy should be considered if the deformity is >15 to 20 degrees.

Established bars can be considered peripheral or central, the latter are more challenging to remove and treat. In order to remove a central bar, a hole is made in the metaphysis and the bone is removed distally until the physeal scar is approached. Circumferential removal of bony bar at the level of the growth plate will continue until growth plate is visualized for 360 degrees; this may be facilitated with the use of an arthroscope camera (170) or a dental mirror. There is the danger that minor components of a central bridge can be left behind. The resected area is then filled with inert material such as fat or cranioplast cement (minimally exothermic bone cement).

Peripheral bars are exposed and the margins of the bar are defined with intraoperative imaging and from preoperative planning. Under loupe magnification and headlight illumination, the bone is burred away until growth plate is present at all areas. Once the bone is removed, inert material (fat or cranioplast) is placed into the defect and will prevent the bone from growing back between the epiphysis and the metaphysis. Placement of k-wires in the epiphysis and the metaphysis will facilitate monitoring of the growth plate function. With time and further development, it is anticipated that image guidance methods and intraoperative CT scanning could improve the ease and results of bridge excision.

Limb Reconstruction/Limb Lengthening. Patients with discrepancies >6 cm may only require *limb lengthening* as a component of their discrepancy. Patients with discrepancies greater than this may require multiple and different operations (including limb lengthening) and are better termed *limb reconstruction* candidates. Correction of projected discrepancies of 15 to 20 cm is possible with recent advances in technology and with improved understanding of mechanisms or strategies to avoid or reduce the morbidity associated with these endeavors (171–173). In order to correct deficiencies of this magnitude, the patient and their families have to be aware that a series of operations may be needed to optimize the limb *(limb optimization)* prior to limb lengthening and that increases in length are best obtained with a series of moderate lengthening attempts (10% to 20% in length at each limb-lengthening attempt). The family needs to recognize that each lengthening attempt will require at least 1 month of fixator time for every centimeter of length gained. Thus, an external fixator period of up to 6 months will be required and will precede many more months of required rehabilitation. Combining these facts with the ubiquitous associated problems or complications and additional surgeries to treat them, it is no wonder that many centers require psychosocial consultation prior to embarking on such rigorous treatment. Finally, it needs to be recognized that some residual length discrepancy may require epiphysiodesis or skeletal shortening of the nonaffected side in order to complete the treatment goal of limb equalization within 2 cm.

Within the scope of this chapter, it is impossible to outline the timing or scope of treatment that is needed for each individual patient. Nor is their uniformity among practitioners regarding the optimal time to begin the limb reconstruction process. Pros and cons exist at every stage of the process; some advocate delaying limb lengthening until patients are older children (10 to 12 years) so that they can comprehend the task ahead; others suggest that lengthening at an earlier age (5 to 7 years) may allow for more normal growth postlengthening and perhaps less ultimate discrepancy.

One reasonable philosophy is to optimize any osseous or joint deformity that would be an obstacle to lengthening prior to school age. For instance, concurrent hip dysplasia in congenital short femur should be corrected well before femoral lengthening in order to avoid hip dislocation. Similarly, correction of ankle valgus may be needed before lengthening for fibular hemimelia. It is also reasonable to plan lengthenings such that when completed an appropriate amount of residual growth on the contralateral leg is available to compensate for incomplete correction via epiphysiodesis of the long leg.

Patient Factors That Affect Limb Lengthening. Certain preexisiting conditions increase the risk of complications during limb lengthening. For instance, the complication rates in limb lengthening for congenital disorders is greater than limb-length discrepancy from shortening due to growth plate injury (infection, trauma, or neoplasia). Perhaps, the easiest lengthenings are in skeletal dysplasia patients who are lengthened for stature. The differences in complication rates from congenital deficiency, growth plate injury, or skeletal dysplasia are likely a result of differences within the adjacent soft tissues. In congenital conditions, the soft tissues are dysplastic and short; in patients with discrepancy from growth plate injury the soft tissues are normal; and in skeletal dysplasia the soft tissues are relatively longer than the bone to be lengthened (thus more amenable to lengthening).

Other comorbidities include irreparable joint instability (such as seen in neonatal septic arthritis) which can preclude lengthening. In addition, neurologic deficits within the leg suggest that the weak leg should be left a little short to facilitate floor clearance during swing phase. Weakness and the need for bracing must be assessed because leg-length discrepancy in patients with paralysis or weakness is usually best handled by undercorrection, leaving the weak leg short to facilitate swing-through. Another important consideration is in those patients where irradiation has been used to treat tumors (29). The absence of mesenchymal precursors and healthy osteoblasts precludes lengthening procedures through the affected bone (impossible for dead bone to form a lengthening regenerate) and the soft tissues that are resistant (fibrotic and inelastic) to lengthening procedures (174, 175). Other conditions to consider include the presence of spinal deformity and spinal imbalance; stiff suprapelvic obliquity which is compensated by discrepancy may lead to trunk imbalance once legs are equalized.

Prior to lengthening, a temporary shoe lift is useful for patients with complex deformities, because they take into account the combined effects of asymmetric feet, angular deformities, contractures, pelvic obliquity, and spinal balance. These lifts can be used to simulate the effect of length gained and prepare the patient for the end result. Finally, the concerns, compliance, and emotional state of the parents and patient must be taken into account. This aspect is particularly important when lengthening is being considered, because this is a long and difficult process requiring understanding and cooperation by all involved. Despite excellent education and preparation, parents and patients always underestimate the duration of lengthening and the later restriction of activities. The surgeon constantly must be aware that patients frequently express concerns about function when they are actually concerned about cosmetic effect, which may be less important when compared with the risks of surgery.

Limb Optimization. Surgery may be needed prior to or at the time of lengthening and include release of contractures, correction of angular deformity, and completion of partial growth arrests causing angular deformity. It is sometimes advisable to correct coexisting deformities before correcting leg-length discrepancy, because the correction of some deformities changes the treatment goal. For example, the correction of angular deformity of the limb usually increases the length of the leg.

Prior to distraction osteogenesis, certain conditions need to be maximized in order to prevent complications from limb lengthening. Most of these involve stabilization of joints above and below the bone to be lengthened. For instance in congenital short femur, hip dysplasia (shallow socket or femoral varus) can lead to hip dislocation during or after femoral lengthening. To improve hip stability, acetabular dysplasia is best corrected via pelvic osteotomy; proximal femoral varus and retroversion can also contribute to hip instability during lengthening and should also be corrected with osteotomy. Patients with congenitally short femur often have associated laxity of the cruciate ligaments (13) and hypoplasia of the lateral femoral condyle (Fig. 28-4). These deformities can predispose to patellofemoral dislocation or subluxation of the tibial plateau during lengthening. Similarly, lengthening of the tibia for fibular hemimelia can be contraindicated in the presence of an unstable.

Limb Lengthening. It is important that families understand that limb lengthening is not just a "procedure" (where the bone is cut and a lengthening device applied) but is more aptly considered a challenging "process," whereby the bone is lengthened after a relatively simple procedure. When considering limb lengthening, certain principles should be considered.

Primarily, limb lengthening is a procedure of last resort and is reserved for situations in which other methods of correction are available. Lengthening is usually not appropriate for patients requiring correction of <6 cm as methods of shortening are of lesser morbidity. Two exceptions to this principle exist; for instance, patients of short stature may not find shortening (skeletal shortening or epiphysiodesis) to be an acceptable alternative. Additionally, for those patients who require an osteotomy to correct deformity, a subsequent limb lengthening for shorter discrepancies can be subsequently performed through the corrective osteotomy site.

Secondarily, it is not uniform to expect to gain distances of >8 to 10 cm at a lengthening without having problems or complications from the lengthening. A goal for a lengthening for most patients is between 15% and 20% of the original bone length. For instance, when lengthening a 30 cm femur, it is reasonable to expect 4.5 to 6 cm of length gain before complications or pain limits further lengthening. Patients requiring large corrections may require staged lengthening of femur and tibia, repeated staged lengthenings of the same bone (176), or supplementary shortening procedures on the long leg. In patients who undergo serial lengthenings, complications rates appear to be similar in bones that are lengthened once or twice (177).

Third, the time required to lengthen a limb and rehabilitate it fully can be extensive; for instance families should expect 1 month of fixator time for every centimeter of length gained and does not include the time required for rehabilitation and to protect the limb from injury. Finally, when the predicted discrepancy approaches 15 to 20 cm, the morbidity, time, and hardship encountered outweigh the benefits of serial lengthening, and this method is abandoned in favor of amputation and prosthetic fitting.

Historical Perspective. Since Codivilla's report, there have been many techniques described for lengthening the leg. These have included step cuts (178), periosteal sleeves (179), onlay cortical grafts (180), slotted plates (181), intramedullary rods (182), and other internal and external devices for gradual controlled lengthening (183–186). The Anderson device, using large pins and an external fixator with threaded rods for lengthening, became widely used but confined the patient to bed. Techniques of instantaneous lengthening of the femur (165, 187–189) and tibia (190) have been reported but have not gained widespread support because the amount of length to be gained is limited. Simultaneous shortening of one femur and lengthening of the other with the excised bone segment from the other-side leg has been recommended (191, 192). Although not considered "limb lengthening," transiliac lengthening up to 2 to 3 cm can be performed in patients with infrapelvic asymmetry that requires concurrent hip stabilization (193–195).

A significant advancement in limb lengthening was made with the Wagner method of limb lengthening which gained popularity in the 1970s and early 1980s. With this method, the diaphysis of a bone was sectioned and soft tissues released as necessary. The bone was acutely lengthened 1 cm while an external fixator device held the bones apart. The Wagner external fixator was then used to lengthen the limb 1.5 mm per day until the desired length was obtained. At that point, the device was removed and a metal plate was fixed to the other side of the osteotomy, thus stabilizing the both ends. The distraction site would then be filled with autogenous bone graft at this operation or during a third operation. Once the bone was consolidated, the plate could then be removed. Although this method was effective, patients suffered complications ranging from device failure, deep infection, poor bone healing, pain, soft-tissue contractures, and even hypertension. Over time, the Wagner method and other methods of lengthening became obsolete with improved understanding of the biology of distraction osteogenesis (Ilizarov) also termed distraction callotasis (DeBastiani). Other advances in technology continue today with newer fixators, computer-guided application, and deformity correction and implantable lengthening devices.

Over the last two decades, the advancement in lengthening has been accompanied with descriptive terms for devices and the methodology of distraction osteogenesis. For instance, the Ilizarov method can be considered distraction osteogenesis which is the same process referred to by DeBastiani as distraction callotasis. The former used a circular fixator with transfixing wires (Ilizarov device) (Fig. 28-41) and the later utilized a monolateral device with half pin fixation (Orthofix device). When choosing between the devices that are available,

FIGURE 28-41. Ilizarov lengthening device. External fixation is accomplished by tensioned wires fixed to circumferential rings. (From Ilizarov G, Deviatov A. Surgical lengthening of the shin with simultaneous correction of deformities. *Ortop Travmatol Protez* 1969;30:32–37.)

the surgeon recognizes that the distinction between the two devices is no longer as sharp as it once was. For instance, ring fixators may be stabilized with half pins as well as transfixing wires; similarly monolateral devices may have rings applied that can utilize the transfixing wires. These latter constructs are more aptly considered hybrid constructs. For the purposes of this chapter, we choose to consider that the biology of distraction osteogenesis is the same whether it is stabilized with a ring or monolateral fixator.

Distraction Osteogenesis. There are three phases in distraction osteogenesis, and these can be characterized as the latency (waiting), distraction, and consolidation periods (Fig. 28-42). After the osteotomy, the bone ends are kept opposed for a period of time that can vary between 3 and 14 days. During this latency period, the osteotomy site passes through the inflammatory phase of fracture healing. The duration of this latency period varies according to factors such as patient age (older-longer latency) and location (diaphysis-longer latency) and further tends to be longer if the bone has had previous surgery, trauma, or is acutely angled. During the latency period, the patient and parents understand and become comfortable with the lengthening mechanism, pin site care, an exercise program to maintain mobility and to attain ambulation with weightbearing.

After the waiting period, the osteotomy then enters the reparative stage of fracture healing and the site is distracted 1 mm per day in differing increments. Most surgeons recommend ¼ mm lengthening steps done four times per day to optimize bone formation. This can be increased if exuberant callus is noted or conversely can be slowed if bone formation seems retarded. It is wise to obtain radiographs at the distraction site 1 week after the distraction is started to make sure the osteotomy is spreading an expected amount. Device malfunction or errant lengthening methodology will become apparent if the bone has not gapped an appropriate distance. During the lengthening period, radiographs of the joint above and below are needed to detect nascent hip or knee subluxation. In addition, radiographs are taken at intervals of 2 to 4 weeks to evaluate alignment and the quality of bone in the lengthening gap (i.e., the regenerate). Alternatively, ultrasonography can be used to measure the lengthening gap (196, 197). The rate of distraction can be modified according to clinical progress or radiologic appearance.

Maintaining motion is extremely important during the lengthening procedure. Patients and parents are instructed in a home exercise program, and the patient's range of motion should be monitored regularly. Stopping the lengthening should be considered if limitation of motion that is resistant to a more intensive motion program develops. Because most patients regain flexion in the first year after lengthening (198), it appears that maintaining knee extension is more important. Thus, many recommend discontinuing lengthening if an extension contracture of >30 degrees develops. Lengthening can be started again if the contracture resolves prior to consolidation of the regenerate.

Distraction is discontinued when the goal has been achieved or when an irresolvable complication, usually loss of motion, supervenes. During the consolidation period, patients are allowed to ambulate with full weightbearing, with aids if necessary. The device is retained until radiographs show consolidation which suggests adequate strength of the regenerate bone. Valid objective radiographic guidelines for what constitutes adequate consolidation and subsequent removal of the lengthening device have not been established. Findings such as corticalization with three cortices visible on two radiographs and the appearance of a medullary cavity are considered to be signs of adequate strength, but the decision to remove the device is still empiric. A good tip is to anticipate regenerative fracture and to leave pins in place for several days while the intervening fixator is removed. If a patient suffers regenerate failure, it is a simple process to reapply the device until fully healed. It is possible to protect the tibia externally with a cast or brace after device removal, allowing removal from the tibia earlier than from the femur. In addition, the mechanical and anatomic axes of the tibia are collinear, and the bone is subject mainly to compressive forces. This is not the case for the femur, in which the regenerate bone is not collinear with the mechanical axis and subject to bending loads.

Some investigators have recommended dual-energy x-ray absorptiometry (DEXA) as a means to assess strength within the regenerate prior to fixator removal (199, 200). In the consolidation period, dynamization of the device will subject the bone to cyclic longitudinal loading and stimulate bone formation. If the bone in the lengthening gap is slow to consolidate, there are several strategies available to increase bone formation or prevent fracture or deformation on fixator removal. Gonzalez et al. (201) compared bone quality in patients who underwent bilateral limb lengthening; those limbs that had a random placement of pulsed electromagnetic field stimulators had shortened the use of the external fixator. Ultrasound has also been used to improve bone formation after limb lengthening (197, 202). Using bisphosphonates in a small series of patients with regenerate insufficiency, Little et al. (203) felt these drugs were effective as a means to slow catabolism and allow the regenerate to mature. Mechanical methods to increase regenerate strength include shortening the device to put the bone under longitudinal compression, either leaving it somewhat shortened or re-lengthening it once the regenerate responds. Alternatively, some investigators have recommended early fixator removal, then intramedullary nailing in order to decrease fixator time and prevent fracture and callus deformation (204). Plate fixation during and after limb lengthening is another method to decrease fixator time and decrease the incidence of fracture: in contrast to intramedullary fixation, this method can be used in children with open growth plates (205, 206).

Patients should be restricted from contact sports until the bone is radiologically normal and the patient's joint range of motion has returned to near normal; fractures through the lengthening gap have been reported years later.

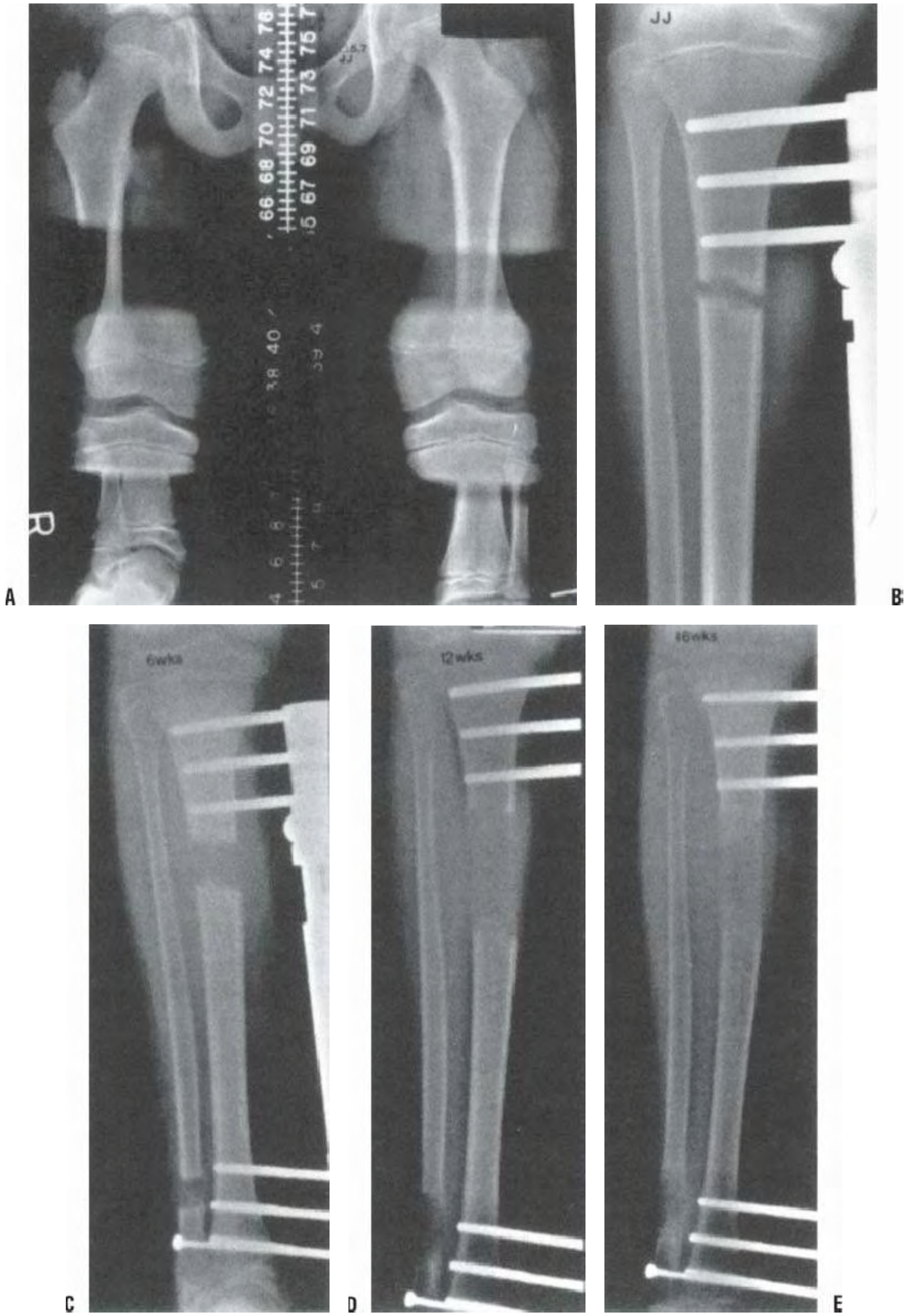

FIGURE 28-42. **A:** Scanogram of a 14-year-old boy with congenital shortening of the tibia and fibula. Note the ball-and-socket ankle joint; as in the normal ankle, the physeal plate of the fibula lies at the level of the plafond. The discrepancy is 5.7 cm, with an anticipated discrepancy at maturity of 6.3 cm. **B:** The osteotomy site 2 weeks after surgery and 1 week after lengthening has begun. **C–E:** Lengthening at 6, 12, and 16 weeks, respectively. The callus remains homogenous without gaps. The fixator remained in place for 21 weeks. The patient's subsequent course was uneventful, and he returned to full athletic activity 18 months after his initial surgery.

Limb Lengthening with External Fixation. Prior to lengthening, the surgeon will propose a lengthening device based upon multiple factors. For instance, half pins and monolateral frames are uniformly better tolerated than transfixing wires and ring fixation applied in the proximal thigh. On the other hand, ring fixators are also more versatile in that they lend themselves to the correction of complex deformities. They can control more than two segments (207), can extend across joints, and can be used to translate segments of bone in the treatment of congenital pseudarthrosis and acquired absences (208). Fixation is accomplished by tensioned through-and-through wires attached to complete or partial rings. Unwillingness to use through-and-through wires in the proximal femur has led to the development of half-pins, which are now gaining favor at all levels. In addition, each device has unique abilities to correct angular and rotational deformity in addition to the length discrepancy. Finally, some devices have companion computer programs which allow one to calculate the deformity and apply the fixator, and the computer can generate recommendations to guide the correction of length and deformity in all three planes.

Occasionally, a patient will have a shortened limb that will also require correction of a deformity; the surgeon has the choice of choosing acute deformity correction followed by gradual lengthening or gradual correction of both problems. There is good evidence to suggest that, if an external device is already in place for lengthening, either gradual or acute correction of coexisting deformity can achieve good results (209). Acute correction has the effect of simplifying the lengthening and widens the selection of devices, whereas gradual correction with the Ilizarov or another ring fixator allows the physician to monitor and modify the correction on an ongoing basis.

When planning the lengthening site, it is considered that greater amounts of bone formation can be expected when the osteotomy is performed in metaphyseal bone (as opposed to diaphyseal bone) that has not been traumatized from previous pathology (210). Additionally greater blood supply and therefore impact on healing is seen in periosteal rather than endosteal blood sources. Thus maintaining the integrity of the periosteum and using low-energy methods to cut the bone (osteotome versus power saw use) decreases thermal injury and improves bone formation (211, 212). Once the bone has been transected, the osteotomy site is not lengthened for a variable period in time that depends on a variety of host factors. On one end of the spectrum, the "latency period" can be as short as 5 days in young children with metaphyseal osteotomies. Yet in the other extreme, this period should be lengthened to 14 days when osteotomy is performed in young adults who undergo diaphyseal osteotomies with acute deformity correction through previously traumatized bone.

The rate of distraction can affect bone formation. Ilizarov (208) recommends distraction of 1 mm per day, a rate that exceeds the ability of the regenerating bone in the gap to effect union but is not so fast that it inhibits bone formation. The rate may have to be slowed if radiographs show inadequate regeneration and a widening lucency in the regenerating bone. Faster rates often induce ischemia and considerably slow the rate of osteogenesis, but some patients who show excellent regeneration radiologically can have their distraction rate increased. The rate of 1 mm per day also appears to be appropriate for the soft tissues that must grow in length in tandem with the bone (213). Increasing the frequency of lengthenings without changing the rate promotes faster consolidation experimentally and reduces the tension stress on the regenerating bone. Lengthening by 0.25 mm four times per day is better than lengthening by 1 mm one time per day, and it appears that gradual, continuous elongation, perhaps by a motorized device as suggested by Ilizarov, is ideal.

Although bone formation can be routinely expected, the distance a bone can be lengthened depends on soft-tissue factors too. In general, the total distance is limited to tightness of the surrounding muscles and tendons and associated decreases in joint motion and increased risk of joint subluxation. Devices that lend themselves to fixation of more than two segments of the same bone make it possible to lengthen a single bone both proximally and distally at the same time. Although this theoretically doubles the rate of bone elongation, the soft tissues do not easily double their elongation rate. In addition, ipsilateral lengthening of the tibia and femur has been shown to increase pressure in the articular cartilage (214); should this method be clinically indicated, the surgeon is well advised to consider fixation across the joint to ameliorate the pathology that could ensue to the articular surfaces and ligaments. Additionally, the surgeon should consider spanning an adjacent joint should that joint be at risk for subluxation or development of a muscle contracture. For example, patients with complete fibular hemimelia may need to have the ankle spanned to prevent ankle subluxation and equinus contracture. Lengthening should be stopped if an untoward joint complication arises, and consideration to muscle lengthenings of the adjacent muscles such as hip adductors, hamstrings, or gastrocsoleus complex can allow further length to be gained.

It is not known whether there is an upper limit to lengthening. Reports on the new techniques suggest that greater lengthening may be possible than was formerly thought. Carroll et al. (215) have shown that permanent changes occur in muscle and joint cartilage with tibial lengthening >11%, and Bell has shown effects on the adjacent joints in animal experiments (214). These effects may be related more to the rate than the magnitude of lengthening, and the degree to which they occur in human patients is uncertain.

Cyclic loading of the regenerate is thought to promote osteogenesis. The fixators with thin wires have the advantage of allowing dynamic loading of the lengthening gap throughout the period of fixation while they simultaneously control length. Their construct of thin wires and circumferential rings provides rigidity against bending in the sagittal and coronal planes but is not so rigid in the axial direction, allowing slight axial movement in response to applied loads. Monolateral devices can also be dynamized by the application of spring mechanisms. In general, the device is removed once there is formation of three cortices out of four cortices seen on anteroposterior and lateral radiographs of the regenerate. The wise surgeon will remove the device and allow the patient to go home for several days before the pins are removed, thus allowing reapplication of the device should a regenerate fracture occurs.

Distraction Epiphysiolysis. Distraction epiphysiolysis was pioneered by Ring and more recently reassessed by Monticelli, Spinelli, and others (216–222) and does not require an osteotomy. A theoretical advantage is the ability to correct growth deformity at the site of the pathology—the growth plate. It is achieved by applying a distraction force across the physis until it fractures. Lengthening can then be obtained by gradual distraction. This method has the disadvantages that the lysis is sudden, painful, and not well tolerated and that the physis can be injured, thereby compounding the leg-length inequality (223, 224). Despite theoretical advantages, the complication rate is high (225) and thus it should be reserved for children who are very near the end of growth to minimize the consequences of physeal damage.

Lengthening over an Intramedullary Rod. In the traditional application of any of the external lengthening devices, the device is responsible for both maintaining alignment and achieving distraction. Numerous unsightly scars result from percutaneous pins or wires used to achieve sufficient stability and because they must be left in place for a prolonged period until the bone is strong. In 1956, Bost and Larsen (182) introduced the concept of lengthening over an intramedullary rod. The rod serves to maintain alignment during both the distraction and the consolidation phases, and the external device serves only to achieve length (226). In this way, the number of percutaneous tracts can be reduced, the external device can be removed at the conclusion of lengthening, and the rate of regenerate fracture may be reduced (227). In addition to getting the external fixator off earlier, patients tend to gain their knee motion quicker in comparison to lengthenings with a fixator for the entire duration (228, 229). Lin described 2 cases of 15 cases of lengthening over a rod that required bone grafting (230), so the effect of rodding on osteogenesis remains unclear. The techniques can also be used in patients with shortened and deformed bones. In a series of patients with deformed and shortened femurs, Kocaoglu et al. (231) recently combined fixator-assisted deformity correction with intramedullary fixation and then lengthening over the nail.

Femoral lengthening over the rod has two disadvantages; one is that the femur must be lengthened along the anatomic axis of the leg, which will medialize the knee during long lengthenings. Second, because the proximal femoral physis is a barrier to blood to the femoral head, a real risk of proximal femoral avascular necrosis exists when the ascending retinacular vessels are injured after placing a rigid reamed nail down the piriformis fossa. Similarly, the presence of the tibial apophysis and risk of growth arrest leading to recurvatum makes tibial lengthening over a nail a challenge in the growing patient. Saraph et al. (232) used modified Ender nails (could be locked at both ends) as a method to avoid the problems of growth abnormalities from rigid tibial nails in children undergoing distraction. Regardless of the nail type, there has been hesitation in using this approach because of the fear of producing a serious intramedullary infection of the rod from infection of pin tracts (233). Early experience with this technique is varied. Some studies report a low infection rate and recommend the technique (229, 234–236) specifically in patients who need standard lengthening in patients with no history of past infection, open fracture bone deformity, or poor soft tissues (237). On the other hand, others are less enthusiastic, mainly because of a higher rate of infection (238–240) and in particular if a past history of an open fracture or infection is present (233).

This procedure involves reaming the medullary canal of the bone and placing a temporary nail in the shaft. Two proximal and two distal external fixator pins are placed in a manner that fixes the bone but does not bind to the nail. The nail is removed, an osteotomy can be performed percutaneously, and then a new nail (one that has not been scored during pin placement) is placed and locked proximally. The limb undergoes standard lengthening, and once the final length is obtained the distal end of the nail is locked and the fixator removed. The limb then can undergo rehabilitation and progressive weight-bearing. This technique has been found to decrease fixator time and patients have a more rapid return of knee motion. Problems with this method include the potential of deep infection of the implant from pin tract infection as well as risk of fat emboli syndrome from reaming an intact bone.

Intramedullary Lengthening Devices. The primary objection to limb lengthening is the use of external fixation with associated pin tract infections, scarring, painful tethering of the tissues by external fixation pins, poor cosmesis, and the extended consolidation period which requires the device to be present for many months. As such, there is tremendous interest in the development of devices or methodology that reduces or eliminates the use of external fixation (241, 242). One potential method would be the use of an intramedullary nail that would simultaneously lengthen and stabilize the bone while it heals. These devices have not found wide clinical applicability in children for several reasons. For instance, children usually heal faster than adults; thus the time for total external fixation is less. In addition, the pediatric patient has obviously smaller bones (which provides engineering difficulties for self-lengthening nails that are strong enough) with growth plates that would be at risk from placement of an intramedullary device. For instance, placement of a femoral intramedullary device through the piriformis fossa would put the femoral epiphysis at risk for avascular necrosis; similarly a tibial nail through the proximal tibia apophysis would raise the risk of recurvatum deformity. Despite the limitations above, there are occasional instances where these devices can be used in children with already compromised growth plates or in adolescents (241–247). Complications are different than those seen in standard distraction osteogenesis and include nail failure, poor bone formation as a result of stresses in the regenerate, need for exchange nailing, and overlengthening or underlengthening as a result of mechanical failure.

Complications from Lengthening. In reality, the success of limb lengthening is less dependent on the amount of length gained than on avoiding complications that may arise as a result of the treatment. It is challenging to evaluate the literature as each paper has a different definition of what constitutes a complication, yet all studies of leg lengthening have reported high complication rates (143, 248–260). Complications are related to the amount of lengthening (252, 261), and if the goal is a modest increase in length, the rate is reduced and the proportion of patients reaching their preoperative goals is increased (171). Complications include technical errors during the execution of the fixator placement and osteotomy. These include neurovascular injury (262), fracture, infection, and compartment syndrome. Complications from the lengthening process include sudden hypertension during lengthening (263–265), device malfunction, pin failure, pin tract infection, osteomyelitis, premature consolidation, poor bone formation, fracture after device removal, decreased growth of the limb (266, 267), malalignment during lengthening, pain, soft-tissue scaring, muscle tightness leading to joint stiffness contracture, or even dislocation (268, 269). The list of complications is long; so it is wise to fully explain them to the parents and patients; we tell patients to expect at least one complication and one additional treatment, operation, or procedure to treat these problems. Treatments range from antibiotic use to repeat osteotomy, sequestrum debridement, bone grafting, fixator adjustment for malalignment, soft-tissue releases for contracture or joint subluxation, and fixator modification to span affected joints.

Because of the frequency of concerns, questions, problems, and complications that surround the care of patients undergoing lengthening, it facilitates their management to form a lengthening team in a program with shared responsibilities. A nurse, physical therapist, social worker, and a skilled technician are important members of the team and should join the surgeon in preparing patients and families for the lengthening procedure. The team can respond to ongoing needs and offer support during the lengthening procedure. Families and patients never fully appreciate the depth and breadth of the hardship they will face, and they will require more support than is needed for most orthopaedic cases.

CONCLUSION

Providing the care of patients with leg-length discrepancy requires familiarity with the disease processes that cause discrepancy: an understanding of the natural history of discrepancy; knowledge in the techniques for patient assessment and the methods for prediction of future growth and discrepancy; familiarity with the factors important in the selection of treatment goals; and expertise in methods of treatment. Maintaining familiarity with up-to-date techniques and philosophies of surgical treatment is challenging, but the improvement in our capabilities should be an adequate reward.

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