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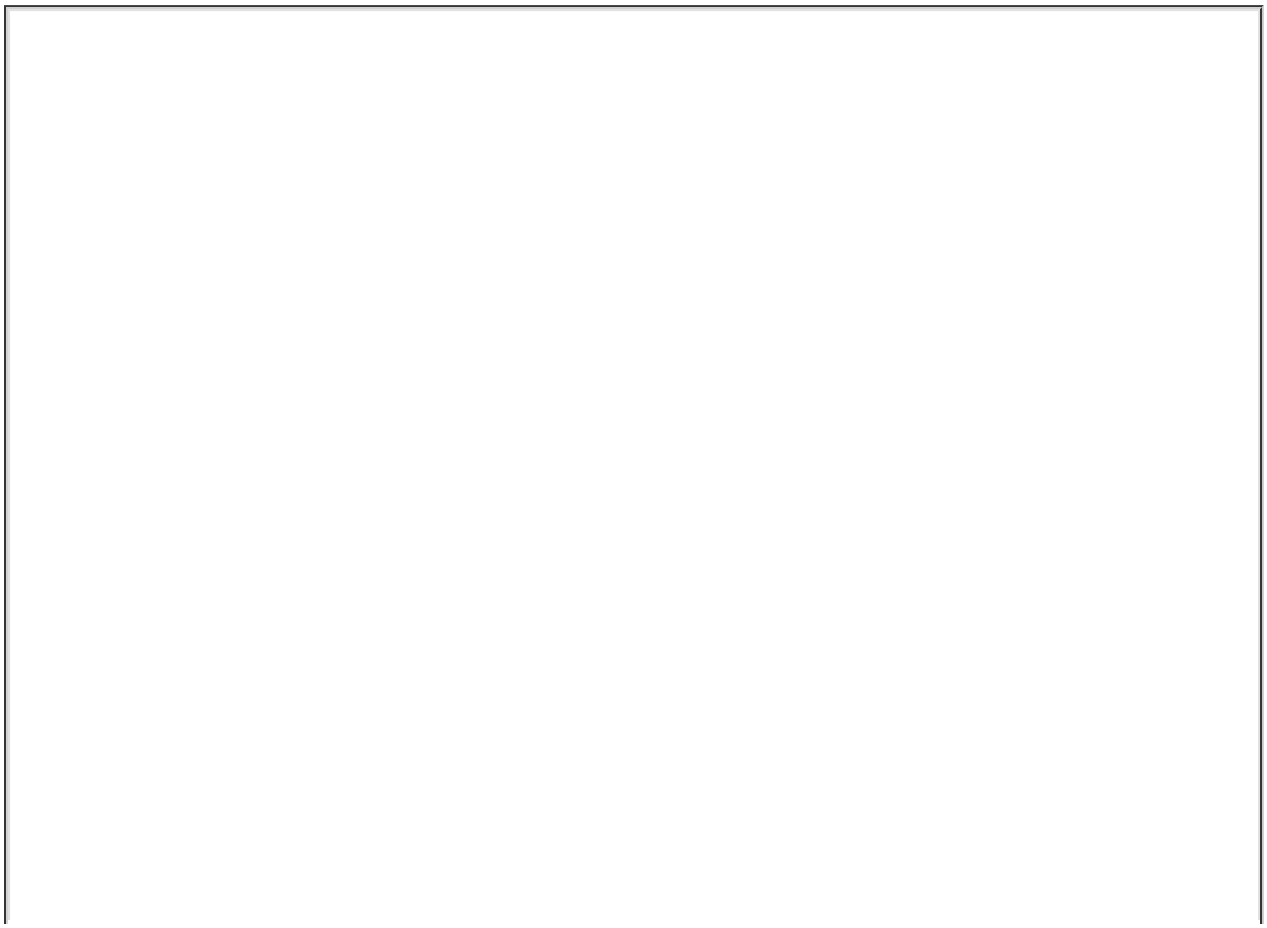
Lovell & Winter's Pediatric Orthopaedics, 5th Edition

ETIOLOGY

Part of "28 - LEG LENGTH DISCREPANCY"

Leg length discrepancy can be classified purely by etiology, but the concepts involved in understanding and treating patients suggest a more logical approach. Leg length discrepancy can result from two types of processes: those that change the length of the leg directly, and those that alter its growth. A fracture that heals with overriding is an obvious example of an effect on length with no effect on growth, and injury to the growth plate from osteomyelitis is an example of an alteration in growth rate with no immediate effect on length. These two effects determine whether a discrepancy is static or dynamic, and greatly influence the choice of treatment. The causes of leg length discrepancy can be classified according to their effect on length and growth, but the classification system breaks down because certain causes can affect different patients differently, and some affect both length and growth. A fracture, for example, can cause shortening in one patient and overgrowth in another, and a congenitally short femur can be thought of as being both short and retarded in its growth. Such a classification is shown in Table 28-2.



Classification	By Growth Retardation	By Growth Stimulation
I. Congenital	Congenital hemiatrophy with skeletal anomalies (e.g., fibular aplasia, femoral aplasia, coxa vara), dyschondroplasia (Ollier disease), dysplasia epiphysealis punctata, multiple exostoses, congenital dislocated hip, clubfoot	Partial giantism with vascular abnormalities (Klippel-Trenaunay, Parkes-Weber) Hemarthrosis due to hemophila
II. Infection	Epiphyseal plate destruction due to osteomyelitis (femur, tibia), tuberculosis (hip, knee joint, foot), septic arthritis	Diaphyseal osteomyelitis of femur or tibia, Brodie abscess Metaphyseal tuberculosis of femur or tibia (tumor albus) Septic arthritis Syphilis of femur or tibia Elephantiasis as a result of soft tissue infections Thrombosis of femoral or iliac veins
III. Paralysis	Poliomyelitis, other paralysis (spastic)	
IV. Tumors	Osteochondroma (solitary exostosis) Giant cell tumors Osteitis fibrosa cystica generalisata Neurofibromatosis (Recklinghausen)	Hemangioma, lymphangioma Giant cell tumors Osteitis fibrosa cystica generalisata Neurofibromatosis (Recklinghausen) Fibrous dysplasia (Jaffe-Lichtenstein)
V. Trauma	Damage of the epiphyseal plate (e.g., dislocation, operation) Diaphyseal fractures with marked overriding of fragments Severe burns	Diaphyseal and metaphyseal fractures of femur or tibia (osteosynthesis) Diaphyseal operations (e.g., stripping of periosteum, bone graft removal osteotomy)
VI. Mechanical	Immobilization of long duration by weight-relieving braces	Traumatic arteriovenous aneurysms
VII. Others	Legg-Calvé-Perthes disease Slipped upper femoral epiphysis Damage to femoral or tibial epiphyseal plates due to radiation therapy	

(From ref. 15, with permission.)

TABLE 28-2. CLASSIFICATION OF CAUSES OF LEG LENGTH DISCREPANCY

Some patients with asymmetry above or below the legs present as having leg length discrepancy and are treated as such, even though their legs may be of equal length. One example is the patient with the neglected, high-riding congenital dislocation of the hip (Fig. 28-11).

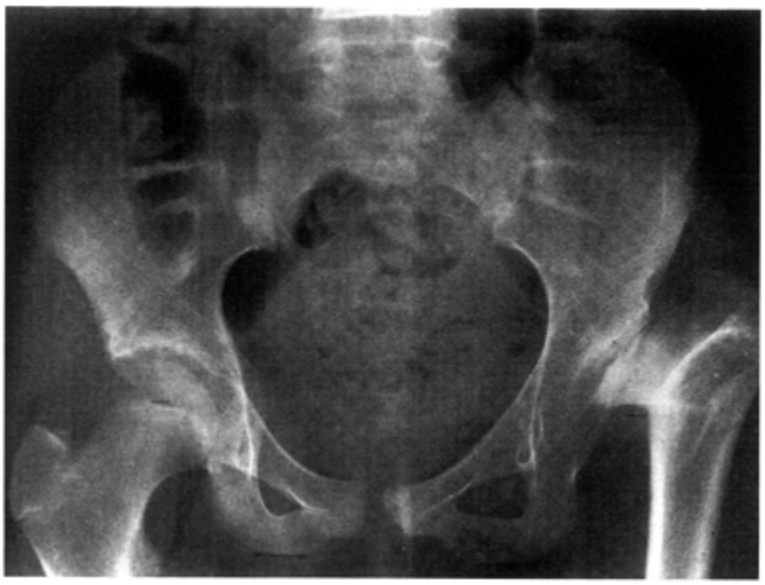


FIGURE 28-11. High-riding dislocation of hip. This patient has a functional leg length discrepancy that is greater than the actual discrepancy in the lengths of the legs, because of the adduction of the short leg.

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Interference with Length

By definition, the only processes that can acutely affect the length of the leg are fractures and dislocations. Whether or not a congenitally short bone has had a direct interference with its length is a moot point, because it occurred before birth, and it is the inhibition of growth that is the important factor. The terminal deletions and proximal focal femoral deficiency and its variants can be thought of as growth inhibition superimposed on a short limb.

Fractures can result in short bones either by overriding or by angular deformity. In the latter case, the shortening often disappears when the angulation is corrected (23). Sugi and Cole have shown that shortening of up to 10% of the femoral length can be accepted in the treatment of femoral fractures by early spica without causing significant discrepancy (24). Overgrowth frequently accompanies healing of fractures, and can spontaneously correct the shortening (25). Excessive length can result when excessive force is applied in traction.

Dislocations have a direct effect on length only if they are unreduced.

Inhibition of Growth

The growth of the physis can be slowed by three mechanisms. First, congenital short bones

grow more slowly than normal bones, as the result of abnormal programming of the genetic mechanism that determines growth rate. Second, the growth plate can be injured in such a way that part or all of it is no longer able to grow, and eventually gets converted to solid bone in the form of a physeal bridge or a prematurely closed plate. Any part of the plate that has retained its ability to grow cannot do so effectively, because of tethering by the fused part. Third, a change in the environment of the plate can influence its growth rate. Unusual vascular malformations can stimulate or inhibit growth (26,27). Children with paralysis usually have shortening of the more severely affected leg, presumably because the growth rate of the plate responds to the decreased compressive forces across it. The concept that pressure might change the direction of the growth of the plate is commonly known as the “Heuter-Volkmann law” (28,29), but the concept was first proposed by Delpech (30,31). He treated an angular deformity of the ankle with casting, to cause the distal tibial plate to change its direction of growth.

Congenital Shortening

When a patient is born with legs of unequal length that are otherwise normal, it is often impossible to know which leg is the abnormal one. Because the more severe cases clearly involve shortening, it is appropriate to think of these cases as hemiatrophy, rather than hemihypertrophy. Beals has stated that the two are separate and distinct clinical syndromes, partly because of associated anomalies (32). The dysplasia usually involves the entire limb, with some shortening of all components, and usually is accompanied by a diminution in girth. Each leg appears to be genetically programmed to be a different size (33).

Congenitally short bones frequently show qualitative as well as quantitative changes (34) (Fig. 28-12). The congenitally short femur also can show coxa vara, bowing, hypoplasia of the lateral condyle (Fig. 28-13), and external torsion (35). It can be associated with anterior cruciate insufficiency (36,37 and 38), a short or missing fibula (39,40), and absence of the lateral rays of the foot. Indeed, the congenital short femur is thought by some to be a variant of proximal focal femoral deficiency (41).

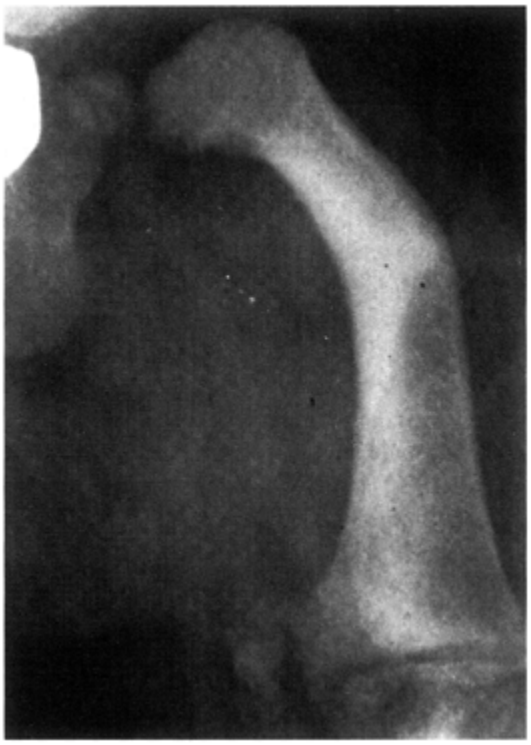

FIGURE 28-12. In proximal focal femoral deficiency, the leg length discrepancy is accompanied by qualitative changes, including coxa vara and bowing.

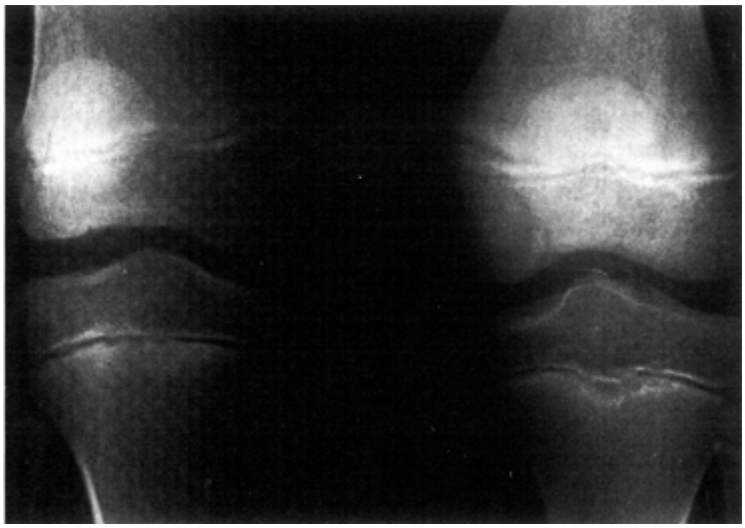

FIGURE 28-13. Hypoplasia of femoral condyle is frequently found in association with congenital shortening of the femur.

Congenitally bowed tibias are frequently accompanied by leg length discrepancy and hypoplastic feet (42,43 and 44). Askins and Ger (45) found that 24% of patients with congenital constriction bands have leg length discrepancy; and Garbarino and colleagues (46) reported short tibias in association with congenital diastasis of the inferior tibiofibular joint.

Trauma

Trauma that injures the physeal plate may slow its rate of growth either by direct injury to the cells responsible for growth or by formation of a bony bridge that tethers the epiphysis to the metaphysis. Salter and Harris provided a classification of fractures of the physeal plate that is useful in anticipating the effect of fractures on future growth (47). This classification is shown diagrammatically in Figure 28-14.

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Fractures can wander through all zones of the plate, but tend to pass through the zone of cell hypertrophy where the material is weakest and the amount of material is least. The material in that zone is cartilage that is weaker than bone, and because the cells there are large, the ratio of matrix volume to cell volume is low (Fig. 28-15). It is important to note that this part of the plate is the site of conversion of cartilage to bone, but is not primarily responsible for growth that occurs by virtue of cell multiplication and matrix production in the zones nearer the epiphysis.

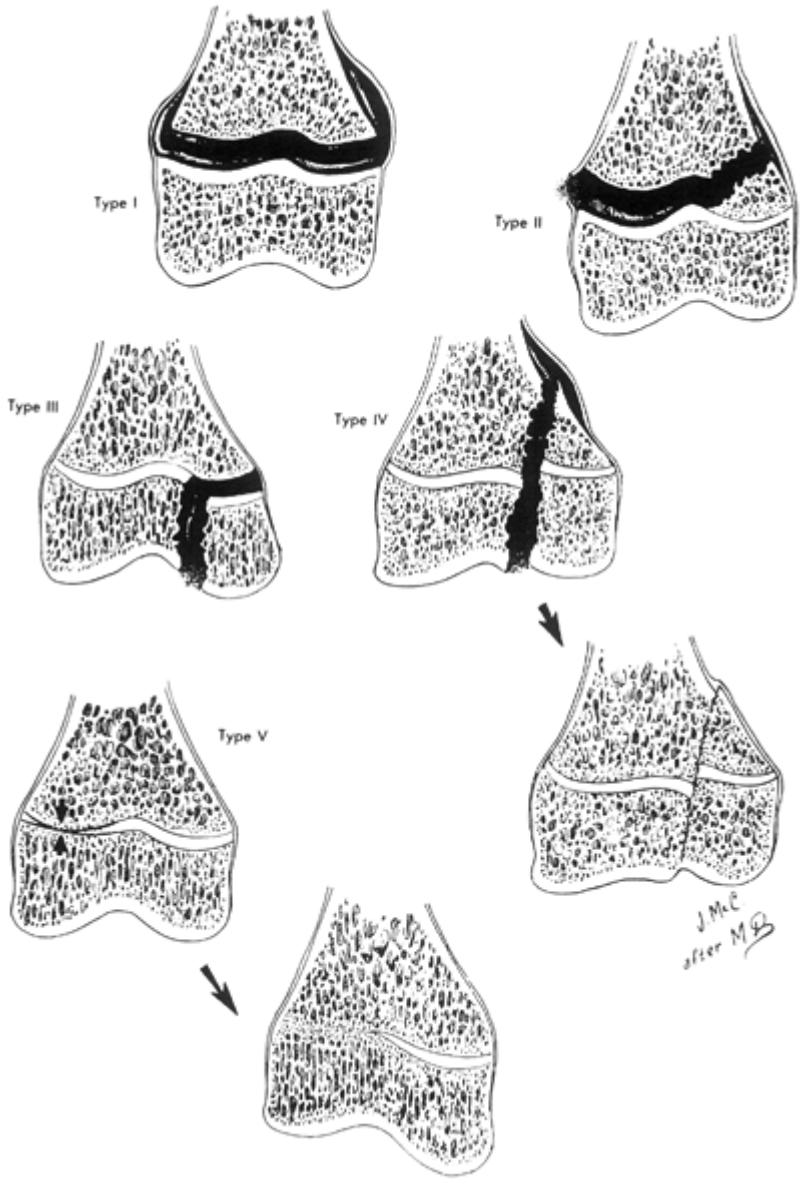

FIGURE 28-14. Salter-Harris classification of epiphyseal fractures. Fractures of types I and II do not cross the part of the growth plate responsible for growth, whereas those of types III and IV do. In the type IV fracture, approximation of epiphyseal bone to metaphyseal bone can result in formation of a bony bridge. (From ref. 46, with permission.)

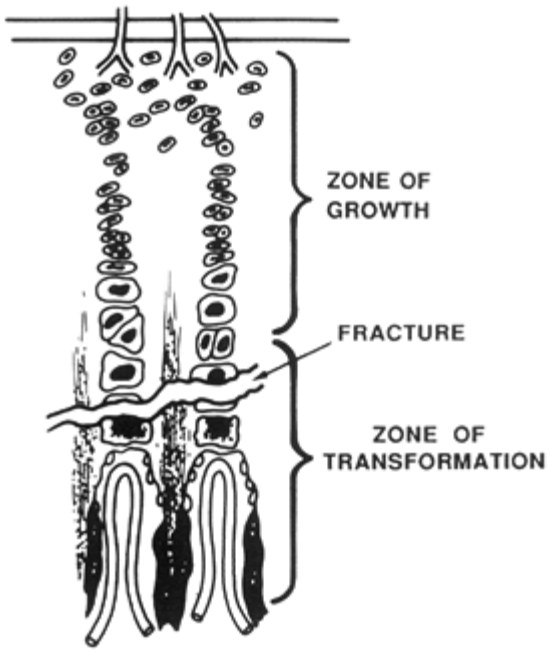

FIGURE 28-15. Location of fractures in the growth plate. Fractures tend to occur through the part of the growth plate where the matrix is least, although individual fractures may wander from zone to zone.

Because the type I and type II fractures do not pass through the growth zone, they are less likely than other fractures to interfere with growth. Both types, however, may be associated with a crush injury that injures the cells by compression. This mechanism may account for the higher-than-expected incidence of growth disturbance in type II fractures of the weight-bearing bones, such as the distal femur, where growth arrest is found in more than one-third of patients (48). Type III and IV fractures do, however, cross the growth zone, and therefore are more likely to result in growth arrest. The type IV fracture, in particular, can result in a bony bridge, when the fracture fragment displaces in the diaphyseal direction (Fig. 28-14). This is one reason why these fractures must be anatomically reduced. Type V fractures can occur in isolation, or can accompany any of the other types. They are insidious because they are not initially recognizable on radiographs, and they always demonstrate their presence by a disturbance of growth—either shortening or a combination of shortening and angulation—usually in the first year after the fracture. Although the fracture classifications provide guidelines about the likelihood of growth arrest, the orthopaedic surgeon must be wary of giving a definite prognosis for a given epiphyseal fracture, until enough time has elapsed to rule out a type V injury.

The bony bridge that causes a growth disturbance following physeal fractures usually is discrete and well defined, and lends itself to excision if small and peripheral. Bridge resections usually are limited to those that involve less than 50% of the plate, in patients who have at least 2 years of growth remaining. Even more extensive resections can be considered in very young children because, if successful, difficult treatment of severe leg

length discrepancy might be avoided. Resection of a bony bridge always should be considered if there is significant growth remaining, even if leg length discrepancy is already present. The angular deformity, which also may be present because of the bridge, can influence treatment of the discrepancy, because both deformities can be corrected at once.

Infection

Osteomyelitis adjacent to the plate can result in destruction of physeal cells and disturbance of growth if not treated early (49). The infection is usually hematogenous osteomyelitis of the metaphysis, but can be epiphyseal in infants, and follow or precede septic arthritis of the joint. The bony bridge that results from infection is more difficult to treat than that following trauma, because it is not so amenable to resection. The bridge tends to be larger, more central, and less discrete than that following trauma, and can even consist of multiple small bridges. It is difficult to define by radiograph, is usually more extensive than it appears, and is more difficult to define during resection. There is the danger that minor components of the bridge can be left behind, because the usual end point of resection, a continuous line of physis around the resection tunnel, can be achieved despite incomplete resection of all components of the bridge.

Infection tends to produce more serious leg length discrepancy problems than trauma, because it occurs so commonly in younger children with so much growth ahead of them. As with trauma, angular deformity and leg length discrepancy can coexist.

Paralysis

Inhibition of growth commonly accompanies weakness or paralysis of the leg, but the mechanism is not clear. It may be true that blood flow to the limb is reduced because of the reduced muscle mass, but this does not necessarily mean that flow to the plate also is reduced. Venous return results partly from muscle activity, and therefore, blood flow to the limb and perhaps to the plate could be reduced as a result of reduced muscle activity and decreased pumping effect. Alternately, abnormal vasomotor control, which is

part of the basic neurologic abnormality, could affect blood flow.

The effect of paralysis and reduced muscle activity can have a more direct effect on the growth rate. The Heuter-Volkmann law suggests that the growth rate of the physis responds to the compression forces across it, and frequently is invoked to explain how a spontaneous reorientation of the physis occurs in contributing to the remodeling of angular deformities in the immature child. This mechanism also can explain the decrease in the overall growth rate that occurs in children with muscle weakness.

The parents of children with cerebral palsy frequently are concerned about leg length discrepancy, and minor degrees of discrepancy can be seen in this condition. More often, however, the discrepancy is more apparent than real, and results from pelvic obliquity due to hip contractures or asymmetric posturing due to asymmetric spasticity. It is likely that serious discrepancies do not occur more often, because even dysfunctional spasticity can be effective, through the Heuter-Volkmann law, in stimulating the physis to grow.

Tumors

Leg length discrepancy can be related to tumors in several ways. The first involves destruction of the plate by direct tumor invasion behaving, in this instance, much like infection.

The second way involves damage to the plate by irradiation used to treat the tumor (50). Irradiation has a particularly harmful effect, because the osteocytes of neighboring bone also are killed, and the bone can take many years to become revascularized and repopulated with healthy osteocytes. The absence of healthy osteoblasts and precursors can complicate the treatment of the ensuing leg length discrepancy, by precluding lengthening procedures through the affected bone. Radiation damage to regional soft tissues also complicates lengthening procedures (51,52).

The third way that leg length discrepancy can be associated with tumors involves those that originate from the cartilage cells of the physis, thereby stealing growth potential from the plate. Examples of this are enchondromatosis and Ollier disease, which can produce growth inhibition of the affected bones (53), and osteochondromatosis, which frequently results in shortening of the ulna with a Madelung deformity. Although unicameral cysts usually do not result in significant leg length discrepancy, some disturbance of growth can result from aggressive attempts to remove the cyst wall, when the cyst is active and immediately adjacent to the plate. Unicameral cysts and fibrous dysplasia cause leg length discrepancy as the result of both growth inhibition and repeated fractures with minimal displacement that produce progressive varus.

Avascular Necrosis

Because the circulation of the physis is derived from the epiphyseal circulation, avascular necrosis of the epiphysis frequently involves the growth plate as well, and these patients can develop leg length discrepancy. This effect may be seen in Legg-Perthes disease, and following treatment of developmental dislocation of the hip. Peterson has reported a case in which a discrepancy resulted from a temporary but significant episode of vascular insufficiency during surgery in an infant (54). This effect, in the case of congenital dislocation of the hip, is maximized by the early age of onset and the years of future growth affected, but is moderated by the fact that the growth plate of the proximal femur contributes only about 15% of the growth of the limb. The likelihood of significant discrepancy has been correlated with the pattern of ischemic damage to the head, and increases with increasing involvement (55). That patients with Legg-Perthes disease do not usually develop significant deformity indicates that the vascular damage to the epiphysis does not always significantly affect the physis (56). Leg length discrepancy also has been reported as a complication of catheterization of the umbilical or femoral artery (57), presumably due to impairment of the arterial supply to the physis.

Stimulation of Growth

Certain conditions are known to stimulate growth, but although the mechanism is popularly thought to be increased circulation, only circumstantial evidence supports this theory. Attempts to stimulate growth in the treatment of leg length discrepancy have been made by

numerous means, including sympathectomy to increase blood flow, insertion of foreign materials next to the physis, stripping and elevation of the periosteum (58,59 and 60), surgical establishment of an arteriovenous fistula (61), short wave diathermy (62), and electrical stimulation (63). None of these methods has consistently produced sufficient growth stimulation to be clinically useful (64,65), but the fact that the arteriovenous fistula does produce stimulation at all supports the hypothesis that increased circulation can be a final common pathway for the conditions that stimulate growth.

Tumor

Vascular malformations, particularly when they involve large portions of the limb, produce growth stimulation that often involves all growth plates of the limb, and not just the ones in proximity or those of the involved bone. This stimulation is seen with hemangiomatosis and the Klippel-Trenaunay-Weber syndrome (66). Stimulation is also seen with certain nonvascular tumors, such as neurofibromatosis, fibrous dysplasia, and Wilms tumor, although an increase in circulation can be the final common pathway in these cases.

Inflammation

Overgrowth of the involved bone is a common feature of chronic osteomyelitis, presumably because of the increased blood flow to the limb as part of the inflammation. Infection therefore can both inhibit and stimulate growth. Overgrowth of the affected limb can be seen in pauciarticular juvenile rheumatoid arthritis (67), particularly in those cases with onset before the age of 3 years (68), and also has been reported in a hemophiliac with chronic knee synovitis (69).

Fracture

Overgrowth usually is seen following fractures of long bones in children, and also is believed to result from the increased blood flow to the limb that is part of the healing process (70). One particularly pernicious example of this effect is the overgrowth of the tibia and valgus deformity that can follow minimally displaced proximal metaphyseal fractures (71). The mechanism involves overgrowth of the medial side of the tibial growth plate (72), possibly as the result of tethering by the fibula or by release of the torn medial periosteum (73,74,75 and 76).

Overgrowth most commonly occurs following femoral fractures in young children (77,78). Some studies have reported that the stimulatory effect can last for years, but it is believed to occur principally during the healing and remodeling periods in the first 2 years after the fracture (79). The stimulation has been reported variously to be greatest in fractures in the proximal third of the femur, the middle third of the femur (80), and in those fractures with greater degrees of overriding (81) and can be accompanied by overgrowth of the fractured tibia on the same side. Conversely, Meals (82) found that the patient's age and the type and location of the fracture did not influence the extent of overgrowth, although, inexplicably, handedness did.