CHAPTER 21

Disorders of the Leg

Introduction

Deformities of the tibia and fibula are probably the most common and obvious abnormalities to affect the child's lower extremity. They can be congenital or acquired, physiologic or pathologic, but all, because of the real or apparent detrimental effect they have on gait and upright activity, draw immediate attention to themselves. Because the majority of lower leg "deformities" are in fact physiologic and resolve spontaneously, early recognition of the benign nature of such deformities is as important as the correct diagnosis of true pathologic conditions. This can reassure parents, avoid unnecessary treatment, and minimize excessive attention to a nonpathologic problem. The various conditions affecting the lower leg are discussed in relation to their anatomic occurrence, going from proximal to distal.

Genu Varum (Bowlegs)

Genu varum (bowlegs) is an extremely common pediatric deformity. Patients uniformly present for evaluation of the *deformity*, as they are rarely symptomatic in the age group (less than 2 years) when bowleg is most common. Determining whether the condition represents *physiologic* genu varum or a pathologic process, such as infantile tibia vara, is critical, because the prognosis and treatment differ profoundly.

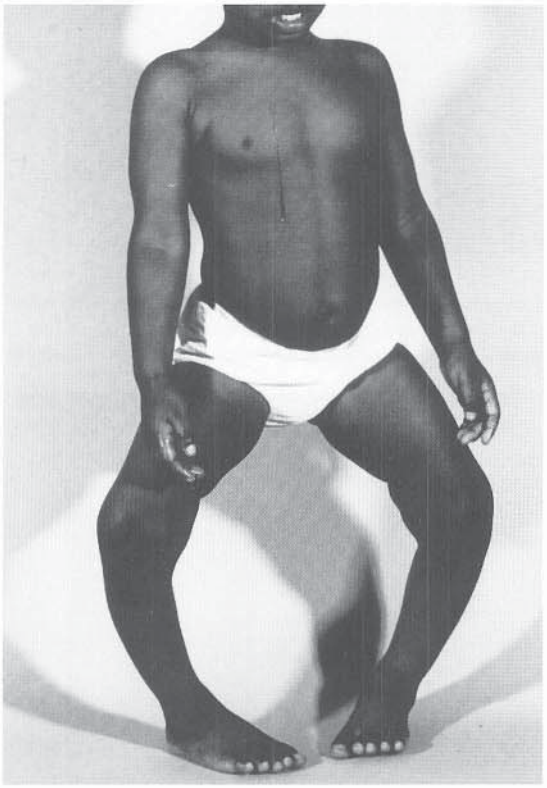
PHYSIOLOGIC GENU VARUM

Physiologic genu varum is a deformity with a tibiofemoral angle of at least 10 degrees of varus, a radiographically normal-appearing growth plate, and medial bowing of the proximal tibia and often of the distal femur.[54,63] The legs of most newborns typically are bowed, with 10 to 15 degrees of varus angulation. When the infant begins to stand and walk, the bowing may appear more prominent with weight-bearing, and often appears to involve both the tibia and the distal femur. A concomitant internal tibial torsion may exacerbate the deformity (Fig. 21–1A). Children with physiologic genu varum and internal tibial torsion typically come to medical attention after standing age (between 12 and 24 months), usually because of the parents' concern about the appearance of the legs, and these children have no other significant findings on clinical examination. In the typical presentation, radiographs are generally unnecessary to determine the physiologic nature of the deformity. Although radiographs at this time may show an apparent delay in ossification of the medial side of the distal femoral and proximal tibial epiphyses (Fig. 21–1B) or flaring of the medial distal femoral metaphysis, the physes have a normal appearance.

Clinical measurements of the knee angle and intercondylar distance in normal children show maximal varus at 6 to 12 months of age, neutral alignment by 18 to 24 months of age, maximal genu valgum at 4 years of age (8 degrees of valgus), and a gradual decrease in genu valgum to a mean of 6 degrees by 11 years of age.[68] The presence of genu varum after 2 years of age can be considered abnormal, but this "expected" pattern of change over time from genu varum to genu valgum is a generalized standard, and variations can be observed (Fig. 21–2).[85] A distinct subset of patients with more severe varus angulation at presentation, slower resolution to neutral alignment by age 3 to 4 years, and radiographic *femora vara* has been described.[27]

Spontaneous resolution of the varus to a neutral tibiofemoral alignment by 24 months of age and to an adult valgus alignment after 3 years of age is well documented (Fig. 21–3),[48,134] as is the variation just noted.[27] Patients can be formally followed to ensure that the varus resolves, but normally parents are reassured that the condition is physiologic and self-correcting and are advised that reevaluation is appropriate and that a radiograph may be indicated if the varus deformity persists beyond 24 months of age or progresses. Nonresolving, asymmetric deformity is the main indication for radiographs.

The differential diagnosis of persistent genu varum still includes physiologic genu varum, which remains the most common etiology, even in a deformity that is slow to resolve and appears to be pathologic (see Fig. 21–2). One must also



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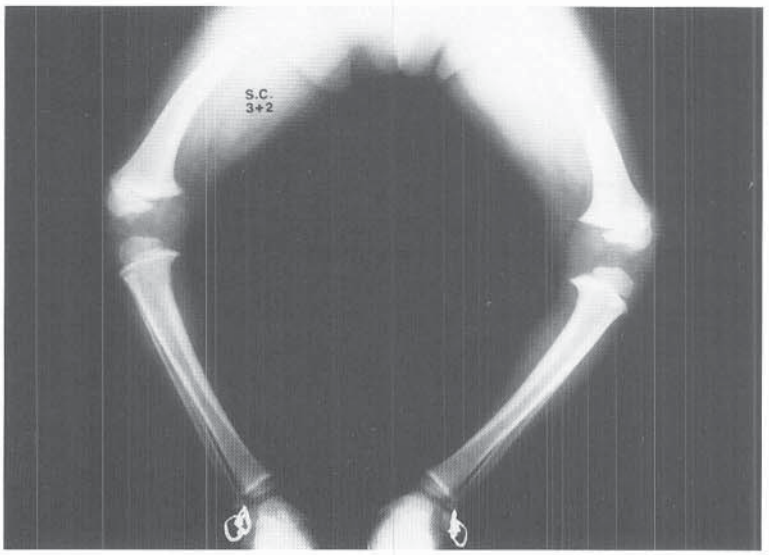

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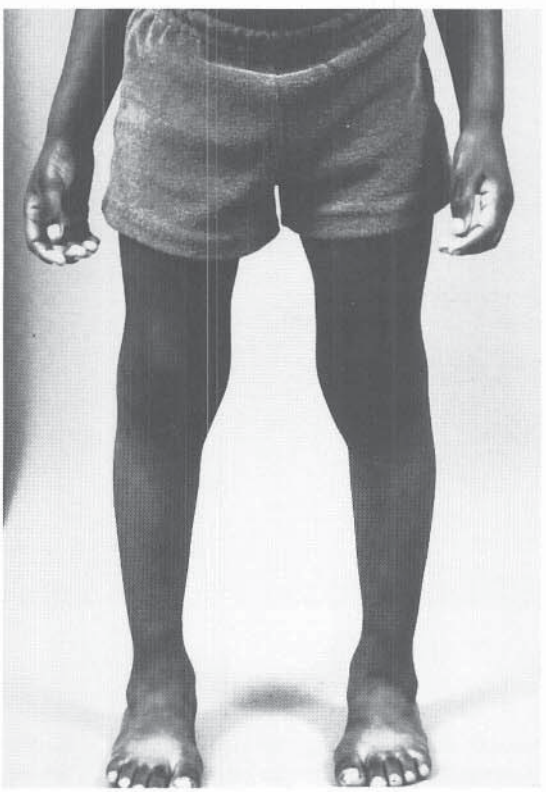

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FIGURE 21–2 An extreme case of physiologic varus in a 3-year-old child. **A,** Clinical appearance. **B,** Radiographic appearance. The medial epiphyseal ossification defects are more severe than in the child in Figure 21–1, but the physes are normal. **C,** Clinical appearance at age 5. The condition resolved spontaneously.

chondral ossification is disrupted, both in the medial aspect of the metaphysis and in the corresponding part of the physis. Varus deformity progresses as long as ossification is defective and growth continues laterally.[96] In later stages of the deformity, an actual bony bridge tethers medial growth, and the medial tibial plateau may appear to be deficient posteromedially. However, actual depression of the posteromedial tibial articular surface is probably not present, as the "deficiency" is one of unossified abnormal fibrocartilage whose delay in ossification is directly related to the underlying histopathology.[148] Ligamentous laxity on the lateral side of the knee frequently develops in the neglected or recurrent deformity.

Clinical Features. The typical child with infantile tibia vara appears similar to a child with physiologic genu varum, with two major differences. First, patients with tibia vara are often obese, exceeding the 95th percentile for weight.[14,40] Finite-element analysis of the knee has shown that a compressive force sufficient to retard physeal growth by the

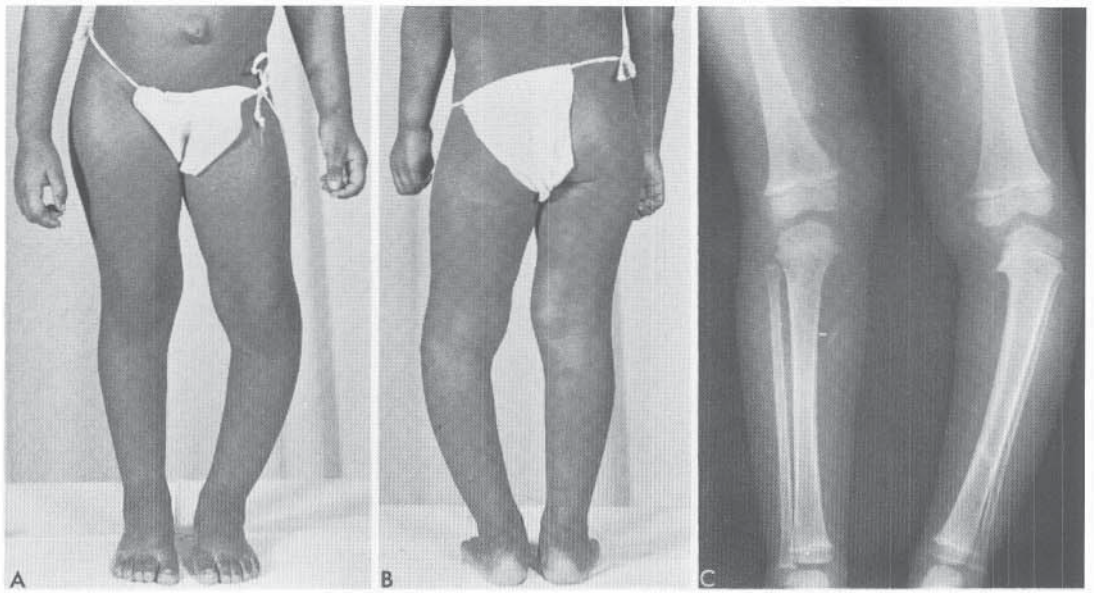


FIGURE 21–4 Blount's disease in a 5-year-old girl. **A** and **B,** Preoperative clinical appearance. Note the abrupt medial deviation of the tibia just below the knee. Lateral "thrust" to the knee during weightbearing exacerbates the "limp." **C,** Radiograph demonstrating abrupt angulation at the epiphyseal-metaphyseal junction and medial metaphyseal lucency and beaking, with apparent lateral subluxation of the proximal tibia.

infantile tibia vara but who have no physeal or metaphyseal changes, Levine and Drennan measured the tibial metaphyseal-diaphyseal angle (i.e., the angle created by the intersection of a line drawn through the transverse plane of the proximal tibial metaphysis and the line drawn perpendicular to the long axis of the tibial diaphysis) (Fig. 21–5). Twenty-nine of 30 patients whose metaphyseal-diaphyseal angle was greater than 11 degrees subsequently developed radiographic Blount lesions, while only three of 58 patients with an angulation of 11 degrees or less subsequently developed the changes.[102] However, subsequent studies measuring the metaphyseal-diaphyseal angle, the tibiofemoral angle, or the mechanical axis have not improved early detection of infantile tibia vara,[106,137] nor have the radiographic measurements been helpful in establishing the severity of disease once the condition is present. Any limb malrotation during radiographic examination can affect the measured metaphyseal-diaphyseal angle and the tibiofemoral angle.[74,153] Thus, although measurement of the metaphyseal-diaphyseal angle may have some prognostic accuracy,[56] it cannot be used by itself to definitively diagnose infantile tibia vara.[47,54,67] That diagnosis requires the unequivocal presence of the characteristic radiographic lesion in the proximal medial tibial metaphysis. If this radiographic finding is not present, the patient by definition has physiologic genu varum.[86] Although

TABLE 21–1 **Radiographic Features of Infantile Tibia Vara (Anteroposterior View)**

- Varus angulation at the epiphyseal-metaphyseal junction
- Widened and irregular physeal line medially
- Medially sloped and irregularly ossified epiphysis, sometimes triangular
- Prominent beaking of the medial metaphysis, with lucent cartilage islands within the beak
- Lateral subluxation of proximal tibia

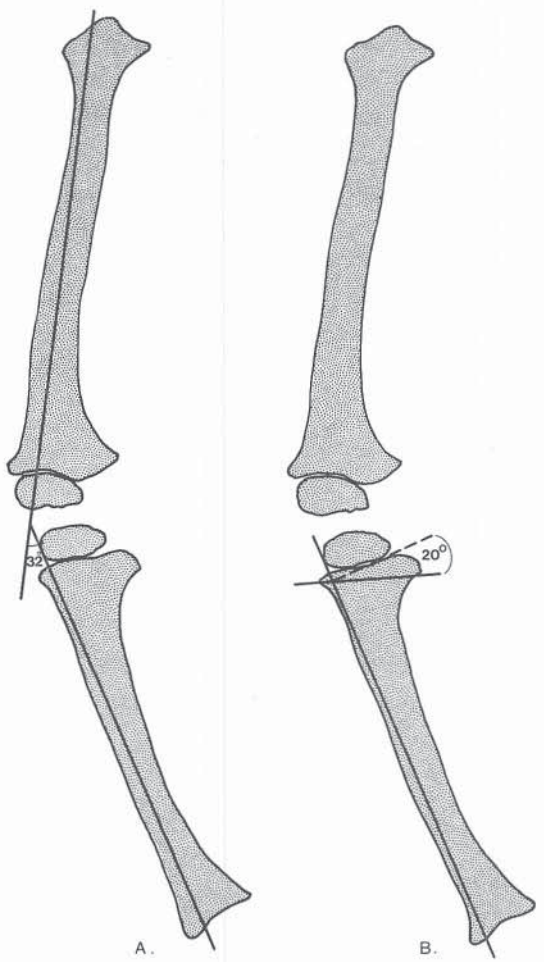


FIGURE 21–5 Radiographic measurement of angular deformity of the lower limb. **A,** Tibiofemoral angle. **B,** Proximal metaphyseal-diaphyseal angle.

vara, prognostic implications have gradually been derived from later studies.[78,106,137] In 1964, Langenskiöld and Riska reported that a simple osteotomy could cure the deformity in patients 8 years of age or younger.[99] In the few cases in which simple osteotomy failed, inadequate surgical correction was implicated. Radiographic stage progression of the deformity was perceived as a consequence of skeletal maturation[97,99] rather than as an indication of progressive inhibition of medial physeal growth and worsening of the condition.[105]

The premise that 8 years is the critical age up to which the condition is surgically curable has undoubtably resulted in a certain complacency in treating young children, particularly those with demonstrable stage progression. A number of investigators have reported difficulty applying the Langenskiöld classification to predict outcome in their own patients.[60,137,152,162] As early as 1963, Golding and McNeil-Smith reported advanced-stage infantile tibia vara appearing at an earlier age in black Jamaican children and requiring aggressive surgical treatment to correct the condition.[60]

The Langenskiöld classification regarding prognosis is inaccurate when applied to a predominantly nonwhite population in the United States.[106] Major inconsistencies are the following: (1) All of the stages can occur earlier than Langenskiöld described (as young as 17 months of age). (2) Stage II and III disease can progress to stage VI disease *despite* treatment (Fig. 21–8), whereas previously it was thought that surgery cured the disease in these patients.[96,99,137] (3) There is a marked tendency for deformity progression among black female children, and thus an even worse prognosis for these patients. (4) Predictably good results from a single tibial osteotomy are achieved only if the surgery is performed by 4 years of age, a notable departure from the previous guideline of 8 years.

It was the unanticipated difficulty in curing stage III and stage IV lesions by osteotomy alone that led to these conclusions, a finding confirmed by other investigators.[63,137] In the non-Scandinavian patient population, infantile tibia vara proved to have a more malignant course, and results of treatment were poorer than Langenskiöld's 1964 or 1981 guidelines suggested.[84,106] The poorer outcomes seemed due, at least in part, to the delay in surgical treatment of younger patients. However, the poorer outcomes may also be attributable simply to the different type and severity of disease that is encountered in the non-Caucasian population. Recent reports from Scandinavian centers[67] continue to attest to a relatively benign course for over 50 percent of their patients and to a condition that will spontaneously resolve, with the varus deformity correcting without treatment in one-third of patients—an experience diametrically opposite to the U.S. and Caribbean experience.[55,60,106,137]

From our experience, it appears that only stage I and II lesions can predictably have full restoration (i.e., cure) with a single osteotomy or bracing. Definitive treatment must be completed before age 4 years. Stage III lesions may be restored, while stage IV to VI lesions cannot be restored with a simple osteotomy and require complex reconstruction and physeal procedures, with a guarded outcome at best.

Treatment. Untreated true infantile tibia vara generally results in a progressive varus deformity, producing joint deformity and growth retardation which then can be corrected only with complex surgical procedures. Even when such surgery is performed, substantial articular disruption of both compartments of the knee may have already occurred.[78] Thus, once the radiographic diagnosis of infantile tibia vara is certain, the orthopaedist should recommend treatment immediately, since patients treated in the early stages of the disease have a better prognosis. There is no justification for simply observing a patient with an unequivocal diagnosis. Treatment choices and prognosis depend greatly on the age of the patient at the time of diagnosis, which should be the same age at which treatment is recommended.

ORTHOTICS. If the child is younger than 3 years of age and the lesion is no greater than Langenskiöld stage II, orthotic treatment is recommended, since 50 percent or more of these patients can be successfully treated with braces, especially if they have only unilateral involvement.[106,131,132,137,173] There may be an inclination to brace patients before a Blount's lesion is visible on radiographs, particularly when the metaphyseal-diaphyseal angle is indicative of varus progression.[102] Thus, when evaluating the reported good outcomes from brace treatment, one must realize that some patients probably had physiologic genu varum rather than true infantile tibia vara. Nevertheless, orthotic treatment appears to affect the natural history favorably.[131,132,173]

The type of orthosis prescribed and the length of time the orthosis is worn during a 24-hour period vary. Raney and associates[131] used a knee-ankle-foot orthosis (KAFO) that produced a valgus force by three-point pressure in 60 tibiae (38 patients), with lesions in 54 tibiae (90 percent) resolving without surgery. Significant risks for failure included ligamentous instability, patient weight above the 90th percentile, and late initiation of bracing. Of the 54 tibial lesions that resolved, 27 were treated by full-time orthotic use, 23 by nighttime use only, and four by daytime use. Three of the six tibiae requiring surgery had been treated with full-time orthotic use and three with nighttime-only use. Based on these findings, the authors conjectured that nighttime-only bracing might be as efficacious as full-time bracing, although they acknowledged that inherently one would expect daytime use (i.e., during weightbearing) to be the most important factor in successful orthotic treatment. On the other hand, Zionts and Shean[173] reported daytime, ambulatory bracing to be successful in altering the natural history of tibia vara in patients younger than 3 years with Langenskiöld stage I or II disease.

We have used conventional KAFOs, conventional hip-knee-ankle-foot orthoses (HKAFOs), and elastic KAFOs in the treatment of infantile Blount's disease.[132] Since 1987, the elastic Blount brace, a medial upright design that uses a wide elastic band just distal to the knee joint (Fig. 21–9),[155] has been used almost exclusively because of its ease of fabrication and smaller profile. With this orthosis, 65 percent of tibiae had successful outcomes at an average follow-up of 5.9 years.[132] However, when bilateral involvement was present, corrective osteotomies for one or both extremities eventually were required in 70 percent of patients, compared with only 6 percent of patients with unilateral involvement. All of the patients were instructed to use the brace during the day (i.e., during weightbearing). Depending on the patient's physician, some patients were encouraged to use the brace for 20 to 24 hours per day.

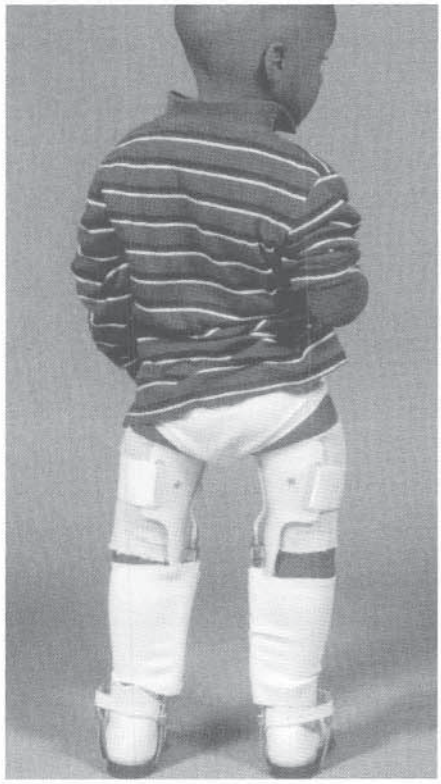


FIGURE 21–9 Elastic Blount's braces. Note the medial upright with droplock knee hinge that can be locked to increase the effectiveness of valgus pressure during weightbearing.

during nonweightbearing,[84,132] although we recognize that part-time wear has been successful as well.

Valgus correction should be increased by bending the medial upright every 2 months until standing radiographs show at least neutral mechanical axis has been achieved. Brace wear can then be gradually tapered off over a period of several months. To ensure permanent correction, the metaphyseal lesion should start resolving radiographically while the mechanical axis is being corrected and the lesion should have nearly resolved by the time the patient is no longer using the orthosis.

Brace therapy usually is not appropriate for children older than 3 years. A maximum trial of 1 year of orthotic treatment to correct the varus deformity is recommended; thus, if correction is not achieved within this time frame in a child younger than 3 years, the orthopaedist can still perform definitive osteotomy before the patient is 4 years old. Good results from this single surgical intervention by this age are seen in almost 90 percent of cases.[106] To begin orthotic treatment after age 3 means that the outcome of treatment will not be known until the patient is more than 4 years old, if brace therapy is given an adequate trial of 1 year. This risks delaying corrective osteotomy past the critical age of 4 years, if 1 year of orthotic treatment is not successful. Even a few months' delay in performing surgery by 4 years can result in failure to achieve permanent reversal of the inhibition of the proximal medial physis. For Langenskiöld stage III lesions, and especially for stage IV lesions, it is debatable whether mechanical realignment with a single osteotomy can ever reverse the physeal inhibition (see Fig. 21–8). The association between age greater than 5 years and biologic physeal arrest (stages IV and greater) cannot be overemphasized.

CORRECTIVE OSTEOTOMY. Surgical treatment in the early stages of the disease (Langenskiöld stage II) is crucial to achieve permanent and lasting correction and to avoid the sequelae of joint incongruity, limb shortening, and persistent angulation. Patients with stage I or II disease have a significantly lower incidence of repeat osteotomy requirement than those with stage III disease.[43,55,106] Surgical overcorrection of the mechanical axis to at least 5 degrees valgus, with lateral translation of the distal osteotomy fragment, achieved by 4 years of age is believed to be optimal.[43,106] Such overcorrection ensures that the supine correction attained at operation will be sufficient to translate the mechanical axis into the lateral compartment of the knee once the patient begins weightbearing. Overcorrection of the mechanical axis offsets the tendency of the knee to go back into varus as a result of sloping of the medial epiphyseal surface and relaxation of the lateral ligaments.

Although Schoenecker and associates reported that correction to within 5 degrees of neutral alignment would prove adequate,[137] most authors recommend physiologic valgus or overcorrection.[97,99,106] Based on the physeal inhibition phenomenon proposed by Cook and associates,[31] overcorrection to absolute valgus alignment is required to reverse the excessive compressive forces medially and to allow a Langenskiöld II or III physis not already irreversibly damaged to respond to such mechanical unloading.

The specific type of proximal tibial osteotomy (e.g., dome, closing or opening wedge) in young children is not important as long as the appropriate valgus and lateral translation are obtained. The osteotomy level should be just distal to the patellar tendon insertion, to avoid the proximal physis and its most distal extent. Internal tibial torsion should be addressed by external rotation of the distal fragment. Fibular osteotomy in the proximal one-third of the diaphysis should be performed routinely through a separate incision. Because long-leg casts often must be split during the postoperative period, some form of internal fixation (e.g., with Kirschner wires) is helpful in maintaining correction. Prophylactic fasciotomy of anterior, lateral, and posterior compartments is recommended, owing to the not insignificant incidence of compartment syndrome.[116,149] Following proximal tibial osteotomy, there may be subtle weakness of the extensor hallucis longus in spite of fasciotomy. This weakness, which is frequently overlooked, is likely due to partial peroneal nerve palsy.

Treatment of Langenskiöld Stage III Lesions. Stage III lesions *can* respond to corrective osteotomy alone in patients older than 4 years. However, the longer the delay in surgery after 4 years of age, the greater the risk of recurrence, even for stage III lesions (see Fig. 21–8). Thus, because of the worsening prognosis, neither observation nor orthotic treatment is recommended beyond this age.

Treatment of Langenskiöld Stage IV and V Lesions. Lesions greater than stage III cannot be definitively corrected by simple mechanical realignment because physiologic physeal arrest has already occurred by stage IV (Fig. 21–10). Even though no *bony* bridge can be visualized by tomographic methods in stage IV or V lesions, physeal damage has progressed to

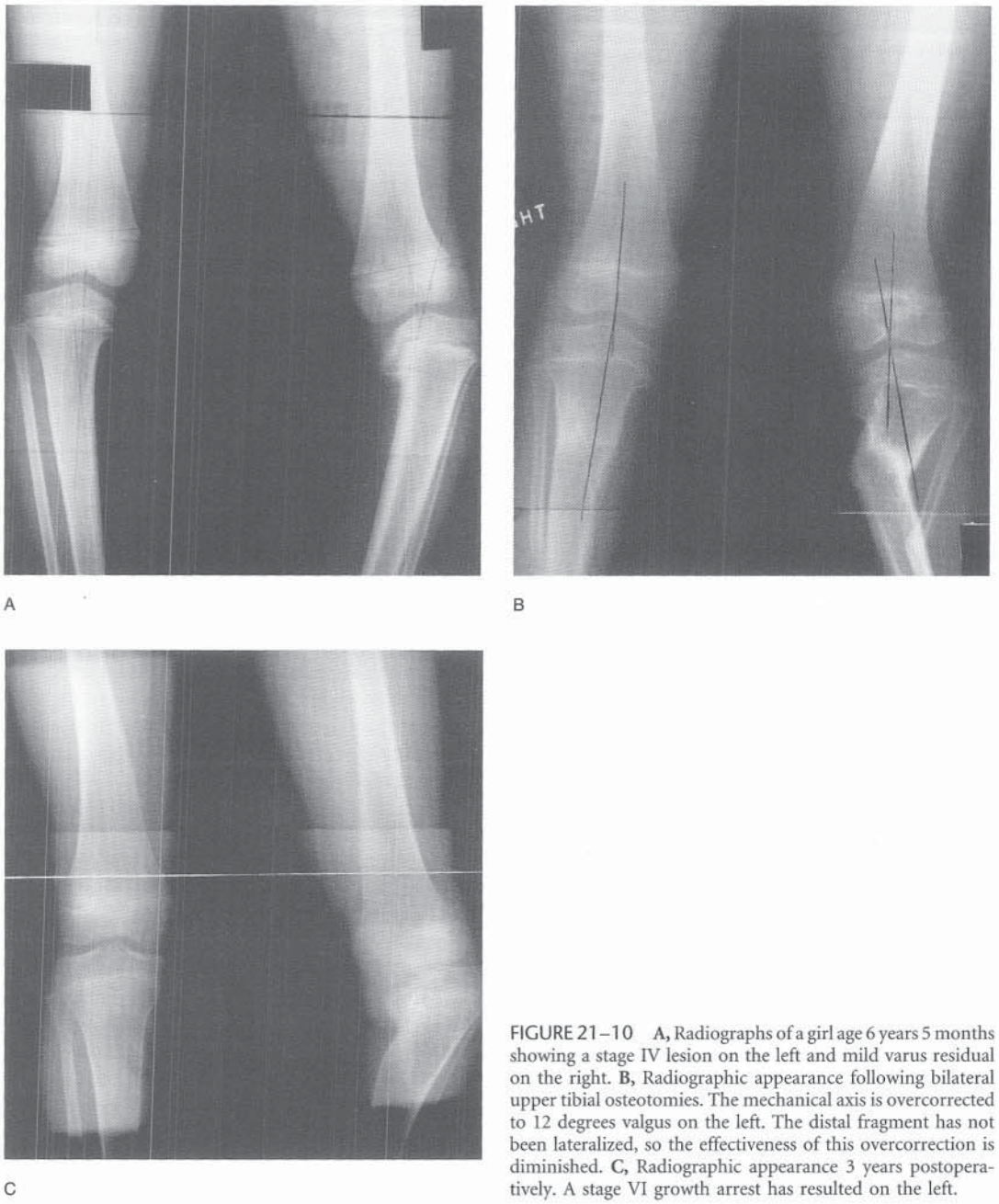


FIGURE 21–10 **A,** Radiographs of a girl age 6 years 5 months showing a stage IV lesion on the left and mild varus residual on the right. **B,** Radiographic appearance following bilateral upper tibial osteotomies. The mechanical axis is overcorrected to 12 degrees valgus on the left. The distal fragment has not been lateralized, so the effectiveness of this overcorrection is diminished. **C,** Radiographic appearance 3 years postoperatively. A stage VI growth arrest has resulted on the left.

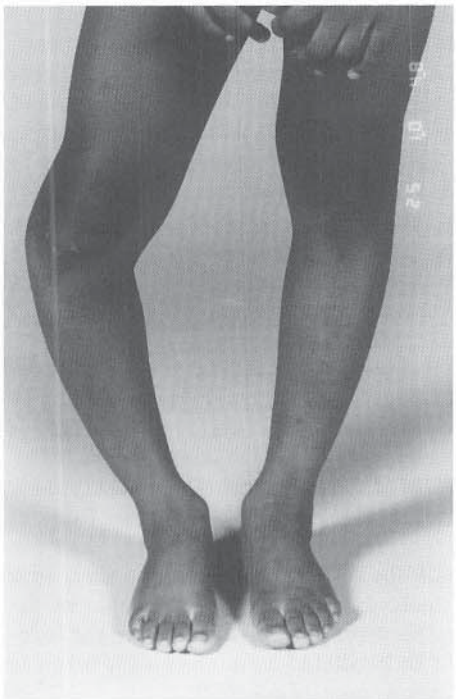
the point where stage IV and V lesions act effectively as medial physeal arrests.[64,136]

Because stage IV lesions can be seen in children as young as 6 years, their treatment presents a significant problem in management. Repeated osteotomies, required because of the predictable and certain recurrence of deformity, may help prevent intra-articular deformity but do not address limb shortening and, because of the repeated neurovascular risks, are clearly an unattractive approach.

Alternatively, some form of lateral epiphysiodesis—permanent or transient—will prevent recurrence by eliminating growth from the lateral side of the proximal tibial physis, but obviously results in significant limb length discrepancy in such a young patient. Total physeal closure at the time of osteotomy prevents recurrence but, by producing unacceptable shortening in young patients, predictably commits them to subsequent limb lengthening procedures.

Thus, treatment must be carefully individualized for stage IV or greater lesions in patients younger than 8 years. Realignment and medial physeal resection with placement of interposition material to prevent rebridging are warranted and reasonable (Fig. 21–11). Because stage IV and V lesions act in a similar manner—they inevitably progress to fully ossified physeal bridges (stage VI)—medial physeal resection with interposition is indicated, provided that 2 years of skeletal growth remains and the limb length discrepancy is not of a magnitude requiring lengthening.

Treatment of Langenskiöld Stage VI Lesions. The treatment of stage VI lesions with established bony bridges must also be individualized. Factors to be considered are patient age and



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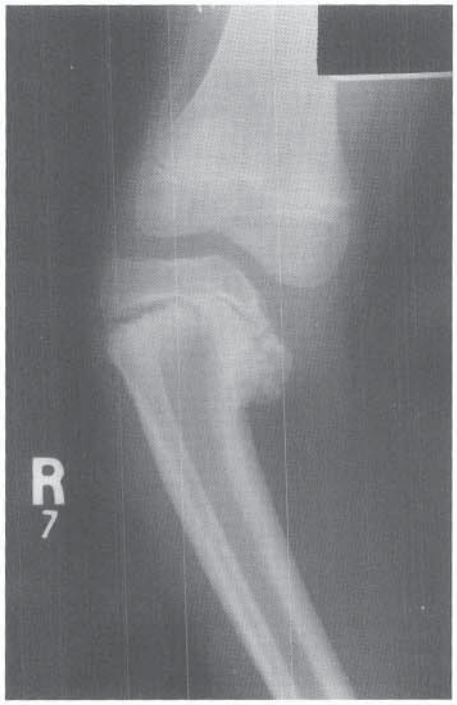

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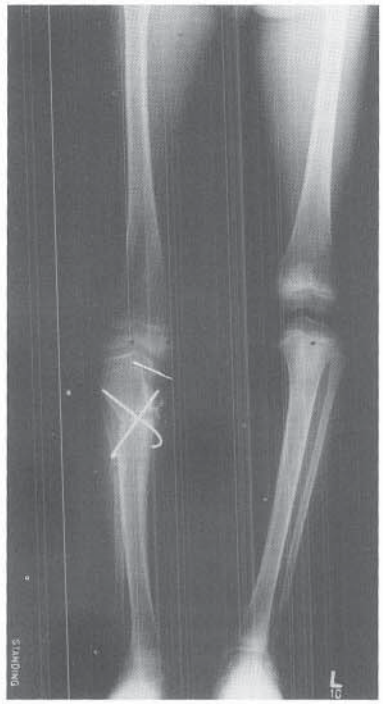

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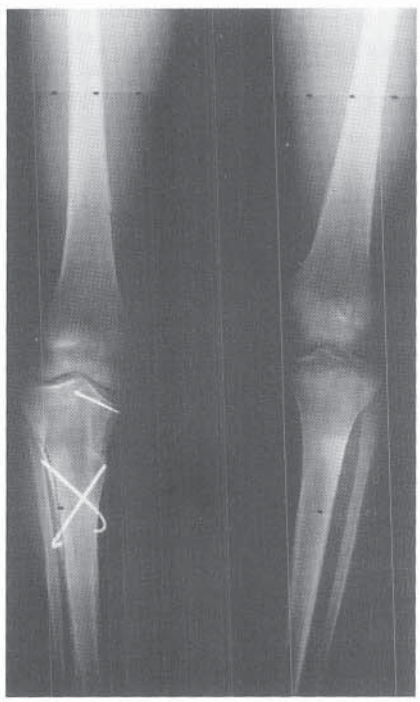

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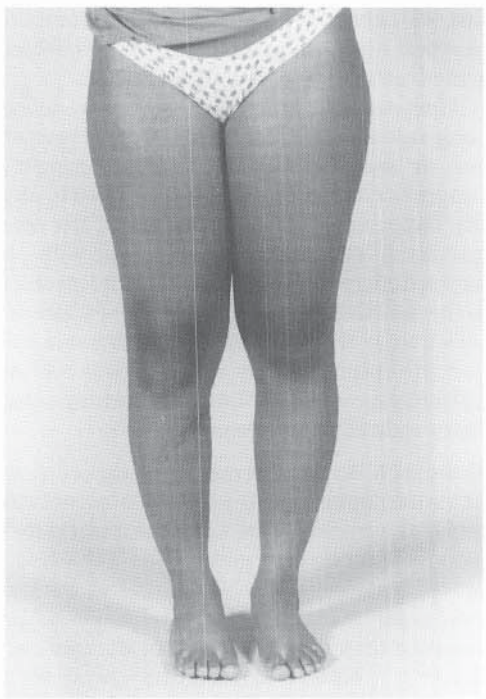

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FIGURE 21–11 **A** and **B,** Clinical and radiographic appearance in a 6-year-old girl with a neglected deformity (stage IV–V). Simple osteotomy in such a case is associated with a 100 percent incidence of recurrence. **C,** Radiographic appearance following physeal resection with a cranioplast interposition graft to prevent rebridging. Pins in the epiphysis anchor the interposed material. A corrective osteotomy to neutral mechanical axis was also performed. **D,** Radiographic appearance 6 years later. The deformity has remained corrected, with growth evident medially. The tibia on the right is 2 cm shorter than on the left. The patient is skeletally mature. **E,** Clinical appearance at age 13. The patient is asymptomatic.

amount of skeletal growth remaining, and the degree of deformity of the joint surface. If the patient has less than 2 years of growth remaining and a relatively normal joint surface, corrective osteotomy with complete physeal closure is a practical means of obtaining and maintaining correction. The osteotomy can performed through the physis so that the mechanical correction is placed as close to the joint as possible and permanent physeal closure occurs.

As previously mentioned, resection of the bony bridge with placement of interposition material is appropriate if 2 years of growth remains. If this procedure is successful, alignment (by osteotomy) and measurable, if not normal, longitudinal growth can be restored (Fig. 21–11).[10] Breaking of a physeal bridge by asymmetric physeal distraction has been described as an alternative approach to resection of the bony bridge in children near skeletal maturity.[25] If significant

incongruity and depression of the joint surface have been demonstrated on preoperative imaging, an intra-articular-directed osteotomy to elevate the medial tibial plateau can be performed to improve joint congruity.[64,97,136,140] Finally, in patients requiring limb length equalization with or without deformity correction, correction using external fixation and distraction osteogenesis is an effective and invaluable method for salvaging a potentially unsatisfactory extremity. Unfortunately, regardless of the method chosen, most patients with stage VI deformity have early degenerative disease because of the degree of intra-articular deformity present.[78]

COMPLICATIONS OF SURGERY. Complications of proximal tibial osteotomy in the growing child can be numerous. The osteotomy must be performed distal to the tibial tubercle to avoid growth arrest. Injury to the proximal tibial physis at the level of the tibial tubercle produces recurvatum of the proximal tibia, with resulting hyperextension instability of the knee. The optimal site of the osteotomy, distal to the tubercle, is near the level of the trifurcation of the popliteal artery. The anterior tibial artery, passing through the interosseous membrane and entering the anterior compartment, can be injured in as many as 29 percent of osteotomy procedures.[149] Prophylactic fasciotomy of all the compartments should be done during all osteotomy procedures, with appropriate postoperative neurovascular surveillance for the first 48 hours.[125,141] Other reported complications include peroneal nerve palsy, deep and superficial infections, iatrogenic fractures, and loss of correction.*

Unexpected recurrence of varus deformity in early Langenskiöld stage lesions may be due to inadequate correction or loss of correction, with subsequent progression of Langenskiöld stage and early asymmetric physeal closure. If this happens within 1 or 2 years of the osteotomy, repeat osteotomy and medial epiphysiolysis with placement of interposition material may correct the problem, particularly in the skeletally immature patient.[10] Failure of physeal bridge resection to at least maintain alignment usually is an indication for epiphysiodesis of the lateral half of the proximal tibial physis, with later limb lengthening equalization procedures utilized as necessary.

Summary. As can be readily discerned from the complex treatment options and numerous complications discussed under the treatment of Langenskiöld IV to VI lesions, and the risks involved in general for any osteotomy or repeat procedure, early treatment aimed at curing infantile tibia vara is far more attractive and likely to produce a good outcome than later treatment of the more advanced condition.[55,106,137] Early diagnosis and corrective treatment (orthotic or osteotomy) by 4 years of age is the most reliable way to avoid a poor outcome in both joint and leg function and cannot be overemphasized.

ADOLESCENT TIBIA VARA

The adolescent form of tibia vara, less common than the infantile form, is a distinctly different entity, owing to the later age at onset and, consequently, the more mature physeal plate and more ossified chondroepiphysis, which are more resistant to mechanical compression and disruption.

In the original description by Blount,[14] the adolescent form was defined as occurring after 6 years of age, and Langenskiöld[97,99] has used the term *adolescent* to describe partial premature closure due to trauma or infection in patients between 6 and 13 years of age. A more widely accepted definition, and the one used at our institution, describes increasing tibia vara after 10 years of age in a patient who usually is male, morbidly obese, and without a history of trauma, infection, or other physeal insult to explain the proximal medial tibial physeal inhibition.

Although some authors have subdivided patients with onset *after* 3 years of age into a juvenile group (ages 4 to 10 years) and an adolescent group (age 11 or older),[161] it can arguably be determined that in most cases, the "juvenile" onset merely represents the lack of definitive radiographic diagnosis prior to age 4, perhaps in the setting of a milder clinical deformity. Furthermore, the adolescent form has little in common with the so-called juvenile tibia vara, with the response to osteotomy usually more favorable in the adolescent form and recurrence more common in the juvenile form, as would be predicted for a child with infantile tibia vara treated after 4 years of age.[161,162]

Finally, some authors have used the term late-onset tibia vara to include both the juvenile and the adolescent types,[28] whereas others use late-onset tibia vara interchangeably with what is normally understood to be adolescent tibia vara.[70,71,73,107] For this discussion, the term *adolescent* will be used exclusively, and we will refer to those with onset after age 8 as described by Wenger and associates[168] and Beskin and associates.[12]

Etiology. Adolescent tibia vara is frequently observed in patients who, as younger children, had a mild degree of physiologic genu varum that never completely resolved to neutral alignment or physiologic valgus.[168] Concurrent with the adolescent growth spurt in children who are significantly obese, a gradual varus of the proximal tibia develops, owing to growth suppression from mechanical causes.[28,162] Although such occult varus can be confirmed by the history in many cases (family photographs are often available), not all investigators have been able to determine either that a mild varus preexisted[12] or that it was required in order for adolescent tibia vara to develop.[35,71] Trauma and infection, sometimes described as etiologic factors and known to produce physeal arrest,[108] are not considered factors in the development of adolescent tibia vara (which technically is idiopathic in nature) unless one wishes to define chronic growth suppression due to obesity as trauma.

Histopathologically, biopsy specimens of the medial physis show evidence of injury, with fissuring and clefts in the physis, fibrovascular and cartilaginous repair tissue at the physeal-metaphyseal junction, and disorganization and sequestered islands of hypertrophic chondrocytes.[28,168] Although these findings cannot be considered pathognomonic of repetitive trauma, in the absence of a history of significant trauma or infection they are consistent with microscopic damage secondary to mechanical compression according to the Hueter-Volkmann principle.[5]

True bony bridges have rarely been demonstrated in specimens from adolescent tibia vara, suggesting that the onset

*See references 72, 100, 116, 125, 130, 141, 149.

of the repetitive "trauma" occurs once the physis and epiphysis are much more developed than in the infantile form. Furthermore, mechanical realignment to unload the compressed medial physis usually is successful in curing adolescent tibia vara, as is gradual mechanical realignment produced by a lateral epiphysiodesis. Thus, even though there may be marked clinical deformity and significant radiographic physeal widening (evidence of disruption), the actual histologic insult to the physis must be relatively moderate, as evidenced by its acute or gradual response to mechanical unloading.

Clinical Features. The typical patient with adolescent tibia vara is a black male teenager whose body weight greatly exceeds 2 standard deviations above the mean (Fig. 21–12). At our institution, patients weighing up to 200 kg in early teenage years have been treated. Henderson has reported that the average weight of patients with adolescent tibia vara exceeds the 95th percentile for age by a mean of 43 kg.[70] Involvement frequently is unilateral, but bilateral cases are also seen. The preponderance of male patients versus female patients has not been explained.

Patients may present either because of the deformity itself or because of the deformity with symptoms. Many patients are essentially asymptomatic but, on close questioning, may describe an aching in the medial or anteromedial portion of the knee associated with activity or occurring toward the end of the day. An area of tenderness along the medial joint line is almost universal, and occasionally there are patellofemoral complaints. Internal tibial torsion is frequently present but variable in severity. In unilateral cases, limb length discrepancy is usually present, and the choice of treatment may be influenced by a discrepancy greater than 2.5 cm.

Radiographic Findings. Radiographically, the shape of the tibial physis is relatively normal, without the depression and beaking in the metaphysis that are typical of infantile tibia vara (Table 21–2). The sine qua non of diagnosis is a widening of the proximal medial physeal plate. This widening may be restricted to the medial one-fourth of the growth plate, or it may actually extend completely across the proximal tibial physis, suggestive of epiphysiolysis, as one might see in slipped capital femoral epiphysis (Fig. 21–12). The widening of the medial tibial physis is significantly greater when compared with the lateral side of the physis or with the physeal width in the normal, contralateral knee.[12]

In addition, there may be widening of the lateral distal *femoral* physis when compared to either the medial femoral physis of the same knee or with the distal femoral physis of the normal knee.[12,34] This *traction* widening on the lateral side of the varus deformity of the femur would appear consistent with Delpech's law, although interestingly, there usually is no localized widening of the lateral proximal tibial physis.

Lack of sloping or inferior beaking of the medial proximal tibia (with absence of medial articular depression) is another important difference from infantile tibia vara. This suggests that the proximal tibial physis and chondroepiphysis formed normally and ossified for a number of years before being compressed later in childhood, coinciding with the weight gain and adolescent growth spurt in a susceptible individual.

Treatment. Treatment is predominantly surgical. Orthotic management in patients with this degree of obesity is impossible and ineffective. Weight loss is undoubtedly desirable and should be recommended. However, it is probably not curative once the deformity is established and, because of the morbid obesity of most patients, is unrealistic to expect. The goal of surgical treatment is to correct the mechanical axis so that normal physeal growth is restored and degenerative arthritis of the medial compartment of the knee can be avoided.

OSTEOTOMY. Overcorrection in adolescent tibia vara is contraindicated,[12,85] as opposed to the recommended intentional overcorrection into valgus for infantile tibia vara. Correction to a neutral mechanical axis is sufficient to restore growth from the medial physis and thus prevent recurrence, although premature closure of the entire physis has been observed following high tibial osteotomy (Fig. 21–12).

There is an interesting dilemma when contemplating correction of the mechanical axis in these patients, because correction to a neutral mechanical axis in patients with morbidly obese thighs actually produces an unsightly cosmetic result that *appears* to be excessive valgus. In addition, when these patients undergo correction to neutral alignment, they may have difficulty ambulating because their thighs impinge during normal gait after mechanical realignment (Figs. 21–12G and H). Although undercorrection may invite recurrence of the deformity, it has been our observation that a final mechanical axis of 0 to 5 degrees varus is probably the best compromise between the mechanical correction desired and the problem posed by the massive proximal thigh girth.

Valgus-producing high tibial osteotomy with internal fixation is probably the most commonly used approach, opting for acute correction of the mechanical malalignment. Again, because of the massive thigh girth, external immobilization with casts is not effective in maintaining osteotomy alignment, and rigid internal fixation is far superior. The obvious challenge with this approach is that correct alignment must be obtained at surgery, as it cannot be changed postoperatively without reoperation. Loss of fixation and inappropriate intraoperative alignment are causes of postoperative under- or overcorrection.[72,107,116] In addition, the well-known complications of high tibial osteotomy (e.g., nerve palsy, compartment syndrome,[107,141,149] infection, delayed union or malunion, and, in this patient population, the possibility of deep vein thrombosis) make acute correction a formidable surgical task. Special large-circumference tourniquets for intraoperative hemostasis are required, and the logistics of performing high tibial osteotomy in a very obese patient must be taken into account before bringing such a case into a pediatric operating room.

LATERAL EPIPHYSIODESIS. Because of the potential complications associated with a high tibial osteotomy, gradual correction by lateral proximal tibial epiphysiodesis is an attractive alternative procedure. The technique is significantly simpler and associated with minimal morbidity, the only real complication being that angular correction is at times not realized. Assuming that the epiphysiodesis is technically

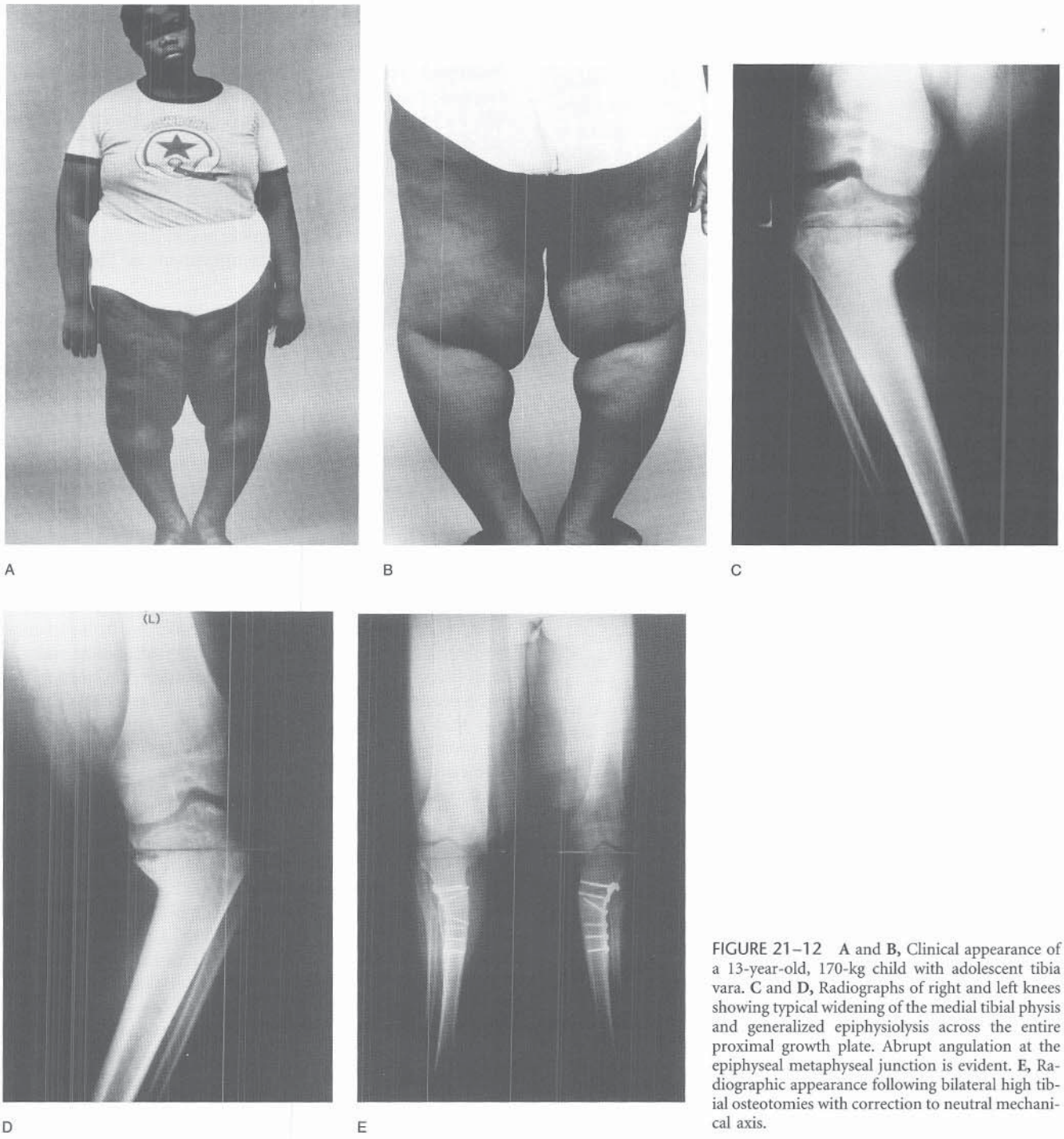
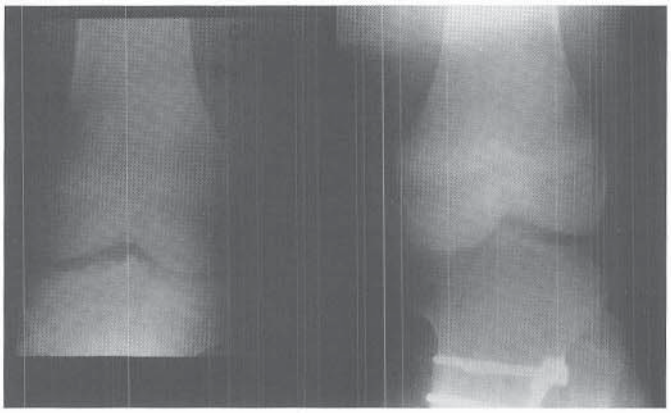


FIGURE 21–12 **A** and **B,** Clinical appearance of a 13-year-old, 170-kg child with adolescent tibia vara. **C** and **D,** Radiographs of right and left knees showing typical widening of the medial tibial physis and generalized epiphysiolysis across the entire proximal growth plate. Abrupt angulation at the epiphyseal metaphyseal junction is evident. **E,** Radiographic appearance following bilateral high tibial osteotomies with correction to neutral mechanical axis.

adequate, an incomplete or inadequate correction may be due to a medial growth plate that is simply too suppressed to respond to the tethering effect of the lateral epiphysiodesis.

Patients treated by this technique have about a 50 percent response rate in which the correction of the varus malalignment is judged to be satisfactory at maturity and no further treatment is necessary (Fig. 21-13).[72,87] Furthermore, performing a lateral epiphysiodesis in no way complicates subsequent correction by high tibial osteotomy should the final correction at maturity be inadequate. The main disadvantage of lateral epiphysiodesis is that rotational deformity cannot be corrected. Nevertheless, because of the minimal morbidity associated with lateral epiphysiodesis, we recommend it as the initial surgical treatment if more than 1 year of growth remains. By doing so, the complications and morbidity of a high tibial osteotomy will be completely avoided in 50 percent of these patients.

A lateral epiphysiodesis can be performed either as an open procedure or percutaneously (Fig. 21–13; see also Fig. 21–19). The percutaneous technique probably has a higher rate of failure, owing to technical inadequacy of the actual



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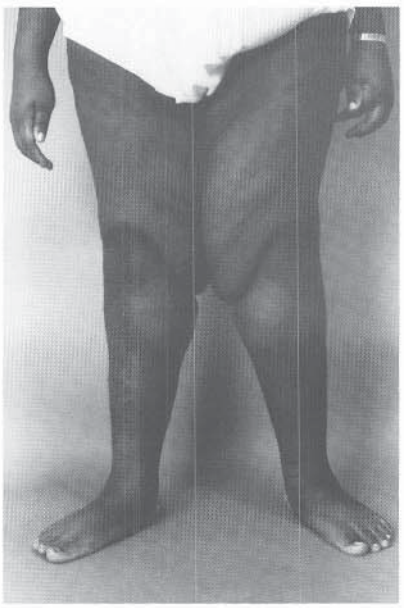

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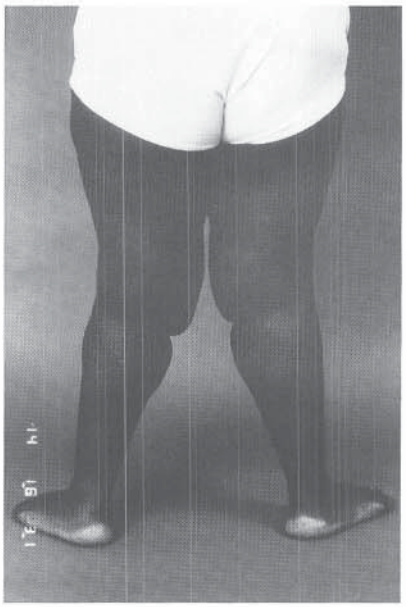


H

FIGURE 21–12 *Continued.* **F,** Close-up view of knees 1 year postoperatively. Both proximal tibial physes are closed, while both distal femora remain open. **G** and **H,** Postoperative clinical appearance. Despite attainment of a radiographic neutral axis, valgus "overcorrection" is evident.

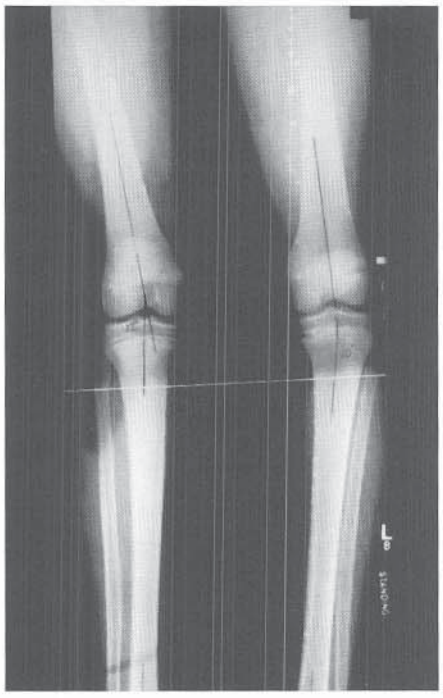
curetting of the physeal plate.[87] In addition, the main advantage of the percutaneous technique, cosmesis, may not be a critical feature of treatment in this patient population. We have found that an open epiphysiodesis performed through a mini-incision under fluoroscopic control is reliable in terms of ensuring that the lateral physeal plate is obliterated by curettage under direct vision. In selected patients, a distal lateral femoral epiphysiodesis can also be performed if there is radiographic evidence of distal femoral varus. We have not found proximal fibular epiphysiodesis necessary as part of the treatment for adolescent tibia vara in the 13- to 15-year-old patient population.

TABLE 21–2 **Radiographic Features of Adolescent Tibia Vara**

- Shape of epiphysis relatively normal
- Lack of beaking of medial tibia metaphysis
- Widening of the proximal medial physeal plate, sometimes extending across to lateral side of physis
- Widening of the lateral distal *femoral* physis compared to either the medial femoral physis of same knee or distal femoral physis of normal knee

REALIGNMENT BY EXTERNAL FIXATION. The problem of achieving accurate neutral alignment intraoperatively in this patient population has been alluded to, and the inability to postoperatively adjust internal fixation is a relative disadvantage. In addition, limb length inequality may exist in unilateral cases, and the ability to achieve equalization by lengthening following angular realignment makes external fixation all the more attractive. The major disadvantages of external fixation are pin or wire complications, which can produce loosening, sepsis, or nerve palsy, and joint stiffness and muscle weakness, which can complicate prolonged treatments. Because the total treatment time with external fixation is definitely longer than the 6 to 8 weeks until union by conventional osteotomy, these disadvantages may have a significant effect on the ultimate outcome.

Three methods of external fixation have been reported. External fixation can be used to align an extremity acutely after complete tibial and fibular osteotomy.[129] Alternatively,



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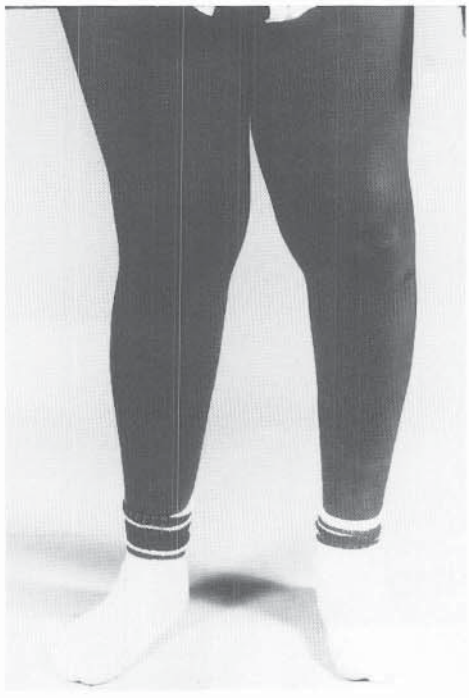

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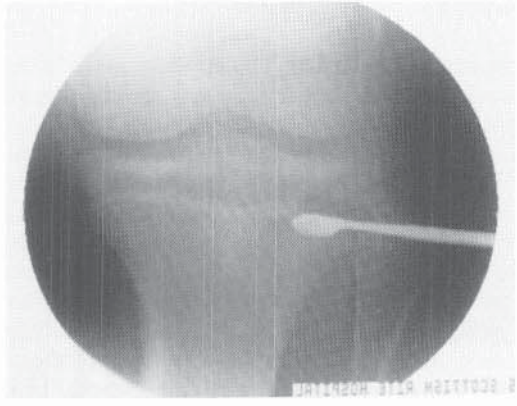

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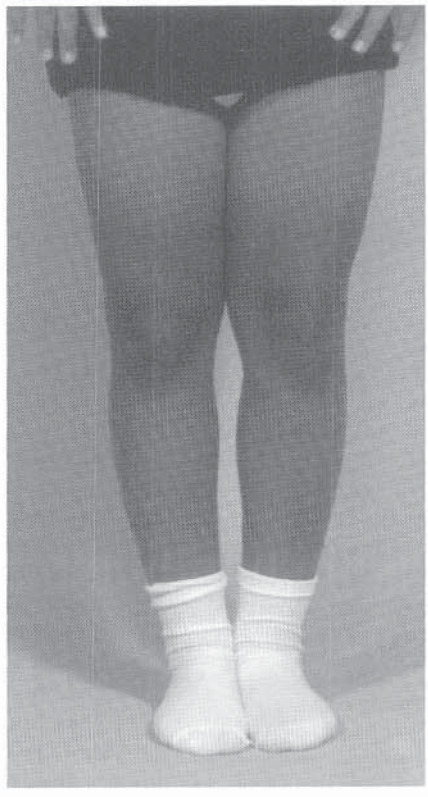


D

FIGURE 21–13 **A,** Radiographic appearance of a boy age 13 years 6 months with adolescent tibia vara on the left. Only the medial one-fourth of the proximal physis appears widened. Excessive physiologic genu valgum is present on the right. **B,** "Windswept" clinical appearance of the combination of deformities. **C,** Fluoroscopic view of lateral proximal tibial epiphysiodesis performed via a mini-incision. The peripheral one-fourth of the physis is curetted (depth = 1.5 cm). **D,** Clinical appearance 1 year following left lateral tibial and right medial distal femoral epiphysiodesis.

a corticotomy technique with gradual correction (distraction osteogenesis) can be utilized, employing either circular or monolateral external fixators (Fig. 21–14).[30,37–39,57,147] Hemichondrodiastasis, or asymmetric physeal distraction, has also been employed, with mixed results, depending on the rapidity and strength of bony consolidation in the physeal distraction gap.[37,39,129] Because of the almost certain physeal closure following physeal distraction methods, the use of this technique is limited to patients nearing skeletal maturity. The time to consolidation with physeal distraction can be prolonged,[129] and thus this approach appears to offer little advantage over conventional metaphyseal corticotomy. De Pablos and Franzreb, however, reported unusually rapid consolidation (average of 2.3 months) after physeal distraction with minimal morbidity, due to the shorter time in the fixator.[39]

The trade-off for the postoperative adjustability of external fixators is a prolonged treatment time, averaging 12 weeks to union or frame removal.[30,57,129,147] Obese patients may have difficulty engaging in postoperative rehabilitation activities with a circular frame and walker or crutches, leading to increased reliance on nonweightbearing mobility. This, in turn, can slow bony consolidation. Knee discomfort due to fixation wires near the joint may compound immobility; thus, half-pin techniques that use more anterior pin placement are better tolerated.

In summary, although correction by external fixation has the unique advantage of postoperative adjustability, the technique is just as formidable as traditional osteotomy, although with a different set of problems related to maintaining function and tolerating longer treatment periods with potential fixation and pin complications.

TIBIA VARA SECONDARY TO FOCAL FIBROCARTILAGINOUS DYSPLASIA

Occasionally, an infant or toddler presents with unilateral varus of the tibia, the deformity appearing slightly more distal than the knee joint itself. If the child has reached standing age, hyperextension of the knee may also be present (Fig. 21–15). This latter finding usually is not present in the toddler with infantile tibia vara, although the lateral thrust seen in stance phase can mimic this deformity. The radiographic findings of focal fibrocartilaginous dysplasia separate this entity from infantile tibia vara, as do the pathologic findings discovered during surgical treatment.

Radiographic Findings. Radiographs of focal fibrocartilaginous dysplasia (Table 21–3) show a characteristic abrupt varus at the metaphyseal-diaphyseal junction of the tibia, clearly not involving the physis (Fig. 21–15C). There is cortical sclerosis in and around the area of the abrupt varus on the medial cortex. A radiolucency may appear just proximal to this area of cortical sclerosis, which probably corresponds to the fibrocartilaginous tissue found at surgery in the area of the insertion of the pes anserine tendons. The etiology of this defect and the pathogenesis of the deformity are unknown.

Treatment. The importance of recognizing this variation of infantile tibia vara is that the deformity often resolves without surgery. Some 28 cases of tibia vara secondary to focal fibrocartilaginous dysplasia have been reported,[2,89] with a slight majority spontaneously correcting because of the normal proximal tibial physeal growth. Surgical treatment may be necessary if the deformity progresses or fails to resolve during a period of observation or orthotic management (Fig. 21–15).[11,117] Other reports note that in the majority of cases, the deformity resolves without osteotomy.[20,75,90]

In two of three patients treated at our institution, the deformity resolved without surgery. One patient, treated several years prior to the first report of this condition in 1985,[11] had radiographic findings characteristic of both infantile tibia vara and, in retrospect, focal fibrocartilaginous dysplasia (Fig. 21–16). The condition responded nicely to orthotic management. The consensus treatment of nonoperative management (observation, orthosis) appears sound, with a corrective high tibial osteotomy becoming necessary only when the deformity fails to resolve or progresses.

Genu Valgum (Knock-Knees)

Valgus alignment of the lower extremities is normal in the child between 2 and 8 years of age (Fig. 21–17).[134] The maximum amount of physiologic valgus occurs between the ages of 2 and 4 years, after which the alignment of the lower extremity assumes a mild valgus femoral-tibial angle, the normal alignment in the adult. Therefore, by age 8 years there should be little or no change in the lower extremity alignment (see Fig. 21–3A), and preparation for treatment of what is deemed excessive physiologic valgus may be made at this age.[143–145]

In the younger child passing through the physiologic valgus phase, radiographs are not indicated unless there is evidence of short stature, a history of trauma or infection, metabolic bone disease, or significant asymmetry suggesting an undetected underlying etiology unilaterally (see discussion under Miscellaneous Causes, below). In addition, the rotational profile of the extremity must be ascertained, as an apparent increase in valgus may result from a rotational profile presenting as increased femoral anteversion, producing an inward turn to the knee during stance phase. Alternatively, an alignment combination with femoral retroversion and adduction, as might be seen in slipped capital femoral epiphysis, can appear to have excessive knee valgus. As with any knee evaluation in the pediatric age group, the hip must also be evaluated.

PHYSIOLOGIC GENU VALGUM

After age 8 years, correction of excessive physiologic genu valgum may be indicated when there is gait disturbance, difficulty running, knee discomfort, patellar malalignment, evidence of ligamentous instability, or excessive cosmetic concern.[151,158] Identification of the malalignment during growth is optimal, so that some form of hemiepiphysiodesis (temporary with staples or transphyseal screws, or permanent using timed hemiepiphysiodesis*) may be planned. Hemiepiphysiodesis procedures have the advantage of producing unilateral physeal inhibition, thus achieving correction gradually and close to the joint, where the deformity correction will be most effective. Hemiepiphysiodesis is a

*See references 17, 18, 26, 81, 112, 126, 151, 174.

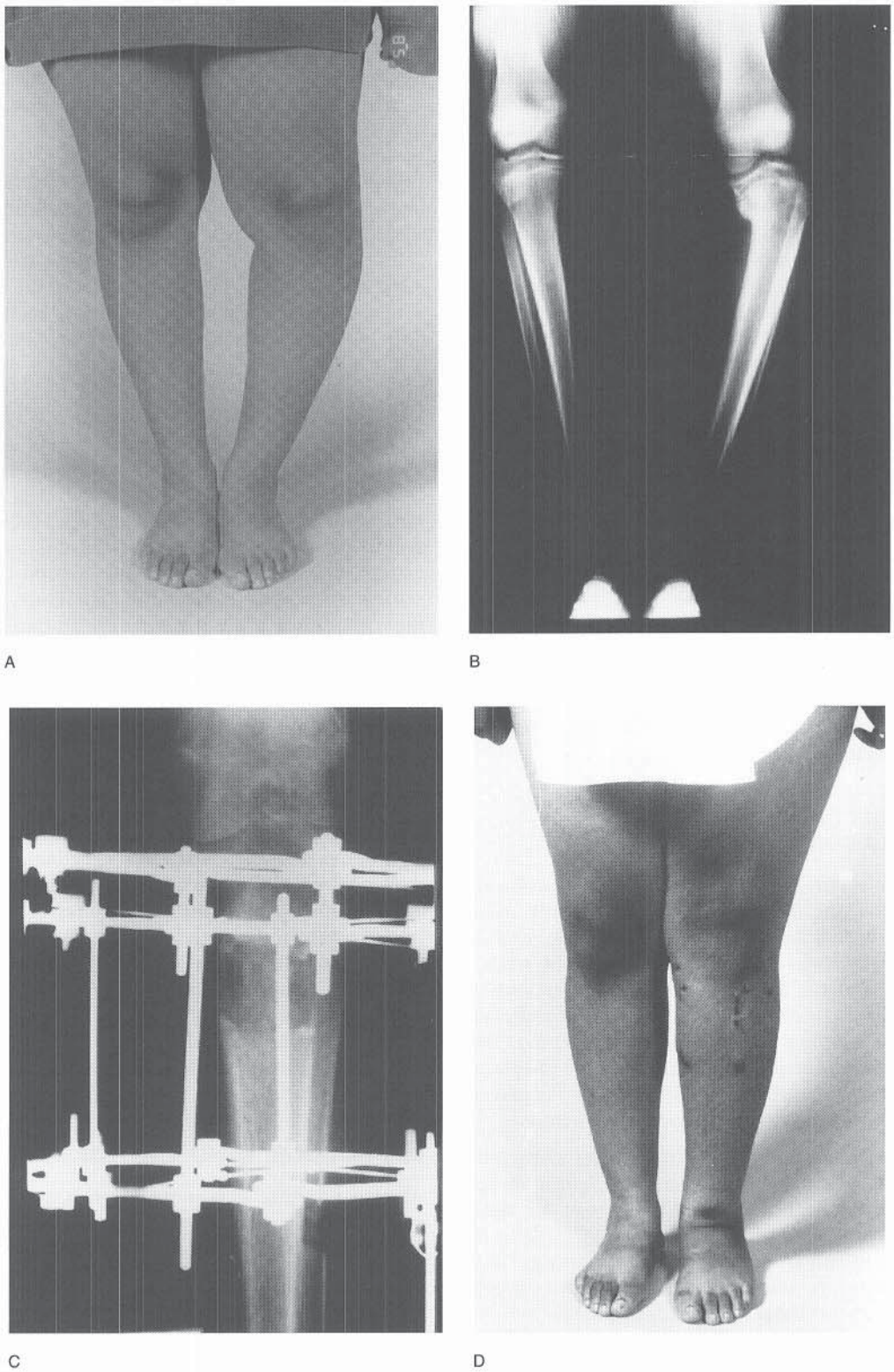
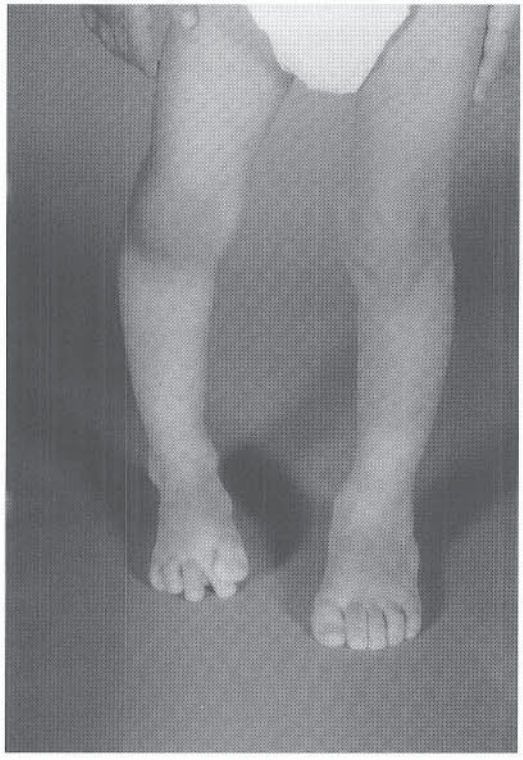


FIGURE 21–14 **A** and **B,** Clinical and radiographic appearance of a 13-year-old with adolescent tibia vara. There is a 2-cm shortening on the left. **C,** Radiograph obtained following gradual correction by external fixation. The regenerated bone was allowed to consolidate. Total time in frame was 14 weeks. **D,** Final result following frame removal.



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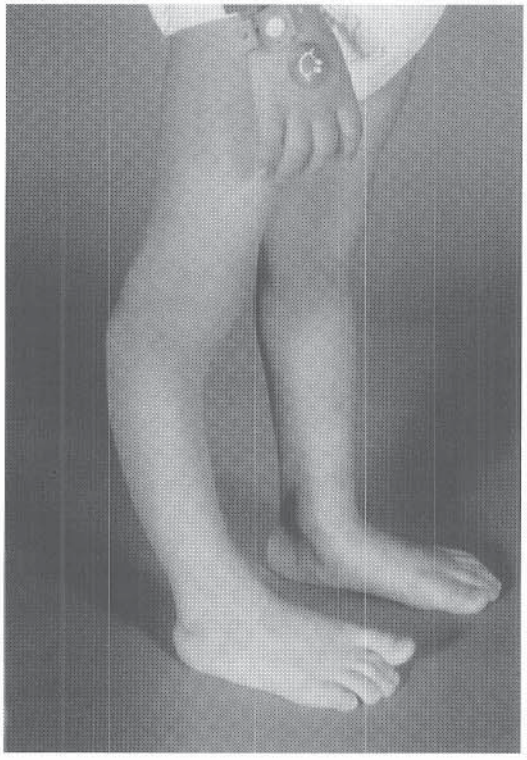

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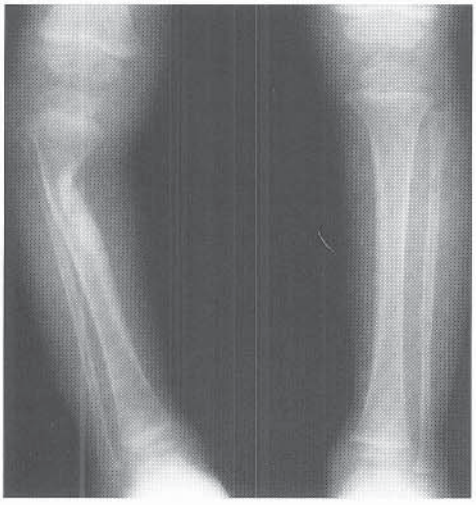

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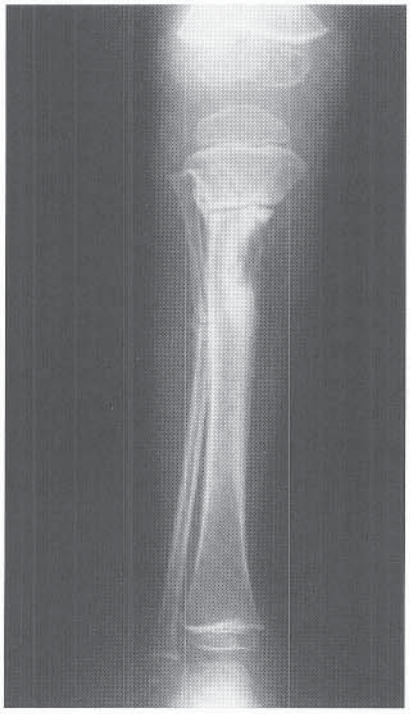

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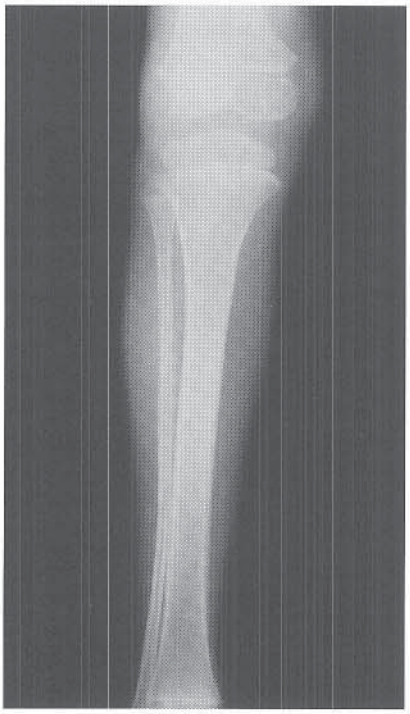

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FIGURE 21–15 **A** and **B,** Clinical appearance of a 15-month-old child with focal fibrocartilaginous dysplasia. Note the recurvatum associated with the varus at the metaphysis. **C,** Radiograph after 1 year of bracing shows no improvement in the deformity. **D,** The patient underwent upper tibial osteotomy and curettage of the focal lesion. Pathology: benign fibrocartilage. **E,** Radiographic appearance 3 years postoperatively.

TABLE 21–3 **Radiographic Features of Tibia Vara Secondary to Focal Fibrocartilaginous Dysplasia**

- Abrupt varus at the metaphyseal-diaphyseal junction of the tibia not involving the physis
- Cortical sclerosis in and around an area of abrupt varus on the medial cortex
- Radiolucency may appear just proximal to the area of cortical sclerosis

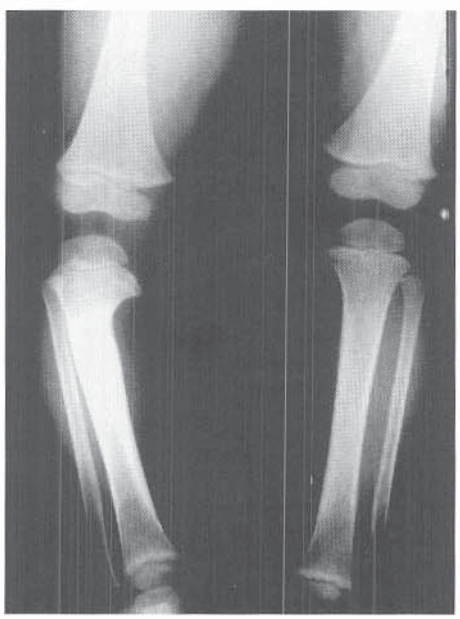
lesser surgical procedure than osteotomy, avoiding the possible neurovascular complications of the latter as well as the complications attendant on delayed union or malunion and surgical infection.

Several forms of hemiepiphysiodesis currently are practiced. The traditional open procedure[123] and the percutaneous method[26] attempt to produce permanent unilateral growth arrest by physeal ablation. Therefore, an element of delay in the timing of correction may be important, as ideally the correction should be done as the patient nears skeletal maturity, so that the possibility of overcorrection due to continued contralateral physeal growth is avoided. To this end, Bowen and associates have recommended use of the Green-Anderson growth charts and measurement of the transverse width of the physis to arrive at a mathematical formula and table to predict the precise timing of a unilateral epiphysiodesis for angular correction.[17,18] The predicted amount of angular deformity is determined from the table once the skeletal age and width of the physis are determined.

The Bowen angular correction table should be used only as a guideline for angular correction. Because of inherent error in determining skeletal age using the Greulich and Pyle atlas and the lack of standardization of standing radiographs to reliably measure physeal width, over- and undercorrection using this technique are common in our experience. In light of the deficiencies in methods to time epiphysiodesis for management of limb length discrepancy[103] it is not surprising that prediction of angular correction is inexact. For this reason, we have tended to use *untimed* hemiepiphysiodesis,[87] in order to ensure that the desired unilateral physeal arrest is achieved and correction produced. The patient is followed closely at 4- to 6-month intervals, and if correction is completed while growth still remains (as indicated by plain radiography and bone age determination), then hemiepiphysiodesis of the remaining lateral portion of the physis is performed to complete the epiphysiodesis and prevent overcorrection. Untimed hemiepiphysiodesis requires patient compliance with follow-up, as failure to return for appropriate follow-up can lead to overcorrection (see Fig. 21–18).

Percutaneous hemiepiphysiodesis performed under fluoroscopic control is widely practiced, following the success noted in early reports.[17,26] Cosmetic outcome was the driving force behind this technique, but the procedure is also preferable to the open Phemister technique because of less postoperative morbidity. A cavitation technique, more than curettage, is recommended to ensure adequate physeal ablation in the setting of undulating physeal anatomy, especially in the distal femur (Fig. 21–19).[87] A mini-incision technique in which the physis is localized fluoroscopically and curetted under direct vision is also reliable. The medial physis should be curetted or excavated to a depth of 1.5 cm peripherally to ensure a peripheral bridge adequate to tether growth while preserving growth of the lateral physis (Fig. 21–13C).

In children who have significant growth remaining (boys



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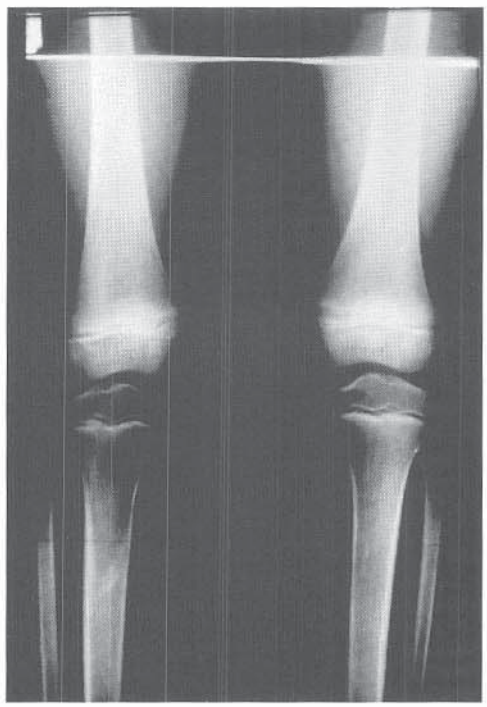

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FIGURE 21–16 **A,** Radiographic appearance in a child age 1 year 10 months with focal fibrocartilaginous dysplasia and a possible Blount's lesion. The diagnosis of focal fibrocartilaginous dysplasia was not made at the time (1978). Bracing was instituted. **B,** Radiographic appearance at age 6 years 3 months. The tibia vara and all bony lesions have resolved.

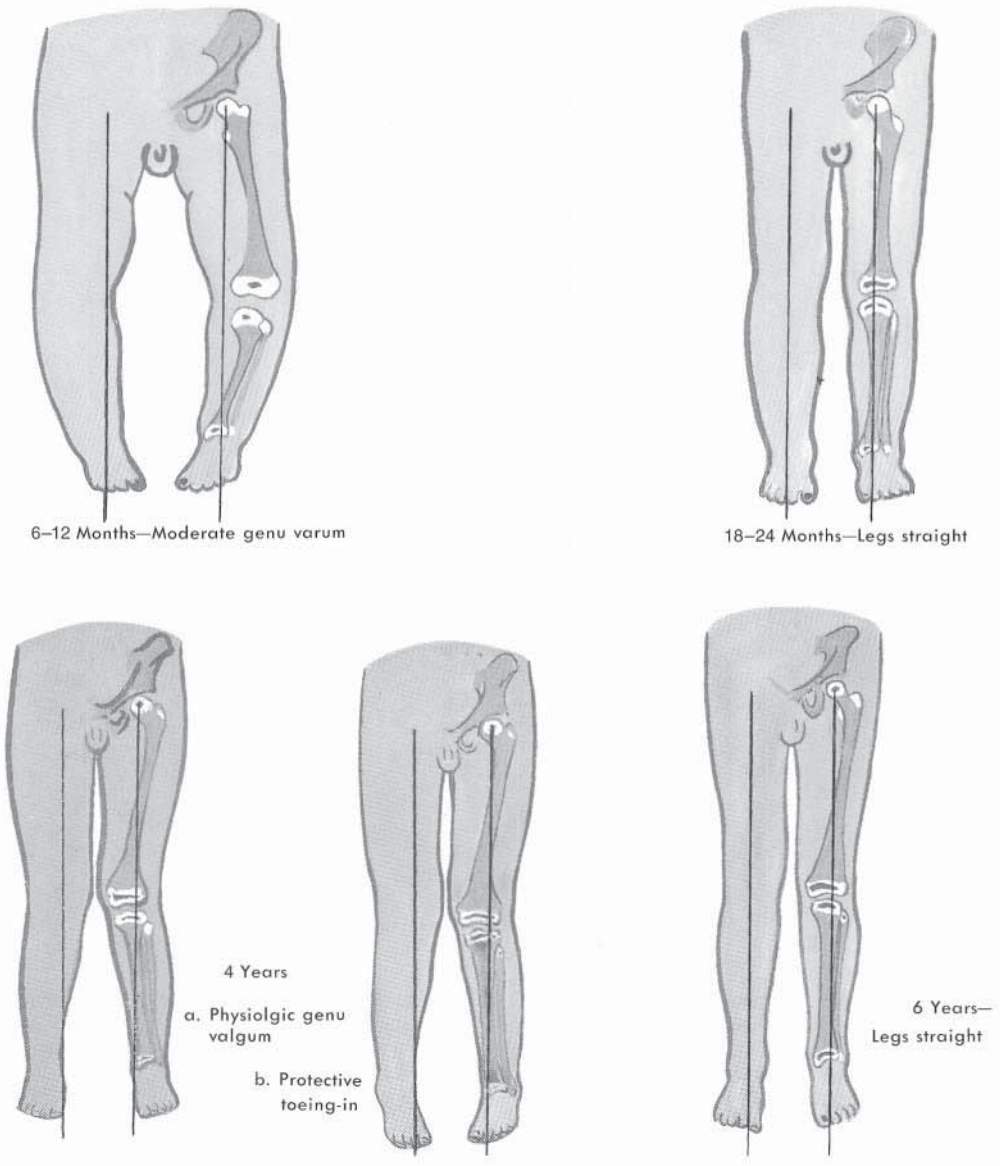
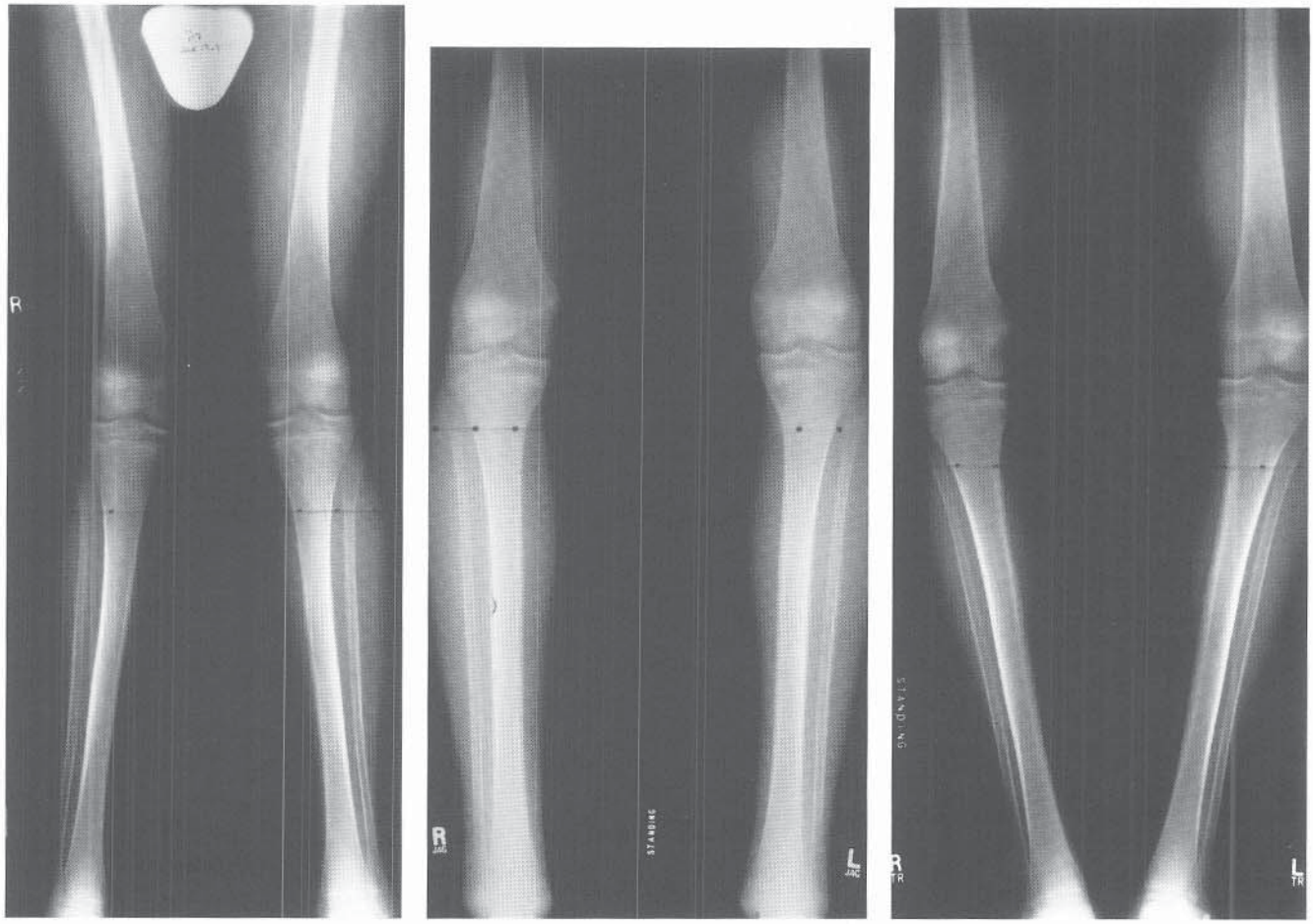


FIGURE 21–17 The evolution of lower limb alignment from varus to valgus to normal alignment.
6 to 12 months: Genu varum
18 to 24 months: Legs straight
4 years: a. Physiologic genu valgum
b. Protective toeing-in
6 years: Legs straight

younger than 12, girls younger than 10), reversible or transient hemiepiphysiodesis approaches are attractive, because correction can be obtained soon after the deformity is diagnosed rather than delaying the permanent hemiepiphysiodesis until near maturity. In addition, once the correction is achieved, removal of the temporary tethering implant (staple, screw) has at least the potential to allow resumption of growth. Blount and Clarke first reported such a method in 1949 using staples,[15] with 87 percent excellent results eventually reported in 82 knees.[174] Blount recommended that slight overcorrection be achieved, as there was a tendency for the original deformity to "rebound" following staple removal. Staple revision due to extrusion or migration was necessary in 12 percent of knees. The actual technique of stapling is crucial to the success of the procedure. Incision of the perichondrial ring requires a large exposure and is actually likely to produce a permanent epiphysiodesis. Thus, stapling should be performed as an extraperiosteal procedure and under fluoroscopic control, using heavy staples that will resist physeal growth without deforming or migrating. At least three staples placed in the anterior, middle, and posterior one-third of the transverse extent of the physis are necessary, and the staple prongs should be centered on the physis and parallel to it.[158] Blount recommended that no staple be removed before a minimum of 1 year in situ, regardless of the rapidity of correction seen. In our experience, it is not uncommon for the staple to remain for 2 years or longer before adequate correction is produced, thus making continued growth on staple removal problematic (Fig. 21–20). Several authors have recently re-



A B C

FIGURE 21–18 **A,** Preoperative bilateral genu valgum in a 14-year-old boy. Because his bone age was only 13 years 6 months (i.e., 2 years of growth remaining), a permanent hemi-epiphysiodesis was delayed an additional 9 months. **B,** Radiographic appearance 6 months after medial distal femoral and proximal tibial hemi-epiphysiodesis. Correction was deemed complete, and closure of the lateral physes was scheduled. **C,** The patient failed to return for closure of the lateral physes until an additional 6 months had passed. Genu varum resulted, owing to continued lateral physeal growth.

ported 90 percent excellent results without any permanent arrests.[61,151]

Besides the unpredictability of growth following staple removal, asymmetric growth retardation in the sagittal plane, producing genu recurvatum or flexion deformity, is an occasional complication. Should the staples migrate or deform, the effectiveness of the hemiepiphysiodesis may be impaired, and a revision will be necessary. Finally, the scars around the knee resulting from such stapling may become a cosmetic issue, as they frequently spread or hypertrophy. Percutaneous methods[18,26,81] are clearly more desirable from a cosmetic standpoint.

Recently a variation on temporary hemiepiphysiodesis was reported that uses screw fixation.[112] Originally used mainly for the distal tibia through the medial malleolus,[35,150] this procedure, performed percutaneously with cannulated implants, is an attractive alternative for correcting angular deformity about the knee. Although the procedure was undertaken primarily to control limb length discrepancy, angular correction by hemiepiphysiodesis was successful in a total of nine knees, including six treated for genu valgum.[112] The issue of overcorrection due to permanent physeal closure was uncertain, however, again emphasizing that with *any* transient hemiepiphysiodesis technique, the surgeon walks a fine line between earlier removal of the implant with incomplete correction and leaving the implant in place too long to achieve correction but possibly producing permanent arrest.

Corrective osteotomy for excessive genu valgum is appropriate when the patient presents near or after skeletal maturity, too late for correction to be obtained by hemiepiphysiodesis. The preferred technique, assuming the deformity is primarily in the distal femur (most common), is a medially based closing-wedge osteotomy performed through a medial incision. If a transosteotomy lag screw technique or an appropriately contoured compression plate (Fig. 21–21) is used, a stable construct can usually be achieved so that postoperative immobilization can be minimized. By performing a closing-wedge (i.e., shortening) osteotomy at the level of the distal femur, the surgeon should be able to avoid the neurovascular complications reported for proximal tibial osteotomy.[158]

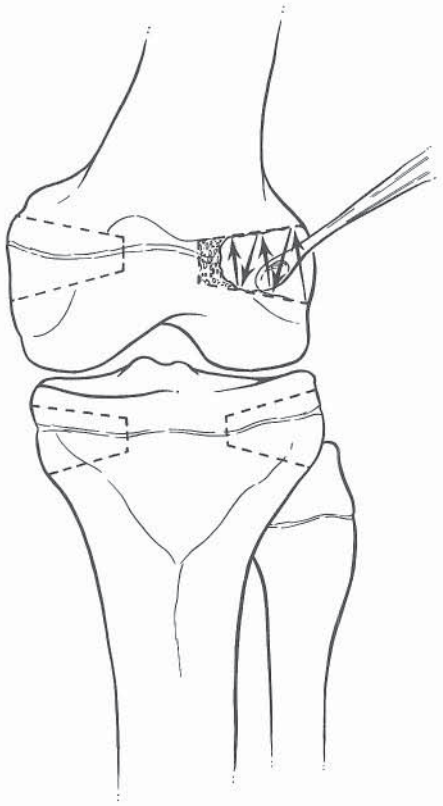


FIGURE 21–19 Cavitation of the peripheral physis, including metaphyseal and epiphyseal extension, to ensure adequate ablation for epiphysiodesis.

An opening-wedge technique[135] is not recommended for routine correction of bilateral physiologic genu valgum, since maintaining length is not an issue. Besides the requirement for iliac crest bone graft for the opening-wedge technique, healing is much slower, and loss of correction with recurrent valgus is a distinct possibility should the graft resorb and the internal fixation be inadequate to resist osteotomy settling. When bilateral procedures are performed, the advantage of early mobilization without casts is impossible with the opening-wedge technique.

Rarely the deformity may be more prominent at the proximal tibia. In these cases there is no superolateral tilt to the knee joint on a standing radiograph, as is typical with the distal femoral deformity. Proximal varus osteotomy of the tibia is associated with the possible significant complications of peroneal nerve palsy, vascular insufficiency, and compartment syndrome, and it should be performed by a medial approach closing-wedge technique to produce a "shortening" procedure. Rarely is such an osteotomy necessary for a case of excessive physiologic knock-knee.

GENU VALGUM SECONDARY TO PREVIOUS PROXIMAL TIBIAL FRACTURE

In 1953 Cozen reported valgus deformity of the tibia following a proximal metaphyseal fracture without an associated fracture of the fibula.[32] Several explanations for this phenomenon have been proposed, including soft tissue interposition in the fracture site[166] and stimulation of the growth of the medial physis[76,88,171] and tethering of the lateral physis by the intact fibula or iliotibial band.[32,160]

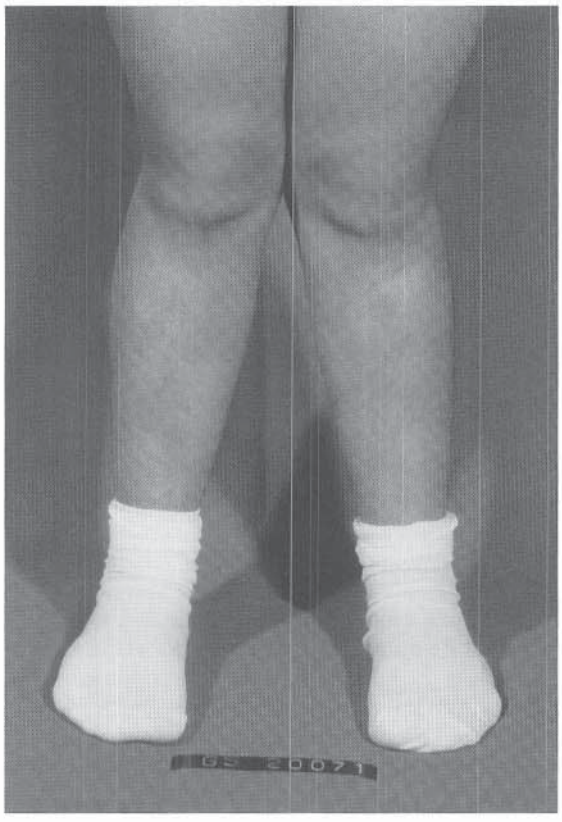
The actual incidence of this deformity is unknown, because the denominator (the total number of tibial fractures) in any series is unknown. A 53 percent incidence has been proposed in a review by Tuten and colleagues.[164] When a nondisplaced proximal metaphyseal fracture is seen, the extremity should be placed in a varus-molded long-leg cast, although there is no evidence that this will prevent valgus from developing. Parental education is mandatory at this stage. *Open* reduction, in order to extract interposed soft tissue, has not been successful in preventing the subsequent valgus.[22]

Treatment of posttraumatic tibia valga is primarily by observation (Fig. 21–22). The maximum deformity is reached at about 1 year post injury. Spontaneous improvement over several years then occurs.[164,172] The deformity is cosmetic, as a functional deficit from the valgus deformity is rare.[164] However, osteotomy to correct the valgus is contraindicated until early adolescence, because a surgical osteotomy in the proximal tibia in a growing child is just as likely to produce the posttraumatic valgus deformity as the original trauma itself (Fig. 21–23).[6,22,76] Hemiepiphysiodesis of the proximal tibia has been used to manage the deformity, should angulatory correction be deemed necessary.[113] Transient hemiepiphysiodesis with staples is essential, and the temporary physeal arrest is more effective if performed within 2 to 3 years after the trauma itself. This is because a late hemiepiphysiodesis will produce a "Z" deformity in the tibia, as the original posttraumatic valgus will have migrated down into the diaphysis of the tibia if several years are allowed to pass before correction (Fig. 21–22C). The later hemiepiphysiodesis will produce a relative varus proximal to the diaphyseal deformity. The controversy juxtaposes the expected spontaneous correction of the deformity over time with the relative risk of complication (i.e., physeal arrest) from a transient hemiepiphysiodesis. Finally, if correction at maturity is elected, usually for symptomatic lateral compartment arthritis, fibular osteotomy should also be performed.

GENU VALGUM SECONDARY TO MISCELLANEOUS CAUSES

The differential diagnosis of genu valgum, besides physiologic and posttraumatic, includes a variety of metabolic disorders and syndromes. Marked genu valgum can result from any of the types of rickets, although renal osteodystrophy classically is most likely to produce valgus.[118] This is explained by the fact that patients with acquired renal failure are generally in the physiologic valgus age group when the metabolic bone disease becomes active, while in patients with familial hypophosphatemic rickets the bone disease is active during early infancy, when physiologic varus is present.

Treatment for excessive valgus associated with rickets includes optimal medical management to decrease the severity of the bone disease. Obviously, the better the etiology (e.g., renal failure) is controlled, the less severe the secondary rickets will be. As the valgus deformity is usually bilateral, orthotic management with KAFOs or reverse Blount-style



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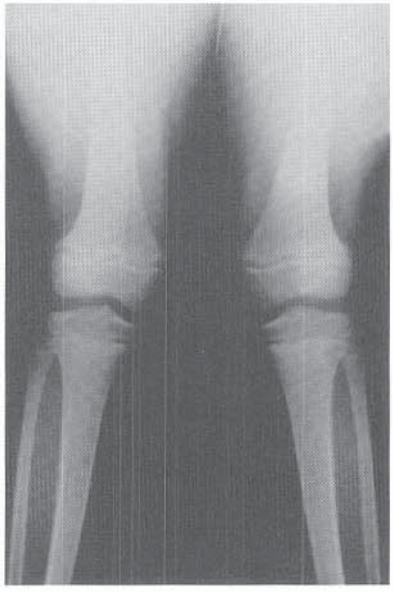

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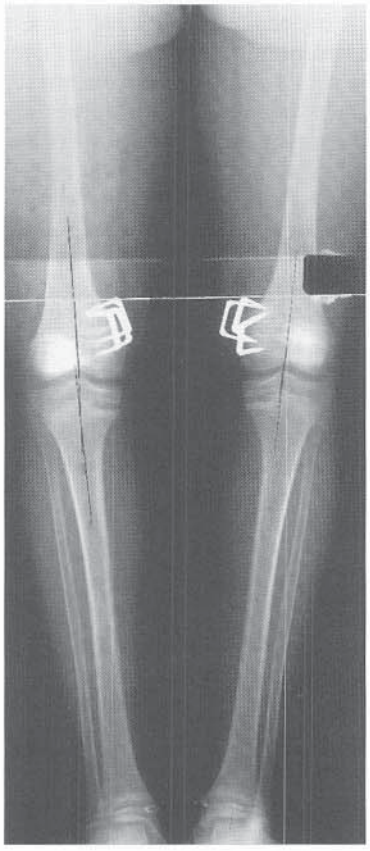

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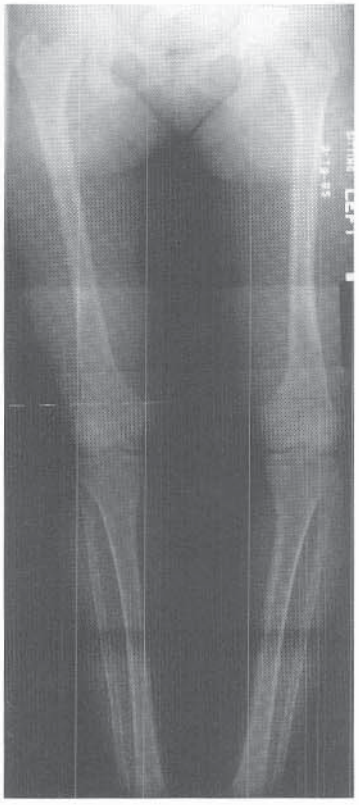


D

FIGURE 21–20 **A** and **B,** Clinical and radiographic appearance in a 9-year-old girl with metaphyseal chondrodysplasia and genu valgum. **C,** Radiograph obtained after medial distal femoral physeal stapling. After 16 months, correction was achieved and the staples were removed. **D,** Radiographic appearance 3 years later. The patient has symptomatic *genu varum* on the left due to permanent physeal closure medially. A distal femoral osteotomy was planned.

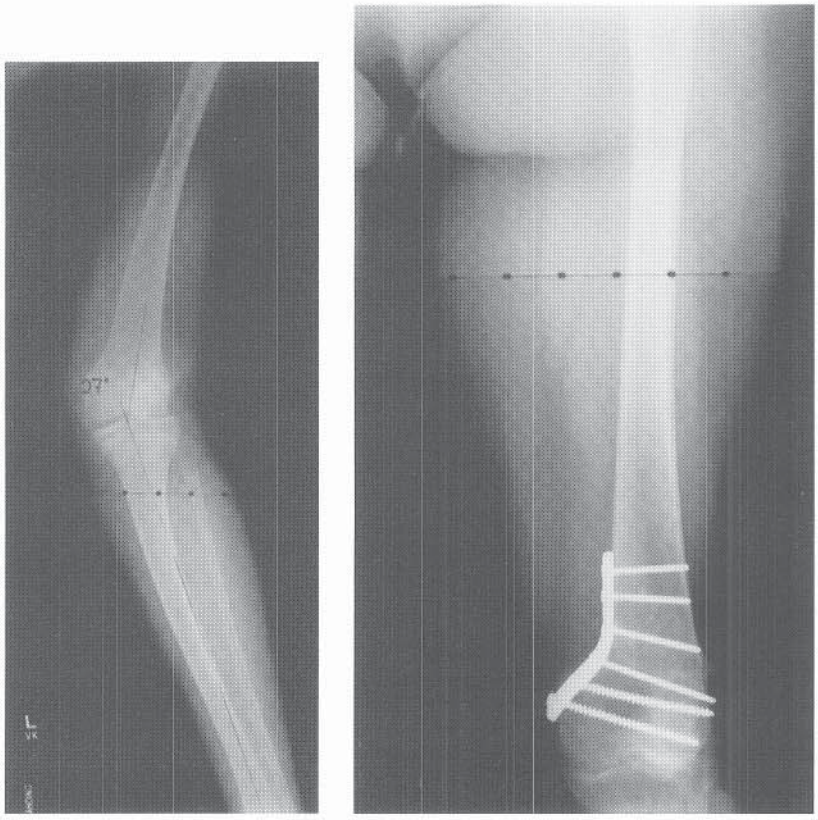


FIGURE 21–21 **A,** Radiographic appearance in a skeletally mature female with neglected genu valgum. Early lateral compartment degeneration was already present. **B,** Acute correction by closing-wedge osteotomy with medially placed internal fixation.

elastic braces (lateral uprights) is poorly tolerated because of the large amount of hardware on the child.

Transient or permanent hemiepiphysiodesis can be a valuable option. A certain unpredictability of effect exists, due to the abnormality of growth of the physis, and thus the response of the physis to unilateral tethering cannot be guaranteed. If the patient's growth is expected to be significantly impaired, physeal tethering is unattractive, as possible growth inhibition should be avoided. Osteotomy is always available, and the surgeon should be prepared to correct deformity at multiple levels at one sitting (see Chapter 31, Metabolic and Endocrine Bone Diseases).

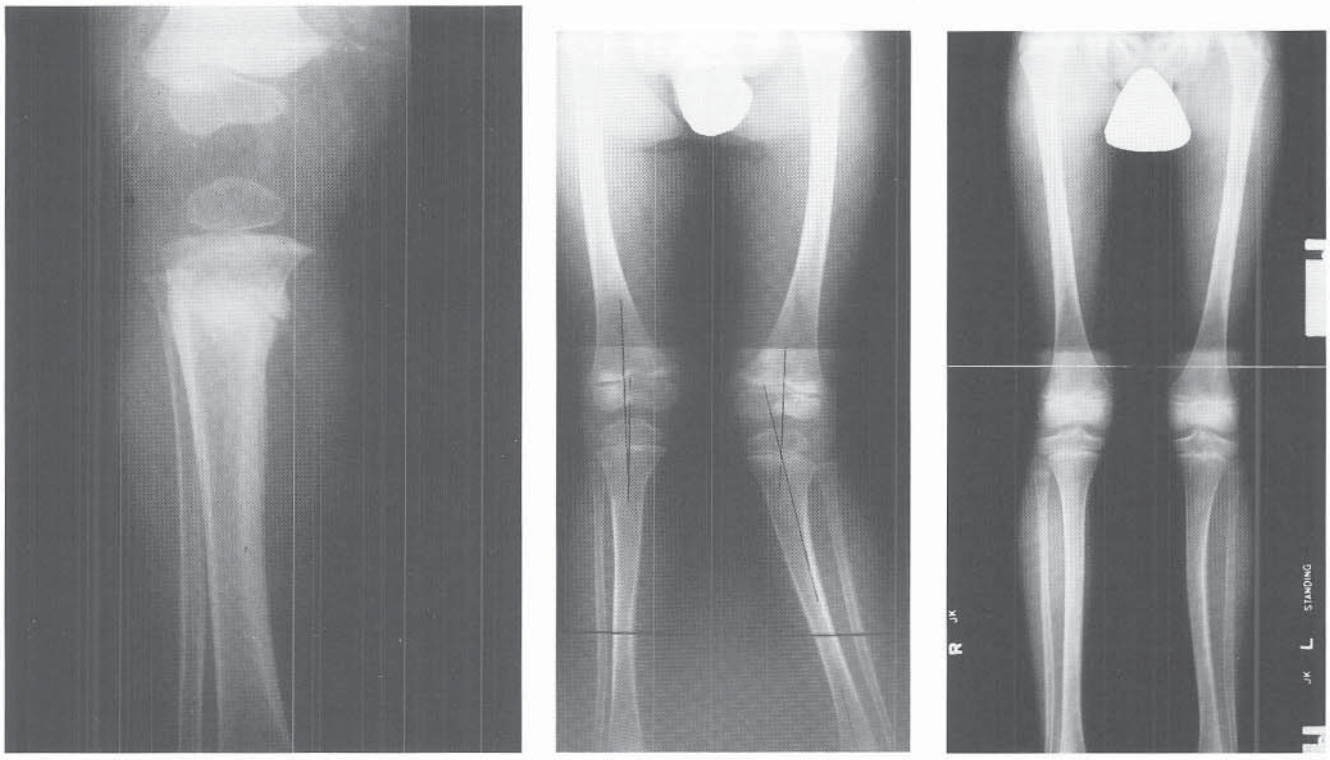
Valgus deformity is frequently seen in *spondyloepiphyseal* and *metaphyseal dysplasias* (Fig. 21–24). The management of limb alignment in these conditions is often complicated by marked joint laxity, and thus both intraoperative arthrography and external fixation may be important adjuncts to obtaining correction. Under anesthesia, the alignment of the nonweightbearing extremity may be so confounded by joint laxity that arthrography to illuminate the articular surface axis is necessary to guide angular correction. For the same reason, the ability to adjust alignment postoperatively when weightbearing alignment can be accurately determined is a valuable advantage of external fixation to manage osteotomy fixation (Fig. 21–24).

Asymmetric or unilateral valgus may be secondary to tumor-like conditions such as *fibrous dysplasia* or *enchondromatosis* (Ollier's disease) (Fig. 21–25). Radiographs generally are all that is required to make the diagnosis. Treatment is individualized and is based not only on progression of angulation but also on limb length discrepancy.

In *multiple hereditary exostoses* (osteochondromatosis), deformity at the knee is typically valgus and symmetric, and frequently does not require treatment. An important caveat is to delay excision of a proximal medial tibial exostosis if possible, to avoid risking an accelerating valgus deformity after excision due to the Cozen's fracture phenomenon. Valgus at the knee may be secondary to disturbed growth of the proximal fibula as a result of a large osteochondroma in this location. Ankle valgus may also be present, again due to a "short" fibula with disturbed distal growth. Hemiepiphysiodesis, either permanent or transient, has been useful in cases requiring realignment, both proximal and distal.

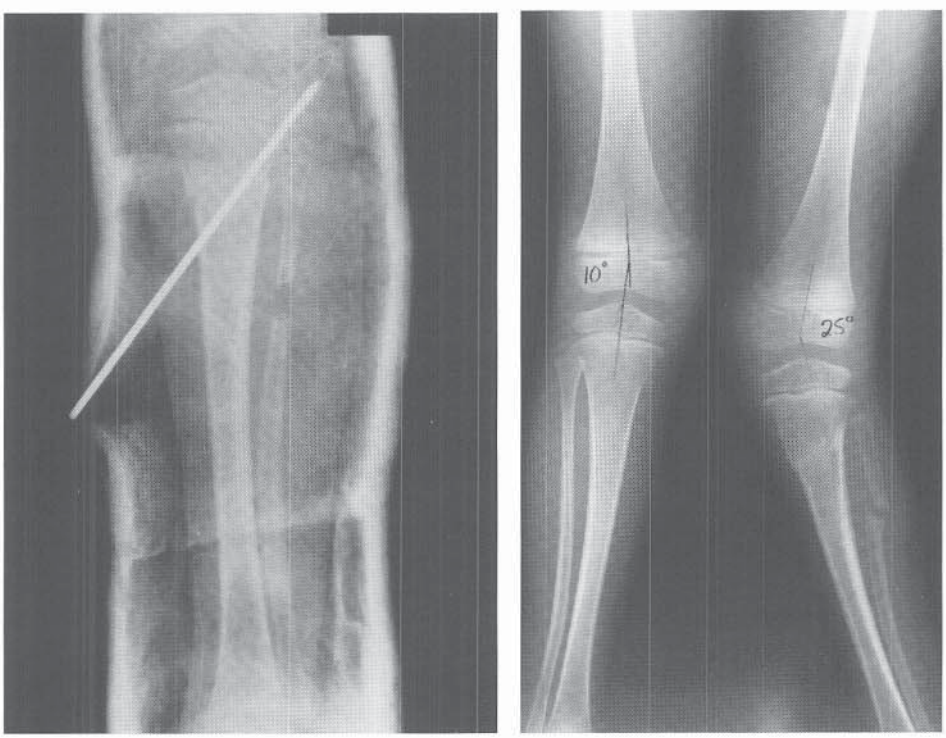
Tibial Torsion

Parents commonly complain of intoeing and outtoeing in toddlers and young children. In this age group there is a wide degree of normal variation, and most deviations from normal are physiologic variants that will spontaneously resolve.[143–145] Both intoeing and outtoeing may be due to deviations of the rotational alignment of the femur or tibia, and medial deviation of the foot (as in metatarsus adductus) is also a cause of intoeing.[128] *Tibial* rotation, or torsion, is the most common cause of intoeing and outtoeing up to the age of 3 to 4 years. The foot progression angle (FPA)—the angle created by the long axis of the foot in relation to



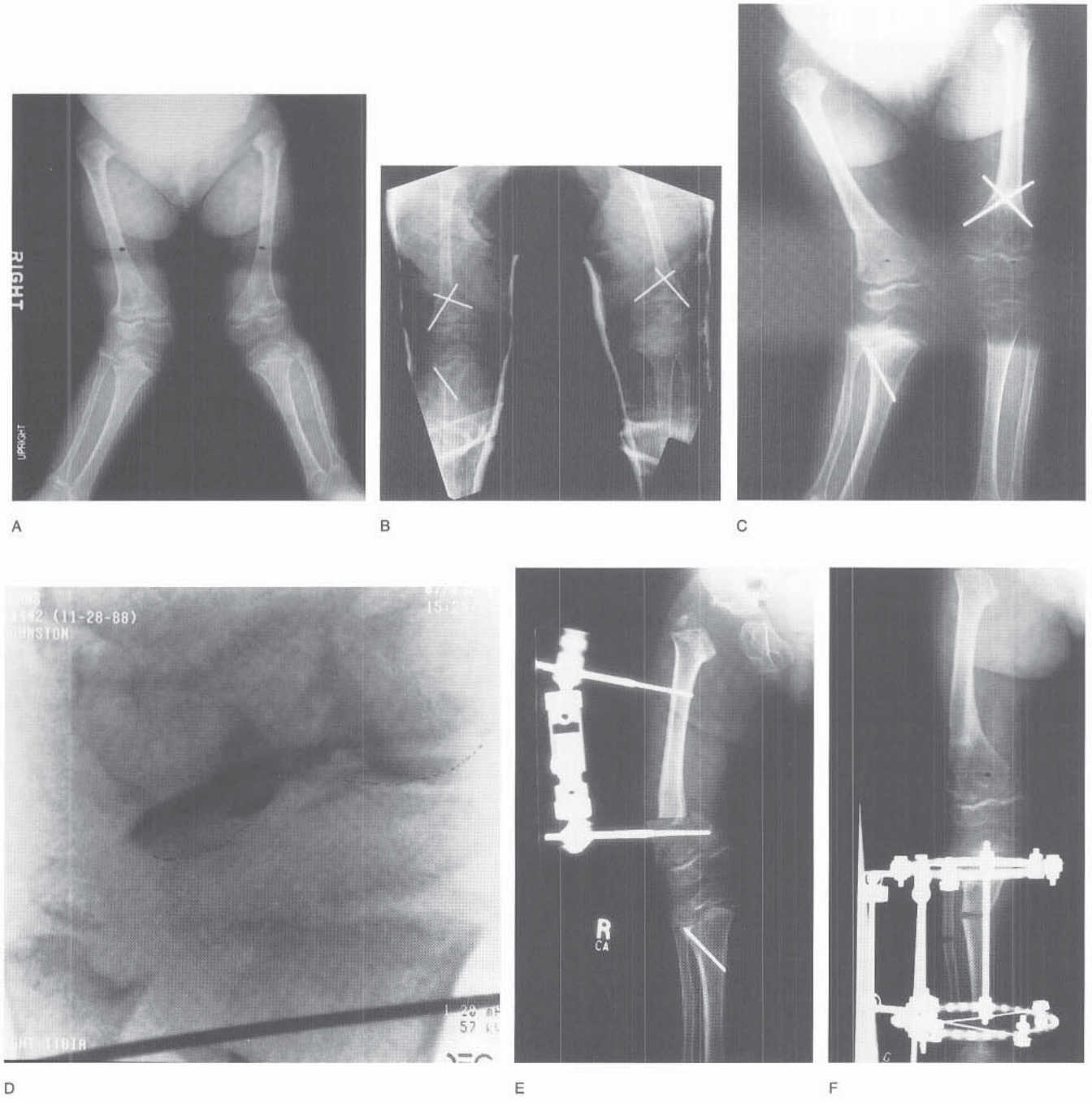
A B C

FIGURE 21–22 **A,** Nondisplaced fracture of the left proximal tibia in a 16-month-old boy. **B,** Radiographic appearance at age 4 years 9 months. The left lower extremity had a 17-degree valgus deformity, which was progressive (and expected) since cast removal 3 years earlier. **C,** Radiographic appearance at age 11. The valgus deformity had completely resolved clinically. A mild zig-zag deformity was seen radiographically. No treatment was prescribed.



A B

FIGURE 21–23 **A,** At age 4, a left proximal tibial rotation osteotomy was performed to correct excessive unilateral external tibial torsion. **B,** Radiographic appearance 8 months postoperatively. A significant valgus deformity has resulted.



A B C

D E F

FIGURE 21–24 **A,** Radiographs of a 6-year-old girl with SED congenita. **B,** She underwent correction by multiple osteotomies. **C,** Marked recurrence on the right 1 year postoperatively. **D,** Intraoperative arthrogram demonstrating valgus deformation within the intra-articular surfaces themselves. **E** and **F,** Because of the need to adjust alignment postoperatively in the weightbearing position, correction of the femoral and tibial deformities was accomplished with external fixation.

the line of progression during walking—is to a great extent determined by the degree of tibial rotation in this age group. After 4 years of age, the medial and lateral rotational profile of the femur becomes the greater determinant of gait.[115] Evaluation for a rotational deformity therefore must consider three levels of examination—the foot, the tibia, and the femur—and in more complex cases may also include evaluation of the hip itself. For most intoeing and outtoeing in the toddler, however, a determination of the tibial torsion by evaluating the angular difference between the axis of the knee and the FPA will be made during the orthopaedic examination. The reader is directed to the section on the focused examination of the lower leg (Chapter 4, The Orthopaedic Examination: Clinical Application).

In normal fetal development, the foot is medially rotated, and lateral rotation occurs with increasing age. The medial malleolus lies posterior to the lateral malleolus in the early fetus, is level with the lateral malleolus at birth, and lies anterior to the lateral malleolus at walking age.[101] The amount of lateral tibial torsion increases from about 5 de-

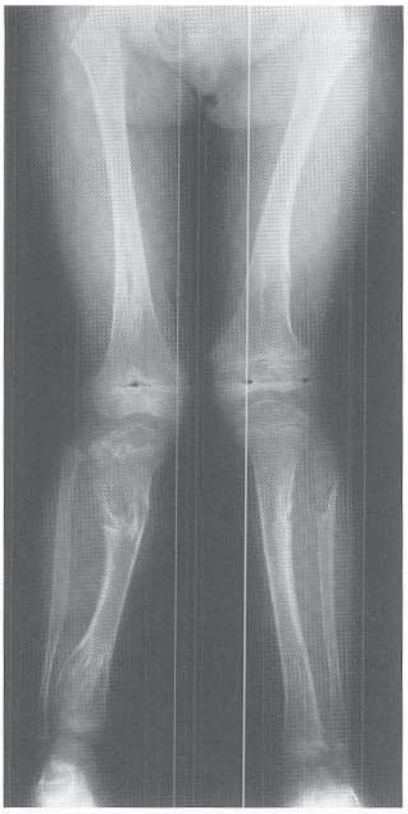


FIGURE 21–25 Asymmetric valgus due to multiple enchondromatosis. Note lesions in the left distal femur laterally and both ends of the left fibula. The right tibia is extensively involved and shortened due to transepiphyseal enchondromas.

grees at birth to an average of 15 degrees at maturity.[145,146] Owing to persistent medial rotation of the leg and to hindfoot rotation in the ankle mortise, commonly termed internal tibial torsion, the foot is rotated inward in the infant and toddler, as is determined by measurement of the thigh-foot angle[145] (see Chapter 4, The Orthopaedic Examination: Clinical Application). A medial thigh-foot angle of 30 degrees falls within 2 standard deviations of the mean in the age group of infants and toddlers.[145,146] An understanding of this normal range and of the natural history of tibial torsion is of utmost importance to avoid unnecessary treatment of a condition that is benign and self-resolving.

CLINICAL FEATURES

Most newborns actually have outtoeing. An external rotation contracture of the hip obscures any inward tibial rotation and posture. Any intoeing seen in infants suggests a possible metatarsus adductus or other deformity within the foot itself.[128] Most intoeing does not become evident until 12 months of age, when the child begins standing and the external rotation contracture of the hip has spontaneously resolved. Intoeing at this time results from medial tibial torsion, with a medially rotated thigh-foot angle. Physical examination of the lower extremities is normal or may show physiologic genu varum (see Figs. 21–1 and 21–17). Lateral tibial rotation will occur normally as the child grows,[143,144] and the natural history of spontaneous resolution should deter any efforts to treat this normal variation. Interestingly, infants with *lateral* tibial torsion as a cause of outtoeing, due to an intrauterine position producing excessive external foot rotation (often associated with a calcaneovalgus foot), do not seem to follow the predicted pattern of *continued* lateral tibial rotation during the first few years of life.

TREATMENT

Efforts aimed at changing the torsional profile of a long bone by an orthosis or other splint are generally ineffective and may simply add to parents' frustration. We have discouraged the use of Denis Browne (DB) splints, or other variations of night splints, for the simple reason that any rotational force applied to the lower extremity of a sleeping child will in all likelihood produce rotation through the points of least resistance, the joints themselves. Genu valgum or excessive lateral tibial rotation can occur with the use of the DB bar,[53] and its efficacy has never been proved.[69] The possible creation of a second deformity through the joints in order to compensate for a spontaneously correcting rotation in a long bone is reason enough to avoid such devices. Other methods used in the past, such as shoe modifications and twister cables, have also been shown to be ineffective in correcting these deformities.[53,93]

Because of the benign natural history of the condition and the generally unattractive and ineffective nature of orthotic management, observation and parental education are the main forms of treatment for tibial torsion. A small segment of the population with severe (more than 2 standard deviations above the mean) medial tibial torsion that does not spontaneously resolve may need treatment. In addition, excessive external tibial torsion might not resolve and therefore might need treatment at a later age. In either case, moderate to severe deformities that fail to resolve should alert the orthopaedist to the possibility of an underlying neurologic problem, for example, cerebral palsy. In the "intoeing clinics" at our institution, it is not uncommon to diagnose subtle spastic diplegia manifesting with either unresolved internal tibial torsion or excessive femoral anteversion with excessive external tibial torsion.

Surgical management by rotational osteotomy may therefore be necessary in the rare situation in which the child has persistent functional or cosmetic problems after age 8 years.[144,145,159] There may be a family history of severe intoeing and medial tibial torsion which can alert the physician to the possibility of future incomplete resolution.

Persistent *lateral* tibial torsion can be a sequela of prematurity when the infant is nursed prone in the neonatal unit for several weeks.[92] Lateral tibial torsion may also be present in a neurologically normal patient who has a torsional malalignment syndrome, associated with excessive medial femoral torsion. In this situation the patient may have an awkward gait or difficulty running and may seek medical attention for patellofemoral symptoms ranging from anterior knee pain to actual subluxation of the patella. This condition appears to be developmental, and it can produce definite disability from either pain or rotational deformity. Fortunately, such cases are rare, and most can be managed conservatively. Operative correction of both femur and tibia appears necessary to correct such a deformity, and the sheer magnitude of such a procedure generally encourages continued conservative management.

Rotational tibial osteotomies should probably be performed only in a child with a persistent deformity exceeding 15 degrees internal FPA or 30 degrees external FPA at age 8 or older—thereby ensuring that no other rotational change or gait accommodation is likely to occur—and in whom definite functional and psychological disturbances appear to be present. Biomechanical analysis of the child's gait may add objective justification, especially for excessive *external* torsion, in which push-off during stance phase may be impaired owing to the relative shortening of the triceps surae produced by the relative heel valgus and hindfoot pronation. Correction of either medial or lateral tibial torsion is probably best done in the supramalleolar region. This avoids compartment syndrome problems and possible peroneal nerve injury, and most supramalleolar osteotomies can be managed with a short-leg cast, especially if internal fixation is used (Fig. 21–26). Small-fragment DCP plates are preferred to ensure maintenance of rotational alignment during healing, which is occasionally slow in the distal tibia. Rotational osteotomies of the *proximal* tibial region are contraindicated in a child under the age of 8 because of the possibility of a late valgus deformity following the osteotomy in this region, similar to the Cozen's fracture situation (see Fig. 21–23).[76,88]

Bowing of the Tibia

Bowing of the diaphysis of the tibia is usually noted at birth or shortly after, and thus the term "congenital" is appropriate when describing these conditions. Three types of deformity are recognized: (1) anterolateral bowing, associated with the limb-threatening condition pseudarthrosis of the tibia, although a benign form also exists; (2) posteromedial bowing, usually benign; and (3) anterior or anteromedial bowing when there is congenital deficiency of the fibula. In this section, anterolateral and posteromedial deformities will be discussed. When bowing accompanies fibular deficiency (discussed in Chapter 23, Limb Length Discrepancy), treatment is directed away from the tibia per se and more toward the foot and limb length discrepancy.

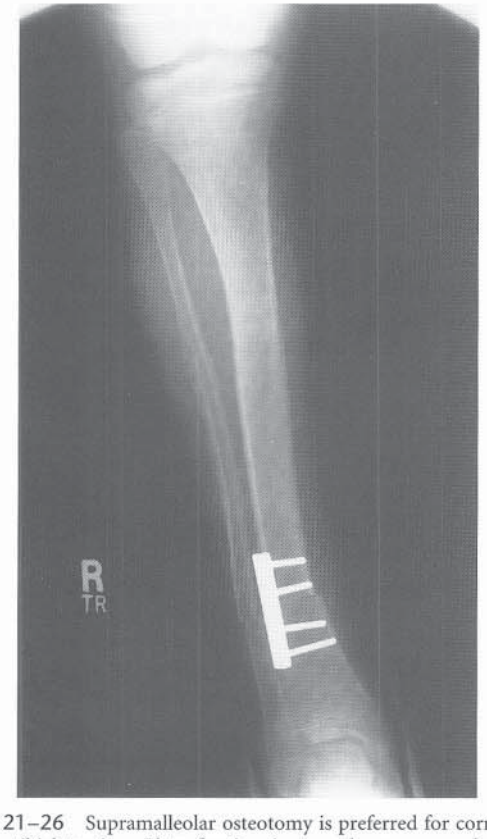


FIGURE 21–26 Supramalleolar osteotomy is preferred for correction of excessive tibial torsion. Plate fixation is strongly recommended due to expected mild delay in solid union at this level.

Congenital Anterolateral Bow of the Tibia

This deformity is closely associated with the development of pseudarthrosis of the tibia. Because most pseudarthroses of the tibia are not present at birth, the term "congenital pseudarthrosis of the tibia" is somewhat inaccurate, but there is no question that the underlying disease process and deformation of the tibia *are* usually present at birth, and it is often merely a matter of time before a first fracture occurs. Management of anterolateral bowing of the tibia and management of the subsequent pseudarthrosis are considered as a continuum. It is striking that such a rare disorder, affecting 1 in 140,000 to 1 in 190,000 population,[3,33] would receive so much attention in the orthopaedic literature. Pseudarthrosis of the tibia poses one of the most challenging treatment problems in all of orthopaedics because of the compounded difficulty of achieving and then maintaining union and simultaneously providing a functional extremity.

ETIOLOGY AND PATHOLOGY

The relationship of anterolateral bowing and neurofibromatosis has been known since 1937,[45] and 5.7 percent of patients with neurofibromatosis type I have the deformity.[33] Up to 55 percent of cases of anterolateral bowing and pseudarthrosis are associated with neurofibromatosis.[3,154,169] Some authors have found anterolateral bowing to be ultimately associated with neurofibromatosis in nearly every instance.[80] Because café-au-lait spots, axillary freckling, or other cutaneous manifestations of neurofibromatosis may not be present in infancy but appear only later, the inability to diagnose neurofibromatosis by NIH criteria at birth, when the anterolateral bowing is first observed, does not mean that the patient may not eventually demonstrate it.[33] The presence of neurofibromatosis does not appear to affect the incidence of union or the ultimate outcome of the tibial pseudarthrosis.[33,142]

Fibrous dysplasia is also seen with anterolateral bowing.[1] Because patients with fibrous dysplasia may also have café-au-lait spots, there is an overlap of the patient population with neurofibromatosis and the patient population with fibrous dysplasia. There may rarely be a relationship between congenital pseudarthrosis and the amniotic band syndrome.[157] Because the pseudarthrosis site typically is surrounded by a rind of thickened hamartomatous tissue that histologically resembles fibrous tissue and appears to constrict the tibia proper, it is not surprising that an amniotic band can produce a pseudarthrosis by an apparent "strangulation" mechanism.

Because of their association with either neurofibromatosis or fibrous dysplasia, one might expect these lesions to be found microscopically in the area of the pseudarthrosis site. Although such a finding has been reported,[62] most investigators have failed to find anything other than thickened fibrous tissue at the pseudarthrosis site,[23] accompanied by a paucity of vascular ingrowth.[66] The fact that the fibrous constriction lesion is universally present has suggested to some investigators that the primary pathologic lesion is in the periosteal structures around the tibia rather than in the bone itself.[1,21] Excision of hamartomatous fibrous tissue around the pseudarthrosis site at the time of surgical treatment is therefore usually emphasized.

CLASSIFICATION

Because of the difficulty in successfully achieving union and the often pessimistic prognosis for union after failed initial attempts, classification systems describing prognostic radiographic characteristics have been proposed by many authors.[3,19,33,157] These classification systems generally describe the untreated appearance of the bone at the pseudarthrosis site (e.g., sclerotic, cystic, dysplastic, or "hourglass" constriction), or may describe the presence of a fracture at birth or the presence of a pseudarthrosis in the fibula. Unfortunately, none of the classification systems provides guidance to management or eventual outcome, primarily because successful union is often a transient phenomenon, with refracture necessitating starting all over with a new round of treatment whose outcome is equally unknown. For this reason, I have relied on two very simple criteria to initially classify anterolateral bowing of the tibia: (1) the presence or absence of fracture, and (2) the age at which fracture first occurs ("early onset" before 4 years of age, "delayed onset" after 4 years[91]). These two features provide the clinician with essentially all the information required to make appropriate treatment decisions initially. An example of the inadequacy of other classification systems is the irrelevance of the type of tibial lesion once fracture has occurred, as it makes no difference whether the prefracture lesion was sclerotic, cystic, an hourglass constriction, or associated with a pseudarthrosis of the fibula. One exception may exist: the type I, or nondysplastic tibia in Crawford's classification,[33] may represent a benign form of anterolateral bowing.[163] Patients with type I can usually be followed without bracing and may never have a fracture.[33] (Crawford's type II is divided into three subtypes: IIA has bowing with failure of tubularization and a widened medullary canal; type IIB has anterolateral bowing with a cystic lesion before fracture or canal enlargement from previous fracture; type IIC has a pseudarthrosis and bone atrophy with "sucked candy" narrowing of the bone ends.)

The benign form of anterolateral bowing, characterized by subperiosteal bone formation in the concavity of the bowed tibia and lack of involvement of the fibula, is not strictly equivalent to the type I described by Crawford. Because of the observation that fracture does not occur and that bracing is unnecessary, these two entities are clinically alike. Thus, Crawford's type I anterolateral bow is indeed prognostic if it truly describes the resolving form of anterolateral bowing, which does not require treatment (see-below).[163]

CLINICAL FEATURES

Anterolateral bowing of the tibia is frequently noted at birth. The deformity is evident from the apical prominence laterally in the leg, with the foot inverted or at least medially displaced in relation to the lower leg. If neonatal fracture has occurred, motion at the pseudarthrosis site will be evident. Because the vast majority of anterolateral deformities are unilateral, the shortening and angulation are easily appreciated when the affected leg is compared with the normal leg. If signs of neurofibromatosis are present, the diagnosis is readily apparent.

On the other hand, a milder deformity unaccompanied by signs of neurofibromatosis may not come to medical attention until much later, when a limp due to deformity or an impending fracture draws attention to the leg. The foot and ankle may be normal or slightly smaller than on the contralateral side, suggesting as an initial diagnosis a neuropathic foot problem such as seen with a unilateral dysraphic lesion. Again, the presence of cutaneous manifestations of neurofibromatosis will rapidly clarify the diagnosis.

The late form of pseudarthrosis[133] presenting as a fracture in an older child will probably be accompanied by none of these findings, even retrospectively. The diagnosis is made radiographically when the child presents with a fracture.

TREATMENT

Except for the resolving form,[163] the natural history of anterolateral bowing is extremely unfavorable, and once fracture occurs, there is little tendency for the lesion to heal spontaneously (Fig. 21–27). Regardless of the treatment method used—internal and external fixation, distraction osteogenesis, bone grafting with or without microvascular transfer of bone, electrical stimulation (either direct current by implantation or via pulsed electromagnetic fields)—there is general pessimism as to the quality and longevity of any union that may be obtained, and the ultimate future function of the leg is uncertain. Although many treatment options exist, the fact that no one option has ever achieved long-lasting success with great frequency indicates that there is no single treatment for pseudarthrosis of the tibia that will produce acceptable results in any predictable fashion. However, based on the experience of multiple investigators and my own experience, there are two basic treatment principles that must be emphasized in order to have any hope of a good functional outcome: (1) the *alignment* of the leg must be maintained, and (2) *permanent* intramedullary fixation to maintain such alignment or to provide internal bracing for a united tibia is desirable. To these ends, treatment should be directed toward correcting the anterolateral deformity once fracture has occurred, and toward aggressively maintaining alignment by intramedullary fixation both during growth and at maturity, in an effort to avoid late fracture and possible failure from nonunion.

Once the diagnosis of nonresolving anterolateral bowing of the tibia has been made, the first step is to prevent fracture if possible. In the infant, prior to walking age, no specific treatment is necessary other than education of the caretakers. Once the child begins weightbearing, prophylactic bracing should be attempted, although there is no documentation that such a program can prevent fracture.[142] A clamshell-

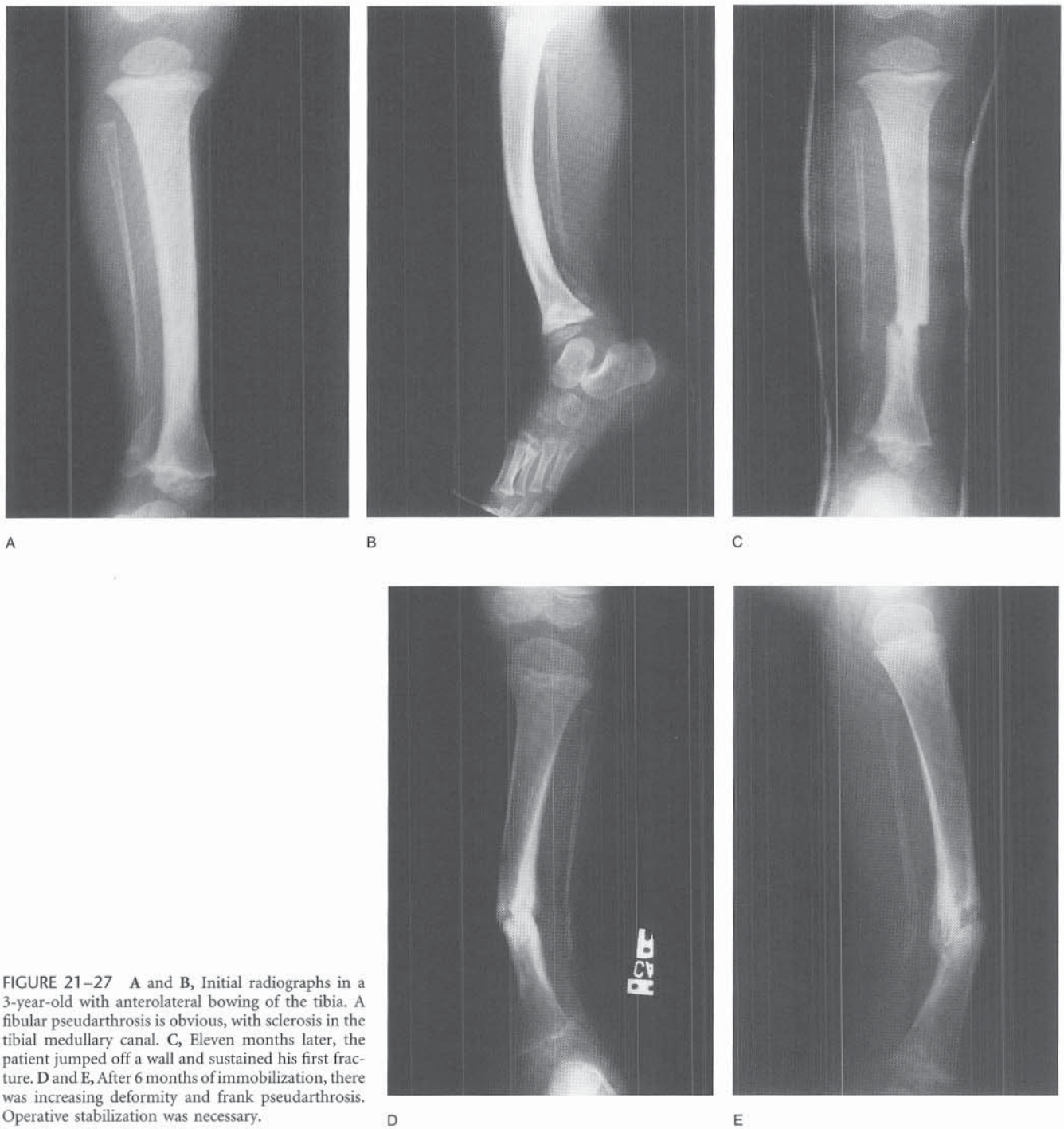


FIGURE 21–27 **A** and **B,** Initial radiographs in a 3-year-old with anterolateral bowing of the tibia. A fibular pseudarthrosis is obvious, with sclerosis in the tibial medullary canal. **C,** Eleven months later, the patient jumped off a wall and sustained his first fracture. **D** and **E,** After 6 months of immobilization, there was increasing deformity and frank pseudarthrosis. Operative stabilization was necessary.

type fracture orthosis that provides circumferential support for the tibial diaphysis is recommended. For later functional considerations,[91] a free ankle joint should be utilized if at all possible, depending on the proximity of the apex of the diaphyseal bowing to the center of rotation of the ankle. Protection of the unfractured bowed tibia may be necessary and should be continued indefinitely until fracture occurs or the patient approaches skeletal maturity (Fig. 21–28).

Prophylactic bypass grafting of the intact deformity has been advocated by Strong and Wong-Chung,[154] modifying the posterior bypass graft originally described by McFarland.[114] Tachdjian, in the second edition of this textbook, proposed that a *delayed* McFarland graft be utilized from the *opposite* tibia, with an interval of 4 to 6 weeks between the initial raising of the graft from the contralateral normal tibia and its subsequent harvesting from the original bed and transfer to the affected side.[157] By raising a cortical graft and replacing it in its original bed, then returning 6 weeks later to harvest it, the osteogenic potential of such a delayed graft was believed to enhance the healing and subsequent hypertrophy of the graft placed posteriorly in the weight-bearing axis of the prepseudarthrotic deformity. Although

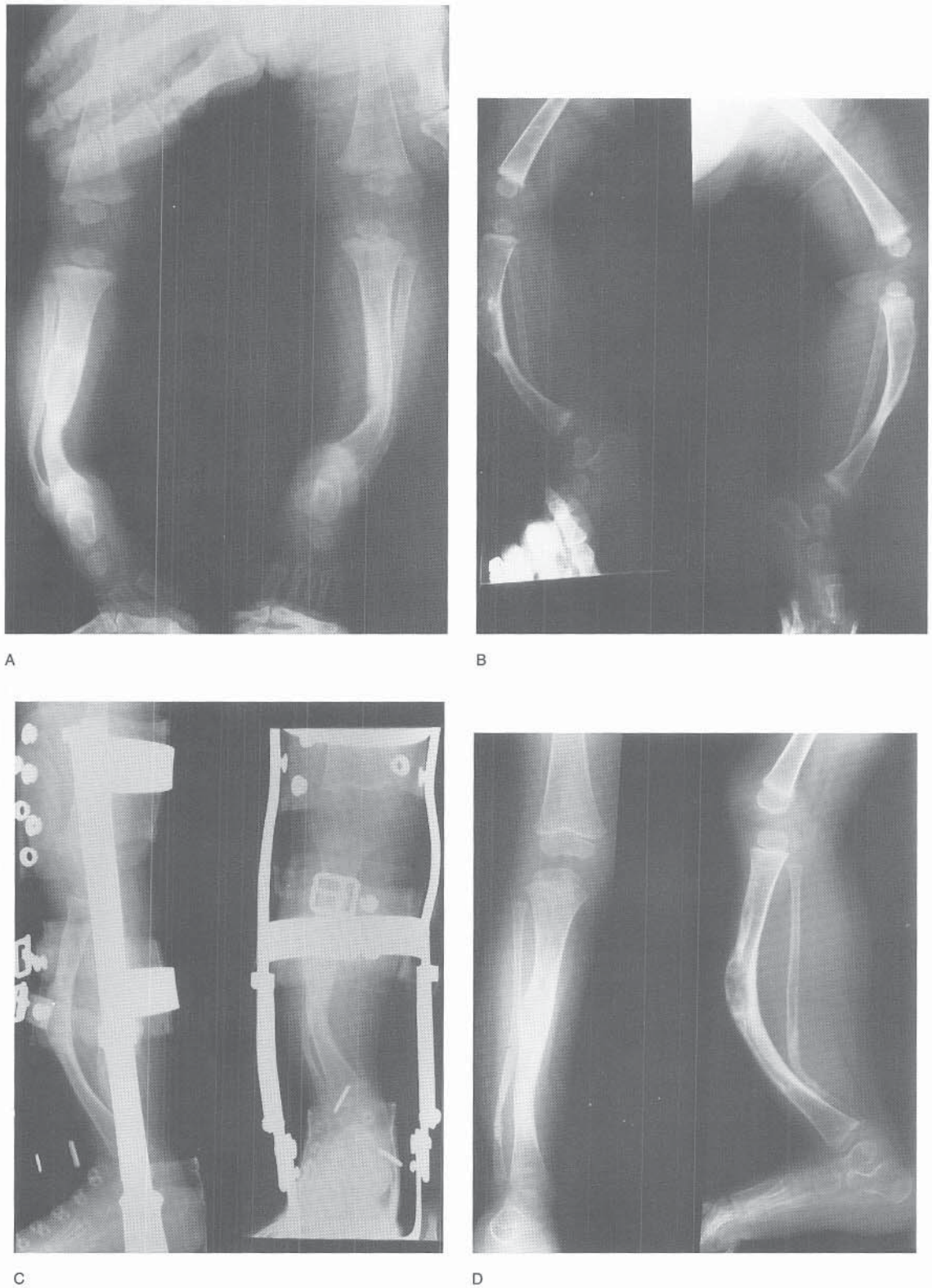
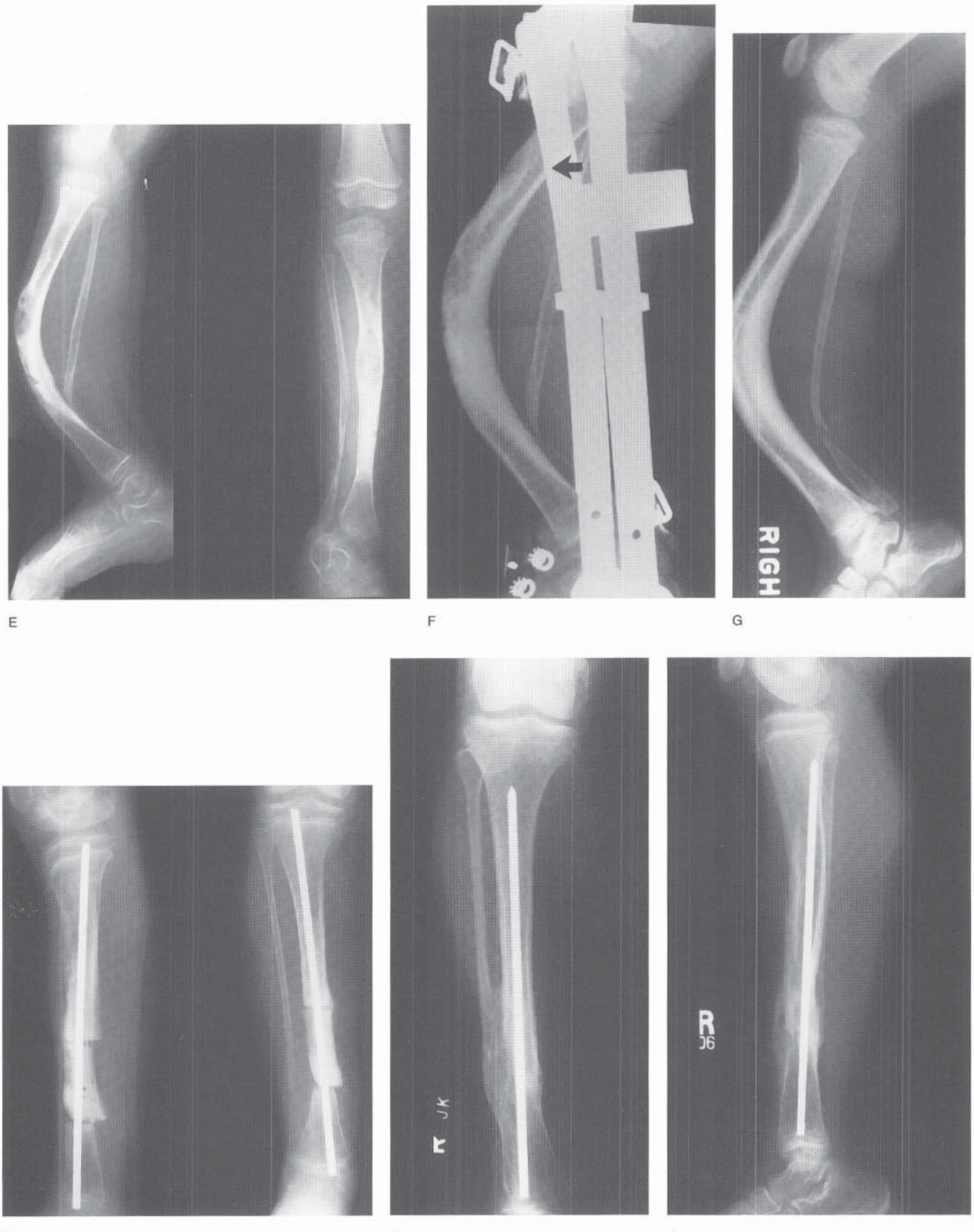


FIGURE 21–28 **A** and **B,** Newborn radiographs in a girl with neurofibromatosis type 1. The tibias are bowed bilaterally. **C,** Prophylactic bracing bilaterally was instituted in 1974 at walking age with KAFOs, using thigh lacers and free ankle joints. **D,** At age 2 the right fibula has fractured, but the tibia remains intact. Bracing was continued.



E F G

H I J

FIGURE 21–28 *Continued.* **E,** At age 3, the right tibia fractured for the first time. The fracture united after several months in a plaster cast. **F,** The right tibia fractured again at age 4 years 6 months (*arrow*). It again healed with casting. **G,** At age 9, the right tibia remained intact, but the patient rejected further bracing, and because of the deformity, osteotomy was performed. The left tibia never fractured. **H,** Appearance 4 months postoperatively. Delayed union of both osteotomy sites occurred. The tibia was bone grafted and re-rodded. **I** and **J,** Final outcome at age 17. The tibia has remained united for 7 years and the patient is fully ambulatory.

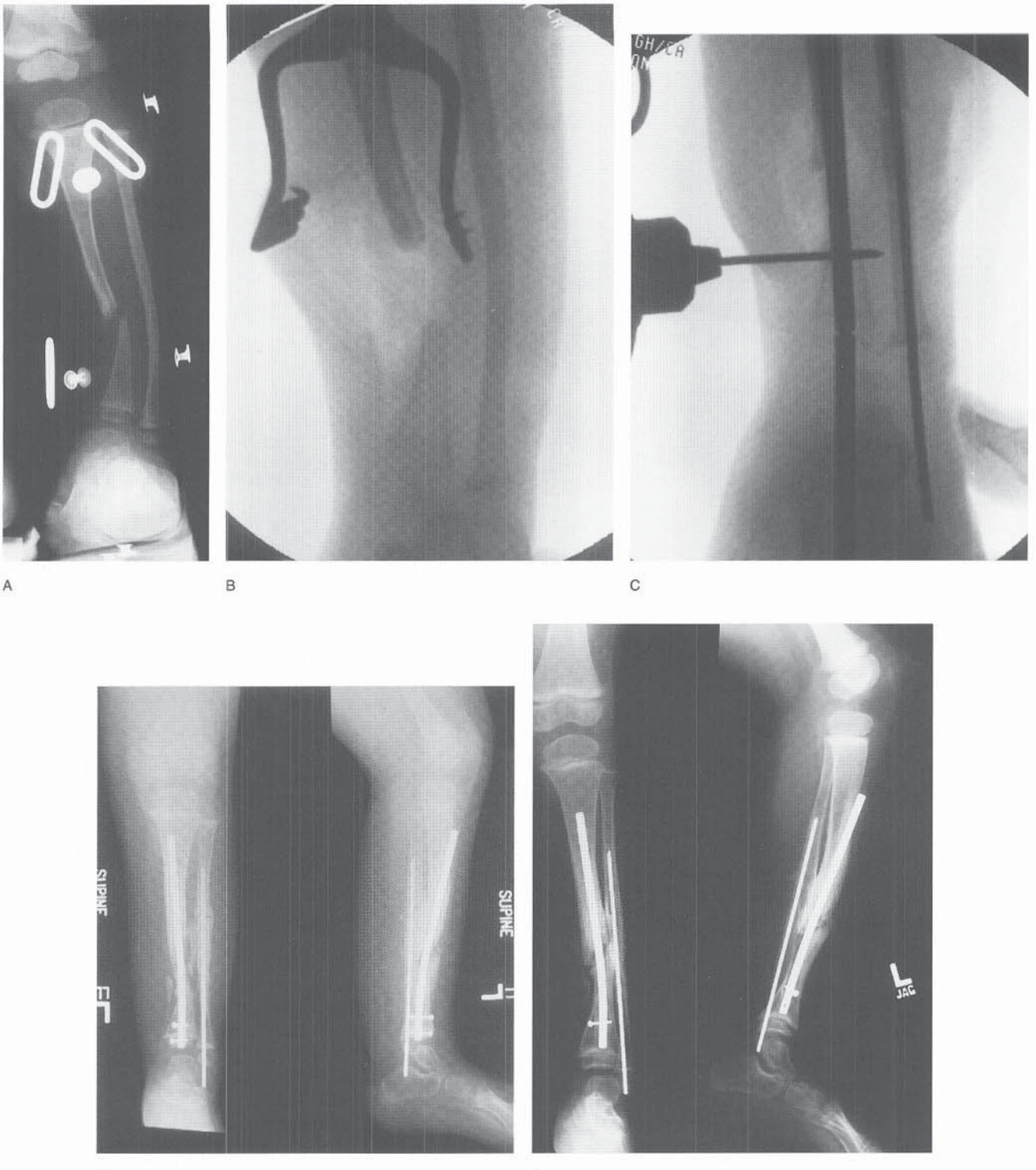
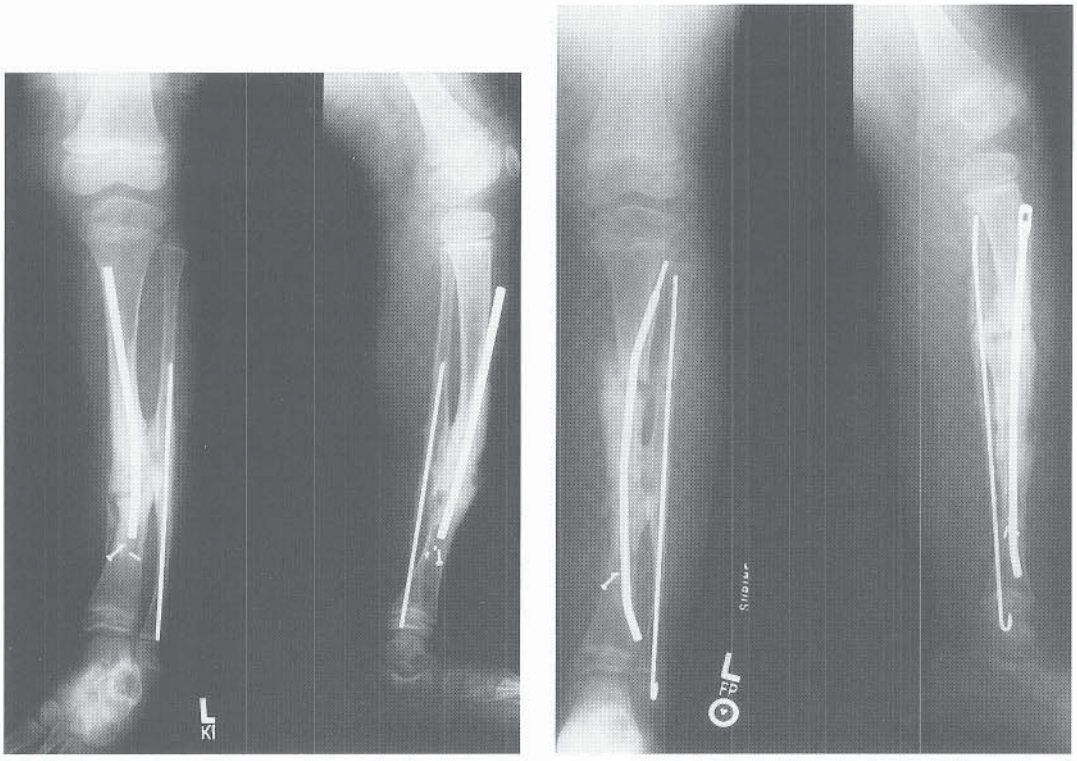


FIGURE 21–29 **A,** Radiographic appearance in an 18-month-old boy with neurofibromatosis type 1 and pseudarthrosis of the tibia only. Bracing allowed full weightbearing activity despite the gross pseudarthrosis. **B** and **C,** At age 3 years 2 months, the pseudarthrosis was widely excised and the fibula shortened. The tibia was fixed with an intramedullary rod and a distal interlocking screw (2.0 mm) so that the rod would not be left across the ankle. The junction of the male and female portions of the Williams rod is seen just proximal to the ankle. The male portion was unscrewed and removed from the plantar surface of the foot. **D,** Six months later, the tibial fixation was revised because of screw migration and loss of distal interlocking. The revision was accomplished from a proximal entry point only, so that the pseudarthrosis site and bone grafting would not be disturbed. Two interlocking screws were placed distally, including one in the epiphysis. **E,** After 3 months, the epiphyseal fixation screw was removed to allow the rod to migrate proximally. In retrospect, the metaphyseal screw should have been removed to allow the rod to migrate distally with epiphyseal growth and thus maintain intramedullary fixation across the pseudarthrosis site longer.



F G

FIGURE 21–29 *Continued.* **F,** Radiographic appearance 2½ years later. The rod has lost its distal interlocking because of broken screws, and recurrent lateral bowing is evident with proximal rod migration. The patient was fully ambulatory without bracing and asymptomatic. The fixation, however, was no longer functioning. **G,** Radiographic appearance 6 months following the second revision. A proximal osteotomy to correct procurvatum was performed to realign the tibia in all planes. The patient was again fully ambulatory without activity restrictions.

Tachdjian reported 100 percent success in his own cases, other reports have been much less sanguine[51,104,114] about either achieving union or preventing amputation. Such a bypass graft has two unattractive features: the normal leg is disrupted, and no attempt to correct the deformity is made. If the patient is to undergo a grafting procedure with a nonvascularized free bone graft, that procedure should be given a greater opportunity to succeed by also correcting the pathologic mechanical axis of the tibia. Once fracture has occurred, the McFarland-type bypass probably has little indication, as the deformity should then be treated aggressively with excision of the pseudarthrosis and mechanical realignment performed simultaneously with cancellous bone grafting.

Thus, prophylactic treatment is primarily orthotic, and should fracture occur before age 4 years (early onset) it is my view that bracing should continue in an effort to allow further growth and hypertrophy of the tibial fragments, particularly the distal one (Fig. 21–29). Fracture frequently occurs insidiously, with no acute pain or inciting event, and although motion at the pseudarthrosis site may be appreciated on physical examination, the patient is still able to bear weight in a well-molded fracture orthosis. Because the surgical outcome in very young patients is likely to be worse,[91] we believe every effort should be made to delay the first surgical attempt at gaining union so that the tibia can enlarge, the amount of autogenous bone available for grafting is greater, and a more secure intramedullary fixation of both tibia and fibula may be possible. However, there is no consensus on the appropriate age for first surgery. Some suggest that the ultimate outcome is improved when union is obtained as early as possible,[4,156] because the associated deformities and shortening of the leg can worsen with growth in the absence of sound stabilization. The fact remains, however, that most surgical series report difficulty in achieving union in infants and young children,[19] and the likelihood of eventual amputation is magnified the earlier that the first procedure is attempted.[91]

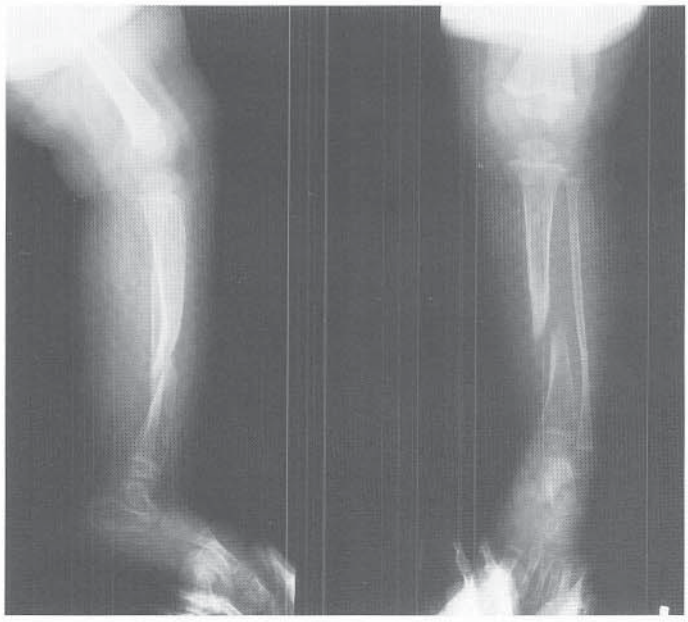
Intramedullary Fixation. The procedure of choice for the first attempt to gain union is thorough resection of the pseudarthrosis, shortening and fixation with an intramedullary rod, and autogenous bone grafting. Rates of union of 90 to 100 percent have been reported with this approach, although solid, lasting union without deformity is another matter. As suggested earlier, because maintenance of intramedullary fixation indefinitely is desirable in most circumstances, this also achieves the goal of maintaining alignment by internal splintage, and in cases with tenuous union it may aid in the effort to prevent refracture. Charnley originally described the technique, which is now most commonly associated with the names of Peter Williams[170] and Sherman Coleman.[13]

Williams conceived the novel approach of threaded male and female components to the rod (Fig. 21–29) which, when joined, can be placed antegrade through the distal

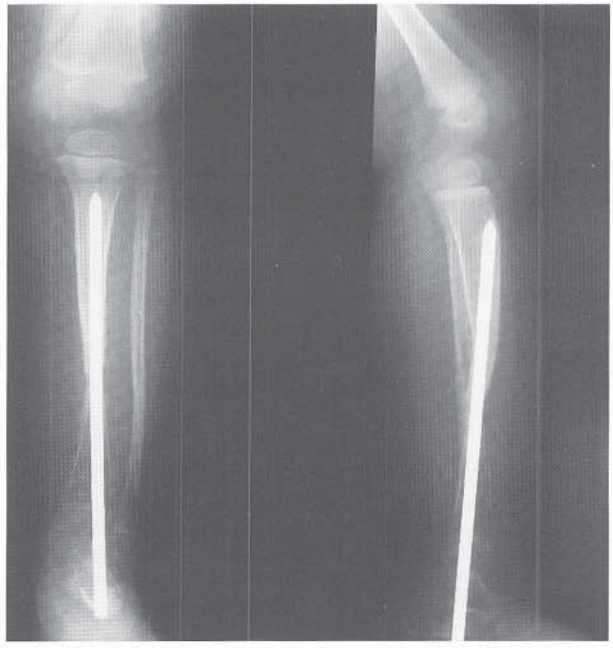
pseudarthrosis site and out the bottom of the foot. After retrograde insertion back into the proximal portion of the tibia, the male end is unscrewed and removed from the bottom of the foot, leaving the female-threaded rod intraosseously in the tibia and talus/calcaneus (Fig. 21–30).

The surgical technique begins with adequate anterior exposure of the pseudarthrosis site, appropriately extensile to allow aggressive longitudinal resection of the fibrotic rind of periosteum and hamartoma circumferentially. Bone should be removed back to normal-appearing medullary canal. Fasciotomies of all compartments are performed. The fibula should be resected through a separate lateral approach if it has a pseudarthrosis. An intact fibula should be shortened to allow impaction of the tibia ends. Failure to shorten an intact fibula means that the resected ends of the tibia will be held in distraction, an almost certain invitation to nonunion (Fig. 21–30). With dissection along the intraosseous membrane the two resection sites should be connectable, so that the bone graft can be placed between them to attempt cross-union at the pseudarthrosis site (Figs. 21–29 and 21–31).

The male-female combined rod is now drilled antegrade down the distal tibial fragment and out the bottom of the foot (see Fig. 21–29C). The position of the distal fragment and ankle is critical, as the foot should be in neutral dorsiflexion and the ankle and subtalar joint in neutral or slight



A



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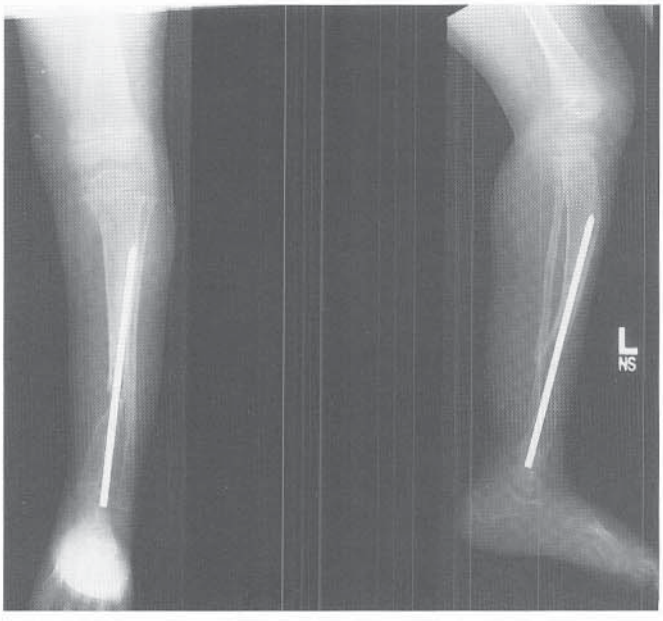
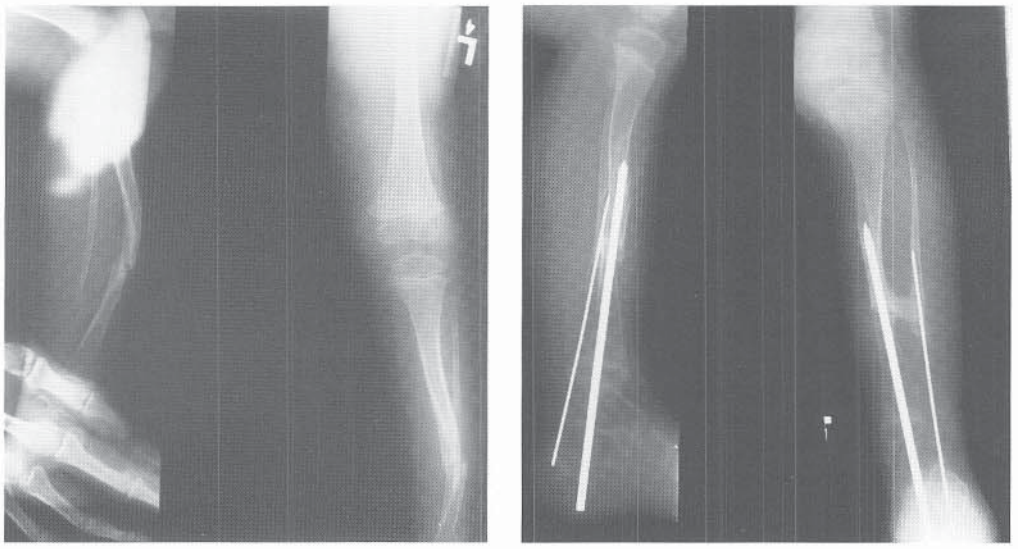
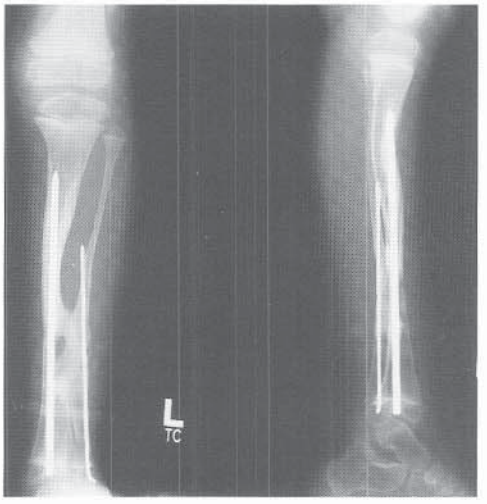


C

FIGURE 21–30 **A,** Radiographic appearance in a 1-year-old child with a nonhealing fracture of the tibia that occurred at age 6 months. **B,** At age 1 year 10 months, Williams rodding and bone grafting were performed *without* a fibular shortening osteotomy. **C,** Appearance 2½ years postoperatively. The rod has migrated into the tibia medullary canal, releasing the ankle from fixation. There is no sign of union across the pseudarthrosis, and anterior bowing at the proximal end of the rod is now apparent. Revision surgery to obtain union and correct alignment will be necessary.



A B



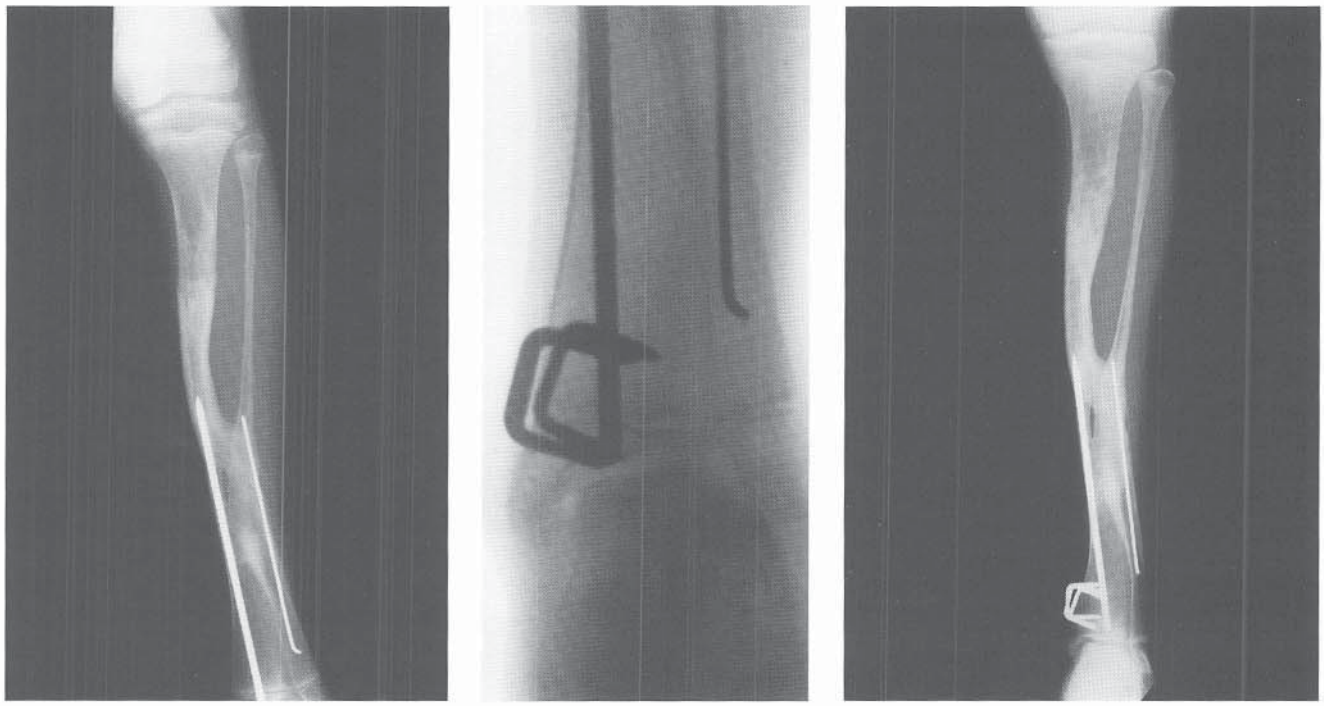
C

FIGURE 21–31 **A,** Radiographic appearance in a 3-year-old child with neurofibromatosis type 1 who sustained a first fracture. He had been braced for 1 year prior to fracture. **B,** Williams rodding and fibular shortening with bone grafting was performed. At 8 months postoperatively the bone appeared to be united, but the rods were migrating out the bottom of the foot. **C,** Two years following rod reinsertion in the tibia by impacting from below. The fibular rod was replaced similarly. The child was fully ambulatory without restriction at age 6.

Illustration continued on following page

varus, to counteract the inevitable valgus that subsequently develops at the ankle. The resection/osteotomy is now shortened and reduced, and the combined rod is reverse-drilled retrograde up the proximal tibial medullary canal. The female rod end ideally should be placed finally in the talus, and the male "inserter" end is unscrewed and removed from the bottom of the foot. By pushing on the plantar aspect of the foot, the surgeon can achieve final impaction of the resection site. The fibula should also be rodded, usually with a smooth K-wire, which can be placed using the same antegrade technique as the tibial rod. The fibula rod can also be placed retrograde from the tip of the distal fibula, after tibial reduction or impaction is achieved. A 180-degree bend in the end of the fibular rod is recommended so that this "hook" can be impacted into the end of the distal fibula to avoid distal migration and rod prominence at the lateral side of the ankle (see Figs. 21–29G and 21–31C). Copious bone grafting around the pseudarthrosis site and over to the fibular site completes the procedure. Postoperative immobilization in a long-leg bent-knee cast or a hip spica cast[13,29] is usually maintained for 6 to 8 weeks, followed by a long-leg weightbearing cast, with the knee in full extension, for an additional 1 to 2 months. Because the ankle joint and sometimes the subtalar joint are crossed by the intramedullary rod, hindfoot immobilization in a rigid AFO is necessary, sometimes supplemented by an anterior tibial shell to add further protection against rotation and bending.

The undesirable effect of ankle immobilization by intramedullary fixation is thought to be necessary to adequately immobilize the small distal fragment.[13,29] As the tibia grows, the foot and ankle may eventually grow off the distal end of the intramedullary rod, allowing the ankle to regain motion. Because indefinite intramedullary fixation within the tibia is desirable and continued even if union is achieved, the rod may need to be replaced at a later date with a longer rod should a new pseudarthrotic lesion at the upper or lower end of the rod develop. Union can be initially achieved,[4] but refracture can occur above or below the rod, requiring



D E F

FIGURE 21–31 *Continued.* **D,** At age 11, marked diaphyseal valgus developed. **E,** Medial distal tibial stapling was performed. It was not known at that time whether the tibial physis was functional or not. **F,** Appearance at age 13. The valgus of the distal half of the tibia was corrected and the staples were removed. Although the alignment is clinically excellent, there is concern about the upper half of the tibia, which has no intramedullary fixation. Observation continues.

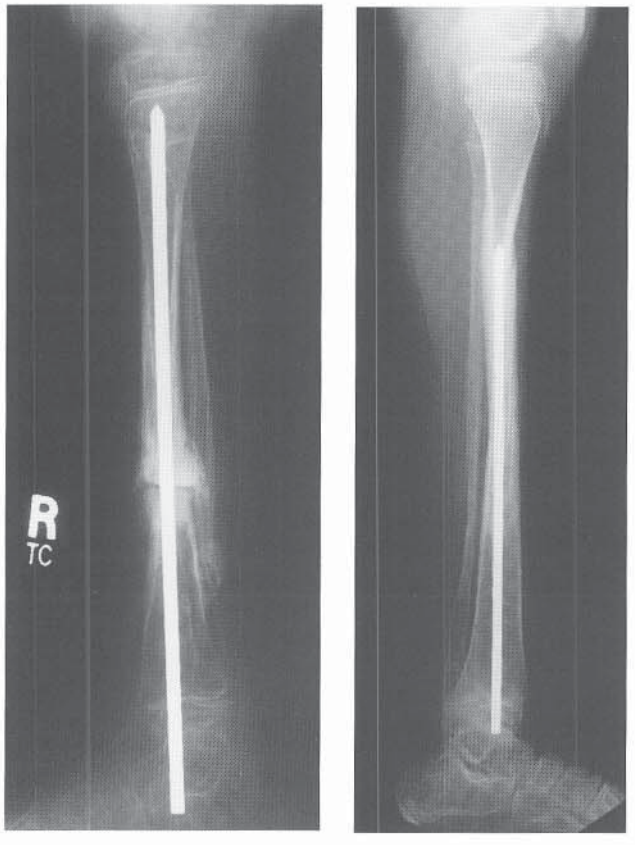
reinsertion with a longer intramedullary device as well as regrafting. It is not uncommon that up to four operations are required in order to finally obtain solid union (see Figs. 21–29 and 21–31).[4,13,91]

Not only is there a possibility of refracture, but the appearance of a valgus deformity somewhere in the leg is almost certain despite the achievement of a solid union (Fig. 21–31). Most commonly the ankle drifts into valgus, owing either to a relative growth inhibition of the lateral portion of the distal tibial physis[114] or to insufficient growth of the distal fibula, especially with persistent pseudarthrosis of the fibula. An additional explanation for the ankle valgus probably includes a muscular imbalance etiology, similar to the ankle valgus seen in patients with spina bifida who lack S1-innervated plantar flexion. Because of transfixation of the ankle by an intramedullary device, or perhaps because of endless immobilization and bracing to protect the tenuously united tibia, there is marked triceps surae atrophy and weakness, not unlike the paralysis seen in the patient with spina bifida. Thus, ankle valgus in response to calf atrophy and triceps surae insufficiency may be the inevitable result of *any* treatment of pseudarthrosis of the tibia. Should the ankle valgus become progressive, treatment of this deformity is strongly recommended during the growth of the limb, usually by temporary or permanent hemiepiphysiodesis of the medial malleolar physis. Staples or screw fixation[150] can be introduced to tether the medial side of the distal tibial physis, or, alternatively, a permanent hemiepiphysiodesis, if appropriate at a more advanced age, can be performed. Distal synostosis of the tibia and fibula[95] has been recommended,[156] but because this procedure requires additional bone grafting and immobilization in order to achieve such a synostosis, I prefer to perform a medial hemiepiphysiodesis (Fig. 21–31), which does not require immobilization and casting. Supramalleolar osteotomy of the distal tibia to correct ankle valgus is probably the least attractive treatment option because of the possibility of a iatrogenic pseudarthrosis being produced should the osteotomy fail to heal. Up to 50 percent of patients undergoing a corrective supramalleolar osteotomy can develop recurrent pseudarthrosis (see Fig. 21–35).[13]

Limb length discrepancy is yet another untoward event, with shortening at maturity averaging as much as 5 cm.[109] Obvious causes of shortening of the tibia include acute shortening at the time of pseudarthrosis resection as well as damage to the distal tibial physis from multiple operations and intramedullary device crossing. The distal tibial epiphysis may simply fail to grow as a consequence of the condition or its treatment.[114] A more subtle cause of limb length discrepancy is the generalized lack of stimulus to growth caused by immobilization of the ankle by intramedullary fixation and indefinite orthotic management. Appropriate and judicious attention to contralateral epiphysiodesis at an appropriate time is the best management for a discrepancy of 5 cm or less. It goes without saying that limb lengthening of a previously united pseudarthrotic tibia may be complicated by a new iatrogenic pseudarthrosis at the lengthening site,[29,156] although this has not been reported as frequently as might be expected using Ilizarov's method (see Fig. 21–35).[16,58,66,119]

The most common untoward result affecting a pseudarthrotic tibia is probably a stiff ankle and subtalar joint producing a poorly functioning foot. Objective follow-up studies using gait analysis and muscle testing of united pseudarthrotic tibias have confirmed what was suspected subjectively for some time: the cost of obtaining and maintaining tibial union is a stiff, atrophic ankle and foot that function little better than a prosthesis.[91]

Stiffness is predictably a result of intramedullary fixation crossing the two joints and of the prolonged immobilization, by cast or by orthosis, to prevent articular damage, rod fracture, or loosening (Fig. 21–32). A functional calcaneus gait occurs because of the significant atrophy and weakness of the plantar flexors.[91] For these reasons, attempts to achieve intramedullary fixation *without* ankle transfixation are indicated, and the results are encouraging. Custom-made interlocking Williams-type rods can be fabricated in a hospital machine shop, allowing the surgeon to interlock a small-diameter nail with 2.0- or 2.5-mm screws and avoid ankle transfixation. An additional benefit is improved rotational stability. Patients are completely ambulatory without an external orthosis and with a normally functioning ankle joint (see Fig. 21–29). Even if union is not achieved at the first operation, effective internal splinting of the tibia without immobilizing the ankle allows the patient a functional leg while growth and hypertrophy of the tissues proceed, setting the stage for a repeat attempt at a later date, when the odds are more favorable that union will be achieved.



A B

FIGURE 21–32 **A,** Williams rodding of the tibia in a 7-year-old with late-onset pseudarthrosis and neurofibromatosis type 1. In retrospect, the distal tibial fragment was large enough to accept an interlocked nail, and ankle transfixation could have been avoided. **B,** Appearance 3 years postoperatively. There is excellent union. However, the ankle is stiff and painful, restricting activity. Damage to the talar dome and distal tibia is occurring because of failure of the rod to migrate completely into the tibial medullary canal. Retrieval of this rod is problematic.

Vascularized Fibular Graft. The transfer of normal bone on a microvascular pedicle would at first glance appear to revolutionize the treatment of pseudarthrosis of the tibia. The failure of nonvascularized bone grafts to achieve definite and lasting union is well documented. The first use of a vascularized transfer of bone tissue, reported by Farmer, was a delayed cross-leg pedicle flap that brought normal vascularized bone from the opposite tibia to the pseudarthrosis site. The success of this procedure was as striking as the cosmetic derangement of the contralateral donor leg.[114] Microvascular transfer of the contralateral, or sometimes ipsilateral, fibula is now the more standard procedure, providing normal tissue with its own blood supply to the lesion.

The special talents of a microvascular surgical team are required, and preoperative arteriography is performed to determine anastomosis sites and rule out anomalies. Often two operative teams work simultaneously, one team harvesting the microvascular fibular graft while the second team resects the pseudarthrosis site, removes previous hardware, and prepares the recipient bed. Technical problems of solid fixation of the transferred fibula to the recipient distal fibula are common but can be overcome by the use of external fixation or limited internal fixation (Fig. 21–33). Intramedullary fixation of the donated fibula is thought to be contraindicated because of possible disturbance of the blood supply of the microvascular graft, although some authors recommend it for distal fixation.[42] The success rate of microvascular transfer is 92 to 95 percent if union alone is evaluated.[42,59,124,165,167] However, additional operative procedures for nonunion at one end of the transferred fibula, stress fracture, and other deformities related to the pseudarthrosis are required in every patient.[167] Osteotomy and lengthening have been reported to be successful following union of a microvascular fibular transfer,[59,167] so that the goal of replacing the pseudarthrotic site with more normal tissue appears to be realized.

Less attention has been paid to the morbidity of the donor leg. This is a major consideration that cannot be dismissed lightly. The distal fibula of the donor site must be synostosed to the tibia, or the fibula reconstructed with bone graft, to prevent ankle valgus. Also, the weakness that may ensue in the donor leg because of resection of the origin of some of the flexor muscles is clinically relevant. Late corrective osteotomy of the donor site has been necessary[165,167] to correct more severe ankle valgus. Thus, the place of microvascular fibular transfer from the contralateral extremity remains somewhat controversial and within the domain of relatively few treatment centers.

The use of the *ipsilateral* fibula, described by Coleman and Coleman,[29] has significant advantages. The ipsilateral fibula is transferred to the tibia with its vascular pedicles intact and is cerclaged to both fragments of the tibial pseudarthrosis (Fig. 21–34). In Coleman's series, a 100 percent union rate was achieved, with avoidance of surgery on the intact limb. Coleman now recommends this reconstruction

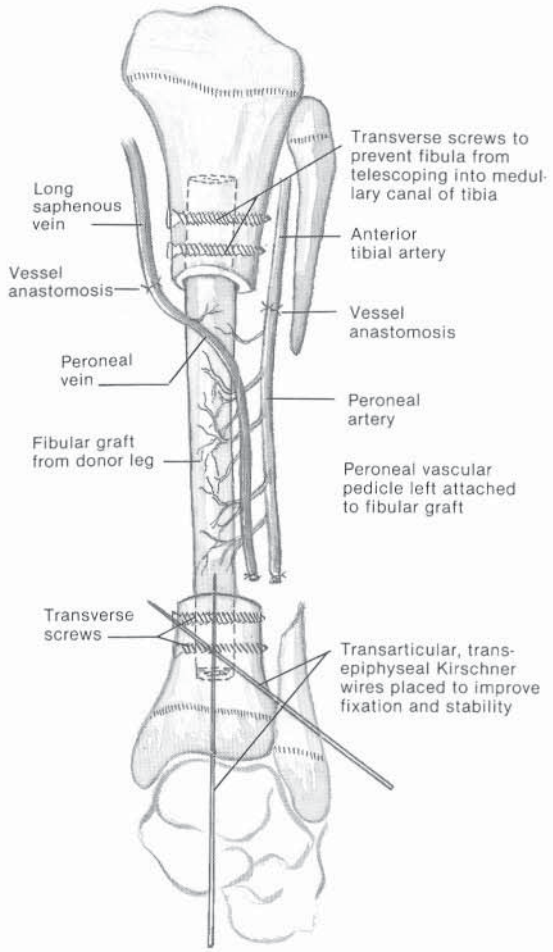


FIGURE 21–33 Free vascularized fibular transfer. (After Pho RWH, Levack B, Satku K, et al: Free vascularized fibula graft in the treatment of congenital pseudarthrosis of the tibia. J Bone Joint Surg 1985;67-B:64.)

as a second-line procedure if intramedullary rodding and conventional bone grafting fail to achieve union.[13] The only technical prerequisite for the successful transfer of the ipsilateral fibula is that the fibular pseudarthrosis, if present, must be at the same level as or distal to the tibial pseudarthrosis, so that the transferred fibular segment will span this pseudarthrosis. In early follow-up, Coleman reported rapid hypertrophy and impressive union of the transferred fibula, but, as always, the function of the extremity will need to be assessed later if the union is maintained.

Electrical Stimulation. Electrical stimulation of bone has several effects that are directly applicable to congenital pseudarthrosis. Bassett and associates[7,8] in early research documented increased calcification of fibrocartilage, increased angiogenesis, and decreased osteoclastic resorption using pulsed electromagnetic fields (PEMFs). Direct current stimulation via an implanted electrode has also been used.[122]

Electrical stimulation is basically an adjunct to conventional bone grafting and internal fixation, and its prophylactic use to prevent fracture is undocumented. Its place in the treatment armamentarium is confounded by inability to separate the effect of the stimulation from the effect of other treatment modalities being used. Electrical stimulation does not correct coexisting deformities, and so its application is probably limited to the earlier phases of pseudarthrosis treatment, when obtaining union is the primary goal.

External Fixation and Distraction Osteogenesis. The application of distraction osteogenesis techniques (Ilizarov method) to pseudarthrosis of the tibia has been widely reported.[16,52,58,65,66,119,127] Because all deformities resulting from the condition can be addressed, it is a natural application of the method. Most commonly a bone transport technique or an acute resection, alignment, and compression with proximal lengthening are utilized (Fig. 21–35).[16,119]

Evaluation of union rates and function following treatment with the Ilizarov method is confounded to a great degree by the poor outcomes of failed previous treatments. Reports of nearly 100 percent union[52,65,119,127] with simultaneous correction of both deformity and shortening have led to tremendous enthusiasm for this treatment method. Recent reports[16,58,66] have tempered the initial wave of enthusiasm, however, because refractures, growth disturbances, and poor foot and ankle function (especially if the foot is included in the frame) remain contentious problems (Fig. 21–35). In addition, even distraction osteogenesis has been unable to achieve union in some tibias or to regain union following refracture in others. Many patients described in the literature, however, were treated after failure of previous conventional bone grafting/fixation and were undergoing an Ilizarov procedure as a final treatment attempt before amputation.

Attempts to achieve union with distraction osteogenesis in toddlers or young children generally do not succeed as

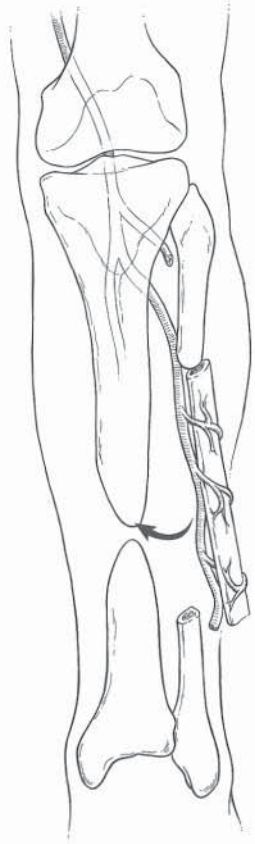


FIGURE 21–34 Transfer of ipsilateral fibula on its vascular pedicle.

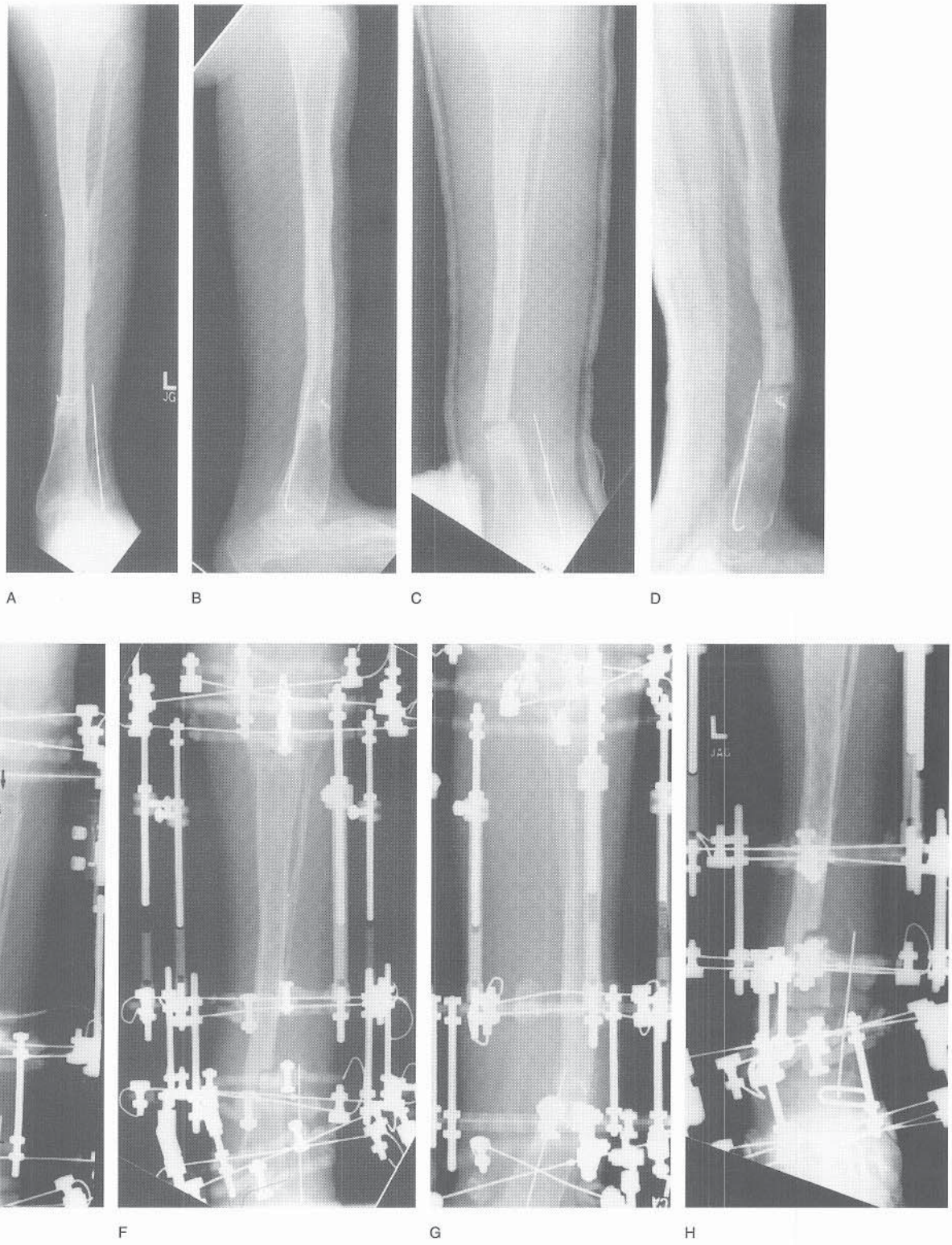


FIGURE 21–35 **A** and **B,** Radiographic appearance in a 13-year-old girl with neurofibromatosis type 1 who had undergone two intramedullary rod placements (at ages 2 and 6) and an Ilizarov procedure (at age 12) to obtain this straight tibia. Although she had poor foot function, she was ambulatory without a brace. **C** and **D,** A pathologic fracture at age 14 years 6 months was treated by another frame to correct deformity distally and compress via a proximal corticotomy-bone transport. **E,** Appearance after proximal lengthening and step-cut compression at the distal fracture site (*arrows*). **F** and **G,** After 4 months the proximal corticotomy had healed but the distal site remained ununited, with valgus and procurvatum deformities. **H,** A new corticotomy was made distal to the nonunion, with provision for angular correction of valgus.

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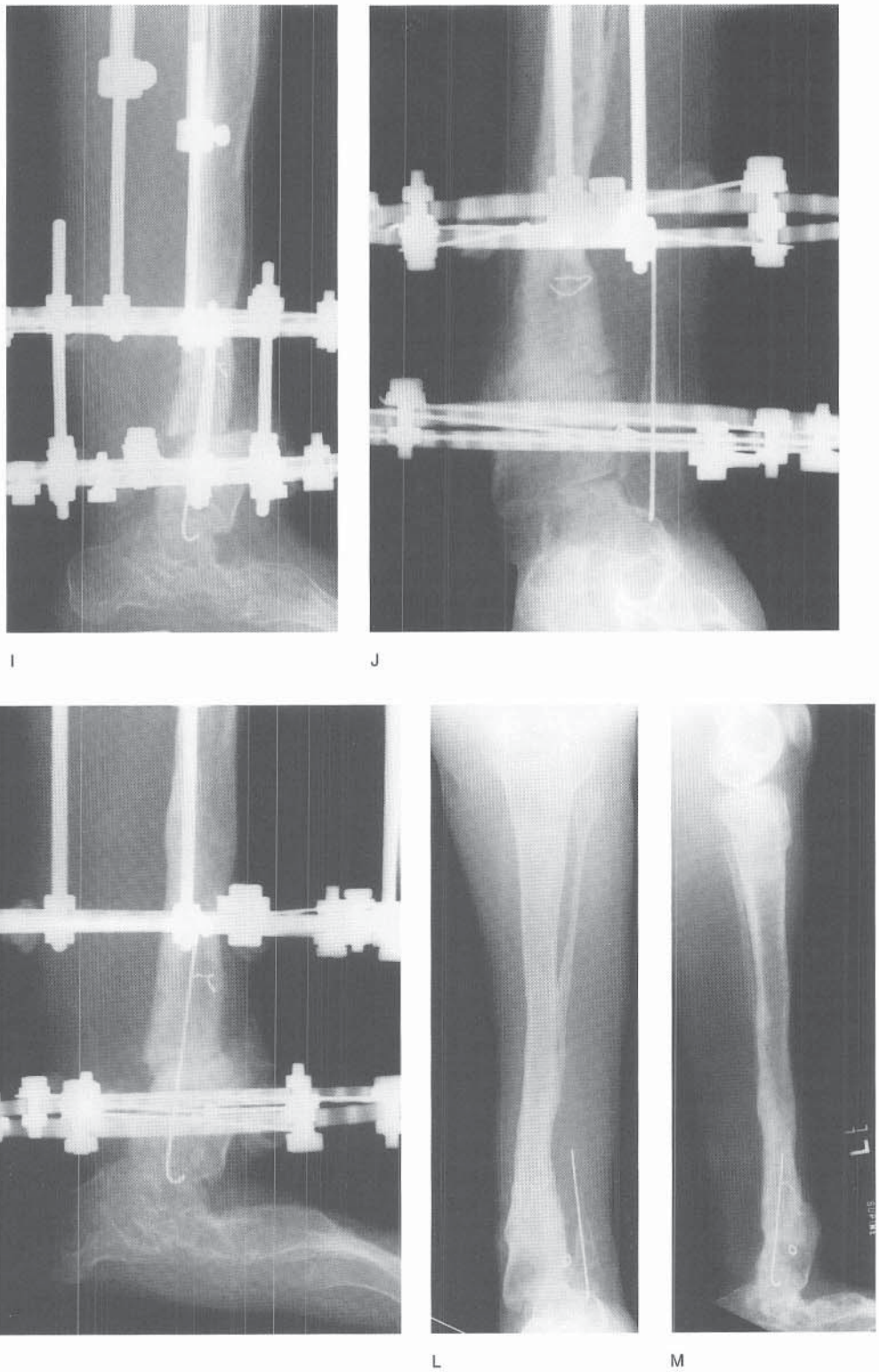


FIGURE 21–35 *Continued.* **I,** The valgus was corrected, but refracture occurred again. A simple compression construct was applied with acute correction of procurvatum. **J** and **K,** Four months later, union was achieved (note removal of rods between rings to test stability on weightbearing). **L** and **M,** Radiographic appearance 3 years following union, at age 19. Although the ankle is still in valgus, the foot is plantigrade and the patient ambulates without orthotics. The ankle and subtalar joints are stiff.

well as in older children. The use of Ilizarov methods in children less than 5 years old either has failed to achieve union or has resulted in almost immediate refracture.[16,58] The ability to compress the pseudarthrosis site does not seem to alter the pathobiology of this bone sufficiently by itself to effectively treat the lesion. The basic principle of intramedullary fixation long term also is not realized by an initial compression technique.

For patients in whom other methods have failed or who have angulation threatening refracture, shortening exceeding 5 cm, late symptomatic ankle or proximal tibial valgus, or procurvatum producing a severe calcaneus-type gait, realignment with external fixation, compression of the pseudarthrosis site, and lengthening or angular correction at another site affords the *possibility* of reconstructing all components of the deformity. Although refracture remains the most likely complication in older patients in whom reconstruction has been successfully performed, this must be weighed in relation to the alternative treatment for unreconstructable deformities—permanent orthosis or amputation. The realistic functional outcome of a heroic attempt at complex reconstruction must also be weighed against the functional and psychological consequences, favorable and unfavorable, of an amputation after years of unsuccessful treatment.

LATE FRACTURE

The previous sections considered the usual presentation of *congenital* or *early-onset* pseudarthrosis and its management. Occasionally a child presents with a tibial fracture that fails to heal following cast treatment. Usually there is no prior history of abnormality of the tibia, or mild bowing may have been noted, but the fractured tibia is radiographically dysplastic.[4,114,133] Typically the patient does not have neurofibromatosis or fibrous dysplasia, there is no family history of these conditions, and the age at presentation can be up to 12 years. Frequently the presenting symptom is pain, with a stress fracture seen radiographically, and may follow trivial trauma or more strenuous activity such as jumping or bicycle accidents. Invariably there is some medullary sclerosis or cortical infraction, and cystic areas have also been observed.[133]

The outcome following treatment of such late fractures in previously undiagnosed dysplastic tibiae is generally more favorable than the outcome of treatment for early-onset pseudarthrosis, following a course typical of many stress fractures. About one-third of late dysplastic fractures will heal with cast immobilization only, although they may heal with abnormal anterior bowing, predisposing to recurrent stress fractures.[133] Another one-third will heal with bone grafting and/or intramedullary fixation, which is the recommended treatment should cast immobilization fail. For the remaining patients, persistence of pseudarthrosis following standard treatment is extremely ominous, leading eventually to amputation in some reports.[4,114] As with early-onset pseudarthrosis, more complex treatment with external fixation devices or vascularized bone-tissue transfer are options that must be weighed in light of the morbidity of ending up with a nonfunctional extremity secondary to atrophy, stiffness, and tenuous union requiring continuous protection.

AMPUTATION

Traditionally amputation is considered the ultimate capitulation and acceptance of failure by surgeon and patient. While amputation is never appropriate as an initial or early treatment, it is entirely appropriate in the resistant pseudarthrosis where other extensive treatments, including fibular transfers or Ilizarov techniques, have not achieved a functional extremity, either because of persistent nonunion or because of dysfunctional angular deformity, shortening, atrophy, and stiffness.[66] Too often amputation is delayed, to the detriment of the functioning of the child, because surgeon and parents have too much invested to abandon the extremity in spite of multiple failed procedures. The final function in a patient who has undergone multiple operations but still must protect the leg in an orthosis may well be worse than if an earlier amputation and prosthetic fitting had been performed.[91] Thus, at some juncture in the continuum of treatment for an ununited pseudarthrosis, the surgeon must consider the dysfunction that will result, physically and psychologically, if further attempts at uniting a resistant lesion are made.

Ankle disarticulation (Syme or Boyd type) rather than amputation through the pseudarthrosis or tibial bone is recommended.[83,110] This prevents spike formation at the transected bone end and subsequent stump revision(s), and covers the stump with end-bearing heel pad skin. Persistent motion at the pseudarthrosis site is managed by the prosthetic socket. Rarely, the pseudarthrosis has healed spontaneously once the foot was removed.[46] Indications for below-knee or transpseudarthrosis amputation would include removal of osteomyelitic bone and a relatively mature patient with little potential for bony overgrowth.

Anterolateral Bow of the Tibia—Benign Form

In 1990 a group of patients at our institution was identified that did not follow the typical course of patients with congenital anterolateral bowing of the tibia.[163] These patients never suffered a fracture, and the anterolateral bowing gradually resolved with growth, a pattern typical of the more common entity of congenital posteromedial bowing (Fig. 21–36). Retrospectively these patients, on their earliest radiographs, demonstrated a presumptive healing response in the concavity of the anterolateral bow, consisting of hypertrophy and subperiosteal new bone. This hypertrophy continued during growth with active remodeling of the deformity, following a pattern similar to remodeling of an angular deformity following a metaphyseal fracture in a young child. Although some of these patients were initially protected in an orthosis, this treatment was gradually abandoned when it was realized that this form of anterolateral bowing did not predispose to pseudarthrosis.

Eventually these patients remodeled their bowing and required only management of a residual limb length discrepancy. Additional radiographic findings that were reliable in identifying this pattern were that the fibula was long and straight and uninvolved with the bowing, while the foot maintained a relative varus position, possibly due to the

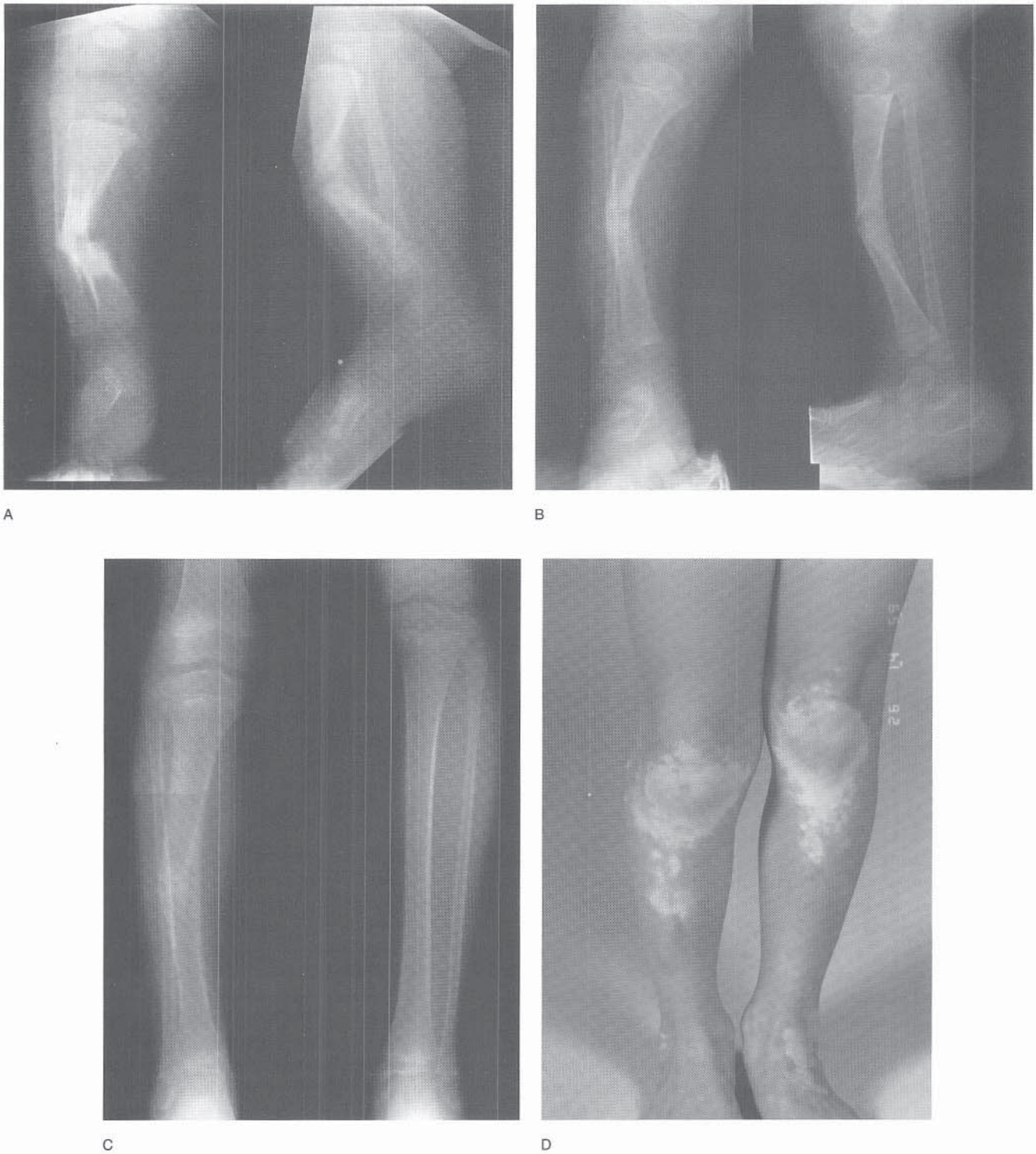


FIGURE 21–36 **A,** Radiographs of a newborn with a benign anterolateral bow. Note the callus in the concavity of the tibia, and the straight uninvolved fibula. **B,** Appearance at age 18 months. Remodeling of the callus in the concavity of the deformity continues. **C,** Appearance at age 9. The deformity has further remodeled. There is 4.5 cm shortening. The patient is fully active. **D,** Clinical appearance (the vitiligo is unrelated).

long fibula, unlike the gradual valgus pattern seen with true pseudarthrosis of the tibia (Fig. 21–36). As already mentioned, no fractures occurred. In retrospect, this pattern of benign resolution appears similar to that described by Crawford in the type I nondysplastic form of anterolateral bowing. The patients with the benign form of anterolateral bowing showed no signs of neurofibromatosis, fibrous dysplasia, or amniotic band syndrome.

Management of the benign form of anterolateral bowing, therefore, is one of early observation with the expectation that the deformity will resolve, and later, management of a residual limb length discrepancy. As is also seen in patients

with posteromedial bowing, discrepancies exceeding 5 cm at maturity are not unusual, and because treatment by epiphysiodesis produces disproportionately short tibias, limb lengthening may be the treatment of choice in a patient with this larger magnitude of shortening (Fig. 21–36).

Congenital Posteromedial Bow of the Tibia

ETIOLOGY AND CLINICAL FEATURES

This deformity, present at birth, is invariably associated with a calcaneus position of the foot and varying degrees of dorsiflexion contracture of the ankle (Fig. 21–37). It is almost certainly due to intrauterine malposition, and although its appearance may be dramatic at birth, with up to 60 degrees of angulation, it is highly likely to resolve spontaneously with stretching and splintage.[77,79,120] The calf is frequently smaller in diameter, however, indicative of the degree of intrauterine compression. A dimple at the apex of the angulation may also be present. Usually a posteromedial bow is a single deformity without associated conditions or anomalies.

The natural history of the bowing is spontaneous resolution, especially during the first 6 months. By age 2, the angulation should be essentially normal, especially the posterior bow. Foot deformity has usually resolved by 9 months. There is no increased risk of fracture or pseudarthrosis. Shortening, however, commonly exceeds 2.5 cm and averages 13 percent of total limb length.[79,120] Thus, the parents can be counseled early about the probable need for limb equalization as the main orthopaedic concern. There is some evidence that the shortening is related to the severity of the initial bowing and dorsiflexion contracture of the ankle. The appropriate use of contralateral epiphysiodesis is therefore commonly necessary.

TREATMENT

Initial treatment of the newborn with a posteromedial bow of the tibia should include gentle stretching of the dorsiflexion contracture as well as stretching of the lateral ankle structures into a supinated or inverted position. In a more severe case, serial casting into plantar flexion and the use of splints or bracing to maintain position until weightbearing have been prescribed. With rare exception, in our practice manipulation by the parents has been quite satisfactory in reducing the dorsiflexion contracture. Stimulation exercises of the plantar flexors has also been helpful. Patients usually do not require orthotic stabilization of the ankle once weightbearing has begun except when the limb discrepancy requires a lift so that the toddler may begin ambulation without resorting to knee flexion on the long side. Because such a lift in a young child is rather bulky, it is best affixed to an AFO, at least initially, so that functional ambulation can begin.

The long-term prognosis for posteromedial bowing is generally benign. There is no increased rate of fracture, and once the foot is in a functional position, the child is usually unimpaired with the exception of eventual limb length discrepancy.

No treatment for the angular deformity should be considered until an adequate period of observation confirms that correction is in fact not occurring. Severe residual bowing after age 3 to 4 years is an indication for corrective osteotomy (Fig. 21–38). Bone healing after osteotomy is normal and does not risk pseudarthrosis.

The eventual limb length discrepancy is usually managed by contralateral epiphysiodesis. If the discrepancy is predicted to exceed 5 cm, a tibial lengthening may be considered to avoid the significant shortening of the tibial segments resulting from epiphysiodesis, which makes the knees appear disproportionately close to the floor.

Congenital Pseudarthrosis of the Fibula

Pseudarthrosis involving only the fibula is an even less common skeletal manifestation of neurofibromatosis than congenital anterolateral bowing of the tibia. The diagnosis is often delayed until childhood or adolescence because of the

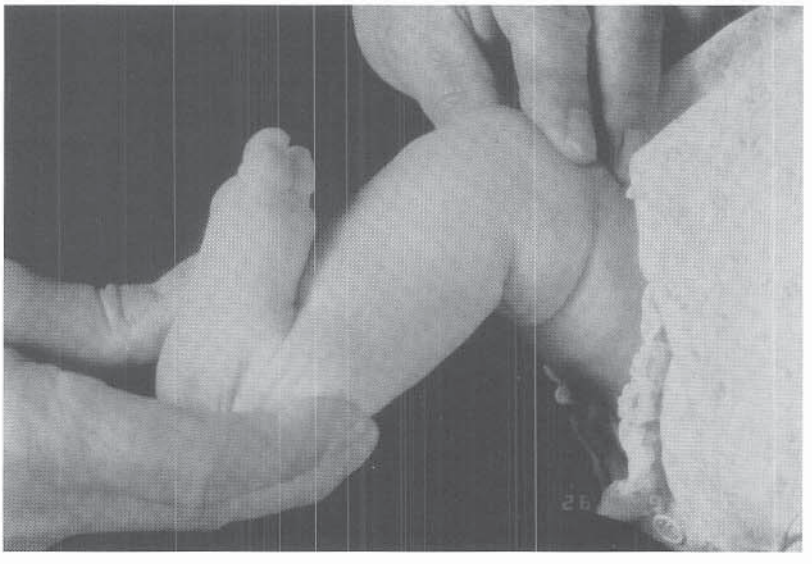
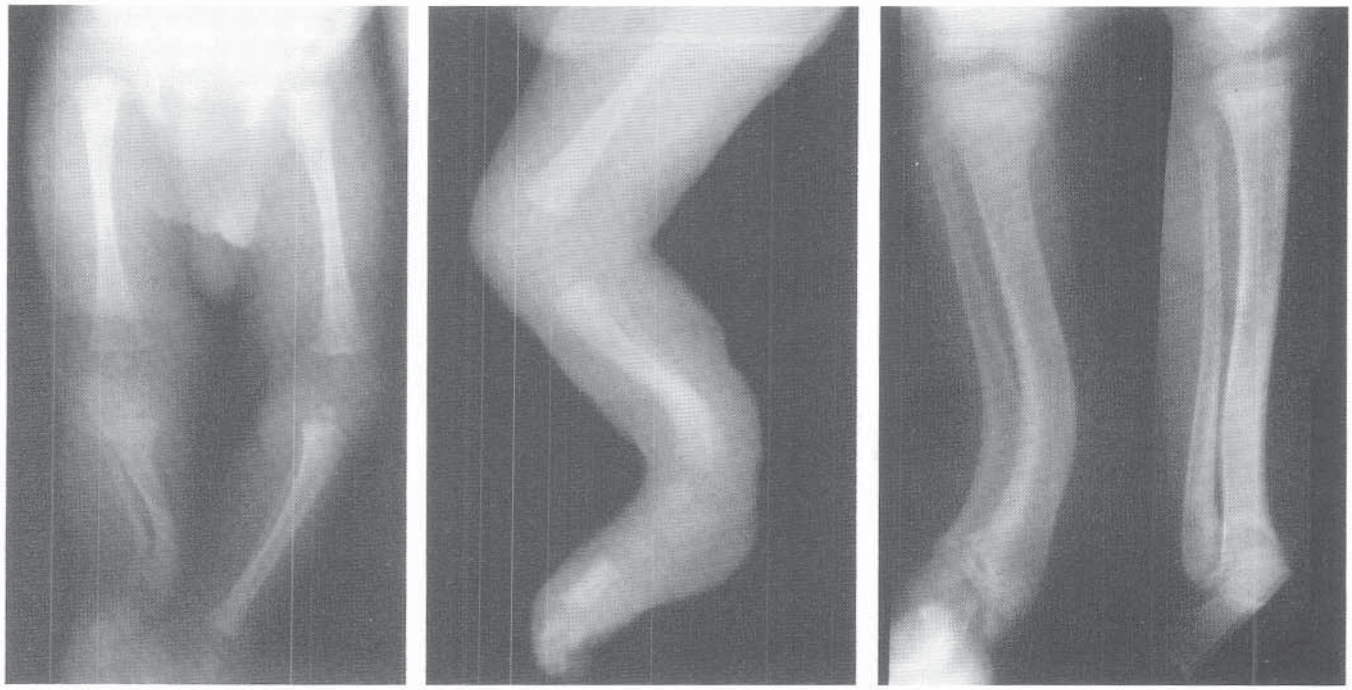


FIGURE 21–37 Clinical appearance of a calcaneus foot deformity associated with a posteromedial bow of the tibia.



A B C

FIGURE 21–38 **A,** Radiograph of a newborn with a severe posteromedial bow. **B,** Lateral radiograph. **C,** Appearance at age 7. The medial bow has not corrected and there is a 6-cm length discrepancy. Angular correction and lengthening are indicated.

lack of symptoms or functional disturbance. Because the fibula bears only one-sixth of the weightbearing stresses in the leg,[94] a pseudarthrosis may not become symptomatic until the leg and ankle are vigorously stressed by the increased activities of schoolchildren.

Isolated fibular pseudarthrosis is rare enough that the radiograph of the tibia must be carefully scrutinized to rule out occult prepseudarthrotic involvement.[156] Medullary sclerosis or an incipient stress fracture, with or without a mild anterolateral bow, indicates that the fibular lesion merely accompanies a heretofore undiagnosed tibial lesion (see Fig. 21–27). Although a bowed fibula without fibular pseudarthrosis has been reported,[41] the actual occurrence of such a lesion must be exceedingly rare, because in the absence of a pseudarthrosis, the diagnosis might never be made, owing to lack of symptoms. This is because the patient with isolated fibular pseudarthrosis may not present until sufficient ankle valgus develops to seek evaluation. The

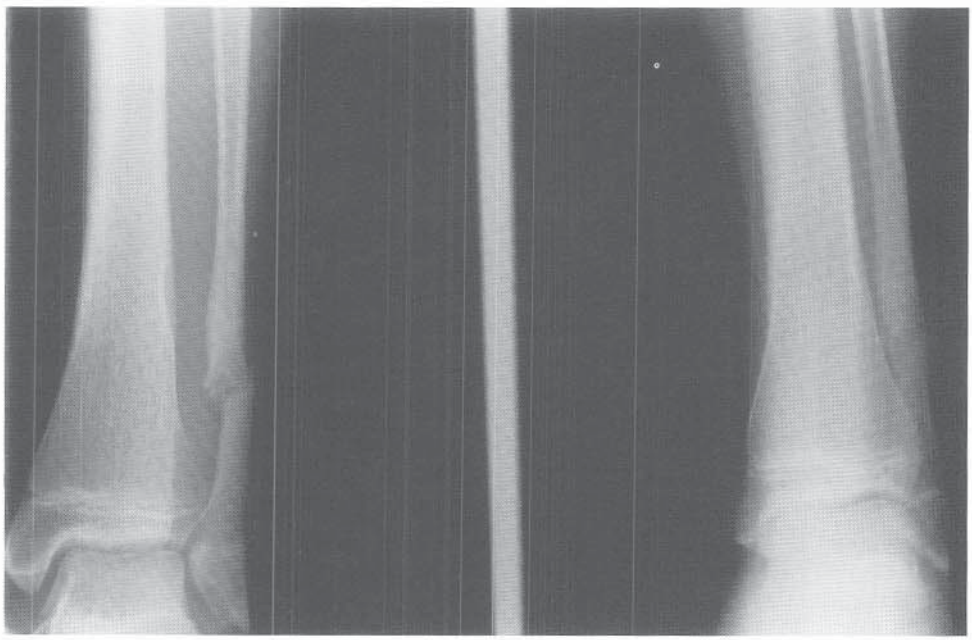
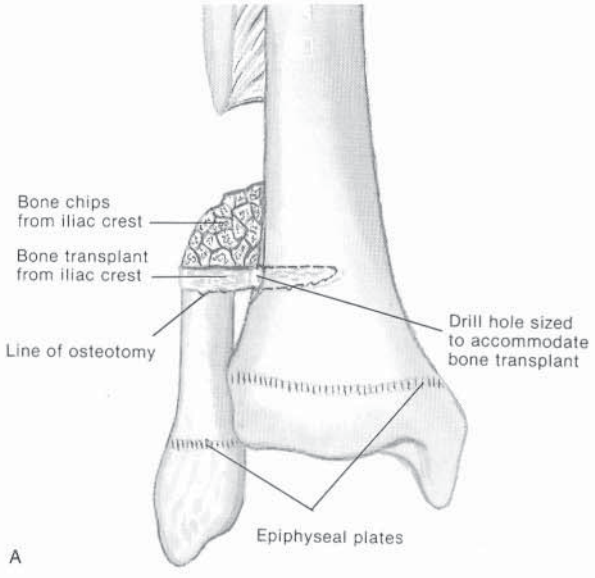


FIGURE 21–39 Radiographic appearance in a 13-year-old, skeletally immature girl with neurofibromatosis type 1 who sought evaluation for increasing ankle pain without deformity. Note the minimal cortical erosion of the tibia associated with the pseudarthrosis of the fibula.



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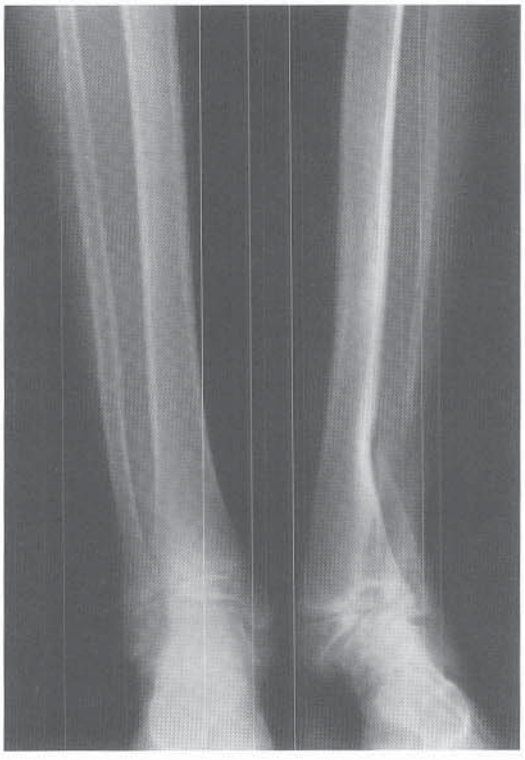

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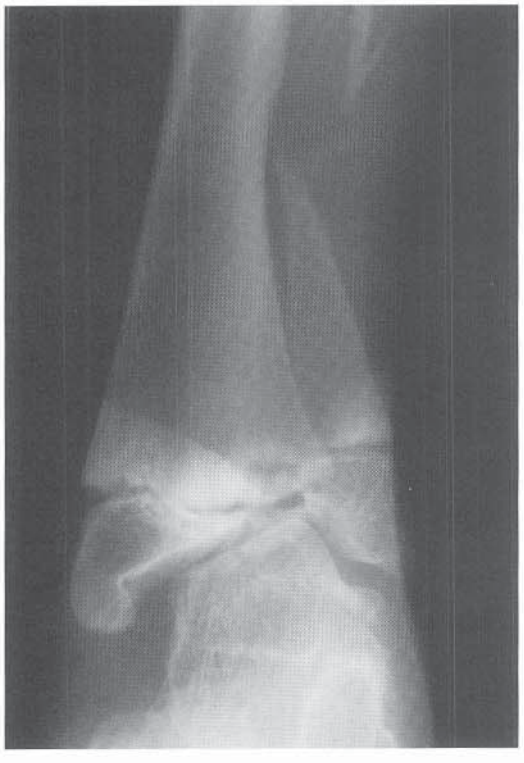

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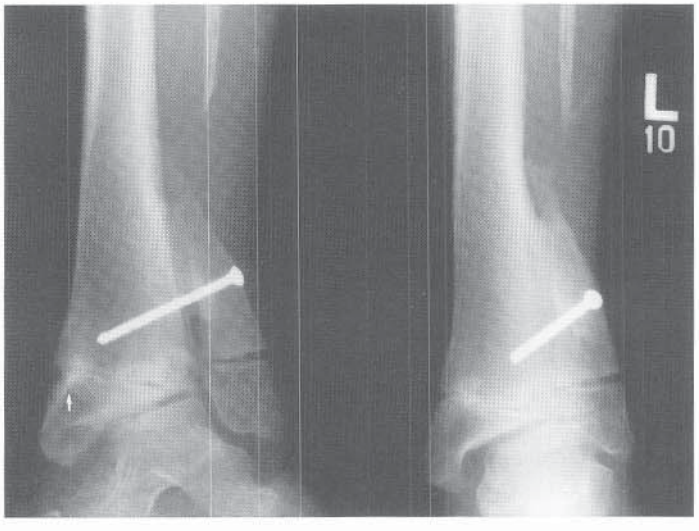

D

FIGURE 21–40 **A,** Distal synostosis of the tibia and fibula (Langenskiöld). **B,** Severe unilateral ankle valgus in a 12-year-old boy with neurofibromatosis type 1. **C,** Close-up of AP radiograph showing fibular pseudarthrosis, lateral wedging of the distal tibial epiphysis, and lateral subluxation of the talus in the mortise with secondary obliquity of the talar dome. **D,** Appearance 9 months after distal-fibular synostosis with syndesmotic screw, and medial distal tibial hemiepiphysiodesis (*arrow*). There is a clear union between tibia and fibula.

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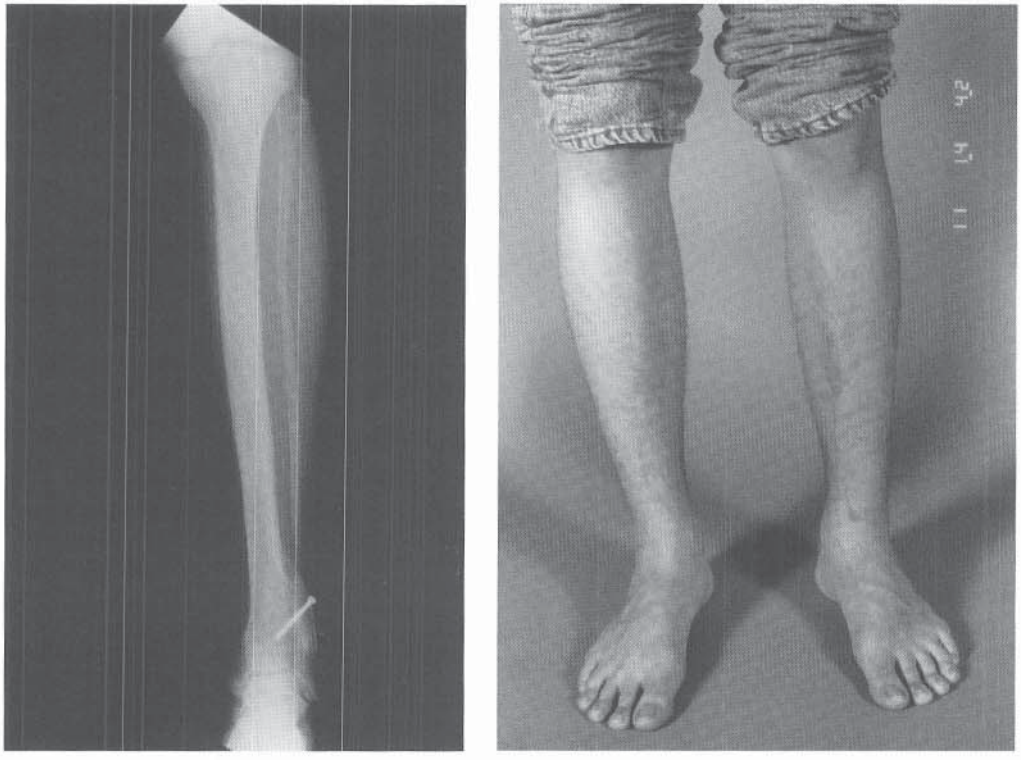

E F

FIGURE 21–40 *Continued.* **E** and **F,** At age 17, the deformity is completely corrected, with congruence of the ankle joint and talar obliquity correction.

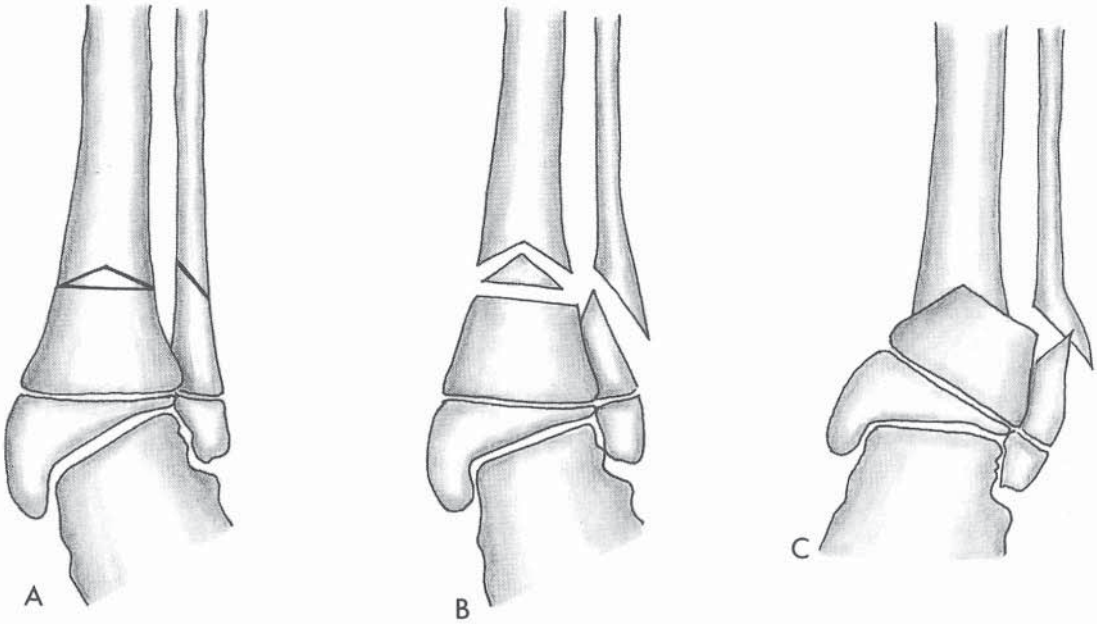

FIGURE 21–41 Wiltse technique of triangular osteotomy of distal tibial diaphysis to correct valgus ankle. **A,** The lines of osteotomy. **B,** Triangular fragment of tibia excised. **C,** Medial rotation and lateral shift of the distal fragment will correct the valgus deformity without undesirable prominence of the medial malleolus; the shortening of the limb will be less than with a simple close-up osteotomy.

pseudarthrosis invariably involves the distal one-fourth of the fibula and is seen almost exclusively with neurofibromatosis type 1 (Fig. 21–39). Although fibular pseudarthrosis has also been reported to be a precursor to late tibial pseudarthrosis,[41] we have not observed such a scenario except when there is a frank lesion in the tibia at the time of diagnosis.

TREATMENT

Treatment of an isolated congenital fibular pseudarthrosis is indicated for relief of symptoms—primarily pain with weightbearing—and to prevent progressive valgus of the ankle. Symptomatic pseudarthrosis is best treated by excision of the lesion, bone grafting, and intramedullary fixation. Osteosynthesis of the excised area is best achieved by an autologous bone graft, and because the tibia is uninvolved, an intercalary tricortical graft from the ilium is attractive, to span the excised segment and maintain length. With intramedullary fixation to stabilize the graft, immediate weightbearing is possible to help achieve union. Successful union alleviates the symptoms and prevents ankle valgus from progressively ascending fibular malleolus.[82,111]

Surgical synostosis of the distal tibial and fibula metaphyses[96] is an alternative to directly treating the fibular pseudarthrosis. According to Tachdjian, the synostosis is simple to perform and heals rapidly while providing control of the ankle valgus (Fig. 21–40A). This procedure also uses iliac bone graft but requires the patient to be nonweightbearing for 2 months, presumably to avoid distal tibial-fibular movement, which might delay the transverse union between the two bones. Internal fixation by transsyndesmotic screw is recommended to allow more rapid mobilization (Fig. 21–40D). This procedure can stabilize ankle position but will not correct a valgus deformity.

Should correction be necessary, medial hemiepiphysiodesis of the distal tibia, either permanent or temporary by means of staple or screw, is an excellent method if sufficient growth remains. Staple hemiepiphysiodesis[24] can be quite successful in correcting the deformity but may be complicated by the well-known problems of staple migration, prominence, and scar hypertrophy. Screw fixation of the medial malleolus[36,150] is our preferred technique in younger children. Permanent hemiepiphysiodesis by partial physeal ablation is indicated in the child closer to skeletal maturity. With a significant deformity, correction by hemiepiphysiodesis *combined with* a distal synostosis will ensure that the fibular malleolus corrects with the distal tibia (Fig. 21–40). In the mature patient, supramalleolar osteotomy is the only option if the valgus must be corrected. An opening-wedge technique[135] is appropriate if further shortening by a closing-wedge technique will exacerbate a limb length discrepancy. In addition, some medialization of the ankle by the tilting technique of Wiltse (Fig. 21–41) may be appropriate if the valgus is severe.

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