:'Ilizarov Technology

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';::'}'n 1951, Professor Gavriil Abramovich Ilizarov from Kurgan in the 80-'/,;, :viet Union developed a circular external flxator forthe treatment of fractures (Fig 1); Dilling the next decade, Ilizarov discovered the tech-niques 'of physeal distraction, corticotomy lengthening, bone transport, and many others. The common basis for all of these methods he called "the, theory of tension stress?"' z Through controlled, mechanically applied tension stress; Ilizarov was able to show that bone and soft tissue can be made to regenerate in a reliable and reproducible manner. Over the next ,*Ai* years until his death in 1992 at the age of71,he developed countless 'clinical applications .ofbone and soft tissue regeneration.

In North America, experience with the technique dates back to 1986. Currently, the most common indications for the Ilizarov technique are as Iollows: (1) limb lengthening; (2) treatment of nonunlons, bone and soft-*suedefects, and osteomyelitis; (3) correction of bony deformities, joint contracture deformities, and even contour deformities of-the, limbs; (4) aitlITodesis; and (5) treatment of 'fractures anddislocatiOIis.

':tLIZAROV APPARAtUS

The Ilizarov apparatus is a circular external fixator that gains fixation to hone through smooth or beaded Kirschner wires of 1.5 or 1.8 rnm in diameter (Fig 2).3 These are placed under tension and are oriented in multiple direcitiOlis and multiple planes. More recently, h~lf-pins' together with wires are used as a hybrid system, or half-pins are used: alone, replacing wires altogether>The key element appears to be the ring and not the wires. In addition to the rings arid wires, the Ilizarov system consists of multiple parts with niuitipurpose designations, such as hinges, plates, andthreaded rods. The frame can 'be assembled in an almost unlimited number of variations and combinations, depending on the task at hand. H is not surprising that this system. has been labeled a human "erector [meeliario) 'set." With the appropriate construct; li:inb 'segments can be moved around in space in any ,diredion to correct problems of length, rotation, translation, and angulation: One level can be placed under com-;pressiOn while another is placed under distraction. In the treatment of bone defects, intercalary segments Of bone can be moved independently ofthe proximal and distal extremities of the bone without changing the

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FIGURE 1.
Professor Gavriil Abramovich Ilizarov, 1921-1992.

bone length. The latter maneuver is called bone transport, and is a technique used to treat bone and soft-tissue defects.

DISTRACTION OSTEOGENESIS

Under controlled mechanical conditions, an osteotomy that is distracted apart will produce bone between the distracted bone ends. Ilizarov showed that there is a variety of factors that will affect this new bone formation. These include the following:

1. The stability of fixation.
2. The type of osteotomy used.
3. The location of the osteotomy in the bone.
4. The presence of a diastasis between the bone ends.
5. The latency period prior to distraction.

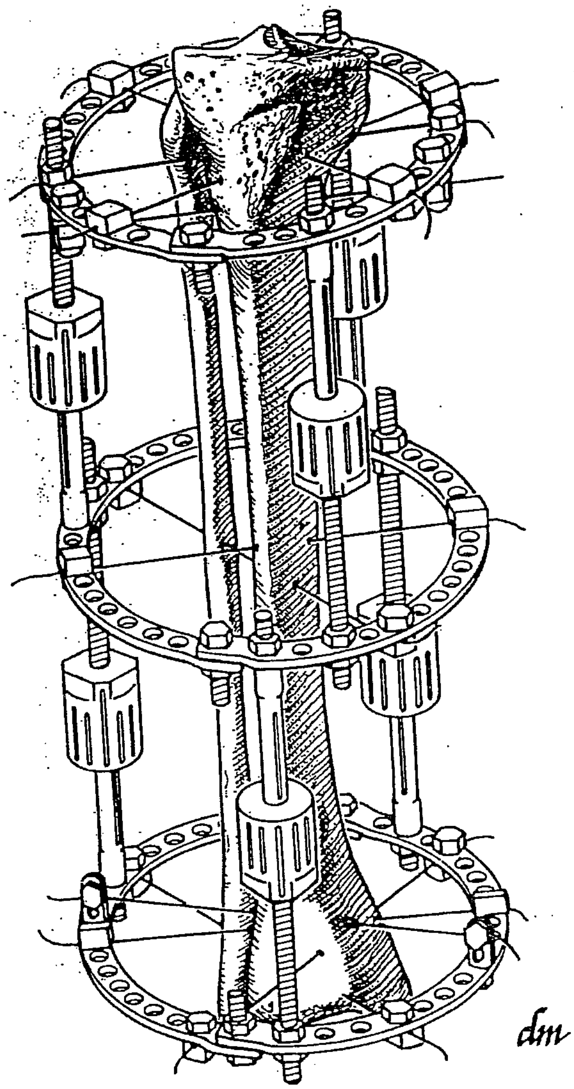


FIGURE 2.
Ilizarov apparatus with multilevel, multiplanar, multidirectional wire fixation. (From Paley D, Rumley TO, Kovelman H: Advances in Plastic and Reconstructive Surgery, vol. 7. 1991, pp 1-40. Used by permission.)

6. The rate of distraction.
7. The rhythm of distraction.

Comparing distraction osteogenesis under different frame stability conditions, Ilizarov showed that more stable fixation produced better bone regeneration? In the same experiment, he also varied the type of osteotomy performed. He compared an open osteotomy that transected the periosteum, cortex, and endosteum, to a percutaneous osteotomy that preservediUost of the periosteum and endosteum, to a completely closed osteoclasis in which there was no damage to the periosteum and the endosteum remainedpredominantly intact. The preservation of these soft tissues decreased the time required for consolidation of the distraction and osteogenesis of new bone (regeneration). The completely closed osteoclasis consolidated only slightly faster than did the percutaneous osteotomy. Based on these results, Ilizarov's recommendations are to use a percutaneous subperiosteal cortical osteotomy called a "corticotomy."

Numerous authors have questioned the relative importance of endosteal vs. periosteal preservation. Kojimoto and coworkers performed selective damage to the periosteum or endosteum. Transection of the periosteum delayed regenerate bone formation, whereas endosteal damage did not. Zemba and colleagues compared corticotomy with endosteal sparing with osteotomy with endosteal transection in dogs and found no difference in consolidation time. Weiner and Paley (unpublished data) compared percutaneous subperiosteal osteotome corticotomy with percutaneous subperiosteal Gigli saw corticotomy in 100 patients undergoing tibial metaphyseal proximal lengthening? In the former, endosteal preservation is attempted; in the latter, the endosteum is transected by the saw. No significant differences were found in bone consolidation time.

The level of the bone cut also is a factor. The metaphyseal region is known to have a very high osteogenetic potential compared to the diaphyseal region. Since the growth plate is located next to the metaphysis, Ilizarov feels that the soft tissues in this region are better adapted to lengthening, since they normally respond to distraction by the epiphyseal plate. For these reasons, a metaphyseal corticotomy is preferable to a diaphyseal corticotomy. Fischgrund and associates recently showed that metaphyseal corticotomies of the tibia consolidate significantly faster than do diaphyseal corticotomies?

Any initial diastasis or translation between the bone ends may be deleterious to bone regeneration. As a result, the corticotomy should remain undisplaced. After performing the corticotomy, there is a latency period prior to distraction. A latency period of 7 to 10 days has been shown to be optimal in a dog experimental model. In general, the two factors that relate to the latency of distraction are the age of the patient and the quality of the corticotomy. The older the patient, the longer the latency period should be prior to distraction. The latency period allows the inflammatory phase of fracture healing to subside, thereby enabling the distraction to begin during the reparative phase when early osteogenesis normally is seen. Distraction of this newly formed callus has been termed "callotasis" (callus distraction).9 In a young child, 3 days is a sufficient latency period, whereas in an older child or a young adult, 5 to 7 days is preferable. In an adult, 7 to 14 days may be opted for. One also must consider how well the comcotomy was performed. In a minimally traumatic corticotomy, distraction may begin earlier than in a corticotomy with more significant vascular damage to the periosteum or endosteum.

Finally, two very important factors are the rate and rhythm of distraction. The optimal rate of distraction usually is 1 mm/day. Slower rates, such as 0.5 mm/day, may lead to premature consolidation; rates that are too rapid, such as 2 mm/day, may cause poor bone formation, with cystic degeneration of the regenerate. Younger patients often can distract faster than older patients. The rhythm of distraction refers to the frequency of applied distraction. Distraction of 1 mm/day can be applied as a single dose or divided into multiple doses applied throughout the day.

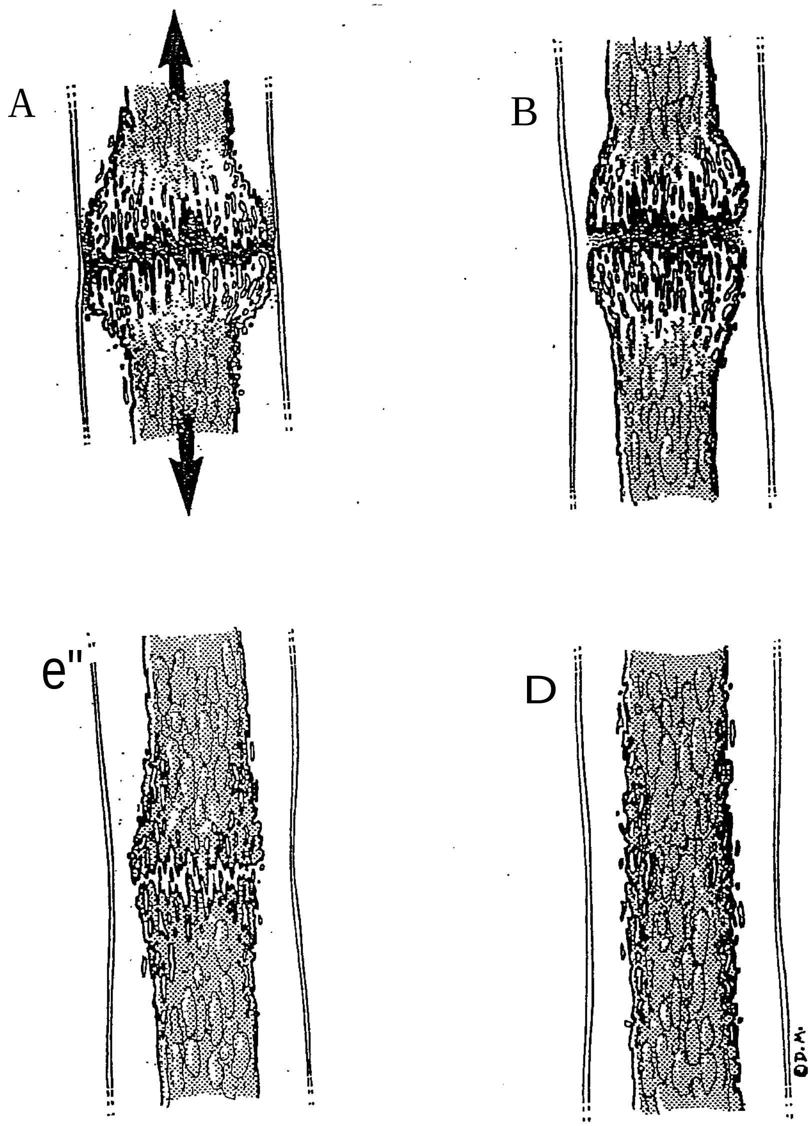


FIGURE 3.
A, distraction phase. Longitudinally oriented trabeculae form from either side of a central lucent zone (the interzone). B, consolidation phase. The distraction complete, the new bone is allowed to mature. Neocorticalization is seen and ossification of the interzone begins. C, removal date. There has been sufficient neocorticalization and obliteration of the interzone to allow safe removal of the fixator and unprotected load-bearing. D, recanalization. The medullary canal is remodeled and the new cortex is the same thickness as the old. It is almost impossible to tell that a lengthening has occurred. (From Paley D, Rumley T Jr. Kovelman H: *Adv* Plcst Reconstr. Surg 1991; 7:1-40. Used by permission.)

Applied in four equal doses, 1 mm (0.25 mm four times per day) leads to more rapid bony consolidation than it does applied in one single dose per day. Ilizarov developed a motorized distractor that performs 60 lengthenings a day to a total (length) of 1mm. Consolidation time using the automated distractor decreased in a dog model.f Human results with an American autoriator (Autogenesis, Inc., Anchorage, AK) are too preliminary to report. There is some evidence that pain may be reduced by this quasicontinuous distraction.

The sequence of events following the performance of a corticotomy involves (1) a latency period, (2) a distraction period (Fig 3, A), and (3) a consolidation phase (see Fig 3, B). The distraction period is the time during which distraction of the bone ends is being performed. The consolidation period is the time following distraction during which the new bone formed is allowed to consolidate prior to removal of the external fixator (see Fig 3, C).[3]

During the distraction period, histologic and radiographic examination reveals trabeculae oriented in the direction of distraction emanating from both bone ends and terminating at a fibrous interzone that separates their proximal and distal tips (Fig 4).[3] This fibrous interzone is the

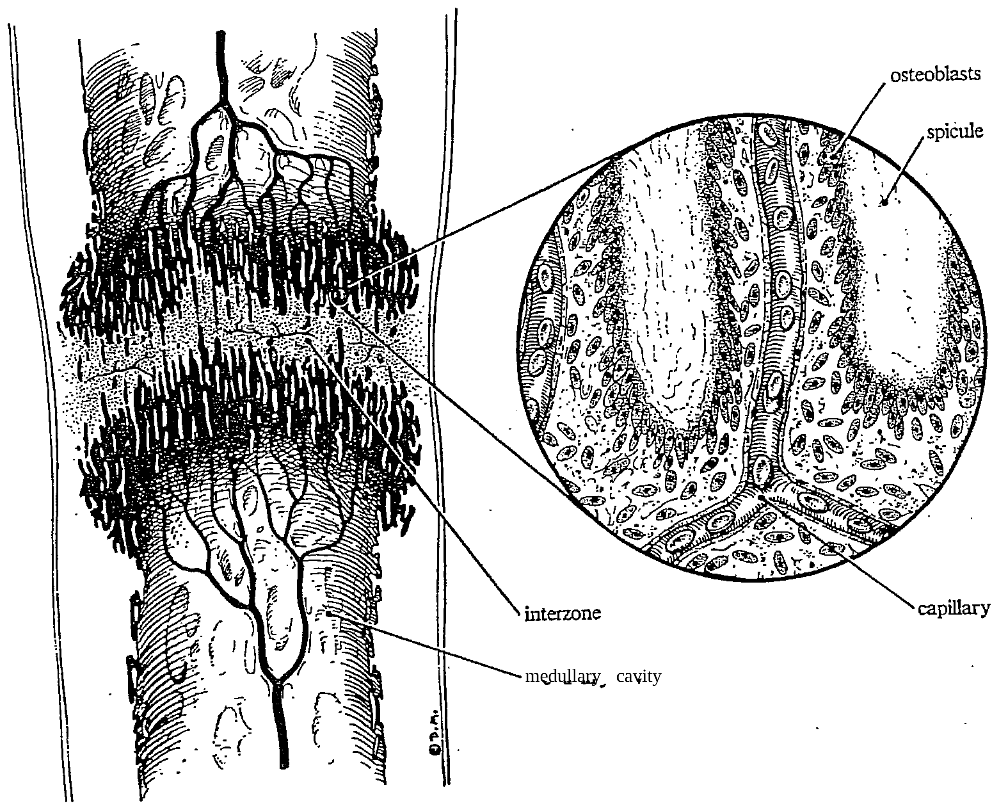


FIGURE 4.
There is continuity between the nutrient artery and vein and the richly vascular trabeculae of new vessels. The interzone is relatively hypovascular compared to the trabeculated regions, which are hypervascular [insert]. The interface between the interzone and the tips of trabeculae is shown. Note that the cells surrounding the trabeculae and the cells of the interzone appear similar. It is presumed that the interzone cells are an undifferentiated mesenchymal type of cell that transforms directly into the osteoblasts of the trabeculae. Therefore, the process is one of intramembranous bone formation. (From Paley D, Rumley T Jr, Kovelman H: *Adv Plast* Reconstr Surg 1991;7:1-40. Used by permission.)

"pseudo-growth plate" of this new bone formation. The trabeculae are conical in shape, with a very wide base near the original bone end and a very narrow tip at the fibrous interzone. Detailed study of the fibrous interzone has revealed it to contain spindle-shaped cells that stream into the tips of the newly formed trabeculae. Collagen formation is seen emanating from the fibrous interzone into the new trabeculae, followed by the deposition of minerals.[11] The spindle-shaped cells from the interzone are seen to differentiate gradually into osteoblasts, which produce mineralized osteoid. They line the outer surface of these conical trabeculae along their entire length. As one proceeds toward the bases of the trabeculae, appositional new bone formation widens their bases. At the fibrous interzone, cartilage intermediary rarely is seen between the spindle-shaped cells and the osteoblasts. This process, therefore, has been termed intramembranous ossification.

Occasionally, one sees small cartilage islands that have been produced from the interzone.[12] These cartilage islands then are converted to bone by the process of endochondral bone formation. These regions may represent areas of increased ischemia or instability. (In rabbits, cartilage formation is part of the normal sequence of events, whereas in higher animals [e.g., dogs, sheep], it is less frequent).[5, 13] Under greater conditions of instability, fibrous tissue formation and, in some cases, degenerative cyst formation is seen.[1, 2] This demonstrates the importance of stability for distraction osteogenesis formation.

The interzone has been shown by vascular injection studies to be relatively hypovascular in comparison to the hypervascular trabecular regions.[1, 2, 11] The latter regions have cascades of trabeculae and wide vascular channels. Between adjacent trabeculae, one can see vascular channels. These emanate from the relatively hypovascular interzone where the new vessels are thought to have originated. The interzone is believed to contain relatively undifferentiated mesenchymal cells that may produce bony, cartilaginous, fibrous, or vascular tissue. The relative hypovascularity of the interzone is thought to reflect its volatile nature, which, under conditions of too rapid distraction, becomes ischemic and leads to cartilage, fibrous tissue, or cystic degeneration.

At the end of the distraction period, one begins to see thickening of the trabeculae at the periphery of the bone tube (see Fig 3, *B).* This is called neocorticalization. During the consolidation phase, the neocorticalization matures. The apparatus can be removed once the new bone formation in the distraction gap demonstrates closure of the interzone and neocorticalization on at least three sides, as judged by anteroposterior and lateral radiographs. Although this method of judging the strength of the newly formed bone is crude, it still is the gold standard. Alternative and more objective methods being investigated include quantitative computed tomography, mechanical stress testing, and acoustic resonance analysis.

After removal, the regenerate bone segment continues to remodel in the junction between the new and the old bone. Remodeling of the medullary trabeculae proceeds until complete recanalization of the medullary canal is seen. At the end of the process, the new bone appears as a normal tube of bone identical to the host bone (see Fig 3, D). This is in contrast to the new bone seen in fracture healing, in which there is a disorganized collagen network of woven bone and the bone often never returns to its original tubular shape.[15]

Distraction osteogenesis also occurs under natural conditions. The bone growth at the perimeter of the growth plate experiences traction forces from the attached periosteum. This new bone formation occurs in a trabecular fashion without an endochondral intermediary. The trabeculae are oriented in the direction of the traction. The bone seen in the sunburst appearance of periosteum elevated by a neoplasm resembles distraction osteogenesis. The tumor acts as the distractor elevating the periosteum, which then produces trabeculae perpendicular to the shaft of the bone in the direction of the distraction. Controlled mechanical distraction osteogenesis is a method developed to reproduce this natural phenomenon and to accelerate it to its maximum potential. To put matters in perspective, the human distal femoral growth plate grows about 50 µm/day; distraction osteogenesis lengthens at 1,000 µm/day.

DISTRACTION HISTOGENESIS OF SOFT TISSUES

The mechanisms of new bone formation under controlled mechanical distraction as pioneered by Ilizarov are relatively well understood and have been reproduced by several investigators.[16-18] In contrast, distraction histogenesis of soft tissues is less well understood. Examples of distraction histogenesis of soft tissues are abundant in nature. A goa-fold increase in the size of the female uterus under the expansion force of the fetus is an excellent example of this process. After birth, there is no question that new soft tissue of the uterus and of the abdominal wall has been regenerated during the gradual distraction from within. Soft-tissue distraction by a rapidly growing neoplasm employs the same mechanism. Controlled mechanical distraction of soft tissues has been performed, employing soft-tissue distractors.[19] It can be assumed that the same mechanisms that are involved in soft-tissue regeneration under conditions of limb lengthening are involved in these other situations. The questions that remain unanswered are as follows:

1. Which soft tissues are amenable to distraction histogenesis?
2. Does cell proliferation occur, or is there simply a stretch phenomenon?
3. Which cells undergo histogenesis under the stimulation of distraction?
4. What are the optimal rate and rhythm parameters for soft-tissue distraction histogenesis?

Ilizarov has investigated the effects of distraction on skin, muscle, tendon, fascia, blood vessels, lymphatic channels, and peripheral nerves.[2] Although different tissues react in different ways, there are two predominant mechanisms at play: reorganization of collagen in response to stretch and neohistogenesis. For example, the initial reaction of fascia[2] to closed distraction is reorientation of its collagen network to stretch. This can be likened to pulling on a fisherman's net. The crosshatches that form square holes in the net reorient to form diamond-shaped holes H:I+d eventually just slits. After collagen fiber reorientation, increased fibroblastic activity is seen.

Iiyacbkova showed that muscle also responds initially by stretching without cell proliferation, followed by a combination of stretching and a cellular response.[20] The cellular response is mixed. First, there is a recruitment of cells, as evidenced by increased numbers of satellite cells seen. These undergo neohistogenesis and contribute to the growth in length of new muscle. Similar findings were reported by Appell and coworkers in athletes.[22] Second, there is an addition of sarcomeres to existing muscle cells. Radiologic markers were placed on both muscle and fascia in order to determine the level at which the lengthening was occurring.[14, 21, 22] For the first 20% of growth and lengthening of the muscle, the radiologic markers moved apart evenly. This implied that the muscle was lengthening evenly between the muscle tendon junctions. After 20% of lengthening of the muscle was achieved, there was greater lengthening seen at the level of the bone distraction than at other levels in the muscle. Between 20% and 25% lengthening, increased damage was noted in the muscle structure.[17, 21, 22]

In another experiment, a double-level bone cut was performed in the proximal and distal tibia. Lesions were not seen in the muscle until it reached the point of 20% to 30% increase in length.[21, 23] This implied that a second level of distraction can redistribute the level of lengthening more physiologically, despite the increased rate of distraction to the muscle.

In other Russian experiments, the first muscle changes were electrophysiologic.[24] This was followed by a change in histology and, finally, by alteration in the total morphology of the muscle. The recovery of the muscle occurred in reverse order. There first was a recovery of the morphology, followed by the histology and, only at a very late stage, the electrophysiology.

Peripheral nerves are seen to undergo change under distraction as well.[5, 25] New Schwann cells and active myelinization can be observed in the peripheral nerves. Electron microscopic morphologic features typical of fetal tissue but atypical of adult tissue are observed in the nerves, muscles, and most of the soft tissues. Ilizarov claims that "tension stress" stimulates tissue to regress into a fetal state with the regenerate potential of fetal tissue.[1, 2] If this hypothesis is correct, then tension stress could prove to be the key to unlocking secrets to limb regeneration.[26]

Needless to say, distraction histogenesis of soft tissues is an area ripe for future research. An example of this type of research is provided by two experiments performed by Ilizarov.[27] In the first one, he created a femur fracture with an associated vascular lesion of the femoral artery. In order to treat the vascular lesion without a bypass graft, he shortened the femoral fracture by overlapping the bone ends. The arterial injury, created by resecting a segment of the artery, was repaired by end-to-end anastomosis after the shortening. Five weeks after shortening, the limb was relengthened without failure of the anastomosis. If relengthening was performed prior to 5 weeks, rupture of the anastomosis or aneurysm formation occurred. In a second experiment, Professor Ilizarov created a nerve defect and treated it by resecting a segment of femur; reopposing it end-to-end, doing a primary anastomosis of the nerve ends, and then lengthening the femur. Again, if there was at least a 5-week latency period, the nerve remained intact at the anastomosis site.

In a lecture on this subject, Ilizarov reported some preliminary findings that nerve regeneration under distraction was faster than that expected following an end-to-end anastomosis without tension. The potential for performing nerve lengthening without the related bone shortening or lengthening may be a treatment consideration in the future, to bridge nerve defects. Ligament regeneration was attempted by Aston in dogs using rapid distraction (2 mm/day) of an intra-articular block of bone. The result was a new anterior cruciate ligament of 75% normal strength.i"

LIMB lENGTHENING

Limb lengthening with the Ilizarov technique uses the biology of distraction osteogenesis for bone regeneration and distraction histogenesis of soft tissues for soft-tissue regeneration.:" 2 This avoids the need for bone grafting and soft-tissue releases or lengthening in the majority of cases. Bone regeneration may arise from distraction of the physis (physeal distraction), distraction of an osteotomy, or distraction of a pseudarthrosis. The latter will be discussed later.

Distraction of the physis leads to physiolysis and is followed by bone regeneration in the same manner as for corticotomy distraction.[29-31] The new bone formed is intramembranous and not endochondral. Although distraction of the physis without fracture theoretically is possible up to 4% of lengthening, it is not practical for the magnitude of most discrepancies. Physeal distraction frequently leads to premature growth plate closure in older children. In young children, as in young animals, this does not seem to occur.[32,33] Nevertheless, there is concern regarding the rate of subsequent growth after physiolysis, even in young children." For this reason, there are very few indications for physeal distraction. Furthermore, physeal distraction of the distal femur requires the fixation pins to be intra-articular, predisposing to knee joint sepsis and knee stiffness. Current indications should be limited to patients with transphyseal bridges that require lengthening and/or angular correction and who are not candidates for bridge resection. Such patients usually are older children or adolescents and require epiphysiodesis as part of their treatment. The physeal distraction is used to break the bridge in cases where the bridge is less than 25%, or is augmented by percutaneous osteotomy of the bridge in cases where it is greater than 25%. This leads to spontaneous groWth plate closure." Conditions such as Blount disease in the older child also may be treated by physeal distraction. The existing and predicted growth discrepancy is made up by lengthening. Recently, resection of a physeal bridge was described using physeal distraction to create a gap through which to do the resection.[37] Physeal distraction also has been used recently to increase the resection margin and preserve the joint and the growth plate in surgery for osteosarcoma.[38]

For the vast majority of lengthening cases, corticotomy distraction is the treatment of choice. The considerations for lengthening vary according to the limb segment being lengthened. Each limb segment, therefore, will be discussed individually. The age of the patient predominantly affects the treatment time. The time in external fixation divided by the number of centimeters lengthened has been called the "healing index" or "lengthening index."[39] It probably is better termed the "external fixation treatment index." Debastiani has reported indices of 1.2 months/em for the femur and 1.4 months/em for the tibia." Paley reported an index of .97 months/em for the tibia in children compared to 1.7 months/em for adults. The index for double-level lengthening of the tibia was .57 months/em;" Both of these studies assumed that the index is a constant, not dependent on the amount of lengthening. In a more recent study, Fischgrund and colleagues demonstrated that the distraction gap was related linearly to consolidation time, but the consolidation index was inversely proportional to the distraction gap." In other words, the index is higher for short distraction gaps, but lower for larger distraction gaps. There were several factors that affected the consolidation index. Diaphyseal osteotomies healed slower than did metaphyseal ones. The tibia healed slower than did the femur. Patients under 20 years of age healed faster than did those over 20 years of age. Similarly, patients 20 to 29 years of age healed faster than did those 30 years of age and older (Fig 5).

The magnitude of lengthening possible varies with the bone being

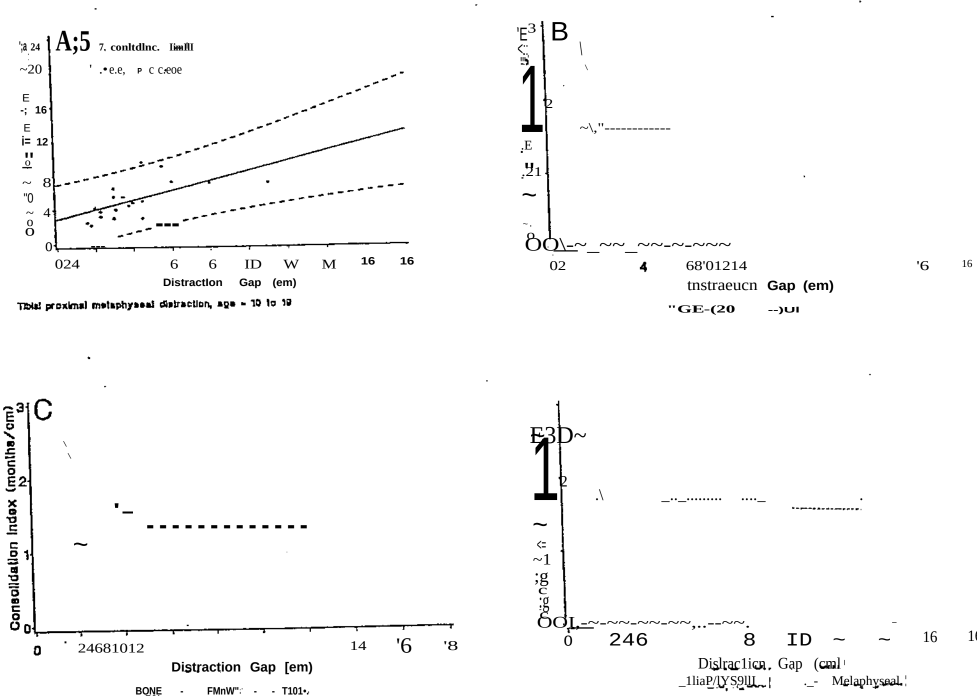


FIGURE 5.

A, consolidation time (from date of osteotomy to radiographic corticalization) is directly proportional to distraction gap. Consolidation index (months per centimeter) is inversely proportional to distraction gap. Individual graphs can be made, taking into account patient age, bone lengthened, and corticotomy level. B, patients over 20 years of age consolidate significantly slower than do those under 20 years. C, the femur has a faster consolidation rate than does the tibia. D, diaphyseal corticotomies consolidate slower than do metaphyseal corticotomies. (From Fischgrund J, Paley D, Suter C: Clin Orthop, in press. Used by permission.)

treated and the etiology of the limb length discrepancy. In general, the femur should not be lengthened more than 6 to 10 ern at one time. It may be safer to perform two short lengthenings than one large lengthening. In the tibia, the upper limits range between 10 and 15 ern. For the humerus, the range is 10 to 15 ern and for the forearm, it is 5 to 10 ern. In a recent study, Paley and associates showed that the rate of complieations is proportional to the complexity of each case.[40] A complexity index was developed. Factors that increase the complexity of a lengthening include (1) magnitude of lengthening; (2) increasing patient age; (3) increasing magnitude and complexity of associated deformities; (4) joint instability; (5) preoperative joint stiffness, contracture, or arthrosis; (6) bone segment (femur, tibia, humerus, forearm); (7) treatment of associated conditions (nonunion, arthrodesis, etc.); (B) pathologic bone (osteoporosis, rickets, infection, etc.): (9) pathologic soft tissues (scarred, radiated, infected); and (10) medical illnesses (e.g., diabetes, immunocompromise). Limb lengthening cannot be lumped together without consideration of the complexity of cases being performed. The treatment chosen must vary according to these various factors. For example, if there is preoperative knee joint instability of congenital origin, a femoral lengthening must include prophylaxis against knee subluxation by extending the apparatus with hinges across the joint to the tibia. Hip instability is dealt with in a similar manner or by preoperative innominate or acetabular osteotomy. Ankle instability may require transport of the fibula distally to buttress a valgus ankle or extension of fixation across the ankle onto the foot.

TIBIAL LENGTHENING

Most tibial lengthehings are performed proximally through a proximal metaphyseal corticotomy. The fixation should include both tibia and fibula to avoid descent of the proximal fibula and ascent of the distal fibula. Care must be taken to avoid injury to the peroneal nerve when fixing the proximal fibula. The more proximal the corticotomy of the tibia, the faster the bone regeneration. The corticotomy must be distal to the tibial tuberosity in adults and the proximal tibial apophysis in children. The more proximal the corticotomy, the less room remains for fixation and. the greater is the problem with axial deviation during lengthening. Distal tibial lengthenings usually are reserved for patients with associated distal tibial deformities 'or those in whom there is a contraindication to proximal lengthening. Foot fixation frequently is recommended with distal tibial lengthening to prevent equinus deformity. Double-level lengthening is more complicated and more painful. It is indicated in cases of discrepancy greater than 5 ern, extensive limb lengthening for stature, or multilevel deformity (Fig 6).

Pin infections aside, the two most common problems that arise during tibial lengthenings are joint contracture and axial deviation. There is a tendency for ankle equinus and knee flexion contractures to develop. Both are due to Increased tension in the gastrocnemius/soleus muscles. To prevent this complication, a dynamic knee extension splint and an ankle dorsiflexion support shoe are worn at night and, if necessary, during the day to keep both ends of the triceps surae muscles under maxi-

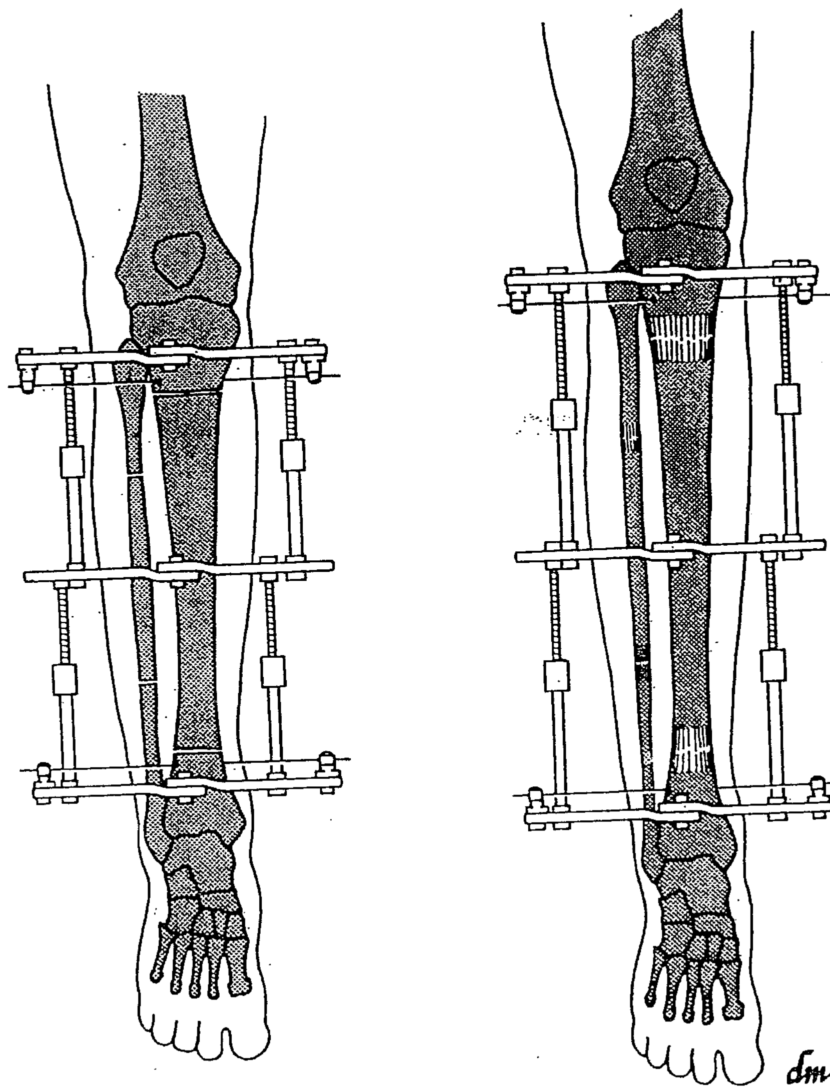


FIGURE 6.
Schematic of double-level tibial lengthening.

mum. stretch. This is felt to stimulate the muscle to regenerate. Our experience with these contractures has shown us that the knee flexion contracture always stretches out, even in severe cases. The ankle equinus contracture usually will not stretch out. Although it may come to neutral with a lot of physical therapy, it usually will not allow return of dorsiflexion. Furthermore, it only may appear to come to neutral, when in fact, the subtalar joint is being forced into valgus. If equinus develops during lengthening, it is preferable to insert wires into the heel both to prevent further contracture and to allow gradual distraction to correct the contracture. Prophylactic tendo-achilles lengthening (percutaneous) can be performed at the index surgery in high-risk or preoperatively contracted cases. Tendo-achilles lengthening should be performed open if done during treatment to avoid a pulmonary embolism from calf manipulation.

Posttreatment residual contracture can be successfully treated by percutaneous or open technique.

Bone deformities also may arise during lengthening. Axial deviation occurs because there is an imbalance in the soft-tissue tension generated by distraction around the bone. Since the bone is not in the center of the soft-tissue mass surrounding it, there is more muscle and soft-tissue mass on some sides of the bone than on others. In the tibia, there is muscle on the lateral and posterior sides, but no muscle on the anterior and medial sides. Furthermore, the intermuscular septum between the tibia and the fibula also acts as a posterolateral tether, especially when the tibia and fibula are cut at different levels. Lengthening produces unequal tensile forces around the bone. If this imbalance of tension is high enough to bend the pins of the apparatus, the bone will deviate in the direction of the larger muscle mass. In the proximal tibia, valgus and procurvatum deviation occur together with lateral and posterior translation, respectively. These deformities often are missed or overlooked during the lengthening process, resulting in fixed bony deformity if the bone consolidates before they are recognized and corrected. Malalignment during lengthening must be expected and looked foron long alignment radiographs. The Ilizarov frame can be adjusted using hinges to achieve the deformity correction. This is one of the major advantages of using circular external fixation over monolateral external fixation for limb lengthening.

Joint instability also must be considered in tibial lengthening. Knee joint instability usually is not a factor, unless the knee is dislocated completely. The main joints to consider are the ankle and subtalar joints. In congenital cases, valgus instability is frequently present. The joint at risk is one with a proximally migrated, hypoplastic, or absent lateral malleolus. Ball-and-socket ankle joints and subtalar coalition often are associated with ankle instability. In these cases, it is important to fix the foot with a lateral olive wire to prevent subluxation. If the fibula is migrated proximally in the ankle mortice, it can be brought down gradually with one wire.

Paley evaluated the results of 67 tibiallengthenings performed on 60 patients: 33 children with 33 ttbinl lengthenlngs and 27 adults with 28 tibiallengthenings. The mean age of the children was 11 years (range, 2 to 19 years) and that of the adults was 26 years (range, 22 to 60 years). The etiologies of the limb length discrepancy or short stature are shown in Table 1.

Using an index designed to evaluate the complexity of each individual case, the patients were grouped according to mild, moderate, and severe difficulty of lengthening. The majority of these patients fell into the moderate-difficulty group. The mean lengthening was 6.7 em in children (range, 2.5 to 16 cm) and 4.3 em in adults (range, 1.5 to 9.6 em). The foot was treated in 6 children and 11 adults, and the femur was treated at the same time in 6 children and 5 adults.

Single-level lengthening was performed in 24 children and 23 adults. Double-level lengthening was performed in 15 children and 5 adults. The mean lengthening for single level was 4.8 em in children (range, 2.5 to 9 em) and 4.3 cm in adults (range, 1.5 to 5 em], The mean double-level

TABLE 1.
Etiologies of Limb Length Discrepancy or Short Stature in 67 Tibial Lengthenings

Etiology	Children, %	Adults, %
Congenital	31	7
Dyplasias	46	0
Posttraumatic	3	79
Miscellaneous	20	14

lengthening was 5 em in children (range, 5 to 16 em) and 7.5 em in adults (range, 4.2 to 9.6 em). The treatment time for single-level lengthening was a mean of 5 months in children (range, 3 to 9.5 months) and 5.8 months in adults (range, 3.6 to 9 months). The mean treatment time for double-level lengthening was 5.8 months in children (range, 3 to 8 months) and 6.8 months in adults (range, 3.6 to 12 months). The external fixation treatment time index measured as the months of treatment in external fixation per centimeter of lengthening was 1.1 month/em for single-level lengthening and 0.6 month/em for double-level lengthening in children (Figs 7 and 8). In adults, these values were 2.3 months/em and 1.1 month/em, respectively.

FIGURE 7.
A, a patient with achondroplasia is seen standing in front of her father for reference (height, 3 ft, 11,5 in.). H, after bilateral double-level lengthening of the tibias, bilateral humeral lengthening, and bilateral lengthening over rods in the femur, her height is 5 ft, .25 in. This course of treatment was achieved over 3 years.

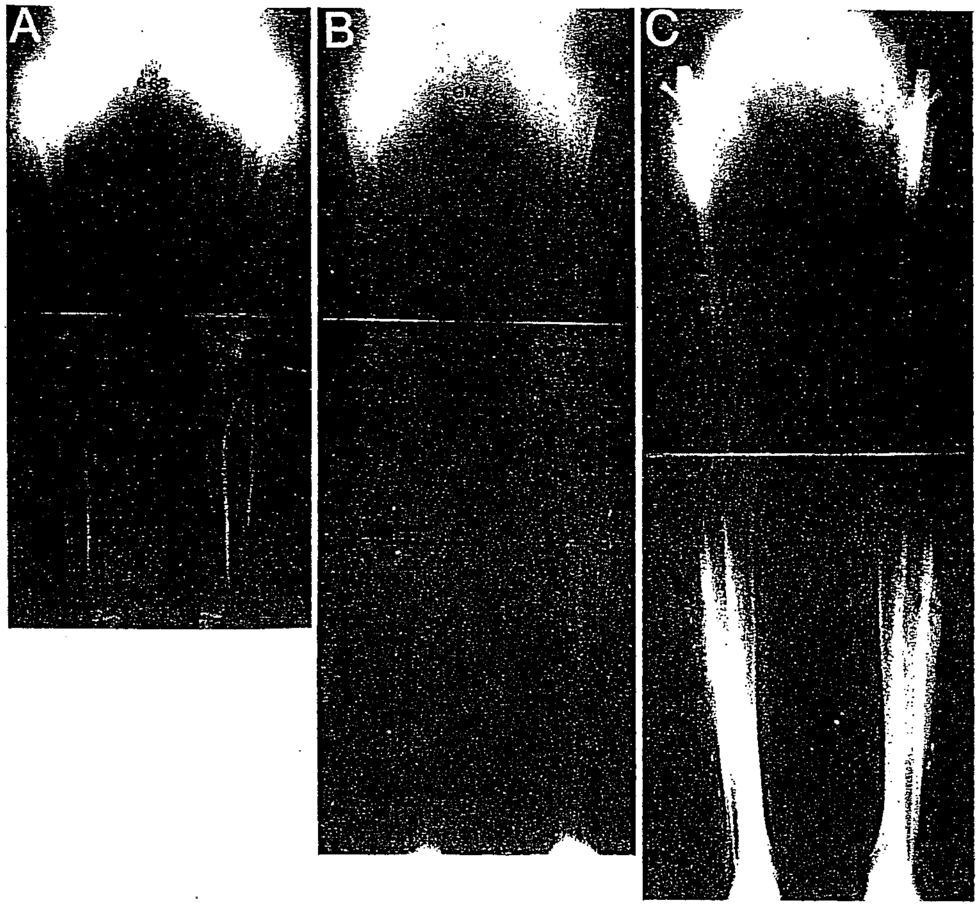


FIGURE 8.
A, Preoperative radiograph. E, after.6V2-in. bilateral double-level tibial lengthening. C, after 4-in. bilateral proximal femoral lengthening over intramedullary rods.

The complications that occurred during treatment included incomplete osteotomy in 1 child and 3 adults, and premature consolidation in 1 child and 5 adults. Eight of these 10 individuals were treated by repeat corticotomy, and 2 spontaneously broke.

Superficial pin infection developed in 5% of the children and 20% of the adults. Pin bone infection occurred in none of the children and 3 of the adults. Superficial pin infections were treated by local pin care or oral antibiotics. In 3 patients with cellulitis, the pin had to be removed.

Peroneal nerve injury occurred in 4 of the children and none of the adults. In 3 of these children, the level of the fibula was abnormally high or low, indicating that the level of the nerve was unpredictable. Two patients recovered fully, but the other two recovered only partially, residua being an EHL palsy in 1 patient and a weakened tibialis anterior in the other. Three peroneal nerve injuries occurred intraoperatively: one from the fibular osteotomy, one from an acute correction of valgus to varus, and one from wire insertion. One peroneal nerve palsy occurred during distraction when the deep peroneal nerve became stretched over a wire that had entered between the branches of the peroneal nerve.

Axial deviation was present in 8 of the children and 1 of the adults. All but 1 of these patients had complete correction of the deformity. Two patients remain with mild residual axial deviation of less than 5 degrees.

Knee flexion contracture developed in 2 of the adults and none of the children, and ankle equinus contracture developed in 1 of the adults and 2 of the children. Almost every patient developed a transient mild knee contracture that resolved during treatment with physical therapy. Only 1 patient with bilateral treatment developed severe contractures that were treated by extending the frame to the femur, followed by gradual distraction to eliminate the contractures. Since this patient was treated, similar contractures have been dealt with by physical therapy alone, with full success. The ankle equinus was treated by a tendo Achillis lengthening in 1 patient and by inserting wires into the foot in the other 2 patients. No residual equinus remained.

One adult patient who mistakenly distracted at a rate of 4 mm/day instead of .25 mm four times per day developed a bone cyst that was bone-grafted.

Two children had fasciotomies, 1 as a prophylactic measure due to an intraoperative hematoma and the other for a suspected compartment syndrome that turned out not to be a compartment syndrome. So far, no true compartment syndromes have been diagnosed following a corticotomy. Watson measured serial compartment pressures following corticotamy and was unable to demonstrate any significant increase in pressure.P Since the corticotomy is subperiosteal, there should be no communication with the compartment.

Refracture was noted in 1 child and 1 adult. The regenerate bone buckled in the child, leading to a l-cm loss of length, but not deformity. The regenerate bent in the adult, causing angular deformity.

Fibular nonunion occurred in congenital hypoplastic fibulas in 3 children. These bones were very thin to begin with and it is thought that the regenerate broke off like pulled taffy.

In total, there were 19 children and 11 adults who had no complications. Nine children required 13 unplanned surgical interventions and 10 adults required 14 unplanned surgeries for treatment of the aforementioned complications. Excluding superficial pin infections, there were a total of 17 soft-tissue-related complications in children compared to 4 in adults. There were 8 bone-related complications in children compared to 13 in adults. The fact that children had a higher propensity for soft-tissue-related complications and adults had a higher risk for bony complications probably reflects the high incidence of congenital disorders in the pediatric group and the slower bone healing and sclerotic posttraumatic bone in the adult group.

The goals of treatment of the patients in this study were (1) to achieve the goals of lengthening within 1 cm, (2) to correct associated deformities to less than 5 degrees; and (3) to maintain (within 15%) or improve the joint range of motion and gait compared to the preoperative state.

These goals were achieved in 38 (97%) of the children and 26 (93%) of the adults. This study demonstrates that the goals of treatment can be achieved despite complications in this difficult group of patients.

FEMORAL LENGTHENING

Femoral lengthening can be carried out proximally, in the mid diaphysis, or distally. Double-level femoral lengthening rarely is indicated and is fraught with complications. The advantages of distal femoral lengthening is that a long leg cast or cast brace can be used to protect the leg after fixator removal. With proximal lengthening, protection would require a spica cast. The femur is broadest at its distal end; therefore, the regenerate is broad. Proximal lengthening is through diaphyseal bone and, in congenital cases, through narrowed, poorly healing bone. The muscle forces on the proximal femur also are much higher, predisposing to axial deviation and late refracture or angulation. The main advantage of proximal femoral lengthening is that it is farther away from the knee joint. In cases of pre-existing knee joint stiffness and pathology, this is advantageous.

The most common and significant problems associated with femoral lengthening (aside from pin infections) are joint instability of the hip and knee, joint stiffness of the knee, and axial deviation. Pin infections are more problematic with this procedure because of the large soft-tissue bulk around the bone. The pins in the fleshy parts of the thigh must be wrapped well to minimize pin skin motion, which promotes infection. The proximal femur does not lend itself well to wire or full ring fixation. For this reason, threaded half-pins are used in the proximal femur and, more recently, in the distal femur. The fixator uses a partial ring called an arc. It is located anterolaterally so that it does not interfere with sitting and lying.

Hip joint stability is assessed radiographically. The center-edge (CE) angle is a good measure of acetabular coverage. If this angle is over 20 degrees, the hip is considered stable. The pelvis must be level to make this assessment. Hip arthrography with push-pull films is helpful in assessing joint stability. Finally, a three-dimensional reconstruction computed tomographic scan is very useful in determining the location of the deficiency. For superior and lateral coverage, a Salter osteotomy is recommended. For posterior coverage, a Dega osteotomy or shelf can be considered. These should be performed prior to limb lengthening.

Knee joint stability can be assessed clinically. If there is anteroposterior or rotatory instability, the apparatus should be extended across the knee to prevent subluxation or dislocation. The hinges allow the knee to flex and extend while preventing any translational or rotatory subluxation. Extending the apparatus across the joint also allows correction of pre-existing or lengthening-related joint contractures, especially knee flexion. Knee joint subluxation during lengthening is due to the pull of the hamstrings on the tibia. The tibia will subluxate posteriorly on the femur. This produces a characteristic ski slope appearance to the front of the knee as a result of the tibia dropping back. A dynamic knee extension splint can be used at night to prevent knee subluxation.

The characteristic loss of motion during femoral lengthening is a knee extension contracture with loss of knee flexion. This is paradoxical, however, since contractures usually occur to the bulkiest muscle groups, which in the thigh, are the hamstrings and the adductors. The hamstring contracture is resisted by the quadriceps contracture, further jamming the knee and increasing the pressure on the joint cartilage. This is further complicated by knee subluxation in some cases. To prevent knee stiffness, it is important to work on knee flexion while maintaining knee extension to prevent subluxation. Rehabilitation is extremely important for all lengthening procedures, but especially for femoral lengthenings. Although most complications are resolvable, the greatest concern is for joint complications that may be irreversible. These include joint stiffness, arthrosis, contracture, and subluxation. Preoperative stiffness or arthrosis may be a relative contraindication to treatment. Lengthening in such patients requires soft-tissue releases to decrease joint pressure and distraction across the joint during lengthening. Carroll and colleagues and, more recently, Bell have shown that lengthenings of 11% to 30% in normal sheep and dogs, respectively, consistently produce articular cartilage damage.[42,43] Loss of hip joint space was noted by Hiroshima in 3 of 26 femoral lengthenings 13 to 24 months after they were compared. Clinically, loss of joint space has not been a complication that we have noticed following limb lengthening. Patients with preoperative joint space narrowing, however, are at a higher risk for worsening of their arthrosis if appropriate precautions are not taken. As previously mentioned, these precautions include extending the apparatus across the joint and maintaining the joint in a decompressed state by distracting across it.

Axial deviation in the femur usually occurs into valgus procurvatum for distal femoral corticotomy lengthenings and into varus procurvatum for proximal femoral lengthenings. Recognition, prophylaxis, and correction are essential to avoid any fixed bony deformities.

Paley and coworkers evaluated the results of 36 femoral lengthenings performed on 35 patients.[40] The average age of the patients was 24 years (range, 6 to 63 years). These patients were broken into two groups: those under 20 years of age (N = 17, group 1) and those over 20 years of age (N = 18, group 2). There were 21 single-level femoral lengthenings and 15 two-level femoral lengthenings, 15 associated tibial lengthenings, and 4 hip osteotomies. The average number of surgical procedures performed per patient was 1.8 and the mean follow-up period was 2.2 years (range, 6 to 36 months). The etiologies of the limb length discrepancies are detailed in Table 2.

TABLE 2.
Etiologies of Limb Length Discrepancy in 36 Femoral Lengthenings

Etiology	Group 1 (<20 yr)	Group 2 (>20 yr)
Congenital	9	4
Dysplasias (Ollier's, MED)	3	0
Growth arrest (traumatic Perthes', postinfection, postirradiation)	2	3
Posttraumatic	2	11
Miscellaneous (pelvic resection for Ewing's sarcoma)	1	0

Using an index designed to evaluate the complexity of each individual case, the patients were divided into groups with mild, moderate, and severe difficulty of lengthening. There were 10 patients in the mild group (scores 1 to 4), 13 in the moderate group (scores 5 to 8), and 12 in the severe group (score 9 or more). The complications per patient by difficulty index were as follows: 4/10 complications per patient in the mild group, 3/13 complications per patient in the moderate group, and 14/12 complications in the difficult group. The complications per patient by diagnosis were congenital 8/13, dysplasia 1/3, growth arrest 1/5, posttraumatic 6/13, and miscellaneous 5/1.

The preoperative leg length difference averaged 8.4 em (range, 2 to 22 em). Group 1 averaged 9.1 em; group 2 averaged 7.6 em. The residual postoperative length difference wast em (range, 0 to 6 em). Three of the patients in this group were excluded, as they were overlengthened intentionally to allow for subsequent limb length discrepancy. Group 1 averaged 1.1 em; group 2 averaged 0.8 em. The percent of femur lengthened was 24.5% (range, 4% to 65%). Group 1 averaged 30.3%; group 2 averaged 15.8%. The length of time treated with the Ilizarov fixator averaged 6.7 months (range, 2.5 to 14.8 months). Groups 1 and 2 both averaged 6.7 months. The lengthening index (months in external fixation per centimeters of lengthening) averaged 1.3 months/em (range, 0.7 to 2.9 month/em). Group 1 averaged 1.1 month/em; group 2 averaged 1.6 month/em.

The complications that occurred during treatment are outlined in Table 3.

Thirty-three of the 35 patients who underwent femoral lengthening had satisfactory (good or fair) results by strict assessment criteria based on physical examination, radiographic results, and functional assessment. Thirty-two of the 35 patients achieved the goal of surgical lengthening

TABLE 3.
Complications Occurring in 36 Femoral Lengthenings*

Type of Complication	Group 1	Group 2
Soft-tissue		
Femoral nerve palsy	1	0
Loss of knee motion	2	0
Loss of hip motion	1	0
Joint subluxation (knee)	1	1
Axial deviation	0	1
Bone		
Premature consolidation	0	1
Buckle or fracture of regenerate	6	1
Other		
Psychiatric reaction	1	0

*Seven complications in patients in group 2 resolved nonoperatively (grade I], four resolved operatively (grade 2), and ten are unresolved (seven of these are grade 3a, five of which will be corrected with planned surgeries; and three are grade 3b).

within 1 cm while maintaining or improving function. There were two poor results due predominantly to loss of knee motion.

FOREARM LENGTHENING

Lengthening of the forearm is indicated primarily for discrepancies between the length of the radius and that of the ulna. With the Ilizarov apparatus, however, the patient can lengthen and correct deformities simultaneously in one or more bones. Furthermore, multiple levels of fixation allow multiple levels of correction. Other applications include bone defects, nonunions, and congenital pseudoarthrosis.

Shortening of the forearm was classified into six types by Paley (Fig 9): type 1, shortening of the radius only; type 2A, shortening of the ulna only; type 2B, shortening of the ulna with dislocation of the radial head; type 3, absent radius; type 4, shortening of both bones to the same proportion; and type 5, shortening of both bones to different proportions.[45] The apparatus configurations and wire placement are specific for each condition.

In the forearm, there are some special considerations. Stability of the elbow usually is not a concern in forearm lengthening. Stability of the wrist is important, especially with radial lengthening. With ulnar lengthening, a wire through the proximal radius is needed to prevent distal migration of the radius. When performing radial lengthening alone, there is no need to fix the ulna. Flexion contracture of the elbow, wrist, and fingers tends to develop with radial lengthening and with one-bone forearm lengthening. This should be countered by vigorous physiotherapy and extension splints. Due to the small diameter of the forearm bones, large lengthenings tend to narrow the diameter of the regenerate bone formation because of the pinch of the surrounding muscles. Inevitably, this leads to the "pulled-taffy effect" (narrowing of the center as the ends are stretched). The rate of 1 mm/day may need to be reduced to .75, .50, or .25 mm per day. Axial deviation usually is not a problem with the radius, since the ulna acts as support to it. Proximal ulnar corticotomies tend to go into flexion, especially in one-bone forearms. This should be recognized and corrected.

Villa and associates reported the use of the Ilizarov technique for lengthening 13 forearms in 12 patients.[45] The lengthening ranged from 2 to 13 cm (10% to 143%). Bone consolidation was achieved in 3 to 19 months without the need for bone grafting. Eleven of the 12 patients were improved functionally and cosmetically. Complications included three temporarily deep radial nerve palsies, one sympathetic dystrophy, one malunion, one delayed malunion, two refractures, and three mild losses of motion.

Despite these complications, the goals of treatment were achieved in all patients, with few permanent residual problems. The Ilizarov method is a reliable, successful alternative for the treatment of forearm length discrepancy and deformity problems (Fig 10).

Tetsworth and colleagues reported their results with 13 forearms treated with the Ilizarov technique.[46] Lengthening of the ulna ranged from 3.4 to 11.7 cm (58% mean increase) and the treatment time averaged 6.1

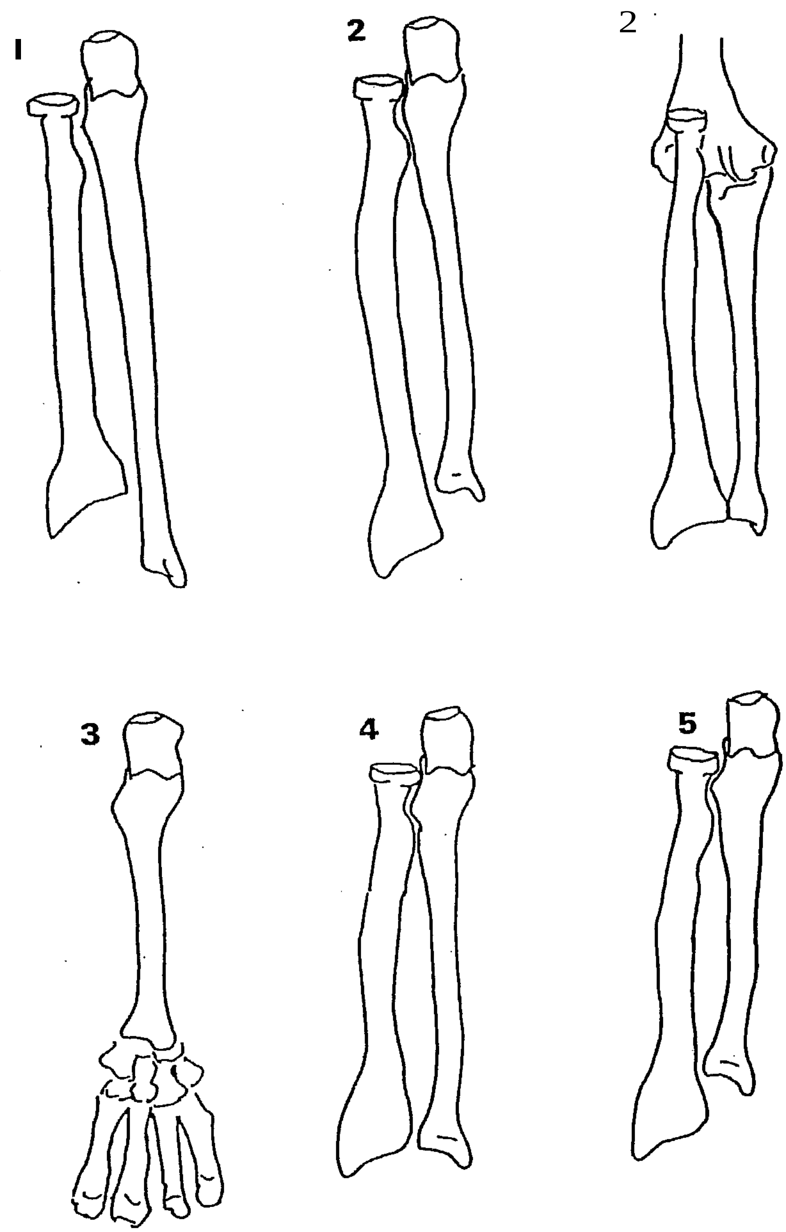


FIGURE 9.
Classification of shortening of the forearm: type 1, shortening of the radius only; type 2A, shortening of the ulna only; type 2B, shortening of the ulna with dislocation of the radial head; type 3, shortening of ulna with absent radius; type 4, shortening of both bones to the same proportion; and type 5, shortening of both bones to different proportions.

months. Lengthening of the radius averaged 1.7 to 6.2 em (23% mean Increase) and the treatment time averaged 5 months. Eleven of the 13 patients had simultaneous correction of two deformities. Complications included superficial pin tract infections, premature consolidation, angulation of the regenerate after the fixator was removed, transient radial nerve injury, and diminished wrist range of motion,

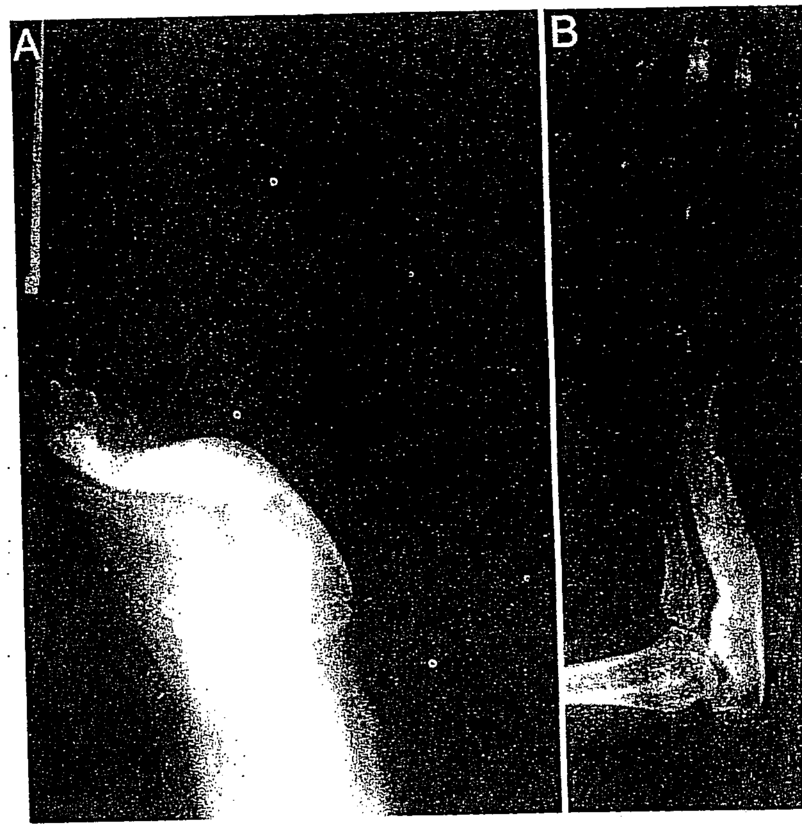


FIGURE 10.
A, radiograph showing secondary deformities following centralization for club hand. **B,** the forearm was treated with osteotomy at two levels to correct angular deformities and lengthen 8 cm. (From Tetsworth K, Krome J, Paley D: Orthop Clin North Am, 1991; 22:689. Used by permission.)

HUMERAL LENGTHENING

Lengthening of the humerus is performed for two indications: limb length discrepancy with or without simultaneous deformity correction or in conjunction with lengthening for stature. The most common cause of humeral shortening is premature physeal arrest. This can be due to trauma, tumor, or radiation, but most often is secondary to infection.

Lengthening of the humerus can be performed with either monolateral or circular external fixators. Recently, hybrid constructs using cantilever half-pins in the proximal humerus and transfixation wires in the distal humerus combined the advantages of circular fixation with those of pin choice. The humerus can tolerate lengthenings of 5 cm or less with little or no functional loss. Discrepancies greater than 5 cm are more obvious from both the functional and cosmetic perspectives.

Complications in humeral lengthening include both bone and soft-tissue considerations. These can develop either intraoperatively or during the course of treatment. As in the forearm, lengthening stability and axial deviation are not often problems with the humerus. In large lengthenings, range of motion in the shoulder usually is not compromised if lengthening is performed distal to the deltoid tuberosity. The main con-

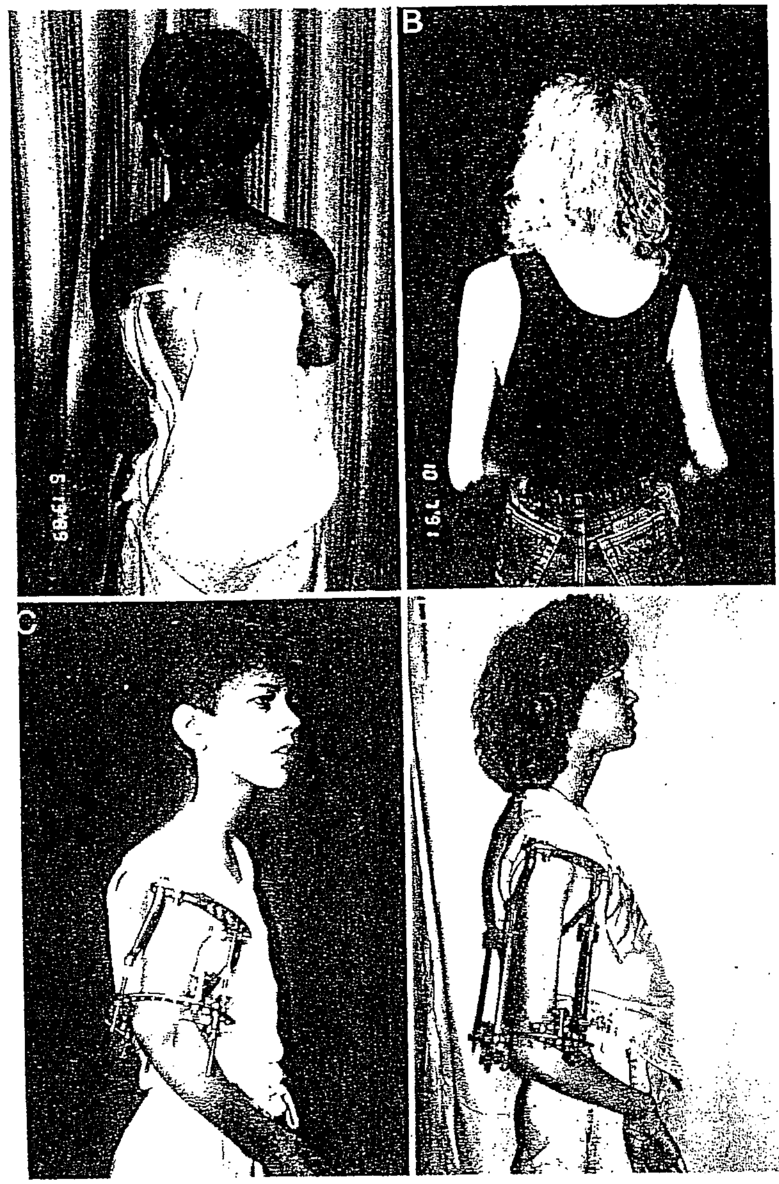


FIGURE 1l.
Seventeen-centimeter humeral lengthening secondary to septic growth arrest. A, before treatment. E, after treatment. C, with Ilizarov apparatus at beginning of lengthening. D, with Ilizarov apparatus at end of lengthening.

sideration in humeral lengthening is the range of motion in the elbow. A dynamic elbow extension splint at night can help prevent elbow flexion contractures as tension on the flexors increases with lengthening. Most complications are related to the soft tissue rather than the bone. The humerus heals faster than most of the bones and is the simplest one to lengthen.

Single-level humeral lengthening heals rapidly and is tolerated well (Fig 11). Double-level lengthening is indicated for large discrepancies or lengthening for stature. This type of lengthening is associated with an increase in pain and risk of complications. Correction of a deformity at one level can be combined with simultaneous lengthening at a second level.

Tetsworth and associates reported a series of humeral lengthenings by single-level distraction in four patients and double-level distraction in two patients.46 The mean lengthening was **11.1** em. The mean increase achieved in the length of the humerus was 62%. Treatment time, defined by the length of time in external fixation, averaged 8.2 months.

The Ilizarov technique for humeral lengthening is a safe and reliable treatment to correct limb length discrepancy with simultaneous correction of deformity.

FEMORAL LENGTHENING OVER AN INTRAMEDULLARY ROD

Limb lengthening by means of external fixation requires distraction and consolidation periods prior to fixator removal. The distraction time is 1 day for every millimeter of lengtheDoing, and the consolidation time ranges from 2 to 4 days for each millimeter. In order to reduce the treatment time significantly, Paley and Herzenberg inserted and proximally locked unreamed nails into the femur and tibia at the same time they applied the fixator (Ilizarov or Orthofix).47 Lengthening was performed at 1 to 2 mmJday until the planned lengthening was completed. The nail then was locked distally and the fixator removed (Fig 12). This technique elimi-

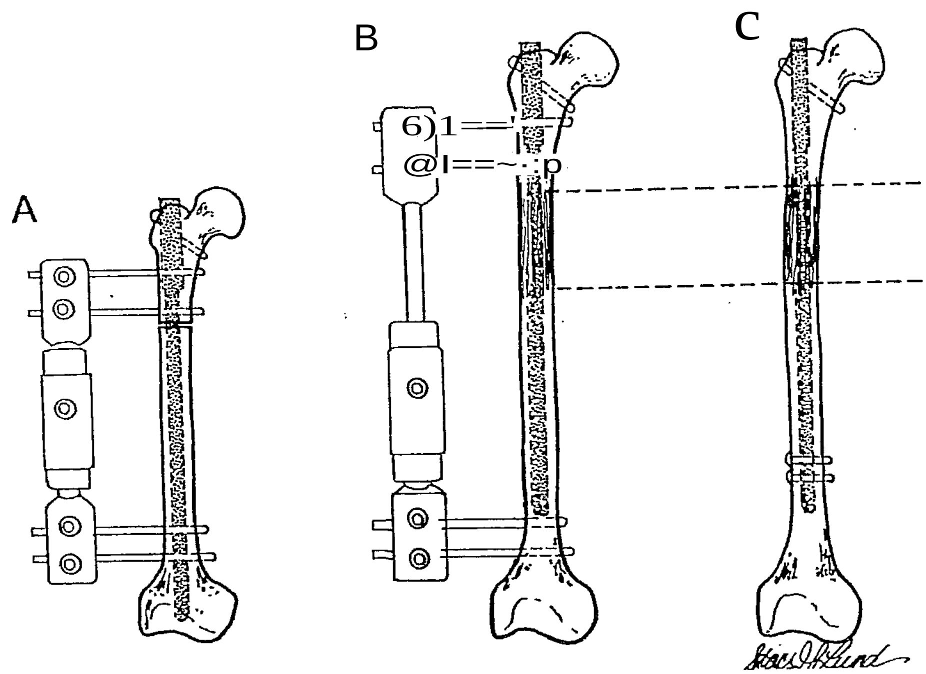


FIGURE 12.
A, lengthening over a proximally locked intermedullary nail using a unilateral external fixator. B, gradual distraction at 1 to 2 mm/day. C, nail is locked distally at the end of the distraction period. The external fixator is then removed. The nail acts as an internal splint during the consolidation phase.

The pseudarthrosis site was treated by closed end-to-end compression in 6 cases (Fig 13, A) and by side-to-side compression after overlapping of the bone ends in 1 case (see Fig 13, B). Open reduction with insertion of one fragment inside the other was performed in 1 case (see Fig 13, C). splitting of the proximal fragment with widening of the bone followed by closed insertion of one fragment into the other was performed in 1 case (see Fig 13, D). The pseudarthrosis site was treated by distraction in 4 cases, 2 for correction of angular deformity and 2 for lengthening. Resection of the pseudarthrosis site was performed in 3 cases. The resulting bone defect was treated by acute shortening and lengthening through a proximal corticotomy in 2 cases and by bone transport in 1 case (Fig 14).

Twelve of the limb segments were treated with lengthening, 8 by distraction through a corticotomy site, 2 by physeal distraction, and 2 by distraction of the pseudarthrosis site. The deformity was corrected in all cases.

Union was achieved with the initial treatment in 15 of the 16 cases. The union rate was 94% with one treatment and 100% with two treat-

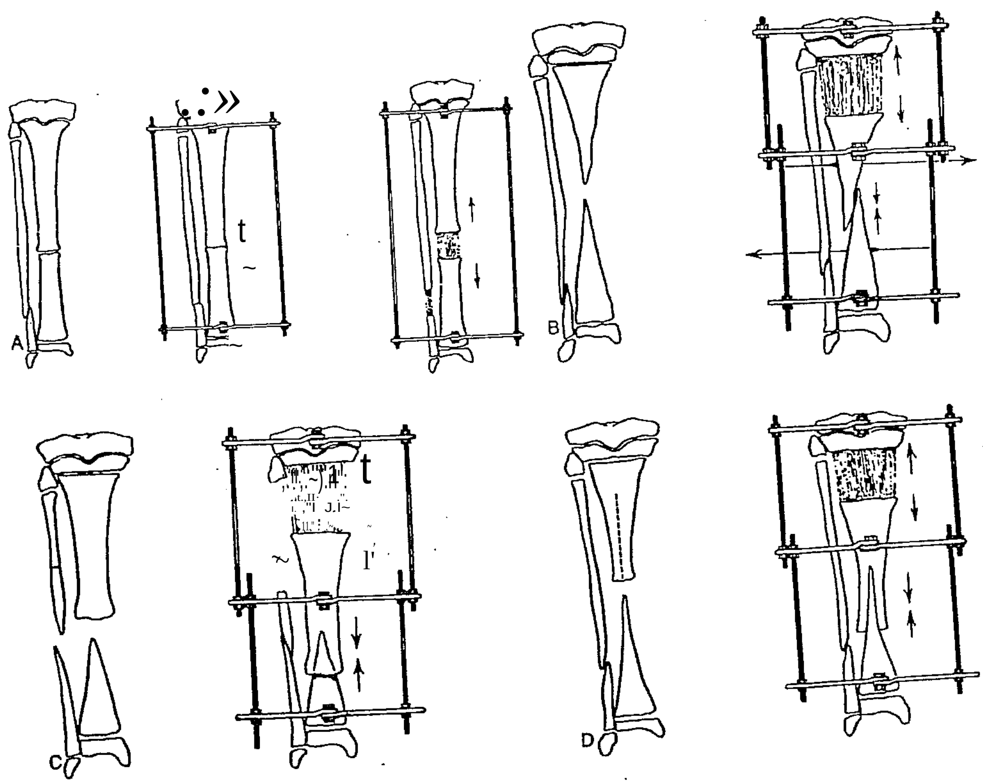


FIGURE 13. A, broad congenital pseudarthrosis of the tibia ends: end-to-end compression; if stiff, may distract through the defect. H, thin, atrophic congenital pseudarthrosis of the tibia ends: side-to-side compression; proximal lengthening. C, broad and thin congenital pseudarthrosis of the tibia ends: insert thin end in broad end open or closed; proximal lengthening. D, broad and thin congenital pseudarthrosis of the tibia ends: split the broad end and insert the thin end to widen; proximal length!3ning. (From Paley D, Catagni M, Argnani F, et al: Clin Orthop 1992; 2BO:81. Used by permission.)

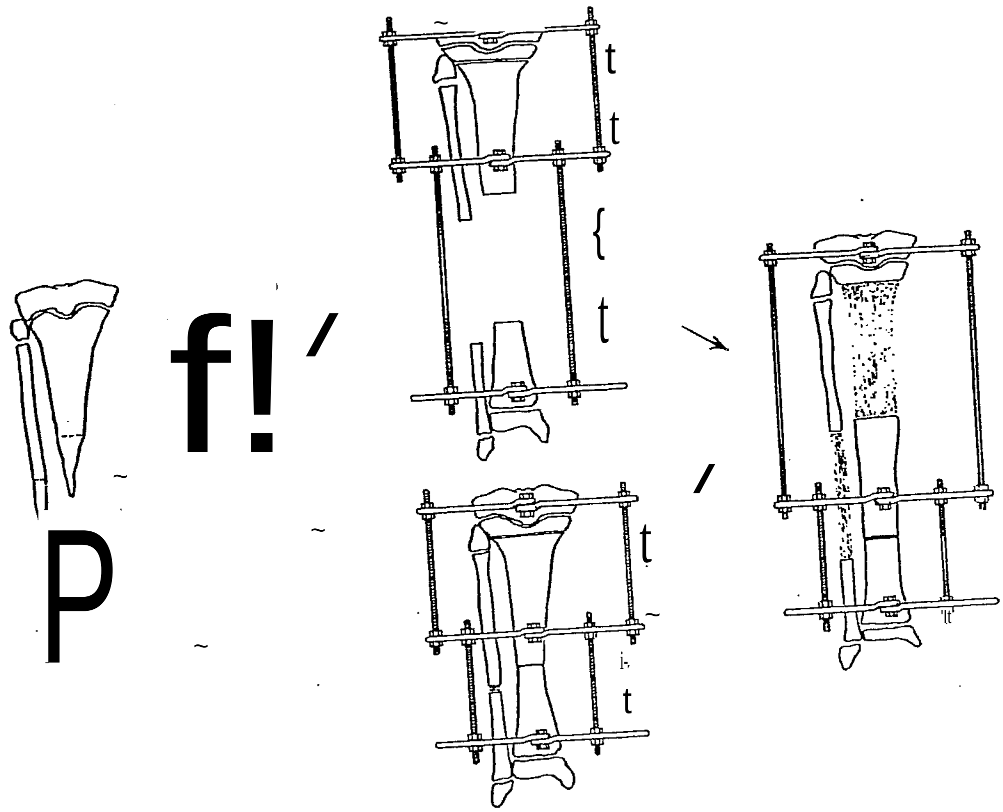


FIGURE 14. Atrophic, mobile thin congenital pseudarthrosis of the tibia ends: resection/bone transport (upper); resection/acute shortening/lengthening (lower). (From Paley D, Catagni M, Argnani F, et al: Clin Orthop 1992; 280:81. Used by permission.)

ments. In 1 case, the initial method of treatment (overlapping the bone ends and side-to-side compression of the pseudarthrosis, and corticotomy lengthening of the leg) failed to achieve union of the pseudarthrosis site and was associated with delayed consolidation of the lengthening site. The apparatus was removed temporarily and reapplied later with resection of the pseudarthrosis and bone transport to fill in the resection gap. Union was achieved by compression of the transported bone ends.

The mean treatment time was 5.6 months (range, 3 to 12 months). Lengthening ranged from 1.5 to 8 cm and was performed on 12 of the 16 cases (Fig 15). Angular deformity was corrected fully at the pseudarthrosis site in all but 1 case. Three cases healed with five-degree valgus axial deviation through the lengthening site.

The original level of the pseudarthrosis maintained a persistent union in 14 of the 16 limb segments throughout the follow-up period. One patient demonstrated a slowly progressive valgus deformity of the proximal tibia, where a five-degree postlengthening deformity through dysplastic bone (fibrous dysplasia) was left uncorrected. One patient developed a valgus buckle fracture of the regenerate, which was manipulated and subsequently healed straight.

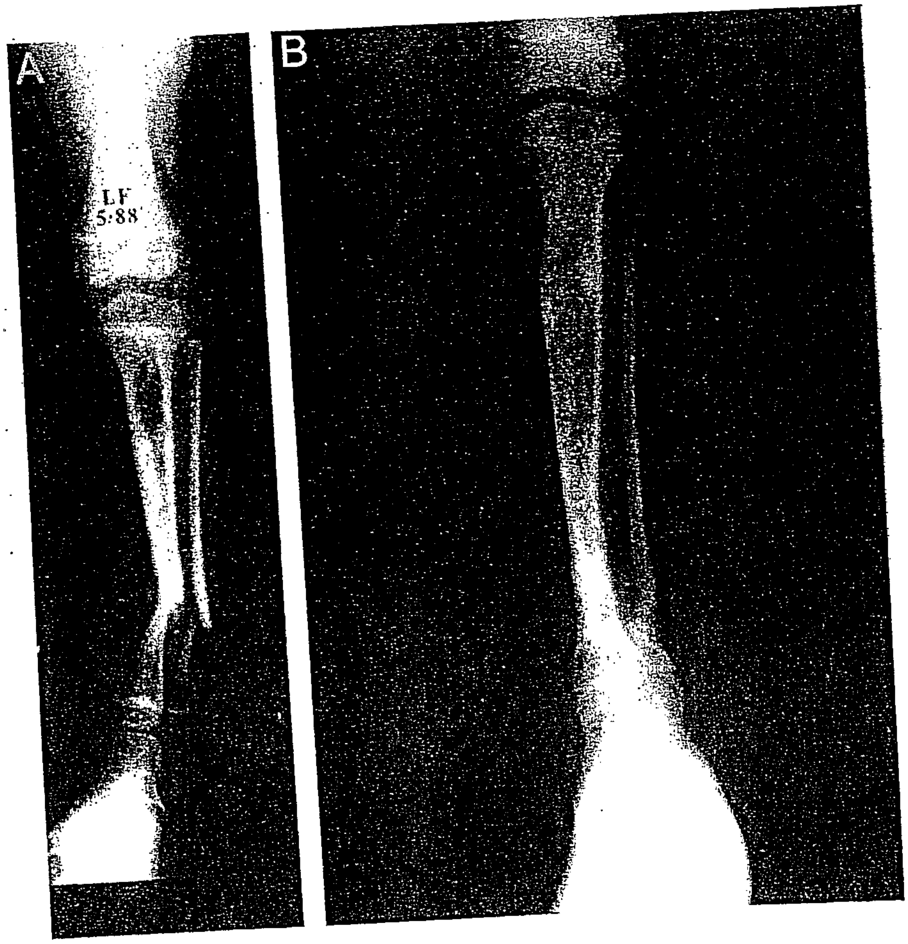


FIGURE 15.
A, preoperative radiograph of congenital pseudarthrosis of the tibia with neurofibromatosis. B, result after resection, shortening, and 8-cm physeal distraction. Treatment time was 5 months. Follow-up was 5 years.

The authors recommend the use of a total-contact patellar tendon-bearing orthosis for added protection until skeletal maturity is reached.

BONE DEFORMITIES AND JOINT CONTRACTURES

The Ilizarov apparatus has modular parts that can be used for deformity correction. The most basic deformity correction unit is the hinge (Fig 16).[3] **With** an osteotomy at the level of an apex of the deformity and the hinge overlying the apex, distraction of the concavity will lead to an opening wedge correction, regenerating a wedge of new bone. If the hinge is placed away from the apex on the convex side, but still at the level of the osteotomy, simultaneous lengthening and deformity correction will occur, with the regeneration of a trapezoid-shaped segment of new bone. Similarly, if the hinge is placed on the concave side of the bone, compression will result from distraction to the concavity. If the hinge is placed prox-

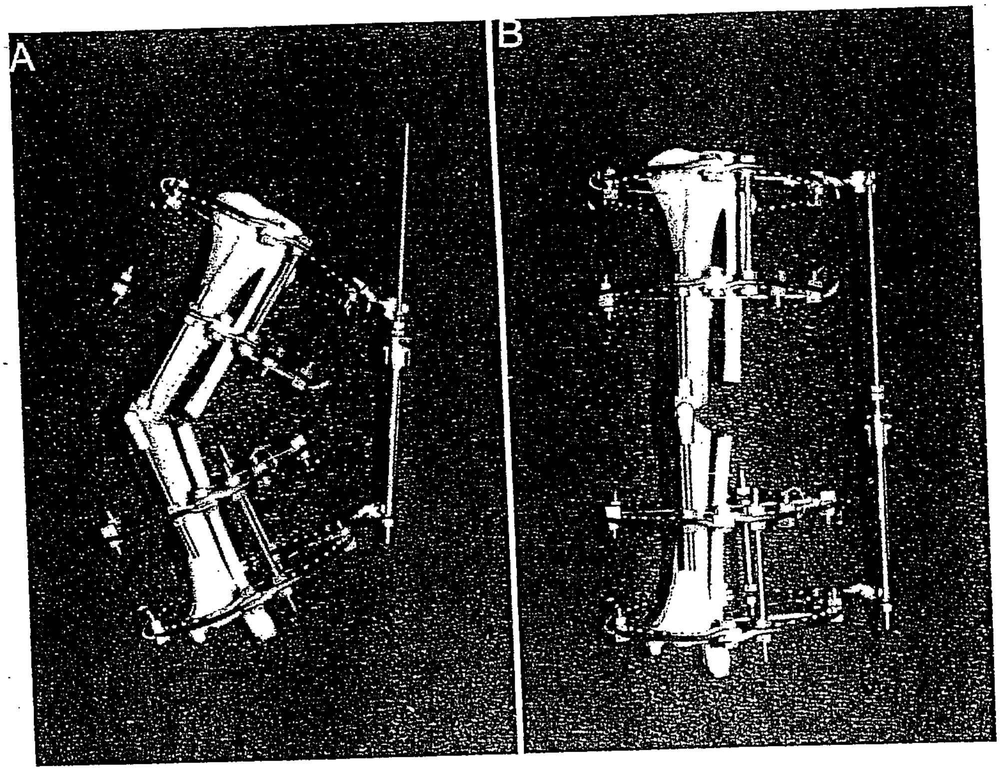


FIGURE 17.
A and B, a standard mounting of the Ilizarov apparatus for an opening wedge angular correction. There are two levels of fixation in each bone segment. Each ring is perpendicular to the shaft of the tibia. The hinge is placed at the apex of the deformity. The distraction rod has a pivot point articulation at each end to allow for auto-adjustment of its angle to the ring. The hinge rods are straight at the end of correction. The distraction rod has a different angle to the ring before and after correction. (From Paley D, Rumley T [r, Kovelman H: Adv Plast Reconstr Surg 1991; 7:1-40. Used by permission.)

an osteotomy is through horizontal threaded rods and posts. This stepwise connection pulls one ring relative to the other along the tangent of the rings. In other words, with three or four of these tangential, horizontal, stepwise connections, the rings will rotate one to the other in a controlled fashion. If all of the horizontal connections are placed parallel to each other such that there are three or four step connections between the rings with horizontal threaded rods, all oriented in one direction, then gradual movement along these threaded rods will translate one ring to the next. In this way, bone segments can be rotated or translated gradually one to the other. More complex configurations even can allow combinations of angulation, rotation, and translation ?imultaneously. The key to deformity correction is preoperative planning. A more detailed discussion of deformity corrections and the methods used to perform them is available in a recent publication.[59]

Any angulation or translation can be defined by four parameters: (1) plane, (2) apical direction, (3) level,, and (4) magnitude. For angular deformities, the Ilizarov apparatus is preconstructed to make sure that the

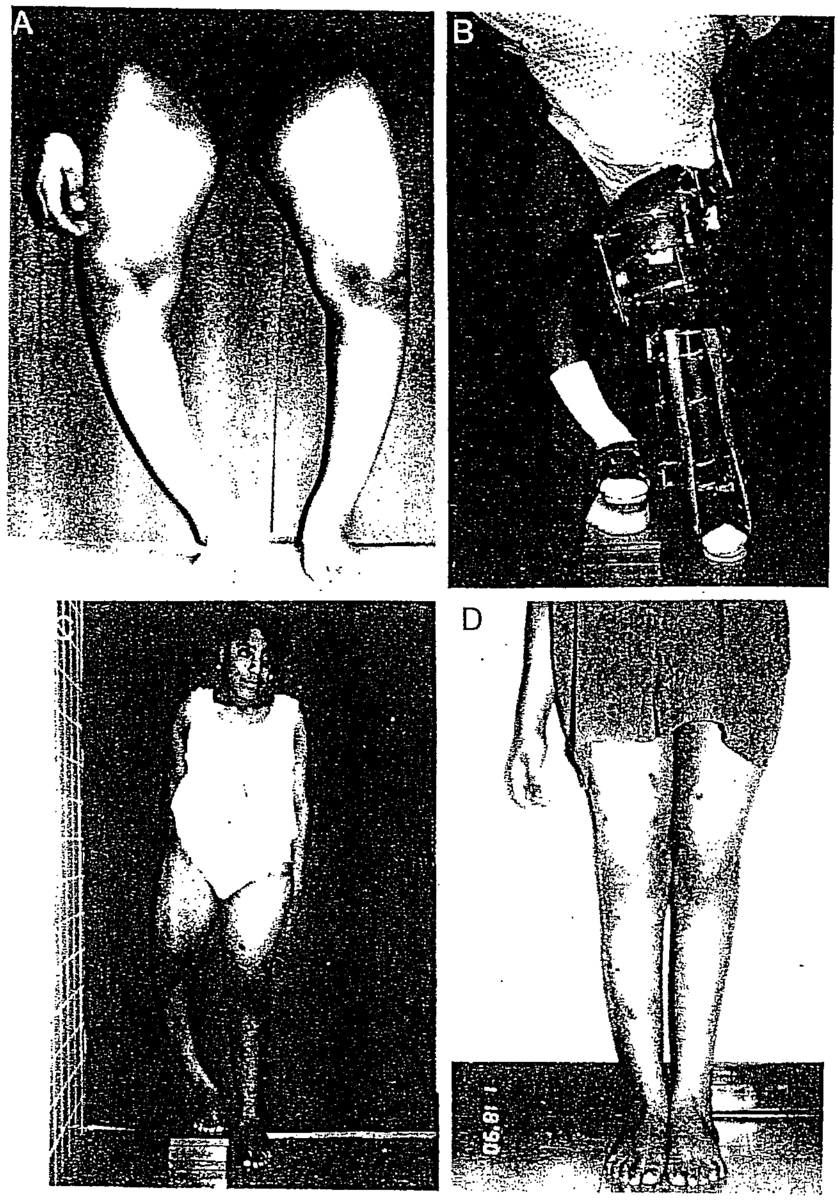


FIGURE 18.

A, severe bony deformities of both lower limbs secondary to hypophosphatemic rickets before treatment. B, simultaneous multilevel correction of left femur and tibia with Ilizarov devices. C, result of correction with 4 in. of length gained due to angular deformity correction. D, final appearance after correction of both lower extremities.

hinges are located at the correct level in the proper plane and are angulated with their apex in the appropriate direction. The true magnitude of angulation at that level in the true plane then is set in the hinges. The apparatus is applied to the limb and the hinges are centered over the correct level of angulation as determined on radiographs. The apparatus is fixed so that it is oriented appropriately on the limb. This places the hinge in fue trne plane of angulation. The osteotomy usually is performed at the same level as the hinges. If the apex of angulation is in the epiphysis or at the joint line, the osteotomy usually is performed in the adjacent metaphysis. In such cases, the hinge still is located at the trne level of deformity. The hinge and osteotomy, therefore, are at two different levels. The hinge will force the osteotomy to translate as the angulation is corrected (Fig 19). This maintains fue proper alignment and joint orien-

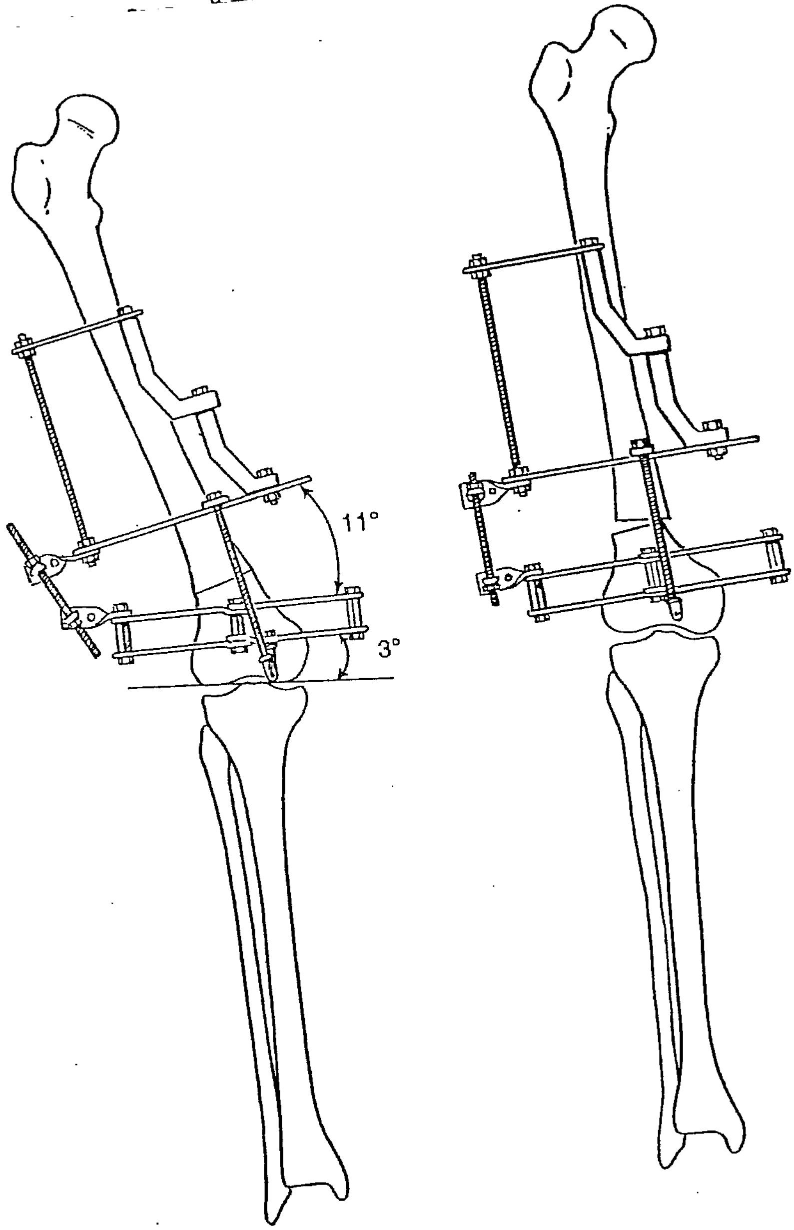


FIGURE 19. ...ic...or hinge. When the center 01 rotation 01 a delonnity Iies at a level not practical for an osteotomy, the osteotomy Is performed at one level while the hinge is placed at the level of the angular center 01 retation. Upon correction, the osteotomY site will angulate and translate. This example demonstrates a distal femoral valgus. **Note that hinges can be placed beloW the rings.**

mechanical axis deviation averaged 9.0 mm. The mean preoperative limb length discrepancy was 2.3 em. The mean postoperative orientation of the knee joint (femoral condyle line) was 87 ± 3 degrees (normal, 87 ± 2 degrees). postoperative orientation of the proximal tibial condyles were 87 ± 3 degrees and 78% (normal, 87 ± 2 degrees). The results following realignment in the earliest 10 cases (those performed before 1989) were compared to the later cases (those performed after 1989). Residual mechanical tibiofemoral angulation averaged 4.6 degrees in the early group and 2.1 degrees in the late group. Residual mechanical axis deviation averaged 13.2 mm in the early group and 4.8 mm in the late group. Statistical analysis demonstrated that the decrease in deformities seen in the late group was statistically significant ($P < .05$). In the late group, the mechanical tibiofemoral angle was accurate to less than 3 degrees of residual deformity in 94% of cases, and the residual mechanical axis deviation was less than 10 mm in 89% of cases. With better preoperative planning and greater experience with the Ilizarov method, gradual angular correction can lead to very accurate deformity correction.

FOOT DEFORMITIES

The Ilizarov apparatus also lends itself well to fixation and correction of foot deformities because of the three-dimensional nature of the foot and of the apparatus. There are two approaches with this apparatus for the correction of foot deformities: with or without osteotomy. In the nonosteotomy technique, the deformity is corrected by distraction of the foot joints and their soft tissues. The correction occurs by eliminating preexisting contractures and bringing joints into new congruous relationships in a plantigrade position. Therefore, one of the prerequisites for nonosteotomy treatment is congruoUS joints with no significant fixed bony deformity. The only exception is in children under the age of 8 years, in whom remodeling of the shape of the foot bones is still possible. These indications are similar to those for soft-tissue release by conventional means. Soft-tissue release relies upon biologic plasticity of cartilaginous bones. Distraction is thought to reshape bones by activation of the circumferential physis of these bones, leading to a new congruouS alignment of the foot bones.

The second method of foot deformity correction is by distraction of foot osteotomies. The indications for correction through bone are fixed bony deformity in patients over the age of 8 years in whom sufficient incongruity of the joints (that would not be expected to remodel) would result from soft-tissue distraction or release. Other indications include patients with neuromuscular imbalance in whom soft-tissue correction would obtain, but not maintain, the correction, and in whom tendon transfer or tenodesis is not possible to maintain the correction. The presence of previous fusions or nonunions is an indication for a bony correction. Finally, some contractures may be judged so stiff that a bony correction is preferable to a soft-tissue one.

Distraction foot osteotomies are classified according to the level of the osteotomy: supramalleolar, hindfoot, forefoot, and combined hindfoot and forefoot.

Twenty-five very complex foot deformities were treated by Ilizarov distraction osteotomies.[53] In addition, the leg was lengthened and widened in the majority of cases. The mean treatment time was 6.4 months. There were 20 minor or major complications related to the foot osteotomies in 18 feet, including deep pin track infection in 3, failure of osteotomy separation in 9, acute postoperative tarsal tunnel syndrome in 2, toe contractures in 3, wire breakage or cutout in 2, and buckle fracture in 1. Nineteen secondary procedures were required in 13 patients to treat these complications. The final result was a plantigrade foot in 22 patients in late follow-up. The 3 nonplantigrade feet were due to unrecognized heel varus (1), ball-and-socket ankle joint (1), and preexisting partial growth arrest with progressive deformity (1). Gait was improved in all cases. Pain was eliminated in all but 2 patients.

Based on these criteria, the results were judged to be satisfactory in 22 patients and unsatisfactory in 3. The Ilizarov method is a successful modality for achieving complex foot deformity correction despite complications.

NONUNIONS, BONE AND SOFT-TISSUE DEFECTS, AND OSTEOMYELITIS

The treatment of nonunions depends upon the nature of the nonunion. Conventionally, nonunions are classified as hypertrophic or atrophic, and infected or noninfected. Ilizarov's approach to nonunions is much more complex and individualized. He considers a wide variety of factors for each nonunion and, based on this, chooses the most appropriate treatment. One of the most important of these parameters is the stiffness of the nonunion. The amount of stiffness present provides information about the tissue between the bone ends. A very stiff nonunion means that the tissue between the bone ends is either dense, fibrous, or fibrocartilaginous, whereas a lax nonunion means that it is either loose connective or synovial. In a stiff nonunion, the potential for bone regeneration from the bone ends is present. The dense fibrous or fibrocartilaginous tissue acts like an interzone and, when put under distraction, regenerates trabeculae of new bone from the nonunion site (Fig 20). In contrast, in a lax nonunion, little or no bone regeneration results upon distraction. Therefore, bone shortening with a stiff nonunion can be treated by lengthening through the nonunion site itself. Lax nonunion requires compression of the bone ends to convert it to a more stiff type and eventual union. Open reduction, freshening of the bone ends, and opening of the medullary canal to allow revascularization may be necessary.

One of Ilizarov's concepts is that of "bone loss" (Fig 21). Most of us think of bone loss as a bone defect. Bone shortening is another type of bone loss. If there is a 6-cm loss of bone, it may be manifested as a 6-cm bone defect, a 6-cm shortening of the limb, or a combination of shortening and bone defect totalling 6 cm. The treatment for each of these situations is different, but similar.[54]

The Ilizarov treatment for a bone defect is a technique called bone transport.[55] In this technique, an intercalary segment is created at one end of the defect by a corticotomy of the bone. The intercalary segment then

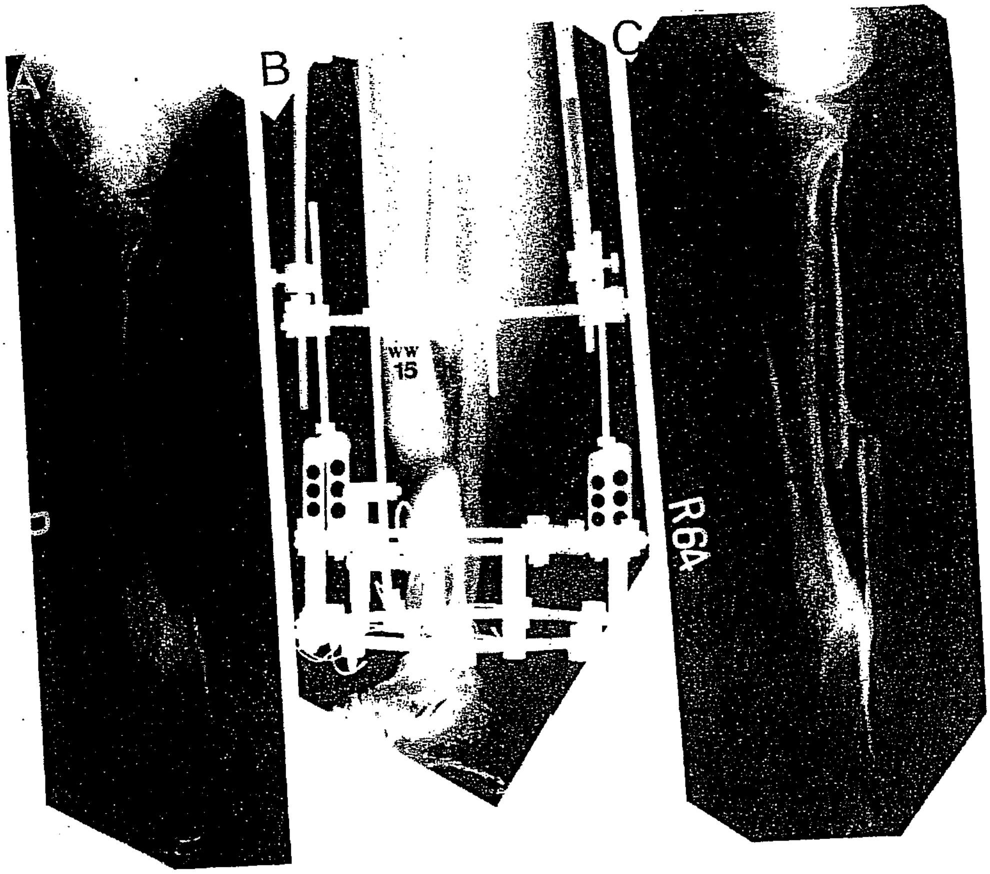


FIGURE 20.
A, stiff hypertrophic nonunion with 3-cm shortening and translational deformities. **B,** new bone formation from distraction of nonunion site. **C,** final result.

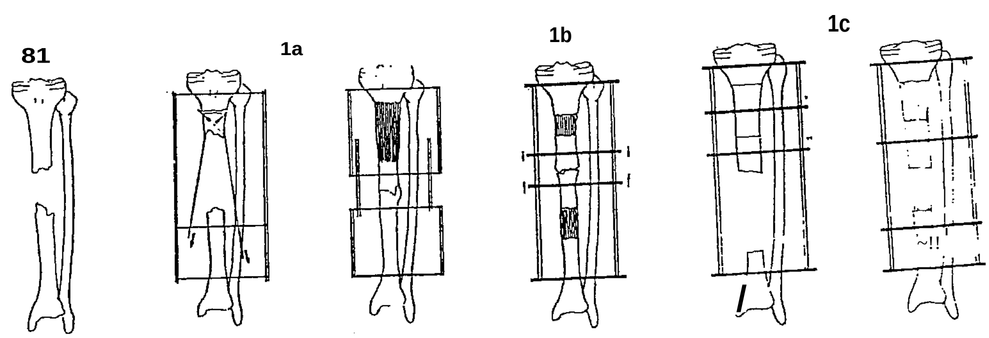


FIGURE 21.
Treatment of type B pseudarthroses. Bl treatment La, internal lengthening (bifocal): bone transport by vertical olive wires or transverse carrier ring followed by compression at nonunion site; sequential distraction-compression osteosynthesis. Bl treatment tb, internal lengthening (trifocal): bone transport by two transverse carrier rings from opposite sides of defect; sequential distraction-compression osteosynthesis. Bl treatment tc, internal lengthening (trifocal): bone transport by two carrier rings from the same side of the defect; sequential distraction-compression osteosynthesis.

is transported at 1 mm/day across the bone defect. Just as in a leg lengthening, bone regeneration occurs between the bone ends. When the migrating fragment reaches the opposite end of the bone defect, it makes contact with the bone there (Fig 22). Compression of the docking site is required to achieve union. A bone graft is required to expedite this union. The original bone defect has been replaced by a distraction gap at a different level, but of the same length. This new bone formation undergoes the same stages as during limb lengthening. When consolidation of both the nonunion site and the distraction osteogenesis site is complete, the apparatus can be removed. In a bone transport, the transported segment is moving within the soft-tissue sleeve. Exactly what is happening at the interface between the transported bone and its surrounding soft tissues is not understood.

The technique of bone transport is very useful in the treatment of osteomyelitis. The infected necrotic bone is resected and the gap is closed treated by bone transport. If there is an associated soft-tissue defect, it is closed by soft-tissue transport. Thus, one can leave the infected wounds of osteomyelitis open to drain, since they will close gradually from the inside as the bone transport carries the soft tissues with it across the gap (Fig 23).[3] This precludes the need for many muscle pedicle or free flaps for the coverage of soft-tissue defects.

Problems of bone transport treatment fall into bony and soft-tissue categories. The bony problems are as follows: (1) Maldocking of the transport fragment with the opposite side. This usually results from malalignment of fixation at the time of frame application. The maldocking usually requires surgery to align the docking fragments so as to ensure a large contact area between the bone ends (Fig 24). (2) Failure of progression of

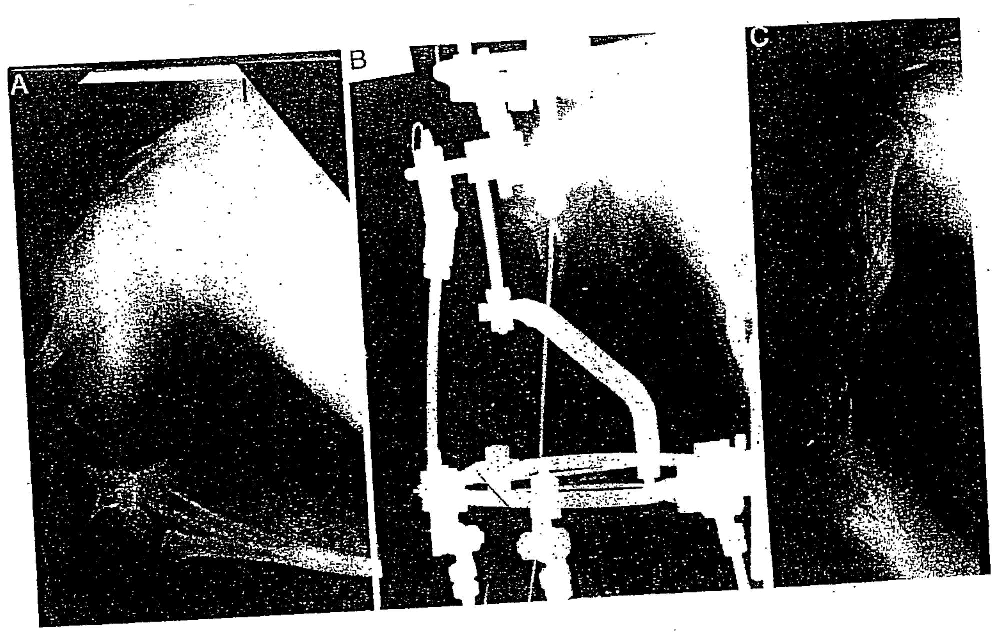
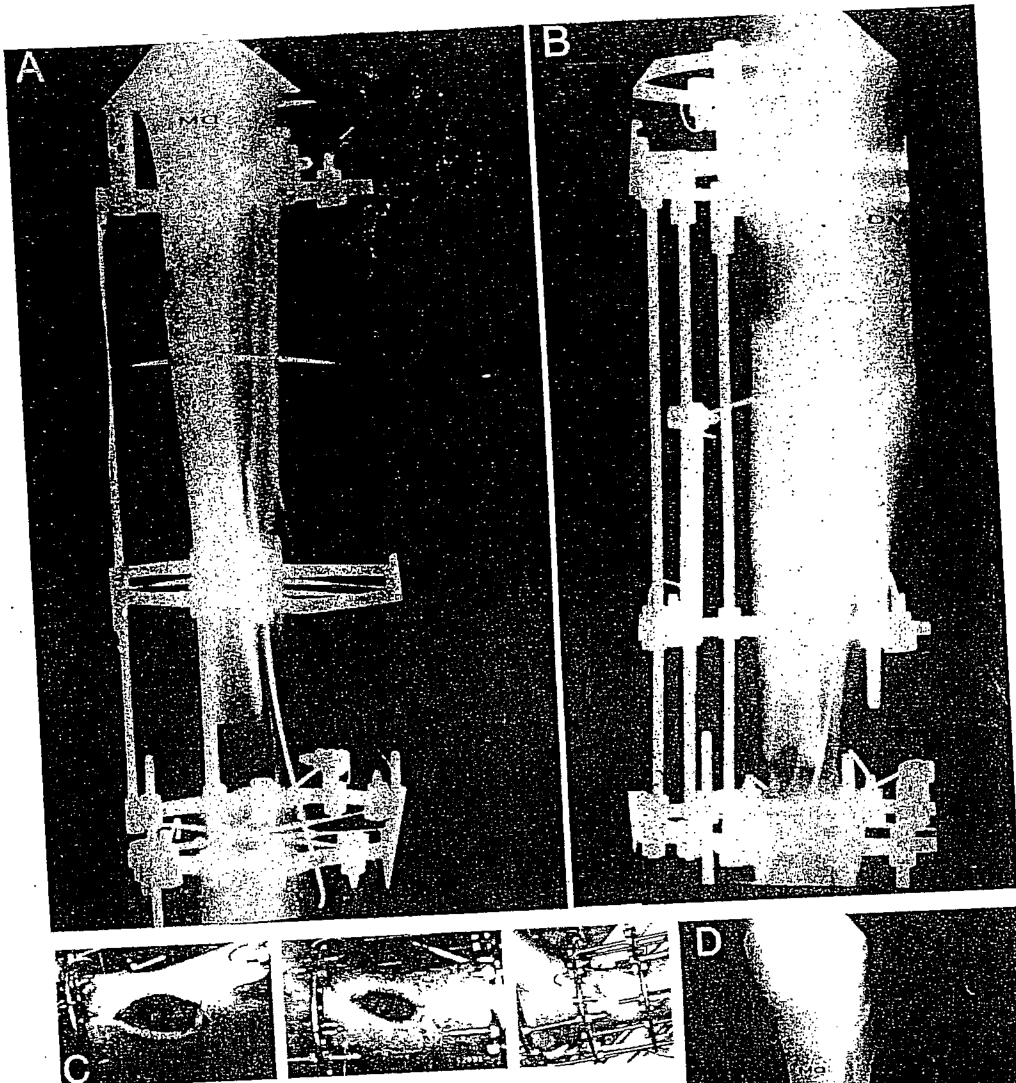


FIGURE 22. A, humeral bone transport with 10-cm bone defect due to neonatal osteomyelitis. B, longitudinal wire transport proximal to distal. C, final radiograph.



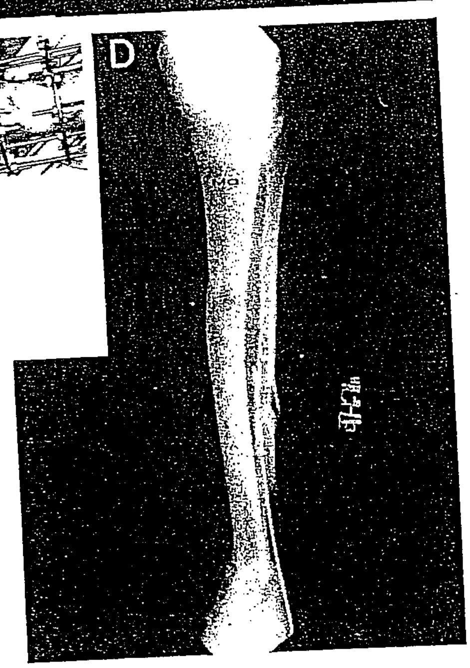


FIGURE 23.
A. 6-cm bone defect secondary to resection of osteomyelitis. **B,** during transport, just before distal docking. **C.** the open wound gradually closed by soft-tissue transport. **D,** final radiograph. (From Paley D, Rumley T Jr, Kovelman H: Adv Plast Reconstr Surg 1991; 7:1-40. Used by permission.)

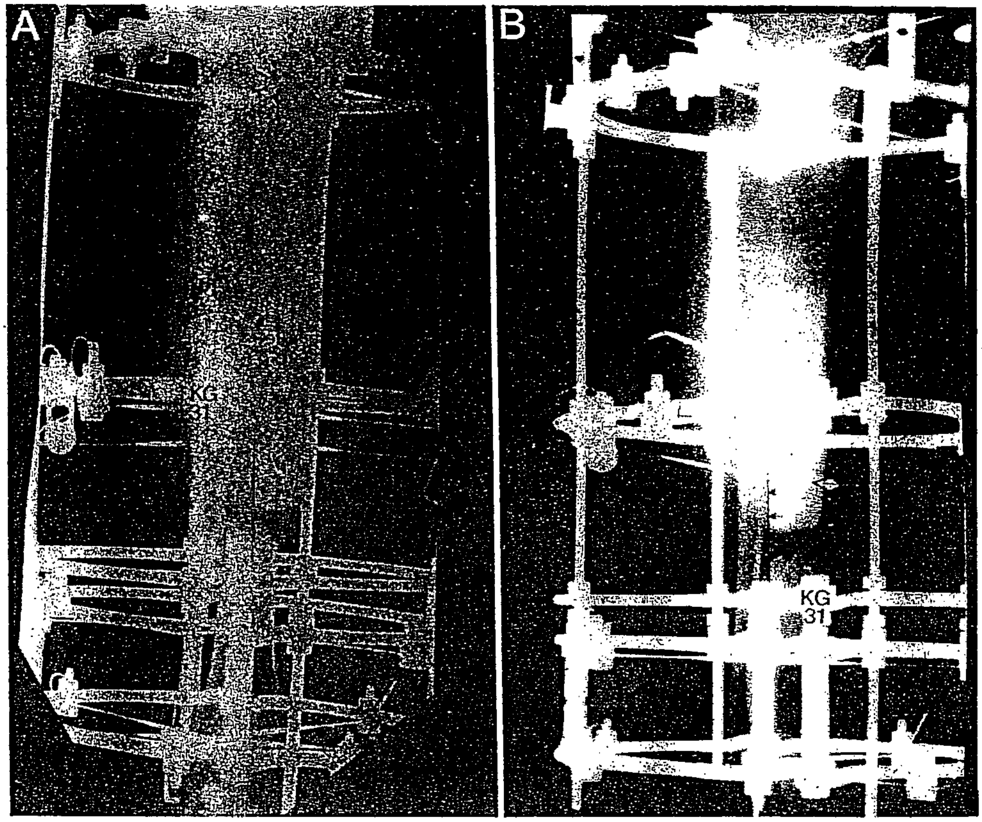


FIGURE 24.
A, docking malalignment during bone transport. B, malalignment corrected using an olive wire.

transport near the time of docking. This usually is due to the dense compressed tissue between the bone ends. This may require a resection with freshening of the bone ends for resolution. (3) Delayed healing of the contact site. These ends usually are atrophic and even may be avascular by the time they reach docking. The ends may need to be freshened and the interposing tissue resected. One end may need to be sharpened and inserted inside the other. A bone graft may be applied. (4) Delayed healing of the regenerate new bone. This may be related to a variety of factors, including a traumatic corticotomy, distraction that is too rapid, a rhythm of distraction that is too low, cyst formation, malnutrition, metabolic disorders, smoking and other substance abuse, chemotherapeutic agents, and other medical disorders. This frequently can be treated by the accordion maneuver, which involves shortening and lenghtening the bone to stimulate new bone formation under compression. In the presence of a cyst, the bone should be shortened down and the cyst aspirated intermittently and then relengthened at half the rate. Alternatively, the site may be bone-grafted.

Soft-tissue problems of bone transport include the following: (1) Obstruction to bone transport. Soft tissues may indent and, therefore, delay transport of the bone segment. The bone segment even may begin to herniate out of the skin. (2) Skin invagination. As the bone transport proceeds, the tissue between the bone ends may invaginate. The bone can be driven right through this tissue and pierce the skin. In order to avoid this problem, it must be recognized and wires inserted to elevate the skin or an open reduction of the bone ends with an acute shortening performed operatively to elevate the soft tissue. A transverse incision should be used. (3) Joint contractures. A proximal-to-distal bone transport will tend to cause a knee contracture to develop due to the pull of the soleus, which then acts upon the gastrocnemius. The transport from distal to proximal may lead to equinovarus contracture in the foot from upward transport of the tibialis posterior muscle (Fig 25). These contracture tendencies must be recognized and treated aggressively with physical therapy. If such

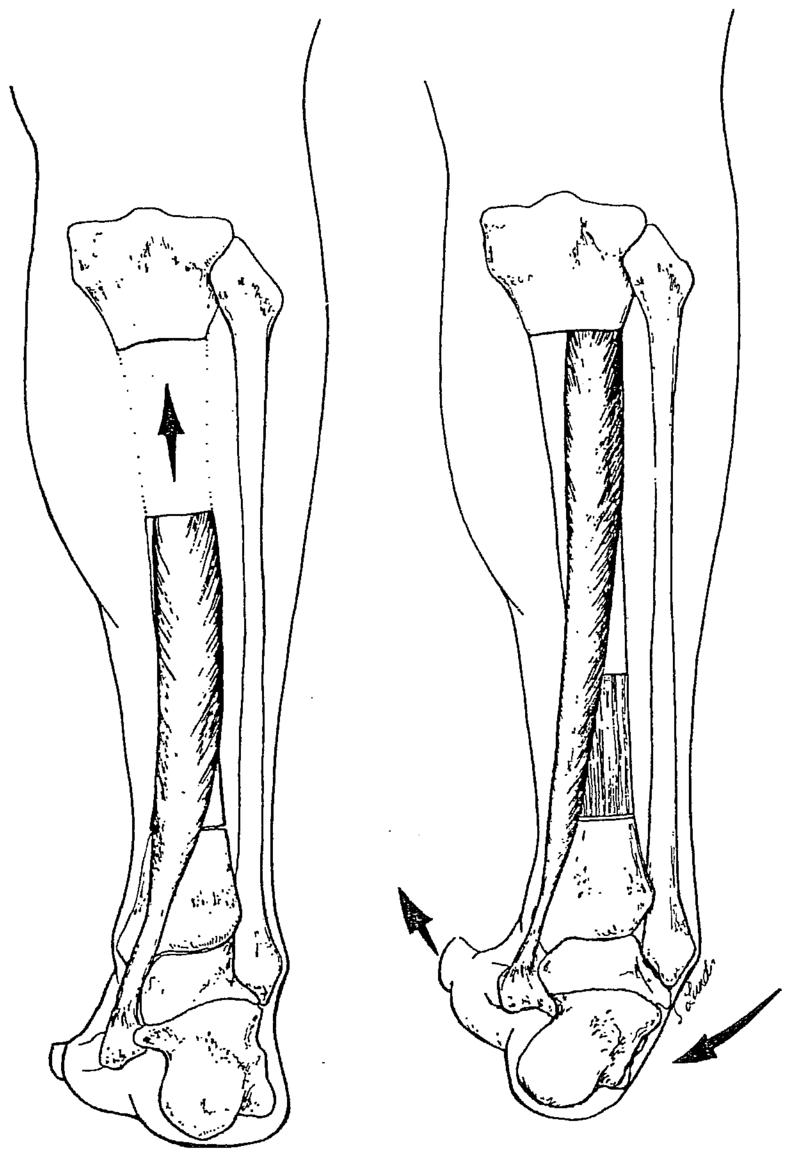


FIGURE 25.
Distal-to-proximal bone transport. Complicated by equinovarus contracture secondary to proximal transport of posterior tibial muscle.

contractures do not respond to therapy, they should be treated by application of the apparatus onto the foot and slow distraction.

Seventeen consecutive tibial bone defects treated by bone transport were evaluated prospectively. The mean age was 37 years (range, 20 to 65 years). The mean bone defect was 8.9 em (range, 2 to 18 em), and the mean limb length discrepancy was 3.6 em. Ten cases initially were infected and 7 still were draining at the time of treatment. The length of treatment time ranged from 4.7 to 22.6 months (mean, 13.9 months). The mean bone regeneration was 9.7 em. Eight patients had soft-tissue defects from debridement. Seven of these defects were closed successfully by soft-tissue transport; one of these required a free flap to cover bone protrusion from transport. All patients were followed-up with clinical and radiographic examinations. The follow-up time since union was an average of 2.1 years. Union was achieved in 16 of the 17 cases. To accelerate union of the docking site, 3 patients had an open decortication of bone ends without bone grafting and 6 had an open decortication with bone grafting. There were no recurrent infections in this series. Leg length discrepancy was less than 2 em in 15 patients. Mechanical axis malalignment greater than 5 degrees remained in 2 cases. The bone results were graded as excellent in 13 cases, good in 3, and poor in 1. The functional results were evaluated as unsatisfactory if the patient had persistent pain or could not return to activities of daily living or work function. One patient had chronic persistent pain due to a preexisting reflex sympathetic dystrophy. This patient requested an elective amputation 2 years after achieving union. There were 15 satisfactory and 2 unsatisfactory functional results. Common problems during treatment included superficial pin infections, distraction pain, and edema. There were no bone infections. Complications included equinovarus contractures (2 patients), soft-tissue invagination between the bone obstructing transport (2 patients), bone protrusion (1 patient), incomplete corticotomy (3 patients), mal-docking (3 patients), and apparatus-related problems (2 patients). Refracture of the docking site occurred in 1 patient and was treated successfully by reapplication of the apparatus. Bending of the regenerate was treated with a cast in 1 patient. Low cross-sectional area of union necessitated prolonged protection in a PTB fracture brace in 2 patients. In total, there were 13 unplanned surgeries to treat complications that arose during transport. There also were 13 second-stage planned procedures such as skin or bone grafts. Only 2 patients required no additional procedures. The average number of surgical procedures was 2.4 per patient. Nonunion of the docking site accounted for 75% of the complications. Docking site healing was the rate-limiting step for treatment time. Patients with reflex sympathetic dystrophy or preexisting severe nerve damage of the posterior tibial nerve should be considered for amputation, even though bony reconstruction is possible with the Ilizarov method.

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