CHAPTER 29

Vincent S. Mosca

The Foot

SURGICAL PROCEDURE INDEX

PRINCIPLES OF ASSESSMENT AND MANAGEMENT

The assessment and management of foot deformities and malformations in children and adolescents are based on principles, not techniques, due to the complexity and variety of the pathologic conditions and the complexity of the foot itself. An orthopaedist managing these conditions must have

1. An appreciation of the age-related physiologic variations in the shape of the foot
2. An understanding of the natural history of each variation and deformity
3. An appreciation of the effect of a chosen intervention on growth and development of the foot as well as the effect of growth and development on a chosen intervention
4. A thorough and working knowledge of the most unique "joint" in the human body, the subtalar joint complex, which is a combination of the talocalcaneal (subtalar) joint, plus the talonavicular and calcaneocuboid joints (transtarsal or Chopart joints)
5. The ability to obtain, and the commitment to evaluate, only weight-bearing or simulated weight-bearing radiographs
6. A dedication to preserving joint motion by utilizing soft-tissue releases and osteotomies instead of arthrodeses
7. A complete understanding of the phrase: "The foot is not a joint" (1)

The first principle to embrace is that "the foot is not a joint" (1), although it is often discussed as if it were another joint in the body, such as the hip, knee, shoulder, or elbow. It is a unique part of the musculoskeletal system comprised of 26 bones with countless articulations. It is extremely unusual for only one portion of the foot or only one joint of the foot or ankle to be congenitally or developmentally deformed. Its many joints are usually deformed or malaligned in rotationally opposite directions, "as if the foot was wrung out like a towel" (1). As examples, note that there is inversion of the subtalar joint and pronation of the forefoot on the hindfoot in a cavus foot and eversion of the subtalar joint and supination of the forefoot on the hindfoot in a flatfoot. And one cannot ignore the adjacent ankle joint as a potential site of additional deformity. The orthopaedist must identify all deformities preoperatively, if possible, and have a treatment plan that addresses each one individually and, usually, concurrently. There is no justification for creating a compensating deformity or incompletely correcting a deformity in order to avoid an additional procedure, particularly one that can usually be carried out during the same operative session.

The child's foot often looks different than that of an adult. In fact, there is so much variation in shape that the foot of one child can look quite different than that of another child. Age-related physiologic variations of the child's foot, such as flexible metatarsus adductus, positional calcaneovalgus, and flexible flatfoot, must be identified as normal, but not average, shapes in order to avoid inappropriate and potentially harmful interventions. This feature of physiologic variation is also seen in the long bones of the child's lower extremities in conditions such as genu varum, genu valgum, femoral anteversion, and tibial torsion (2, 3). There are age-related average shapes and normal ranges of shapes. The natural history is for spontaneous change

from the normal shapes of the child to those of the adult through normal growth and development. Externally applied forces cannot modify these physiologic shape variations of the long bones. And the long-term health consequences of persistent physiologic variations of the long bones have yet to be proven.

An understanding of the natural history of each foot shape variation and deformity is of paramount importance. Eighty-five to ninety-five percent of feet with metatarsus adductus correct spontaneously with little if any long-term disability even with mild to moderate residual deformity (4–6). Essentially all calcaneovalgus "deformities" correct spontaneously (7). Flexible flatfoot is almost ubiquitous at birth and is present in approximately 23% of adults, most of whom are asymptomatic (8). The height of the longitudinal arch increases spontaneously during the first decade of life in most children (9, 10). There is a wide range of normal arch heights at all ages (particularly in young children) (9, 10). Most feet with accessory naviculars (11, 12) and approximately 75% of feet with tarsal coalitions (13) are asymptomatic and do not need treatment, whereas one can expect the onset of symptoms from the rest to develop in late childhood or early adolescence. Conversely, all congenital clubfoot and congenital vertical talus deformities persist and cause disability unless treated.

The natural history of an intervention must also be fully appreciated and considered in relation to the natural history of the deformity or condition. Unfortunately, although there are few good natural history studies on deformities and variations of the child's foot, there are fewer good long-term follow-up studies on operative intervention for these conditions. It seems most reasonable that the default should logically go to the natural history of the condition.

Unique challenges facing those who manage foot deformities in children are the consideration of the effect of a chosen intervention on growth and development of the foot as well as the effect of growth and development on a chosen intervention. Early reconstruction of foot deformities in children normalizes the stresses on the bones and joints to allow more normal development. Delay results in the development or persistence of abnormalities in the shapes of the bones and joints that makes reconstruction more difficult. Furthermore, procedures that affect or potentially affect growth in a positive or in an adverse way must be used judiciously. Conversely, one must consider how the early positive result of an intervention may change as the child grows. Cavus foot deformity is most commonly a manifestation of muscle imbalance from an underlying neuromuscular disorder. In some cases, the disorder is static (cerebral palsy) or can be stabilized but may recur (tethered cord in myelomeningocele). In others, the disorder is progressive and the rate and extent of neuromuscular deterioration may not be predictable (Charcot-Marie-Tooth [CMT]). It is difficult to establish precise muscle balance in any cavus foot, and it is well known that growth as well as progressive neurologic deterioration can undo an excellent early result of intervention. The child and family must be made aware that there are no panaceas and more surgery may be needed in the future. The surgeon must also remember this admonition, avoid burning bridges, and keep reasonable options available for future surgeries.

Although most congenital clubfeet and many congenital vertical talus deformities respond to nonsurgical or minimally invasive management, some undergo operative releases in the first year of life when the foot is 8 to 9 cm in length. The hope is that the correction of these deformities, located at the foundation of the human body, will be maintained through 14 to 16 years of growth and a doubling to tripling in the length of the foot. Problems, including recurrence, overcorrection, pain, and stiffness, as well as plans for their management, should be anticipated.

There is no other "joint" in the human body with the unique anatomy and three-dimensional motion of the subtalar joint complex. This complex consists of two components, the talocalcaneal or subtalar joint, plus the talonavicular and calcaneocuboid or transtarsal joints. These four bones, several important ligaments, and multiple joint capsules function together as a unit. Terms that apply to sagittal and coronal plane alignment and motions, such as varus, valgus, abduction, adduction, flexion, extension, supination, and pronation, do not necessarily apply to the subtalar complex because its axis of motion is in neither the sagittal nor coronal plane. Inversion and eversion are terms that, in my opinion, define the motions of this complex, but they need to be better defined and understood by all that use them.

Almost 200 years ago, Scarpa (14) saw similarities between the subtalar joint complex and the hip joint. He compared the femoral head to the talar head and the pelvic acetabulum to his so-called acetabulum pedis (AP). The latter is a cup-like structure made up of the navicular, the spring ligament, and the anterior end of the calcaneus and its facets. Although it is not a perfect comparison, I believe that the two anatomic areas share certain features that make the comparison both valid and worthwhile. The hip, a pure ball-and-socket joint with a central rotation point, is comprised of two bones, one intra-articular ligament, and a joint capsule. The subtalar joint is not an independent ball-and-socket joint, though the combined motions of the subtalar joint and the immediately adjacent ankle joint give the impression of a ball-and-socket joint. In fact, the subtalar joint has an axis of motion that is in an oblique plane that is not frontal, sagittal, or coronal, thus creating motions that are best described with the unique terms inversion and eversion. The stable structure in the hip joint is the acetabulum (the socket), while the stable structure in the subtalar joint complex is the talus (the ball). Inversion is comprised of plantar flexion, supination, and internal rotation of the AP around the head of the talus (15). Eversion is a combination of dorsiflexion, pronation, and external rotation of the AP around the talar head. The static position of inversion of the subtalar joint is called hindfoot varus and is found in cavovarus feet and clubfeet. Hindfoot valgus is the static position of the everted subtalar joint and is seen in flatfeet and skewfeet. It is essential that all who manage foot deformities have a thorough and working knowledge of this most unique joint complex.

It is important to evaluate deformity both clinically and radiographically with the foot in the weight-bearing position. That is the baseline against which the corrected foot will be judged. A flexible flatfoot appears to have an arch, and a normal foot may appear to have a cavus or clubfoot deformity when dangling in the air.

Deformities of the child's foot should be corrected by means of soft-tissue releases to align the joints and osteotomies to correct residual deformities. Arthrodesis should be reserved for the older child, adolescent, or adult with established degenerative arthrosis of a joint or with such severe deformity that correction cannot be achieved with soft-tissue releases and osteotomies. Long-term follow-up studies have demonstrated that arthrodesis of even the small joints of the child's foot should be avoided because of the risk of developing degenerative arthrosis at the adjacent unfused joints (16–18). Arthrodesis of the subtalar joint, particularly triple arthrodesis, leads to stress transfer to the ankle (19–27). The development of degenerative arthrosis at that important joint is a potentially disastrous outcome.

Correction of foot deformities must be combined with balancing of muscle forces in order to help prevent recurrence. Balancing muscle forces in a mobile foot is much more challenging than in one that has undergone arthrodesis. This challenge must be accepted.

There is a great need for more natural history studies on deformities and variations in the shape of the child's foot, as well as long-term follow-up studies on the interventions used to treat these conditions. The message must be to exhibit caution with interventions until it is clear that the treatment is not potentially worse than the condition.

All of these principles apply to the congenital and developmental deformities and other conditions that will now be presented individually and alphabetically, not in order of importance, incidence, or complexity.

CONGENITAL DEFORMITIES AND MALFORMATIONS OF THE FOOT

Accessory Navicular

Definition. Accessory navicular is a term applied to a plantar-medial enlargement of the tarsal navicular beyond its normal size. It may consist of a separate ossicle connected to the main body of the navicular by fibrous tissue or cartilage, or it may be a solid bony enlargement of the bone. Other terms that have been used to describe the separate ossicle are the os tibiale externum, the navicular secundum, or the prehallux (Fig. 29-1).

Epidemiology. The accessory tarsal navicular is the most common accessory bone in the foot, occurring in between 4% and 14% of the population (8, 11, 28). It is frequently bilateral and occurs more commonly in females. Geist (28) recognized a higher incidence of accessory naviculars in young patients evaluated radiologically than in cadaver studies.

Etiology. McKusick (29) believed that the accessory navicular was inherited as an autosomal dominant trait. Geist (28) reported that there are three types: (a) a sesamoid bone within the substance of the posterior tibial tendon, (b) a separate bone with a true articulation (synovial joint) with the navicular, and (c) an ossicle with a synchondrosis to the main navicular.

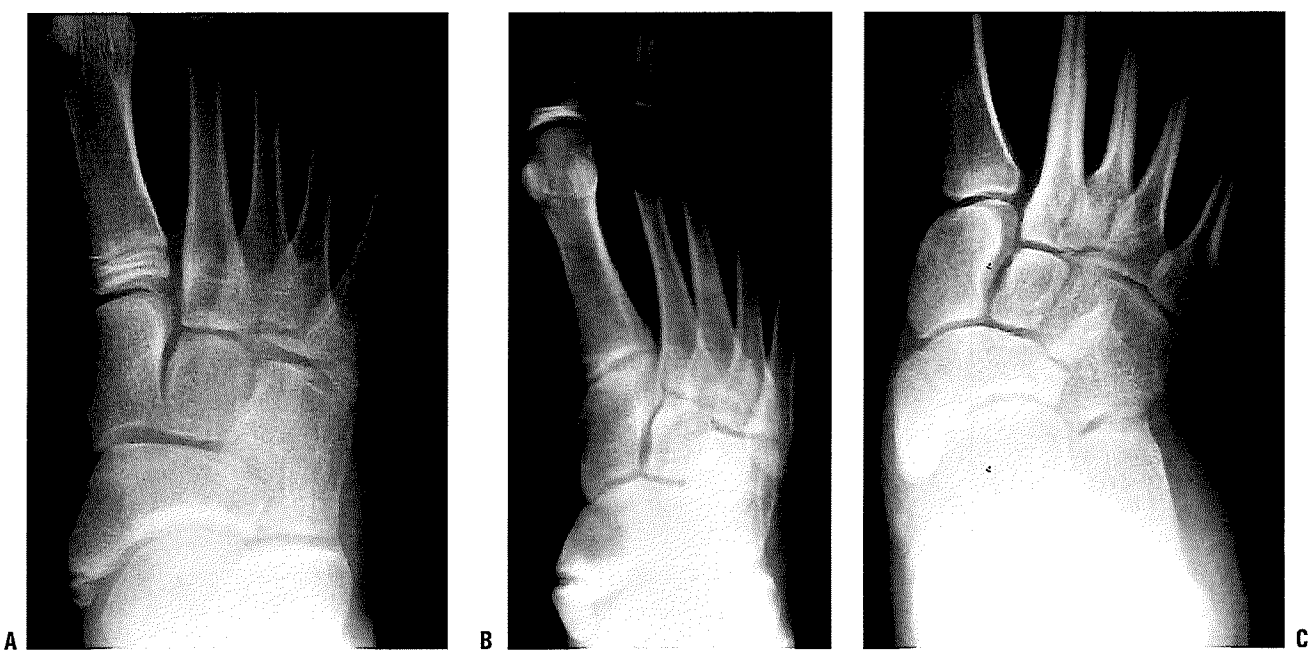


FIGURE 29-1. Accessory navicular. **A:** Type I. **B:** Type II. **C:** Type III. (From the private collection of Vincent S. Mosca, MD.)

Clinical Features. Pain, tenderness, and callus formation may develop over the firm prominence distal to the talar head on the plantar-medial aspect of the midfoot starting in adolescence. There may be a coexistent flexible flatfoot (30–33), but there is not conclusive evidence that there is a cause-and-effect relationship between the two conditions as was historically believed (12). The prominence of an accessory navicular is in close proximity to the head of the talus, which is prominent in a flexible flatfoot. Inverting and everting the subtalar joint with one's thumb on the prominence is helpful for differentiating the two. If the prominence moves, it is an accessory navicular. If it remains stationary, it is the head of the talus.

Individuals with an accessory navicular may present for evaluation because of the prominence, but more commonly they present because of pain at the site. The typical patient is an active adolescent girl with a history of minor trauma who presents with pain, callus formation, tenderness, redness, and, occasionally, swelling over the bony prominence. Maximum tenderness is elicited by upward pressure under the prominence. Because of the frequency of this anatomic variation in the general population, one must be careful not to assume that a radiographic finding of an accessory navicular is the cause of the foot pain without thorough evaluation (34).

Radiographic Features. An accessory navicular can usually be seen on standing AP and lateral radiographs, but a lateral oblique view (opposite to the standard medial oblique view that is generally obtained) may be necessary for identification. There are three types of accessory naviculars (35) (Fig. 29-1). Type I is a rarely symptomatic, small pea-sized sesamoid bone located in the center of the most distal portion of the tibialis posterior tendon. Type II, the most frequently symptomatic type, is a bullet-shaped ossicle joined to the tuberosity of the navicular by a syndesmosis or synchondrosis. Type III is a large, horn-shaped navicular that probably results from fusion of a type II with the body of the navicular over time.

Pathoanatomy. There is proliferating vascular mesenchymal tissue, cartilage proliferation, and osteoblastic and osteoclastic activity in the tissue between the ossicle and the main body of the navicular in painful type II accessory naviculars (35). These histologic findings are consistent with healing microfractures, substantiating the opinion that pain at this site is related to chronic, repetitive stress. There are at least two other possible sources of pain. One is pain from pressure on the skin overlying the bony prominence. The other is tendinitis in the tibialis posterior, the tendon in which the ossicle resides. Any or all of these sources may exist in the same painful foot.

Natural History. Accessory naviculars are for the most part asymptomatic. If symptoms do occur, a period of protection from stress and injury generally returns the patient to the asymptomatic state (32).

Treatment. Nonsurgical treatment of a painful accessory navicular often relieves symptoms. Pain is due to inflammation of skin, tendon, and/or cartilage. Strenuous, pain-exacerbating activities should be curtailed. Shoes should be examined to assess the site of contact with the bony prominence. Shoes can be stretched. Alternatively, shoes that provide softness or no contact in that area should be purchased. An over-the-counter orthosis can be used to elevate the arch and change the site of contact between the prominence and the shoe. Orthoses may also decrease the stress on the tibialis posterior tendon and relieve the tendinitis. If pain is acute and persistent, the foot can be immobilized in a below-knee cast for 4 to 6 weeks.

Surgery is indicated if prolonged attempts at conservative management fail to relieve symptoms. The Kidner procedure combines removal of the ossicle with advancement of the tibialis posterior tendon (30, 31). This was based on Kidner's belief that the height of the longitudinal arch was related to its muscular support, a theory that has since been disproved (36, 37). Several studies have shown good to excellent results in 90% or more of cases by simple excision of the ossicle and shaving of the medial enlargement of the main body of the navicular through a tendon splitting approach (12, 28, 37–41). The incision should be placed slightly dorsomedially to avoid creating a painful scar.

Cavus Foot Deformity

Definition. Cavus refers to a fixed equinus (plantar-flexion) deformity of the forefoot in relation to the hindfoot resulting in an abnormally high arch. The high arch may be along the medial border of the foot or across the entire midfoot. The heel may be in a neutral, varus, valgus, calcaneus (dorsiflexed), or equinus position. There may be an accompanying clawing of the toes. There are two common patterns of cavus deformity (42). Cavovarus, or anterior cavus, is the most common pattern. It consists of pronation of the forefoot on the hindfoot and inversion (or varus) deformity of the hindfoot, rotationally opposite direction deformities that give the impression that the foot has been "wrung out like a towel" (1). Calcaneocavus, or posterior cavus, occurs less frequently. It is also known as transtarsal cavus, because the entire arch is elevated from the ground from medial to lateral.

Etiology. Cavus is a manifestation of a neuromuscular disorder with muscle imbalance, until proven otherwise. At least two-thirds of patients who seek treatment for a painful high arch will have an underlying neurologic problem, and over half of these will have Charcot-Marie-Tooth (CMT) disease (43, 44). There are many other causes of cavus foot deformity. It is helpful to consider those that cause unilateral versus bilateral deformity when developing a differential diagnosis for your patient (Table 29-1). The number of cases termed "idiopathic" cavus foot continues to decrease as diagnostic methods improve.

Calcaneocavus deformity is seen almost exclusively in children with myelomeningocele and poliomyelitis due to a specific pattern of muscle imbalance seen in many of these children.

Epidemiology. The incidence of cavus is variable and is related to the prevalence of neuromuscular disorders at any point in time.

TABLE 29-1 Causes of Cavus Foot Deformity

Bilateral
Charcot-Marie-Tooth disease
Friedreich ataxia
Dejerine-Sottas interstitial hypertrophic neuritis
Polyneuritis
Roussy-Lévy syndrome
Spinal muscular atrophy
Myelomeningocele
Syringomyelia
Spinal cord tumor
Diastematomyelia
Spinal dysraphism (tethered cord)
Muscular dystrophy
Cerebral palsy—paraparesis or quadriparesis (although usually planus deformities)
Familial (consider Charcot-Marie-Tooth disease)
Clubfoot/recurrent clubfoot
Idiopathic cause (diagnosis of exclusion)

Unilateral
Traumatic injury of a peripheral nerve or spinal root nerve
Poliomyelitis
Syringomyelia
Lipomeningocele
Spinal cord tumor
Diastematomyelia
Spinal dysraphism (tethered cord)
Tendon laceration
Overlengthened Achilles tendon
Cerebral palsy—hemiparesis
Clubfoot/recurrent clubfoot
Compartment syndrome of the leg
Severe burn of the leg
Crush injury of the leg

Clinical Features. The clinical features depend on the underlying etiology. Some of the underlying neurologic abnormalities are known at the time of presentation and some are not, some are treatable and some are not, and some are static and some are progressive. The resultant muscle imbalance, however, always leads to progressive foot deformity. The clinical manifestations are instability of gait with frequent falling, a feeling that the ankle is "giving out," and a history of repeated ankle sprains. Instability may be secondary to muscle weakness, sensory loss, or deformity (45). Painful callosities develop under the metatarsal heads, lateral to the base of the fifth metatarsal, and over the dorsum of the proximal interphalangeal (PIP) joints of the progressively clawing toes. In the calcaneocavus foot, callus formation occurs primarily under the calcaneus; however, exaggerated callus formation under all metatarsal heads can occasionally be seen.

A history of progressive change in foot shape and function must be ascertained, even in situations of a known underlying neurologic abnormality. A sudden increase in cavus deformity in a child with myelomeningocele or lipomeningocele could represent evidence of a tethered spinal cord.

Medical, birth, and developmental histories, as well as a review of systems, are mandatory in searching for underlying causes of cavus foot. A family history of cavus foot deformity should be investigated as an aid to diagnosis. Charcot-Marie-Tooth disease, an autosomal dominant disorder with variable phenotypic expression, is one of the most common causes of cavus foot deformity (43). Examination of other family members can often be revealing, because a subtle cavus foot may be present.

A detailed neurologic examination, including motor, sensory, and reflex testing, of the upper and lower extremities is mandatory. The spine must be examined for deformity, midline defects, hairy patches, dimples, or other evidence of spinal dysraphism. The Trendelenburg test should be performed.

The foot is examined with the child seated, standing, and walking. Cavovarus is the most common specific form of cavus foot deformity and can be caused by many different underlying neurologic disorders. The longitudinal arch is excessively elevated along the medial border of a weight-bearing foot (Fig. 29-2). The lateral border of the foot is convex and plantigrade, and the base of the fifth metatarsal is prominent and often callused. The hindfoot is in varus alignment. Atrophy of the intrinsic muscles is apparent. There may or may not be contracture of the gastrocnemius or the triceps surae. In many cases, as in most cavovarus deformities secondary to CMT, the forefoot equinus gives the false impression of a hindfoot equinus (46). The thigh–foot angle is usually neutral, despite the internal rotation deformity in the subtalar joint. The reason is that external tibial torsion is always associated with a developmental cavovarus deformity. The two rotational deformities are in opposite directions and cancel each other out. It is important to discuss this with the patient and family, because the external tibial torsion will become obvious after correction of the foot deformity.

Despite differences in the patterns of muscle imbalance that create cavovarus, the pattern of deformity development is fairly constant (47–49). The first metatarsal becomes plantar-flexed, giving the appearance of pronation of the forefoot on the hindfoot. The deformity is flexible at first but becomes rigid with time. The plantar fascia and the other

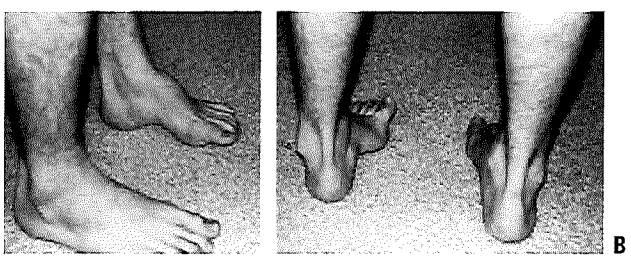


FIGURE 29-2. Cavovarus deformity in this individual with Charcot-Marie-Tooth disease. **A:** The arch is elevated only along the medial border of the foot. **B:** Varus and adduction can be appreciated. (From the private collection of Vincent S. Mosca, MD.)

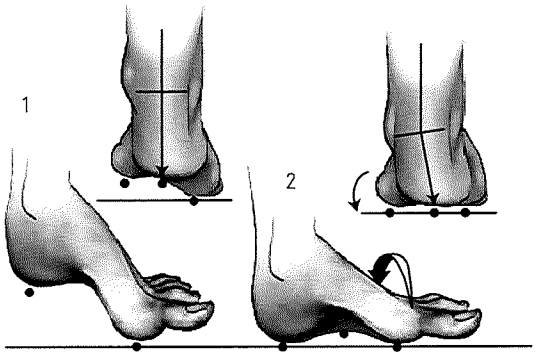


FIGURE 29-3. The tripod effect. The hindfoot must assume a varus position when weight bearing if the first metatarsal is fixed in plantar flexion (9). Initial contact of plantar-flexed first metatarsal (4). Fifth metatarsal makes contact through supination of the forefoot (*arrow*), which also drives the hindfoot into varus. (From Paulos L, Coleman SS, Samuelson KM. Pes cavovarus. Review of a surgical approach using selective soft-tissue procedures. *J Bone Joint Surg Am* 1980;62: 942–953, with permission.)

plantar soft tissues become contracted. The incompletely ossified bones change shape due to excessive compression on their plantar aspects (Hueter-Volkmann law). The normal tripod structure of the foot becomes unbalanced. Bearing weight on the plantar-flexed first metatarsal causes the forefoot to supinate in relation to the tibia, thereby allowing the fifth metatarsal head to touch the ground. Because the forefoot is rigidly pronated in relation to the hindfoot, the subtalar joint is thereby driven into inversion, or varus (48) (Fig. 29-3). This flexible hindfoot varus deformity eventually becomes rigid as the plantar-medial soft tissues of the subtalar joint contract. The cavovarus foot, therefore, has two major rotational deformities in opposite directions from each other: pronation of the forefoot and supination (varus and inversion are other descriptive terms) of the hindfoot. It appears as if the foot is wrung out (1).

Determination of the flexibility or rigidity of each deformity is important when planning an operation. Flexible deformities are treated with tendon transfers, and inflexible deformities are treated with soft-tissue releases, osteotomies, and, occasionally, arthrodeses. Coleman and Chestnut (50) devised the block test to help evaluate the flexibility of the hindfoot (Fig. 29-4). The patient stands with a block of wood under the lateral border of the foot to recreate the tripod while allowing the first metatarsal to plantar-flex. A flexible varus deformity of the hindfoot will correct to valgus alignment. One that is already contracted and rigid will not. In the first situation, surgery for deformity correction is confined to the forefoot. In the latter case, forefoot and hindfoot procedures are needed.

The arch is elevated across the entire midfoot in the calcaneocavus deformity (Fig. 29-5). The calcaneus is dorsiflexed and vertically aligned, giving it the appearance of posterior truncation. The plantar heel pad is thickly callused from excessive pressure over a small surface area.

Radiologic Features. Standing AP and lateral radiographs of the foot are indicated on initial evaluation. According to Meary (51), there is normally a straight-line relationship between the axis of the talus and that of the first metatarsal on the lateral view. In essentially all cavovarus foot deformities, those two lines intersect within the body of the medial cuneiform, indicating that site as the center of rotation of angulation (CORA) (52), with the apex of the angulation directed dorsally (Fig. 29-6A). A calcaneal pitch (CP) >30 degrees is also indicative of a cavus deformity. The hyperdorsiflexion of the calcaneus seen on the radiographs confirms that the apparent clinical equinus deformity is, in fact, forefoot equinus, that is, cavus (Fig. 29-6). Inversion or varus deformity of the hindfoot is indicated on the AP radiograph by parallelism between the talus and calcaneus and by adduction at the talonavicular joint. It is also manifest as the intersection of the axis of the talus and that of the first metatarsal (the CORA) (52) at the talonavicular joint or within the head of the talus (Fig. 29-7A).

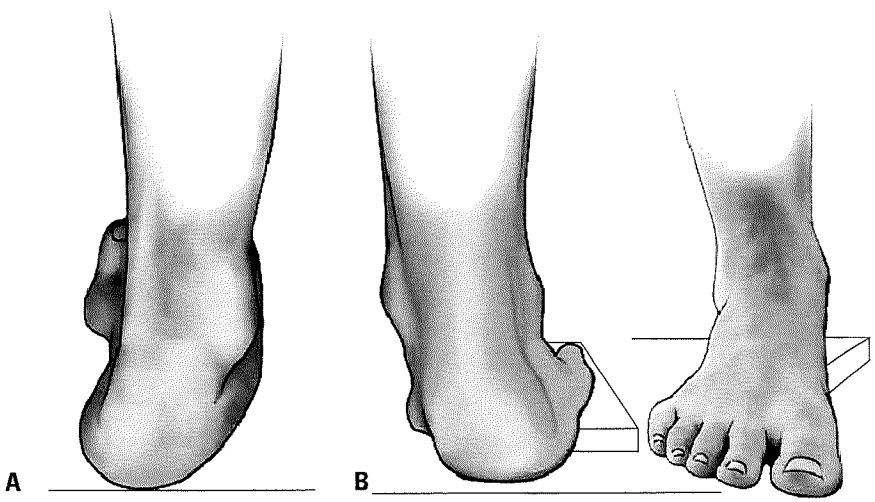


FIGURE 29-4. The Coleman block test for determination of hindfoot flexibility. The flexible varus deformity of the hindfoot **(A)** corrects to valgus **(B)** when the plantar-flexed first metatarsal is allowed to drop down off the edge of the block of wood as in this example. Failure to correct to valgus indicates the need for surgical correction of the hindfoot deformity, in addition to the procedures on the forefoot. (From Coleman SS, Chesnut WJ. A simple test for hindfoot flexibility in the cavovarus foot. *Clin Orthop* 1977;123:60–62, with permission.)

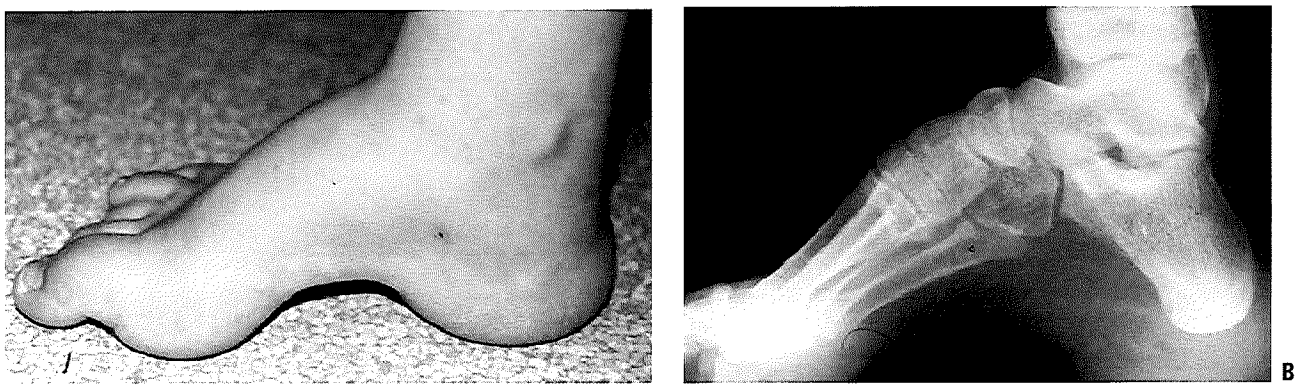


FIGURE 29-5. Calcaneocavus deformity in a child with myelomeningocele. **A:** Transtarsal cavus with thick callosities under the calcaneus and the metatarsal heads. **B:** Radiograph of the calcaneocavus "pistol-grip" deformity. (From the private collection of Vincent S. Mosca, MD.)

I obtain a standing AP Coleman-type block test radiograph to document the flexibility of the hindfoot. I find that the clinical and radiographic block tests are more reliable if a Plexiglas block is placed under the lateral metatarsal heads alone with the heel on the X-ray plate/platform (Fig. 29-7).

An AP pelvis radiograph is indicated because of the known association of Charcot-Marie-Tooth disease with progressive hip subluxation and dysplasia. Standing AP and lateral radiographs of the spine are indicated if any physical examination findings suggest spinal dysraphism, spine tumor, or diastematomyelia.

Other Diagnostic Studies. Consultation by a pediatric neurologist is in the best interest of the child. Although most cavus foot deformities are caused by CMT, a progressive genetically determined neuromuscular disorder for which there is no known treatment, it is vital to identify and correct a treatable neurologic disorder before treating the foot. An MRI of the spine along

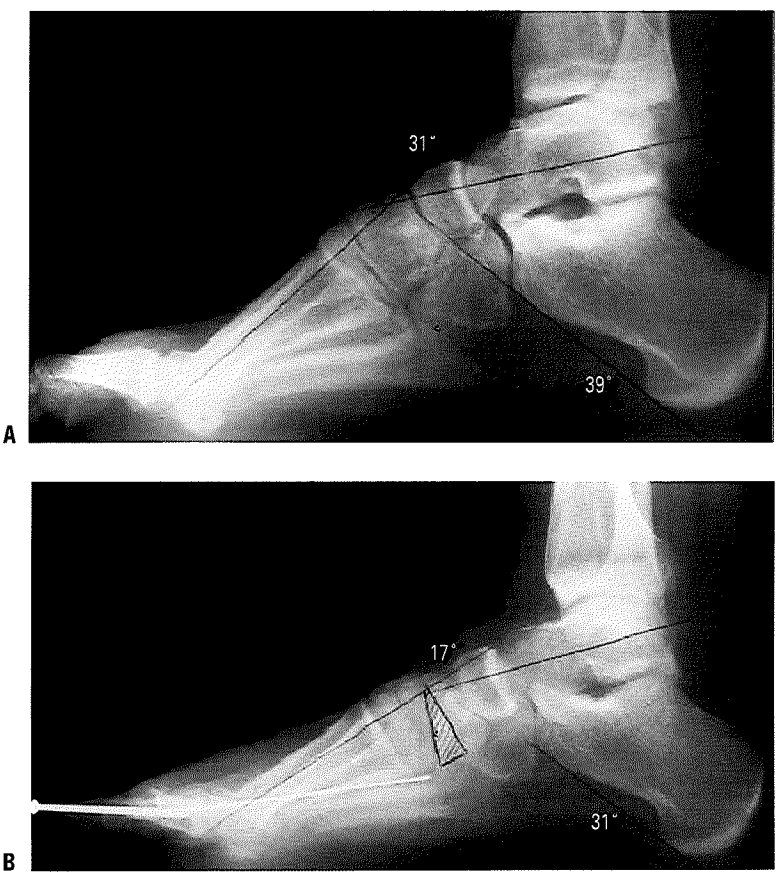


FIGURE 29-6. Lateral radiograph of a cavovarus foot deformity before **(A)** and after **(B)** a medial cuneiform plantar-based opening-wedge osteotomy. The axis lines of the first metatarsal and the talus cross each other in the body of the medial cuneiform, indicating that as the site of deformity, that is, the CORA. (From VS. Ankle and foot: pediatric aspects. In: Beaty J, ed. *Orthopaedic Knowledge Update 6.* Rosemont, IL: American Academy of Orthopaedic Surgeons, 1999:583, with permission.)

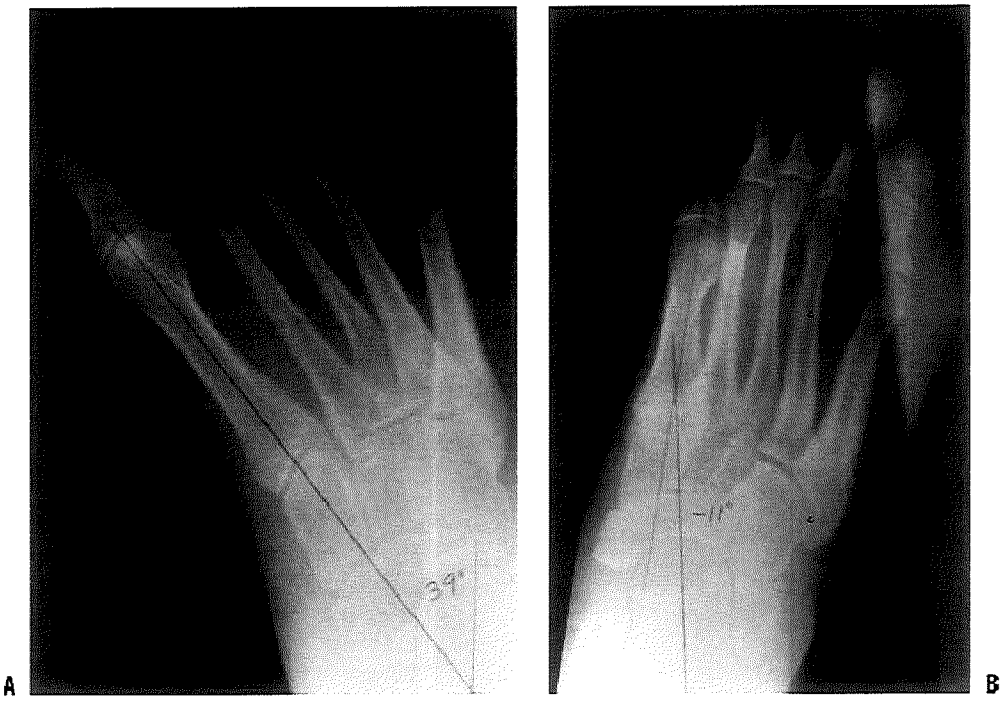


FIGURE 29-7. Standing block test radiograph with Plexiglas under the lateral metatarsal heads. The flexibility or rigidity of the subtalar joint can be documented by assessing alignment at the talonavicular joint using the talus–first metatarsal angle. **A:** Without block. **B:** With block, the hindfoot varus is corrected as indicated by abduction of the 1st metatarsal axis in relation to the axis of the talus. (From the private collection of Vincent S. Mosca, MD.)

with DNA blood tests for CMT, electromyogram with nerve conduction studies, and muscle biopsy may be indicated.

Pathoanatomy. Several patterns of muscle imbalance can create the cavovarus deformity. A common pattern is that seen in CMT (47, 49). Denervation begins in the intrinsic muscles of the foot (49). The weakened lumbricals allow the long toe extensors to extend the metatarsophalangeal joints and the long toe flexors to flex the interphalangeal joints, thereby creating claw toe deformities. These same forces create elevation of the longitudinal arch during gait by the windlass effect of the plantar fascia (53) (Fig. 29-8). The intrinsic muscles undergo atrophy, fibrosis, and shortening that lead to secondary contracture of the plantar fascia. This creates a bowstring between the anterior and posterior pillars of the arch that draws them closer and produces equinus of the forefoot on the hindfoot. The tibialis anterior, a dorsiflexor of the first metatarsal, becomes weak, while the peroneus longus, a plantar flexor of the first metatarsal, remains relatively strong (42). The extensor hallucis longus is involuntarily recruited in an attempt to provide additional dorsiflexion strength along the medial column of the foot, but it creates a paradoxical effect of plantar flexion due to the windlass effect of the plantar fascia. The first metatarsal starts to plantar-flex, and, with time, this creates more contracture and shortening along the plantar-medial than the plantar-lateral border

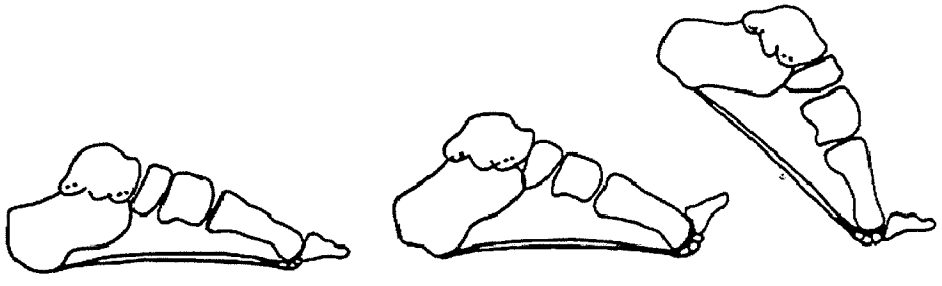


FIGURE 29-8. The windlass effect of the plantar fascia. The drum, or pulley, of the windlass is the head of the metatarsal. The handle is the proximal phalanx. The cable that is wound under the drum, through its attachment to the plantar pad of the metatarsophalangeal joint, is the plantar fascia. Dorsiflexion of the toes creates elevation of the longitudinal arch. (From Hicks JH. The mechanics of the foot. II. The plantar aponeurosis and the arch. *J Anat* 1954;88:25–30, with permission.)

of the foot. The forefoot becomes rigidly pronated in relation to the hindfoot. The tripod effect (48) accounts for the varus position that the hindfoot must assume during weight bearing due to the fixed pronation of the forefoot. Also contributing to the varus deformity of the hindfoot is the muscle imbalance between the tibialis posterior, an invertor of the subtalar joint, that remains strong and the peroneus brevis, an evertor of the subtalar joint, that becomes weak (47). The subtalar joint eventually becomes rigidly deformed in varus because of contracture of the plantar-medial soft tissues, including those of the subtalar joint complex. Although the triceps surae does not become contracted in CMT, it does in some of the other diseases that cause cavus.

The calcaneocavus deformity develops when there is little or no strength in the triceps surae, but strength exists in the muscles that plantar-flex the forefoot and toes. The tibialis posterior, peroneus brevis and longus, flexor hallucis longus, and flexor digitorum longus (FDL) bypass the calcaneus and plantar-flex the entire forefoot on the hindfoot without creating varus. Contracture of the plantar fascia, elongation of the paralyzed triceps surae, and preservation of functional strength in the tibialis anterior contribute to the dorsiflexion posture of the calcaneus.

Natural History. Muscle imbalance from both static and progressive neuromuscular disorders leads to progressive increase in the severity and stiffness of cavus foot deformities, though the rate of progression varies considerably.

Treatment. Treatment of the underlying neurologic disorder should precede treatment of the foot deformity. In most cases, cavus deformity is the result of the problem (a neurologic disorder), not the primary problem itself. Unfortunately, there is no known treatment or cure for many of the neurologic conditions that create cavus deformity.

Established muscle weakness and imbalance are not reversible, even with successful arrest of the neurologic deterioration. The foot deformity, therefore, progresses. There is little role for nonoperative management of the cavus deformity because most are progressive and of an advanced degree of severity at the time of diagnosis. The complexity of reconstruction increases with the severity and rigidity of the deformities (48, 54, 55). Inexpensive, accommodative arch supports and shoe modifications may be used to ameliorate symptoms during the time it takes to diagnose and, if possible, to treat the underlying etiology.

Operative indications include evidence of a progressive deformity, painful callosities under the metatarsal heads or base of the fifth metatarsal, and hindfoot/ankle instability. The main principles of surgical management of a cavus foot are to (a) correct all of the segmental deformities and (b) balance the muscle forces. Correcting the deformities without balancing the muscle forces (through tendon transfers) will lead to recurrence of the deformities. Conversely, balancing the muscle forces (through tendon transfers) without correcting the structural deformities will create well-balanced deformities. Neither of these outcomes is desired. Correction of deformities lends itself more readily to an algorithm than does muscle balancing. And balancing muscles in a mobile foot is more challenging than in one that has undergone subtalar or triple arthrodesis. The challenge must be accepted. The third principle of surgical management is to leave reasonable treatment options available for the possible recurrence of deformity and pain. Because the foot deformity is the result of the (neurologic) problem and not the primary problem, recurrence of deformity is likely. This leads to an important principle of patient management. Educate the patient and family that there are no treatment panaceas and that more surgery may be required in the future (19–21, 42, 48, 56–58).

There is a very long list of operative procedures that can be employed to correct the individual deformities and balance the muscle forces in the cavus foot (42, 48, 56–74). No single procedure can accomplish both goals. Although some authors have tried (48, 55, 56, 74–76), no one has yet developed a reliable algorithm to help one choose the combination of procedures that will best address the unique pathology in any particular case that is based on the severity and rigidity of the deformities and the underlying etiology.

The cavus foot deformity must be classified based on the flexibility of the segmental deformities. Correction of deformities begins with soft-tissue releases, such as fasciotomies, capsulotomies, and muscle or tendon lengthenings, with the goal of realigning joints (48, 55, 58, 60). For the cavovarus foot, this means a proximal release of the abductor hallucis, a tibialis posterior tendon lengthening and possibly a talonavicular joint capsulotomy (48), and a plantar fasciotomy (60) (Figs. 29-9 to 29-12).

Although percutaneous division of the plantar fascia has often been recommended, this alone is not sufficient to correct the deformity in most feet with significant cavus deformity. In most feet it is necessary to release not only the plantar fascia but also the origin of the intrinsic muscles of the foot and the abductor hallux from the calcaneus. An osteotomy of the medial cuneiform can be accomplished through a distal extension of the incision.

Some of the bones in the child's foot will grow abnormally when unbalanced muscle forces are placed on them for a prolonged period of time (48). Osteotomies are employed to correct these bone deformities that can only be identified following soft-tissue releases and realignment of the joints. The first ray becomes plantar-flexed early in the course of development of the cavovarus foot deformity. The site of that cavus deformity, the so-called CORA (52), is within the body of the medial cuneiform (see *Radiologic Features*) (Fig. 29-6A). Treatment with a plantar-based opening-wedge osteotomy of the medial cuneiform is safe, effective, reliable, and inherently stable without the need for internal fixation (48, 55, 74–77) (Fig. 29-6B). An osteotomy of the first metatarsal (70, 74) is less desirable because it puts the growth plate at risk for arrest, requires internal fixation, puts the second metatarsal head at risk for a stress transfer lesion,

Text continued on page 1399

Plantar-Medial Release for Cavovarus Foot (Figs. 29-9 to 29-12)

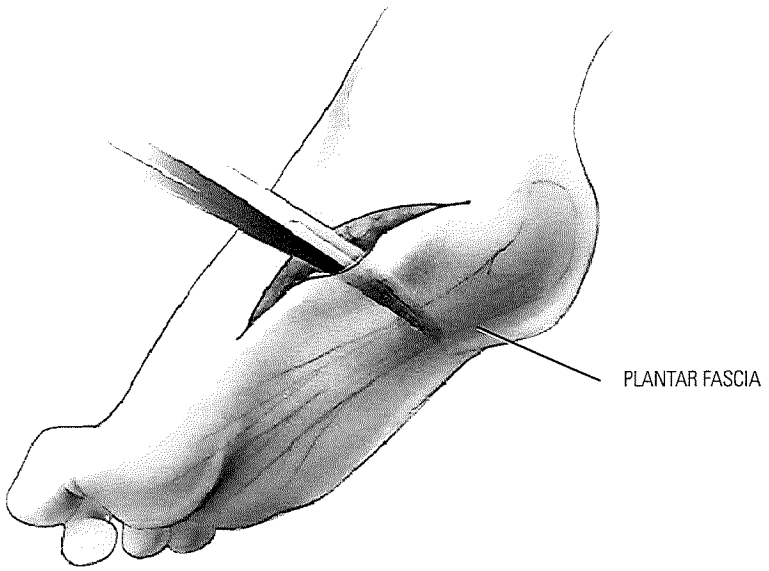


FIGURE 29-9. Plantar Release for Cavus Foot. A plane is developed between the plantar fascia and the subcutaneous fat of the heel pad at the transverse level of the lateral plantar neurovascular bundle. This must extend from the medial to the lateral side of the foot.

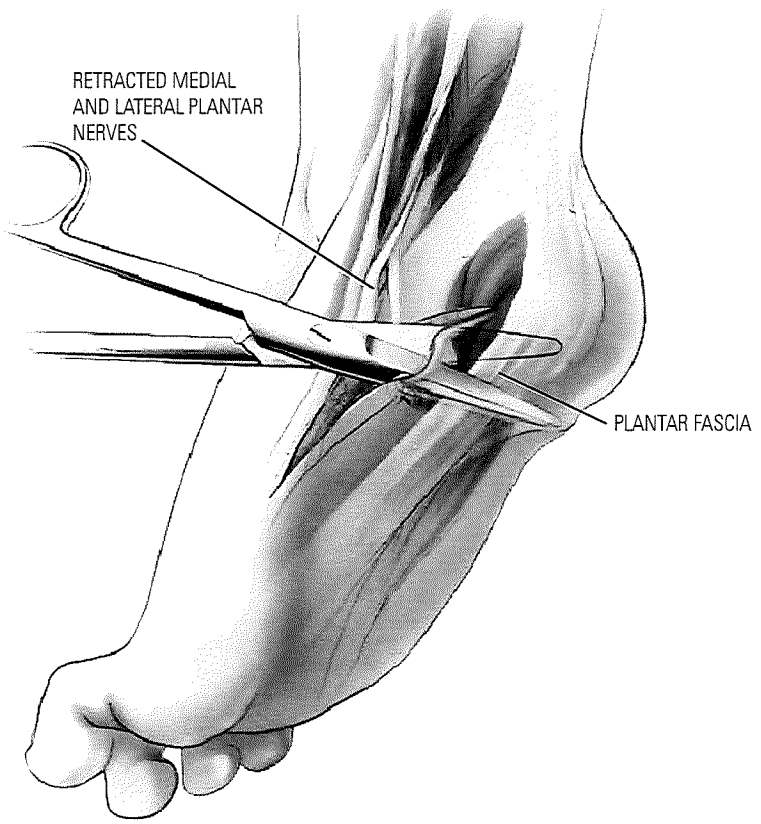


FIGURE 29-10. With the medial and lateral plantar nerves retracted distally and dorsally, a heavy scissors can be used to divide the plantar fascia, the flexor digitorum brevis, and the quadratus plantae (flexor accessorius). One blade of the scissors is passed in the plane that was developed between the plantar fascia and the subcutaneous fat, and the other blade is passed over the dorsal surface of the muscles in the interval plantar to the lateral plantar neurovascular bundle. The surgeon should feel the blades of the scissors near the lateral skin. After these structures are divided, a finger can be passed into the gap to ensure that no tight attachments are left behind.

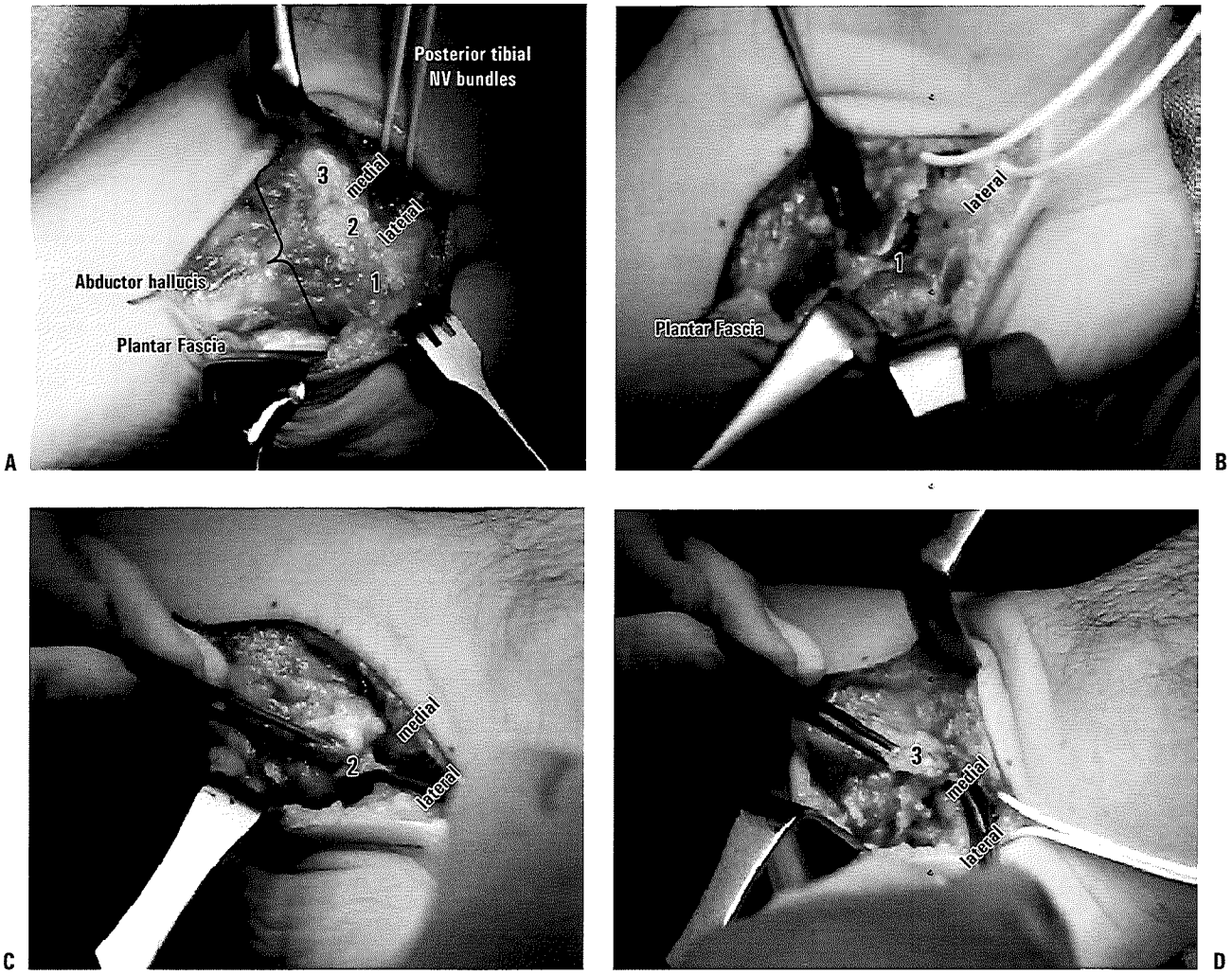


FIGURE 29-11 Superficial plantar-medial release. A: The abductor hallucis muscle has 3 origins on the medial surface of the calcaneus (labeled 1, 2, and 3 from plantar to dorsal). The posterior tibial neurovascular bundle (with white vessel loop around it) divides into medial and lateral plantar neurovascular bundles immediately before passing into the muscle. The laciniate ligament (flexor retinaculum) has been incised vertically in line with the NV bundles to expose the bundles. The plantar fascia is seen as a white band of dense collagen plantar-lateral to the abductor hallucis. It is sharply separated from the thick layer of plantar fat. **B:** After release of the lowest/largest origin of the abductor hallucis muscle (1) and the plantar fascia and short toe flexors, the lateral plantar neurovascular bundle can be seen traversing the foot in a distal-lateral direction. Release of those soft tissues using the tunnel of the NV bundle for guidance obviates injury to those important structures. **C:** The thin septum (and 2nd origin) of the abductor hallucis that separated the medial and lateral plantar NV bundles is divided under direct vision. **D:** The most dorsal origin (3) of the abductor hallucis, which is dorsal to the medial plantar NV bundle, is released. This completes a superficial plantar-medial release for a cavovarus foot deformity with flexible hindfoot varus. (From the private collection of Vincent S. Mosca, MD.)

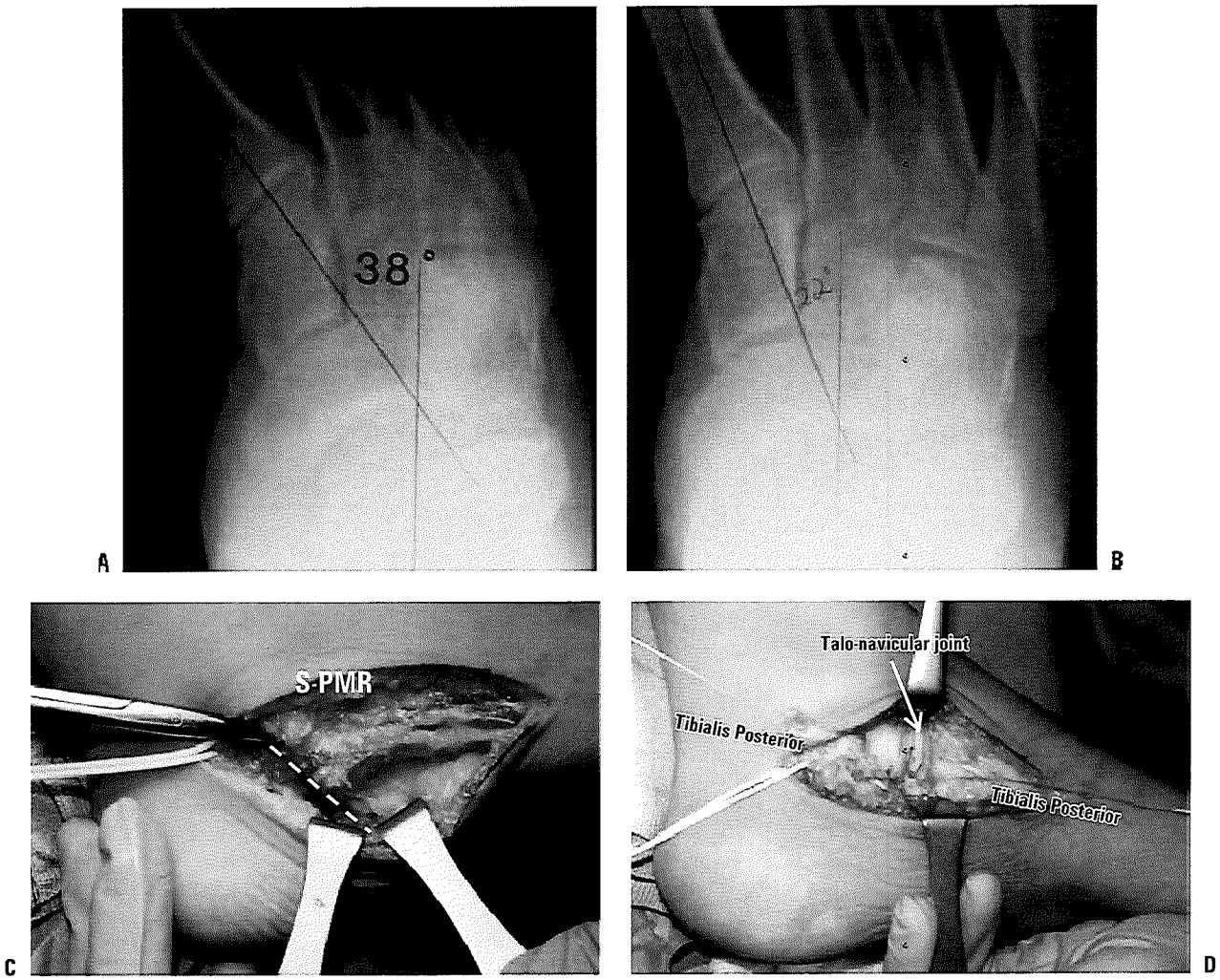


FIGURE 29-12 Superficial and deep plantar-medial releases. A: Standing AP x-ray of a cavovarus foot. The foot-CORA is in the talonavicular joint, confirming that the deformity is hindfoot varus and not midfoot adductus. **B:** The varus deformity does not correct fully, as confirmed by a standing Coleman-type block test x-ray. The foot-CORA is still in the talonavicular joint, but the forefoot axis (and acetabulum pedis) is still medially deviated. This is the indication for a deep plantar-medial release. The subtalar joint inversion requires release, just as it would if this were a clubfoot. **C:** The superficial plantar-medial release is performed first. Besides providing the necessary release of the contracted more superficial structures, it provides access to the deep structures **D:** Deep plantar-medial release. The tibialis posterior tendon is Z-lengthened and the talonavicular joint is released dorsal to plantar, including release of the spring (calcaneo-navicular) ligament. Again consider the analogy to a clubfoot release. (From the private collection of Vincent S. Mosca, MD.)

and, most importantly, is not located at the site of deformity. Osteotomy at the base of all lesser metatarsals might obviate the stress transfer to the second metatarsal; however, there are reported risks of malunion, delayed union, and nonunion (68–71). The lateral column of the foot may require shortening if adductus and varus persist following the procedure(s) on the medial column (78) (see Fig. 29-62). Alternatively, a posterior calcaneus lateral displacement (19, 71, 74) or closing-wedge osteotomy (75, 79, 80) can be employed. This procedure is identical to that used for valgus deformity of the hindfoot (see Figs. 29-86 to 29-90), except the posterior fragment is translated and possibly angled laterally in the varus hindfoot. Using these techniques, all but the most severe and rigid cavovarus deformities can be corrected with preservation of motion in the subtalar joint.

Midfoot osteotomies, such as those popularized by Cole (63), Japas (65), Jahss (66), and Wilcox and Weiner (67), correct the cavus deformity by removing a dorsally based wedge from the midtarsal bones while sacrificing the midtarsal joints to arthrodesis. These procedures should, therefore, be considered second-line treatment for the severe, rigid, neglected, or recurrent cavovarus foot. Preservation of the important metatarsal–tarsal joints (Lisfranc joints) distally and the talonavicular and calcaneocuboid joints (Chopart joints) proximally diminishes the extent of foot rigidity that is created (Figs. 29-13 to 29-17).

Text continued on page 1403

Dorsal Tarsal Wedge Osteotomy for Cavus Deformity (Figs. 29-13 to 29-17)

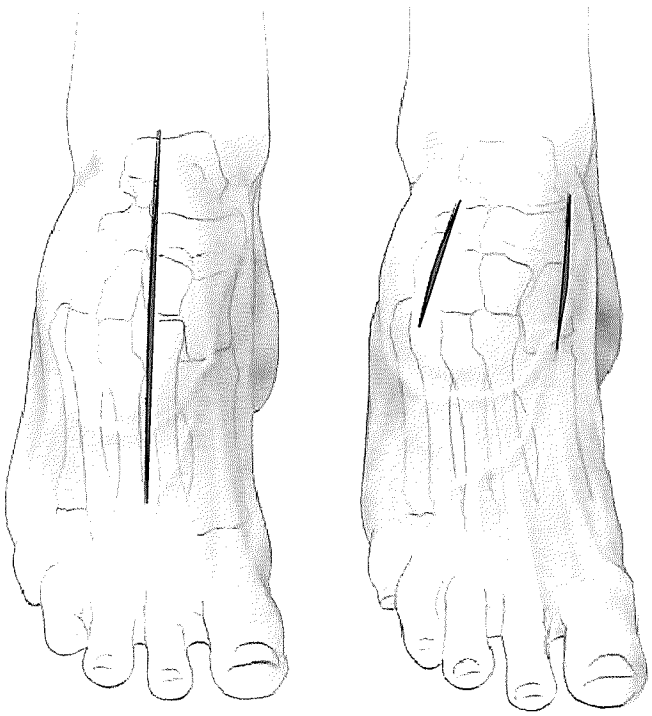


FIGURE 29-13. Dorsal Tarsal Wedge Osteotomy for Cavus Deformity. The midfoot osteotomy operation may be performed through either one long midline incision or two separate incisions, one over the dorsomedial aspect of the navicular and first cuneiform bone and the second over the cuboid bone in line with the fourth metatarsal. In the severe cavus foot, the single incision makes it difficult to reach the lateral extent of the cuboid bone. The incision must extend from the dorsal aspect of the talar neck distally as far as the middle of the metatarsals. Through this incision the entire area of the osteotomy can be exposed extraperiosteally without interference from the anterior or posterior tibial tendons. It is also easier to see the osteotomy through this single incision. It is important that the operation be preceded by a plantar release.

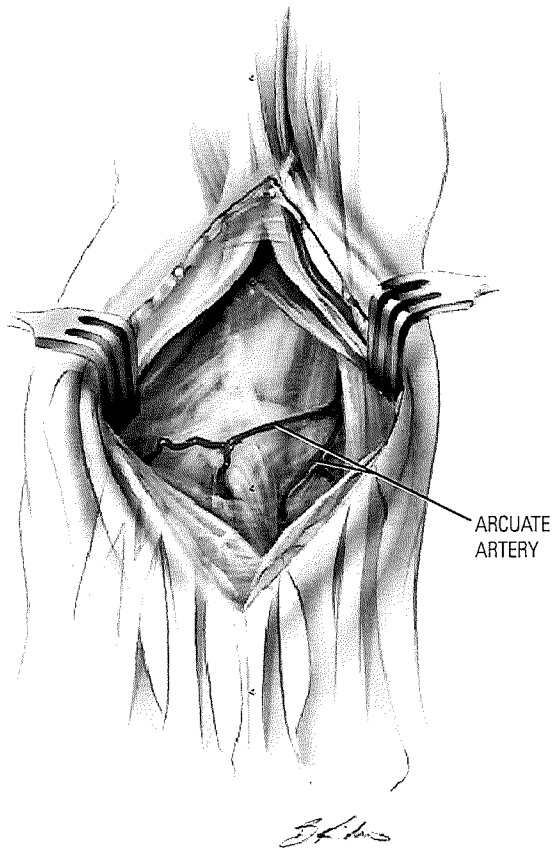


FIGURE 29-14. After the skin and subcutaneous tissues are divided, the interval between the extensor tendons to the second and third toes is developed. The neurovascular bundle lies between the extensor tendons to the second and great toes. In developing this interval, care should be taken to interrupt as few vessels as possible. The arcuate artery coming off the dorsalis pedis artery runs laterally at the level of the tarsal–metatarsal joints. If this is identified, an effort to preserve it should be made.

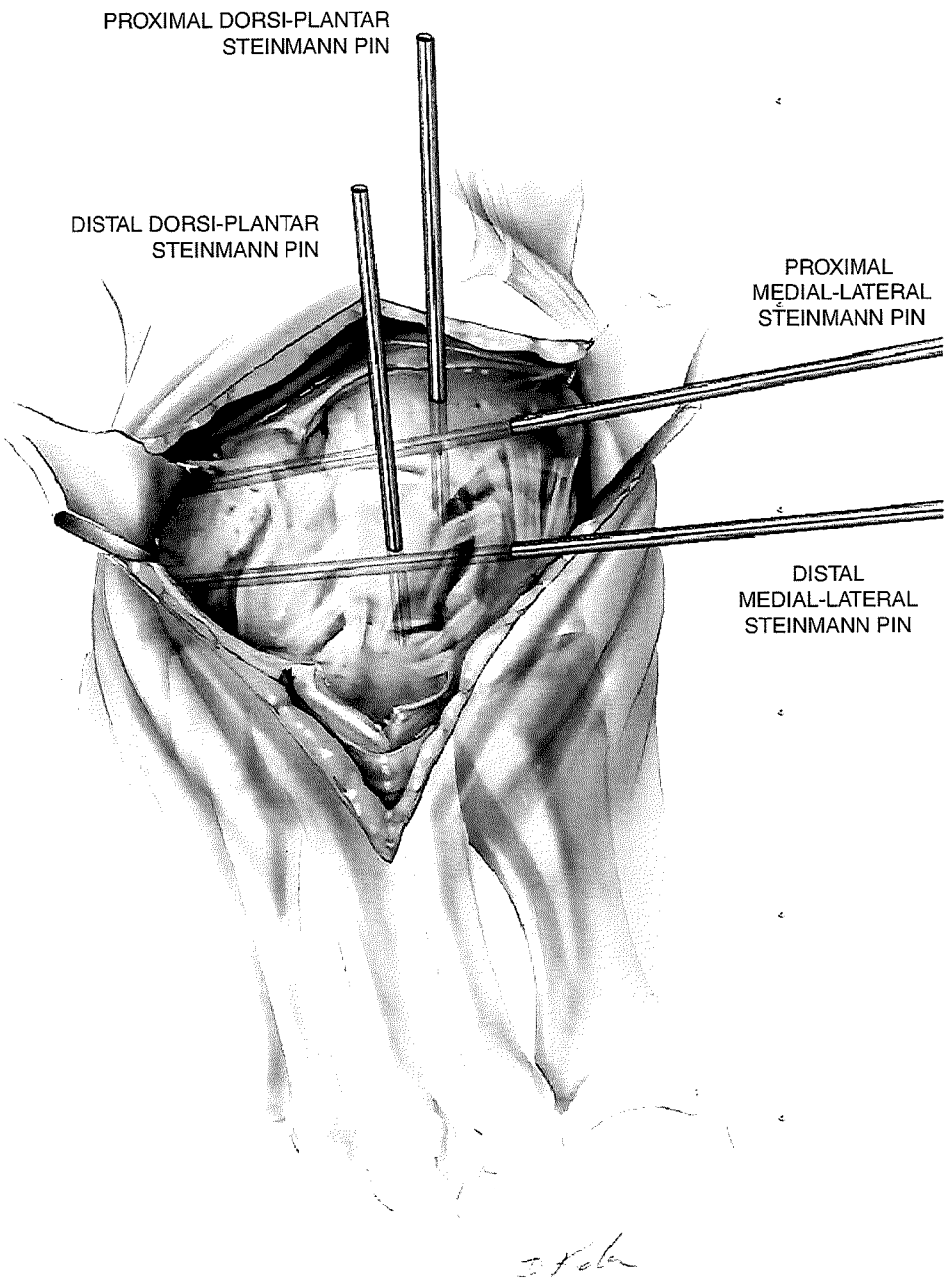


FIGURE 29-15. After this interval is developed, the midtarsal bones should be exposed extraperiosteally between Chopart joints proximally and Lisfranc joints distally, while preserving and protecting those joint capsules. Medially, the dissection should go completely around the navicular first cuneiform joint; laterally, it should go completely around the cuboid bone. Most of the cuboid bone should be exposed, but the joints proximal and distal to it do not need to be entered. Steinmann pins can be used as guide wires to mark the proximal and distal limits of the bone wedge that is to be removed. Insert one from medial-to-lateral parallel with, and immediately distal to, Chopart joints through the navicular and cuboid. Insert another one from dorsal-to-plantar at the level of this transverse pin perpendicular to the desired longitudinal axis of the hindfoot. Insert a third pin parallel with, and immediately proximal to, Lisfranc joints through the three cuneiforms and the cuboid. A fourth pin is inserted from dorsal-to-plantar at the level of the third pin perpendicular to the desired longitudinal axis of the forefoot.

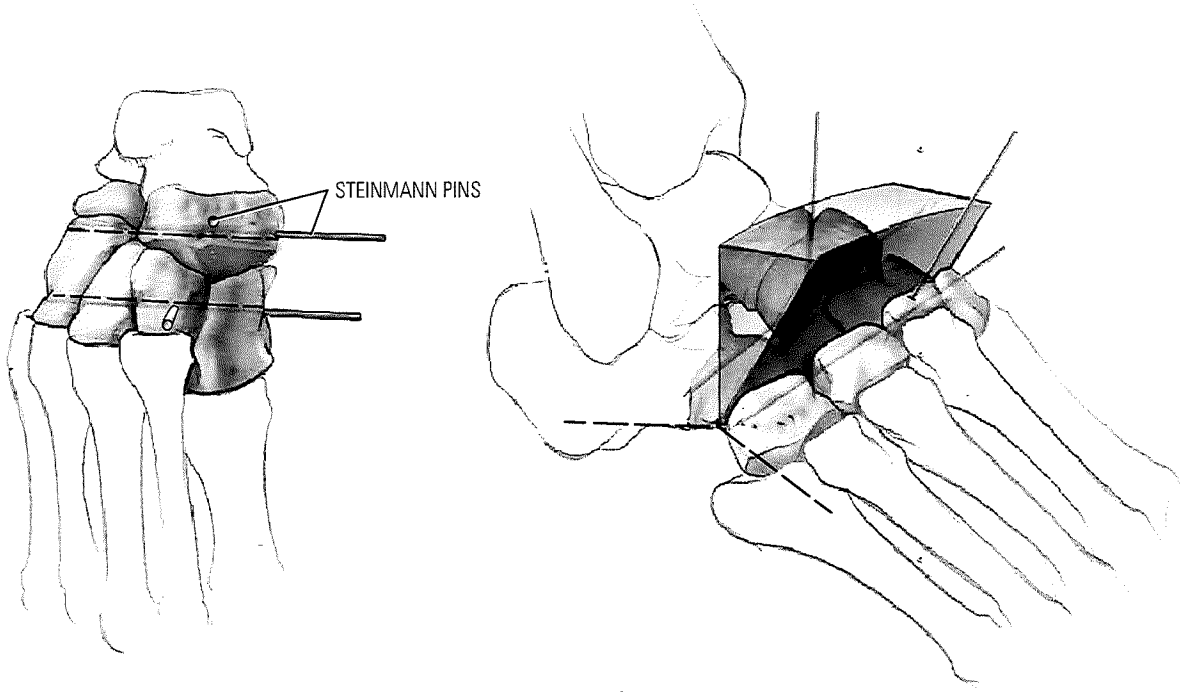
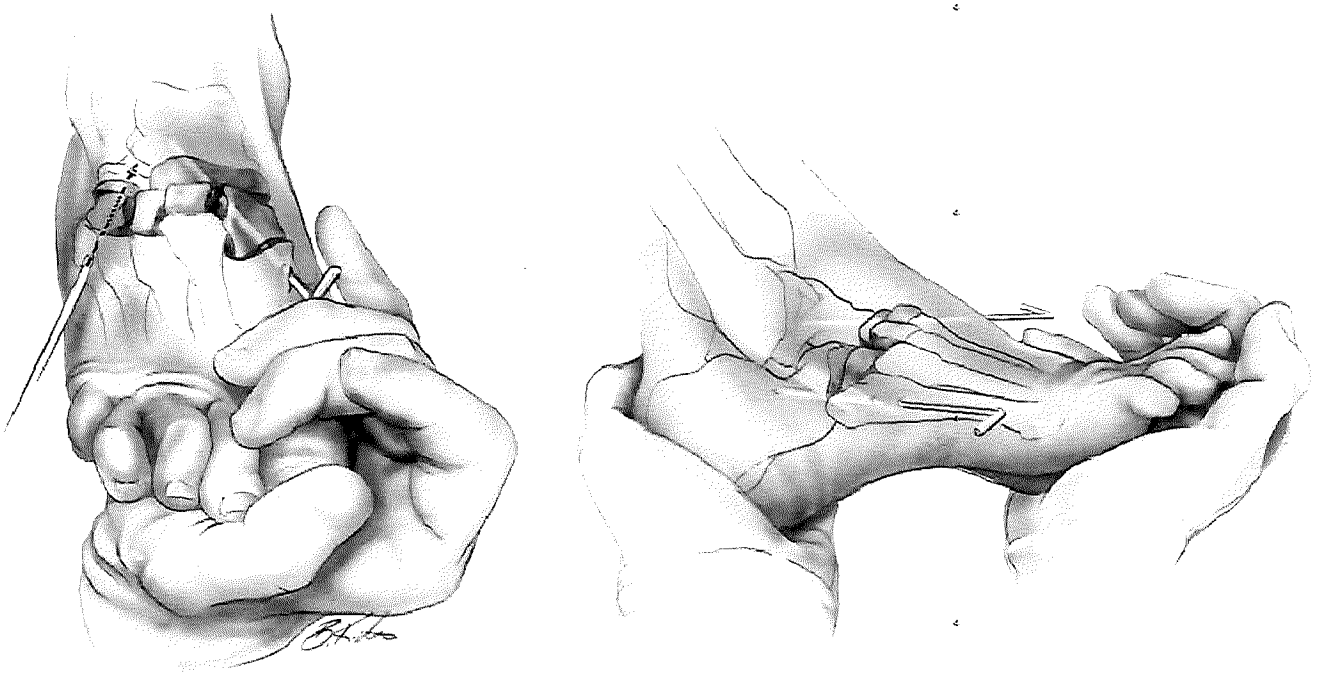


FIGURE 29-16. The osteotomy is performed using a large 1/2-inch osteotome, chisel, or sagittal saw. The plantar soft tissues are protected with wide, curved Crego retractors. The proximal cut is made immediately distal to, and parallel with, the plane created by the two proximal guide pins. It passes through the mid-body of the navicular and the proximal end of the cuboid. This cut is estimated to be perpendicular to the hindfoot axis. The distal osteotomy is made immediately proximal to, and parallel with, the plane created by the two distal guide pins . It passes through the mid-body of each of the three cuneiform bones and the distal end of the cuboid. It is made perpendicular to the axis of the forefoot. It is to be noted that unlike the medial half of the osteotomy, the joints on either side of the cuboid bone are not entered. Rather, the wedge is removed entirely from the cuboid bone. To avoid excessive shortening of the foot, the osteotomies should be fashioned so that no gap of bone is present at the plantar apex of the wedge.



A B

FIGURE 29-17. The osteotomy is closed by elevating the forefoot **(A)**. It is possible to rotate the distal segment, if needed, to correct pronation deformity. Often the first metatarsal will be more depressed than the others. This can be corrected by supinating the forefoot; however, care should be taken not to produce an unintended malrotation. Much depends on the angulation and flexibility of the hindfoot. The osteotomy can be fixed with either two Steinmann pins or multiple staples. The dorsal surface of the cuneiform bones is usually higher than the navicular, and this may make staple fixation more difficult. Secure fixation with Steinmann pins is not as easy as it may first appear **(B)** as the medial pin may pass too far plantarward. The medial pin is inserted first. It must start in the first metatarsal at an oblique angle directed dorsally and laterally. This pin should engage the first metatarsal, the first cuneiform bone, the navicular, and the talus. The lateral pin is started distal to the flare at the base of the fifth metatarsal and is aimed medially and slightly dorsally, crossing the cuboid bone and entering the calcaneus. The ends of the pins are left protruding outside the skin. A well-padded, non–weight-bearing short-leg cast is applied. The foot is kept elevated for the first few days. The patient is then ambulated with a three-point, non–weight-bearing crutch gait for 6 weeks. After 6 weeks the cast and the pins are removed in the office. A short-leg walking cast is applied, and the patient is permitted partial weight bearing for an additional 4 to 6 weeks, at which time healing should be complete.

Removal of a dorsally based wedge from the tarsal bones to correct a fixed cavus deformity with its apex in the midfoot has been described by Cole (63). Because it shortens the foot, its best used only when the problem is bilateral. This disadvantage is offset by preservation of the metatarsal–tarsal joints distally and the talonavicular and calcaneocuboid joints proximally. Because this operation corrects only the cavus deformity, the absence of fixed-heel varus is a prerequisite. The operation should be preceded by a plantar release.

There are variations on this procedure. Jhass (66) has described a similar osteotomy that removes the wedge distally, excising the metatarsal–tarsal joints. A common variation is that described by Japas (65). The results of the Cole procedure have been reported (6).

Triple arthrodesis (Figs. 29-18 to 29-23) originally described by Hoke (81), has been used historically to correct and stabilize severe, rigid, neglected, or recurrent cavovarus foot deformities. The operation is deceptively simple. The talocalcaneal, talonavicular, and calcaneocuboid joints are resected, separating the foot into three movable segments: the forefoot, the calcaneus, and the talus and ankle mortise. If the correct wedges of bone are resected, the position of the foot will be correct when the bony surfaces are apposed. In practice this can be a difficult task. More importantly, it has been shown in many long-term follow-up studies that triple arthrodesis causes stress shifting and premature degenerative arthrosis (19–21, 26, 27, 55) or Charcot arthropathy (82) in adjacent unfused joints, such as the ankle joint. Furthermore, Schwend and Drennan (72) have shown that, even after solid triple arthrodesis, there can be progressive deterioration and recurrence of deformity over time secondary to muscle imbalance. Additionally, it cannot by itself correct the coexisting deformities of the forefoot

Text continued on page 1408

Triple Arthrodesis (Figs. 29-18 to 29-23)

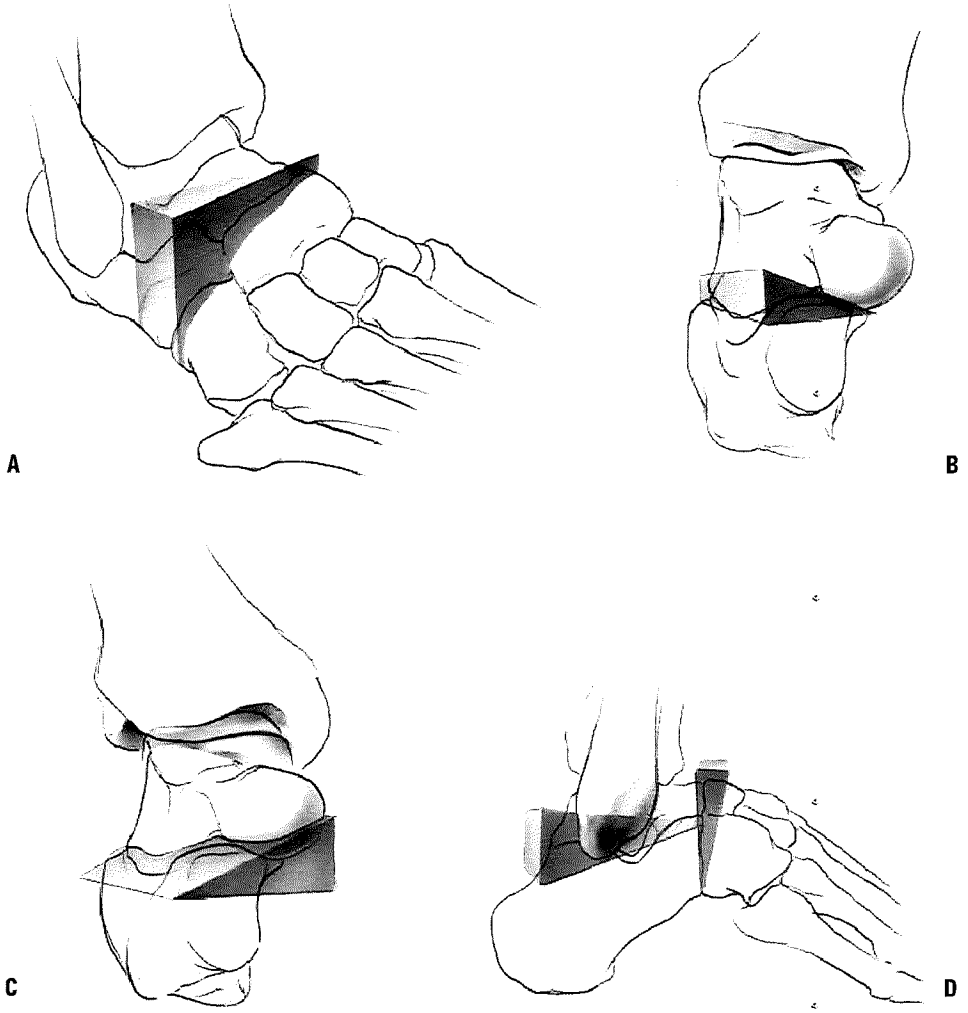


FIGURE 29-18. Triple Arthrodesis. Before beginning the triple arthrodesis operation, the surgeon should give some thought and planning regarding the wedges of bone to be removed and, in particular, the amount of bone to be removed. Simplify the cuts to parallel and perpendicular in relationship to obvious large bony landmarks. It is not particularly beneficial to preoperatively plan precise wedges with cutouts, since the three-dimensional nature of the deformities makes such planning imprecise. Visualizing the foot at surgery and making the osteotomy cuts to create the wedges, as described in the subsequent discussion, seems much more practical and accurate. The most common deformity for which triple arthrodesis is performed is fixed cavovarus deformity. To correct this deformity, a laterally based wedge of bone is removed from each of the joints to be resected. Conceptually, two wedges of bone at right angles to each other are removed. The wedge that will allow correction of the forefoot will excise the talonavicular and calcaneocuboid joints. To achieve correction to a neutral position, the distal cut is perpendicular to the long axis of the forefoot and the proximal cut is perpendicular to the longitudinal axis of the calcaneus **(A)**. When these two surfaces are opposed, the forefoot should be straight. To correct the varus of the hindfoot, a laterally based wedge must be removed from the subtalar joint. To correct the heel to a neutral position, the proximal cut from the undersurface of the talus should be perpendicular to the long axis of the tibia (or parallel with the ankle mortise), whereas the distal cut from the superior surface of the calcaneus should be parallel to the bottom of the heel **(B)**. When these two surfaces are apposed, the heel should be in neutral. A triple arthrodesis for fixed valgus deformity is extremely difficult. This is because the medially based wedges that are created using the espoused principles must be removed from the lateral side **(C)**. This task is simplified if all the joints are widely released by extensive capsulotomies and the interosseous ligament of the subtalar joint is sectioned. A laminar spreader can be used to hold the joints open. Calcaneocavus deformity is the most uncommon indication for triple arthrodesis. In this circumstance a posteriorly based wedge is removed from the subtalar joint, which allows correction of the calcaneus deformity. A dorsal wedge is removed from the talonavicular and calcaneocuboid joints to allow the forefoot to be dorsiflexed **(D)**. A slightly different technique is used for mild deformities. The joint surfaces are simply removed with osteotomes and curettes until there is sufficient resection to gain the desired correction.

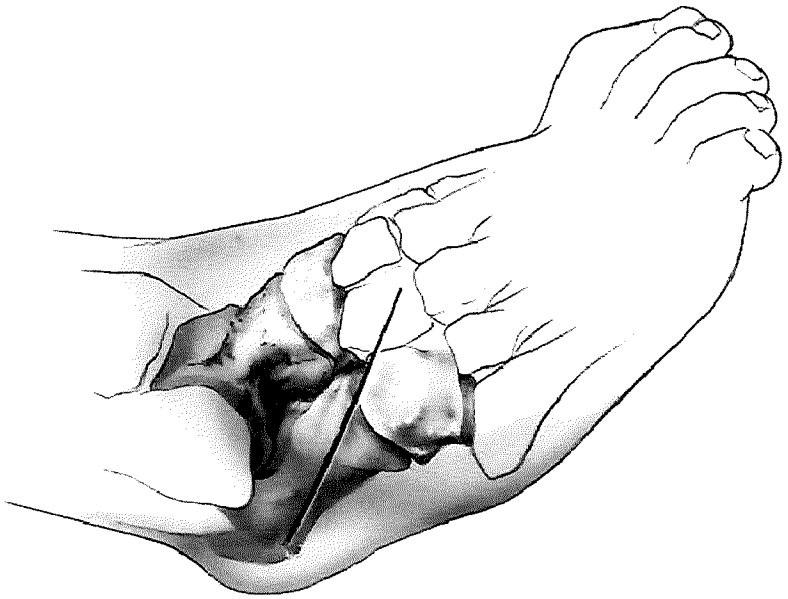


FIGURE 29-19. The triple arthrodesis operation is illustrated for the most common deformity: cavovarus. The patient is placed on the operating table with a sandbag under the hip on the side to be operated, thus bringing the lateral side of the foot into better position. A small, sterile sandbag or other support is placed under the medial side of the foot. This supports the foot while the joint surfaces are cut. The incision is a straight lateral incision that crosses the lateral side of the talonavicular joint and the distal end of the calcaneus. It should extend from just medial to the most lateral extensor tendons dorsally to just past the peroneal tendons volarly. There should be no undermining of the skin edges. The superficial peroneal and sural nerves are retracted and protected. After the fascia over the extensor brevis muscle is incised, the proximal insertion of this muscle is identified and the muscle is elevated to expose the lateral capsules of the calcaneocuboid and talonavicular joints. The fibrofatty tissue is removed from the sinus tarsi, exposing the lateral aspect of the subtalar joint.

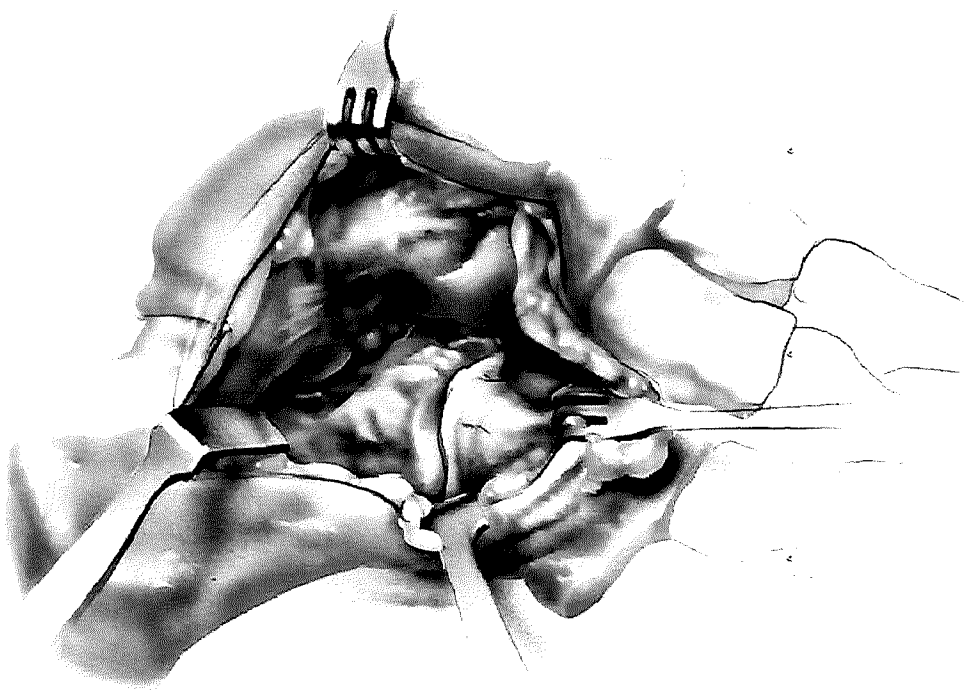


FIGURE 29-20. The talonavicular and calcaneocuboid joint capsules are incised circumferentially, exposing the joint surfaces. It will assist removal of the bone wedges from the subtalar joint if the capsule of the subtalar joint is also nearly circumferentially released. This can be done by sliding a curved periosteal elevator (e.g., a Crego elevator) around the posterior and then medial aspect of the subtalar joint until it rests along the medial side of the joint. At this point, almost the entire capsule of the subtalar joint can be visualized and incised, the interosseous ligament can be divided, and a large bone skid can be used to pry the joint open. This will give the surgeon an excellent view of the two bony surfaces of the subtalar joint that are to be excised.

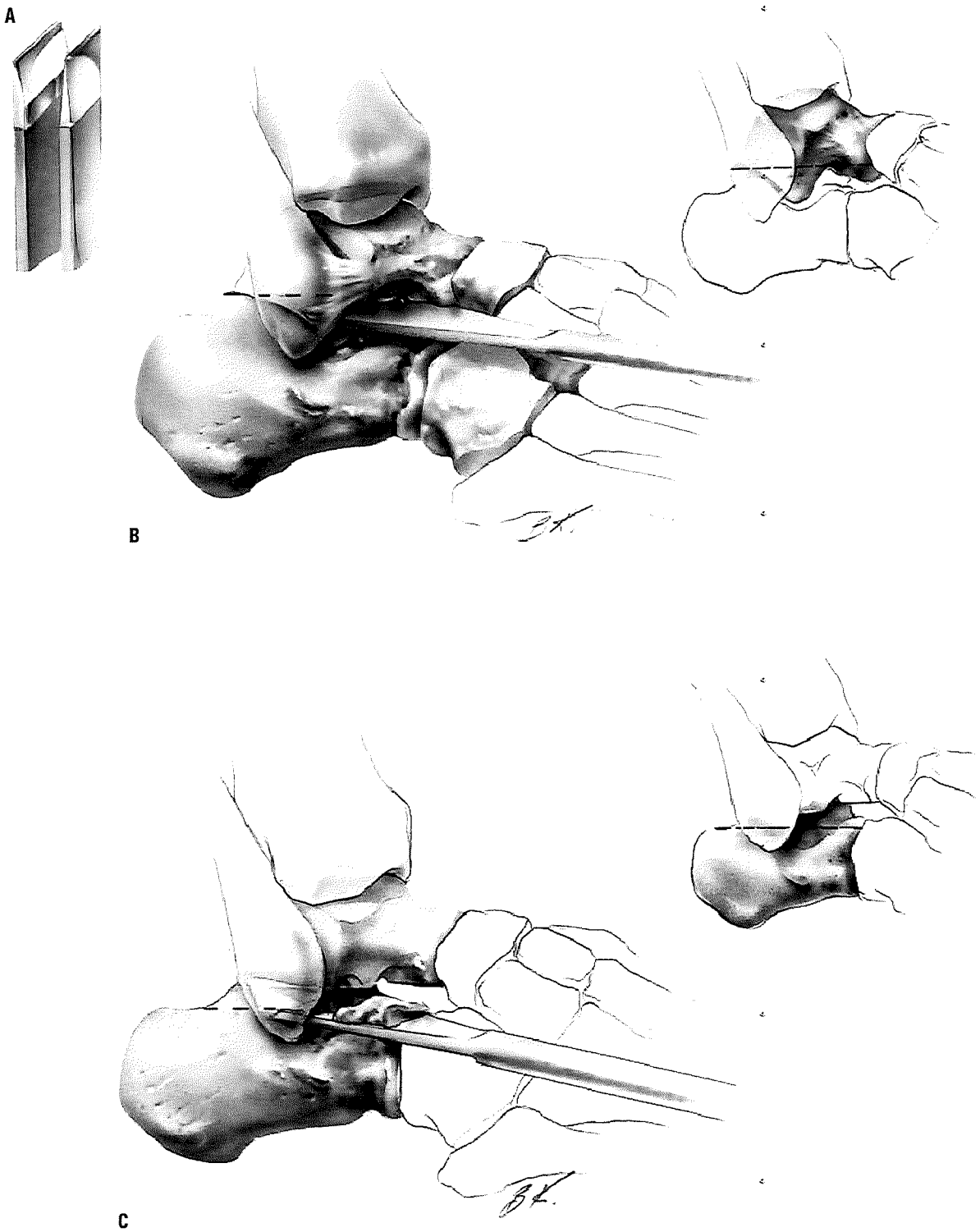


FIGURE 29-21. The wedges of bone are now excised. The subtalar joint is resected first. Most of the bone for the correction should be removed from the calcaneus. It is better to use a chisel than an osteotome for these cuts. The chisel, with its flat surface as opposed to the double-beveled surface of an osteotome, is easier to keep on a straight course **(A)**. The cut in the bottom of the talus should be parallel with the ankle mortise from lateral to medial **(B)**. The cut into the dorsal surface of the calcaneus should be parallel to the bottom of the heel **(C)**. It is best to make the most proximal and distal aspects of these cuts first and the middle portion in between them last. This is because the middle part will be the most difficult to remove with remaining capsule attached to the prominent sustentaculum tali and the most worrisome to cut through with the neurovascular bundle in close proximity. If these cuts are made correctly, the heel will be in neutral alignment regarding varus and valgus when the two cut surfaces are apposed.

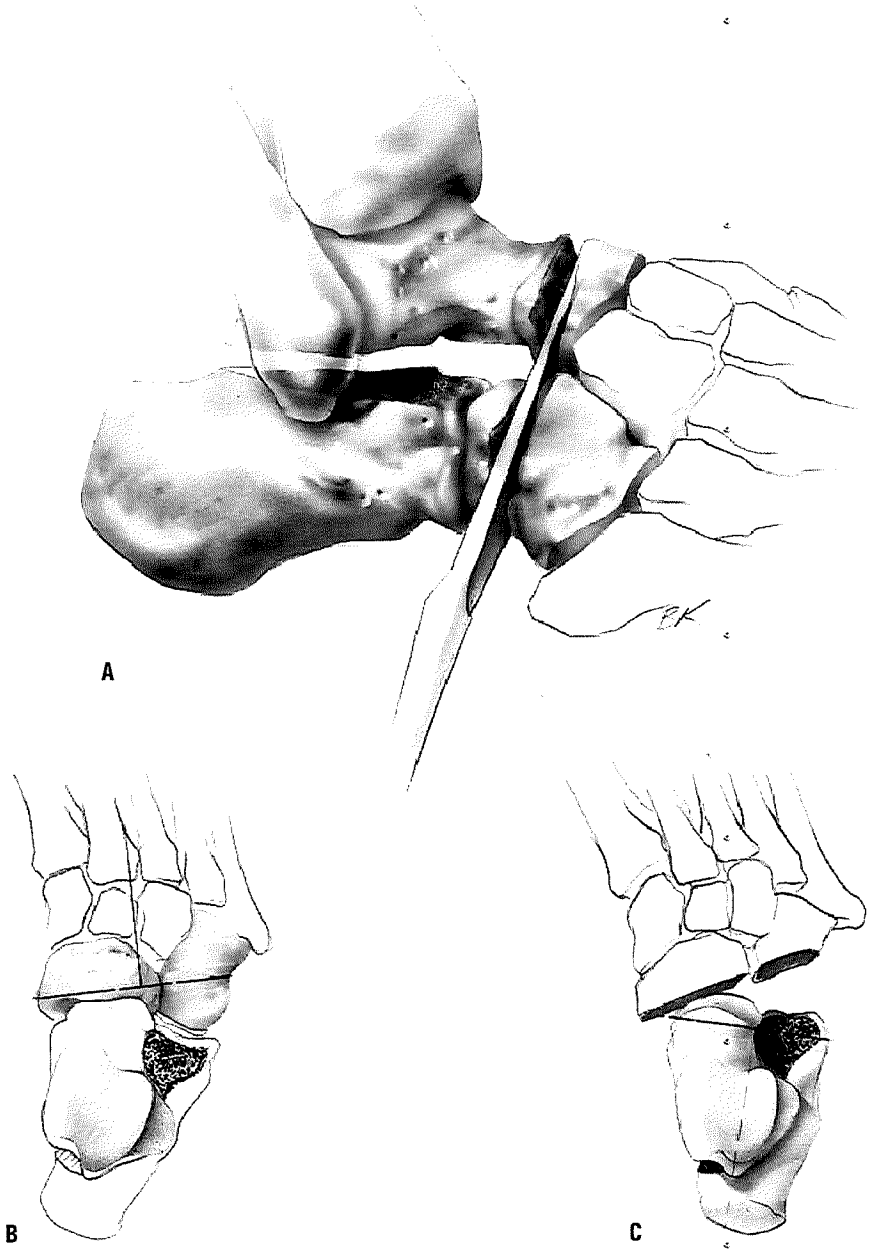


FIGURE 29-22. The same principle is used in aligning the forefoot. The cuts in the navicular and the cuboid should be perpendicular to the longitudinal axis of the forefoot **(A, B)**, whereas the cuts in the distal talus and calcaneus should be perpendicular to the longitudinal axis of the hindfoot or calcaneus **(C)**.

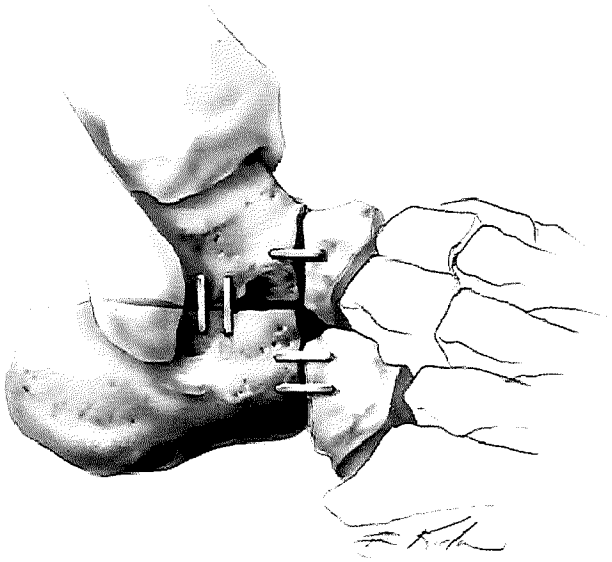


FIGURE 29-23. When the wedges are removed, the foot is placed in the corrected position and the surfaces are inspected. Good coaptation should be present to ensure prompt healing. Trim additional bone as needed. The external contour of the foot should be inspected to ascertain that the desired three-dimensional alignment of the foot has been achieved. If so, the resected joint surfaces are held together with staples, screws, wires, or combinations of these internal fixation devices. A well-padded short-leg non–weight-bearing cast is applied and bivalved to allow for the expected significant swelling that will occur over the next few days. Radiographs are obtained, and the cast is changed to a weight-bearing cast at 6 weeks. At 12 weeks, healing is usually complete, and no further cast protection is needed.

and the hindfoot. Therefore, it should not be used as a primary, or perhaps even a secondary, reconstructive technique in a child or adolescent with a cavus foot deformity. Triple arthrodesis should be reserved as a salvage procedure for existing severe arthritis in the subtalar joint complex or recurrent deformity in older individuals. Mild recurrences in younger individuals are often amenable to additional tendon transfers with or without deformity-correcting osteotomies. If indicated, the operation should be performed within 1 year or at most 2 years of skeletal maturity. In the young child, much of the bone is composed of cartilage. Resection of enough cartilage to obtain good bony apposition is difficult in the young child. In addition, this resection slows the growth of the resected bones. The resulting growth arrest, combined with an operation that in itself shortens the foot, may result in unacceptable shortening of what may already be a short foot.

There are several principles of tendon transfers. A transfer should eliminate a deforming force and bring strength to a site of weakness. It should not leave behind weakness that could shift the balance of forces in the opposite direction. Phasic transfers are better than those that are out of phase. A dynamic tendon transfer cannot correct static bony deformity; the deformity must be corrected concurrently. One must know the existing strength of all muscles. A tendon will lose at least one grade of strength when transferred. When choosing tendons to transfer, one must also anticipate the future strength of the muscles in cases of underlying neurologic disorders in which there will be progressive deterioration. A summary statement about tendon transfers, based on these principles, is that one should move the right tendon to the right location and anchor it at the right tension. This is an art that can be perfected with practice.

Although there are often two or more tendon transfers that are indicated for a particular cavus foot (48, 59, 61–64, 74), the one with the greatest benefits is the transfer of the peroneus longis to the peroneus brevis (62, 74, 76). The peroneus longus, which is the plantar flexor of the first metatarsal (and therefore the pronator of the forefoot), is the primary deforming force in a cavovarus foot. Releasing it will immediately potentiate the strength of the weakened tibialis anterior, the dorsiflexor of the first metatarsal. By then transferring the peroneus longus to the brevis, additional pure eversion strength is concentrated at the base of the fifth metatarsal. In combination with a lengthening, and thereby a weakening, of the tibialis posterior, hindfoot muscle balance is improved.

Deformity correction in the calcaneocavus foot requires radical plantar fasciotomy and release, as well as bony procedures. Dorsal displacement of a posterior calcaneal osteotomy lowers the longitudinal arch by elevating the heel. It is an effective procedure that has the added benefit of preserving joint motion. Alternatively, the midfoot wedge resection osteotomies (63, 65–67) can be employed (Figs. 29-13 to 29-17). Joint preservation is the goal when treating all deformities in the child's foot. The elimination of midtarsal joint motion has less negative long-term consequences than the elimination of subtalar joint motion.

Finally, Ilizarov management has been used in a number of cases of severe foot deformity with reasonable improvement. A combination of distraction osteotomy and joint correction is used in order to achieve satisfactory alignment. This may represent a valuable alternative to triple arthrodesis, but more data are required in this area. Kucukkaya et al. (73) have recommended a V osteotomy using the Ilizarov frame and method for management of foot deformity in poliomyelitis and CMT. In a series of patients treated in this manner, they documented nearly uniform achievement of a painless plantigrade foot. It appears that there is a role for a combination of soft-tissue stretching and osteotomy in the correction of severe deformity using an Ilizarov frame (73).

Cleft Foot (Ectrodactyly)

Definition. A cleft foot, or split foot, is characterized by varying degrees of central longitudinal fissuring and deficiency involving the soft tissues and bones (83). The term "lobster claw foot" used by Cruveilhier (83) in 1829 is no longer appropriate as a description of this clinical condition. Ectrodactyly, which

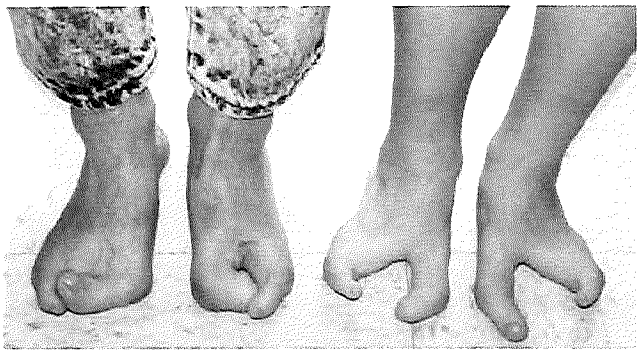


FIGURE 29-24. Father and son with cleft feet. (From the private collection of Vincent S. Mosca, MD.)

means congenital absence of all or part of a central digit, is an acceptable synonym.

Epidemiology. The cleft foot is a rare condition with an overall occurrence rate of 1 per 90,000 live births (84). In the most common form of this condition, the malformation is bilateral, usually associated with cleft hands, and has an autosomal dominant pattern of inheritance with incomplete penetrance (84–86) (Fig. 29-24). In the less common form, the cleft foot is unilateral without associated hand malformation, and there is no evidence of familial inheritance. The incidence of this form is 1 in 150,000 (84). Approximately 10% of cleft feet have no family history (87). Boys are affected more frequently than girls.

Etiology. The apical ectodermal ridge (AER) induces normal development of limb buds by interaction with the underlying mesenchyme (88). A defect of the AER, by genetic or toxic influences, could induce osseous syndactyly by deficient differentiation, polydactyly by excessive differentiation, or a central defect by another mechanism. Watson and Bonde (89) proposed that cleft formation was the result of selective damage to a specific region in the AER localized to the second or third ray, which is the typical area for deficiency in a cleft foot. They further proposed that the extent and duration of damage to the AER would relate to both the width and depth of the defect.

Clinical Features. Variations of the malformation known as cleft foot range from a mere deepening of the interdigital commissure to the typical central ray deficiency to the monodactylous foot (90). The width of the foot at the metatarsal heads is excessive in comparison with the hindfoot, particularly in those feet with a greater deficiency of rays. And in these feet with greater ray deficiencies, there are often exaggerated hallux valgus and fifth toe varus orientations (Fig. 29-24). Anomalies associated with cleft foot include cleft hand, cleft lip and palate, deafness, urinary tract abnormalities, triphalangeal thumb, and tibial hemimelia.

Radiographic Features. Blauth and Borisch (90) have classified cleft foot into six groups based on the number of metatarsal bones (Fig. 29-25). They identified two additional forms: a polydactylous type and a diastatic type. The latter is characterized by monodactyly with lower leg diastasis or tibial aplasia or both. Other variations of morphology in this condition, such as crossbones and synostoses, are also described by the authors (Fig. 29-26). In addition to radiographs of the foot, renal ultrasonography is indicated to rule out renal abnormalities, as they are frequently associated with cleft foot.

Pathoanatomy. The pathoanatomy is one of variable degrees of failure of formation with occasional duplication and malorientation of bones. Synostoses may be seen at the margins of the cleft and in the tarsals (90). Often a hallux valgus deformity results from the pull of conjoined flexor and extensor tendons, resulting in an abduction force.

Natural History. Many patients with cleft foot function well and can wear regular shoes without pain or compromise of function, while others do well with accommodative shoe wear. For such individuals, no surgical treatment is required.

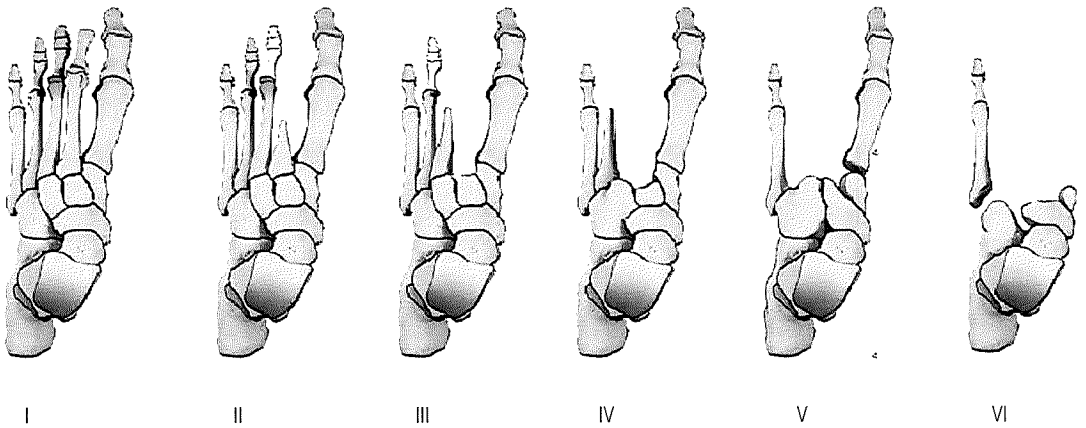


FIGURE 29-25. Cleft foot classification according to Blauth and Borisch. (From Blauth W, Borisch NC. Cleft feet. Proposals for a new classification based on roentgenographic morphology. *Clin Orthop* 1990; 258:41–48, with permission.)

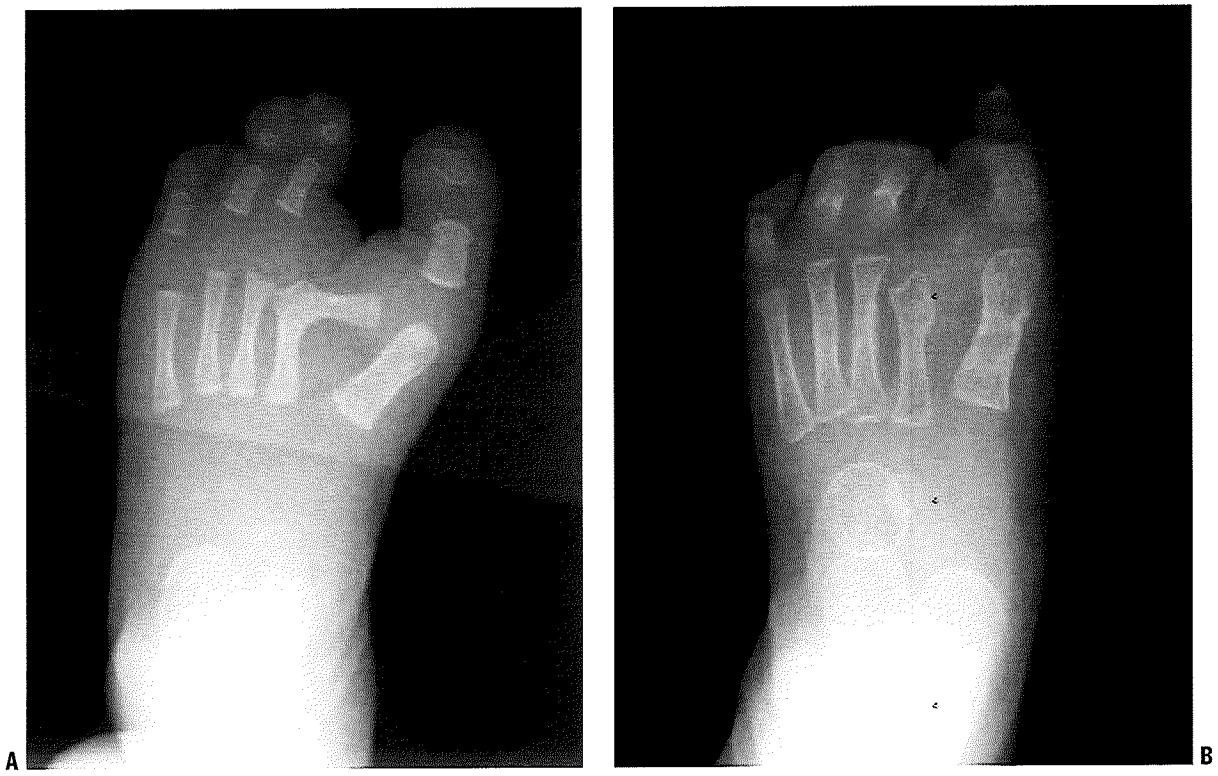


FIGURE 29-26. **A:** This patient had a mild cleft foot with a transverse metatarsal bridging the cleft and widening the foot. **B:** With repair of the cleft and removal of the transverse metatarsal, a healthy foot resulted. (From the private collection of Vincent S. Mosca, MD.)

However, in many cases of cleft foot, there is a marked increase in the width of the forefoot (Fig. 29-24), making it difficult or impossible to find shoes that fit comfortably. Painful callosities develop over the medial and lateral metatarsal heads.

Treatment. The primary goals of cleft foot treatment are comfort and good function in standard shoes. A secondary goal is socially acceptable appearance. Treatment begins with education of the physician on the treatment used in the family member(s) from whom the malformation was inherited (Fig. 29-24).

Surgical techniques for the symptomatic cleft foot must be individualized. The problem of symptomatic splayfoot can be addressed effectively with an osteotomy or osteotomies at the base of the lateral, and occasionally the medial, ray of the foot (Fig. 29-27). The soft tissues in the cleft can be partially approximated to improve the appearance and help maintain the corrected deformity (91). Hallux valgus and digiti minimi varus deformities are often severe. Soft-tissue reconstructions, such as those used to correct hallux valgus, are ineffective. Distal osteotomies of the metatarsals can be employed to improve the malalignment, but recurrence of these distal deformities is common even following osteotomies. Complex plastic surgical procedures that create the appearance of toes have not added sufficient functional benefit or cosmetic improvement to justify their implementation (92).

Clubfoot (Congenital Talipes Equinovarus)

Definition. Clubfoot is the term used to describe a complex, congenital, contractural malalignment of the bones and joints of the foot and ankle. The individual deformities include equinus of the hindfoot, varus (or inversion) of the subtalar joint complex, cavus (plantar flexion of the forefoot on the hindfoot), and adductus of the forefoot on the midfoot (Fig. 29-28). Congenital talipes equinocavovarus is a more descriptive term, though certainly more cumbersome.

There are four recognized classes of clubfoot. This section will focus on the idiopathic variety, the most common type, which is found in otherwise normal children and does not resolve without intensive treatment. A second class is the postural variety. This type resolves completely without intervention, or with manipulation alone, or with one or two casts. Neurogenic clubfoot refers to that seen in children with myelomeningocele. Syndromic clubfoot is seen in children with other anomalies (Table 29-2). The latter two types tend to be rigid and quite resistant to treatment.

Epidemiology. The incidence of clubfoot is 0.93 to 1.5 per 1000 live births in Caucasians (93–95), 0.6 per 1000 in Asians, 0.9 per 1000 births in Western Australia (96), and 6.8 per 1000 in Hawaiians, Polynesians, and Maorians (97). Boys are affected twice as often as girls are, and bilateral involvement is seen in approximately 50% of cases (93, 94, 98).

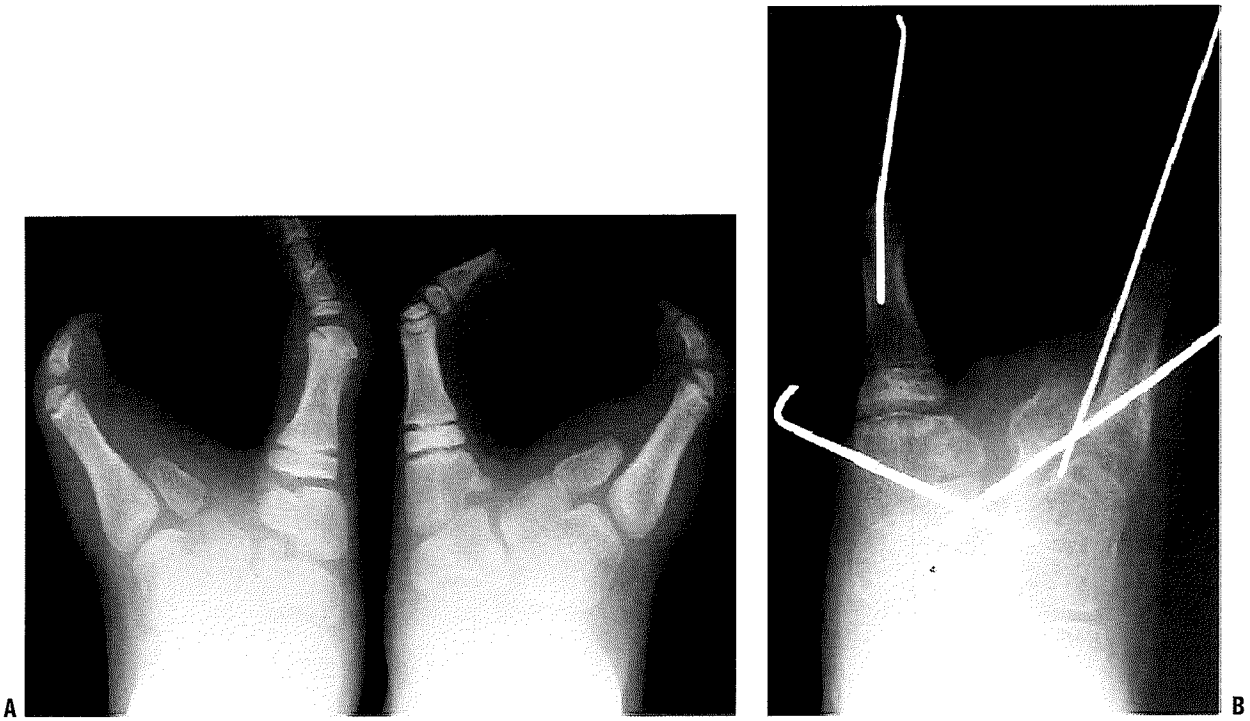


FIGURE 29-27. Preoperative **(A)** and postoperative **(B)** radiographs of symptomatic cleft foot treated with osteotomies. (From the private collection of Vincent S. Mosca, MD.)

Wynne-Davies (94, 95) determined that the occurrence rate is 17 times higher than the normal population for first-degree relatives, is 6 times higher for second-degree relatives, and approximates the normal population risk for third-degree relatives. Unaffected parents with an affected son have a 1 in 40 (2.5%) chance of having another son with the disorder, while the risk to a subsequent daughter is very low. Unaffected parents with an affected daughter have a 1 in 16 (6.5%) chance of having a son with clubfoot and a 1 in 40 (2.5%) chance of seeing the deformity in another daughter. The chances are about one in four that a subsequent child will have a clubfoot if a parent and child have the disorder. There is a 32.5% rate of concordance in monozygotic twins and a 2.9% rate in heterozygotic twins.

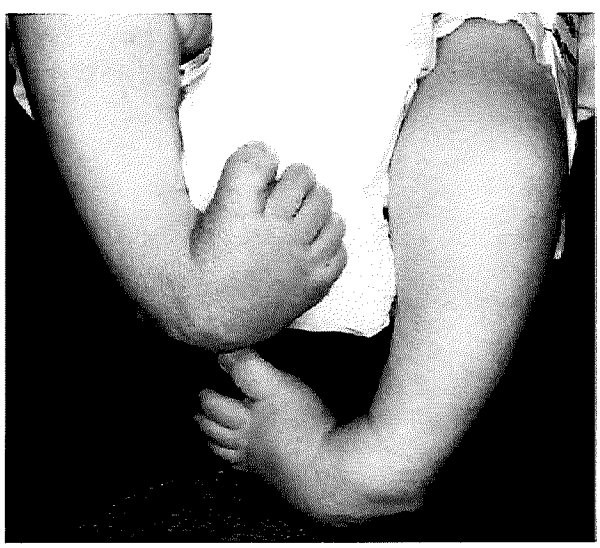


FIGURE 29-28. Clubfoot deformity is associated with forefoot supination, deep medial creases, and equinovarus of the hindfoot. (From the private collection of Vincent S. Mosca, MD.)

Etiology. Clubfoot is probably etiologically heterogeneous. Genetics clearly plays a part. The observations of Wynne-Davies (94, 95) on occurrence rates led her to propose that clubfoot is inherited as a dominant gene with reduced penetrance or multifactorial inheritance. Cowell and Wein (98) concluded that the data could be accounted for using a multifactorial inheritance model. Complex segregation analysis using a regressive

TABLE 29-2 Syndromes with Which Clubfoot Is Commonly Associated

Arthrogryposis
Constriction bands (Streeter dysplasia)
Prune belly
Tibial hemimelia
Möbius syndrome
Freeman-Sheldon syndrome (whistling face) (autosomal dominant)
Diastrophic dwarfism (autosomal recessive)
Larsen syndrome (autosomal recessive)
Opitz syndrome (autosomal recessive)
Pierre Robin syndrome (X-linked recessive)

logistic model was used by Rebbeck et al. (99) to conclude that the probability of having clubfoot was explained by the mendelian segregation of a single gene with two alleles plus the effects of some other factors that are yet to be elucidated.

The proximate cause of the genetic message is debated with fervor, based on a small number of personal observations by a large number of investigators. There may be validity in many of the discordant studies if, in fact, clubfoot is a clinical manifestation that can result from multiple causes and mechanisms. Among the proposed theories are *in utero* molding (100), primary muscle lesion (101), primary bone deformity (germ plasm defect) (102), primary vascular lesion (103), intrauterine enteroviral infection (104), developmental arrest (105), primary nerve lesion (106), abnormal tendon insertion, retracting fibrosis (107), and abnormal histology (108).

Environmental factors may modulate the genetic expression of the disorder. Skelly et al. (109) demonstrated an increased risk of clubfoot for mothers who smoked during pregnancy. The data were particularly convincing because the risk increased with increased numbers of cigarettes smoked per day. Honein et al. (110) also identified maternal smoking as a significant etiologic factor. The joint effect of family history of clubfoot and maternal smoking during pregnancy was more than additive, suggesting a genetic–environmental interaction.

Other associations with clubfoot have been identified, such as increased ligament laxity in families of children with clubfoot (111) and increased internal hip rotation in a limb with a clubfoot (112). The implications of these findings are yet unknown.

Clinical Features. The deformities of the clubfoot fit the acronym CAVE. They are cavus (plantar flexion of the forefoot on the hindfoot), adductus of the forefoot on the midfoot, varus (or inversion) of the subtalar joint complex, and equinus of the ankle. These deformities are not passively correctable. The severity of the deformities and associated findings vary from foot to foot, even in bilateral cases. There is a single (occasionally double) posterior ankle skin crease (Fig. 29-29).

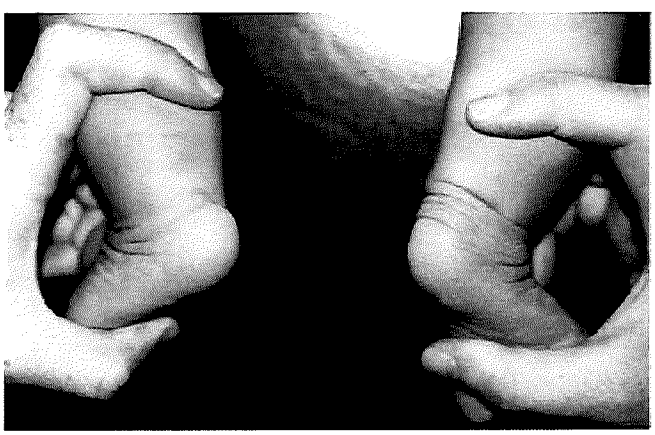


FIGURE 29-29. Clubfoot (left) with single heel crease and healthy foot (right) with multiple heel creases. (From the private collection of Vincent S. Mosca, MD.)

The calcaneus is difficult to palpate within the fatty heel pad, resulting on the so-called empty heel pad sign. A deep transverse skin crease crosses the midfoot and extends under the longitudinal arch. Assessment of tibial torsion in a newborn with a clubfoot is unreliable. The head of the talus can be seen and palpated on the dorsolateral aspect of the midfoot/hindfoot just anterior to the ankle joint. This is due to excessive inversion of the subtalar joint complex around the talus. An examiner's thumb can be placed on the dorsolateral aspect of the head of the talus as a solid fulcrum around which one can attempt to evert the subtalar joint. In an idiopathic clubfoot, the navicular will not fully align with the head of the talus and displace the examiner's thumb.

The foot and calf are smaller than the contralateral side. Little and Aiona (113) noted a leg length discrepancy of >0.5 cm in 18% of children with unilateral clubfoot and 4% with bilateral deformities. In those with unilateral deformity and discrepancy, the tibia is short in 89% and the femur is short in 43%. Spiegel and Loder (114) found a leg length discrepancy of >0.5 cm in 32 of 47 patients with a unilateral clubfoot, a frequency that is perhaps more accurate.

In 2009, Howlett and Mosca (112) reported increased internal hip rotation in limbs with clubfoot deformity. Because this was a clinical study, it was not possible to determine if this finding was due to femoral anteversion, acetabular anteversion, or both. There was at least 10 degrees greater internal hip rotation, and 10 degrees less external rotation, ipsilateral to a unilateral clubfoot compared to the nonaffected limb in >80% of cases. Children with bilateral clubfoot deformities had at least 10 degrees greater internal hip rotation than that reported for age-matched normal controls (115). There might be genetic implications for this finding that are yet to be elucidated, but the clinical implications are significant. An in-toeing gait in a child with a clubfoot could represent persistence or recurrence of the clubfoot deformity, but, if the in-toeing is due to increased internal hip rotation, it must be identified and differentiated before considering inappropriate treatment of a well-corrected clubfoot.

A complete physical examination of the child is indicated to rule out a neurogenic or syndromic etiology for the deformity (Table 29-2). Muscle testing and sensory examination should be part of the initial examination in all patients, as clubfoot has been found to be associated with absent anterior compartment muscles and lesions involving the innervation to the anterior and lateral compartment muscles. A finding as subtle as adducted and contracted thumbs across the palms will identify a child with clubfeet as having arthrogryposis. Examination of the hips in a newborn is an important part of the musculoskeletal screening exam, but there is no reported increased risk for developmental dysplasia of the hip in children with clubfoot (116, 117).

Classification of the severity and rigidity of the clubfoot is important for the comparison of treatment modalities. Several classification systems have been proposed (120-122, 125, 163, 173, 528), but none has been universally accepted. They all suffer from excessive subjectivity and only fair reproducibility.

Clinical Classification of Clubfoot. With respect to classification of clubfeet, important clinical features must be documented. Included in these features are

1. The severity and rigidity of the deformities
2. Depth of the skin creases (Fig. 29-29)
3. Tightness and contractility of the muscles

Although there are a number of classification systems in use, two of them seem to be of particular value in attempting to classify clubfeet at the initiation of treatment. One of these classification systems was defined by Dimeglio et al. (121) and the second by Pirani (122, 123). The classification systems apply a point score to a number of physical findings, which, when totaled, leads to a "grade of involvement." Flynn et al. (124) showed a good correspondence between the classification systems of Dimeglio and Pirani. They found a correlation coefficient of 0.83 with the Dimeglio system and 0.9 with the Pirani system when applied by three individuals. The correlation improved in both systems after the initial 15 feet were scored. Wainwright et al. (125) compared four classification systems: Dimeglio, Catteral, Harold and Walker, and Ponseti and Smoley, and determined that the Dimeglio system was the most reproducible. Both the Dimeglio and the Pirani point systems attempt to differentiate between mildly affected feet requiring little treatment and those that are extremely severe. If the outcomes of treatment are to be compared, a valid classification system must be employed before the initiation of treatment.

One should create a checklist with either or both of the systems and attempt to score clubfeet at the initiation of treatment. Each of the scoring systems can be applied in <2 minutes. In the Pirani system, isolated physical findings, including the severity of deformity, the depth of skin creases, and the degree of certain pathoanatomic variations of the midfoot and the hindfoot, are each given severity scores of 0, 0.5, or 1 with a maximum possible score of 6. In the Dimeglio system (Fig. 29-30), the deviations of the foot—equinus, varus, adductus, and foot rotation—are scored based on the degree of deformity. Scores for the depth of skin creases, presence of cavus, and the condition of the muscles are combined to give a maximum possible score of 20. Choose one and attempt to grade clubfeet on the basis of these clinical criteria.

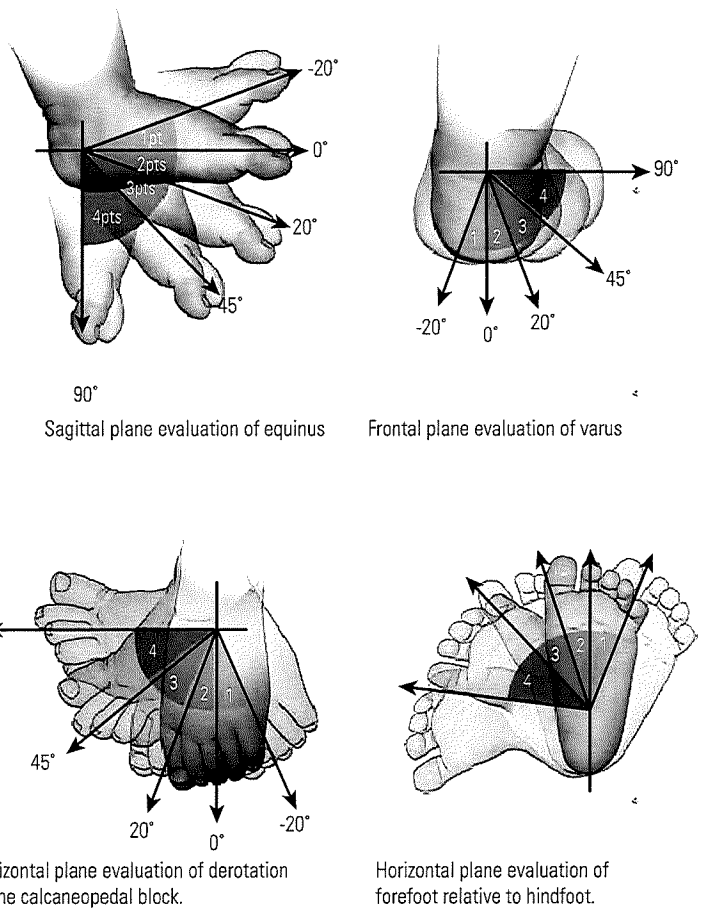


FIGURE 29-30. Dimeglio's classification system for clubfoot deformity rates the position of the foot relative to equinus, varus, foot rotation, and forefoot medial deviation. These are scored from 0 to 4 on the basis of severity. Finally, the depths of posterior crease, medial crease, cavus, and muscle condition are each assigned a 0 or 1 point score. Total score ranges from 0 to 20 points, correlating with the severity of the clubfoot deformity. (From Dimeglio A, Bensahel H, Souchet P, et al. Classification of clubfoot. *J Pediatr Orthop B* 1995;4:129–136, with permission.)

Radiographic Features. There is no consensus on the role of radiographs in the diagnosis and management of the clubfoot (121, 122). However, it should be stated that the diagnosis of clubfoot in the newborn can and should be based solely on clinical findings. The intended role of radiographs in the assessment of foot deformities is to demonstrate the relationships between bones. This is accomplished by first drawing the axis of each bone, and herein lies the limitation of this imaging modality. There is little ossification of the bones in the normal newborn foot, and there is a delay in ossification in the clubfoot (126). The ossific nucleus of the talus is not centrally located in the cartilaginous anlage (127, 128). The ossific nucleus of the talus is between the head and neck and may be spherical in shape for the first several weeks of life. Ossification of the navicular does not begin until age 3 to 4 years in children with clubfoot and even then is eccentric. Brennan et al. (129) identified an even more fundamental problem when they showed very poor reproducibility in positioning the clubfoot for the radiograph. These factors make it unrealistic to consider radiographs of the newborn and infant clubfoot as objective data. The age or point at which radiographs become reliable is unclear.

Despite these limitations, there might be a role for radiographs to confirm correction of the clubfoot deformities or to help identify the site(s) of residual deformity in the child who is several months old and has been undergoing serial manipulation and casting. In the latter situation, the information can be helpful for surgical planning, particularly if one ascribes to à la carte surgery (118, 119, 130). The AP view is obtained with the foot pressed against the radiographic plate with a dorsiflexion and external rotation force (120, 131) (Fig. 29-31A). This will place the subtalar joint in its most everted and corrected position. The lateral radiograph is obtained with the foot dorsiflexed and maximally everted, but also with the leg internally rotated to assure a true lateral view of the ankle (manifest by the projection of the fibula within the posterior half of the tibia) (Fig. 29-31B). The talocalcaneal and talus–first metatarsal angles are measured on both views (10). The axis of the talus and calcaneus normally diverge from each other and the axis of the talus and the first metatarsal normally form a nearly straight line on both views. The tibiotalar and tibiocalcaneal angles are measured on the lateral view (10). The axis of the talus normally aligns almost perpendicular to the tibia and the calcaneus dorsiflexes above a right angle with the tibia. The alignment of the calcaneus and cuboid are assessed on the AP view (132).

A second point at which radiographs may be helpful is intraoperatively to confirm the adequacy of correction of the deformities. The low dose radiation and convenience of minifluoroscopy make that technology desirable. The third indication for radiographs could be at some substantial time after surgery to confirm maintenance of deformity correction. Alternatively, the third point at which radiographs are obtained is when recurrence or other secondary deformities are identified.

Other Imaging Studies. In response to the limitations of radiographs, ultrasound techniques are evolving for the assessment of the infant clubfoot during nonoperative and operative treatment (133–137). Early results seem promising; however, availability of the technology in orthopaedic outpatient clinics

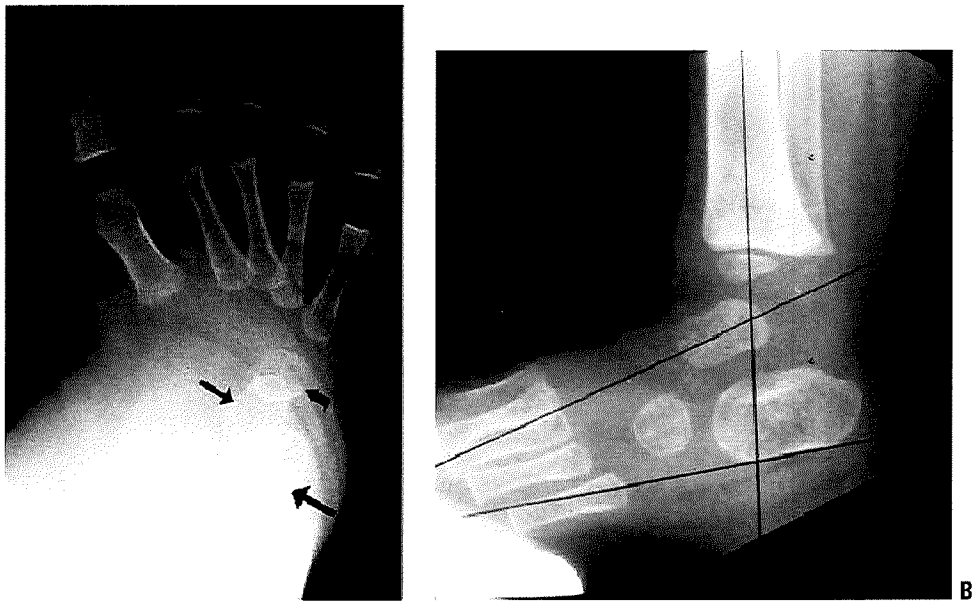


FIGURE 29-31. A: Simulated weight-bearing anteroposterior radiograph of clubfoot. The talus (*small straight arrow*) and calcaneus (*large straight arrow*) are parallel, rather than divergent. The metatarsals are markedly adducted in relation to the talus. The cuboid ossification center (*curved arrow*) is medially aligned on the end of the calcaneus, rather than in the normal straight alignment. **B:** Maximum dorsiflexion lateral radiograph of clubfoot. The talus and calcaneus are somewhat parallel to each other and plantar-flexed in relation to the tibia. (From the private collection of Vincent S. Mosca, MD).

is currently limited. Arthrography, computerized tomography (138), and magnetic resonance imaging (127, 139, 140) may have a role in research or in the evaluation of postsurgical deformities, but do not have a role in the routine assessment of the idiopathic clubfoot.

Intrauterine Diagnosis. The intrauterine diagnosis of clubfoot has become increasingly frequent with the routine use of fetal ultrasonography during pregnancy. This has implications for the orthopaedist, who is being consulted by prospective parents regarding the diagnosis, possible relationship to syndromes, treatment options, and prognosis. It appears that the earliest that a clubfoot can be diagnosed by ultrasound with accuracy is 12 weeks of gestational age. With sequential studies, there is an increased ability to visualize the deformity, relating either to the progressive development of a clubfoot deformity or perhaps the accuracy with which it can be seen. According to Keret et al. (141), the clubfoot deformity has been detected in routine studies in approximately 60% of cases, which is indicative of some degree of false-negative assessment. In 86% of cases, the deformity is identified by 23 weeks of gestational age, but still others are recognized up to 33 weeks. The diagnosis is made on ultrasound by the fixed position of the foot in an equinovarus position, not deviating from this on sequential observations (Fig. 29-32). Three-dimensional ultrasound may provide a more accurate diagnosis than standard ultrasound studies (142).

In studies of large populations using routine *in utero* ultrasound (143), the recognition of clubfoot deformity varies from 0.1% to 0.4% (144). Because postnatal studies suggest an occurrence rate at birth between 0.1% and 0.6%, one can assume a rather low false-negative rate. The false-positive rate for *in utero* diagnosis of clubfoot using ultrasound varies from 30% to 40%, depending on the series and the criteria (144–147). A term functional false-positive rate has been used in cases in which a foot may have the appearance of remaining in a plantar-flexed, varus, and medially deviated position but can passively be corrected to neutral during exam just following birth. The foot is characterized with a score of 0, 1, or 2 using the Dimeglio classification system and has been classified by some authors as a positional clubfoot. Such a foot requires only parent-administered exercise, and no long-term deformity results. With the advancement of ultrasound and increase in experience, the accuracy of diagnosis will steadily increase.

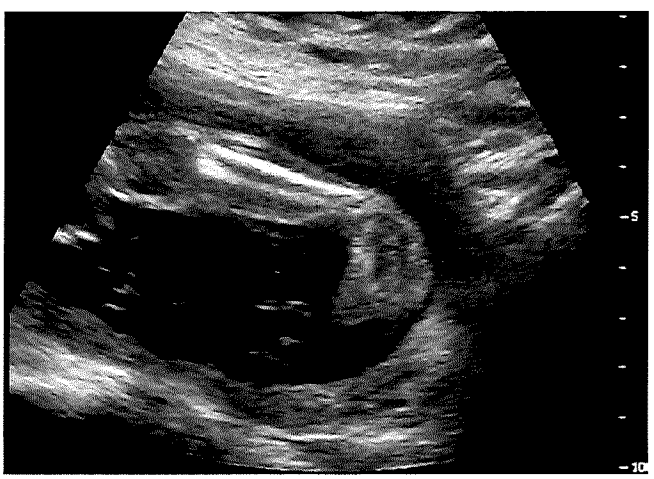


FIGURE 29-32. The clubfoot is diagnosed by ultrasound *in utero* when there is persistent medial deviation and equinus of the foot relative to the tibia. (From the private collection of James R. Kasser, MD.)

The ability to recognize syndromes associated with skeletal malformations is also increasing with time (142, 148). The combination of technologic advances and improved expertise in obtaining and interpreting images will certainly lead to further progress in recognizing fetal structural abnormalities. This brings one to the question of the need for amniocentesis and karyotyping if an isolated clubfoot deformity is found. In 1998, Shipp and Benacerraf (149) and Rijhsinghani et al. (144) suggested that amniocentesis and karyotyping were necessary to identify associated syndromes when clubfoot was identified. Malone et al. (150), in the year 2000, showed that in 57 cases of isolated clubfoot deformity out of 27,000 prenatal exams, there were no unrecognized associated abnormalities. Therefore, the recommendation is that karyotyping not be done in cases where a diagnosis of isolated clubfoot deformity was made. This still appears controversial, and a geneticist should be consulted about the need for amniocentesis if the question arises.

There is no attempt to provide any therapeutic intervention once an intrauterine diagnosis of clubfoot is made. The orthopaedist is only involved in counseling the family about the etiology, treatment, and prognosis, which generally alleviates fears and guilt, dispels myths, allows the parents to make personal decisions concerning the pregnancy, and allows for an improved emotional state for the family during the remainder of the pregnancy and the delivery of their child.

Pathoanatomy. The deformities of the clubfoot are created, in part, by malalignment of the bones at the joints and, in part, by deformation in the shapes of the bones (102, 127, 138–140, 151–160). The neck of the talus is short and deviated plantar-medially on the body of the talus (102, 153, 157–160). This directs the articular cartilage of the head of the talus in the same plantar-medial direction. Anatomic dissections (158) and MRI scan images (139, 140, 153) confirm that there is a varus deformity of the distal end of the calcaneus creating a medial tilt of its articular surface at the calcaneocuboid joint. Howard and Benson (157) and Epeldegui and Delgado (156) found that the anterior facet of the calcaneus is tilted medially in relation to the middle facet in the clubfoot, indicating that the location of the varus deformity is between the two facets (Fig. 29-33). Epeldegui and Delgado (156) performed elegant microdissections of the feet of 75 stillborns, some of which had clubfoot deformities. He specifically studied the bony and soft-tissue anatomy of the talocalcaneonavicular joint, which he, like Sarrafian, called the AP and Scarpa (14) termed the pes acetabulum. The AP is an ellipsoid articular cavity that holds and rotates around the head of the talus, comparable to the relationship between the pelvic acetabulum and the femoral head at the

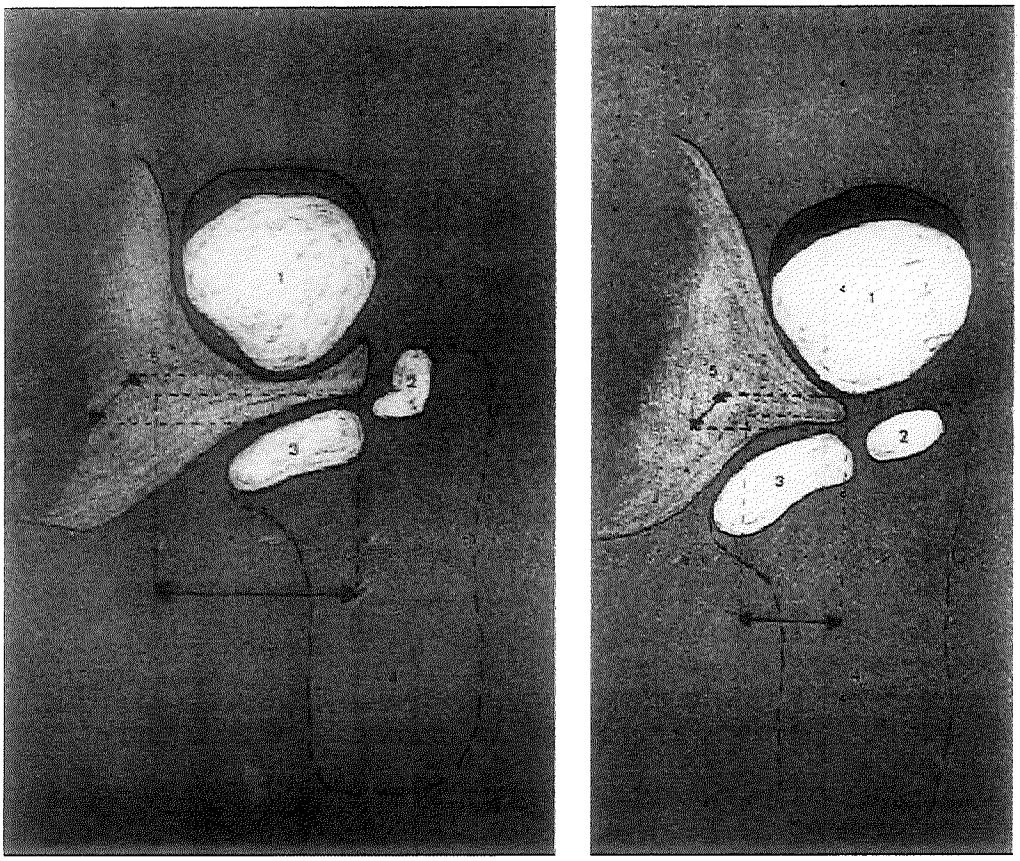


FIGURE 29-33. Navicular (1). The axis of the anterior facet of the calcaneus (2) is tilted medially in relation to the axis of the middle facet (3) in the clubfoot (image on *left*) compared with the normal foot (image on *right*). (From Epeldegui T, Delgado E. Acetabulum pedis. Part I: talocalcaneonavicular joint socket in normal foot. *J Pediatr Orthop B* 1995;4:9, with permission.)

hip joint. Its bony elements are the posterior articular surface of the navicular and the anterior and posterior articular facets of the calcaneus. Epeldegui found that the soft tissues of the AP were likewise markedly different in shape and orientation in the clubfoot from the normal foot. The shape of the medial cuneiform has not been studied in the newborn, but it is trapezoid shaped in the older child with residual forefoot adductus deformity. The subtalar joint complex is severely inverted, a combination of internal rotation and plantar flexion. The axis of rotation is in the interosseus talocalcaneal ligament. The AP is inverted around the plantar-medially-deviated head and neck of the talus, thereby aligning the navicular at or near the medial malleolus. The calcaneus is rotated downward and inward resulting in parallel alignment with the talus in the frontal and sagittal planes. The posterior part of the calcaneus is tethered to the fibula by the calcaneofibular ligament. There is a varus deformity of the distal end of the calcaneus with medial deviation of a congruous calcaneocuboid joint in many clubfeet (78, 139, 140, 153, 155–159, 161). There may be medial subluxation of the cuboid on the distal calcaneus in some feet (152, 162). The plantar fascia, short plantar muscles, and spring ligament are contracted. The Achilles, tibialis posterior, flexor hallucis longus, and flexor digitorum communis tendons are contracted. The posterior capsules of the ankle and talocalcaneal joints are contracted.

Tibial torsion and the position of the talus in the ankle are debated. McKay (154) believes the talus is in neutral alignment, Goldner (163) believes that it is internally rotated, and Carroll (151, 152, 155) believes that it is externally rotated. A recent three-dimensional MRI study of clubfeet revealed an externally rotated position of the talus in clubfoot compared with normal feet (127).

The muscles are abnormal in both anatomical insertion and intrinsic structure (101, 106). Muscles in clubfoot are smaller than normal and there is an increase in intracellular connective tissue within the gastrocsoleus and posterior tibial muscles. A predominance of type I muscle fiber has been seen in posterior and medial muscle groups. Electron microscopic studies have shown loss of myofibrils and atrophic fibers, suggesting a regional neuronal abnormality as well (108).

The ligaments are thick, with increased collagen fibers and increased cellularity (107). This is particularly true of the calcaneonavicular ligament or spring ligament and the posterior tibial tendon sheath (164). An electron microscopic study of medial ligaments in clubfoot identified myofibroblasts, which could be responsible for fibroblastic contracture in the postoperative clubfoot. Fukuhara et al. (165) showed myofibroblast-like cells in the deltoid and spring ligaments. Together, the thickened and shortened ligaments with contractile fibroblasts may

produce a significant component of the clubfoot pathology. Sano et al. (166) confirmed these findings, showing that cells of the medial ligamentous structures contained vimentin uniformly and myofibroblasts in some cases. More recently, Khan et al. (167) were unable to show myofibroblast-like cells in clubfeet, and van der Sluijs and Pruys (168) demonstrated normal collagen cross-linking in clubfeet.

Finally, abnormal vasculature in the foot is frequently present (169). The dorsalis pedis artery in many cases is absent or altered. Katz et al. (170) showed deficient dorsalis pedis flow in 45% of clubfeet compared to 8% of normal controls. In the more severely affected feet requiring surgery, the incidence of dorsalis pedis abnormality was 54%, whereas those successfully treated with cast therapy had an abnormality in dorsalis pedis flow in only 20% of cases. These data suggest that the severity of clubfoot may in some way relate to the vascular abnormality frequently seen in this condition.

Natural History. The untreated clubfoot persists as a rigid, unsightly deformity. A large, callused bursa develops over the dorsolateral aspect of the hyperflexed midfoot which functions as the weight-bearing surface of the foot (Fig. 29-34). In the most extreme cases, the toes point backward during ambulation. Specially made footwear is required to accommodate the deformity. Surprisingly, untreated adults in certain cultures and environments will have little pain for many years and can function adequately. Their function is similar to that of individuals with Syme amputations when not wearing their prostheses. City-dwelling adolescents and adults with untreated clubfoot experience pain and disability with ambulation on paved sidewalks and hard floors.

Treatment of Clubfoot

Nonoperative Treatment. The goal of treatment is to achieve a plantigrade, supple, painless foot that looks normal, although it is not technically normal, and provides good function. There should be good pressure distribution on the skin and no need for special or modified shoe wear. Treatment methods have varied considerably since the deformity was first described by Hippocrates around 2300 years ago. Poor results have been consistently observed following the many aggressive and traumatic operative and nonoperative methods that were employed during the past two centuries, though these techniques dominated the treatment armamentarium until quite recently. Kite (171, 172), in 1939, presented his method of cast correction of clubfoot with a plea for gentle nonoperative management. His method of cast treatment required a lengthy period of immobilization, often >1.5 to 2 years. Ponseti and Smoley (173) developed a casting method for clubfoot in the late 1940s that differed significantly from Kite's, yet it never caught on for reasons that remain unclear. Most orthopaedists during those years attempted to use the method proposed by Kite and were unsuccessful with it. Lovell et al., in 1979, reported long-term follow-up results using Kite's method. In 47 patients with 67 affected feet, the average duration of casting was 20.4 months, while the improvement of 43 feet was rated to be good, 12 feet were rated fair, and 12 feet were rated poor (174). Because of the rather poor results with Kite's method as well as the serial introduction of increasingly popular methods of surgical management (154, 163, 175–181), use of corrective cast treatment was deemphasized through the 1970s, 1980s, and 1990s, that is, all except in Iowa City.

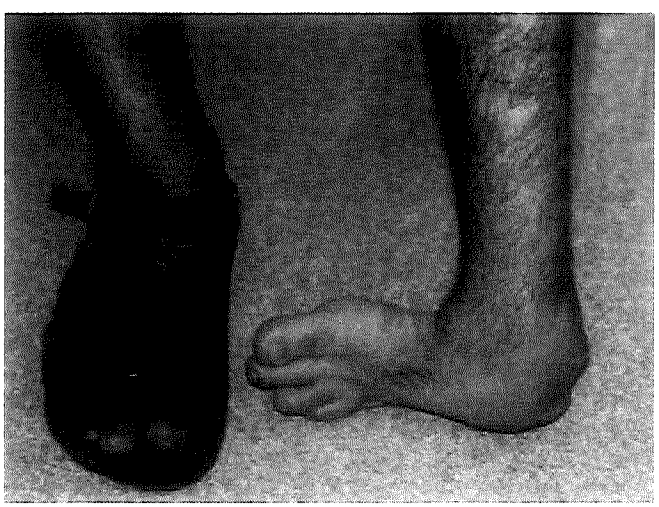


FIGURE 29-34. Untreated clubfoot in an adult migrant farm worker. (From the private collection of Vincent S. Mosca, MD.)

Dr. Ignacio Ponseti, at the University of Iowa, continued to utilize and study the efficacy of his method and to periodically report the excellent results (103, 173, 182, 183), yet it was not until the mid-1990s that the superiority of his method was widely recognized and acknowledged. The publication of his book on this subject in 1996 followed soon after the 1995 publication of the landmark article by Cooper and Dietz (184) in which they reported the only truly long-term results of a single treatment method for clubfoot. They evaluated 45 patients with 71 clubfeet at an average age of 34 years (range 25 to 42 years) who had been treated from infancy using Ponseti's method (185–187). All feet underwent a series of five to eight manipulations and castings at intervals of 5 to 7 days. A percutaneous Achilles tenotomy was performed in the clinic in over 90% of the feet at between 1.5 and 3 months of age after the manipulations and casts, initiated soon after birth, had corrected all but the equinus deformity. Three months of full-time bracing was initiated when the final cast was removed 3 weeks after the Achilles tenotomy. The foot abduction brace (FAB) was continued at nighttime for 3 to 5 years. Somewhat <50% of patients underwent transfer of the tibialis anterior to the lateral cuneiform after walking age, when muscle imbalance on the dorsum of the foot was identified. Cooper and Dietz (184) used pedobarographic and electrogoniometric analyses in addition to clinical and radiographic assessment. With the assessment of pain and functional limitations as the outcome criteria, 35 (78%) of the 45 patients had an excellent or good outcome compared with 82 (85%) of 97 individuals who did not have congenital foot deformity. They reported that excessive weakening of the triceps surae might predispose patients to a poor result and that the final treatment outcome could not be predicted from the

radiographic result. This study, along with subsequent studies from Iowa and elsewhere, has established the Ponseti method as the gold standard for conservative management of clubfoot not only in the United States but around the world.

The basis of Ponseti's treatment (182, 183, 188) was his knowledge of the pathoanatomy of clubfoot deformity based on studies done on affected stillborn infants. The efficacy of manipulation and casting is due to the viscoelastic, or rate-dependent, behavior of the collagen in the ligaments and tendons (189). The creep phenomenon occurs during manipulation. A constant stress (force per unit area), or load, is applied. The tissues undergo deformation (strain), relatively quickly at first and then more slowly until there is little further elongation possible at that time. The cast is applied with the same load used during manipulation. Stress relaxation takes place in the cast. The tissues undergo a small additional amount of elongation in the cast. As soon as that occurs, the stress on the collagen effectively decreases and will not increase again until the next manipulation. Using serial MRI scans during cast correction of infant clubfeet, Pirani et al. (153) showed that there is a third biologic process occurring during cast treatment of clubfeet in infants. The mildly misshapen cartilage anlagen of the bones change to more normal shapes, as if they were clay objects undergoing molding. The ability to correct the clubfoot deformity completely by the Ponseti method depends on the unique stiffness of the tissues of the individual child's foot, the age at which the treatment is initiated, the skill of the practitioner, and the definition of complete.

Manipulation and casting should begin as early as is feasible. There is no evidence that a delay of several days makes a difference in the rate of success. In fact, anecdotal reports suggest that initiation of this method within several months of birth leads to outcomes very similar to those in feet treated from birth. And a study by Lourenco and Morcuende (190) indicates that it is worthwhile to attempt Ponseti management even as late as 9 years of age in neglected cases. The technique of manipulation and casting described by Ponseti (123, 173, 182, 183, 188) should be studied in detail and with supervision and practiced regularly. The manipulations and cast moldings are gentle above all else, a concept actually first espoused by Hippocrates.

Although plaster of Paris casts are the old standard, semirigid fiberglass has been shown by Coss and Hennrikus (193) to be statistically superior to plaster of Paris in its durability, convenience, performance, and ease of removal. Proper molding of fiberglass casts is a bit more challenging than plaster of Paris, but the technique can be learned easily, so that there is no difference in efficacy or efficiency between the two materials. The parents can remove the casts and bathe the child immediately before returning to clinic for remanipulation and casting, which should occur every 5 to 7 days until no further improvement is seen. If good correction is not achieved by 3 months of casting, it is unlikely that further nonoperative treatment will be successful (Figs. 29-35 to 29-40).

Full correction of the cavus, adductus, and varus deformities using the Ponseti method of manipulation and casting is achievable in approximately 90% of cases. Full cast-correction of the equinus deformity, with the achievement of at least 10 degrees of ankle dorsiflexion, is achievable in <10% of those cases. For the others, a percutaneous complete transection of the Achilles tendon is required to achieve this position (Fig. 29-41).

The tenotomy can be performed in the clinic or the operating room, based on local health care facility considerations and regulations. The final cast following tenotomy is left in place for 3 weeks. This is the time it takes for infants to reconstitute a sound and strong Achilles tendon that will go on to look and perform normally, as has been documented both by clinical experience and by observation at the time of revision surgery. In this final cast, the foot is held in a position of 15 to 20 degrees of dorsiflexion with 70 to 75 degrees of external rotation of the foot relative to the thigh. This ends the active phase of treatment, better defined as the phase that the orthopaedist controls.

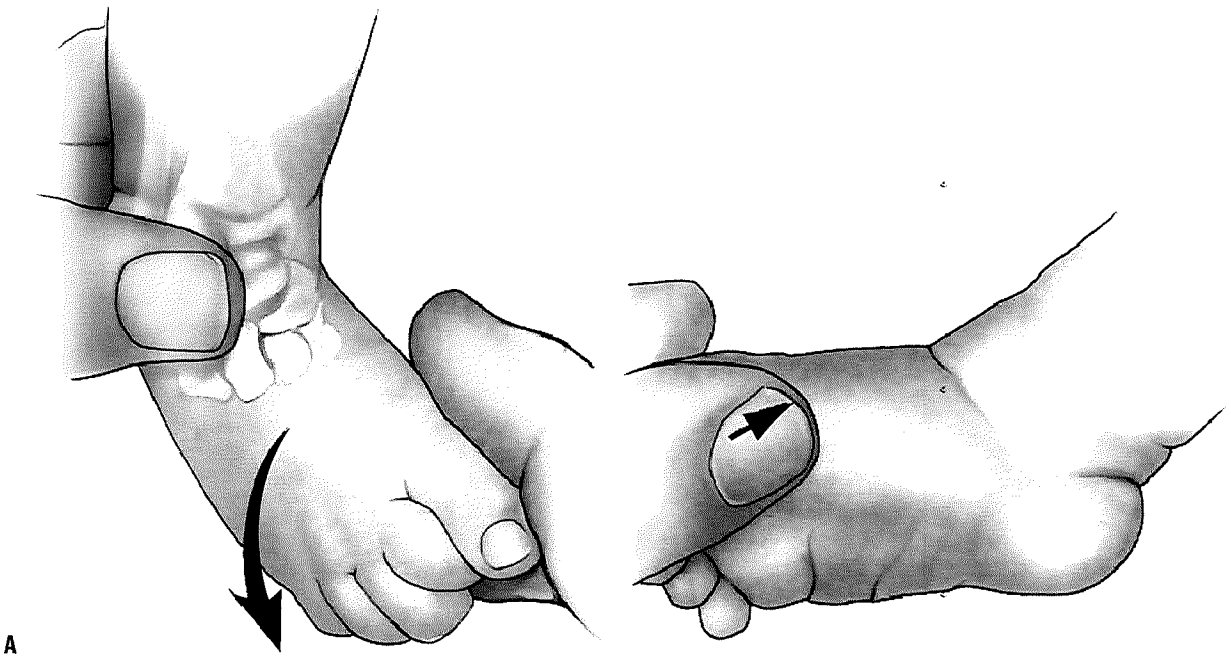
After removal of the final cast, the maintenance phase of treatment begins, the phase perhaps better defined as that which the parents control and the orthopaedist monitors. The hallmark of this phase is the FAB (Fig. 29-42).

There are now many makes and models, but the basic design is a pair of semirigid shoes connected together by a bar. The corrected clubfoot is held by the shoe in a controlled fashion maintaining a straight lateral foot border and slight valgus of the hindfoot. The shoes are connected to the bar with 70 degrees of external foot rotation (45 degrees for a contralateral normal foot), and the bar is bent with its apex facing away from the child to create slight dorsiflexion of the ankles. Dorsiflexion cannot be achieved adequately without bracing the foot in a position of severe external rotation and abduction. Padding in the shoe proximal to the heel can prevent the foot from rising out of the shoe and can help to provide relief for the calcaneus so that blistering does not occur. There is rarely a problem with compliance for the first 3 months during which the FAB is worn by the child 23 hours per day, but maintaining nighttime brace usage (12 out of 24 hours per day) for 4 years is quite challenging. Ponseti (182) believed that in the absence of a good bracing program, there is up to a 70% rate of recurrence of the clubfoot deformity. In such cases, it is difficult to reinstitute this method of management, and surgical treatment might be required. Therefore, parent education is important and should start with the first encounter with the orthopaedist to ensure that parents are knowledgeable about their role in the treatment plan long before it begins. This author also asks the parents to perform ankle dorsiflexion stretching exercises for at least 1 minute at least three times per day, because equinus is not only the last deformity to achieve correction, but is also the first to recur.

A number of individuals have tried to reproduce the results achieved by Ponseti using his method. Herzenberg et al. (194), in 2002, compared a group of 27 patients treated with the Ponseti method to 27 patients treated with traditional casting techniques. One of the patients in the Ponseti group required posteromedial release, whereas 91% of patients in Herzenberg's control group required surgical treatment. Foot abduction bracing with a Denis-Browne bar was crucial to the success of this method of treatment, just as Ponseti

Text continued on page 1424

Cast Treatment for Congenital Clubfoot: The Ponseti Method (Figs. 29-35 to 29-40)



A B

FIGURE 29-35. Cast Treatment for Congenital Clubfoot: The Ponseti Method. A, B: When beginning the Ponseti clubfoot manipulation technique, the position of the bones of the foot is first identified in relation to the medial and lateral malleoli and the head of the talus. Although the whole foot is in extreme supination, the forefoot is pronated in relation to the hindfoot. This creates the cavus deformity that is manifest by the first metatarsal being more plantarflexed than the lateral metatarsals. The first manipulation strives to correct the cavus deformity by supinating the forefoot and dorsiflexing the first metatarsal. This maneuver stretches the plantar fascia and "unlocks" the subtalar joint. Counterpressure is applied to the dorsolateral aspect of the head of the talus with the thumb of the other hand. The third point of pressure and stabilization is the medial malleolus, not the calcaneus. Supinating the forefoot places the forefoot in proper rotational alignment with the hindfoot, that is, matched supination of the hindfoot and the forefoot. Since the cavus is usually not a fixed deformity at birth, correction often occurs with the first cast. Correction of severe cavus in a stiff foot will need two or three cast changes with the forefoot in forced supination.