performed. Wrist fusion is not performed in young patients because this leads to loss of wrist motion and potential loss of ulnar physeal growth. However, Catagni et al. (228) performed wrist fusion in conjunction with distraction lengthening in adolescent and young adult patients with recurrent deformity.

Generally, wrist reconstruction is performed before thumb reconstruction. Ideally, thumb reconstruction should be performed before the child is 18 months of age because the learning ability for the pinch movement becomes limited once the central nervous system matures to this stage. In mild forms of radial dysplasia, the thumb hypoplasia causes functional problems involving decreased first web space, MCP joint instability, and weak thenar muscles. The first web space can be deepened with Z-plasties or rotation flaps (274, 275). Release of adductor and first dorsal interosseous fascia is often necessary. The MCP joint can be stabilized with local fascia or use of extra flexor digitorum superficialis tendon length for ligament reconstruction. On occasion, MCP joint chondrodesis (fusion of the proximal phalanx epiphysis to the metacarpal head) or arthrodesis is appropriate. Opponensplasty is performed simultaneously with use of the abductor digiti quinti (276), ring-finger flexor digitorum superficialis, or accessory digital extensors. All have had reported success in providing opposition strength. Thumb aplasia is best addressed with pollicization (275, 277). Toe-to-thumb microvascular transfers have been reported, but to date the results are less successful than those of index-finger pollicization. Overall, the quality of the index-finger donor determines the quality of the subsequent thumb. If there is significant camptodactyly, the thumb will be stiffer, weaker, and less often used in pinch activities than if the index has full passive mobility and intrinsic and extrinsic strength. In a well-performed pollicization, the results are functionally and cosmetically pleasing to the patient, family, and surgeon.

Complications. Recurrent deformity and premature closure of the distal ulnar physis are the two major complications of wrist reconstruction (278–280).The occurrence of these problems depends on the procedure performed (centralization versus radialization) and the quality of the preoperative musculoskeletal and soft-tissue anatomy. With radialization, the goal is to dynamically rebalance the wrist and maintain motion. If this fails to occur, radial deviation and flexion deformity will recur with growth. In addition, if there is limited elbow flexion, excessive flexion and radial deviation of the wrist will be used by the patient to compensate while carrying out activities of daily living such as oral hygiene and feeding. This contributes to the recurrence rate.

Physeal arrest is more common with centralization procedures. The forearm is already foreshortened, and this is further exacerbated by loss of distal growth. Because 70% to 80% of forearm growth comes from the distal physis, postoperative growth arrest is a major aesthetic and functional problem (281).

Finally, in patients with radial dysplasia, pollicization procedures can have poorer results in terms of opposition strength and active range of motion (282). The opposition weakness may be improved by opponensplasty transfer (283, 284), but there should be a strong donor if the procedure is to succeed. Otherwise, the patient will continue to compensate with lateral digital pinch on the ulnar side of the hand.

Ulnar Dysplasia/Ulnar Clubhand. Ulnar, or postaxial, longitudinal deficiency is less common than either radial or central longitudinal deficiency. It is classified as a failure of formation of parts. The incidence was found by Birch-Jensen to be 1 in 100,000 live births (240). Ogden et al. (185) cited a male-to-female ratio of 3:2, with only 25% of the patients showing bilateral involvement.

Most cases are sporadic, but there are reports of familial occurrence (285–288). It also occurs as a part of rare, identified, inheritable syndromes, such as ulnar mammary (Schnitzel) syndrome, Klippel-Feil syndrome (289–291), and some nongenetic syndromes such as Cornelia de Lange syndrome. It is associated with musculoskeletal system malformations in up to 50% of cases. Contralateral upper extremity deficiencies of phocomelia, transverse arrest, radial deficiency, and aphalangia occur commonly. Similarly, lower extremity deficiencies, such as proximal femoral focal and fibular deficiencies, occur in almost one-half of the cases. Unlike those with radial dysplasia, it is uncommon for patients with ulnar deficiency to have associated major organ system malformations. Ogino and Kato's experimental data may explain this finding (291). They produced major deficiencies in rat fetuses by injecting the mothers with the antimetabolite Myleran. The timing of injection during the gestational period determined the limb malformation produced. For example, ulnar deficiencies were produced by earlier injections than were radial deficiencies. Fetuses that had ulnar deficiencies had more lethal cardiac malformations. This may explain why there are fewer major organ system malformations and a lower incidence of ulnar deficiency among live births.

Clinical Features. Bayne classified ulnar deficiency into four groups based on the musculoskeletal abnormalities of the elbow and forearm (292) (Fig. 22-32). Most clinicians use this system to define and establish treatment plans for these patients. Type I deficiency is hypoplasia of the ulna. Both distal and proximal physes of the ulna are present, but decreased in growth. There is minimal, nonprogressive bowing of the radius, and a variable presentation of hand malformations. Type II deficiency is the most common type and involves partial absence of the ulna. There is a fibrous anlage extending from the distal ulna to the carpus. The hand is ulnarly deviated, with bowing of the radius, and these deformities may be progressive with growth. The elbow is stable if there is sufficient proximal ulna present. Again, digital malformations or absences are variable. Type III deficiency involves complete absence of the ulna. There is no ulnar anlage. The radius, wrist, and hand are usually straight. The elbow is unstable as a result of the lack of an olecranon. Hand malformations and absences are common. Type IV deficiency involves synostosis of the distal humerus to the proximal radius. There is an ulnar anlage present from the distal humerus to the carpus, with marked bowing of the radius and ulnar deviation of the hand. Hand anomalies are common also in type IV deformities.

In addition, in these patients the limb is foreshortened and usually internally rotated. The glenoid may be dysplastic. The radial head is often dislocated, and range of motion of the elbow is limited in up to 40% of cases (295). These abnormalities make placement of the hand in space difficult. The hand malformations limit pinch, grasp, and release functions. Reconstructive surgery is indicated for improving hand and wrist orientation, thumb opposition, and digital motion and strength.

Treatment. There is a scarcity of data regarding the natural history of untreated ulnar dysplasia. In 1927, Southwood stated, "From the functional viewpoint, therefore, the deformed limb is much more useful than its anatomical condition would lead one to expect" (296). This malformation is not associated with central nervous system deficiencies. As with all congenital malformations in individuals with normal brains, the patients will perform activities well, but differently. Treatment has to improve function and aesthetics, if it is to be warranted.

Nonsurgical treatment has predominantly involved physical therapy and corrective casting or splinting. In type I and III deficiencies, the mild ulnar deviation of the wrist and hand may be improved with serial casting, splinting, and passive exercises starting in infancy. In type II and IV deficiencies, the ulnar anlage may make nonsurgical correction of the severe ulnar deviation of the hand and wrist impossible.

There is considerable debate regarding the treatment of the hand that is severely ulnarly deviated and that does not respond to casting/splinting. There is limited information to allow for critical evaluation of the options of (a) leaving the patient alone, (b) performing excision of the ulnar anlage, and (c) corrective radial osteotomy. Some of the confusion exists because not all of these deformities are progressive (297). As Flatt (1) makes clear, it is difficult to critically evaluate the literature because of limited objective measurements in previous studies. He correctly points to the low incidence of this disorder as hampering objective assessment of the therapeutic options. As with many rare conditions, only multicenter, prospective studies can definitively answer the questions.

The lack of this information allows for subjective interpretation of the treatment options, leading to reluctance to pursue aggressive surgical intervention. Within these limits, an attempt is made to outline treatment options and recommendations for wrist deformity, elbow instability, and digital and thumb deficiencies.

Resection of the ulnar anlage is indicated for progressive ulnar deviation of the wrist and hand of >30 degrees. This can occur in type II and IV deficiencies (1, 298). Through an ulna-based incision, the anlage is identified as it inserts into the carpus. The ulnar artery and nerve should be protected. Resection should be performed until neutral positioning of the wrist can occur intraoperatively. If there is associated marked ulnar deviation of the radius, concomitant radial osteotomy can be performed. However, it is imperative to assess the location of the radial head and the status of forearm rotation before proceeding with anlage excision and consideration of radial osteotomy.

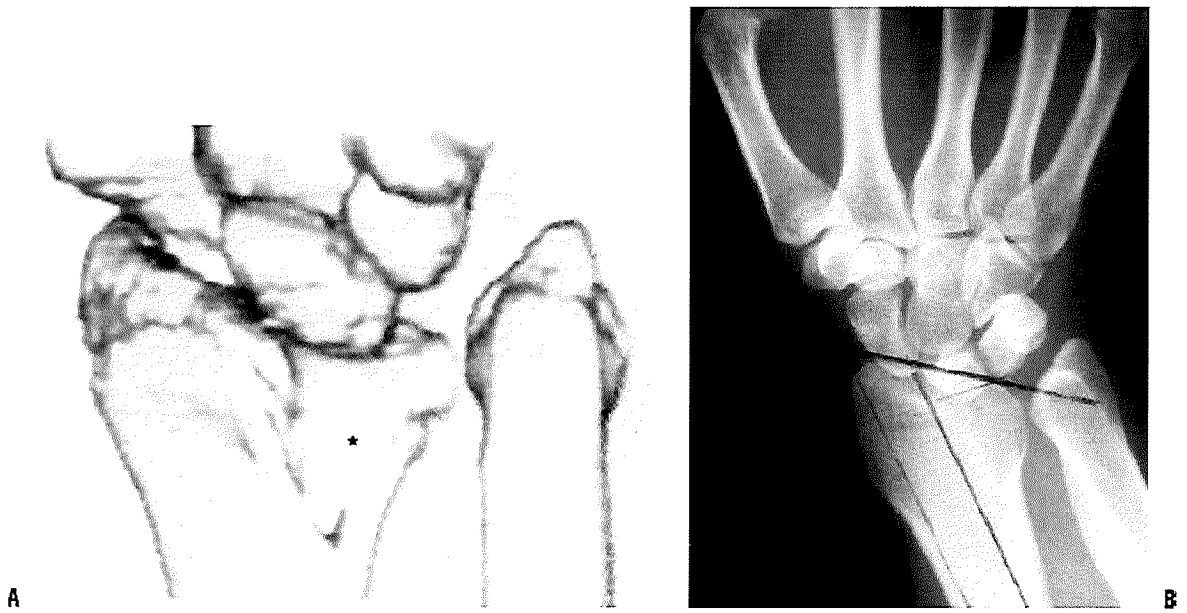
If there is a dislocated radial head and limited forearm rotation preoperatively in the type II deformity, anlage excision, resection of the radial head, and creation of a single-bone forearm should be carried out simultaneously. If there is acceptable forearm rotation preoperatively, it is best to correct only the wrist deformity and to monitor the status of the forearm and elbow with growth. Resection of the radial head for cosmetic reasons should be performed cautiously because even the dislocated head may be providing some elbow stability in these patients.

Similarly, creation of a single-bone forearm may result in improved cosmesis, but the loss of forearm rotation may cause an unacceptable loss of function. In patients with type IV deficiency, there may be associated internal rotation posture to the arm that limits placement of the hand in space. If this is present, simultaneous external rotation osteotomy of the limb and ulnar anlage excision should be performed. This is clearly the case with patients with bilateral deformity who are unable to reach their mouths preoperatively.

Repair of digital and thumb deficiencies is indicated. Syndactyly is common and should be corrected in infancy. Thumb hypoplasia or absence should also be repaired in infancy. Broudy and Smith (295) described a modified pollicization procedure for the malpositioned thumb in the plane of motion of the other fingers. Tendon transfers for intrinsic and extrinsic muscle deficiencies of the thumb and fingers are indicated if there are adequate donors available.

Madelung Deformity. Madelung (299), in 1878, described a growth deformity of the distal radius. For reasons that are still unknown, the volar, ulnar aspect of the distal radial physis slows or stops growing prematurely. The continued normal growth of both the ulnar physis and the remaining dorsal, radial aspect of the radial physis results in ulnar overgrowth, carpal subluxation, and radial articular deformity (Fig. 22-33). Madelung deformity usually occurs in girls and is most often bilateral (300). It may not become clinically apparent until the adolescent growth spurt, which is when most patients present. It is generally sporadic in presentation. It is also associated with Leri-Weill syndrome, a dyschondrosteosis form of mesomelic dwarfism that is inherited in an autosomal dominant manner due to mutation in the SHOX gene (298, 301, 302). In addition, Madelung deformity has been associated with Hurler mucopolysaccharidosis, Turner syndrome, osteochondromatosis, achondroplasia, and Ollier disease (303). True Madelung deformity should be distinguished from a posttraumatic or postinfectious physeal arrest.

Clinical and Radiographic Feature. The clinical and radiographic picture is dependent on the age at presentation and the severity of the growth arrest (304). Generally, by the time the affected children are brought for treatment, there is marked deformity, limitation of motion, and activity-related pain. Because the condition is usually bilateral, the subtle growth deformity that occurs before the adolescent growth spurt is often ignored. However, with early presentation there is a slight positive ulnar variance and loss of the volar, ulnar aspect



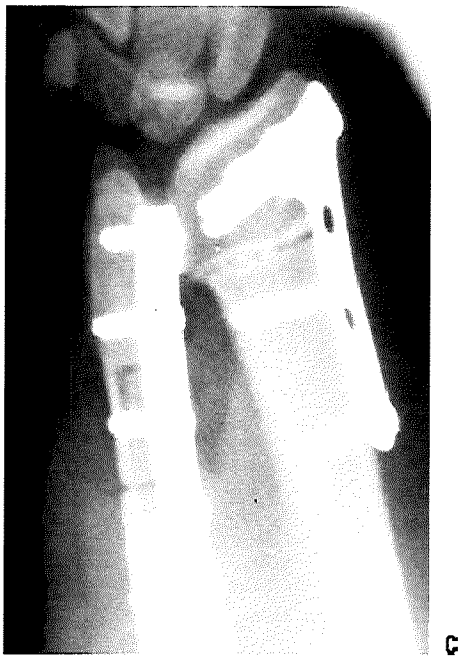


FIGURE 22-33. **A:** CT reconstruction of severe Madelung deformity. Note that the lunate fossa (*) is markedly deficient and oriented ulnarly. **B:** Preoperative three-dimensional CT of a patient with Madelung deformity and debilitating ulnocarpal and radioulnar pain. There is nearly complete deficiency of the lunate fossa and subluxation of the carpus ulnarly, volarly, and proximally. The ulna has a positive variance. (Figure courtesy of Children's Orthopaedic Surgical Foundation (COSF), © 2010.) **C:** Postoperative radiograph of a radiodorsal closing-wedge osteotomy of the radius and ulnar Z-shortening osteotomy in this patient. This procedure restored radial articular alignment, corrected the ulnocarpal impaction, and reduced the distal radioulnar joint. (Figure courtesy of Children's Orthopaedic Surgical Foundation (COSF), © 2010.)

of the radial lunate fossa (Fig. 23-33). The carpus subluxates volarly and into the gap between the radius and the ulna. These patients may have mild symptoms of ulnocarpal impaction with power grip activities, and distal radioulnar joint incongruity with forearm rotation. More often, individuals with Madelung deformity present late with marked deformity. There is an increased tilt of the radial articular surface from the dorsal radial corner of the styloid to the volar, ulnar aspect of the depleted lunate fossa (305). The ulnar variance is more positive, with carpal overlap and dorsal subluxation. The carpus migrates more proximal into the increasing diastasis between the radius and the ulna on anteroposterior radiographs. These patients have more pain and limitation of motion, especially forearm rotation and wrist extension.

Pathoanatomy. The skeletal features are well described. As mentioned previously, the arrest of the volar, radial aspect of the distal radial physis causes subsequent deformity of the radiocarpal, radioulnar, and ulnocarpal joints. Vickers and Nielsen (306), Linscheid (307), and Ezaki (300) have described abnormal tethering of soft tissues from the distal radius to the carpus and the ulna. These have included aberrant ligaments (303, 306) and pronator quadratus muscle insertions (307). It is unclear whether these structures are responsible for or a consequence of the growth deformity of the radius. Vickers and Nielsen's successful treatment of Madelung deformity by excision of the volar tethering soft tissues and prophylactic physiolysis of the volar radial physis indicates that there may be a causal relationship.

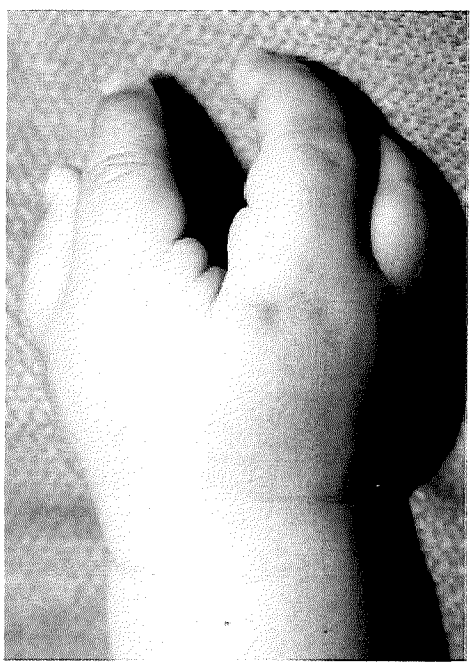
Treatment. The early descriptions of the treatment of Madelung deformity advocated treatment only for symptomatic patients at skeletal maturity (308). Originally, the mere presence of the deformity was not an indication for operative intervention in the asymptomatic patient, regardless of age. However, the growth discrepancy is easier to treat if it is addressed early. Young patients become symptomatic and the range of function of the limbs becomes restricted with increasing growth deformity. Vickers and Nielsen (306) advocated early intervention with physiolysis. The treatment principle is similar to that for Blount disease, with resection of the abnormal volar, ulnar physeal region of the radius and fat interposition. At the same time, any aberrant, tethering anatomic structures are excised. Their case series indicates restoration of radial growth and prevention of progressive deformity. Some patients with Madelung deformity can present at a very young age with marked deformity and complete lack of a lunate fossa for carpal support. In these patients early radial dome osteotomy and ulnar shortening is warranted but there is a high rate of recurrent deformity with growth. An alternative treatment for the patient presenting early is to perform ulnar and radial epiphysiodesis at the same time as corrective osteotomies in order to prevent progressive or recurrent deformity and symptoms. Later length issues may or may not need to be addressed. In the patient with bilateral disease, this treatment leads to foreshortened upper limbs without side-to-side discrepancy.

The treatment for a patient presenting late with marked deformity and symptoms is more common. The radial deformity can be addressed by an osteotomy. Techniques described include a dome osteotomy, dorsal radial closing-wedge osteotomy, or volar opening-wedge radial osteotomy and bone grafting (309). Dome osteotomy is preferred. The ulnar positive variance may be corrected by the dome osteotomy. If not, an ulnar-shortening procedure is required (310). Alternative methods of ulnar shortening include resection of the distal ulna and a Sauve-Kapandji procedure. However, these are not usually performed in the young unless there is already significant deterioration of the articular cartilage, wrist ligaments, or triangular fibrocartilage.

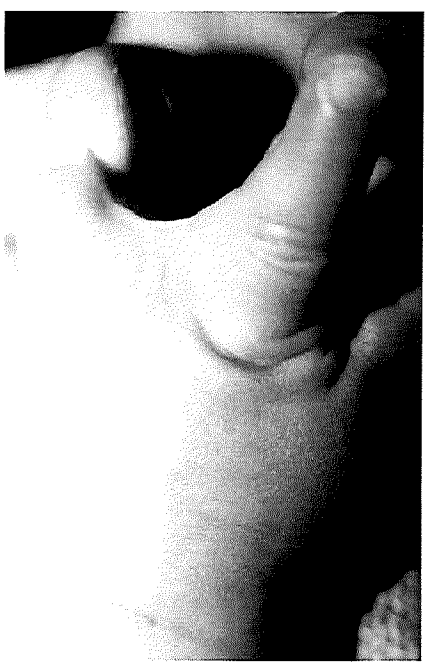
HAND REGION

Congenital

Cleft Hand and Symbrachydactyly. Central defects of the hand have been described in the past as typical or atypical (311). Since 1992, the International Federation of Societies of Surgery of the Hand has classified typical cleft hands as *cleft hands* and atypical cleft hands as part of *symbrachydactyly*. Cleft hands represent a partial or complete longitudinal deficiency in the central portion of the hand (Fig 22-34). The elbow, forearm, and wrist are usually normal. There are often ulnar and radial-sided syndactylies and digital hypoplasia. Cleft hands often occur in conjunction with cleft feet. In that situation, there is an autosomal dominant inheritance pattern, mediated by the SHFM genes. However, the penetrance is variable, with approximately one-third of the known carriers of the gene having no malformations (312, 313). In addition, the phenotypic expression in affected individuals is variable. Cleft hands are also associated with other syndromes and malformations such as cleft lip/palate (ectrodactyly, ectodermal dysplasia, cleft syndrome), other craniofacial syndromes, Cornelia de Lange syndrome, congenital heart disease, ocular malformations, and imperforate anus (313, 314). The incidence is estimated at between 1 in 90,000 and 1 in 100,000 live births (240, 315, 316). Various classification schemes have been used. Most have focused on the nature of the deformities (317, 318).



A



B

FIGURE 22-34. **A:** Cleft hand with absent middle ray. **B:** Incomplete syndactyly of the first web space in the same patient. Closure of the cleft must include deepening of the first web space to maintain maximum hand function. (Figures courtesy of Children's Orthopaedic Surgical Foundation (COSF), © 2010.)

Manske and Halikis (319) proposed a classification system of cleft hands based on the thumb and the first web space. This scheme aids the surgeon in surgical reconstruction decisions and therefore may be the most useful classification.

Symbrachydactyly is defined by unilateral central digital deficiencies and simple syndactylies. It is a sporadic event without genetic inheritance. There are no associated anomalies. The feet are normal. It is a unilateral process, often with multiple absent digital rays. There are often finger nubbins present, which is a situation not seen in cleft hands. Symbrachydactyly is a transverse deficiency that may or may not be a separate entity from transverse absence of digits (313). These entities are distinct from the amputations associated with constriction band syndrome. Symbrachydactyly should be viewed as a clinical entity distinct from cleft hand, with a very different treatment plan.

Treatment

Cleft Hands. As Flatt (1) poignantly stated, "The cleft hand is a functional triumph and a social disaster." The wide central cleft allows for outstanding grasp, release, and pinch functions. Sensibility is normal. The cleft hand, therefore, usually functions without limits.

Treatment of the cleft hand centers on closure of the cleft. However, surgical closure of the cleft must be accompanied by appropriate treatment of the first web space and thumb to avoid functional compromise. The skin flaps designed for cleft closure, however, must take into account the status of the first web space. If the first web space is normal or mildly narrowed (Manske types I and IIA), simple cleft closure, such as with a Barsky flap (311), can be performed. If necessary, a simultaneous but separate Z-plasty widening of the first web space can be performed (Manske types IIA and IIB). If the first web space has a marked syndactyly (Manske type III), the flap designs use the redundant skin of the cleft closure to create a first web space. The adduction contracture of the thumb is released, and the index ray is transposed ulnarly at the same time. The Snow-Littler, Ueba, and Miura flaps (320–322) all involve transposition of the cleft skin to the first web space, with simultaneous transposition of the index ray ulnarly. If there is a transverse bone across the cleft, this must be removed in order to prevent progressive deformity. Often there is a conjoined flexor and extensor across the base of the cleft that has to be released. Sometimes, carpal closing-wedge osteotomy is necessary to close the cleft. In addition, the stability of the index- and ring-finger MCP joints should be maintained or restored. Associated fourth web space syndactylies are separated with Z-plasties and skin grafts. There may be associated camptodactyly or clinodactyly of the adjacent digits requiring corrective splinting or surgery.

Symbrachydactyly. The treatment of symbrachydactyly in the United States is probably the most individualized of that of any of the congenital malformations. The range of options include (a) leaving the child alone, (b) nonvascularized transfers to the soft-tissue nubbins of the phalanges (323, 324), (c) microvascular toe transfer(s) (268, 325–328), (d) web-space deepening, (e) digital distraction lengthening or bone grafting (329), and (f) use of a prosthesis. In addition, families and patients are very interested in the possibilities of transplantation and laboratory cellular growth of digits. There is no definitive answer at present. The choice is greatly influenced by the family's desires and the surgeon's experience and biases. There are few peer-reviewed published studies regarding functional and cosmetic outcomes that would guide the decision more objectively. However, there are clear principles to help guide all parties as to the best choice for them.

The primary goal is to improve pinch. In the presence of a normal thumb and web space, all of the choices for treatment will work. In this situation, treatment options focus on the quality of the other digital rays. If the soft-tissue pockets of the digits are adequate, nonvascularized transfer of the proximal phalanx of the toes is a very good choice. Although it will not provide normal digital length, it will provide stability for lateral pinch. This must be performed before 18 months of age and include the periosteum and collateral ligaments (323, 330). The proximal phalanx is harvested through an extensor-tendon-splitting dorsal approach. The proximal phalanx is harvested extraperiosteally, while protecting the neighboring tendons and neurovascular structures. At the metatarsophalangeal joint, the collateral ligaments, dorsal capsule, and volar plate are detached proximally from the metatarsal, while leaving intact their distal attachments to the phalanx. At the PIP joint, those soft tissues are left attached to the middle phalanx. With transfer to the hand, the proximal soft tissues of the toe phalanx are sutured to the corresponding soft tissues of the recipient site. The best results for phalangeal survival and growth are realized when this procedure is performed before 1 year of age. The quality of the soft-tissue pocket clearly affects the outcome. Multiple phalangeal transfers can be performed simultaneously. In the presence of a normal thumb and first web, digital lengthening is another option. In addition, digital lengthening has been performed successfully after nonvascularized toe phalangeal transfer (329). Finally, prosthetic use has been tried. The major problems with prostheses are that children function as well or better without them because the prostheses are insensate and at times cumbersome. In the adolescent and adult, a cosmetic prosthesis may be used for social reasons (317). It should be noted that the finest aesthetic prostheses are very expensive.

If there is a deficient first web space, deepening of the web with release of the adduction contracture is appropriate. At times, this may require resection or transfer of the index metacarpal in order to achieve a useful web for pinch and grasp functions. If there is absence of the thumb, then digital transposition or microvascular transfer is indicated.

Microvascular toe transfer should be performed only if the patient is a child older than 2 years; the family is well informed about all aspects of the surgery and possible outcomes; there are proximal nerves, vessels, tendons, and muscles available for creating a viable and functional transfer; there is carpal or metacarpal support for the transfer; and there is an experienced surgical team (291). Unfortunately, although this procedure is being performed more commonly nowadays, objective data regarding functional, cosmetic, and psychologic outcomes are still minimal in relation to children.