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.

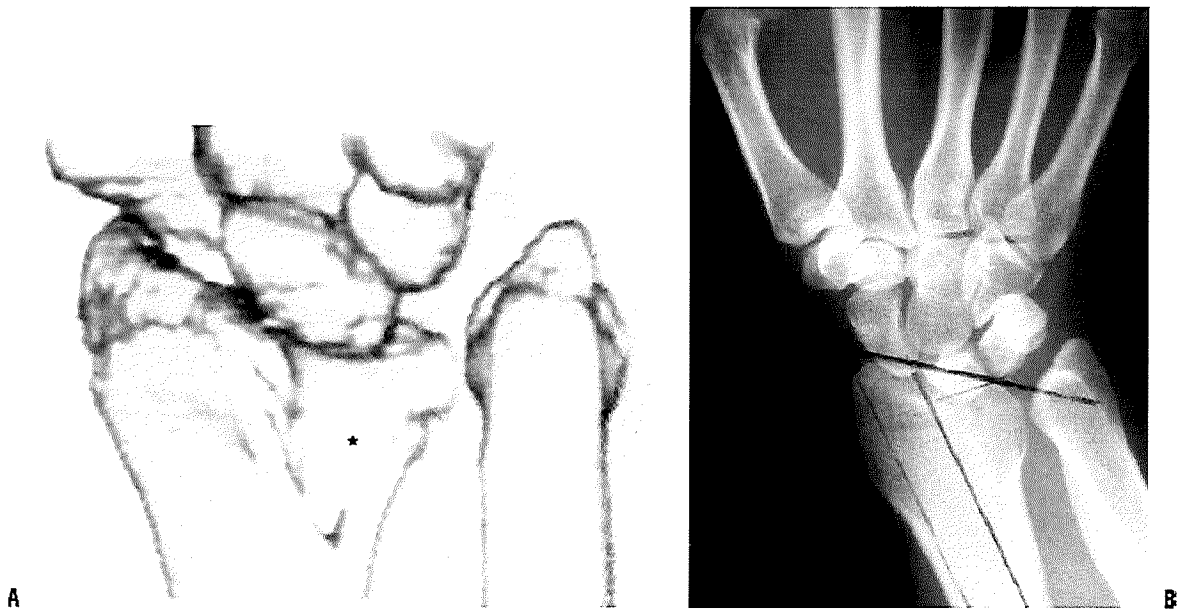
If there is a dislocated radial head and limited forearm rotation preoperatively in the type II deformity, anlage excision, resection of the radial head, and creation of a single-bone forearm should be carried out simultaneously. If there is acceptable forearm rotation preoperatively, it is best to correct 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



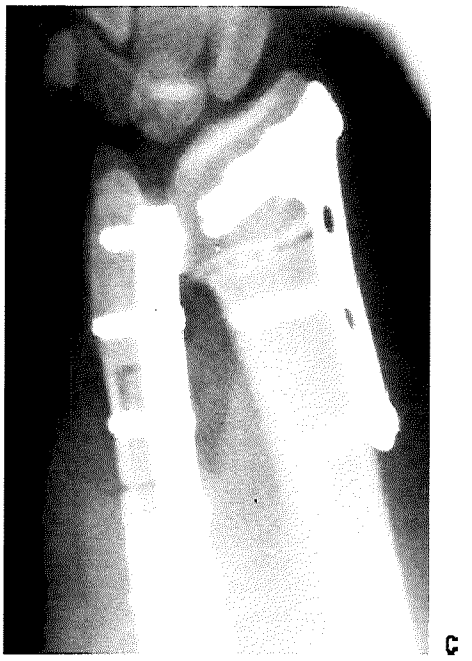


FIGURE 22-33. **A:** CT reconstruction of severe Madelung deformity. Note that the lunate fossa (*) is markedly deficient and oriented ulnarly. **B:** Preoperative three-dimensional CT of a patient with Madelung deformity and debilitating ulnocarpal and radioulnar pain. There is nearly complete deficiency 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.

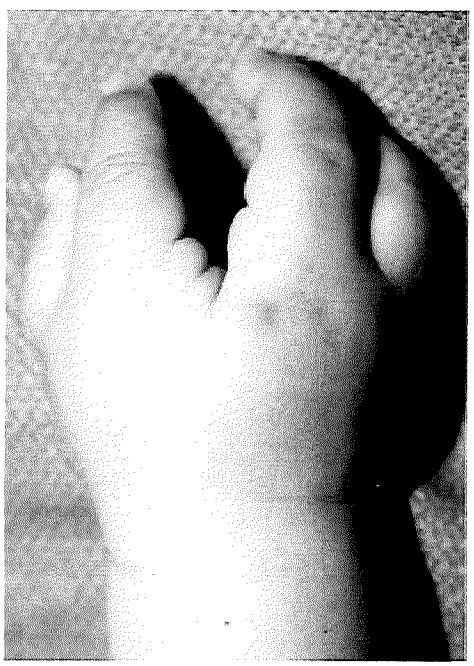
Treatment. The early descriptions of the treatment of Madelung deformity advocated treatment only for symptomatic patients at skeletal maturity (308). Originally, the mere presence of the deformity was not an indication for operative intervention in the asymptomatic patient, regardless of age. However, 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.

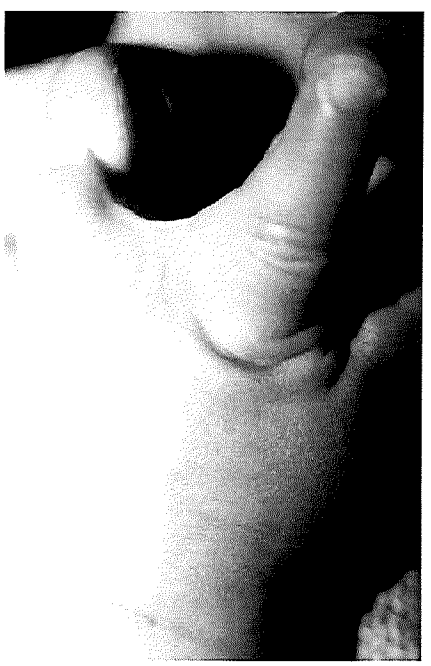
HAND REGION

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.

Constriction Band Syndrome. *Constriction band syndrome,* also known as *amnionic band syndrome,* or *amnion disruption sequence* is likely the result of disruption of the inner placental wall, the amnion. This early amnionic rupture often results in oligohydramnios and amnionic bands. The fibrous bands from the amnionic wall wrap around the digits, causing constricting digital bands, amputations, and syndactylies (313, 331–333). Streeter (334) was the first to propose that this syndrome is a mechanical deformation rather than a malformation.

There is no inheritance pattern. This syndrome occurs in approximately 1 in 15,000 live births (335). It is associated with other musculoskeletal deformations in 50% of cases, the most common being clubfeet. There may be devastating cleft lip and facial deformations as a result of deforming amnionic bands. There are no associated major organ system malformations.

In the hand, the ring finger is most frequently affected. This may be because in a clenched fist, the ring finger is the longest. The band may merely cause an indentation. However, if it is circumferential, the constriction ring may lead to distal edema or cyanosis. Intrauterine amputations are the result of vascular insufficiency caused by the tourniquet-like bands (335). At times, this can be noted at birth with a necrotic or severely compromised phalanx distal to a constricting band. Syndactyly occurs when the bands attach adjacent digits. There are often skin clefts proximal to the syndactyly, indicating the embryonic formation of a web space before the amnionic rupture and subsequent deformation. The extremity proximal to the band is normal. The development of the underlying tendons, nerves, vessels, and muscles is also normal.

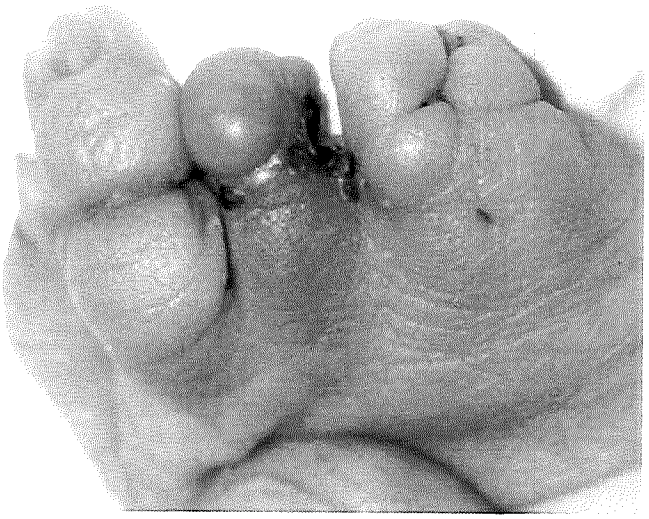


FIGURE 22-35. An amnionic band causing digital ischemia in a neonate in the newborn nursery. This condition is rare and requires immediate removal of the band in order to prevent further soft-tissue digital loss. (Figure courtesy of Children's Orthopaedic Surgical Foundation (COSF), © 2010.)

Clinical Treatment. Impending tissue necrosis is an indication for emergent removal of the band to relieve vascular compromise. This is a rare situation, usually seen only in the neonatal period (Fig. 22-35). Removal of neonatal constricting bands that are causing vascular compromise can generally be performed outside the operating room. The band will literally unravel or debride like an eschar. Improved venous drainage is almost immediate. With recent advances in prenatal diagnosis and less invasive fetal surgery, there have even been reports of fetoscopic constriction band release (336, 337). While these advances hold promise, at present prenatal intervention is not yet at the stage in which it may considered standard treatment.

Multiple minor band indentations without vascular compromise or functional or cosmetic problems do not require treatment. Constricting rings that cause distal deformity are treated with excision of the constriction ring and concomitant or staged Z-plasty reconstruction (Fig. 22-36). Complete excision of the ring is necessary to recontour the digit or limb. The depth of excision can extend to the periosteum. Such digits usually have chronic impaired venous outflow with marked distal swelling. In these situations, it is imperative to preserve distal venous drainage and the deep neurovascular structures. Careful dissection of the veins, arteries, and nerves is performed on both sides of the deep constricting band. These structures are then delicately freed from the band to preserve their longitudinal integrity. It is recommended that complete circumferential excision not be performed in one procedure. Rather, excision up to 270 degrees at one time may be safest for preservation of vascular inflow and outflow. Z-plasties or flap and fat mobilization are performed after ring excision, so as to prevent recurrence.

Syndactyly release with Z-plasties and skin grafts follows the basic principles outlined in the section on syndactyly. The unique features of amnionic band syndrome are acrosyndactyly secondary to constricting bands and the presence of epithelialized incomplete web-space proximal to the syndactyly.

In the rare situation of constricting bands causing progressive deformity in digits of unequal length, early digital separation is necessary. More often, the acrosyndactyly separation can be performed after 6 months of life. There is usually limited skin

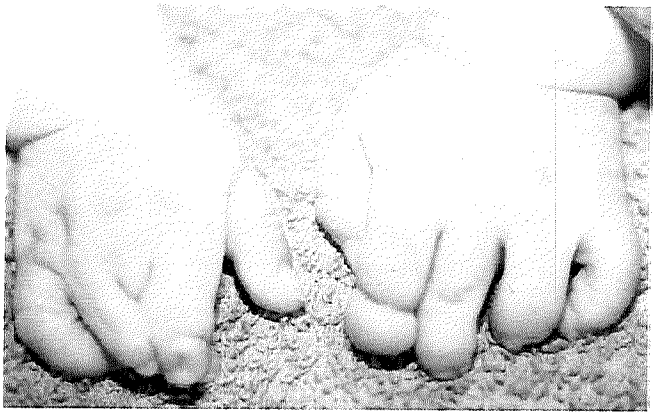


FIGURE 22-36. Bilateral amnionic band syndrome with deep constriction rings on the left hand and partial acrosyndactyly and amputations on the right hand. (Figure courtesy of Children's Orthopaedic Surgical Foundation (COSF), © 2010.)

for coverage, and creative flap design is needed to cover all the web spaces. Abundant skin graft is necessary. Distal release of complex syndactylies may require excision of osseous or cartilaginous synostoses. The embryologic remnant of the web is usually too distal and small to serve as an acceptable web reconstruction. Excision of that epithelial tract is usually performed. If that primitive web is used, it often must be deepened secondarily.

The most severe cases involve absence or deficiency of the thumb. The reconstruction can include metacarpal or phalangeal transposition from the index finger (158, 338), nonvascularized toe phalangeal transfer, or vascularized toe transfer (Fig. 22-37). It is imperative to reconstruct the thumb for pinch and proper grasp and release functions in these patients.

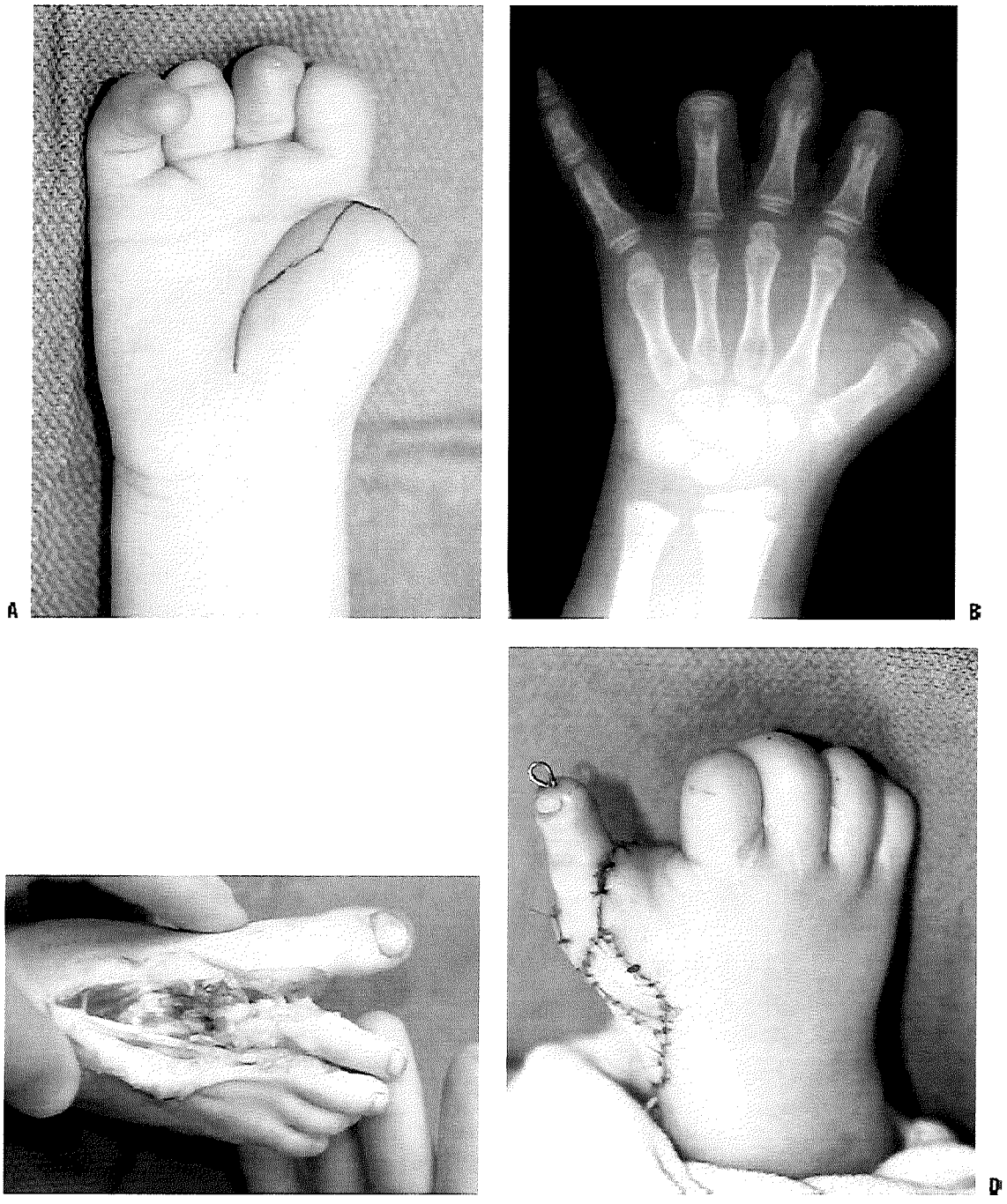


FIGURE 22-37. **A:** Preoperative clinical photograph of a toddler with constriction band syndrome and congenital amputation of the thumb at the metacarpal–phalangeal joint level. **B:** Preoperative radiograph revealing the level of the congenital amputation through the proximal phalanx just beyond the epiphysis. **C:** Intraoperative anatomy of second toe donor harvest for microscopic transfer and thumb reconstruction. **D:** Immediate postoperative clinical photograph of toe transfer. This process led to normal thumb functioning and to a remarkable cosmetic result for the patient.

Because the underlying tissues are normal, as is the central nervous system, these patients have outstanding hand function after reconstruction. There are clear cosmetic differences, but minimal functional differences between them and their peers.

Release of Congenital Constriction Band Congenital constriction bands can occur in any location on a limb. They occur with varying degrees of severity, ranging from incomplete partial rings that may require no treatment to deep rings that completely encircle the part, creating distal edema and cyanosis. This ring of abnormal constriction has breadth as well as depth and consists of abnormal, dense, scar-like tissue. For this reason, the constriction band must be excised rather than merely incised. If not excised, the dense scar tissue is merely rotated into the flaps. In excising the constriction ring, especially in areas such as the fingers or when it appears to go down to the bone, great care must be taken not to divide vital structures that lie beneath. This is especially troublesome in the fingers. Further, no more than half the circumference of the constriction ring should be excised at one time to avoid complete disruption of the lymphatic and vascular drainage from the distal part. An interval of 2 to 3 months is usually adequate for the reestablishment of the lymphatic and vascular drainage of the operated half of the finger.

It is not acceptable simply to excise the ring, no matter how minor it appears, because attempts to repair the defect in a linear manner cause the resultant scar to contract, creating a cosmetic problem. Thus, the treatment of congenital constriction bands lies in the application of the principles of Z-plasty (Figs. 22-38 and 22-42).

Syndactyly. Syndactyly is one of the most common congenital deformities. It occurs because of a failure of separation of the digital rays *in utero*. Normal differentiation of digits occurs during the fifth to eighth weeks of gestation. Failure of normal programmed cell death results in *syn* (together) *dactylos* (digits). The incidence is between 1 in 2000 and 1 in 2500 live births (1). It can occur in isolation or as part of a syndromic condition. It is often an inheritable condition, whether in isolation or as part of a syndrome. It is bilateral and symmetric in up to 50% of patients. It is more common in boys than in girls.

Syndactyly is classified by the extent of, and the tissues involved in, the webbing. Digital separation *in utero* starts distally and proceeds proximally. Normally, the third web space is the most distal web, followed by the second, fourth, and first web spaces. The normal commissure of the web extends over 30% to 35% of the length of the proximal phalanx (339). The bones, joints, tendons, and neurovascular structures separate before the skin does. If separation fails to occur or ends prematurely, syndactyly results. If it extends over the entire length of the phalanges, then it is deemed complete. Incomplete syndactyly is when the web is more distal than is normal but does not extend to the digital tips (Fig. 22-40); complex syndactyly is when there is osseous connection between the digits; and simple syndactyly is when the digits are joined by skin only (335). Acrosyndactyly involves webbing of the tips of all the digits. Syndactyly can also be a part of other major developmental problems in the hand that affect hand function, such as brachydactyly, camptodactyly, clinodactyly, symphalangism, and polydactyly. These are the most complicated syndactylies in terms of surgical decisions and care. As noted in the preceding section, syndactyly secondary to amnionic band syndrome is not a malformation, but an *in utero* disruption, and will be considered separately.

Clinical and Radiographic Features. Syndactyly most often affects the third web space of the hand. It is sequentially less common in the fourth, second, and, finally, first web spaces (1). It may be associated with syndactyly of the toes. In isolated syndactyly, often one of the parents will have an incomplete syndactyly of the fingers and/or toes. As mentioned in the preceding text, there are many chromosomal, craniofacial syndromic, and generalized syndromic conditions associated with syndactyly. There are rare reports of cardiac conduction defects associated with syndactyly. These all need to be evaluated before treating the syndactyly. The most important aspect of the hand evaluation for syndactyly is determination of the quality of the affected digits. In simple syndactyly, these digits are usually normal except for their skin union. In more complex situations, the digits may have malalignment, limited motion, and limited strength after surgical separation. Plain radiographs will reveal osseous union and marked joint and bony malalignment. However, in infancy, the areas of chondral abnormalities in the joints, physes, and between digits exhibiting syndactyly will not be visible on plain radiographs. MRI and arteriography are used only in very complex situations to define digital anatomy preoperatively.

Treatment. Patients with incomplete syndactyly may choose not to undergo surgical separation. If the syndactyly does not extend to or beyond the PIP joint, this will not limit function. However, it may affect wedding ring wear in the third web space or the use of gloves during manual labor or sports-related activities; for this reason, some patients request separation.

Most parents and children with complete syndactyly desire separation of the digits for functional and aesthetic reasons. There are rare situations in which a family declines surgery for complete syndactyly. Because of the discrepancy in the lengths of the adjacent digits, there may be some degree of bony malalignment and joint contracture. This is most marked in the border digit syndactylies (first and fourth web spaces), and least marked in the third web space. Leaving the digits joined also precludes independent function of the affected digits. There are also syndromic and chromosomal situations in which the overall health or the mental capacity of the patient does not warrant the risks of surgical separation. Finally, there

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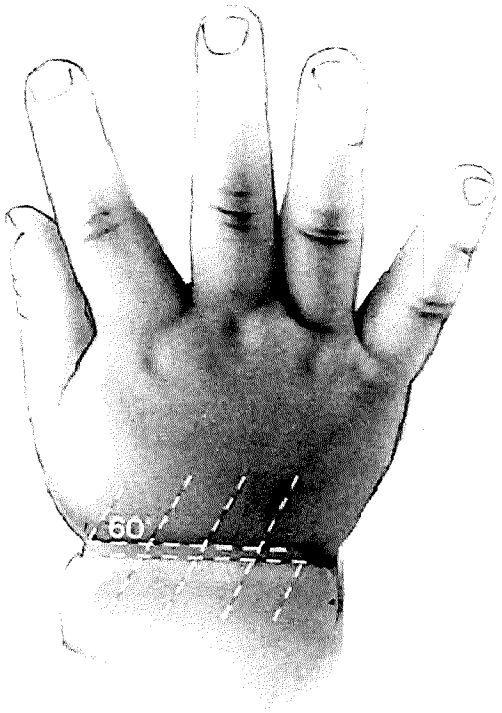
FIGURE 22-38. Release of Congenital Constriction Band. The excision of the constriction band can be marked, although it is usually so obvious that this is not necessary. The flaps of the Z-plasty are then planned. They should be as large as is feasible. The angle of 60 degrees is believed to provide the optimal balance between the vascular supply to the tip of the flap and the mobility of the base of the flap. Ideally, the length of the flap should be no more than two times the width of the base of the flap.



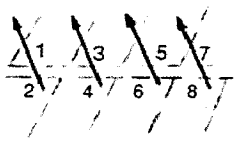
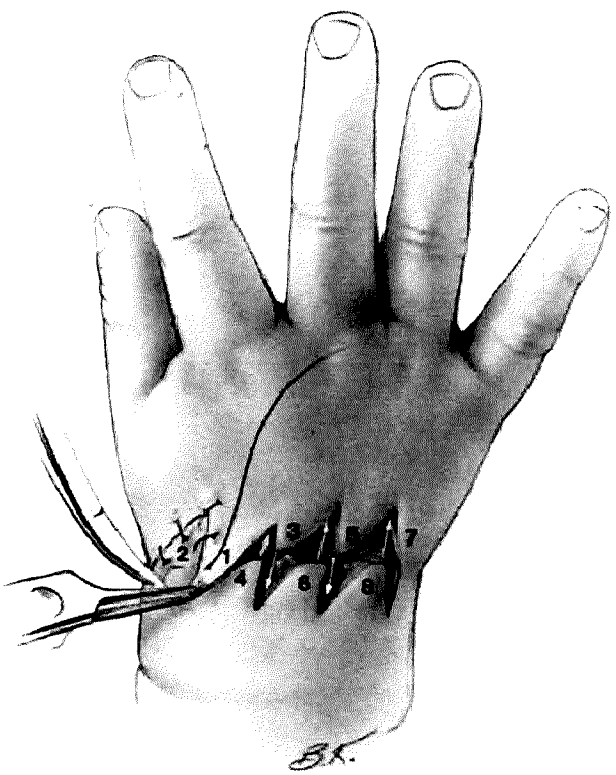
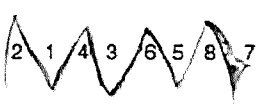


FIGURE 22-39. After the constriction band is excised and the flaps are mobilized, they are transposed. In children, it is best to use fine absorbable sutures to avoid the arduous task of suture removal. Great care should be exercised in handling the flap tips to avoid necrosis. When placing the corner stitches, the subcutaneous tissue, not the skin, should be grasped in the flap tip region to avoid tip necrosis.





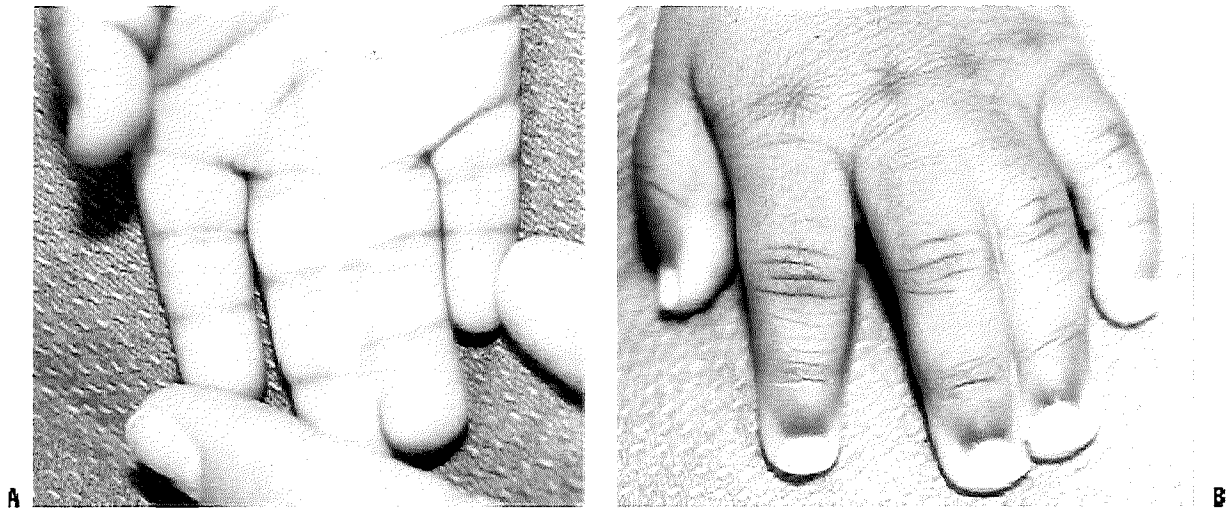


FIGURE 22-40. **A,B:** A 1-year-old child with complete simple third web-space syndactyly. In this patient, the distal eponychial folds and nail plates are already separate. The underlying joints, tendons, nerves, and blood vessels should be separate and normal. (Figures courtesy of Children's Orthopaedic Surgical Foundation (COSF), © 2010.)

are situations of complex syndactyly in which the affected digits are too hypoplastic, malaligned, or stiff to warrant separation. Otherwise, the standard treatment is surgical separation of the affected digits.

Unfortunately, separation is not as simple as parents wish; simple division of the conjoined skin is not sufficient. The uncovered soft tissues result in linear scars with long-term joint contractures, digital malalignment, and loss of motion and function (297, 340). Standard treatment now consists of (a) vascularized local rotation flap coverage for web commissure reconstitution; (b) zig-zag incisions, avoiding the interdigital creases to prevent scar contractures; and (c) full-thickness skin grafts to cover all areas of the digits not covered by local flaps (Fig. 22-41). In addition, special attention is given to the eponychial reconstruction with either local flaps or composite grafting (341).

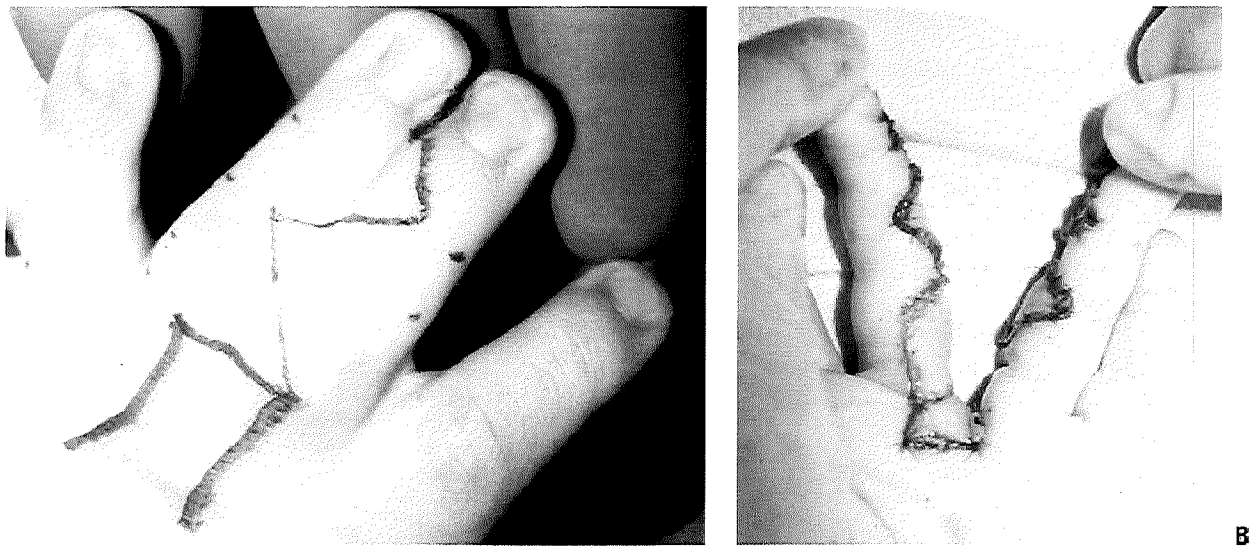


FIGURE 22-41. **A:** Intraoperative photograph of a 1-year-old child with complete syndactyly treated with dorsal rotation flap coverage and Z-plasties, as outlined. Note the skin marks on the lateral borders of the ring and long fingers to outline the apex and base of each Z-plasty. This allows for precision placement of corresponding volar and dorsal Z-plasties. **B:** Intraoperative photograph after dorsal-to-volar rotation flap coverage for web space, Z-plasties, and full-thickness skin grafting. (Figures courtesy of Children's Orthopaedic Surgical Foundation (COSF), © 2010.)

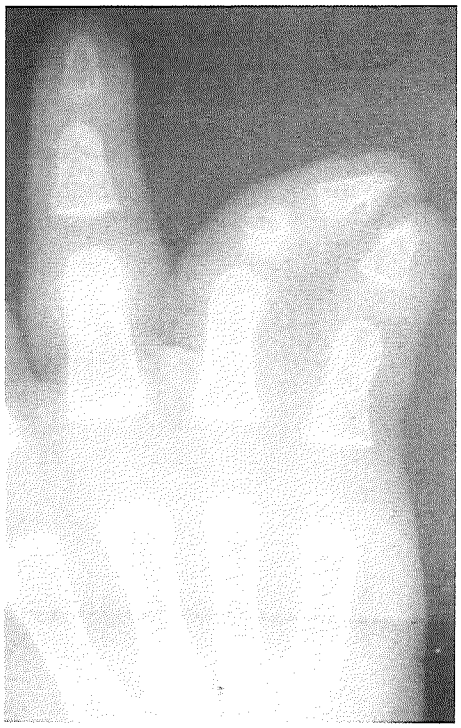


FIGURE 22-42. Radiograph of complex syndactyly of the fourth web space with progressive deformity of the ring finger. This should be released early in infancy to prevent progressive deformity. The abnormal middle phalanx of the ring finger may still require corrective osteotomy. (Figure courtesy of Children's Orthopaedic Surgical Foundation (COSF), © 2010.)

Surgery is generally performed in infancy, when anesthesia and surgical handling of the tissues are safe. There is some controversy regarding the best age for surgery, but in most institutions, it is performed at approximately 12 months of age (342). After 6 months of age, the anesthesia risk is equivalent throughout childhood. With magnification, surgery can be performed safely and skillfully during infancy. The only controversy concerns surgical healing and scarring. Neonatal releases result in more scarring. There is some evidence that surgical release performed at approximately 18 months of age may result in less scarring and recurrent web contractures than release during infancy (343). However, this is a very difficult developmental age for elective surgical intervention. Border digits of unequal length need to be separated earlier to lessen angular and rotatory deformity in the longer digit (Fig. 22-42). Complete separation of the digits in the neonatal period has had a higher rate of complications.

In incomplete syndactyly that is proximal to the PIP joint, surgery usually involves the use of local flaps such as double-opposing Z-plasties and "stickman" or "dancing girl" flaps. Separation may not be to normal depth, but patients often prefer to avoid skin grafting (344). If the incomplete syndactyly extends to the middle phalangeal region, full-thickness skin grafting is necessary.

In simple, complete syndactyly, surgery involves the use of a dorsal rotation flap into the web, Z-plasty flaps the length of the digits, and full-thickness skin grafts to cover the defects. It is important to have a vascularized flap for web commisure reconstruction. This is usually done with a dorsal rectangular flap but may also involve a dorsal metacarpal island flap (345–347). The fascial connections between the digits extending from Grayson and Cleland ligaments need to be separated. Any synostosis or synchondrosis union of the distal phalanges should be divided. Conjoined nails are divided, and the exposed eponychial and paronychial regions are reconstructed with local flaps or composite grafts (348, 349). If the common digital nerve extends beyond the desired web deepening, epineural separation is performed proximally. If the common digital artery bifurcates distally, ligation of one of the proper digital arteries may be necessary for obtaining the desired separation. This is one of the major reasons why surgery is not simultaneously performed on both sides of a single digit in multiple syndactylies.

Recently there has been interest in "graftless" techniques of digital separation, utilizing pedicled dorsal metacarpal artery flaps or random pattern advancement flaps from the dorsum of the hand (350, 351). At present, the superiority of these techniques over traditional release with full thickness skin grafts has not been demonstrated.

Complex syndactylies are more likely to have abnormal underlying joints, bones, neurovascular structures, muscles, or tendons. The separation of skin follows the same principles as in complete syndactyly. If there is significant digital malalignment, skin incisions may need to be modified so as to maximize coverage. After separation of the skin, all abnormal connections of fascia, tendons, bones, joints, nerves, and arteries need to be addressed individually. Phalangeal deformity may require osteotomy. Instability of the joints may require ligamentous reconstruction. Stiffness of the joints, camptodactyly, or symphalangism may need to be dealt with subsequently. Neural, vascular, and nail problems are managed in a manner similar to those described for complete syndactyly. Tendon reconstruction is performed primarily if possible. Brachydactyly is usually addressed subsequently, if at all.

Acrosyndactyly separation begins early in life, especially if it is bilateral. Adjacent webs are not separated simultaneously. Generally, the first and third webs are separated together, as are the second and fourth webs. Abundant full-thickness skin graft is usually required. Sufficient time between procedures (3 to 6 months) lessens worries about flap necrosis and scar contracture. In syndromic conditions, such as Apert syndrome, the acrosyndactyly is more complex, and normalcy may never be achieved (352, 353).

Complications. Fortunately, the most worrisome complication—an avascular digit—is almost never encountered and should

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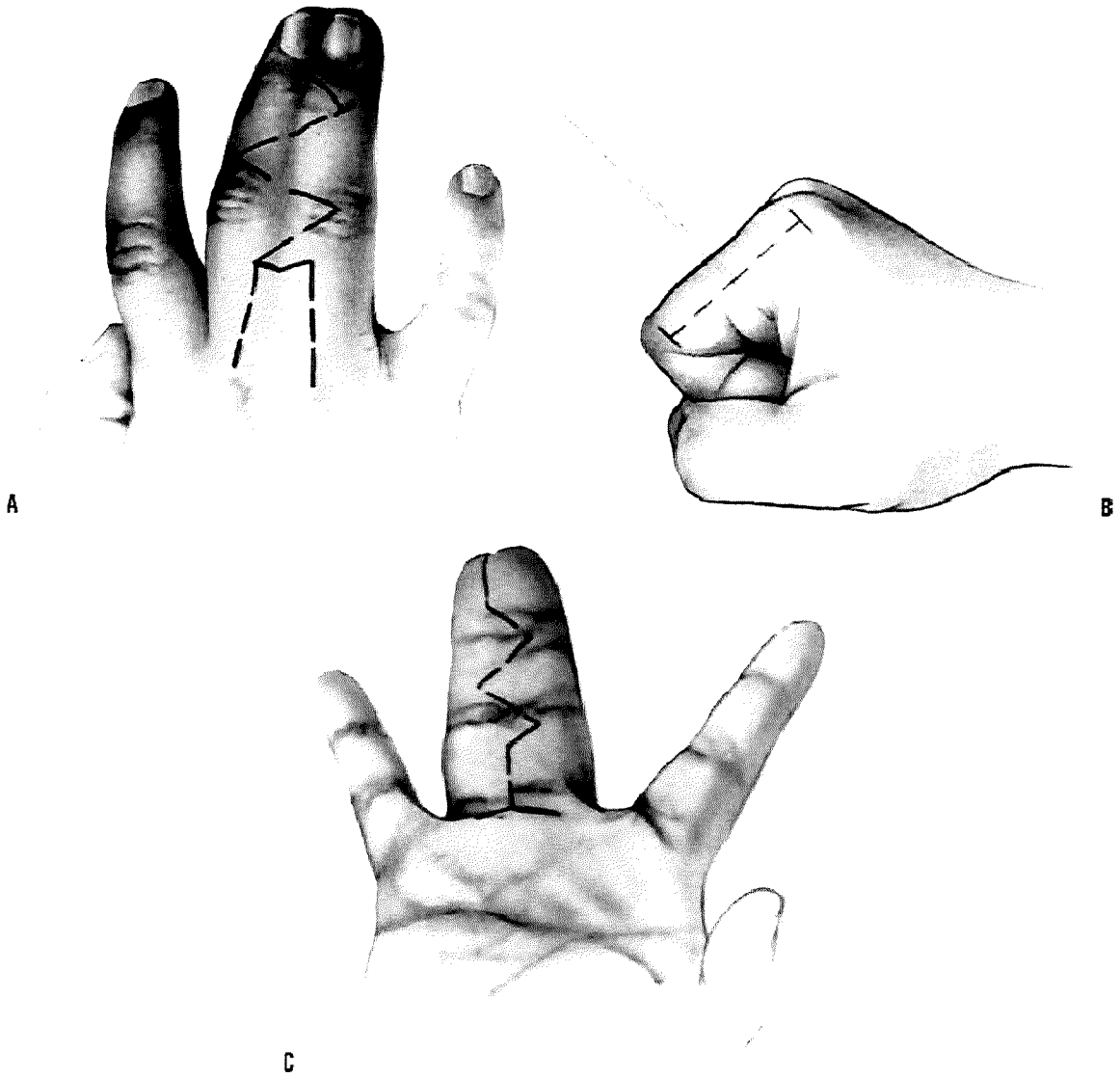
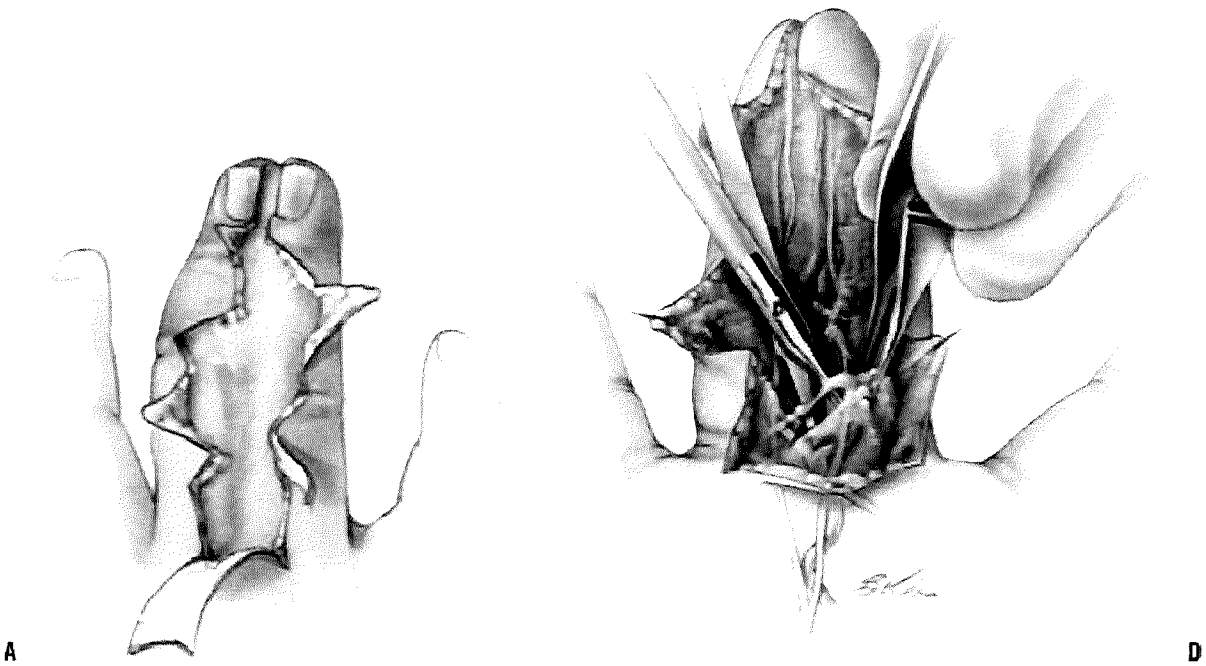


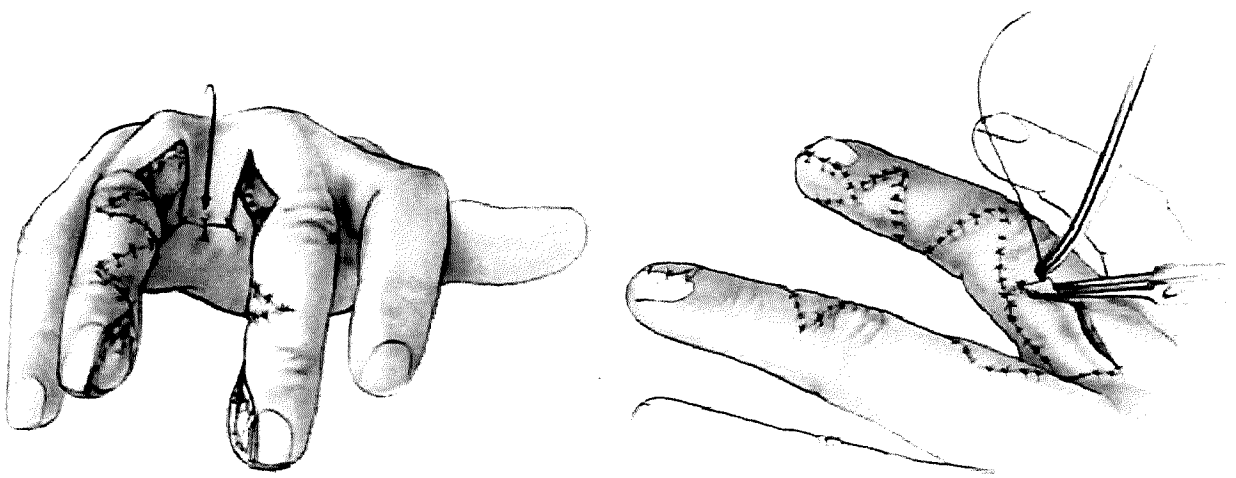
FIGURE 22-43. Release of Simple Syndactyly. The first part of the operation is to plan the incisions. Many different incisions work well, and surgeons often prefer to use the one that they were taught. The planning begins with the flap that will be used to reconstruct a commissure. The broad dorsal flap, first described by Bauer and colleagues (3), has received wide acceptance and is described here. The flap begins at the metacarpophalangeal joint and extends about two-thirds of the way to the proximal interphalangeal joint. It is about 1 cm in width and should taper slightly from the base to the tip **(A)**. Before the volar incisions are made, it is first necessary to determine the location of the commissure on the volar surface. The location can be determined by examining the hand from the radial side in the clenched-fist position. In the normal hand **(B)**, the commissure is about midway between the metacarpal head and the distal condyle of the proximal phalanx. This point can be marked by passing a small needle from the dorsal to the volar surface along this midway mark. At this point, a transverse incision can be made to provide the area where the dorsal flap will be sutured. After this, dorsal and volar zigzag incisions are made out to the distal interphalangeal joint **(C)** in such a manner that the base of the triangle of the volar flap matches the tip of the dorsal flap, and vice versa. The planning of this interdigitation can be aided by passing a small 27-gauge needle through the dorsal and volar skin to mark the tips of the flaps.

From the distal interphalangeal joint to the tip of the fingers, a straight longitudinal incision is made to complete the separation of the skin. Making one clean, sharp, decisive incision through the nail plate, if it is joined, will help avoid damaging it. As these flaps are developed, it is extremely important that they are defatted. This increases their mobility and decreases the postoperative swelling.



A D

FIGURE 22-44. There is usually a clear line of separation between the two fingers in a simple syndactyly **(A)**, with little crossing of fibrous bands or blood vessels. In some cases, however, the digital vessels or nerves can divide more distally than usual. It is therefore necessary to isolate the neurovascular bundles. This isolation should start distally **(B)**, following these structures proximally until their junction is found. If the nerve divides more distally than is desirable, it can be split. If the vessels divide more distally, the surgeon must decide whether to divide one of these to allow the commissure to be moved more proximally to the correct location. Some surgeons do not advocate this, but it is commonly done. If one vessel is divided, it should be recorded carefully in the operative note so that on further surgical planning these records can account for the fact that only one artery supplies the digit. This is especially important when there is a syndactyly on the other side of the affected finger.



A B

FIGURE 22-45. The flaps are now sutured into place, starting with the dorsal flap. Before this is done, it is important to be sure that the flaps are defatted. The flaps should be sutured with a fine, absorbable material **(A)** so that these sutures will not have to be removed.

This leaves two areas on each finger to be grafted: the most distal and most proximal portion of each finger. Thin, full-thickness skin **(B)** should be used for this coverage. It can be obtained most easily from the groin crease; however, the physician must be careful to stay far enough laterally to avoid skin that will later grow hair. This full-thickness skin should be obtained lateral to the femoral artery and preferably even more laterally, from an area below and just medial to the anterosuperior iliac spine. The defect from the donor area of the graft is closed primarily, and the graft is sutured into the recipient areas. A pressure dressing that applies gentle compression to the flaps and the skin grafts is essential. This should be covered with a rigid plaster that extends above the elbow to immobilize the child's arm.

be avoidable. Adherence to the axiom of never operating on both sides of a digit during the same procedure prevents the occurrence of this devastating complication. Careful dissection of the digital vessels in complex situations lessens the risk of avascularity at the initial or subsequent operations. Preoperative vascular studies in complicated situations prepare the surgeon and allow him or her to avoid intraoperative surprises and dangers. Flap necrosis and scar contracture are more common. The flaps should be secured without tension, and their vascularity should be checked with deflation of the tourniquet at the completion of the procedure. If in doubt, lessen the tension and use skin graft. Skin graft failure is usually caused by inadequate immobilization and excessive shear forces applied to the grafts. Secure immobilization with a compressive dressing, long-arm cast, and sling and swathe bandages is necessary for protecting the grafts. Infection is rare but will result in marked scar contractures that require reoperation. Long-term issues that have been reported regarding skin graft sites include contracture formation, graft breakdown (both incidences are seen more often with split thickness grafts), hyperpigmentation, and hair growth (both incidences are seen more often with full-thickness grafts) (354). Nail deformity and poor eponychial coverage are common when there is an initial conjoined nail. The use of composite grafts seems to lessen the incidence of these complications. Web-space creep is common with growth, but often does not require reoperation unless digital contractures develop. The use of local rotation flaps and skin grafts will resolve this problem. Keloid formation is rare but has been shown to be associated with primary digital enlargement before syndactyly separation (355). Finally, angular deformity may require osteotomy or joint reconstruction.

Polydactyly. Polydactyly is a common congenital malformation. It can occur on the radial (preaxial), central, or ulnar (postaxial) portion of the hand. Preaxial, or thumb, polydactyly usually occurs in isolation and will be addressed in the section dealing with the thumb. Central polydactyly is very rare and is usually associated with syndactyly; the underlying digits are rarely normal. It can be inherited in an autosomal dominant manner. It affects girls more than boys and is often bilateral. Postaxial or small-finger polydactyly has a variable racial incidence, with the occurrence in African Americans estimated to be as high as 1 in 230 live births, and that in whites estimated to be 1 in 3000 live births (356–358). It is often bilateral. Postaxial polydactyly has been classified by Stelling (359, 360) and Turek into three types. Type I involves soft tissue alone and is very common in the African American population. Type II involves phalangeal duplication articulating with a single or bifid metacarpal head. Type III involves a complete ulnar ray duplication, including the metacarpal. There is also a universal classification of polydactyly proposed by Buck-Gramcko and Behrens (335) that denotes the digit involved by a Roman numeral (I to V) and the extent of bifurcation by abbreviation [DIST (distal), DIP (distal interphalangeal), MID (middle), PIP (proximal interphalangeal), PROX (proximal), MCP (metacarpophalangeal), MET (metacarpal), CMC (carpometacarpal), C (carpal), IC (intercarpal), and RUD (rudimentary polydactyly)]. The major issue for the surgeon is not the classification of the polydactyly, but whether excision with reconstruction, or excision alone, is warranted. Only postaxial soft-tissue polydactyly (type I, or rudimentary) can be treated by excision alone. All other forms require reconstruction. Central polydactyly is usually an isolated malformation, and postaxial polydactyly in African Americans is almost always an isolated malformation. Postaxial polydactyly in whites without a positive family history may be associated with chromosomal abnormalities, other syndromes, or other malformations.

Treatment. Soft-tissue polydactyly on the ulnar side can be treated with excision during the newborn period (Fig. 22-46). Unfortunately, too often this has been performed with a suture ligature by inexperienced hands. The result is a persistent soft-tissue nubbin caused by incomplete excision of the base of the digit. If a suture ligature is used, it must bring about necrosis of the entire digit. Otherwise, it may be best to perform an elliptical excision under local anesthesia. Care is taken to ligate the digital vessels with this excision. Other than failure to completely excise the digit, complications are rare, and the hand is normal afterward. There have been reports of traumatic neuromas along with hypertrophic scars that have led to late problems and the need for repeat surgery (361, 362). The parents should be aware of the future genetic implications for them and their children.

More complex postaxial polydactylies require excision and reconstruction in the operating room. In addition to excising

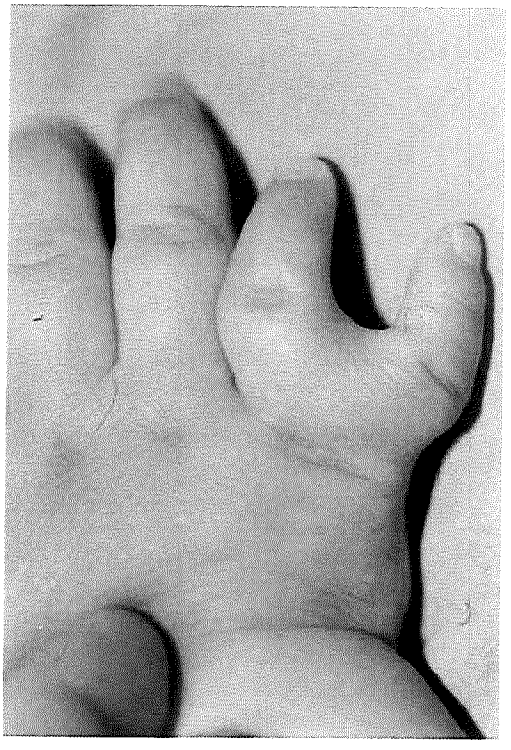


FIGURE 22-46. A: Complete postaxial polydactyly with phalangeal duplication with a conjoined metacarpal.

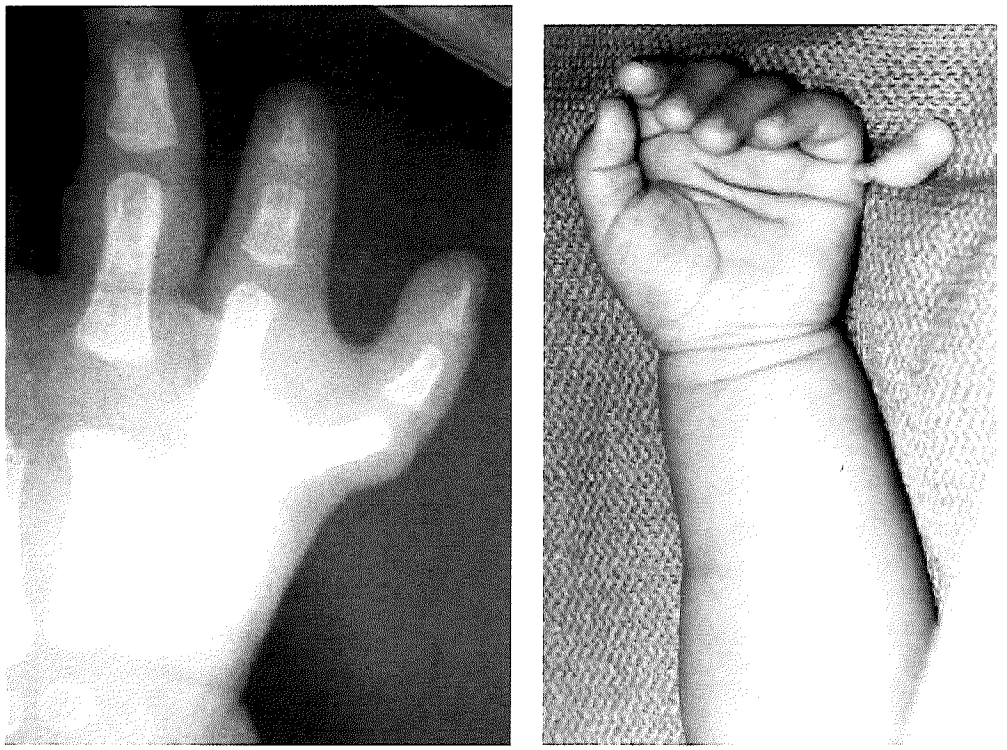


FIGURE 22.46 (*continued*) **B:** Radiograph of the same patient. Reconstruction will consist of excision of the duplicate phalanges, contouring of the bifid metacarpal head, and transfer of the metacarpophalangeal joint ulnar collateral ligament and the hypothenar musculature to the reconstructed fifth digit. **C:** Simple polydactyly with only soft-tissue attachment. This can be excised in the newborn nursery under local anesthesia. Care must be exercised while performing suture ligature; the entire stalk should be excised so as to avoid leaving a residual nubbin. (Figures courtesy of Children's Orthopaedic Surgical Foundation (COSF), © 2010.)

the redundant parts, transfer of the hypothenar muscles (abductor digiti quinti, flexor digiti quinti) from the sixth to the fifth digit is necessary. In type II polydactyly, the MCP joint collateral ligaments are also transferred to the reconstructed fifth digit. If the metacarpal head is enlarged or bifid, intra-articular osteotomy is appropriate. In this procedure, the origins of the collateral ligament and the metacarpal physis should be preserved. In type III polydactyly, the entire ray is resected, and the basilar joint is stabilized. The outcome of surgery in both type II and III malformations is usually cosmetic and functional normalcy.

Treatment of central synpolydactyly is much more complex (363, 364). The major decision is whether it is feasible to achieve independently functioning digits. The choices are to leave the digits conjoined, to attempt reconstruction to a five-digit hand, or to perform ray resections of part or all of the synpolydactyly. Often the involved digits have bone and joint malalignment, hypoplasia, and poor motor, nerve, and vascular supplies. The reconstructed digit is usually smaller, stiffer, weaker, and malaligned. The family needs to be well aware of this, so that their expectations are realistic as far as surgical reconstruction and digital function are concerned. Treatment decisions in this condition are often personal, based on the family's desires and the surgeon's preferences.

Camptodactyly. Camptodactyly translates from Greek to mean *bent finger*. It involves a flexion deformity of the PIP joint, most commonly in the small finger. It may present in infancy or in adolescence. It may involve a single digit or multiple digits (Fig. 22-47).

Camptodactyly may be associated with multiple systemic malformations. Its incidence is unknown but has been cited at <1% of the general population (313, 332, 365). Most cases appear in infancy, and there is equal gender distribution (366). Less commonly, it may first appear in adolescence, usually in girls (367). Some cases are familial, with an autosomal dominant inheritance etiology and variable penetrance patterns. Most cases are isolated, without a positive family history. Up to two-thirds of the patients have bilateral small-finger involvement. It is important to distinguish camptodactyly from neurologic causes of clawing or from posttraumatic butonniere deformities (368).

The etiology is unknown and continues to be debated. In simple terms, there is an anatomic imbalance between the flexor and the extensor mechanisms (366). This may be secondary to an abnormal insertion of the lumbricals, hypoplastic or foreshortened flexor digitorum superficialis, or retinacular ligament anomalies (369–371)). The volar skin is usually tight. With growth, these anatomic abnormalities cause PIP joint contracture and phalangeal bony abnormalities. The distal proximal phalanx becomes narrowed volarly and flattened

FIGURE 22-47. An adolescent patient with marked camptodactyly of the small, ring, and long fingers. There are proximal interphalangeal joint flexion contractures in each digit, and the patient is actively hyperextending the metacarpophalangeal joints to compensate for those contractures. (Figures courtesy of Children's Orthopaedic Surgical Foundation (COSF), © 2010.)

dorsally, with loss of the normal contour of the head. The articular surface can become incongruous, with notching of the base of the middle phalanx (372). Initially, the patient can compensate for the PIP joint flexion contracture by MCP and distal interphalangeal (DIP) joint hyperextension (Fig. 23-24). This keeps the digital pulp of the affected fingers in line with the other digital rays. Therefore, the mild contracture in the older child may not need treatment. However, as the contracture continues to progress beyond 30 degrees and toward 90 degrees, it becomes more difficult for the patient to compensate. Camptodactyly is usually progressive with growth.

Treatment. Treatment should attempt to restore normal flexor–extensor balance. The options for treatment are splinting and surgical reconstruction. Most clinicians recommend an initial treatment program of progressive passive extension and splinting with dynamic or progressive static splints. Parents are instructed to perform frequent home exercises for their infants; affected adolescents are similarly instructed to perform a home program for themselves. The goal is to achieve full or normal passive extension. It is hoped that active extension will follow. Many clinicians (367, 373–375) report success with a splinting program in most cases. The best results are in mild cases in young patients. The patients are followed until the end of growth in order to treat recurrence if it occurs.

There is significant debate regarding the indications for surgery. McFarlane et al. strongly recommend surgical intervention to reconstruct the aberrant insertion of the lumbrical (371, 376). However, the published surgical results are disappointing in terms of outcome (371, 374, 376, 377). Surgery is reserved for severe contractures that are not amenable to splinting treatment. Specifically, surgery shows best results in patients in whom the finger is flexed into the palm and obstructs use of the hand. The principle of surgical intervention is to correct the abnormal anatomy. This involves release of the aberrant lumbrical (313, 332, 371, 376) or flexor digitorum superficialis (370) insertion in conjunction with volar Z-plasties and PIP joint release (378). Local flaps or full-thickness skin grafts are often necessary for volar skin coverage. Tendon transfer to the extensor mechanism is performed in patients with full passive extension of the PIP joint, but no active extension (379). The results of soft-tissue reconstruction often merely change the arc of motion rather than normalize it. Preoperatively, patients have significant flexion contracture. Postoperatively, they generally have difficulty achieving full active and passive flexion.

There are frequently bony changes present at the PIP joint, and these preclude restoration of normal motion or alignment. Oldfield (380) and Flatt (1) have stated that, in the presence of marked radiographic evidence of bone and joint changes, corrective extension osteotomy may be most effective at improving alignment and function. The published data about this salvage operation are too limited to enable objective evaluation (381).

In summary, with treatment of camptodactyly, it is unusual to achieve a perfectly aligned and mobile digit. Parents and patients should be aware of this from the outset. In addition, deformity can recur with growth and persistent muscle imbalance.

Clinodactyly. Clinodactyly is abnormal angulation (>10 degrees) of the digit in the radioulnar plane. It is usually caused by abnormalities of the middle phalanx. The middle phalanx is trapezoidal, with less height on the radial side. This results in deviation at the DIP joint. Clinodactyly is most often seen in the small finger and is usually bilateral. This form of clinodactyly has an autosomal dominant inheritance (382–385). Clinodactyly is also frequently associated with syndromes (Holt-Oram, Turner, Silver, and Cornelia de Lange) and chromosomal abnormalities (trisomies 18 and 21), and should alert the primary neonatal examiner to look for associated malformations or problems. For example, clinodactyly of the thumb is seen in Rubinstein-Taybi syndrome (386, 387) and diastrophic dwarfism (388, 389). In addition, it is common with other congenital abnormalities of the hand. Mild clinodactyly, however, is commonplace in otherwise healthy individuals. In these situations, the major issue is aesthetic. Function is affected only when the deformity is severe enough to impinge on the adjacent digit during flexion.

Treatment. Treatment is based on the degree of deformity. Most cases are mild and nonprogressive, and therefore do not warrant surgical intervention. Progressive, severe clinodactyly

may interfere with flexion and grip. In these rare situations, the progressive deformity is secondary to altered physeal growth. The middle phalangeal physis may be a bracketed delta phalanx. Treatment options are epiphyseal bracket resection and surgical realignment. Physeal bar resection and fat graft interposition have been reported by Vickers (390, 391) to restore longitudinal growth and provide correction (392). Surgical realignment can be in the form of opening-wedge, closing-wedge, and reverse-wedge osteotomies. Osteotomy should be delayed until there is sufficient ossification of the middle phalanx to allow for precise cuts. Generally, this is around school age. The complications of osteotomy are persistent deformity and loss of interphalangeal motion. Loss of motion in a patient whose indication for surgery was purely aesthetic is unacceptable to most patients, their families, and surgeons.

HAND: THUMB

Congenital deformities of the thumb occur in all categories of congenital hand anomalies described by the American Society and the International Federation of Societies for Surgery of the Hand. Failure of formation occurs with aplasia of the thumb, and this is often associated with other radial-sided longitudinal deficiencies (Fig 22-48). Failure of separation occurs with thumb–index syndactylies, which are common with other syndactyly syndromes. Duplication is seen in the form of thumb polydactyly, undergrowth as thumb hypoplasia, and overgrowth as macrodactyly and triphalangeal thumbs. Also, thumb abnormalities are common as part of constriction band syndrome or generalized musculoskeletal disorders. The list is exhaustive and this section will cover the more common congenital thumb malformations.

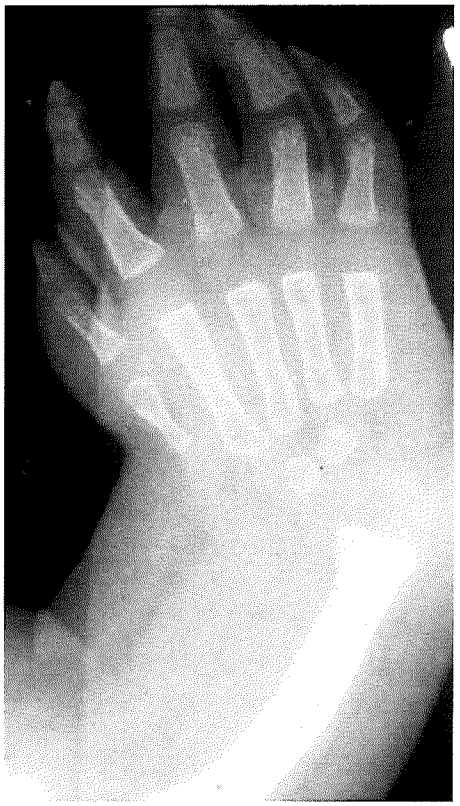


FIGURE 22-48. Radiograph of a thumb with type IIIB hypoplasia without a carpometacarpal joint and proximal thumb metacarpal. This is usually treated with pollicization. Less often, treatment is by reconstruction, potentially including a microvascular toe metatarsophalangeal transfer to form a thumb carpometacarpal joint. Note the radial dysplasia in this patient.

Trigger Thumbs/Digits. Trigger thumb represents an abnormality of the flexor pollicis longus and its tendon sheath at the A1 pulley, where there is a palpable mass (Notta nodule), representing the flexor pollicis longus constriction at the A1 pulley. In the past, trigger thumbs were thought to be congenital. However, this condition is *almost always* acquired in the first 2 years of life, as indicated by a prospective screening of neonates, which failed to record any trigger thumbs (393–395). The cause appears to be a size mismatch between the flexor pollicis longus and the A1 pulley, leading to progressive constriction. Unlike adult trigger digits, there does not appear to be an inflammatory component (396); 30% of the cases are bilateral. Isolated trigger thumbs have no associated syndromes. However, trigger digits are seen with neurologic syndromes (trisomy 18) and mucopolysaccharidoses (119). There is no familial inheritance pattern. Patients with trigger thumb present at ages ranging from infancy to school age. Often, the diagnosis is missed until local trauma brings attention to the thumb. In the emergency setting, the flexed interphalangeal joint can be mistaken for an interphalangeal joint dislocation. Radiographs are misleading because of limited phalangeal ossification. A palpable nodule at the A1 pulley is diagnostic. If the trigger is longstanding, compensatory hyperextension of the MCP joint develops to effectively bring the thumb out of the palm. In addition, mild radial deviation of the interphalangeal joint may develop, secondary to eccentric flexor pull.

Treatment. In infants younger than 9 months of age, Dinham and Meggit (397) found that 30% of trigger thumbs may resolve spontaneously. In infants older than 1 year of age, <10% of trigger thumbs resolved spontaneously. Ger et al. (398) found lack of resolution in their patients after observation for 3 years. There is limited evidence that splinting is of benefit (399), and often it is not well tolerated. For these reasons, surgical release of the constricting A1 pulley and the flexor tendon sheath has historically been the treatment of choice in infants who show no spontaneous resolution by 1 year of age, and in any toddler or older child presenting with a locked trigger thumb (Figs 22-49 to 22-52). Incision is made transversely in the digital crease to lessen scarring. Care must be taken to avoid injury to the superficial digital neurovascular bundles. The oblique pulley has to be preserved so as to prevent flexor tendon bowstringing. Recurrence is extremely rare. Recently, recommendations for earlier surgical release have been reexamined given new information regarding the natural history and potential for spontaneous improvement in IP joint extension (400). In a prospective evaluation of 71 thumbs

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Release of Congenital Trigger Thumb (Figs. 22-49 to 22-52)

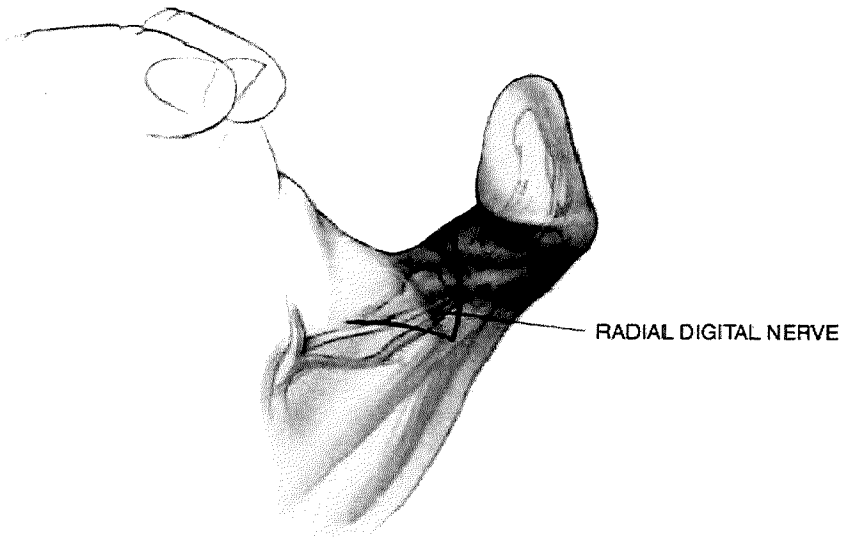

FIGURE 22-49. Release of Congenital Trigger Thumb. Although this release can be done through a simple transverse incision, an ulnar-based zigzag incision over the flexor crease of the metacarpophalangeal joint provides better exposure. The incision is perhaps the most difficult part of the operation because the radial digital nerve crosses the midline at exactly this point. Also, because the radial digital nerve lies just beneath the skin, it can be divided easily while making the skin incision.

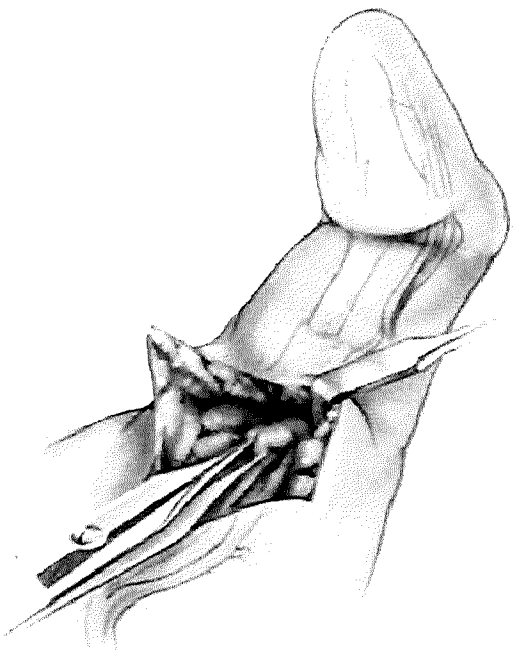

FIGURE 22-50. After the dermis is divided, small, sharp scissors or a hemostat is used to dissect out the radial digital nerve.

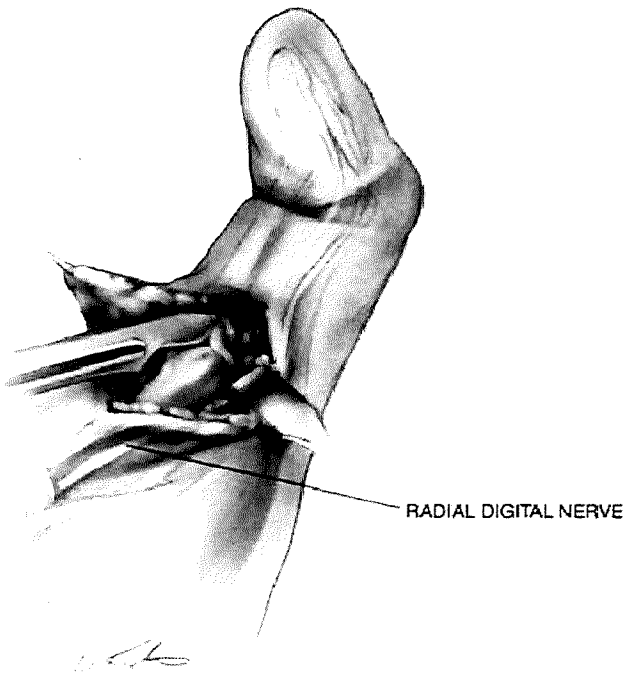

FIGURE 22-51. After it is safely extracted out of the way, the A-1 pulley is incised. It is usually not necessary to excise a portion of this pulley or shave the nodule, which will disappear after the release.

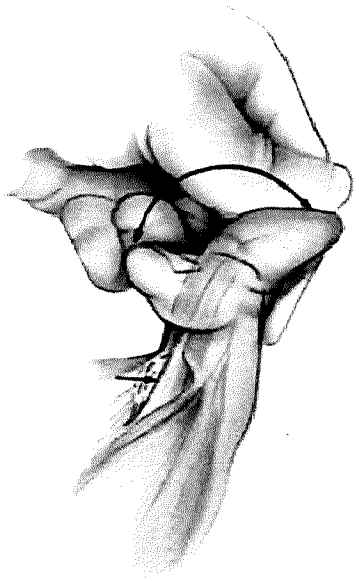

FIGURE 22-52. The thumb is extended fully to be certain that the release is complete. The release should not be extended too far distally so that the bow stringing of the flexor pollicis longus tendon is avoided. Only the skin is closed.

TABLE 22-3 Modified Blauth Classification of Congenital Thumb Hypoplasia

Type	CT scan/MRI findings
I	Small thumb with hypoplasia of abductor pollicus brevis and opponens pollicus
II	Narrow first web space, laxity of the ulnar collateral ligament of the metacarpophalangeal joint, thenar muscle hypoplasia
III	Global thenar weakness and metacarpophalangeal joint collateral ligament instability, partial aplasia of the first metacarpal, extrinsic weakness
IIIA	As in the previous category, with proximal metacarpal and carpometacarpal joints present
IIIB	Absent proximal metacarpal, with deficient carpometacarpal joint
IV	Pouce flottant (floating thumb); no bony support V Thumb aplasia

Findings are additive with increasing severity.
Adapted from Tada K, Kurisaki E, Yonenobu K, et al. Central polydactyly—a review of 12 cases and their surgical treatment. *J Hand Surg [Am]* 1982;7(5):460–465; Wood VE. Treatment of central polydactyly. *Clin Orthop Relat Res* 1971;74:196–205; Kay S. Camptodactyly. In: Green DP, Hotchkiss RN, Pederson WC. eds. *Green's operative hand surgery*, 4th ed. New York, Churchill-Livingstone, 1999:510, with permission.

in 53 children, 63% achieved spontaneous full IP joint extension with observation alone at 4 years after presentation.

Trigger digits are more often multiple, and can be associated with central nervous system disorders and syndromes (trisomy 18, mucopolysaccharidoses). The pathology appears to predominate at the decussation of the flexor tendons under the A2 pulley, and not at the A1 pulley alone. The triggering appears to occur as the flexor digitorum profundus passes through the chiasm of the flexor digitorum superficialis. Surgical recurrence following A1 pulley release alone is high in pediatric trigger digits. Further opening of the chiasm or resection of a slip of the flexor digitorum superficialis is often necessary to prevent recurrence (401, 402).

Release of Congenital Trigger Thumb Congenital trigger thumb is usually noted by the parent after birth (Figs. 22-49 to 22-52). In some cases it may be possible to extend the thumb, in which case a trial of nonoperative treatment is indicated (256). A course of observation is indicated in most circumstances because about 40% of the cases resolve and full motion results after surgery if it is performed in the first 3 years of life (398)

Hypoplasia/Aplasia of the Thumb. Children with thumb hypoplasia or aplasia will have deficient prehension and grasp. Thumb hypoplasia or aplasia can occur in isolation, or be associated with other radial-sided deficiency syndromes (Holt-Oram and Fanconi syndromes). It is seen universally in radial dysplasia. It is also common in other congenital malformations (277, 403), including those of the cardiac, craniofacial, musculoskeletal, renal, gastrointestinal, and hematopoietic systems. It may involve hypoplasia of the metacarpals (Cornelia de Lange syndrome and diastrophic dwarfism) or phalanges (Rubinstein-Taybi and Apert syndromes). The finding of a thumb deficiency in a neonate should prompt a thorough multiple-system examination for other malformations. In addition, a DEB screening for Fanconi anemia is recommended for all infants with radial-sided defects including thumb hypoplasia.

In general terms, the clinical manifestations of thumb hypoplasia include a contracted first web space, unstable MCP joint, thenar weakness, and interphalangeal joint stiffness or instability. Buck-Gramcko (404) and Manske et al. (405) have modified the Blauth (406) classification for thumb hypoplasia. This classification system is useful from the point of view of treatment considerations. There are five types of thumb hypoplasia in the Buck-Gramcko modification of the Blauth classification (Table 22-3). A type I thumb is essentially a normal thumb, except for diminished size. A type II thumb is even smaller and narrower, with a contracted first web space and thenar atrophy. Type III thumbs have marked atrophy or absence of both the intrinsic and the extrinsic musculature. The thumb is globally unstable and underdeveloped. Manske et al. (405) further subdivided type III thumbs into A and B categories. Type IIIA thumbs have a stable carpometacarpal (CMC) joint, whereas type IIIB thumbs have absence of the proximal metacarpal and trapezium. Type IIIB thumbs have no basilar joint stability (Fig. 22-53). Type IV thumbs are the classic *pouce flottant* or "floating thumbs." A type V demonstrates complete aplasia of the thumb.

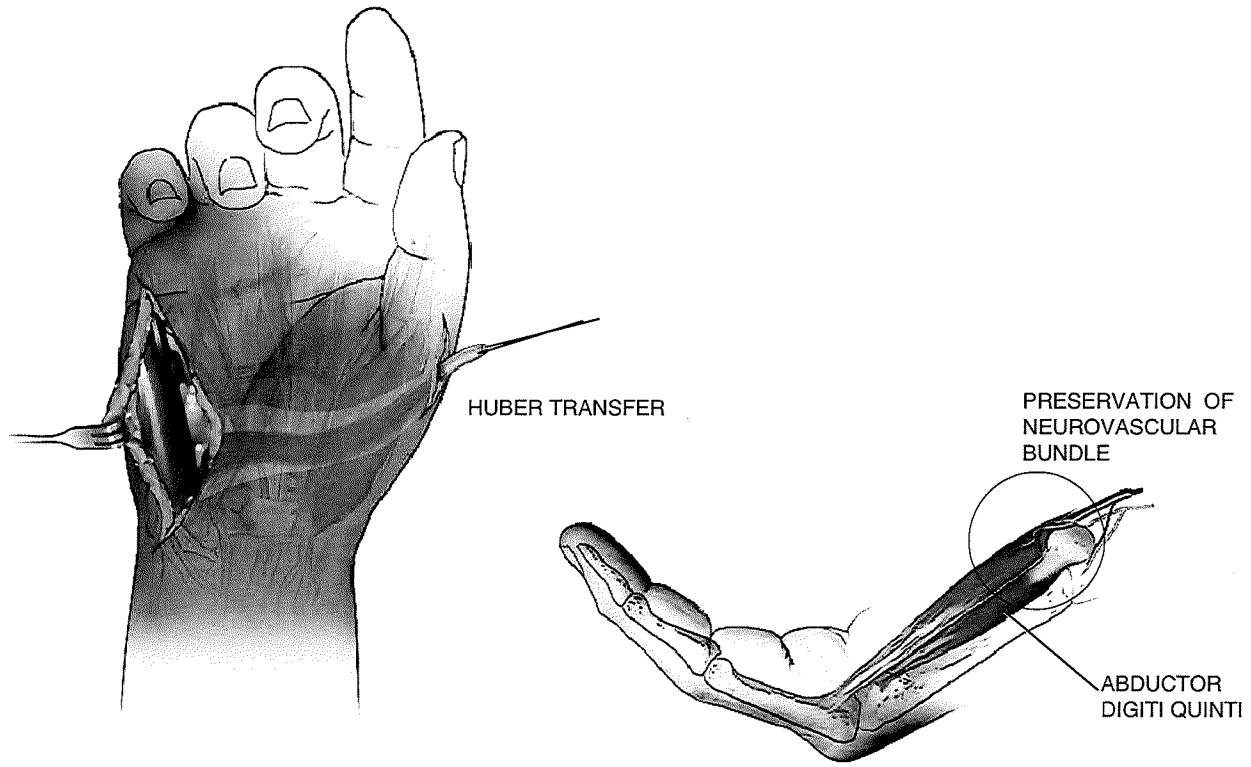

FIGURE 22-53. **A:** Illustration of Huber opponensplasty transfer of abductor digiti quinti to the abductor pollicus brevis for a hypoplastic thumb reconstruction. It is imperative that the neurovascular bundle be preserved proximally during the transfer. **B:** Postoperative clinical photograph of opposition pinch in a patient with a Huber opponensplasty reconstruction. (Figure courtesy of Children's Orthopaedic Surgical Foundation (COSF), © 2010.)

The pathoanatomy is dependent on the severity of the thumb hypoplasia. Universally, the thumb ray bones are smaller and narrower. The interphalangeal joint is usually underdeveloped and stiff. The first web space is contracted in all except the rare type I thumb. The MCP joint usually has ulnar collateral ligament insufficiency, but may be globally unstable. The thenar intrinsics are deficient, and they may be completely absent in the more severe forms of hypoplasia. The thumb's extrinsic musculature is progressively deficient in the classification scheme. The major anatomic determining factor for reconstruction is the status of the CMC joint. Type IIIB and type IV thumbs have no basilar joints. Plain radiographs are helpful in distinguishing the skeletal development, including the carpus and distal radius.

Treatment. The choice in the treatment of children with thumb hypoplasia is whether to do surgery or not. If left alone,

these children will adapt. They will use lateral pinch between the long and the index fingers. However, the deficiencies in pinch, grasp, and fine motor activities will be significant. Surgical reconstruction can improve function and aesthetics and is advised (407).

Reconstruction of the hypoplastic thumb, therefore, includes first web space deepening; opponensplasty, with either the abductor digiti quinti accessory extrinsic extensor (EIP or EDQ) the ring-finger flexor digitorum superficialis tendon; and MCP joint stabilization, with either ligamentous reconstruction or chondrodesis/arthrodesis. Blauth types I to IIIA should be reconstructed according to these principles (Fig. 22-53). The choice of first web-space deepening procedure includes two-part and four-part Z-plasties or the use of dorsal rotation flaps from the index finger, thumb, or hand. The degree of contracture determines the amount of skin necessary to provide a normal depth and breadth to the web for pinch and grasp activities. The abductor digiti quinti transfer for opposition can be used in the infant with a relatively stable MCP joint. Care must be taken to protect the proximal neurovascular pedicle to the abductor digiti quinti muscle, avoiding overzealous dissection near the pisiform (260). In older children, or in patients with marked instability of the MCP joint, the ring-finger flexor digitorum superficialis is used for opposition. After the tendon is sewn into the proximal phalanx and extensor hood, the additional tendon length can be used for MCP ligamentous reconstruction. In addition to the flexor digitorum superficialis tendon, local fascia from the adductor can be mobilized for ligamentous augmentation at the MCP joint. If soft-tissue procedures fail, or the instability at the MCP joint is too severe, fusion can be performed. The physis of the proximal phalanx should be preserved so as to maximize growth of the thumb ray. Chondrodesis of the metacarpal head to the epiphysis of the proximal phalanx is desirable in the young (408).

Thumb hypoplasia with the absence of a basal joint (type IIIB), *pouce flottant* (type IV), or aplasia of the thumb (type V) is a candidate for pollicization of the index finger. The major area of controversy is still the type IIIB thumb. In the absence of a CMC joint, the results of reconstruction have been disappointing (409). It is a difficult decision for the parents to accept pollicization in these children because of the relatively normal appearance of the thumb. However, reconstruction without a CMC joint leads to continued lateral pinch of the index and long fingers, rather than use of the reconstructed thumb. CMC joint reconstruction, with microvascular transfer of a toe metatarsophalangeal joint, has rarely been performed in children with type IIIB thumbs (409) as an alternative to pollicization in these children. Surgery is performed later and is quite extensive. Pollicization involves the conversion of the triphalangeal index finger, without a basilar joint and a narrow web space, to a biphalangeal thumb with a CMC joint and a deep first web space. It results in a four-digit hand (three fingers and thumb). The removal of the index metacarpal and the use of the index metacarpal epiphysis as the trapezium enable the surgeon to properly position and cover the thumb. The technique described by Buck-Gramcko is used most commonly (274, 275) utilized (Fig. 22-54). In the congenital absence of the thumb, this is better than microvascular toe transfer. Thumb reconstruction or pollicization is generally performed between 6 and 18 months of age. The quality of the pollicized digit is dependent on the quality of the original index finger, in terms of tendon function and joint motion. Patients with thumb aplasia and radial dysplasia generally do more poorly because the involved index finger has deficient musculature and camptodactyly. Manske and McCarroll (284) performed secondary opposition transfers in children with poor pollicizations. The best results are seen in children who have aplasia alone and normal index fingers (Fig. 22-55) (408, 410–413).

Thumb Duplication

Preaxial Polydactyly. Thumb duplication may be a misnomer because it implies that there are two normal thumbs whereas, in fact, both thumbs are hypoplastic. In isolation, thumb duplication is usually a sporadic occurrence. It is rarely associated with genetic syndromes, such as acrocephalopolysyndactyly (Nocack and Carpenter types) and Holt-Oram or Robinow syndromes. If it is associated with a triphalangeal thumb or with duplication of the great toe, it may be autosomal dominant, with variable penetrance (414). The locus for preaxial polydactyly has been mapped to chromosome 7q36 (415). Preaxial polydactyly is a rare occurrence, with an incidence estimated at 0.08 in 100,000 live births. There are many different classification systems, including the universal (416), Marks and Bayne (417), and Wassel (418) systems. Classification by the Wassel system is dependent on the number of bifid or duplicated phalanges or metacarpals, starting distally and progressing proximally (Fig. 22-55). A bifid distal phalanx is Wassel type I. A duplicated distal phalanx is type II (Fig. 22-56), constituting approximately 20% of all thumb duplications. A duplicated distal phalanx with a bifid proximal phalanx is type III. A thumb with duplicated proximal and distal phalanges is type IV, which is the most common type (approximately 40%) (Fig. 22-57). Duplicated proximal and distal phalanges with a bifid MCP (or MET) is type V. Duplication of all phalanges and metacarpals is type VI. Any duplication with a triphalangeal thumb is type VII, which accounts for approximately 20% of thumb duplications.

The pathoanatomy is dependent on the type of polydactyly. The nails, bones, joints, ligaments, muscles, tendons, nerves, and blood vessels are split between the two digits. In addition, there can be hypoplasia or aplasia of any of the normal anatomic elements of a thumb. Plain radiographs will generally provide definitive information regarding skeletal pathoanatomy. Careful surgical exploration will define the soft-tissue anatomy.

Treatment. Surgical reconstruction is the treatment of choice to improve function and aesthetics. Unlike postaxial polydactyly, ablation in the nursery is not recommended, even for the simplest preaxial polydactyly. Too often, the thenar musculature and collateral ligaments insert on the radius-based digit and can be lost with simple excision (419–421).

Treatment involves excision of the more hypoplastic thumb and reconstruction of the more developed thumb. Generally,

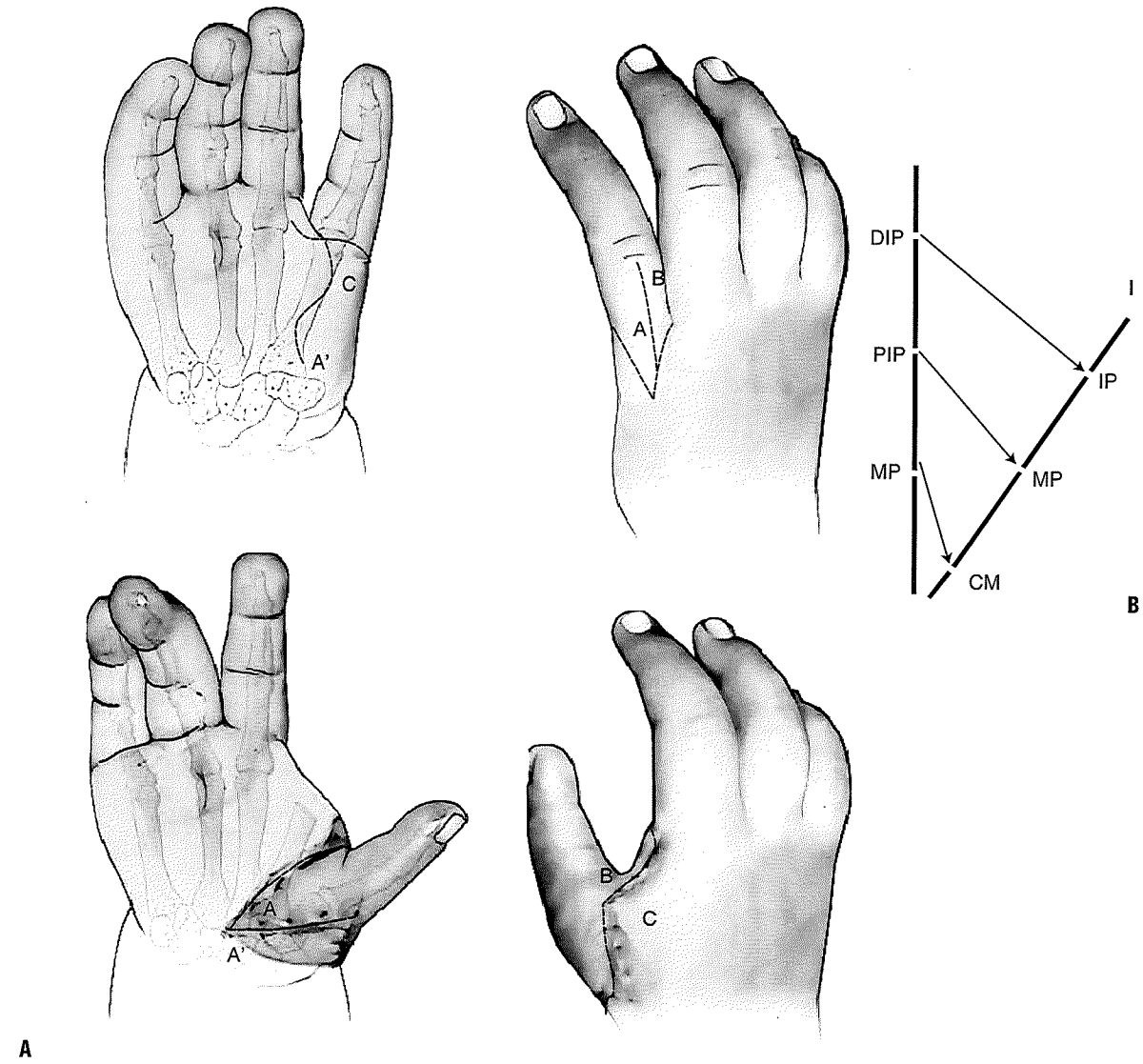

FIGURE 22-54. Illustrations of the pollicization procedure as popularized by Buck-Gramcko. **A:** Outlines show the skin incisions that provide first web-space flap coverage as the pollex is positioned for opposition. The triphalangeal index finger is converted into a biphalangeal thumb. The index metacarpal is excised, except for the distal epiphysis. **B:** The changes in joints from the index finger to the thumb and the tendon transfers to provide opposition and pinch function are as follows: extensor indicis proprius → extensor pollicis longus; extensor digitorum II → abductor pollicis longus; interosseus palmaris I →adductor pollicis; interosseus dorsalis I → abductor pollicis brevis. (*DIP*, distal interphalangeal; *PIP*, proximal interphalangeal; *MP*, metacarpophalangeal; *IP*, interphalangeal; *CM*, carpometacarpal; *I*, number of finger.)

the radius-based digit is excised. The soft-tissue elements usually bifurcate at the level of the skeletal split. Transfers of the shared or aberrant tendons, nerves, and ligaments to the reconstructed thumb are necessary in order to maximize outcome.

The extensor pollicis longus tendon is usually bifid. The flexor pollicis longus usually inserts on the ulnar thumb, although it can be bifid or insert on the radial thumb. The thenar musculature and the radial collateral ligament to the MCP joint usually insert radially, especially in the common type IV polydactyly. These need to be transposed. The radial digital nerve may be present only in the radial thumb and should be transposed to the radial aspect of the reconstructed thumb. The bifid proximal phalanx and the metacarpal should be excised in types II and IV, respectively. Primary phalangeal or metacarpal osteotomy is indicated if there is axial malalignment. The more developed thumb often has a larger nail and distal phalanx. However, some Wassel type II malformations have almost equally sized distal phalanges and nails (Fig. 22-46). Surgical recombination of the distal phalanx and nail beds (Bilhaut-Cloquet procedure) in type I and II malformations has been disappointing because of poor nails, loss of joint motion, and physeal closure. It still appears best to accept the

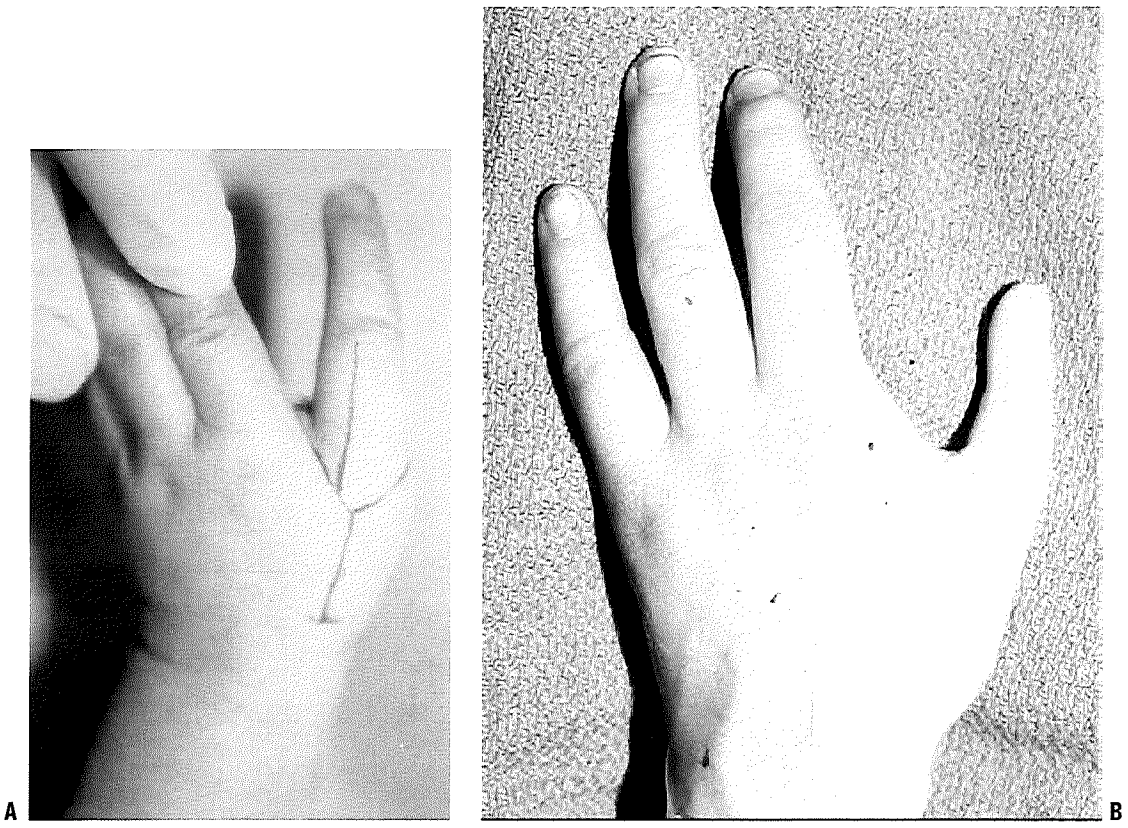

FIGURE 22-55. **A:** A 1-year-old child with thumb aplasia. The surgical incisions for index pollicization are outlined. These flaps provide for deep first web space. **B:** Long-term follow-up clinical photograph of a pollicization. (Figure courtesy of Children's Orthopaedic Surgical Foundation (COSF), © 2010.)

more hypoplastic distal phalanx than to consider recombination of the nail and phalanx (422). Adduction contracture of the first web space should be treated with Z-plasty to deepen the web space. The need for additional surgery with growth may be as high as 40% of cases.

Angular deformity of the proximal and distal phalanges into a zig-zag posture is the most common problem (423–426). Reconstruction with osteotomy, tendon transfers, ligament reconstruction, or arthrodesis may be necessary to improve both function and cosmesis.

Excision of Duplicate Thumb. Because polydactyly is the most common congenital anomaly of the hand, most orthopaedic surgeons encounter this anomaly. Duplication of the

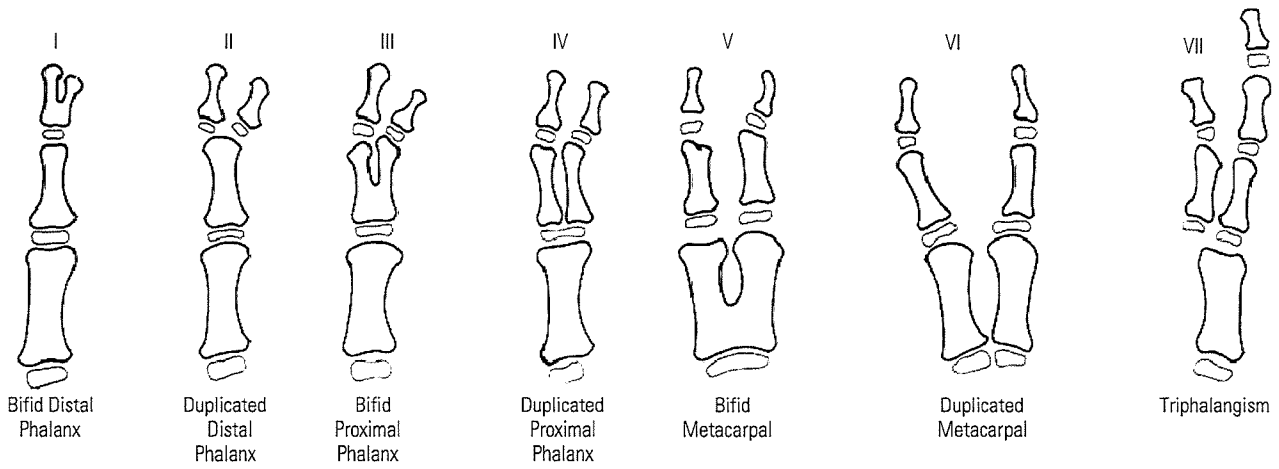

FIGURE 22-56. Wassel classification of thumb duplications. Type IV is the most common, with an incidence of 40%. Types II and VII have an incidence of approximately 20% each.

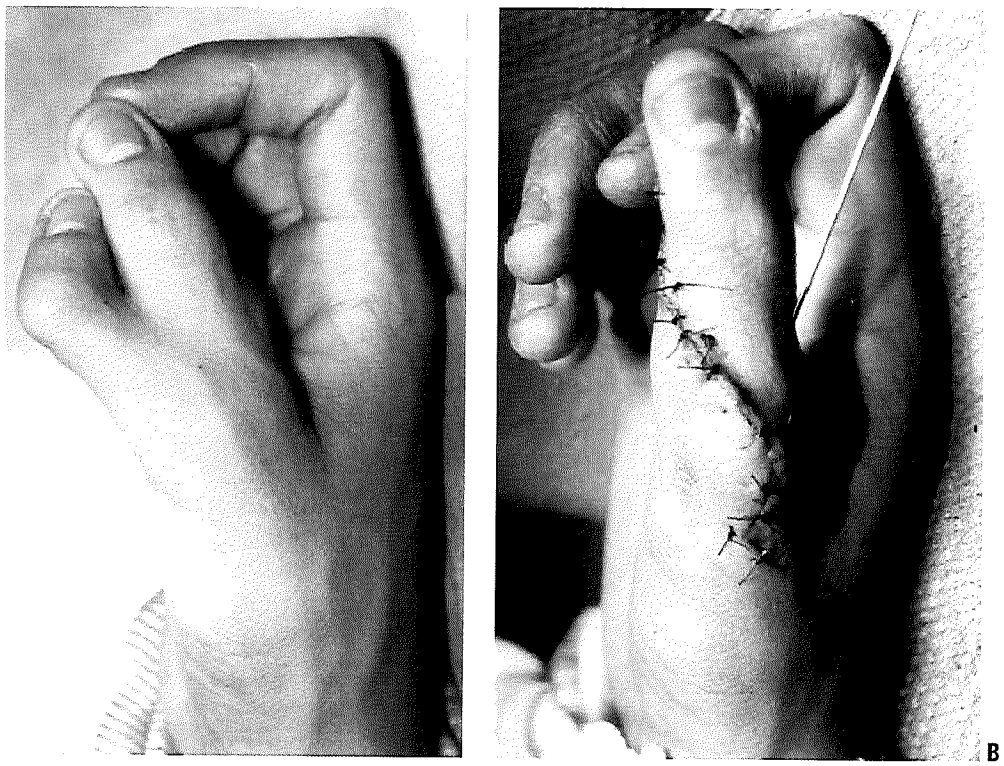

FIGURE 22-57. **A:** An adolescent with the more common Wassel type IV thumb duplication. **B:** Results at the conclusion of the surgical reconstruction. Surgery included excision of the radial proximal and distal duplicated digits, transfer of the thenars and metacarpophalangeal joint radial collateral ligament to the reconstructed thumb, and contouring of the skin by excision of redundancy and Z-plasties.

thumb, or preaxial polydactyly, however is not common, and the treatment of this condition may hold a trap for the physician who is unaware of its unique anatomy. Because of the anatomic reasons discussed in this section, simple excision of the smaller thumb rarely produces a satisfactory result (427).

Selecting which digit to remove may be difficult if both are of nearly the same size and function. Both function and cosmesis should be considered. In general, the radial digit is the smaller digit and is removed. This confers the advantage of leaving the ulnar collateral ligament intact for stability during pinch. Even though both thumbs have their own flexor and extensor motor action, the thenar musculature attaches to the most radial digit. It is therefore necessary that these attachments be preserved and reattached to the remaining digit. If the ulnar digit is removed, it is necessary to preserve a periosteal and ligamentous flap from the radial border of this digit to reconstruct an ulnocollateral ligament for the remaining digit (Figs. 22-58 to 22-63).

The wide variety of anomalies encountered under the term *duplicate thumb* makes careful assessment and planning imperative. Wassel (418) has classified these anomalies into seven types, whereas Marks and Bayne (417) have devised a more simple classification based on the level of duplication. These authors and others have discussed the principles involved as well as the results (426, 428). The surgeon must plan an incision that will not leave a linear scar that will later contract, correct all rotational and angular deformity in the retained digit, carefully plan a reconstruction of the joint that will impart lasting stability, centralize the flexor and extensor tendons, and plan to keep the attachments of the thenar muscles on the retained digit. These principles are illustrated on the most common type of thumb duplication: a Wassel type IV or a Marks and Bayne type I, in which there is complete duplication of the proximal phalanx.

Triphalangeal Thumbs. Triphalangeal thumbs are usually inherited in an autosomal dominant manner. This is true whether they are associated with thumb polydactyly (429) or are seen in isolation. In inherited isolated triphalangeal thumbs, the genetic marker has been localized to chromosomal region 7q36 (430). The extra phalanx is the middle phalanx. It may be wedge-shaped or rectangular. The thumb may be in a position of opposition or in the plane of motion of the other fingers. The latter situation may indicate an index finger duplication with an absent thumb. This concept is supported not only by the clinical and radiographic appearance of the most radial digit but also by the dermatoglyphics (431). The triphalangeal thumbs that were studied contained the radial loops normally seen in the index finger and not in the thumb.

Triphalangeal thumbs may be associated with musculoskeletal malformations, such as cleft feet and preaxial polydactyly (432); congenital heart disease, including Holt-Oram syndrome (433); hematopoietic abnormalities such as Fanconi and Blackfan-Diamond syndromes (434, 435); and imperforate anus (436).

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Excision of Duplicate Thumb (Figs. 22-58 to 22-63)

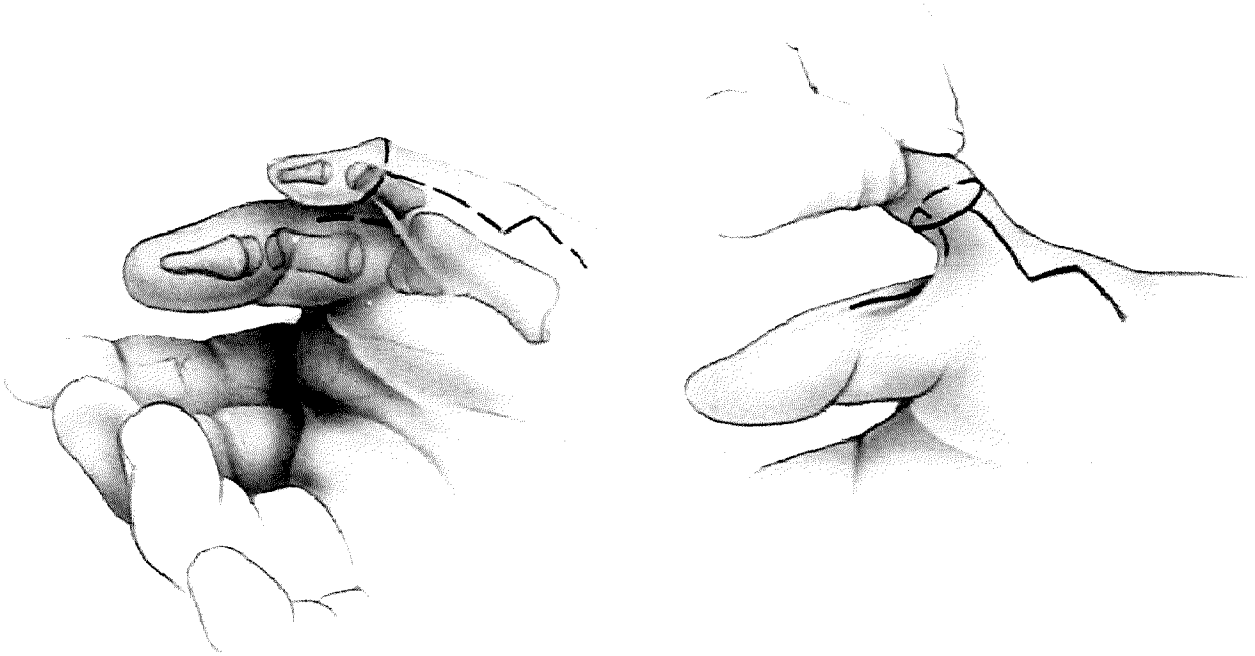
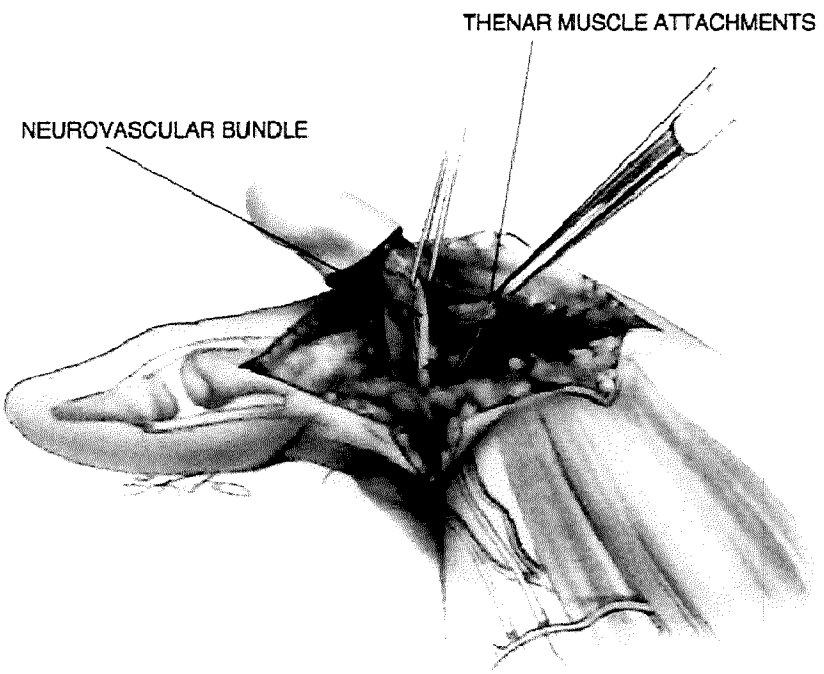


FIGURE 22-58. Excision of Duplicate Thumb. The incision is planned so that a straight scar is avoided. The incision illustrated has committed the surgeon to removal of the radial thumb. This incision permits exposure of all the structures to both thumbs. If the surgeon is uncertain at the beginning of the case about which thumb is to be retained, a different incision should be planned. This situation might arise if both digits are small or there is a question of the blood supply to the digit that has to be retained.

FIGURE 22-59. After the flaps are developed, the neurovascular bundles are identified and traced to their respective digits. This is to ensure that they are protected and the digit to be retained is innervated and vascularized. At this point, the thenar muscles are detached from the base of the radial digit. These will attach to the radial side of the radial digit by a broad tendon. Sufficient tendon and, if necessary, periosteum should be retained with the muscles to provide strong attachment to the retained digit.



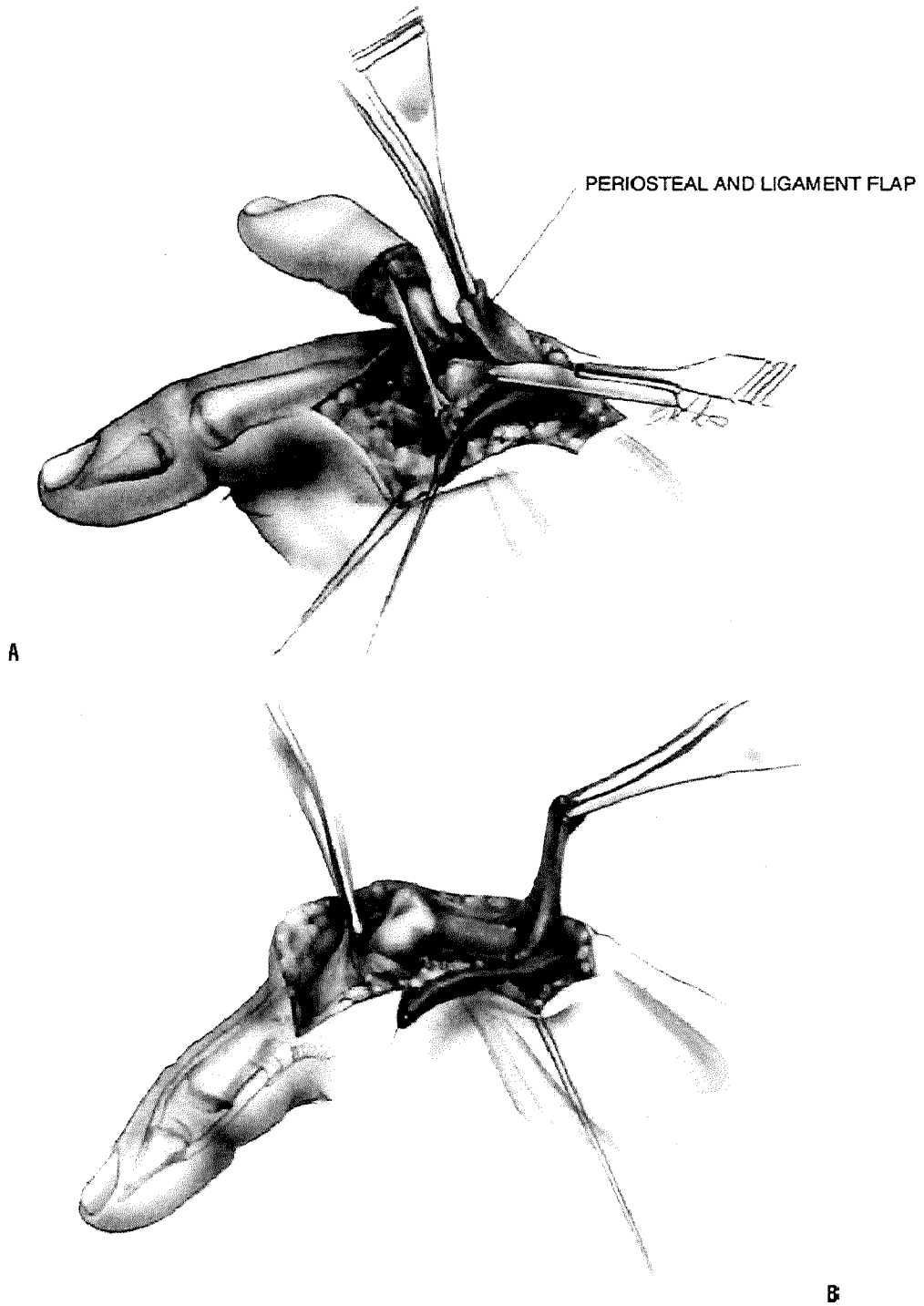


FIGURE 22-60. With the thenar muscles detached and retracted out of the way, a flap of periosteum and ligament from the radial digit is raised. This flap **(A)** is then dissected proximally off the metacarpal and sutured into the radial side of the retained digit to reconstruct the radial collateral ligament. This step is important to avoid ulnar deviation, which is one of the most common complications of this procedure. The radial digit is now removed and the remaining ulnar digit subluxated to demonstrate the condyle of the metacarpophalangeal joint **(B)**. This condyle is broader than normal and must be narrowed to provide good cosmesis as well as good stability for the retained digit. There is usually a small ridge on the articular surface of the condyle that identifies that portion of the condyle on which each of the thumbs articulated. In addition, there is frequently an ulnar deviation of the metacarpal head. In keeping with the principles outlined, it is necessary to correct this deviation at this time.

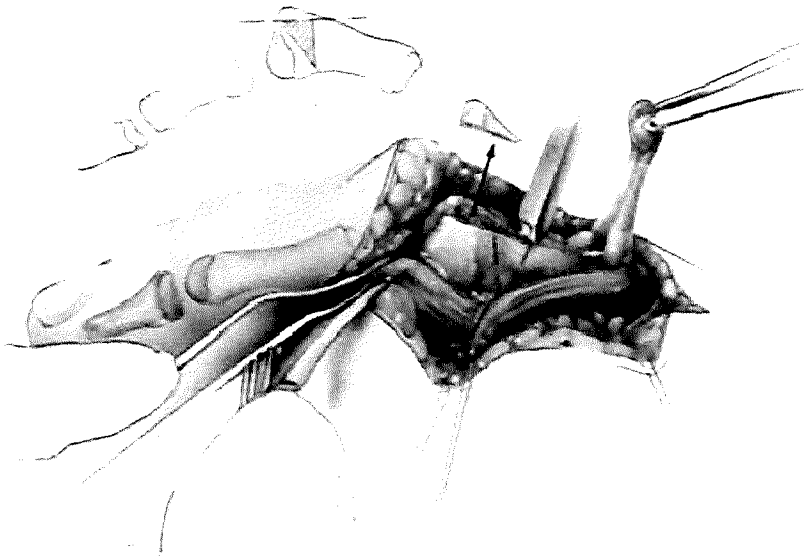


FIGURE 22-61. The excess radial portion of the condyle is removed with a small osteotome. A small closing wedge osteotomy is made just behind the condyle of the metacarpal. This is designed to correct the ulnar deviation. Any rotational malalignment can also be corrected. This osteotomy usually is fixed with one small Kirschner wire passed from the tip of the distal phalanx.

FIGURE 22-62. When the osteotomy is fixed, the periosteal and ligamentous flap that was raised from the discarded digit and radial side of the metacarpal is sutured to the radial side of the retained digit. Care should be taken in adjusting the tension of this repair. Subsequently, the thenar muscles are sutured over this to the base of the retained digit.

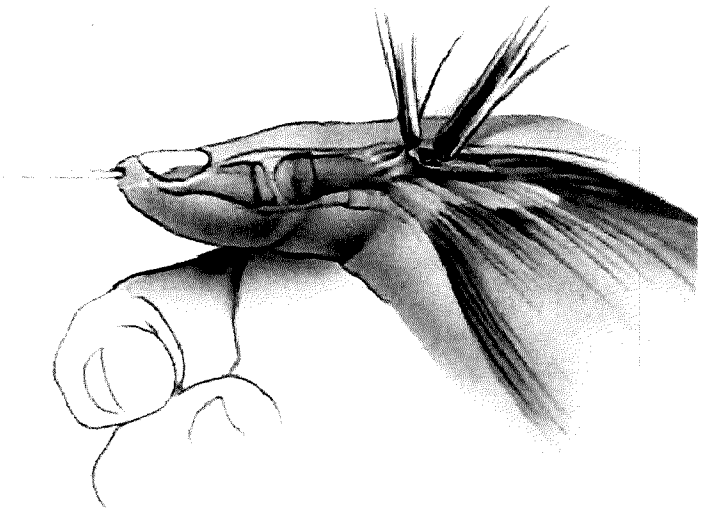
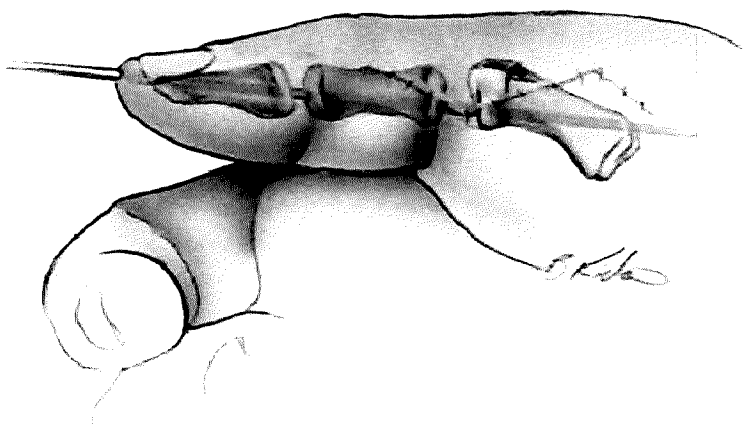


FIGURE 22-63. The skin is closed. There is no problem with a shortage of skin. There is often excess skin that may be trimmed. The resulting suture line should not be linear. A rigid dressing and a long arm cast are applied.



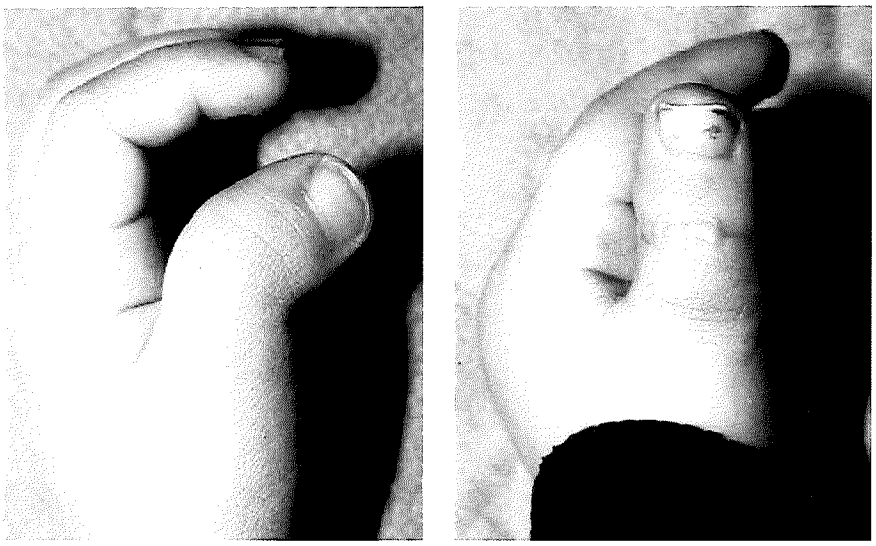


FIGURE 22-64. **A:** Triphalangeal thumb with a delta phalanx and a marked radial deviation deformity. **B:** Postoperative photograph after excision of the delta phalanx and rotation flap reconstruction of the soft tissues.

Classification and pathoanatomy are dependent on the type of triphalangism. Type I involves a delta middle phalanx with radial deviation deformity (Fig. 22-64). Type II involves a normal middle phalanx, but an opposable thumb. Type III is an index-finger duplication with all digits in the same plane. In type I and II triphalangism, the first web space is normal. In the five-fingered hand (type III), there is a contracted first web space that limits prehension. Similarly, usually the thenar musculature is normal in type I and II triphalangeal thumbs, whereas it is absent in type III triphalangism. In addition, the triphalangeal thumb may be hypoplastic and have associated extrinsic musculature weakness.

Treatment. Rarely, patients with triphalangeal thumbs prefer not to undergo surgical reconstruction. Patients with significant cosmetic abnormalities and limitation of pinch prefer reconstruction for both functional and cosmetic reasons. A malaligned and elongated triphalangeal thumb in the same plane as the other digits is cosmetically anomalous and functionally limited for prehension activities.

Depending on the type of triphalangeal thumb, surgery may involve web-space deepening, excision of the extra phalanx (437), opposition transfer (438), or a modified pollicization procedure (437, 439, 440). The delta phalanx is usually excised to correct the length and the angular deformity of the type I triphalangeal thumb (Fig. 22-64). If this procedure is performed in infancy, usually a stable interphalangeal joint can be reconstructed (441). In older children, or in children with abnormal phalanges and interphalangeal joints, a combination of shortening osteotomy and arthrodesis is preferred. In these situations, physeal growth of a biphalangeal digit should be preserved. In the five-fingered hand, a modified pollicization procedure is necessary so as to provide a deep first web space and an opposable thumb.

TRAUMATIC INJURIES

Fractures to the pediatric hand are commonplace, accounting for approximately one-fourth of all childhood fractures (442). The two peak ages for these fractures are adolescence (from sport-related activities) and infancy (from crush injuries). Most fractures are nondisplaced, nonphyseal injuries. Physeal injuries, however, can account for up to 40% of finger fractures (443, 444), with a Salter-Harris II fracture of the small-finger proximal phalanx being the most common. Most pediatric hand fractures do well regardless of treatment (445). Malunion and growth disturbance are rare (443, 446). However, there is a subset of pediatric hand injuries that will do poorly if not recognized and treated appropriately. The purpose of this section is to review fractures that are problematic and require surgical treatment.

Overview. The bones of the digits have only one secondary center of ossification. These appear between birth and 3 to 4 years of age. The epiphyses of the phalanges are proximal. The epiphyses of the metacarpals are distal, except for the thumb, in which the physis is proximal. Distally in the thumb, there can be a second epiphysis or pseudoepiphysis. Ossification of the phalangeal condyles is progressive with growth, but in preschool children the condyles may be predominantly cartilaginous. Radiographic evaluation of injuries in young children may be difficult because of the chondral nature of the epiphysis and the intra-articular portions of the condyles. It is important to obtain true anteroposterior and lateral radiographs of the injured digits. In a diagnostically confusing situation, MRI scans of the fingers and hand should be performed.

To protect the anatomic healing of a traumatized digit in a young child, maximal protection is necessary. It is often necessary to protect the preschool-age child with a long-arm mitten cast. The older child often needs a short-arm mitten cast. Single-finger splinting is difficult to maintain, even in an adolescent. If the fracture is painful, or if it requires immobilization to maintain reduction, casting of the entire hand is appropriate. Fortunately, most pediatric fractures are nondisplaced and stable (447). The outcome will be successful regardless of immobilization technique. In these situations, the change is made to simple buddy taping as soon as

it is comfortable. However, it is imperative not to mistake a problematic injury for a simple one and treat it with benign neglect. Such a mistake will lead to long-term loss of alignment, motion, and function, which may not be salvageable by secondary surgical reconstruction.

Distal Phalanx Injuries

Nail-plate Injuries, Physeal Fractures, Mallet Fingers, and Tip Amputations. Most injuries to the distal phalangeal region are secondary to crush injury. These injuries are most common in the toddler and preschool age groups. The mechanism of injury is usually a digit caught in a door. Adults, often parents, are frequently involved in the accident, which makes the situation emotionally charged. The injuries can include partial or complete amputation, nail-bed laceration, and distal phalangeal fracture. All of these sites of trauma need to be addressed in the care of the child. In addition, time and energy need to be spent in helping the family cope with the emotional trauma.

Tip Amputations. A distal phalangeal crush injury in a toddler usually includes a partial amputation with a nail-bed laceration and a distal phalangeal tuft fracture (Fig. 22-65). The fractures are generally minor avulsions that heal without problems. The partial amputation often extends dorsally through the nail bed, leaving some volar pulp intact. The volar soft-tissue attachments maintain the vascularity to the distal tip. Meticulous repair under conscious sedation and/or local anesthesia usually leads to normal long-term outcome. The technique involves initial repair of the eponychial folds to properly align and stabilize the digit. It is imperative to then meticulously repair the nail-bed laceration with fine, absorbable suture (i.e., 6-0 chromic suture) and loupe magnification to prevent long-term nail deformity. The dorsal roof of the eponychium is preserved by placement of a spacer for several days after repair. Even when there is nearly complete amputation, with apparently nonviable distal tissue, soft-tissue repair almost always leads to survival of the tip. Uncomplicated repair of the partial amputation and nail-bed laceration will generally heal without permanent damage to the nail or phalanx. However, neglected nail-bed injuries are associated with a high rate of permanent deformity (441, 448).

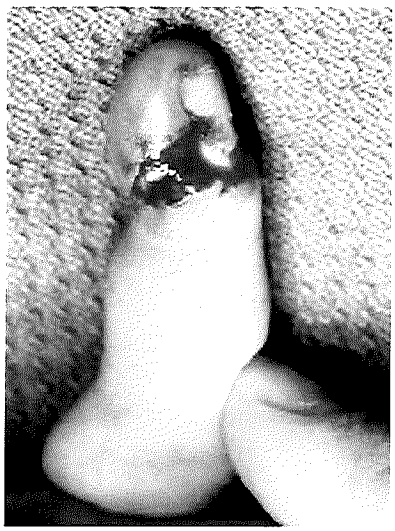


FIGURE 22-65. Typical distal phalangeal crush injury with nail and pulp laceration. Repair requires nail-plate removal, absorbable suture repair of the lacerated nail bed, and anatomic closure of the eponychial skin. (Figure courtesy of Children's Orthopaedic Surgical Foundation (COSF), © 2010.)

Care of the complete amputation at the level of the nail, or distally, is more controversial. First, this is a much more difficult situation emotionally for the child and the family. The injury often occurs while the child is under adult supervision. There is usually significant stress and guilt associated with the amputation. The parents universally want the tissue replaced. However, this injury is beyond the trifurcation of the digital arteries near the level of the DIP joint, and so the tissue is not typically replantable. The debate concerns whether to suture the amputated part back on without vascular anastomoses (a composite graft) or to allow healing by secondary intention (449). Replacement of the amputated soft tissue initially makes the family feel better, but usually is not necessary. The amount of soft tissue amputated is small. The physis is proximal and not affected. Subsequent growth will be normal. The overall digital length, therefore, will be nearly normal. The cosmetics of composite grafting and healing by secondary intention are generally equal in the long term. Long-term sensibility and function seem to be equivalent. In addition, in the short term, replacement of the amputated part may be more stressful for the family and the child, with multiple dressing changes, superficial necrosis of the distal tip, and slow healing. Therefore, if the soft-tissue loss is minor, reassurance to the family of the long-term result and treatment with serial dressing changes are best. This is true as well for the situation in which a minimal portion of exposed bone is debrided. However, if the piece includes the eponychium or the entire sterile and germinal matrix of the nail, composite grafting of the amputated part is preferred. There is a chance that this will heal and preclude secondary reconstructive surgery.

Physeal Fractures and Nail-bed Entrapment. Some children will present with distal laceration and flexion posture of the distal phalanx, or mallet appearance. Radiographs will reveal a displaced physeal fracture with dorsal widening. Too often, these children are diagnosed as having mallet finger and treated with splinting. This is not an extensor tendon disruption. The extensor mechanism is intact because the terminal tendon inserts into the more proximal epiphysis. The deformity is caused by entrapment of the proximal nail bed (germinal matrix) in the physeal fracture site (450, 451), the so-called Seymour's fracture. If not recognized early, the open injury can become secondarily infected. The clinical appearance of the finger and the radiographic physeal changes may be interpreted as distal phalangeal osteomyelitis. However, antibiotic treatment alone or surgical debridement of the distal phalanx is not the proper treatment

for this late-presenting fracture. Surgical repair of the nail bed is necessary in order to prevent long-term nail and distal phalanx growth problems. Under local, regional, or general anesthesia, the nail plate should be removed. By flexing the distal phalanx, the surgeon can gently extract the entrapped nail bed. The germinal matrix should be meticulously repaired so as to avoid long-term nail-plate problems. At times, this requires more proximal exposure by raising an eponychial flap. After repair of the nail bed, the nail plate can be replaced to preserve the dorsal roof of the eponychium and to provide an internal splint for repair of the fracture (450, 452). Placement of a cautery hole in the nail plate will lessen the risk of subsequent hematoma and paronychial infection.

Mallet Fingers. True mallet fingers are rare in the preadolescent child. In this age group, physeal fracture is more common. In the adolescent, mallet injuries with disruption of the extensor mechanism are more common and adult-like. As long as there is no entrapment of the germinal matrix, as noted in the preceding text, these injuries can be treated with immobilization. Rarely, the entire epiphysis can be displaced with the extensor mechanism (453, 454). If recognized early, this injury should be treated with open reduction of the dislocated epiphysis. The rare, chronic mallet finger in the young child has been treated successfully with tenodermodesis (455, 456).

Phalanx Fractures

Phalangeal Neck Fractures. Phalangeal neck fractures are problematic. They are usually caused by crush injury. As the child attempts to extract the affected digit, the condyles become entrapped and a fracture occurs in the subcondylar region (433, 442, 443, 447, 457, 458). The condylar fragment displaces into extension and often malrotates. The fragment is tethered by the collateral ligaments as it rotates dorsally up to 90 degrees. The condylar fragment is small and has a precarious blood supply through the collateral ligaments. The subcondylar fossa is obliterated, blocking interphalangeal flexion (Fig. 22-66). If not properly recognized and treated, complications of malunion, loss of motion, and avascular necrosis can occur. Too often, the severity of the fracture is underappreciated in the urgent care setting. The patient presents late, with inappropriate immobilization and a significantly healed fracture. In addition, the fracture is unstable and will often displace after closed reduction (459). The treatment of choice is closed reduction and percutaneous pinning (460). In a young child, this can be accomplished with a single oblique pin. In an older child, crossed pins prevent malrotation. Placement of the pins in the distal fragment requires careful localization of the fragment and avoidance of the extensor mechanism. If open reduction is necessary, the collateral ligaments should not be dissected from the distal fragment. Careful dissection lessens the risk of avascular necrosis.

When there is marked callus, open reduction may cause avascular necrosis. The fracture has generally been healing too long for successful closed reduction and percutaneous pinning to be carried out. If there is still lucency along the fracture line, percutaneous osteoclasis can be performed (461). A pin is placed dorsally in the fracture site under fluoroscopic control and used for reducing the fracture. The subcondylar fossa can be reconstituted, and the fracture can be pinned percutaneously.

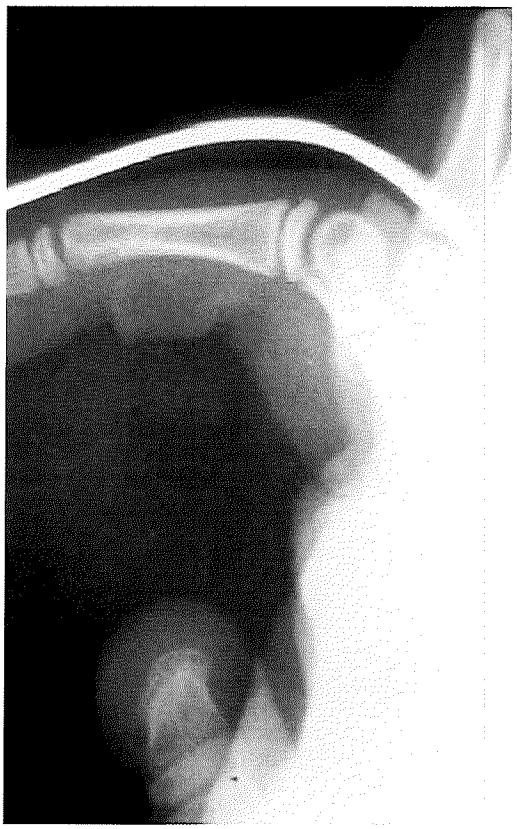


FIGURE 22-66. Displaced phalangeal neck fracture with loss of the subchondral fossa. This leads to loss of digital flexion. This fracture requires prompt attention, anatomic reduction, and pin stabilization. If the fracture is left in the position shown here in the splint, there will be a problematic malunion. (Figure courtesy of Children's Orthopaedic Surgical Foundation (COSF), © 2010.)

If the fracture is completely healed upon referral, late subchondral fossa reconstruction can be performed if there is a bony block to flexion (457). An average of 90 degrees of PIP joint flexion has been obtained with subchondral fossa reconstruction (457). Remodeling of the fracture is rare because of the significant distance from the physis, but it has been described in case reports of both proximal and middle phalanx phalangeal neck fractures (462). Observation may be appropriate, but only if the patient is young, there is only malangulation and not malrotation, and the family is willing to wait for 1 or 2 years.

Intercondylar Fractures. Intercondylar fractures in young children are often small osteochondral fractures. These carry a high risk of nonunion, malunion, and avascular necrosis. This is particularly true in the middle phalanx if the injury is a crush injury that alters the local blood supply. The fracture is intraarticular, generally displaced, and requires anatomic reduction for a successful outcome. Most often, the fracture has to be

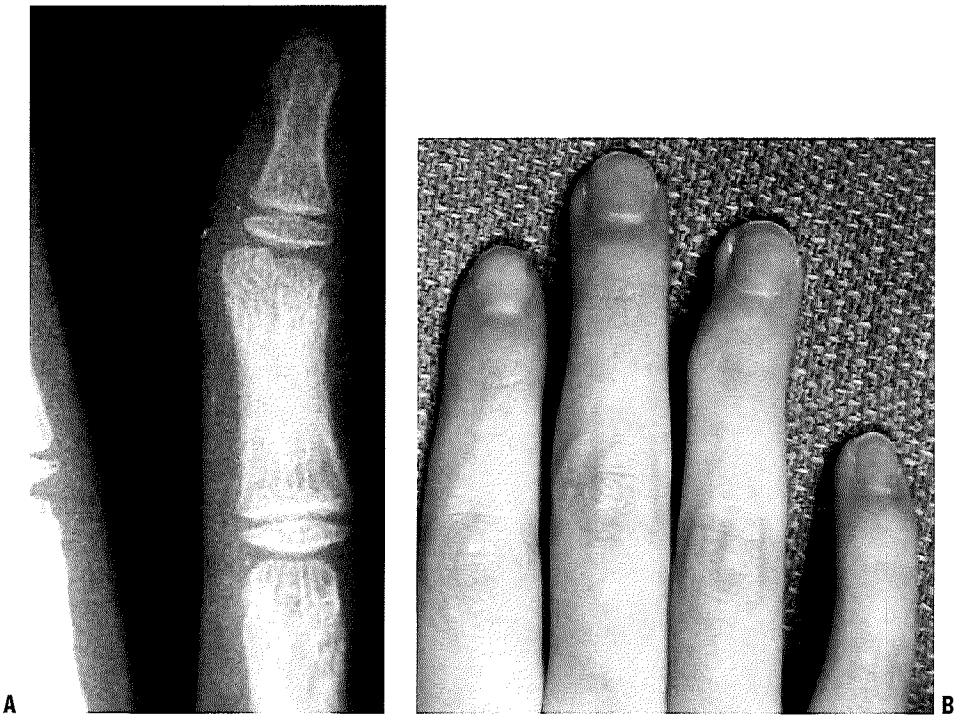


FIGURE 22-67. A: Radiograph of a displaced intercondylar fracture with articular malunion. **B:** Clinical photograph of a similar patient. This injury requires acute anatomic reduction and pin stabilization to prevent long-term loss of motion, malalignment, and potential pain and arthritis. (Figure courtesy of Children's Orthopaedic Surgical Foundation (COSF), © 2010.)

treated with open reduction. The collateral ligament attachments to the fragment are preserved so as to lessen the risk of avascular necrosis. On occasion, bone grafting is necessary for maintaining articular congruity and to prevent collapse. Even with well-performed open reduction, complications of this fracture can occur in the young patient. Avascular necrosis usually resolves by revascularization, but often not before collapse. Articular malunion can occur (Fig. 22-67). Loss of motion may not limit function.

In the adolescent, treatment of intercondylar fractures is similar to that in adults. Anatomic reduction and pin fixation are necessary to restore the joint surface and to prevent loss of reduction that can occur with this unstable fracture. Often, the procedure can be performed closed, using distraction and a percutaneous towel clip to obtain reduction (463). Open reduction can be performed with a volar, midaxial, or dorsal approach. This is appropriate for fractures that cannot be reduced closed or for comminuted fractures. Restoration of an anatomic joint is the mandatory goal and will lessen the risk of loss of motion, malalignment, or long-term arthritis.

Diaphysis-level phalangeal fractures are rare in the young child and more common in the teenager. The major issue in these fractures is malrotation (443) (Fig. 22-68). Frequently, children will not actively move the finger in the acute setting to allow for accurate assessment of digital alignment. However, close inspection of the nail plates will reveal the digital malalignment. In addition, the examiner can test digital alignment by tenodesis of the wrist. With passive wrist extension, the fingers flex and point toward the volar scaphoid tubercle. Digital alignment is generally symmetric. Children will tolerate this test, even when they are in too much pain or are too frightened to actively move their digits. Tenodesis assessment should be performed on all phalangeal and metacarpal fractures, regardless of radiographic appearance. If closed treatment is chosen, a finger should never be immobilized by itself, but should be secured to the adjacent digits to prevent subsequent loss of reduction and malrotation.

If the fracture is malrotated and unstable, reduction with pin or screw stabilization is necessary (463, 464). Although malrotation is uncommon, it is a major problem if missed until after healing. The malrotated digit impairs the function of the adjacent digits because the digits will overlap in flexion. At that stage, malrotation can be corrected only with osteotomy.

Physeal Fractures. Physeal fractures constitute 30% to 70% of pediatric finger fractures (442–444, 446). A Salter-Harris II fracture of the small finger is the most common of these fractures. Closed reduction of the abducted fracture is performed in MCP flexion to tension the collateral ligaments and effectuate ligamentotaxis. The surgeon's thumb or a cylindrical object such as a pencil can be used as a fulcrum. Postreduction stability is maintained by taping the digit loosely to the adjacent digit and applying a short-arm mitten cast. The less common type III physeal fracture requires open reduction if there is more than 2 mm of diastasis or articular step-off.

Metacarpal Fractures. Distal metacarpal metaphyseal (boxer) fractures are common in adolescence. The mechanism is axial load to a closed fist, most often to the small-finger metacarpal. These fractures are usually juxtaphyseal and malaligned, with apex dorsal angulation. In the acute setting, closed reduction and cast immobilization with three-point mold for

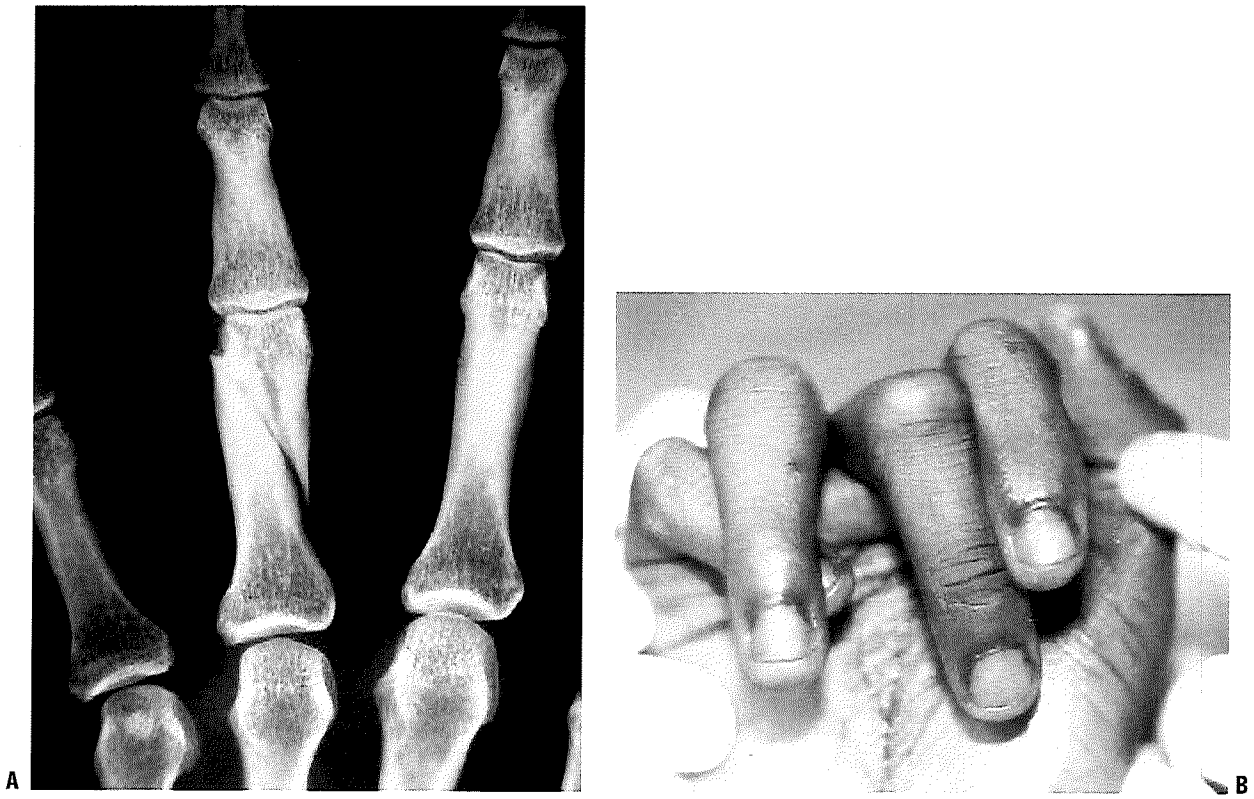


FIGURE 22-68. **A:** Displaced proximal phalanx fracture with malrotation. **B:** Tenodesis testing reveals malrotation. By passively extending and flexing the wrist, the digits will passively flex and extend, respectively. Malalignment is evident. (Figure courtesy of Children's Orthopaedic Surgical Foundation (COSF), © 2010.)

3 weeks are preferred for displaced fractures. This includes volar-to-dorsal pressure on the metacarpal head and dorsal-to-volar pressure on the more proximal shaft. A number of prospective randomized studies have demonstrated that casting with the MCP and IP joints extended ("pancake casts") is efficacious and safe, with minimal risks of contractures and stiffness when used in younger patients for 3 to 4 weeks duration (465, 466). Often, these patients will not seek medical attention until there is significant healing. Fortunately, the fracture is adjacent to the distal metacarpal physis, and the flexion malunion can remodel if there is sufficient growth remaining. This fact has led some clinicians to approach these fractures with neglect to allow for remodeling. Indeed, depending on the age of the patient, remodeling of the flexion deformity can occur. However, malrotation will not remodel. In addition, if remodeling is slow or fails to occur, the prominence of the metacarpal head in the palm can be limiting, an apparent extensor lag may develop, and a pseudoclaw appearance due to MCP joint hyperextension can occur.

Unstable or multiple metacarpal fracture(s) are rare. They should be treated with closed reduction and percutaneous pinning. Two smooth pins are placed, under fluoroscopic control, from ulnar to radial, distal to the fracture site(s), from the 5th to the 4th or the 3rd metacarpal, as necessary. Late open reduction of the juxtaphyseal fracture carries the risk of physeal injury and should be avoided. Diaphyseal fractures of the fifth metacarpal carry a higher risk of a malunion that will not remodel. Closed reduction and transverse percutaneous pinning to the adjacent metacarpals, or intramedullary pin fixation, is the treatment of choice. Open reduction with interfragmentary compression screws or plate-and-screw constructs may also be considered in appropriate fracture patterns. Corrective osteotomy may be necessary in the severe, malunited diaphyseal fracture that fails to remodel the flexion deformity with growth.

The major issue in other diaphyseal metacarpal fractures, especially if there are multiple metacarpal fractures, is malrotation. Active digital motion or passive tenodesis of the wrist will reveal malrotation. Anatomic reduction and pin, screw, or plate fixation will correct the malrotation.

Thumb Fractures. The unique features of fractures of the thumb are seen in Salter-Harris III fractures of the proximal phalanx and at the base of metacarpal fractures. The Salter-Harris type III physeal fracture of the thumb proximal phalanx is the skeletally immature equivalent of an adult ulnar collateral ligament disruption (467) (Fig. 22-69). These fractures require open reduction and internal fixation in order to restore stability of the joint and anatomic alignment of the joint and physis. During surgical exposure, the surgeon should remember that the ligament is intact. Therefore, after adductor takedown, the MCP joint should be exposed through the fracture site rather than through inadvertent incision of the ligament. The long-term results of anatomic open reduction are excellent.

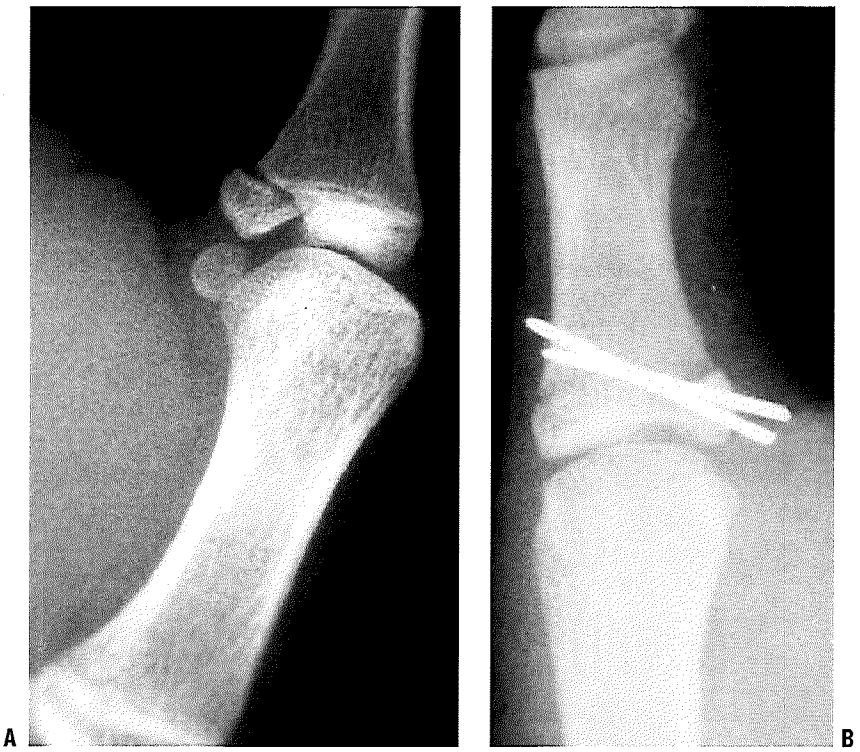


FIGURE 22-69. A: Salter-Harris III displaced proximal phalanx fracture of the thumb. This fracture requires open reduction and internal fixation in order to restore articular congruity and ligamentous stability. **B:** Postoperative radiograph of a similar patient with pins in place. (Figures courtesy of Children's Orthopaedic Surgical Foundation (COSF), © 2010.)

Metaphyseal fractures at the base of the thumb metacarpal often displace (468). Immobilization and observation, even in displaced fractures, are appropriate as long as there are at least 2 years of growth remaining because the malunited dorsoradial prominence will remodel during the ensuing 6 to 12 months (Fig. 22-70). Most parents would prefer to wait for remodeling rather than have operative reduction and pinning. In the markedly displaced and unstable fracture, closed reduction and percutaneous pinning are appropriate. The presence of a Bennett fracture with articular malalignment requires anatomic alignment of the intra-articular component and pin fixation of the thumb metacarpal to the adjacent second metacarpal and carpus.

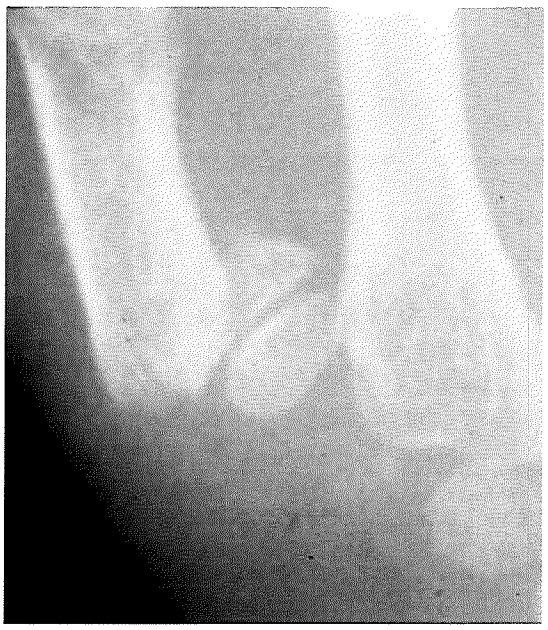


FIGURE 22-70. Displaced base in a metacarpal thumb fracture. Because this fracture is juxtaphyseal, adjacent to the carpometacarpal joint with universal motion, its remodeling potential is almost unlimited if the patient is younger than 10 years. Treatment choices are closed reduction and pinning, or immobilization in a cast to allow for biologic remodeling for the ensuing 6 to 12 months of growth. This patient was treated in a cast. (Figure courtesy of Children's Orthopaedic Surgical Foundation (COSF), © 2010.)

Wrist Injuries

Scaphoid Fractures. Scaphoid fractures occur from a fall on an outstretched wrist. The pain is often mild, rather than the severe pain a child or a parent expects from an acute fracture. This leads some patients and families to ignore the acute injury and not present until late. When the child presents at the acute stage of the injury, there will be tenderness to palpation over the anatomic snuff-box (the region dorsoradially at the wrist between the extensor pollicis longus and the extensor pollicis brevis tendons), over the volar scaphoid tubercle, and upon axial compression of the thumb CMC joint. Radiographs may reveal the fracture. The best view is an anteroposterior view in 30 degrees of ulnar deviation (scaphoid view). If the radiographs are diagnostic for the fracture, long-arm immobilization in a thumb spica cast can be considered

for nondisplaced injuries. If the radiographs are negative, but the child has tenderness at the site of the injury, as noted in the preceding text, protected immobilization for 2 weeks is advised. Repeat clinical and radiographic examination should be performed out of cast 2 weeks later if tenderness persists. If there is still doubt about the diagnosis, MRI or CT scan is advised. The CT scan shows the bony alignment better, but the MRI gives added information about the vascularity to the proximal pole, and better information about the cartilage surfaces in a young child. CT scans are used for assessing acute fractures in adolescents. MRI scans are used for assessing the acute injury in the child younger than 10 years of age. In the acute setting, MRI scans have been diagnostic for the exact site of injury, and can lead to appropriate treatment early (469, 470).

Previously, it was reported that most scaphoid fractures in the skeletally immature patient were distal pole or avulsion fractures (471). Distal pole fractures heal readily with cast immobilization without risk of nonunion or avascular necrosis. However, scaphoid wrist fractures are now more common in the adolescent age group. These fractures can displace and carry the same risks of nonunion and avascular necrosis in the child as they do in the adult. Therefore, nonunions are becoming more commonplace (472, 473). Treatment of an established nonunion in a child should be with open reduction, bone grafting, and internal fixation. Internal fixation screws have been used in children with both acute displaced wrist fractures and established nonunions (462, 472, 473) (Fig. 22-71). It is difficult to determine whether a bipartite scaphoid, even if bilateral, is congenital or posttraumatic. However, if it is symptomatic, it should be treated in the same way as a traumatic nonunion. The success of open reduction, bone grafting, and internal fixation for a scaphoid nonunion is high in children (472, 473). Proximal pole fractures and nonunions have now been described in children and adolescents (108). These fracture nonunions have been treated successfully with vascularized bone graft from the distal radius to the scaphoid.

Wrist Pain, Triangular Fibrocartilage Complex Tears, and Ligamentous Injuries

Atraumatic Ligamentous Instability. Most adolescents with chronic wrist pain have overuse injuries. These patients often have generalized ligamentous laxity or a hyperelasticity syndrome. The wrist pain is similar to the patellofemoral knee pain and multidirectional shoulder instability pain seen in this age group. Overuse, growth, and resultant muscle weakness all contribute to instability of the joints. On physical examination, there is often systemic evidence of generalized laxity (elbow cubitus valgus, pes planus, knee hyperextension, passive hyperextension of the index finger parallel to the dorsal forearm, and thumb abduction to the volar forearm). At the wrist, there will be increased midcarpal translation to volar and dorsal applied stress. This passive midcarpal instability will be equivalent on both the affected and the unaffected wrist. However, the affected side may often have painful clicking. Plain radiographs may reveal a volar tilt to the lunate on the lateral view. MRI scans and arthrograms appear normal. These children respond to alteration of activities and strengthening, as in other growth-related overuse injuries in the teenager. It is the authors' experience that resistive strengthening in a neutral wrist position with therapeutic putty is best.

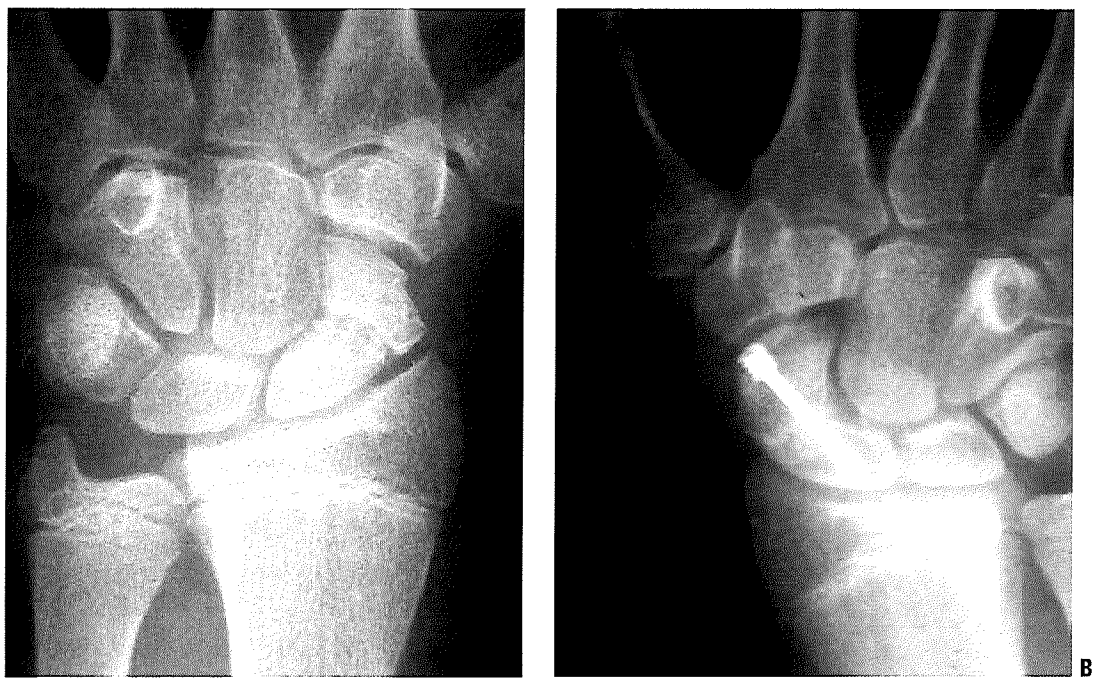


FIGURE 22-71. **A:** Scaphoid nonunion in an adolescent. **B:** Postoperative iliac crest bone grafting and internal fixation for a similar scaphoid fracture nonunion. (Figures courtesy of Children's Orthopaedic Surgical Foundation (COSF), © 2010.)

Traumatic Scapholunate Ligamentous Injuries. Posttraumatic injuries to the scapholunate ligament rarely occur in children. Most descriptions of these injuries have been case reports (474). However, traumatic ligamentous injuries are occurring more frequently with today's higher level of athletic competition in this age group. On physical examination, the tenderness is usually more focal than in atraumatic ligamentous laxity wrist pain. Asymmetric clicking with applied ligamentous stress testing is common. Static injuries are more common in adults, and will show an increased scapholunate distance and a flexed scaphoid on plain radiographs. Most scapholunate injuries in children are dynamic injuries, with normal plain radiographs. In addition, the scapholunate distance in children has been difficult to interpret because of eccentric ossification and the chondral nature of the carpus in the young. There is an age standard for scapholunate distances in both sexes on pediatric wrist radiographs (475). MRI scans may reveal the ligamentous injury and will reveal chondral to chondral distances. Arthroscopic examination of the wrist is diagnostic and often therapeutic. Many of these injuries are partial tears of the ligament, with an associated chondral lesion of the lunate fossa of the radius. This leads to mechanical impingement that often responds favorably to arthroscopic debridement. It is rare that there will be a complete ligamentous disruption in this age group. However, symptomatic complete scapholunate disruptions need to be treated with ligamentous reconstruction.

Triangular Fibrocartilage Complex Tears. Posttraumatic chronic wrist pain that does not respond to prolonged rest and therapy should be evaluated for intra-articular pathology. As in scaphoid fractures and intercarpal ligamentous injuries, the epidemiology of TFCC tears has changed to include the adolescent. Physical examination for TFCC tears includes ulnocarpal compressive testing, lunate-triquetral stress testing, and stress testing of the distal radioulnar joint. Painful clicking with these maneuvers, especially if asymmetric, may be indicative of a tear. However, many children with nondissociative laxity show similar findings on physical examination. Plain radiographs are normal. The MRI may be diagnostic, but there has been a high incidence of false-negative readings for adolescent TFCC tears. In skilled hands, arthroscopy is definitive for diagnosis, and often for treatment (Fig. 22-72).

TFCC tears do occur in the skeletally immature patient. Most tears are associated with nonunion of ulnar styloid fractures, radial growth, ulnar overgrowth, and/or ulnocarpal impaction syndrome. Isolated tears also occur. In adolescents and children, these tears are usually peripheral tears that respond well to surgical repair (476). Isolated tears can be repaired arthroscopically. Tears associated with radial and ulnar bony deformities are repaired at the time of corrective osteotomy (Fig. 22-73). Treatment should also include appropriate excision of the ulnar styloid nonunion, shortening of the impacting ulna, osteotomy of the deformed radius, and stabilization of the distal radioulnar joint, if necessary (476).

Dislocations. Most hyperextension injuries to the interphalangeal joints in children result in a tear of the volar plate. There may be associated minimal Salter-Harris III physeal avulsions of the adjacent phalanx (Fig. 22-74). Most injuries are stable. Treatment should be brief immobilization for comfort, followed by buddy taping and early range-of-motion exercises until the patient is asymptomatic. Prolonged treatment with splints or casts can lead to PIP joint stiffness. True interphalangeal dislocations occur less often. The dislocation is usually dorsal, and it occurs more commonly at the PIP joint than at the DIP joint. Closed reduction with distraction and dorsal-to-volar manipulation are generally successful. Rarely, a displaced epiphysis, flexor tendon, or interposed volar plate can block reduction, demanding an open reduction procedure.

MCP joint dislocation of the thumb or the index finger can be simple or complex (460). Simple dislocations are reducible in the emergency setting. Complex dislocations are irreducible and have an interposed volar plate blocking closed reduction. Plain radiographic evidence of widening and lateralization of

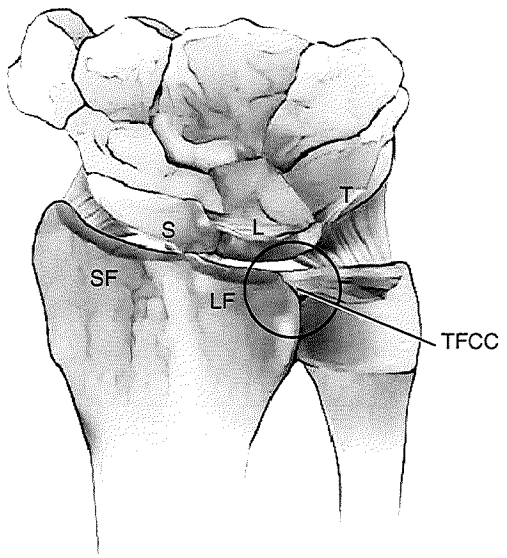


FIGURE 22-72. A: Illustration of intra-articular wrist anatomy, as seen from the dorsal 3/4 portal. (*S*, scaphoid; *L*, lunate; *SF*, scaphoid fossa; *LF*, lunate fossa; *T*, triquetrum; *TFCC*, triangular fibrocartilage complex.)

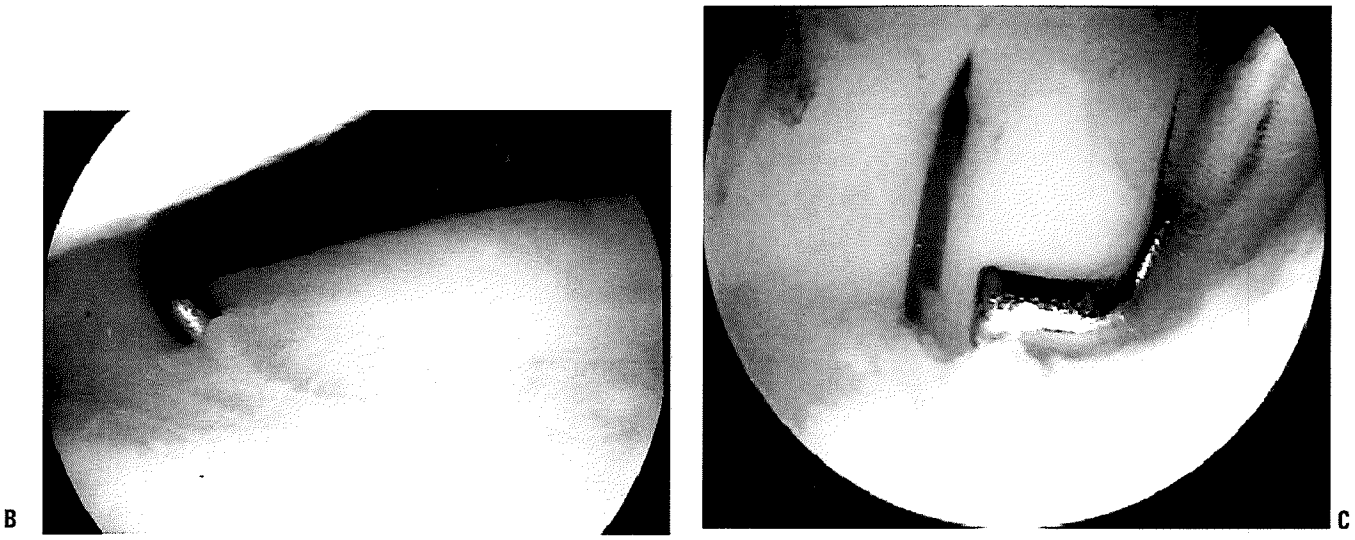


FIGURE 22-72. *(Continued)* **B:** Arthroscopic photograph of a triangular fibrocartilage complex (TFCC) peripheral tear. The tear is along the peripheral edge of the TFCC, where the blood supply enters and aids in healing. In this situation, repair is by arthroscopic suture techniques. (Figure courtesy of Children's Orthopaedic Surgical Foundation (COSF), © 2010.) **C:** Intraoperative photograph of arthroscopic suture repair of peripheral TFCC tear. (Figure courtesy of Children's Orthopaedic Surgical Foundation (COSF), © 2010.)

the joint, as well as bayonet apposition of the proximal phalanx and metacarpal, indicates an irreducible situation.

Open reduction of a complex dislocation can be performed through a volar (477, 478) or a dorsal (479, 480) approach. In the volar approach, the radial neurovascular bundle is tented just beneath the skin by the metacarpal head. It is imperative to be cautious with the skin incision so as not to cause laceration. In either approach, the volar plate should be incised so as to allow reduction of the joint and anatomic realignment of the flexor tendons, sesamoids, and collateral ligaments. Postoperative treatment is by early protected motion with buddy taping and extension block splinting. Chronic instability is rare, but

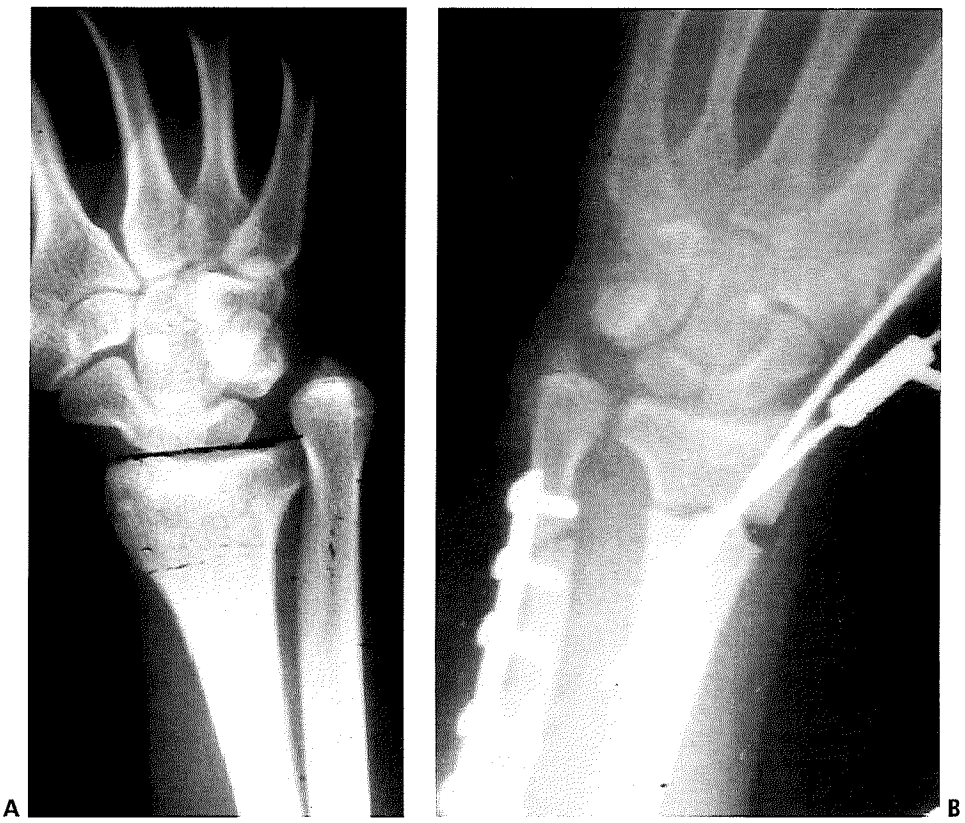


FIGURE 22-73. A: Radiograph of ulnocarpal impaction associated with radial growth arrest, ulnar overgrowth, and a TFCC tear. **B:** Repair of the TFCC, ulnar shortening, and radial osteotomy with bone grafting were performed. (Figures courtesy of Children's Orthopaedic Surgical Foundation (COSF), © 2010.)

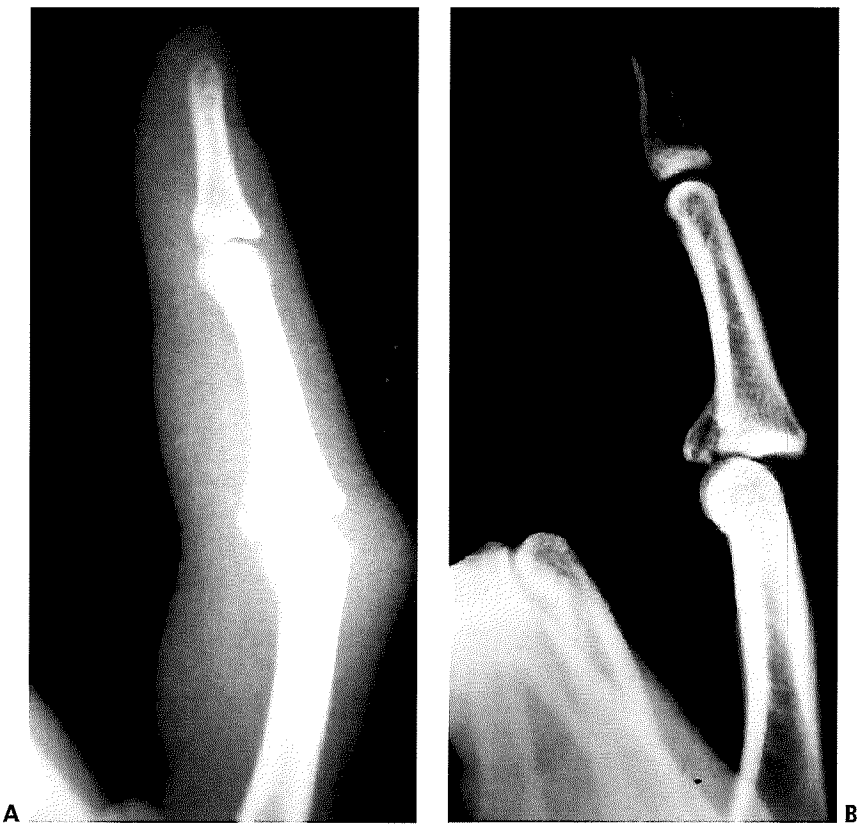


FIGURE 22-74. Volar plate injuries. **A:** Nondisplaced minor volar and dorsal avulsion injuries. This injury requires minimal immobilization and prompt initiation of range-of-motion exercises to minimize the risk of permanent flexion contracture. **B:** This injury has a more significant fracture fragment and joint subluxation. It requires anatomic reduction of the proximal interphalangeal joint and an extension block splint to allow for motion within the range of joint stability. (Figures courtesy of Children's Orthopaedic Surgical Foundation (COSF), © 2010.)

limitation of MCP motion is not. The digital neurapraxia secondary to the dislocation will resolve in this age group.

Tendon Lacerations. The classification system, in terms of zones of injury for flexor tendon lacerations, is the same for the child as it is for the adult. The diagnosis of flexor tendon injury, operative care, and postoperative rehabilitation may be more difficult in the child. This is especially true in the toddler and preschool-aged child, whose ability for cooperation is limited. Often, the presenting digital cascade and digital excursion with wrist tenodesis serve as the basis for the diagnosis of flexor tendon laceration (Fig. 22-75). If in doubt, the clinician should explore the wound under anesthesia. Repair of the tendon lacerations in zones I and II requires meticulous technique with fine sutures. Repair can be performed electively in the first 1 to 2 weeks with equivalent results. However, if there is any concern regarding the vascular status of the digit, repair should be emergent, with exploration of the digital neurovascular bundles. In the infant, the core suture may be as fine as 6-0 and the epitenon suture may be 8-0. Postoperative immobilization in a cast for 4 weeks is effective protection (481, 482). Subsequent rehabilitation is necessary in order to regain maximal passive and active motion. Recently, there has been growing enthusiasm for early motion rehabilitation protocols when appropriate repairs are performed (483).

There have been no differences in total active motion (TAM) between early mobilization protocols and cast immobilization for 4 weeks in children younger than 15 years (481). The results of isolated profundus tendon lacerations in zone I averaged 90% to 94% of normal TAM. Isolated profundus lacerations in zone I averaged 71% to 78% TAM. Combined superficialis and profundus lacerations in zone II averaged 72% TAM. However, if cast immobilization continued beyond 4 weeks, there was a significant decrease in TAM, to 40% by 6 weeks. There was no difference in the results according to age groups from birth to 15 years.

Associated nerve or palmar plate injuries diminished the results slightly. Postoperative tendon rupture is rare. Two-stage reconstruction of unrecognized zone II lacerations in children younger than 6 years has had poorer results than in adults, with a higher rate of complications and a mean TAM of approximately 60% of normal. Results were better with supervised rehabilitation (484).

The principles of the treatment of extensor tendon lacerations in the adult apply to the child as well. Direct repair in both the emergency room under sedation and the operating room under general anesthesia is feasible. Cast immobilization in a protected position of wrist dorsiflexion and digital extension is continued for 4 weeks after repair. Results are excellent with primary repair. Associated fractures, dislocations, and flexor tendon injuries impair the results.

Amputations. Complete or partial amputations of the distal fingertip are very common in children, and have been discussed in detail in the section on distal phalangeal injuries. Treatment of more proximal, complete digital amputations with replantation in children as young as 1 year of age is now standard. In children, the indications for replantation are more liberal than in adults, and include multiple-digit, thumb, midpalm, hand, and

FIGURE 22-75. Photograph of an altered digital flexion cascade with passive wrist extension tenodesis. This is diagnostic of a flexor tendon laceration in the long finger. (Figure courtesy of Children's Orthopaedic Surgical Foundation (COSF), © 2010.)

distal forearm amputations as well as single-digit amputations in zones I and II. Crush amputations from doors, heavy objects, or bicycle chains have a peak incidence at 5 years, whereas sharp amputations occur more commonly in adolescence. Digital survival rates from replantation range from 69% to 89% in pediatric series. More favorable digital survival was seen with sharp amputations, body weight >11 kg, more than one vein repaired, bone shortening, interosseous wire fixation, and vein grafting of arteries and veins. Vessel size generally exceeds 0.8 mm in digital replants in children and is not a technical problem for the skilled microvascular surgeon. Index- and long-finger replants have better survival than small-finger replants in children. A finger survival rate of 95% was seen in children if prompt reperfusion occurred after arterial repair with at least one successful venous anastomosis, compared with zero survival if one or both of these factors was absent (485). Neural recovery rates far exceed those cited in adults, with return of two-point discrimination of <5 mm often present. Tenolysis may rarely be necessary after tendon repair. Two-stage flexor tendon reconstruction in children has a higher rate of complications than in adults. Growth arrest or deformity is more common if there is a crush component to the amputation. These digits are rarely normal after replanting, although the results in children are better than in adults, in terms of sensibility and recovery of range of motion. Microvascular toe-to-thumb transfer is a very successful alternative to pollicization in the case of a failed thumb replant in a young child (486) (Fig. 22-76).

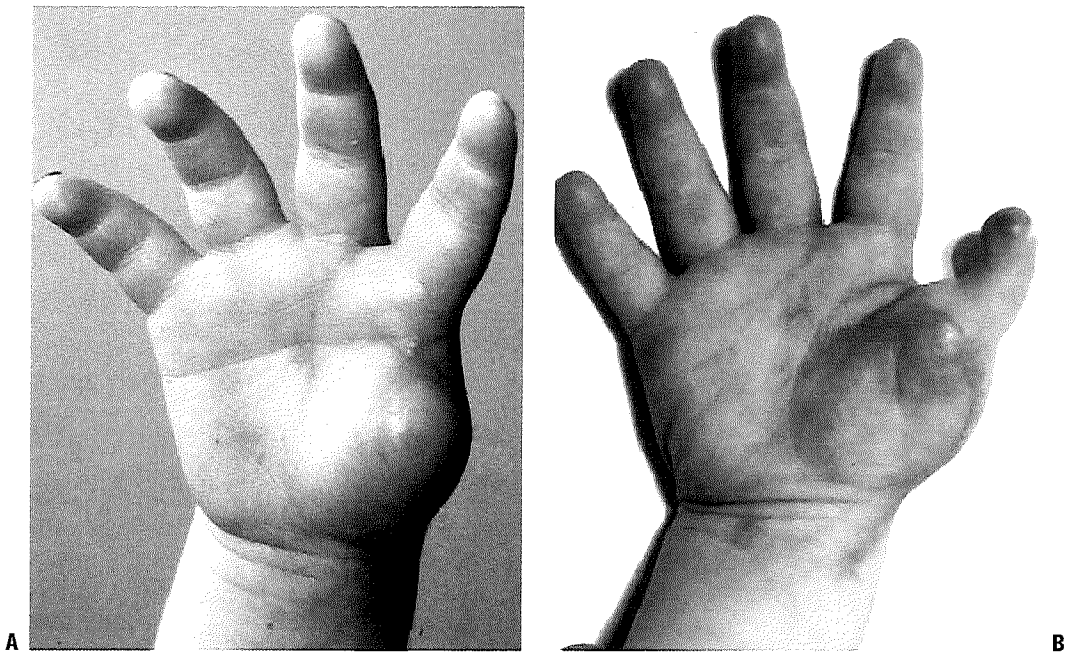


FIGURE 22-76. **A:** Traumatic amputation of the thumb in a 2-year-old child. **B:** Postoperative photograph of the same patient after microvascular toe-to-thumb transfer for thumb reconstruction. (Figures courtesy of Children's Orthopaedic Surgical Foundation (COSF), © 2010.)

REFERENCES

1. Flatt AE. *The care of congenital hand anomalies.* St. Louis, MO: Quality Medical Publishing, 1994.
2. McCarroll HR. Congenital anomalies: a 25-year overview. *J Hand Surg Am* 2000;25:1007–1037.
3. Saunders JWJ, Cairns JM, Gasseling MT. The role of the apical ridge of ectoderm in the differentiation of the morphological structure and inductive specificity of limb parts in the chick. *J Morphol* 1957;101:57.
4. Ulthoff HK. *The embryology of the human locomotor system.* Berlin, Germany: Springer-Verlag, 1990.
5. Tabin C. The initiation of the limb bud: growth factors, Hox genes, and retinoids. *Cell* 1995;80:671–674.
6. Omi M, Ude H. Effects of digit tissue on cell death and ectopic cartilage formation in the interdigital zone in chick leg buds. *Dev Growth Differ* 1996;38:419.
7. Saunders JW Jr. The proximo-distal sequence of origin of the parts of the chick wing and the role of the ectoderm 1948. *J Exp Zoo* 1998;282: 628–668.
8. Solursh M. Ectoderm as a determinant of early tissue pattern in the limb bud. *Cell Differ* 1984;15:17–24.
9. Summerbell D. A quantitative analysis of the effect of excision of the AER from the chick limb-bud. *J Embryol Exp Morphol* 1974;32:651–660.
10. Summerbell D. The zone of polarizing activity: evidence for a role in normal chick limb morphogenesis. *J Embryol Exp Morphol* 1979;50: 217–233.
11. Tickle C. Experimental embryology as applied to the upper limb. *J Hand Surg Br* 1987;12:294–300.
12. Goodman FR. Limb malformations and the human HOX genes. *Am J Med Genet* 2002;112:256–265.
13. Muragaki Y, Mundlos S, Upton J, et al. Altered growth and branching patterns in synpolydactyly caused by mutations in HOXD13. [see comment]. *Science* 1996;272:548–551.
14. Lamb DW, Wynne-Davies R, Soto L. An estimate of the population frequency of congenital malformations of the upper limb. *J Hand Surg Am* 1982;7:557–562.
15. Conway H, Bowe J. Congenital deformities of hand. *Plast Reconstr Surg* 1956;18:286.
16. Philipp T, Philipp K, Reiner A, et al. Embryoscopic and cytogenetic analysis of 233 missed abortions: factors involved in the pathogenesis of developmental defects of early failed pregnancies. *Hum Reprod* 2003;18:1724–1732.
17. Swanson AB. A classification for congenital limb malformations. *J Hand Surg Am* 1976;1:8–22.
18. Swanson AB, Swanson GD, Tada K. A classification for congenital limb malformation. *J Hand Surg Am* 1983;8:693–702.
19. Luijsterburg AJ, van Huizum MA, Impelmans BE, et al. Classification of congenital anomalies of the upper limb. *J Hand Surg Br* 2000;25:3–7.
20. De Smet L. Classification for congenital anomalies of the hand: the IFSSH classification and the JSSH modification. *Genet Couns* 2002;13: 331–338.
21. Luijsterburg AJ, Sonneveld GJ, Vermeij-Keers C, et al. Recording congenital differences of the upper limb. *J Hand Surg Br* 2003;28:205–214.
22. Bradbury ET, Hewison J. Early parental adjustment to visible congenital disfigurement. *Child Care Health Dev* 1994;20:251–266.
23. Bradbury ET, Kay SP, Tighe C, et al. Decision-making by parents and children in paediatric hand surgery. *Br J Plast Surg* 1994;47:324–330.
24. Gath A. The impact of an abnormal child upon the parents. *Br J Psychiatry* 1977;130:405–410.
25. Netscher DT, Scheker LR. Timing and decision-making in the treatment of congenital upper extremity deformities. *Clin Plast Surg* 1990;17:113–131.
26. Varni JW, Setoguchi Y. Effects of parental adjustment on the adaptation of children with congenital or acquired limb deficiencies. *J Dev Behav Pediatr* 1993;14:13–20.
27. Volpe JJ. *Neurology of the newborn.* Philadelphia, PA: WB Saunders, 1987.
28. Smith RJ. *Tendon transfers of the hand and forearm.* Boston, MA: Little, Brown, 1987.
29. Zancolli EA, Goldner LJ, Swanson AB. Surgery of the spastic hand in cerebral palsy: report of the Committee on Spastic Hand Evaluation (International Federation of Societies for Surgery of the Hand). *J Hand Surg Am* 1983;8:766–772.
30. Manske PR, McCarroll HRJ, Hale R. Biceps tendon rerouting and percutaneous osteoclasis in the treatment of supination deformity in obstetrical palsy. *J Hand Surg Am* 1980;5:153.
31. Geutjens G, Gilbert A, Helsen K. Obstetric brachial plexus palsy associated with breech delivery. A different pattern of injury. *J Bone Joint Surg Br* 1996;78:303–306.
32. Waters PM, Van Heest A. Spastic hemiplegia of the upper extremity in children. *Hand Clinics* 1998;14:119–134.
33. Bleck EE. *Orthopaedic management of cerebral palsy.* Philadelphia, PA: WB Saunders, 1979.
34. Sakellarides HT, Mital MA, Lenzi WD. Treatment of pronation contractures of the forearm in cerebral palsy by changing the insertion of the pronator radii teres. *J Bone Joint Surg Am* 1981;63:645–652.
35. Van Heest AE, House JH, Cariello C. Upper extremity surgical treatment of cerebral palsy. *J Hand Surg Am* 1999;24:323–330.
36. Mital MA. Lengthening of the elbow flexors in cerebral palsy. *J Bone Joint Surg Am* 1979;61:515–522.
37. Pletcher DF, Hoffer MM, Koffman DM. Non-traumatic dislocation of the radial head in cerebral palsy. *J Bone Joint Surg Am* 1976;58:104–105.
38. House JH, Gwathmey FW, Fidler MO. A dynamic approach to the thumb-in palm deformity in cerebral palsy. *J Bone Joint Surg Am* 1981;63: 216–225.
39. Phelps WM. Long-term results of orthopaedic surgery in cerebral palsy. *J Bone Joint Surg Am* 1957;39:53.
40. Koman LA, Gelberman RH, Toby EB, et al. Cerebral palsy. Management of the upper extremity. *Clin Orthop Relat Res* 1990:62–74.
41. Van Heest AE, House J, Putnam M. Sensibility deficiencies in the hands of children with spastic hemiplegia. *J Hand Surg Am* 1993;18:278–281.
42. Mowery CA, Gelberman RH, Rhoades CE. Upper extremity tendon transfers in cerebral palsy: electromyographic and functional analysis. *J Pediatr Orthop* 1985;5:69–72.
43. Randall M, Carlin JB, Chondros P, et al. Reliability of the Melbourne assessment of unilateral upper limb function. *Dev Med Child Neurol* 2001;43:761–767.
44. Bourke-Taylor H. Melbourne Assessment of Unilateral Upper Limb Function: construct validity and correlation with the Pediatric Evaluation of Disability Inventory. *Dev Med Child Neurol* 2003;45:92–96.
45. Davids JR. Quantitative gait analysis in the treatment of children with cerebral palsy. *J Pediatr Orthop* 2006;26:557–559.
46. Krumlinde-Sundholm L, Holmefur M, Kottorp A, et al. The Assisting Hand Assessment: current evidence of validity, reliability, and reponsiveness to dhange. *Dev Med Child Neurol* 2007;49:259–264.
47. Manske PR. Cerebral palsy of the upper extremity. *Hand Clinics* 1990;6:697–709.
48. Pagliano E, Andreucci E, Bono R, et al. Evolution of upper limb function in children with congenital hemiplegia. *Neurol Sci* 2001;22:371–375.
49. Sterr A, Freivogel S, Voss A. Exploring a repetitive training regime for upper limb hemiparesis in an in-patient setting: a report on three case studies. *Brain Inj* 2002;16:1093–1107.
50. Willis JK, Morello A, Davie A, et al. Forced use treatment of childhood hemiparesis. *Pediatrics* 2002;110:94–96.
51. Sterr A, Freivogel S, Schmalohr D. Neurobehavioral aspects of recovery: assessment of the learned nonuse phenomenon in hemiparetic adolescents. *Arch Phys Med Rehabil* 2002;83:1726–1731.
52. Wright PA, Granat MH. Therapeutic effects of functional electrical stimulation of the upper limb of eight children with cerebral palsy. *Dev Med Child Neurol* 2000;42:724–727.
53. Goldner JL, Koman LA, Gelberman R, et al. Arthrodesis of the metacarpophalangeal joint of the thumb in children and adults. Adjunctive treatment of thumb-in-palm deformity in cerebral palsy. *Clin Orthop Relat Res* 1990:75–89.
54. Botte MJ, Keenan MA. Percutaneous phenol blocks of the pectoralis major muscle to treat spastic deformities. *J Hand Surg Am* 1988;13:147–149.

55. Gelberman RH. The upper extremity in cerebral palsy. In: Bora W, ed. *The pediatric upper extremity*. Philadelphia, PA: WB Saunders, 1986:323.
56. Keenan MA, Tomas ES, Stone L, et al. Percutaneous phenol block of the musculocutaneous nerve to control elbow flexor spasticity. *J Hand Surg Am* 1990;15:340–346.
57. Braun RM, Hoffer MM, Mooney V, et al. Phenol nerve block in the treatment of acquired spastic hemiplegia in the upper limb. *J Bone Joint Surg Am* 1973;55:580–585.
58. Autti-Ramo I, Larsen A, Taimo A, et al. Management of the upper limb with botulinum toxin type A in children with spastic type cerebral palsy and acquired brain injury: clinical implications. *Eur J Neurol* 2001;8:136–144.
59. Rameckers EA, Speth LA, Duysens J, et al. Botulinum toxin-a in children with congenital spastic hemiplegia does not improve upper extremity motor-related function over rehabilitation alone: a randomized controlled trial. *Neurorehabil Neural Repair* 2009;23:218–225.
60. Rosblad B, Andersson G, Pettersson K. Effects of botulinum toxin type A and a programme of functional activity to improve manual ability in children and adolescents with cerebral palsy. *Scand J Plast Reconstr Surg Hand Surg* 2007;41:250–258.
61. Sajzewski L, Ziviani J, Boyd R. Systematic review and meta-analysis of therapeutic management of upper-limb dysfunction in children with congenital hemiplegia. *Pediatrics* 2009;123:1111–1122.
62. Tonkin M, Gschwind C. Surgery for cerebral palsy: part 2. Flexion deformity of the wrist and fingers. *J Hand Surg Br* 1992;17:396–400.
63. Waters PM, Zurakowski D, Patterson P, et al. Interobserver and intraobserver reliability of therapist-assisted videotaped evaluations of upper-limb hemiplegia. *J Hand Surg Am* 2004;29:328–334.
64. Dahlin LB, Komoto-Tufvesson Y, Salgeback S. Surgery of the spastic hand in cerebral palsy. Improvement in stereognosis and hand function after surgery. *J Hand Surg Br* 1998;23:334–339.
65. Eliasson AC, Ekholm C, Carlsted T. Hand function in children with cerebral palsy after upper limb tendon transfer and muscle release. *Dev Med Child Neurol* 1998;40:612–621.
66. Manske PR, Langewisch KR, Strecker WB, et al. Anterior elbow release of spastic elbow flexion deformity in children with cerebral palsy. *J Pediatr Orthop* 2001;21:772–777.
67. Green WT, Banks HH. Flexor carpi ulnaris transplant and its use in cerebral palsy. *J Bone Joint Surg Am* 1962;44:1343.
68. Tachdjian MO. *Pediatric orthopaedics*, 2nd ed. Philadelphia, PA: WB Saunders, 1990:1744.
69. Hoffer MM, Perry J, Melkonian GJ. Dynamic electromyography and decision-making for surgery in the upper extremity of patients with cerebral palsy. *J Hand Surg Am* 1979;4:424.
70. Hoffer MM, Lehman M, Mitani M. Long-term follow-up on tendon transfers to the extensors of the wrist and fingers in patients with cerebral palsy. *J Hand Surg Am* 1986;11:836.
71. Hoffer MM, Perry J, Garcia M, et al. Adduction contracture of the thumb in cerebral palsy. A preoperative electromyographic study. *J Bone Joint Surg Am* 1983;65:755–759.
72. Goldner JL. Reconstructive surgery of the hand in cerebral palsy and spastic paralysis resulting from injury to the spinal cord. *J Bone Joint Surg Am* 1955;37:1141.
73. Rang M. Cerebral palsy. In: Morrissy RT, ed. *Lovell and Winter's pediatric orthopaedics*, 3rd ed. Philadelphia, PA: JB Lippincott, 1990:495.
74. Page CM. An operation for relief of flexion contracture in the forearm. *J Bone Joint Surg Am* 1923;5:233.
75. Van Heest AE, House JH. Lateral band rerouting in the treatment of swan neck deformities due to cerebral palsy. *Tech Hand Up Extrem Surg* 1997;1:189–194.
76. Hardy AE. Birth injuries of the brachial plexus: incidence and prognosis (abstract). *J Bone Joint Surg Br* 1981;63-B(1):98–101.
77. Adler JP, Patterson RL Jr. Erb's palsy. Long-term results of treatment in eighty-eight cases. *J Bone Joint Surg Am* 1967;49:1052–1064.
78. Foad SL, Mehlman CT, Ying J. The epidemiology of neonatal brachial plexus palsy in the United States. *J Bone Joint Surg Am* 2008;90:1258–1264.
79. Wickstrom J. Birth injuries of the brachial plexus. Treatment of defects in the shoulder. *Clin Orthop Relat Res* 1962;23:187–196.
80. Pearl ML, Edgerton BW. Glenoid deformity secondary to brachial plexus birth palsy. *J Bone Joint Surg Am* 1998;5:659–667.
81. Waters PM, Smith GR, Jaramillo D. Glenohumeral deformity secondary to brachial plexus birth palsy. *J Bone Joint Surg Am* 1998;80:668–677.
82. Waters PM. Comparison of the natural history, the outcome of microsurgical repair, and the outcome of operative reconstruction in brachial plexus birth palsy.
83. Gilbert A, Tassin JL. Reparation chirurgicale du plexus brachial dans la paralysie obstetricale. *Chirurgie* 1984;110:70–75.
84. Gilbert A, Brockman R, Carlioz H. Surgical treatment of brachial plexus birth palsy. *Clin Orthop Relat Res* 1991;26:39–47.
85. Mayer VE. Wounds of the extensor tendons proximal to the metacarpophalangeal articulation (zones VI to VIII). *Ann Chir* 1986;41:671–674.
86. Michelow BJ, Clarke HM, Curtis CG, et al. The natural history of obstetrical brachial plexus palsy. *Plast Reconstr Surg* 1994;93:675–680; discussion 681.
87. Lejman T, Strong M, Michno P, et al. Septic arthritis of the shoulder during the first 18 months of life. *J Pediatr Orthop* 1995;15:172–175.
88. Bae DS, Waters PM, Zurakowski D. Reliability of three classification systems measuring active motion in brachial plexus birth palsy. *J Bone Joint Surg Am* 2003;85-A:1733–1738.
89. Kawai H, Tsuyuguchi Y, Masada K. Identification of the lesion in brachial plexus injuries with root avulsion: a comprehensive assessment by means of preoperative findings, myelography, surgical exploration and intraoperative electrodiagnosis. *Neuro Orthop* 1989;7:15–23.
90. Kozin SH. Correlation between external rotation of the glenohumeral joint and deformity after brachial plexus birth palsy. *J Pediatr Orthop* 2004;2:189–193.
91. Pearl ML. Arthroscopic release of shoulder contracture secondary to birth palsy: an early report on findings and surgical technique. *Arthroscopy* 2003;19:577–582.
92. Waters PM. Comparison of the natural history, the outcome of microsurgical repair, and the outcome of operative reconstruction in brachial plexus birth palsy. *J Bone Joint Surg Am* 1999;81:649–659.
93. Moukoko D, Ezaki M, Wilkes D, et al. Posterior shoulder dislocation in infants with neonatal brachial plexus palsy. *J Bone Joint Surg Am* 2004;86:787–793.
94. Lin JC, Schwentker-Colizza A, Curtis CG, et al. Final results of grafting versus neurolysis in obstetrical brachial plexus palsy. *Plast Reconstr Surg* 2009;123:939–948.
95. Oberlin C, Beal D, Leechavengvongs S, et al. Nerve transfer to biceps muscle using a part of ulnar nerve for C5-C6 avulsion of the brachial plexus: anatomical study and report of four cases. *J Hand Surg Am* 1994;19:232–237.
96. Troum S, Floyd WE, Waters PM. Posterior dislocation of the humeral head in infancy associated with obstetrical paralysis. A case report. *J Bone Joint Surg Am* 1993;75:1370–1375.
97. Hoffer MM, Wickenden R, Roper B. Brachial plexus birth palsies. Results of tendon transfers to the rotator cuff. *J Bone Joint Surg Am* 1978;60:691–695.
98. Waters PM. Update on management of pediatric brachial plexus palsy. *J Pediatr Orthop B* 2005;14:233–244.
99. Waters PM, Bae DS. The early effects of tendon transfers and open capsulorrhaphy on glenohumeral deformity in brachial plexus birth palsy. *J Bone Joint Surg Am* 2008;90:2171–2179.
100. Kozin SH, Boardman MJ, Chafetz RS, et al. Arthroscopic treatment of internal rotation contracture and glenohumeral dysplasia in children with brachial plexus birth palsy. *J Shoulder Elbow Surg* 2010;19(1):102–110.
101. Pedowitz DI, Gibson B, Williams GR, et al. Arthroscopic treatment of posterior glenohumeral joint subluxation resulting from brachial plexus birth palsy. *J Shoulder Elbow Surg* 2007;16:6–13.
102. Pearl ML, Edgerton BW, Kazimiroff PA, et al. Arthroscopic release and latissimus dorsi transfer for shoulder internal rotation contractures and

glenohumeral deformity secondary to brachial plexus birth palsy. *J Bone Joint Surg Am* 2006;3:564–574.
103. Waters PM, Bae DS. The effect of derotational humeral osteotomy on global shoulder function in brachial plexus birth palsy. *J Bone Joint Surg Am* 2006;88:1035–1042.
104. Leffert RD. Brachial plexus. In: DP G, ed. *Operative hand surgery*. 3rd ed. New York, NY: Churchill Livingstone, 1993:1509.
105. Aitken J. Deformity of elbow joint as sequel to Erb's obstetrical paralysis. *J Bone Joint Surg Br* 1952;34:352.
106. Zancolli E. Paralytic supination contracture of the forearm. *J Bone Joint Surg Am* 1967;49:1275.
107. Waters PM, Simmons BP. Congenital abnormalities: elbow region. In: Peimer CA, ed. *Surgery of the hand and upper extremity*. New York, NY: McGraw-Hill, 1996:2049.
108. Waters PM, Stewart S. Surgical treatment of non-union and avascular necrosis of the proximal scaphoid in adolescents. *J Bone Joint Surg Am* 2002;84:915–920.
109. Hankins SM, Bezwada HP, Kozin SH. Corrective osteotomies of the radius and ulna for supination contracture of the pediatric and adolescent forearm secondary to neurologic injury. *J Hand Surg Am* 2006;31:118–124.
110. Bevan WP, Hall JG, Bamshad M, et al. Arthrogryposis multiplex congenita (amyoplasia): an orthopaedic perspective. *J Pediatr Orthop* 2007;27:594–600.
111. Fahy MJ, Hall JG. A retrospective study of pregnancy complications among 828 cases of arthrogryposis. *Genet Counsel* 1990;1:3–11.
112. Weeks PM. Surgical correction of upper extremity deformities in arthrogrypotics. *Plast Reconstr Surg* 1965;36:459–465.
113. Kang PB, Lidov HG, David WS, et al. Diagnostic value of electromyography and muscle biopsy in arthrogryposis multiplex congenita. *Ann Neurol* 2003;54:790–795.
114. Gibson DA, Urs ND. Arthrogryposis multiplex congenita. *J Bone Joint Surg Br* 1970;52:483–493.
115. Lloyd-Roberts GC, Lettin AWF. Athrogryposis multiplex congenital. *J Bone Joint Surg Br* 1970;52-B:483–493.
116. Meyn M, Ruby L. Arthrogryposis of the upper extremity. *Orthop Clin North Am* 1976;7:501–509.
117. Ezaki M. Treatment of the upper limb in the child wtih arthrogryposis. *Hand Clinics* 2000;16:703–711.
118. Sells JM, Jaffe KM, Hall JG. Amyoplasia, the most common type of arthrogryposis: the potential for good outcome. *Pediatrics* 1996;97(2):225–231.
119. Van Heest A, Waters PM, Simmons BP. Surgical treatment of arthrogryposis of the elbow. *J Hand Surg Am* 1998;23:1063–1070.
120. Bunnell S. Restoring flexion to paralytic elbow. *J Bone Joint Surg Am* 1951;33:566.
121. Carroll RE, Kleinman WB. Pectoralis major transplantation to restore elbow flexion to the paralytic limb. *J Hand Surg Am* 1979;4:501–507.
122. Zancolli E, Mitre H. Latissimus dorsi transfer to restore elbow flexion. An appraisal of eight cases. *J Bone Joint Surg Am* 1973;55:1265–1275.
123. Williams PF. The elbow in arthrogryposis. *J Bone Joint Surg Br* 1973;55(4):834–840.
124. Van Heest AE, Kallemeier P. Thumb carpal metacarpal arthritis. *J Am Acad Orthop Surg* 2008;16:140–151.
125. Smith DW, Drennan JC. Arthrogryposis wrist deformities: results of infantile serial casting. *J Pediatr Orthop* 2002;22:44–47.
126. Clark J. Reconstruction of biceps brachii by pectoralis major transplantation. *Br J Surg* 1946;34:180.
127. Doyle JR, James PM, Larsen LJ, et al. Restoration of elbow flexion in arthrogryposis multiplex congenita. *J Hand Surg Am* 1980;5:149–152.
128. Sprengel O. Die angeborene Verschiebung des Schulterblattes nach oben. *Arch Klin Chir* 1891;42:545–549.
129. Carson WG, Lovell WW, Whitesides TE Jr. Congenital elevation of the scapula. Surgical correction by the Woodward procedure. *J Bone Joint Surg Am* 1981;63:1199–1207.
130. Cavendish ME. Congenital elevation of the scapula. *J Bone Joint Surg Br* 1972;54:395–408.
131. Hensinger RN. Orthopedic problems of the shoulder and neck. *Pediatr Clin North Am* 1986;33(6):1495–4509.
132. Greitemann B, Rondhuis JJ, Karbowski A. Treatment of congenital elevation of the scapula. 10 (2–18) year follow-up of 37 cases of Sprengel's deformity. *Acta Orthop Scand* 1993;64:365–368.
133. Schrock RD. Congenital elevation of the scapula. *J Bone Joint Surg Am* 1926;8:207–215.
134. Jeannopoulos CL. Observations on congential elevation of the scapula. *Clin Orthop* 1961;20:132–138.
135. Neer RM. Effect of season on activity and bone mineral density. *J Bone Miner Res* 1990;5(12):1271–1272.
136. Robinson DW, Masters FW. The management of complicated avulsion injuries of the extremities. *South Med J* 1967;60:1039–1343.
137. Orrell KG, Bell DF. Structural abnormality of the clavicle associated with Sprengel's deformity. A case report. *Clin Orthop Relat Res* 1990;258: 157–159.
138. Klisić P, Filipović M, Uzelac O, et al. Relocation of congenitally elevated scapula. *J Pediatr Orthop* 1981;1:43–45.
139. Woodward JW. Congenital elevation of the scapula. Correction by release and transplantation of the muscle origins. A preliminary report. *J Bone Joint Surg Am* 1961;43A:219–228.
140. Green WT. The surgical correction of congenital elevation of the scapula [Sprengel's deformity]. *J Bone Joint Surg Am* 1957;39:1439
141. Leibovic SJ, Ehrlich MG, Zaleske DJ. Sprengel deformity. *J Bone Joint Surg Am* 1990;72:192–197.
142. Wilkinson JA, Campbell D. Scapular osteotomy for Sprengel's shoulder. *J Bone Joint Surg Br* 1980;62-B:486–490.
143. Grogan DP, Stanley EA, Bobechko WP. The congenital undescended scapula: surgical correction by the Woodward procedure. *J Bone Joint Surg Br* 1983;65:598
144. Tachdjian MO. *Pediatric orthopedics*, 2nd ed. Philadelphia: WB Saunders; 1990:148.
145. Crenshaw AH, ed. *Campbell's operative orthopaedics*, 7th ed. St. Louis, MO: CV Mosby; 1987:2764.
146. Bellemans M, Lamoureux J. Results of surgical treatment of Sprengel deformity by a modified Green's procedure. *J Pediatr Orthop B* 1999;8:194.
147. Borges JL, Shah A, Torres BC, et al. Modified Woodward procedure for Sprengel deformity of the shoulder: long-term results. *J Pediatr Orthop* 1996;15:508.
148. Farsetti P, Weinstein SL, Caterini R, et al. Sprengel's deformity: long-term follow-up study of 22 cases. *Pediatr Orthop B* 2003;12:202–210.
149. Chung SMK, Nissenbaum MM. Congenital and developmental defects of the shoulder. *Orthop Clin North Am* 1975;6:381.
150. Ross DM, Cruess RL. The surgical correction of congenital elevation of the scapula: a review of seventy-seven cases. *Clin Orthop* 1977;125:17.
151. Jinkins WJ. Congenital pseudarthrosis of the clavicle. *Clin Orthop* 1969;62:183.
152. Gibson DA, Carroll N. Congenital pseudarthrosis of the clavicle. *J Bone Joint Surg Br* 1970;52:629.
153. Quinlan WR, Brady PG, Regan BF. Congenital pseudarthrosis of the clavicle. *Acta Orthop Scand* 1980;51:489.
154. Schnall SB, King JD, Marrero G. Congenital pseudarthrosis of the clavicle: a review of the literature and surgical results of six cases. *J Pediatr Orthop* 1988;8:316.
155. Lorente Molto FJ, Bonete Lluch DJ, Garrido IM. Congenital pseudarthrosis of the clavicle: a proposal for early surgical treatment. *J Pediatr Orthop* 2001;21:689–693.
156. Grogan DP, Love SM, Guidera KJ, et al. Operative treatment of congenital pseudarthrosis of the clavicle. *J Pediatr Orthop* 1991;11:176.
157. Bergsma LA. Long-term results of nonsurgical management of condylar fracture in children. *Int J Oral Maxillofac Surg* 2001;30:365–366.
158. Dobyns JH. Congenital abnormalities of the elbow. In: Morrey BF, ed. *The elbow and its disorders*. Philadelphia, PA: WB Saunders, 1985:161.
159. Fidalgo Valdueza A. The nail-patella syndrome. A report of families. *J Bone Joint Surg Br* 1973;55(1):145–162.

160. Mital MA. Congenital radioulnar synostosis and congenital dislocation of the radial head. *Orthop Clin North Am* 1976;7:375–383.
161. Wynne-Davies R. *Heritable disorders in orthopedic practice*. Oxford, UK: Blackwell Science, 1973.
162. Campbell CC, Waters PM, Emans JB. Excision of the radial head for congenital dislocation. *J Bone Joint Surg Am* 1992;74:726–733.
163. Bucknill TM. Anterior dislocation of the radial head in children. *Proc R Soc Med* 1977;70:620–624.
164. Danielisz L. Congenital dislocation of the head of the radius and elbow injury. *Arch Chir Neerl* 1971;23:163–171.
165. Danielsson LG, Theander G. Traumatic dislocation of the radial head at birth. *Acta Radiol Diagn (Stockh)* 1981;22(3B):379–382.
166. Almquist EE, Gordon LH, Blue AI. Congenital dislocation of the head of the radius. *J Bone Joint Surg Am* 1969;51:1118–1127.
167. Lloyd-Roberts GC, Bucknill TM. Anterior dislocation of the radial head in children: aetiology, natural history and management. *J Bone Joint Surg Br* 1977;59-B:402–407.
168. Mardam-Bey T, Ger E. Congenital radial head dislocation. *J Hand Surg Am* 1979;4:316–320.
169. McFarland B. Congenital dislocation of the radial head of the radius. *J Bone Joint Surg Br* 1936;24:41–49.
170. Hirayama T, Takemitsu Y, Yagihara K, et al. Operation for chronic dislocation of the radial head in children. Reduction by osteotomy of the ulna. *J Bone Joint Surg Br* 1987;69:639–642.
171. Exarhou EI, Antoniou NK. Congenital dislocation of the head of the radius. *Acta Orthop Scand* 1970;41:551–556.
172. Bell SN, Morrey BF, Bianco AJ Jr. Chronic posterior subluxation and dislocation of the radial head. *J Bone Joint Surg Am* 1991;73: 392–396.
173. Sachar K, Mih AD. Congenital radial head dislocations. *Hand Clinics* 1998;14:39–47.
174. Yamazaki S, Majima T, Yasui K, et al. Reconstruction of chronic anterior tibial tendon defect using hamstring tendon graft: a case report. *Foot Ankle Int* 2007;28:1190–1193.
175. Brennan, et al. 1963
176. Villa A, Paley D, Catagni MA, et al. Lengthening of the forearm by the Ilizarov technique. *Clin Orthop Relat Res* 1990;250:125–137.
177. Lewis and Thibodeau 1937
178. King 1939
179. Søjbjerg JO, Ovesen J, Gundorf CE. The stability of the elbow following excision of the radial head and transection of the annular ligament. An experimental study. *Arch Orthop Trauma Surg* 1987;106:248–250.
180. Kelly DW. Congenital dislocation of the radial head: spectrum and natural history. *J Pediatr Orthop* 1981;1(3):295–298.
181. Sutro CJ. Regrowth of bone at the proximal end of the radius following resection in this region. *J Bone Joint Surg Am* 1935;17(4):867–878.
182. Mead C, Martin M. Aplasia of the trochlea: an original mutation. *J Bone Joint Surg Am* 1963;45:379.
183. Wood V. Congenital elbow dislocations. In: Buck-Gramcko D, ed. *Congenital malformations of the hand and forearm*. London, UK: Churchill Livingstone, 1998:487.
184. Bayne LG. Ulnar club hand. In: Green DP, ed. *Operative hand surgery*, 3rd ed. New York, NY: Churchill Livingstone, 1993:298.
185. Ogden JA, Watson HK, Bohne W. Ulnar dysmelia. *J Bone Joint Surg Am* 1976;58:467–475.
186. Riordan CD. Congenital absence of the radius, 15 year follow up. *J Bone Joint Surg Am* 1955;37:1129.
187. Lloyd-Roberts GC. Treatment of defects of the ulna in children by establishing cross-union with the radius. *J Bone Joint Surg Br* 1973;55:327–330.
188. Wood VE. Ulnar dimelia. In: Green DP, ed. *Operative hand surgery*, 3rd ed. New York, NY: Churchill Livingstone, 1993:490.
189. Tsuyuguchi Y, Tada K, Yonenobu K. Mirror hand anomaly: reconstruction of the thumb, wrist, forearm, and elbow. *Plast Reconstr Surg* 1982;70:384–387.
190. Wilkie DPD. Congenital radio-ulnar synostosis. *Br J Surg* 1914;1:366.
191. Green WT, Mital MA. Congenital radio-ulnar synostosis: surgical treatment. *J Bone Joint Surg Am* 1979;61:738–743.
192. Ogino T, Hikino K. Congenital radio-ulnar synostosis: compensatory rotation around the wrist and rotation osteotomy. [erratum appears in *J Hand Surg Br* 1987;12(3):402]. *J Hand Surg Br* 1987;12:173–178.
193. Hansen OH, Andersen NO. Congenital radio-ulnar synostosis. Report of 37 cases. *Acta Orthop Scand* 1970;41:225–230.
194. Cleary JE, Omer GE Jr. Congenital proximal radio-ulnar synostosis. Natural history and functional assessment. *J Bone Joint Surg Am* 1985;67: 539–545.
195. Simmons BP, Southmayd WW, Riseborough EJ. Congenital radioulnar synostosis. *J Hand Surg Am* 1983;8:829–838.
196. Giuffre L, Corsello G, Giuffre M, et al. New syndrome: autosomal dominant microcephaly and radio-ulnar synostosis. *Am J Med Genet* 1994;51:266–269.
197. Robinson GC, Miller JR, Dill FJ. Klinefelter's syndrome with the XXYY sex chromosomes complex. *J Pediatr* 1964;65:266.
198. Sachar K, Akelman E, Ehrlich MG. Radioulnar synostosis. *Hand Clin* 1994;10:399–404.
199. Morrison J. Congenital radio-ulnar synostosis. *Br J Med* 1892;2:1337.
200. Cross AR. Congenital bilateral radioulnar synostosis. *Am J Dis Child* 1939;58:1259.
201. Kelikian H. *Congenital deformities of the hand and forearm*. Philadelphia, PA: WB Saunders, 1974.
202. Stretton JL. Congenital synostosis of radio-ulnar articulations. *Br Med J* 1905;2:1519.
203. Tagima T, Ogisho N, Kanaya F. Follow-up study of joint mobilization of proximal radio-ulnar synostosis. *American Society of Surgery of the Hand Meeting*. Cincinnati, OH, 1994.
204. Boireau P, Laville JM. Rotational osteotomy technique for congenital radio-ulnar synostosis with central medullary nailing and external fixation. *Rev Chir Orthop Reparatrice Appar Mot* 2002;88:812–815.
205. Nieman KM, Gould JS, Simmons BP. Injuries to and developmental deformities of the elbow in children. In: Bora FW, ed. *The pediatric upper extremity: diagnosis and management*. Philadelphia, PA: WB Saunders, 1986:213.
206. El-Adl W. Two-stage double-level rotational osteotomy in the treatment of congenital radioulnar synostosis. *Acta Orthop Belg* 2007;73:704–709.
207. Murase T, Tada K, Yoshida T, et al. Derotational osteotomy at the shafts of the radius and ulna for congenital radioulnar synostosis. *J Hand Surg Am* 2003;28:133–137.
208. Fujimoto M, Kato H, Minami A. Rotational osteotomy at the diaphysis of the radius in the treatment of congenital radioulnar synostosis. *J Pediatr Orthop* 2005;25:676–679.
209. Ramachandran M, Lau K, Jones DH. Rotational osteotomies for congenital radioulnar synostosis. *J Bone Joint Surg Br* 2005;87:1406–1410.
210. Kadiyala K, Waters P. Upper extremity compartment syndromes. *Hand Clinics* 1998;14:467.
211. Dobyns J, Wood V, Bayne L, et al. Congenital hand deformities. In: Green DP, ed. *Operative hand surgery*, 3rd ed. New York, NY: Churchill Livingstone, 1993:251.
212. Murphy HS, Hanson CG. Congenital humeroradial synostosis. *J Bone Joint Surg Am* 1945;27(4):712–713.
213. Leisti J, Lachman RS, Rimoin DL. Humeroradial ankylosis associated with other congenital defects (the "boomerang arm" sign). *Birth Defects Original Article Series* 1975;11:306–307.
214. Solomon L. Hereditary multiple exostosis. *J Bone Joint Surg Br* 1963;45:292–304.
215. Jaffe HL, Bodansky A. Diagnostic significance of serum alkaline and acid phosphatase values in relation to bone disease. *Bull N Y Acad Med* 1943;19:831–848.
216. Shapiro F, Simon S, Glimcher MJ. Hereditary multiple exostoses. Anthropometric, roentgenographic, and clinical aspects. *J Bone Joint Surg Am* 1979;61:815–824.
217. Bock GW, Reed MH. Forearm deformities in multiple cartilaginous exostoses. *Skeletal Radiol* 1991;20:483–486.
218. Fogel GR, McElfresh EC, Peterson HA, et al. Management of deformities of the forearm in multiple hereditary osteochondromas. *J Bone Joint Surg Am* 1984;66:670–680.

219. Masada K, Tsuyuguchi Y, Kawai H, et al. Operations for forearm deformity caused by multiple osteochondromas. *J Bone Joint Surg Br* 1989;71:24–29.
220. Akita S, Murase T, Yonenobu K, et al. Long-term results of surgery for forearm deformities in patients with multiple cartilaginous exostoses. *J Bone Joint Surg Am* 2007;89:1993–1999.
221. Arms DM, Strecker WB, Manske PR, et al. Management of forearm deformity in multiple hereditary osteochondromatosis. *J Pediatr Orthop* 1997;17:450–454.
222. McCornack EB. The surgical management of hereditary multiple exostosis. *Orthop Rev* 1981;10:57.
223. Rodgers WB, Hall JE. One-bone forearm as a salvage procedure for recalcitrant forearm deformity in hereditary multiple exostoses. *J Pediatr Orthop* 1993;13:587–591.
224. Waters PM, Van Heest AE, Emans J. Acute forearm lengthenings. *J Pediatr Orthop* 1997;17:444–449.
225. Lowe HG. Radio-ulnar fusion for defects in the forearm bones. *J Bone Joint Surg Am* 1991;45-B:351–359.
226. Murray RA. The one-bone forearm: a reconstructive procedure. *J Bone Joint Surg Am* 1955;37-A(2):366–370.
227. Tetsworth K, Krome J, Paley D. Lengthening and deformity correction of the upper extremity by the Ilizarov technique. *Orthop Clin North Am* 1991;22:689–713.
228. Catagni MA, Szabo RM, Cattaneo R. P Preliminary experience with Ilizarov method in late reconstruction of radial hemimelia. *J Hand Surg Am* 1993;18:316–321.
229. Dahl MT. The gradual correction of forearm deformities in multiple hereditary exostoses. *Hand Clinics* 1993;9:707–718.
230. Pritchett JW. Lengthening the ulna in patients with hereditary multiple exostoses. *J Bone Joint Surg Br* 1986;68:561–565.
231. Cheng JC. Distraction lengthening of the forearm. *J Hand Surg Br* 1991;16:441–445.
232. Irani RN, Petrucelli RC. Ulnar lengthening for negative ulnar variance in hereditary multiple osteochondromas. *J Pediatr Orthop B* 1992;1: 143–147.
233. Paley D. Lengthening and deformity of the upper arm. *American Society of Surgery of the Hand Congenital Differences Workshop*. Salt Lake City, UT, 1994.
234. Wood VE. Congenital pseudarthrosis of the forearm. In: Green DP, Hotchkiss RN, Pederson WC, eds. *Green's operative hand surgery*, 4th ed. New York, NY: Churchill Livingstone, 1999:541.
235. Bell DF. Congenital forearm pseudarthrosis: report of six cases and review of the literature. *J Pediatr Orthop* 1989;9:438–443.
236. Allieu Y, Gomis R, Yoshimura M, et al. Congenital pseudarthrosis of the forearm–two cases treated by free vascularized fibular graft. *J Hand Surg Am* 1981;6:475–481.
237. Bae DS, Waters PM, Sampson CE. Use of free vascularized fibular graft for congenital ulnar pseudarthrosis: surgical decision making in the growing child. *J Pediatr Orthop* 2005;25:755–762.
238. Weiland AJ, Weiss AP, Moore JR, et al. Vascularized fibular grafts in the treatment of congenital pseudarthrosis of the tibia. *J Bone Joint Surg Am* 1990;72:654–662.
239. Tsai TM, Ludwig L, Tonkin M. Vascularized fibular epiphyseal transfer. A clinical study. *Clin Orthop Relat Res* 1986:228–234.
240. Birch-Jensen A. *Congenital deformities of the upper extremities*, Copenhagen, Denmark: Ejnar Munksgaard Forlag, 1950.
241. Bayne LG, Klug MS. Long-term review of the surgical treatment of radial deficiencies. *J Hand Surg Am* 1987;12:169–179.
242. Manske PR, McCarroll HR Jr, Swanson K. Centralization of the radial club hand: an ulnar surgical approach. *J Hand Surg Am* 1981;6:423–433.
243. James MA, McCarroll HRJ, Manske PR. The spectrum of radial longitudinal deficiency: a modified classification. *J Hand Surg Am* 1999;24:1145–1155.
244. Goldberg MJ, Bartoshesky LE. Congenital hand anomaly: etiology and associated malformations. *Hand Clinics* 1985;1:405–415.
245. Barry JE, Auldist AW. The Vater association; one end of a spectrum of anomalies. *Am J Dis Child* 1974;128(6):769–771.
246. Beals RK, Rolfe B. VATER association. A unifying concept of multiple anomalies. *J Bone Joint Surg Am* 1989;71:948–950.
247. Carroll RE, Louis DS. Anomalies associated with radial dysplasia. *J Pediatr* 1974;Mar;84:409–411.
248. Goldstein R. Hypoplastic anemia with multiple congenital anomalies (Franconi syndrome). *Am J Dis Child* 1955;89:618.
249. Basson CT, Bachinsky DR, Lin RC, et al. Mutations in human TBX5 [corrected] cause limb and cardiac malformation in Holt-Oram syndrome. *Nat Genet* 1997;15:30–35. [Erratum in: *Nat Genet* 1997;15:411].
250. Hall JG. Thrombocytopenia and absent radius (TAR) syndrome. *J Med Genet* 1987;24:79–83.
251. Esmer C, Sanchez S, Ramos S, et al. DEB test for Fanconi anemia detection in patients with atypical phenotypes. *Am J Med Genet* 2004;124A:35–39.
252. Klopocki E, Schulze H, Strauss G, et al. Complex inheritance pattern resembling autosomal recessive inheritance involving a microdeletion in thrombocytopenia-absent radius syndrome. *Am J Hum Genet* 2007;80(2):232–240.
253. Geddis AE. Congenital amegakaryocytic thrombocytopenia and thrombocytopenia with absent radii. *Hematol Oncol Clin North Am* 2009;23:321–331.
254. Fleischman RA, Letestu R, Mi X, et al. Absence of mutations in the HoxA10, HoxA11 and HoxD11 nucleotide coding sequences in thrombocytopenia with absent radius syndrome. *Br J Haematol* 2002;116:367–375.
255. Hedberg VA, Lipton JM. Thrombocytopenia with absent radii. A review of 100 cases. *Am J Pediatr Hematol Oncol* 1988;10:51–64.
256. Bayne LG, Costas BLL, Lourie GM. The upper limb. In: Morrissy RT, Weinstein SL, eds. *Lovell and Winter's pediatric orthopaedics*, 4th ed. Philadelphia, PA: Lippincott-Raven, 1996:781.
257. Skerik SK, Flatt AE. The anatomy of congenital radial dysplasia. Its surgical and functional implications. *Clin Orthop Relat Res* 1969;66: 125–143.
258. Lamb DW. Radial club hand. A continuing study of sixty-eight patients with one hundred and seventeen club hands. *J Bone Joint Surg Am* 1977;59:1–13.
259. Bora FW Jr, Nicholson JT, Cheema HM. Radial meromelia. The deformity and its treatment. *J Bone Joint Surg Am* 1970;52:966–979.
260. Bora FWJ, Carniol PJ, Martin E. Congenital anomalies of the upper limb: radial club hand. In: Bora FWJ, ed. *The pediatric upper extremity: diagnosis and management*. Philadelphia, PA: WB Saunders, 1986:28.
261. Kessler I. Centralisation of the radial club hand by gradual distraction. *J Hand Surg Br* 1989;14:37–42.
262. Nanchahal J, Tonkin MA. Pre-operative distraction lengthening for radial longitudinal deficiency. *J Hand Surg Br* 1996;21(1):103–107.
263. Tu YK, Yeh WL, Sananpanich K, et al. Microsurgical second toe-metatarsal bone transfer for reconstructing congenital radial deficiency with hypoplastic thumb. *J Reconstr Microsurg* 2004;20(3):215–225.
264. Albee FH. Formation of radius congenitally absent, condition seven years after implantation of bone graft. *Ann Surg* 1928;87:105.
265. Starr DE. Congenital absence of the radius: a method of surgical correction. *J Bone Joint Surg Am* 1945;27:572.
266. Entin MA. Reconstruction of congenital aplasia of radial component. *Surg Clin North Am* 1964;44:1091.
267. Vilkki SK. Distraction lengthening and microvascular bone transplantation in the treatment of radial club hand. *American Society for Surgery of the Hand Congenital Differences Workshop*. Salt Lake City, UT, 1994.
268. Buck-Gramcko D. Radialization as a new treatment for radial club hand. *J Hand Surg Am* 1985;10:964–968.
269. Geck MJ, Dorey F, Lawrence JF, et al. Congenital radius deficiency: radiographic outcome and survivorship analysis. *J Hand Surg Am* 1999;24:1132–1144.
270. Nanchahal J, Tonkin MA. Pre-operative distraction lengthening for radial longitudinal deficiency. *J Hand Surg Br* 1996;21:103–107.
271. Goldfarb CA. Reconstruction of radial polydactyly. *Tech Hand Up Extrem Surg* 2006;10:265–270.
272. Taghinia AH, Al-Sheikh AA, Upton J. Preoperative soft-tissue distraction for radial longitudinal deficiency: an analysis of indications and outcomes. *Plast Reconstr Surg* 2007;1205:1305–1312.

273. Sabharwal S, Harten RD, Sabatino C, et al. Selective soft tissue release preserves growth plate architecture during limb lengthening. *J Pediatr Orthop* 2005;25:617–622.
274. Lister G. Reconstruction of the hypoplastic thumb. *Clin Orthop Relat Res* 1985:52–65.
275. Buck-Gramcko D. Pollicization of the index finger. Method and results in aplasia and hypoplasia of the thumb. *J Bone Joint Surg Am* 1971;53:1605–1617.
276. Huber E. Hilfsoperation bei Medianuslahmug. *Dtsch Z Chir* 1921;162:271.
277. Carroll RE. Pollicization. In: Green DP, ed. *Operative hand surgery*, 2nd ed. New York, NY: Churchill Livingstone, 1988:2263.
278. Damore E, Kozin SH, Thoder JJ, et al. The recurrence of deformity after surgical centralization for radial clubhand. *J Hand Surg Am* 2000;25:745–751.
279. Niessen FB, Meek MF, Nicolai JA. The recurrence of deformity after surgical centralization for radial clubhand. *J Hand Surg Am* 2001;26: 363–364.
280. Goldfarb CA, Stern PJ. Indications and techniques for thumb carpometacarpal arthrodesis. *Tech Hand Up Extrem Surg* 2002;6:178–184.
281. Sestero AM, Van Heest A, Agel J. Ulnar growth patterns in radial longitudinal deficiency. *J Hand Surg Am* 2006;31:960–967.
282. Gilbert A. Pollicizations in radial club hand. *Second International Workshop on Congenital Differences of the Upper Limb*. Salt Lake City, UT, 1994.
283. Manske PR, McCaroll HR Jr. Index finger pollicization for a congenitally absent or nonfunctioning thumb. *J Hand Surg Am* 1985;10:606–613.
284. Manske PR, McCarroll HR. Abductor digiti minimi opponens-plasty in congenital radial dysplasia. *J Hand Surg Am* 1978;3:552.
285. Bamshad M, Krakowiak PA, Watkins WS, et al. A gene for ulnar-mammary syndrome maps to 12q23–q24.1. *Hum Mol Genet* 1995;4: 1973–1977.
286. Robert M, Moulies D, Longis B, et al. Fractures of the upper part of the radius in children. *Chir Pediatr* 1986;27:318–321.
287. Turnpenny PD, Dean JC, Duffty P, et al. Weyers' ulnar ray/oligodactyly syndrome and the association of midline malformations with ulnar ray defects. *J Med Genet* 1992;29:659–662.
288. Wulfsberg EA, Mirkinson LJ, Meister SJ. Autosomal dominant tetramelic postaxial oligodactyly. *Am J Med Genet* 1993;46:579–583.
289. Chemke J, Nisani R, Fischel RE. Absent ulna in the Klippel-Feil syndrome: an unusual associated malformation. *Clin Genet* 1980;17: 167–170.
290. Franceschini P, Vardeu MP, Dalforno L, et al. Possible relationship between ulnar-mammary syndrome and split hand with aplasia of the ulna syndrome. *Am J Med Genet* 1992;44:807–812.
291. Ogino T, Kato H. Clinical and experimental studies on ulnar ray deficiency. *Handchir Mikrochir Plast Chir* 1988;20:330–337.
292. Bayne LG. Ulnar club hand (ulnar deficiencies). In: Green DP, ed. *Operative hand surgery*, 3rd ed. New York, NY: Churchill Livingstone, 1993:288.
293. Cole RJ, Manske PR. Classification of ulnar deficiency according to the thumb and first web. *J Hand Surg Am* 1997;22:479–488.
294. Broudy AS, Smith RJ. Deformities of the hand and wrist with ulnar deficiency. *J Hand Surg Am* 1979;4:304–315.
295. Johnson J, Omer GE Jr. Congenital ulnar deficiency. Natural history and therapeutic implications. *Hand Clinics* 1985;1:499–510.
296. Southwood AR. Partial absence of the ulna and associated structures. *J Anat* 1926–1927;61:346.
297. Raus EE. Repair of simple syndactylism in the healthy neo-newborn. *Orthop Rev* 1984;13:498.
298. Beals RK, Lovrien EW. Dyschondrosteosis and Madelung's deformity. Report of three kindreds and review of the literature. *Clin Orthop Relat Res* 1976:24–28.
299. Madelung V. Die spontane subluxaion der hand nach vorne. *Verh Dtsch Ges Chir* 1878;7:259.
300. Ezaki M. Madelung's deformity. In: Green DP, Hotchkiss RN, Pederson WC, eds. *Green's operative hand surgery*. New York, NY: Churchill Livingstone, 1999:528.
301. Felman AH, Kirkpatrick JA Jr. Dyschondrosteose. Mesomelic dwarfism of Lwei and Weill. *Am J Dis Child* 1970;120:329–331.
302. Shears DJ, Vassal HJ, Goodman FR, et al. Mutation and deletion of the pseudoautosomal gene SHOX cause Leri-Weill dyschondrosteosis. *Nat Genet* 1998;19(1):70–73.
303. Nielsen JB. Madelung's deformity. A follow-up study of 26 cases and a review of the literature. *Acta Orthop Scand* 1977;48:379–384.
304. Zebala LP, Manske PR, Goldfarb CA. Madelung's deformity: a spectrum of presentation. *J Hand Surg Am* 2007;32:1393–1401.
305. McCarroll HR Jr, James MA, Newmeyer WL III, et al. Madelung's deformity: quantitative assessment of x-ray deformity. *J Hand Surg Am* 2005;30:1211–1220.
306. Vickers D, Nielsen G. Madelung's deformity: treatment by osteotomy of the radius and Lauenstein procedure. *J Hand Surg Am* 1987;12:202–204.
307. Linscheid RL. Madelung's deformity. *Am Soc Surg Hand Corresp Newslett* 1979;2:10.
308. Ranawat CS, DeFiore J, Straub LR. Madelung's deformity. An end-result study of surgical treatment. *J Bone Joint Surg Am* 1975;57:772–775.
309. Harley BJ, Brown C, Cummings K, et al. Volar ligament release and distal radius dome osteotomy for correction of Madelung's deformity. *J Hand Surg Am* 2006;31:1499–1506.
310. Bruno RJ, Blank JE, Ruby LK, et al. Treatment of Madelung's deformity in adults by ulna reduction osteotomy. *J Hand Surg Am* 2003;28: 421–426.
311. Barsky AJ. Cleft hand: classification, incidence, and treatment. review of the literature and report of nineteen cases. *J Bone Joint Surg Am* 1964;46:1707–1720.
312. Scherer SW, Poorkaj P, Allen T, et al. Fine mapping of the autosomal dominant split hand/split foot locus on chromosome 7, band q21. 3-q22.1. [see comment]. *Am J Human Genet* 1994;55:12–20.
313. Kay SPJ. Congenital hand deformities: cleft hand. In: Green DP, Hotchkiss RN, Pederson WC, eds. *Green's operative hand surgery*. New York, NY: Churchill Livingstone, 1999:402.
314. Buss PW. Cleft hand/foot: clinical and developmental aspects. *J Med Genet* 1994;31:726–730.
315. Calzolari E, Manservigi D, Garani GP, et al. Limb reduction defects in Emilia Romagna, Italy: epidemiological and genetic study in 173,109 consecutive births. *J Med Genet* 1990;27:353–357.
316. Froster-Iskenius UG, Baird PA. Limb reduction defects in over one million consecutive livebirths. *Teratology* 1989;39:127–135.
317. Ogmno T. Cleft hand. *Hand Clinics* 1990;6:661–671.
318. Watari S, Tsuge K. A classification of cleft hands, based on clinical findings: theory of developmental mechanism. *Plast Reconstr Surg* 1979;64:381–389.
319. Manske PR, Halikis MN. Surgical classification of central deficiency according to the thumb web. *J Hand Surg Am* 1995;20:687–697.
320. Snow JW. The use of a volar flap for repair of fingertip amputations: a preliminary report. *Plast Reconstr Surg* 1967;40:163–168.
321. Ueba Y. Plastic surgery for the cleft hand. *J Hand Surg Am* 1981;6: 557–560.
322. Miura T, Komada T. Simple method for reconstruction of the cleft hand with an adducted thumb. *Plast Reconstr Surg* 1979;64:65–67.
323. Goldberg NH, Watson HK. Composite toe (phalanx and epiphysis) transfers in the reconstruction of the aphalangic hand. *J Hand Surg Am* 1982;7(5):454–459.
324. Buck-Gramcko D. Progress in the treatment of congenital malformations of the hand. *World J Surg* 1990;14:715–724.
325. Duclos and Gilbert 1995
326. Buck-Gramcko D, Pereira JA. Proximal toe phalanx transplantation for bony stabilization and lengthening of partially aplastic digits. *Ann Chir Main Memb Super* 1990;9:107–118.
327. Cowen NJ, Loftus JM. Distraction augmentation manoplasty: technique of lengthening digits or entire hands. *Orthop Rev* 1978;7:45–53.
328. Entin MA. Reconstruction of congenital abnormalities of the upper extremities. *J Bone Joint Surg Am* 1959;41-A(4):681–701.
329. Vilkki SK. Advances in microsurgical reconstruction of the congenitally adactylous hand. *Clin Orthop Relat Res* 1995;314:45–58.

330. Seitz WH Jr, Dobyns JH. Digital lengthening. With emphasis on distraction osteogenesis in the upper limb. *Hand Clin* 1993;9:699–706.
331. Pillet J. Esthetic hand prostheses. *J Hand Surg Am* 1983;8(5 Pt 2): 778–781.
332. Kay S. Camptodactyly. In: Green DP, Hotchkiss RN, Pederson WC, eds. *Green's operative hand surgery*, 4th ed. New York, NY: Churchill-Livingstone, 1999:510.
333. Kino Y. Clinical and experimental studies of the congenital constriction band syndrome, with an emphasis on its etiology. *J Bone Joint Surg Am* 1975;57:636–643.
334. Streeter GL. *Focal deficiencies in fetal tissue and their relation to intrauterine amputation. Contributions to embryology*. Washington, DC: Carnegie Institution; 1930:126.
335. Patterson TJS. Congenital ring-constrictions. *Br J Plast Surg* 1961;14:1.
336. Ickowicz V, Patrier S, Lechevallier J, et al. Favourable outcome of a tight constriction band secondary to amniotic band syndrome. *Prenat Diagn* 2004;24:198–201.
337. Keswani SG, Johnson MP, Adzick NS, et al. In utero limb salvage: fetoscopic release of amniotic bands for threatened limb amputation. *J Pediatr Surg* 2003;38:848–851.
338. Soiland H. Lengthening a finger with the "on the top" method. *Acta Chir Scand* 1961;122:184–186.
339. Light TR. Congenital anomalies: syndactyly, polydactyly and cleft hands. In: Peimer CA, ed. *Surgery of the hand and upper extremity*. New York, NY: McGraw-Hill, 1996:2111.
340. Jones NF, Upton J. Early release of syndactyly within six weeks of birth. *Orthop Trans* 1992;17:360–361.
341. Sommerkamp TG, Ezaki M, Carter PR, et al. The pulp plasty: a composite graft for complete syndactyly fingertip separations. *J Hand Surg Am* 1992;17:15–20.
342. Percival NJ, Sykes PJ. Syndactyly: a review of the factors which influence surgical treatment. *J Hand Surg Br* 1989;14:196–200.
343. Keret D, Ger E. Evaluation of a uniform operative technique to treat syndactyly *J Hand Surg Am* 1987;12(5 Pt 1):727–729.
344. Shinya K. Dancing girl flap: a new flap suitable for web release. *Ann Plast Surg* 1999;43:618–624.
345. Withey SJ, Kangesu T, Carver N, et al. The open finger technique for the release of syndactyly. *J Hand Surg Br* 2001;26:4–7.
346. Aydin A, Ozden BC, Erer M. Functional reconstruction of the thumb after resection of a slowly growing chondrosarcoma. *Plast Reconstr Surg* 2004;114:1683–1684.
347. Dao KD, Shin AY, Billings A, et al. Surgical treatment of congenital syndactyly of the hand. *J Am Acad Orthop Surg* 2004;12:39–48.
348. Takagi S, Hosokawa K, Haramoto U, et al. A new technique for the treatment of syndactyly with osseous fusion of the distal phalanges. *Ann Plast Surg* 2000;44(6):660–663.
349. Golash A, Watson JS. Nail fold creation in complete syndactyly using Buck-Gramcko pulp flaps. *J Hand Surg Br* 2000;25:11–14.
350. Sherif MM. V-Y dorsal metacarpal flap: A new technique for the correction of syndactyly without skin graft. *Plast Reconstr Surg* 1998;101:1861–1866.
351. Ekerot L. Syndactyly correction without skin-grafting. *J Hand Surg Br* 1996;21:330–337.
352. Fearon JA. Treatment of the hands and feet in Apert syndrome: an evolution in management. *Plast Reconstr Surg*. 2003;112:1–12.
353. Chang, et al. 2002
354. Deunk J, Nicolai JP, Hamburg SM. Long-term results of syndactyly correction: full-thickness versus split-thickness skin grafts. *J Hand Surg Br* 2003;28:125–130.
355. Muzaffar AR, Rafols F, Masson J, et al. Keloid formation after syndactyly reconstruction: associated conditions, prevalence, and preliminary report of a treatment method. *J Hand Surg Am* 2004;29:201–208.
356. Frazier TM. A note on race-specific congenital malformation rates. *Am J Obstet Gynecol* 1960;80:184–185.
357. Nathan PA, Keniston RC. Crossed polydactyly. Case report and review of the literature. *J Bone Joint Surg Am* 1975;57(6):847–849.
358. Woolf CM, Myrianthopoulos NC. Polydactyly in American negroes and whites. *Am J Hum Genet* 1973;25(4):397–404.
359. Stelling F. The upper extremity. In: Ferguson AB, ed. *Orthopaedic surgery infancy and childhood*. Baltimore, MD: Williams & Wilkins, 1963:304.
360. Turek SL. *Orthopaedic principles and their applications*. Philadelphia, PA: JB Lippincott, 1976.
361. Heras L, Barco J, Cohen A. Unusual complication of ligation of rudimentary ulnar digit. *J Hand Surg Br* 1999;24:750–751.
362. Leber GE, Gosain AK. Surgical excision of pedunculated supernumerary digits prevents traumatic amputation neuromas. *Pediatr Dermatol* 2003;20:108–112.
363. Tada K, Kurisaki E, Yonenobu K, et al. Central polydactyly—a review of 12 cases and their surgical treatment. *J Hand Surg Am* 1982;7: 460–465.
364. Wood VE. Treatment of central polydactyly. *Clin Orthop Relat Res*. 1971;74:196–205.
365. Jones KG, Marmor L, Lankford LL. An overview on new procedures in surgery of the hand. *Clin Orthop Relat Res* 1974;Mar-Apr(99):154–167.
366. Engber WD, Flatt AE. Camptodactyly: an analysis of sixty-six patients and twenty-four operations. *J Hand Surg Am* 1977;2:216–224.
367. Benson LS, Waters PM, Kamil NI, et al. Camptodactyly: classification and results of nonoperative treatment. *J Pediatr Orthop* 1994;14:814–819.
368. Larner AJ. Camptodactyly in a neurology outpatient clinic. *Int J Clin Pract* 2001;55:592–595.
369. Millesi H. Surgical treatment of injured peripheral nerves. *Hefte Unfallheilkd* 1974;0(117):366–371. [Article in German].
370. Smith RJ, Kaplan EB. Camptodactyly in similar atraumatic flexion deformities of the proximal interphalangeal joints of the fingers. *J Bone Joint Surg Am* 1968;50:1187.
371. McFarlane RM, Curry GI, Evans HB. Anomalies of the intrinsic muscles in camptodactyly. *J Hand Surg Am* 1983;8(5 Pt 1):531–544.
372. Buck-Gramcko D. Camptodactyly. In: Bowers WH, ed. *The interphalangeal joints*. Edinburgh, UK: Churchill Livingstone, 1987:200.
373. Courtemanche AD. Campylodactyly: etiology and management. *Plast Reconstr Surg* 1969;44:451–454.
374. Hori M, Nakamura R, Inoue G, et al. Nonoperative treatment of camptodactyly. *J Hand Surg Am* 1987;12(6):1061–1065.
375. Ogino T, Kato H. Operative findings in camptodactyly of the little finger. *J Hand Surg Br* 1992;17:661–664.
376. Miura T, Nakamura R, Tamura Y. Long-standing extended dynamic splintage and release of an abnormal restraining structure in camptodactyly. *J Hand Surg Br* 1992;17:665–672.
377. McFarlane RM, Classen DA, Porte AM, et al. The anatomy and treatment of camptodactyly of the small finger. *J Hand Surg Am* 1992;17:35–44.
378. Siegert JJ, Cooney WP, Dobyns JH. Management of simple camptodactyly. *J Hand Surg Br* 1990;15:181–189.
379. Siegert JJ, Wood MB. Blood flow evaluation of vascularized bone transfers in a canine model. *J Orthop Res* 1990;8:291–296.
380. Carneiro RS. Congenital attenuation of the extensor tendon central slip. *J Hand Surg Am* 1993;18:1004–1007.
381. Oldfield MC. Dupuytren's contracture. *Proc R Soc Med* 1954;47: 361–365.
382. Hersh AH, DeMarinis F, Stecher RM. On the inheritance and development of clinodactyly. *Am J Dis Child* 1940;60:1319.
383. McKusick VA. *Mendelian inheritance in man: catalogs of autosomal dominant, autosomal recessive, and X linked phenotypes*, 2nd ed. Baltimore, MD: The Johns Hopkins University Press, 1968.
384. Poznanski AK, Pratt GB, Manson G, et al. Clinodactyly, camptodactyly, Kirner's deformity, and other crooked fingers. *Radiology* 1969;93:573–582.
385. Poznanski AK. Anomalies of the hand—general considerations. *The hand in radiographic diagnosis*. Philadelphia, PA: Saunders WB, 1984:173–480.
386. Poznanski AK, Macpherson RI, Dijkman DJ, et al. Otopalatodigital syndrome: radiologic findings in the hand and foot. *Birth Defects Orig Artic Ser* 1974;10:125–139.
387. Rubinstein JH. The broad thumbs syndrome—progress report 1968. *Birth Defects Original Article Series* 1969;2:25–41.
388. Amuso SJ. Diastrophic dwarfism. *J Bone Joint Surg Am* 1968;50:113.

389. Taybi H. Diastrophic dwarfism. *Radiology* 1963;80:1.
390. Vickers DW. Langenskiold's operation (physiolysis) for congenital malformations of bone producing Madelung's deformity and clinodactyly. *J Bone Joint Surg Br* 1984;66:778.
391. Vickers D. Clinodactyly of the little finger: a simple operative technique for reversal of the growth abnormality. *J Hand Surg Br* 1987;12:335–342.
392. Caouette-Laberge L, Laberge C, Egerszegi EP, et al. Physiolysis for correction of clinodactyly in children. *J Hand Surg Am* 2002;27:659–665.
393. Slakey JB, Hennrikus WL. Acquired thumb flexion contracture in children: congenital trigger thumb. *J Bone Joint Surg Br* 1996;78:481–483.
394. Rodgers WB, Waters PM. Incidence of trigger digits in newborns. *J Hand Surg Am* 1994;19:364–368.
395. Moon WN, Suh SW, Kim IC. Trigger digits in children. *J Hand Surg Br* 2001;26:11–12.
396. Buchman M, Gibson T, McCallum D. Transmission electron microscopic pathoanatomy of congenital trigger thumbs. *J Pediatr Orthop* 1999;19:411.
397. Dinham JM, Meggitt BF. Trigger thumbs in children. A review of the natural history and indications for treatment in 105 patients. *J Bone Joint Surg Br* 1974;56:153–155.
398. Ger E, Kupcha P, Ger D. The management of trigger thumbs in children. *J Hand Surg [Am]* 1991;16:994.
399. Tsuyuguchi Y, Toda K, Kawaii H. Splint therapy for trigger fingers in children. *Arch Phys Med Rehabil* 1983;64:75.
400. Baek GH, Gong HS, Chung MS, et al. Modified Bilhaut-Cloquet procedure for Wassel type-II and III polydactyly of the thumb. *J Bone Joint Surg Am* 2007;89:534–541.
401. Van Heest AE, House J, Krivit W, et al. Surgical treatment of carpal tunnel syndrome and trigger digits in children with mucopolysaccharide storage disorders. *J Hand Surg Am* 1998;23:236–243.
402. Bae DS, Sodha S, Waters PM. Surgical treatment of the pediatric trigger finger. *J Hand Surg Am* 2007;32:1043–1047.
403. Kleinman WB, Strickland JW. Thumb reconstruction. In: Green DP, Hotchkiss RN, Pederson WC, eds. *Green's operative hand surgery*, 4th ed. New York, NY: Churchill Livingstone, 1999:2068.
404. Buck-Gramcko D. Congenital absence and hypoplasia of thumb. In: Strickland JW, ed. *The thumb, the hand, the upper limb*. Edinburgh, UK: Churchill Livingstone, 1971:1605.
405. Manske P, McCarroll H, James M. Type IIIA hypoplastic thumb. *J Hand Surg Am* 1995;20:246.
406. Blauth W. Der hypoplastiche Daumen. *Arch Orthop Unfall Chir* 1967;62:225.
407. Abdel-Ghani H, Amro S. Characterisitics of patients with hypoplastic thumb: a prospective study of 51 patients with the results of surgical treatment. *J Pediatr Orthop Part B* 2004;13:127–138.
408. Kowalski MF, Manske PR. Arthrodesis of digital joints in children. *J Hand Surg Am* 1988;13:874–879.
409. Shibata M, Yoshizu T, Seki T. Reconstruction of a congenital hypoplastic thumb with use of a free vascularized metatarsophalangeal joint. *J Bone Joint Surg Am* 1998;80:1469.
410. Manske PR, Rotman MB, Dailey LA. Long-term functional results after pollicization for the congenitally deficient thumb. *J Hand Surg Am* 1992;17:1064–1072.
411. Staines KG, Majzoub R, Thornby J, et al. Functional outcome for children with thumb aplasia undergoing pollicization. *Plast Reconstr Surg* 2005;116:1314–1323; discussion 1324–1315.
412. Goldfarb CA, Monroe E, Steffen J, et al. Incidence and treatment of complications, suboptimal outcomes, and functional deficiencies after pollicization. *J Hand Surg Am* 2009;34:1291–1297.
413. Kozin SH, Weiss AA, Webber JB, et al. Index finger pollicization for congenital aplasia or hypoplasia of the thumb. *J Hand Surg Am* 1992;17:880–884.
414. Orioli IM, Castilla EE. Thumb/hallux duplication and preaxial polydactyly type I. *Am J Med Genet* 1999;82:219–224.
415. Heus HC, Hing A, van Baren MJ, et al. A physical and transcriptional map of the preaxial polydactyly locus on chromosome 7q36. *Genomics* 1999;57:342–351.
416. Light T, Buck-Gramcko D. Polydactyly: terminology and classification. In: Buck-Gramcko D, ed. *Congenital malformations of the hand and forearm*. London, UK: Churchill Livingstone, 1998:217.
417. Marks TW, Bayne LG. Polydactyly of the thumb: abnormal anatomy and treatment. *J Hand Surg Am* 1978;3:107–116.
418. Wassel HD. The results of surgery for polydactyly of the thumb. *Clin Orthop Relat Res* 1969;64:175–193.
419. Benetar N. Thumb duplication: simple surgery for a common problem? *Curr Trends Hand Surg* 1995:473.
420. Mih AD. Complications of duplicate thumb reconstruction. *Hand Clinics* 1998;14:143–149.
421. Seidman GD, Wenner SM. Surgical treatment of the duplicated thumb. *J Pediatr Orthop* 1993;13:660–662.
422. Masuda T, Sekiguchi J, Komuro Y, et al. "Face to face": a new method for the treatment of polydactyly of the thumb that maximises the use of available soft tissue. *Scand J Plast Reconstr Surg Hand Surg* 2000;34:79–85.
423. Kawabata H, Masatomi T, Shimada K, et al. Treatment of residual instability and extensor lag in polydactyly of the thumb. [see comment]. *J Hand Surg Br* 1993;18:5–8.
424. Lourie GM, Consts BL, Bayne LG. The zig-zag deformity in preaxial polydactyly. *J Hand Surg Br* 1995;20:561.
425. Miura T. An appropriate treatment for postoperative Z-formed deformity of the duplicated thumb. *J Hand Surg Am* 1977;2:380–386.
426. Tada K, Yonenobu K, Tsuyuguchi Y. Duplication of the thumb: a retrospective view of two hundred and thirty-seven cases. *J Bone Joint Surg Am* 1983;65:584.
427. Dobyns JH, Lipscomb PR, Cooney WP. Management of thumb duplication. *Clin Orthop* 1985;195:26.
428. Cheng JCY, Chan KM, Ma GFY, et al. Polydactyly of the thumb: a surgical plan based on ninety-five cases. *J Hand Surg Am* 1984;9:155
429. Jaeger M, Refior HJ. The congenital triangular deformity of the tubular bones of hand and foot. *Clin Orthop Relat Res* 1971;81:139–150.
430. Dobbs MB, Dietz FR, Gurnett CA, et al. Localization of dominantly inherited isolated triphalangeal thumb to chromosomal region 7q36. *J Orthop Res* 2000;18:340–344.
431. Shiono H, Ogino T. Triphalangeal thumb and dermatoglyphics. *J Hand Surg Br* 1984;9:151–152.
432. Phillips RS. Congenital split foot (lobster claw) and triphalangeal thumbs. *J Bone Joint Surg Br* 1971;53:247.
433. Newbury-Ecob RA, Leanage R, Raeburn JA, et al. Holt-Oram syndrome: a clinical genetic study. *J Med Genet* 1996;33:300–307.
434. Aase JM, Smith DW. Congenital anemia and triphalangeal thumb. *J Pediatr* 1969;74:471.
435. Diamond LR, Allen DM, Magill FB. Congenital (erythroid) hypoplastic anemia: a 25 year study. *Am J Dis Child* 1961;102:403.
436. Townes PL, Brocks ER. Hereditary syndrome of imperforate anus with hand, foot, and ear anomalies. *J Pediatr* 1972;81:321–326.
437. Milch H. Triphalangeal thumb. *J Bone Joint Surg Am* 1951;33:692.
438. Buck-Gramcko D. Hand surgery in congenital malformations. In: Jackson IT, ed. *Recent advances in plastic surgery*. New York, NY: Churchill Livingstone, 1981:115.
439. Barsky AJ. *Congenital anomalies of the hand and their surgical treatment*. Springfield, IL: Charles C. Thomas, 1958.
440. Wood VE. Treatment of the triphalangeal thumb. *Clin Orthop Relat Res* 1976;00:188–200.
441. Horii E, Nakamura R, Makino H. Triphalangeal thumb without associated abnormalities: clinical characteristics and surgical outcomes. *Plast Reconstr Surg* 2001;108:902–907.
442. Landin LA. Fracture patterns in children. Analysis of 8,682 fractures with special reference to incidence, etiology and secular changes in a Swedish urban population 1950–1979. *Acta Orthop Scand* 1983;202:1–109.
443. Hastings H II, Simmons BP. Hand fractures in children. A statistical analysis. *Clin Orthop Relat Res* 1984:120–130.
444. Barton NJ. Fractures of the phalanges of the hand in children. *Hand* 1979;11:134–143.
445. Rang M. *Children's fractures*, 2nd ed. Philadelphia, PA: JB Lippincott, 1983.

446. Fischer MD, McElfresh EC. Physeal and periphyseal injuries of the hand. Patterns of injury and results of treatment. *Hand Clinics* 1994;10: 287–301.
447. Leonard MH, Dubravcik P. Management of fractured fingers in the child. *Clin Orthop Relat Res* 1970;73:160–168.
448. DaCruz DJ, Slade RJ, Malone W. Fractures of the distal phalanges. *J Hand Surg Br* 1988;13:350–352.
449. Louis DS, Jebson PJ, Graham TJ. Amputations. In: Green DP, Hotchkiss RN, Pederson WC, eds. *Green's operative hand surgery*, 4th ed. New York, NY: Churchill Livingstone, 1999:48.
450. Seymour N. Juxta-epiphyseal fracture of the terminal phalanx of the finger. *J Bone Joint Surg Br* 1966;48:347.
451. Al-Qattan MM. Extra-articular transverse fractures of the base of the distal phalanx (Seymour's fracture) in children and adults. *J Hand Surg Br* 2001;26:201–206.
452. Wood VE. Fractures of the hand in children. *Orthop Clin North Am* 1976;7:527–542.
453. Waters PM, Benson LS. Dislocation of the distal phalanx epiphysis in toddlers. *J Hand Surg Am* 1993;18:581–585.
454. Michelinakis E, Vourexaki H. Displaced epiphyseal plate of the terminal phalanx in a child. *Hand* 1980;12:51–53.
455. DeBoeck H, Jaeken R. Treatment of chronic mallet finger in children by tenodermodesis. *J Pediatr Orthop* 1992;12:351.
456. Kardestuncer T, Bae DS, Waters PM. The results of tenodermodesis for severe chronic mallet finger deformity in children. *J Pediatr Orthop* 2008;28:81–85.
457. Dixon GL Jr, Moon NF. Rotational supracondylar fractures of the proximal phalanx in children. *Clin Orthop Relat Res* 1972;83:151–156.
458. Simmons BP, Peters TT. Subcondylar fossa reconstruction for malunion of fractures of the proximal phalanx in children. *J Hand Surg Am* 1987;12:1079–1082.
459. Green DP. Hand injury in children. *Pediatric Clin North Am* 1977;24:903.
460. Al-Qattan MM. Phalangeal neck fractures in children: classification and outcome in 66 cases. *J Hand Surg Br* 2001;26:112–121.
461. Waters PM, Taylor B. Percutaneous osteoclasis and reduction of malangulated phalangeal neck fractures in children. *J Hand Surg Am* 2004;29:707–711.
462. Mintzer CM, Waters OM, Brown DJ. Remodeling of a displaced phalangeal neck fracture. *J Hand Surg Br* 1994;19:594.
463. Campbell RM Jr. Operative treatment of fractures and dislocations of the hand and wrist region in children. *Orthop Clin North Am* 1990;21:217–243.
464. Green DP, Anderson JR. Closed reduction and percutaneous pin fixation of fractured phalanges. *J Bone Joint Surg Am* 1973;55:1651–1654.
465. Hofmeister EP, Kim J, Shin AY. Comparison of 2 methods of immobilization of fifth metacarpal neck fractures: a prospective randomized study. *J Hand Surg Am* 2008;33:1362–1368.
466. Tavassoli J, Ruland RT, Hogan CJ, et al. Three cast techniques for the treatment of extra-articular metacarpal fractures. Comparison of short-term outcomes and final fracture alignments. *J Bone Joint Surg Am* 2005;87:2196–2201.
467. Gabuzda G, Mara J. Bony gamekeeper's thumb in a skeletally immature girl. *Orthopedics* 1991;14:792–793.
468. Jehanno P, Iselin F, Frajman JM, et al. Fractures of the base of the first metacarpal in children. Role of K-wire stabilisation. *Chirurgie de la Main* 1999;18:184–190.
469. Bretlau T, Christensen OM, Edstrom P, et al. Diagnosis of scaphoid fracture and dedicated extremity MRI. *Acta Orthop Scand* 1999;70: 504–508.
470. Brydie A, Raby N. Early MRI in the management of clinical scaphoid fracture. *Br J Radiol* 2003;76:296–300.
471. Light TR. Injury to the immature carpus. *Hand Clinics* 1988;4:415–424.
472. Mintzer CM, Waters PM. Surgical treatment of pediatric scaphoid fracture nonunions. *J Pediatr Orthop* 1999;19:236–239.
473. Mintzer CM, Waters PM, Simmons BP. Nonunion of the scaphoid in children treated by Herbert screw fixation and bone grafting. A report of five cases. *J Bone Joint Surg Br* 1995;77:98–100.
474. Gerard FM. Posttraumatic carpal instability in a young child. A case report. *J Bone Joint Surg Am* 1980;62:131–133.
475. Kaawach W, Ecklund K, Di Canzio J, et al. Normal ranges of scapholunate distance in children 6 to 14 years old. *J Pediatr Orthop* 2001;21: 464–467.
476. Terry CL, Waters PM. Triangular fibrocartilage injuries in pediatric and adolescent patients. *J Hand Surg Am* 1998;23:626–634.
477. Becton JL, Christian JD Jr, Goodwin HN, et al. A simplified technique for treating the complex dislocation of the index metacarpophalangeal joint. *J Bone Joint Surg Am* 1975;57:698–700.
478. Bohart PG, Gelberman RH, Vandell RF, et al. Complex dislocations of the metacarpophalangeal joint. *Clin Orthop Relat Res* 1982: 208–210.
479. Green DP, Terry GC. Complex dislocations of the metacarpophalangeal joint. *J Bone Joint Surg Am* 1973;55:1480.
480. McLaughlin HL. Complex "locked" dislocation of the metacarpophalangeal joint. *J Trauma* 1965;5:683.
481. O'Connell SJ, Moore MM, Strickland JW, et al. Results of zone I and zone II flexor tendon repairs in children. *J Hand Surg Am* 1994;19:48–52.
482. Berndtsson L, Ejeskar A. Zone II flexor tendon repair in children. A retrospective long term study. *Scand J Plast Reconstr Surg Hand Surg* 1995;29:59–64.
483. Moehrlen U, Mazzone L, Bieli C, et al. Early mobilization after flexor tendon repair in children. *Eur J Pediatr Surg* 2009;19:83–86.
484. Amadio PC. Staged flexor tendon reconstruction in children. *Annales de Chirurgie de la Main et du Membre Superieur* 1992;11:194–199.
485. Baker GL, Kleinert JM. Digit replantation in infants and young children: determinants of survival. *Plast Reconstr Surg* 1994;94:139–145.
486. Devaraj VS, Kay SP, Batchelor AG, et al. Microvascular surgery in children. *Br J Plast Surg* 1991;44:276–280.