Reconstruction of Congenital Differences of the Hand

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Summary: Congenital differences of the upper limb occur in approximately 0.16 to 0.18 percent of live births. These patients provide a unique challenge for the reconstructive hand surgeon. The correct and timely diagnosis of hand and upper limb congenital differences will lead to appropriate care and rehabilitation. The International Federation of Societies for Surgery of the Hand has classified congenital upper limb differences based on abnormalities of embryogenesis: failure of formation of parts, failure of differentiation of parts, duplication, overgrowth, undergrowth, constriction ring syndrome, and general skeletal abnormalities. This classification scheme is used as a basis for discussion of the most common upper limb anomalies. Both surgical and nonsurgical treatments are discussed, as is appropriate timing of intervention. (*Plast. Reconstr. Surg.* 124 (Suppl.): 128e, 2009.)

Congenital differences of the upper limb occur in approximately 0.16 to 0.18 percent of live births.[1] Approximately 10 percent of these infants will have partial or complete absence of the involved limb, leading to serious loss of function.[2,3] Because limb formation occurs concurrently with other organ development, it is important to be aware of associated abnormalities, including cardiac, hematopoietic, or tumorous conditions. Communication with the pediatrician is important in establishing a comprehensive diagnosis and for staging and planning of any reconstructive procedures. A multispecialty approach will provide superior outcomes by addressing all aspects of the physical and emotional state of both the patient and the family.[4–6] Because congenital anomalies of the upper limb are a significant challenge, the hand surgeon or reconstructive surgeon, as the team leader and primary decision maker, has a unique opportunity to positively and directly affect the child's growth and development.

A child achieves bimanual palmar grasp by the age of 9 months and learns three-digit pinch between 1 and 2 years. Patterns of hand-eye coordination have been established by age 3. Therefore, successful technical reconstruction may fail to alter already established fixed functional or psychological patterns if reconstruction is not completed by approximately 4 years of age.[2] Ideally, reconstruction should be completed by school age to allow for easier social transitioning.

EMBRYOLOGY OF LIMB DEVELOPMENT

During embryonic development, the upper extremity develops from the arm bud, a mass of mesoderm-derived mesenchyme covered by ectoderm. The *Hox* genes (*HoxA*, *HoxB*, *HoxC*, and *HoxD*) are responsible for regulating limb development in the human embryo. Sonic hedgehog, fibroblast growth factor, and Wnt-7a are some of the known signaling proteins that control *Hox* gene expression. *Hox* gene products act on competent mesenchymal cells within the limb bud, guiding these cells to form condensations at the appropriate time and location. These condensations form the precartilaginous skeletal foundation of the limb.

The limb must form simultaneously across three anatomical axes: proximal to distal axis, dorsal to palmar axis, and anteroposterior (preaxial/postaxial) axis. The apical ectodermal ridge forms as a thickening of ectoderm at the leading edge of the limb bud and, through its interactions with the underlying mesenchymal cells, is responsible for proximal to distal differentiation of the limb.[7] The

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dorsal ectoderm helps to control the dorsal to palmar axis of differentiation, leading to distinct flexor and extensor surfaces of the hand and arm.[8] The zone of polarizing activity is a condensation of mesenchymal cells on the preaxial surface of the limb bud. This zone signals the anteroposterior formation of the limb bud by setting up a gradient of signaling proteins along this axis.[9]

The arm bud begins as an outgrowth from the ventrolateral wall of the developing embryo and appears at approximately 30 days' gestation.[10] Located opposite the fifth through seventh cervical somites, the arm bud precedes lower extremity development throughout embryogenesis. At 33 days' gestation, blood circulation develops within the bud, which has established a flipper-like appearance. By 38 days, blood vessels have become apparent growing from proximal to distal, and a constriction marks the separation of the forearm from the upper arm. Finger development is apparent by day 44, with five distinct mesenchymal separations. By day 52, the digits are completely separated because of apoptosis of the intervening mesenchymal tissue. This orderly resorption of tissue occurs through the release of lysosomal enzymes as cells migrate toward the digital condensations to participate in chondrogenesis.[11] By approximately the seventh week of gestation, the limb bud rotates 90 degrees on its long axis with the elbow positioned dorsally. By the eighth week of gestation, embryogenesis is complete. After the eighth week, the small but completely formed upper limb continues to grow in size and primary ossification centers replace areas of cartilage to complete development.

CLASSIFICATION

Several classification schemes for congenital upper limb malformations have been devised.[12–14] The current classification scheme has been agreed on by the American Society for Surgery of the Hand and the International Federation of Societies for Surgery of the Hand and was first published by Swanson.[15] This classification comprises seven groups based on abnormalities of embryogenesis: failure of formation of parts, failure of differentiation of parts, duplication, overgrowth, undergrowth, constriction ring syndrome, and general skeletal abnormalities. Many of these groups are further subdivided by the anatomical level of the malformation (Table 1). The International Federation of Societies for Surgery of the Hand classification scheme is used to organize the discussion of congenital hand surgery for this review.

Table 1. Swanson Classification of Congenital Hand Deformities

Congenital Hand Deformities
Failure of formation of parts
Transverse arrest
Longitudinal arrest
Failure of differentiation of parts
Soft-tissue involvement
Skeletal involvement
Congenital tumorous conditions
Duplication
Overgrowth
Undergrowth
Congenital constriction ring syndrome
Generalized skeletal abnormalities

Failure of Formation of Parts: Transverse Arrest

Transverse Deficiencies

Transverse deficiencies of the upper limb may occur at any level from the shoulder to the phalanges. Transverse arrest most commonly occurs at the level of the proximal third of the forearm and at the wrist. Digital appendages, or nubbins, are often found at the end of the limb. Transverse deficiencies are usually isolated, unilateral, and sporadic.[16] These defects are thought to be the result of vascular disruption at some point during embryogenesis of the upper limb.[17] Transverse deficiency differs from constriction ring syndrome at the same level in that proximal parts tend to be hypoplastic.

Proximal Transverse Deficiencies

With proximal transverse deficiencies, treatment is usually a prosthetic device.[18] These devices may be static or dynamic and may be controlled by remaining skeletal structures or myoelectric impulses. For children with transverse deficiency at the wrist or metacarpal level, a volar paddle prosthesis may act as a post against which the remaining carpus or metacarpals may be flexed. In bilateral deficiencies, children often become adept at using their lower extremities to perform activities of daily living.

Surgical options for proximal transverse deficiencies may include removal of functionless digital nubbins, stump revision to allow for prosthetic fitting, and excision of excess or functionless parts.[19] In children with bilateral deficiencies and visual impairment, the Krukenberg procedure is advocated. This procedure separates the distal ulna from the radius, allowing for opposition of the two bones during supination of the forearm.[20]

Transverse Arrest of the Digits Distal to the Metacarpal Level

Transverse arrest of the digits distal to the metacarpal level, sometimes referred to as symbrachydactyly, has been treated by conventional techniques of distraction lengthening[21] and nonvascularized toe phalangeal bone grafting.[22,23] Metacarpal or phalangeal lengthening uses the principles of distraction osteogenesis to form new bone. A distractor is placed spanning a metacarpal or phalangeal osteotomy or corticotomy and the bone is distracted 0.5 to 1 mm per day for 3 to 6 weeks until the desired digit length is achieved. The bone gap may consolidate with regenerate bone or may require secondary autogenous or allograft bone grafting.

Transverse deficiencies of the digits may also be treated with nonvascularized toe phalangeal bone grafting from the proximal phalanges of the second, third, or fourth toe. Up to 1.5 cm of length can be achieved with each proximal phalanx graft (Fig. 1). Whether the epiphysis of a toe phalangeal bone graft continues to grow remains controversial. It has been recommended that toe phalangeal bone grafts be performed before 15 months of age, that the bone be harvested extraperiosteally, and that the collateral ligaments and tendons be reattached to provide the optimal conditions for the physis to remain open and thus maintain continued growth.[22,23]

Free microvascular toe-to-hand transfer is becoming an increasingly accepted method for treatment of these patients. The first toe-to-hand transfer was performed by Nicoladoni in 1897 for a traumatic thumb amputation[24] and required multiple stages to preserve the blood supply to the transferred toe. In 1955, Clarkson reported the first series of congenital toe-to-thumb transfers with 15 transfers in six patients.[25] Because multiple stages required immobilization of the hand to the foot, the procedure fell out of favor. The first successful microvascular toe-to-hand transfer was reported by Cobbet in 1969 and led to the possibility of free toe transfers for congenital malformations.[26] The first toe-to-hand transfer to reconstruct a congenital anomaly was performed by O'Brien et al. in 1978.[27] In 1995, Vilkki reported a series of 18 successful congenital toe transfers, with an 11-year follow-up proving that toe transfer was beneficial in this population.[28]

Several congenital anomalies have been treated with toe transfer, including transverse deficiency, longitudinal deficiency, traumatic amputation, vascular malformations, and constriction ring syndrome. Studies have shown that growth potential is retained in the transferred toe. Epiphyseal plates remain open and bone growth is comparable to that of the corresponding toe on the contralateral foot.[29] A long-term study of toe-to-hand transfers in posttraumatic deformities has shown good hand function and acceptance of the transferred digit up to 20 years after the procedure.[30] Transverse deficiency of the thumb is an ideal indication for free microvascular toe-to-hand transfer. Unlike longitudinal thumb deficiencies, the proximal thumb remnant tends to retain some normal anatomy, including a mobile carpometacarpal joint, thenar muscles, and proximal stumps of the flexor pollicis longus and extensor pollicis longus tendons. In such cases, a microsurgical second toe-to-thumb transfer is a better option than pollicization of the index finger (Fig. 2). In children with a thumb but absence of all four fingers or with complete absence of all five digits, bilateral second toe transfers can be performed. The two toe transfers can provide three-post pinch to a remaining thumb or one toe transfer can be used to reconstruct the thumb and the other toe transfer used to create a digit for pinch activity. The child's family should be carefully counseled regarding the limitations and potential complications before proceeding with this extremely difficult reconstruction.[6,28]

Failure of Formation of Parts: Longitudinal Arrest

Radial Longitudinal Deficiency

Radial longitudinal deficiency, or radial club hand, involves approximately one in 30,000 to one in 100,000 live births[31] and is more common in boys than in girls and affects Caucasians more often than other races. The deformity is bilateral up to 50 percent of the time and, when unilateral, affects the right side more often than the left. Radial longitudinal deficiency is often found in association with other malformations of the hematopoietic, cardiac, genitourinary, and skeletal systems, including Fanconi anemia, TAR syndrome (*t*hrombocytopenia, *a*bsent *r*adius), Holt-Oram syndrome (cardiac defects), and the VATER association (*v*ertebral anomalies, *a*nal atresia, *t*racheo*e*sophageal fistula, *r*enal defects).

Radial longitudinal deficiency has been classified into four types based on the severity of the involvement of the radius (Table 2).[32] Thumb hypoplasia is often present ranging from slight to total absence. The scaphoid and trapezium may also be absent. Radial sided digits often exhibit a flexion deformity or camptodactyly. The small fin-

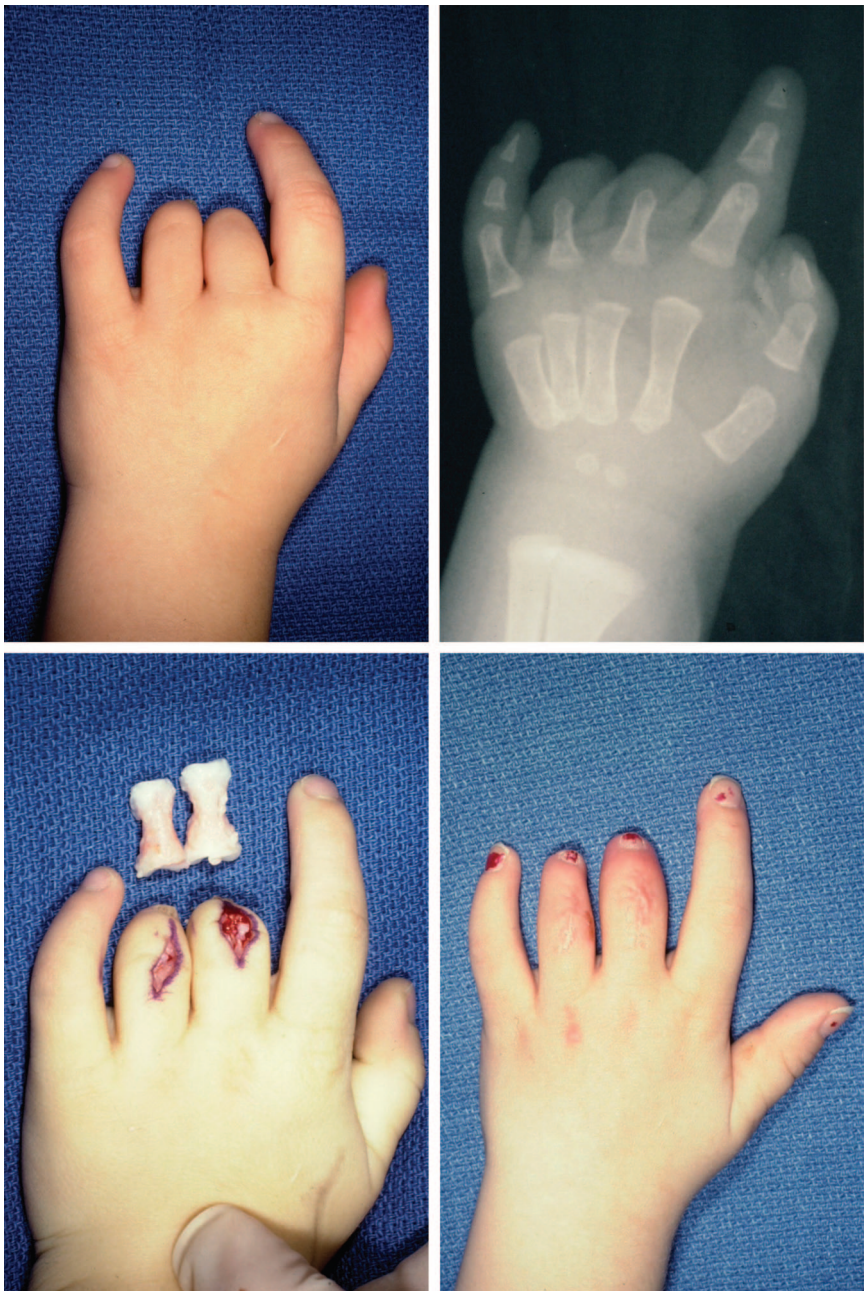

Fig. 1. Nonvascularized toe phalangeal bone grafts. (*Above*) Transverse arrest of the middle and ring fingers at the proximal interphalangeal joint level. (*Below, left*) Second and third toe nonvascularized proximal phalanx grafts. (*Below, right*) Postoperative result after nonvascularized toe phalangeal grafting.

ger is often unaffected, with relatively normal function. Short, fibrotic muscles run along the radial side of the forearm and insert into the ulna, causing severe bowing (Fig. 3). The ulna may also be short and the distal humerus is often hypoplastic, leading to stiffness of the elbow. Both the radial artery and radial nerve may be absent. The median nerve is always present and courses superficially below the skin in the radial concavity, making it prone to injury during operative exposure.

Radial longitudinal deficiency results in very poor function in the affected hand because of the flexed position of the radially deviated hand, loss of wrist support, poor flexor/extensor tendon excursion, hypoplasia of the thumb, and stiffness of the elbow. These deformities increase with age,

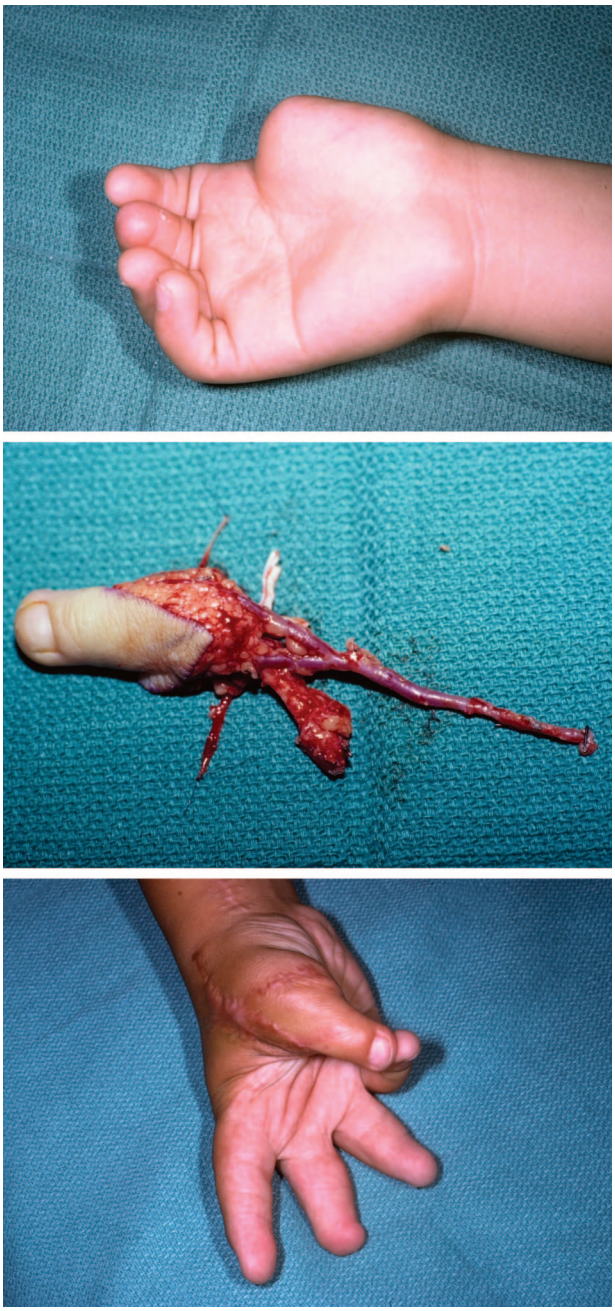

Fig. 2. Microvascular second toe transfer. (*Above*) Constriction ring syndrome with absence of the thumb at the distal metacarpal level. (*Center*) Completed dissection of the second toe. (*Below*) Postoperative appearance and function of the transferred second toe. Note the excellent opposition to the fifth digit.

leading to deteriorating function. Psychologically, the appearance of the hand can be quite troubling for both the child and the parents. For these reasons, treatment is advocated immediately after birth and consists of passive stretching exercises and serial casting to begin, centralizing the wrist and hand on the remaining ulna. Contraindications to treatment include older patients who have adapted to their deformity and children with stiff elbows. In these circumstances, the radial angulation of the hand and carpus makes feeding and hygiene possible in the presence of a stiff elbow.

Surgical treatment of radial longitudinal deficiency attempts to improve the appearance and function of the hand by stabilizing the carpus on the end of the ulna. Historically, centralization of the carpus over the ulna and bone grafting of the absent radius have been attempted,[33] but centralization of the carpus on the ulna remains the definitive treatment. Centralization is usually performed in the first year through a Z-plasty incision over the radial aspect of the wrist to release the tight skin envelope. After identifying the median nerve, the carpus is freed from the radial fibrotic muscle mass and then centralized over the ulna, after transecting the brachioradialis, flexor carpi radialis, and extensor carpi radialis longus tendons. The lunate may need to be excised to fit the carpus over the end of the ulna. A longitudinal Steinmann pin is used to hold the middle finger metacarpal and carpus over the ulna for several months. Radial deforming tendons such as the flexor carpi radialis may then be transferred to the extensor carpi ulnaris to help rebalance the carpus. Buck-Gramcko has advocated radialization of the carpus in which the deformity is overcorrected to the ulnar side by placing the ulna along the axis of the index finger metacarpal.[34] Preoperative distraction may be performed initially to allow the carpus to be radialized or centralized without the need for resection of carpal bones.[35,36] If significant ulnar bowing is present, a corrective osteotomy or multiple osteotomies may also be performed and fixed with the same Steinmann pin to help straighten the long axis of the forearm.

Ulnar Longitudinal Deficiency

Ulnar longitudinal deficiency occurs in one in every 100,000 live births.[37] The deformity is often sporadic and does not have the syndromic associations of radial longitudinal deficiency. However, approximately 50 percent of patients will have some type of musculoskeletal abnormality, including the contralateral upper limb or the lower limbs. There is a clinical spectrum from hypoplasia of the ulna with an intact epiphysis to total absence of the ulna with radiohumeral synostosis. In all cases, a fibrous anlage tether replaces the missing ulna and inserts into the ulnar aspect of the carpus or the distal radius epiphysis. The flexor carpi ulnaris is absent, the ulnar and me-

Table 2. Classification of Radial Longitudinal Deficiencies

Type	Description	Anatomical Features
I	Short distal radius	Distal radial epiphyseal attenuation; mildly shortened radius; hypoplastic thumb; adequate carpal support
II	Hypoplastic radius	Proximal and distal epiphyseal attenuation; substantially shortened radius; ulnar bowing and poorly supported carpus
III	Partial absence of radius	Radial absence: proximal, middle, or distal third; ulnar bowing and poorly supported carpus
IV	Absent radius	Complete absence of radius; unsupported hand with severe radial displacement

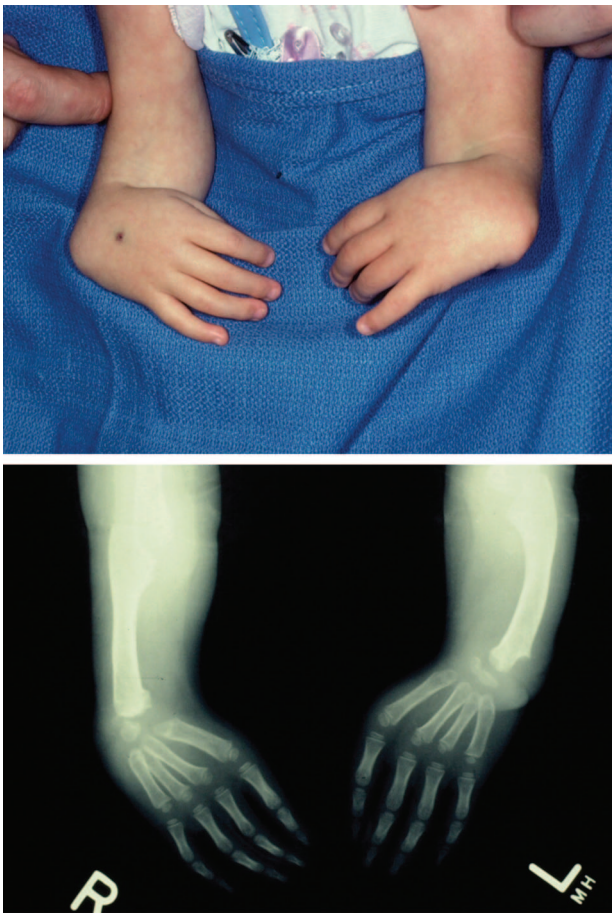

Fig. 3. Clinical and radiographic views of radial club hand with severe ulnar bowing at the wrist.

dian nerves are present, but the ulnar artery is often absent.[38] Unlike radial longitudinal deficiency, the wrist is stable, allowing for relatively normal digital function. The radial head may be dislocated, leading to pain or loss of function at the elbow. With the most severe deficiencies, the humerus is internally rotated and the forearm pronated, compromising positioning of the hand.

Treatment of ulnar longitudinal deficiency consists of serial casting to improve the wrist and elbow positions.[31] Excision of the anlage is indicated for greater than 30 degrees of angulation or when the deformity is progressive. The anlage is approached through a lazy-S incision and resected off of the carpus or distal radius. Kirschner wires may be used to hold the wrist in a neutral position. Tendon transfers are not required; however, in severe bowing, a radial osteotomy may be required to help straighten the long axis of the forearm. When there is loss of function at the elbow, the proximal radial head is resected and a one-bone forearm is created by osteosynthesis of the distal radius to the proximal ulna. In the case of radiohumeral synostosis, a derotational osteotomy of the humerus may be required to place the hand into a more functional position. Arthroplasty of the elbow is not advised because of the low likelihood of success.[2]

Central Ray Deficiency

Central ray deficiency, or cleft hand, was originally classified as either typical or atypical. Typical (true) cleft hand is caused by failure of development of the central digit of the hand, the middle finger, including the metacarpal, which leads to a deep V-shaped cleft. The border digits are occasionally involved in a syndactyly with a tight first web space. Transverse bones separating the index and ring fingers are often present (Fig. 4). Atypical cleft hand is now considered to be a variant of symbrachydactyly. The central digits of the hand are shortened or absent, with vestigial nubbins remaining. The "cleft" is broad and flat, unlike the V-shaped cleft of typical central ray deficiency, usually leaving a thumb and ulnar border digits.

Cleft hand is usually inherited as an autosomal dominant trait, with reduced penetrance and variable expressivity among family members. There may be associated abnormalities, including cardiac, visceral, ocular, auditory, and musculoskeletal,[39] including cleft feet. Manske and Halikis have classified cleft hands based on the involvement of the first web space, which is the most predictive of hand function and therefore helps guide surgical treatment.[40]

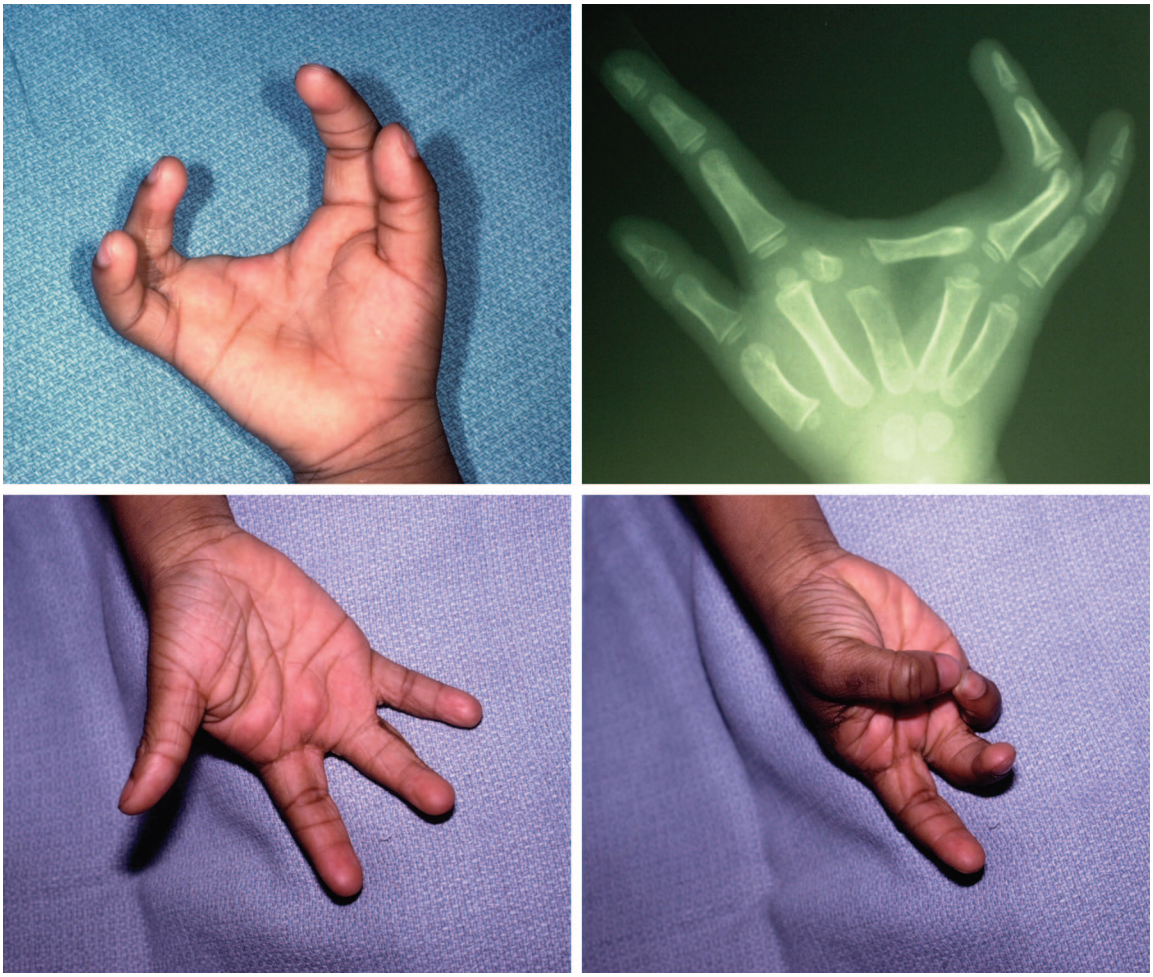

Fig. 4. Typical cleft hand. (*Above*, *left*) Clinical appearance of the typical cleft hand with a V-shaped cleft. (*Above*, *right*) Radiographic appearance of the typical cleft hand. Note the anomalous transverse bones. (*Below*) Postoperative views showing improved appearance and function. Note the widened first web space and excellent thumb opposition.

Flatt described cleft hand as a "functional triumph but a social disaster."[41] Treatment is directed at closing the cleft to improve the appearance of the hand and to treat any syndactyly that may exist. Transverse bones are removed from within the cleft, as these will continue to grow and push the cleft farther apart with age. If the ulnar border digits are syndactylized, they can be released at the time of cleft closure. The Snow-Littler procedure may be used to release the first web space syndactyly by releasing the thumb from the index finger and then transposing the index finger ray onto the middle finger metacarpal remnant, thereby achieving web release and cleft closure simultaneously.[42] Alternatively, closure of the cleft by transposition of the index finger into the middle finger position as described by Miura and Komada may be technically simpler.[43]

Undergrowth

Hypoplastic Thumb

Hypoplastic thumb may also be characterized as a variant of radial longitudinal deficiency and is often associated with radial club hand. Children with a hypoplastic thumb may begin to develop widening of the second web space to achieve rudimentary pinch between the index and middle fingers. Children with complete absence of the thumb may develop pronation of the index finger for the same reason. Therefore, treatment is often recommended by the second year to establish more normal prehensile patterns.[2]

The Blauth classification is used to categorize thumb hypoplasia[44] (Table 3). Type I is a slightly shorter normal functioning thumb and does not require any treatment. Types II and IIIA in which the carpometacarpal joint is stable can be treated

Table 3. Classification of the Hypoplastic Thumb

Type	Clinical Features
I	Gross size diminished
II	Narrow first web space; hypoplastic thenar muscles; MCP joint instability
III	Narrow first web space; hypoplastic thenar muscles; MCP joint instability; abnormal extrinsic tendons; hypoplastic metacarpal
IIIA	Stable CMC joint
IIIB	Unstable CMC joint
IV	Pouce flottant (floating thumb); rudimentary phalanges; skin bridge with neurovascular pedicle
V	Absent thumb

MCP, metacarpophalangeal; CMC, carpometacarpal.

with deepening of the first web space and a tendon transfer to improve opposition. The web space is deepened by a traditional four-flap Z-plasty procedure. The Huber transfer uses the abductor digiti minimi to recreate the thenar eminence and replace the hypoplastic intrinsic muscles.[45] The flexor digitorum superficialis from the ring finger may also be transferred through a window in the transverse carpal ligament to restore thumb opposition.

Types IIIB, IV, and V require complete reconstruction because of an unstable or absent carpometacarpal joint. Index finger pollicization is the treatment of choice for these children. Pollicization was originally described by Littler and modified by Buck-Gramcko.[46,47] In this technique, skin flaps are designed to widen the web space between the new thumb and middle finger. The index finger is elevated as an island flap on its radial and ulnar neurovascular pedicles, dorsal veins, and tendons. The metacarpal is osteotomized at the level of the distal epiphyseal plate and the metaphyseal flare at its base and the intervening shaft removed. The finger is pronated between 140 and 160 degrees and the metacarpal fixated in 45 degrees' abduction palmar to the base of the index finger metacarpal. The metacarpal head then becomes the new carpometacarpal joint. The first dorsal interosseus muscle is reattached to become the abductor pollicis brevis, the first palmar interosseus muscle becomes the adductor pollicis, the index extensor digitorum communis functions as the abductor pollicis longus, and the extensor indicis proprius becomes the extensor pollicis longus (Fig. 5).

Failure of Separation of Parts

Syndactyly

Syndactyly resulting from failure of digital separation is one of the most common congenital hand malformations and occurs in approximately one in 2000 births and is common in white male children.[48] It occurs bilaterally in 50 percent of cases, and 10 to 40 percent of these cases demonstrate a family history of inheritance as an autosomal dominant trait.[49] Inherited forms are associated with genetic defects involving particular candidate regions on the second chromosome.[50] In isolated syndactyly, the long-ring finger web space is most commonly affected, whereas the thumb-index finger web space is the least commonly affected. Syndactyly is classified as either complete or incomplete and as either simple or complex, depending on the degree of skin and/or bone involvement.[51] In complete syndactyly, the web extends out to the nails, whereas incomplete syndactyly stops short of the fingertips. Simple syndactyly involves only the soft tissues, whereas complex syndactyly involves the phalanges, most commonly involving fusion of the distal phalanges. Syndactyly associated with other anomalies such as polydactyly, constriction rings, toe webbing, brachydactyly, spinal deformities, and heart disorders is termed complicated syndactyly (i.e., Apert syndrome).[52]

Surgical release of syndactyly is recommended early to allow normal growth of the digits and normal grasp and pinch. Timing of syndactyly release is based largely on surgeon preference, although most begin separation by 12 months of age with the goal of finishing all releases by the time the child is of school age. Early release of syndactyly involving the thumb-index finger web space, complex syndactyly involving the distal phalanges, and syndactyly producing a flexion contracture of the longer digit may require release by 3 to 6 months of age.[53] Syndactyly involving more than one web space such as in Apert or Poland syndrome requires a decision on the sequence of staged releases, because usually only one side of a digit should be released at one time to avoid the vascular compromise of the digit that would occur if both sides of the digit were released simultaneously. The border digits, thumb, and small finger are usually released first, followed by the central three digits several months later. In complete syndactyly, the web space is reconstructed with a proximally based dorsal rectangular flap. The design of interdigitating skin flaps must be planned carefully, and triangular, zigzag, and rectangular incisions have all been advocated.[54,55] Separation will not usually provide sufficient skin to resurface the circumference of each digit, and thus skin grafts are required. Full-thickness skin grafts are preferred to split-thickness grafts, as they are less

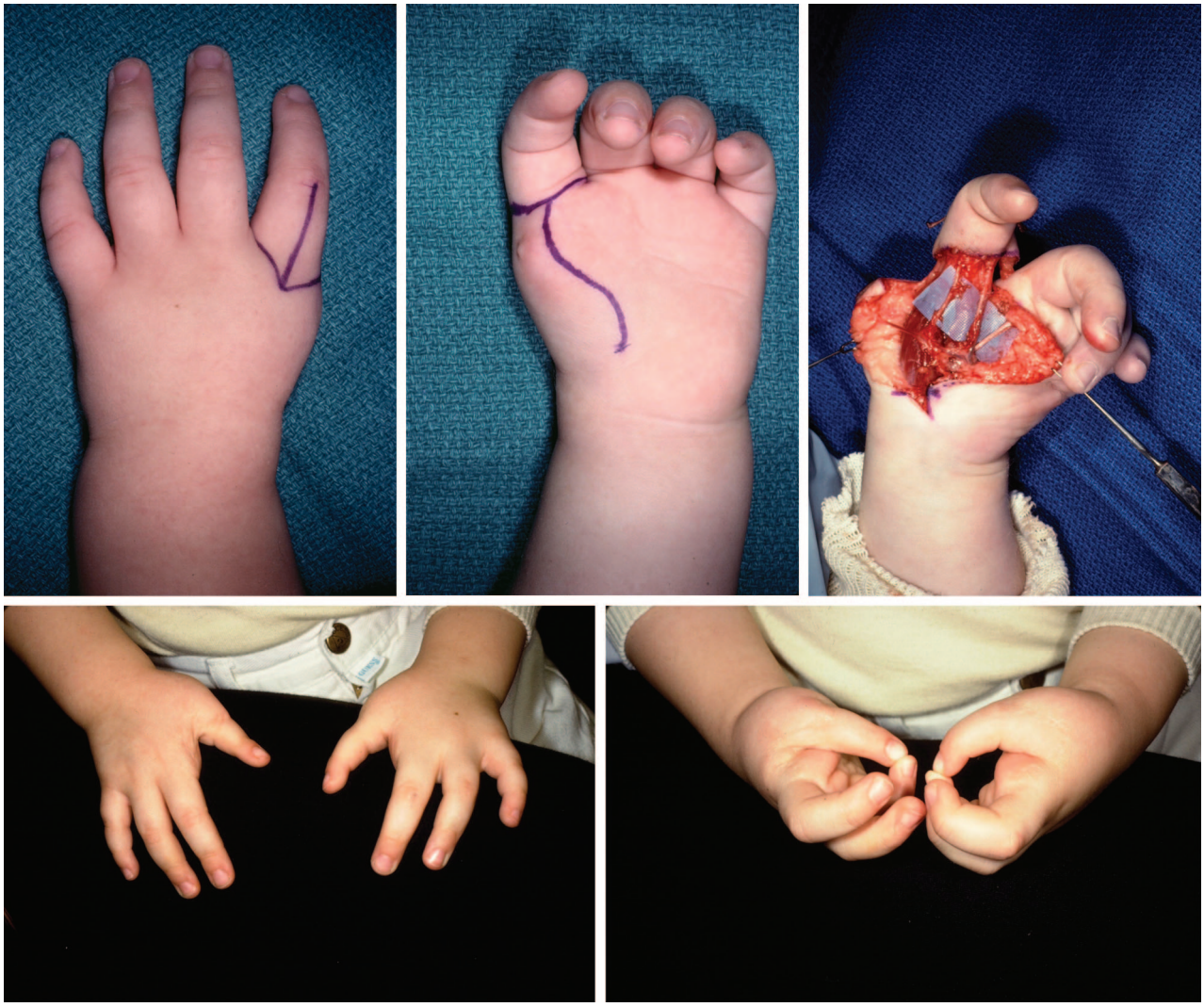

Fig. 5. (*Above*, *left* and *center*) Type V hypoplastic thumb in preparation for index finger pollicization. (*Above*, *right*) Intraoperative view of pollicization procedure showing isolated index finger neurovascular bundles and middle finger radial neurovascular bundle. (*Below*) Postoperative views of bilateral pollicization with intact opposition.

prone to contracture[56] (Figs. 6 through 8). In incomplete syndactyly, various other techniques, including simple Z-plasty, four-flap Z-plasty, or double-opposing Z-plasty, may allow separation of the digits and deepening of the web space without requiring full-thickness skin grafts.[57]

Radioulnar Synostosis

Congenital proximal radioulnar synostosis results from the failure of developing cartilaginous precursors of the forearm to separate late in the first trimester and is bilateral in 60 percent of all patients.[58] The incidence is unknown and most cases occur sporadically, but it can be inherited as an autosomal dominant trait with variable penetrance.[59] Children usually present between 2 and 6 years of age, with absence of forearm rotation and a slight elbow flexion contracture. The child is often school age before diagnosis is made because of the ability of the wrist to compensate for the lack of pronation/supination of the forearm.[60,61] Clinical suspicion warrants radiographs of the forearm, which reveal the proximal radioulnar synostosis. The radial head is often subluxed or dislocated.

Surgical management depends on the severity of the synostosis and the resulting functional impairment. Extreme pronation or supination that interferes with function is an indication for surgery. In addition, a forearm fixed in greater than 60 degrees of pronation generally requires surgery.[60] Derotational osteotomy either at the site of synostosis or in the diaphysis of the radius and

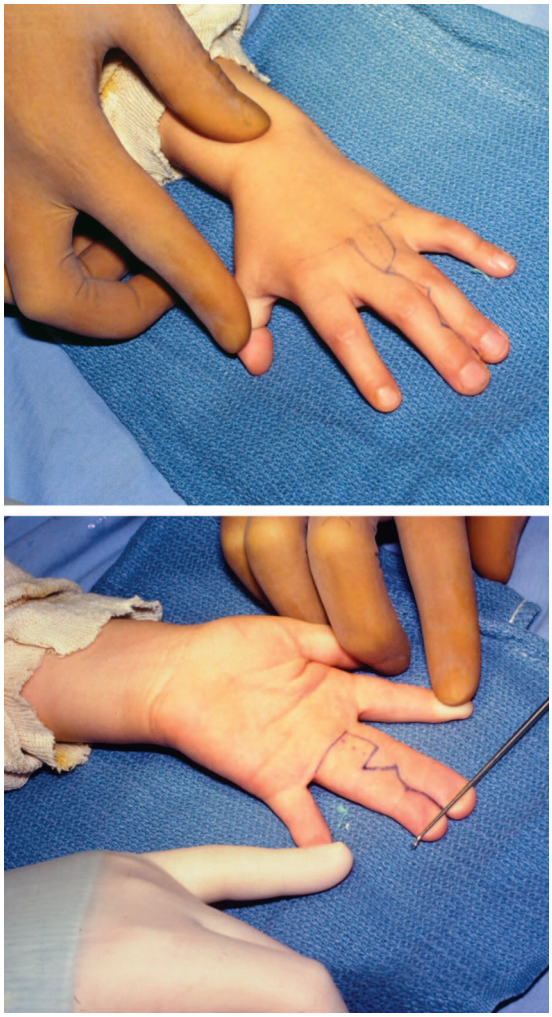

Fig. 6. Syndactyly release. Preoperative flap design.

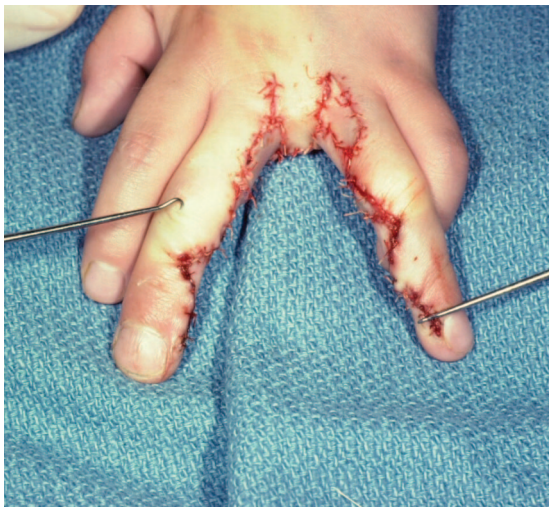

Fig. 7. Syndactyly release. Immediate postoperative view after insetting of flaps and full-thickness skin grafts.

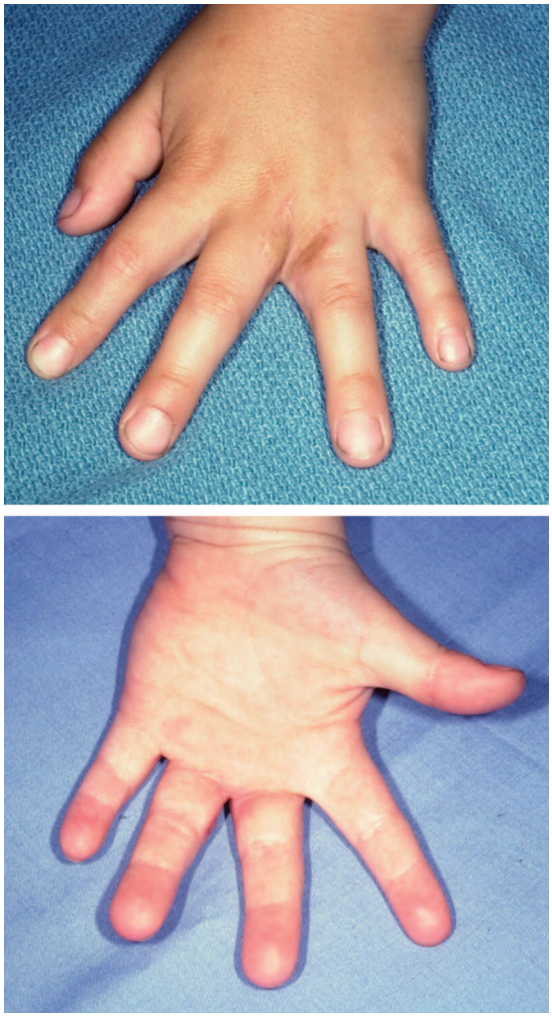

Fig. 8. Syndactyly release. Postoperative views showing complete release and reconstruction of web space.

ulna to fix the forearm in neutral or slight pronation has been advocated.[62] However, resection of the synostosis and interposition of autologous tissue or allograft between the radius and ulna is favored.[58] However, separation is tenuous, as the synostosis tends to recur, and many interposition materials placed at the time of separation have been studied, including synthetic materials, autologous tissues, and allograft tissue. Synthetic materials have included silicone and polyethylene sheeting, and autologous tissues have included nonvascularized or vascularized tissue such as free fat grafts, the radial forearm fascial flap, and a free lateral arm adipofascial flap.[63] In addition, some surgeons have recommended perioperative irradiation, although this is usually used for posttraumatic radioulnar synostosis.[64]

Symphalangism

Symphalangism is the term used to describe failure of interphalangeal joint development and fusion of the proximal phalanges to the middle phalanges and was first described by Cushing in 1916.[65] This condition constitutes 1 percent of all congenital upper extremity anomalies and is frequently transmitted as autosomal dominant.[66] Flatt and Wood classified symphalangism as true symphalangism without additional skeletal abnormalities, symphalangism associated with symbrachydactyly, or symphalangism with syndactyly.[67] Clinically, there is absence of motion and there are skin creases in the affected digits. The proximal interphalangeal joint does not develop with growth. The affected fingers do have some flexion, as the metacarpophalangeal and distal interphalangeal joints are present and have normal range of motion. Attempts have been made to reconstruct or replace the proximal interphalangeal joints, but results have not been favorable.[68] If a child has poor grasp secondary to symphalangism, a wedge of bone can be removed from the level of the proximal interphalangeal joint and the digit fused in 45 degrees of flexion.

Duplication (Polydactyly)

Polydactyly can occur on the preaxial (radial) or postaxial (ulnar) side of the limb or centrally, with postaxial polydactyly being the most common type. Preaxial polydactyly is more common in the white population, and postaxial polydactyly is more common in African Americans.[69,70]

The supernumerary digit in postaxial polydactyly is either well developed (type A) or rudimentary and pedunculated (type B).[71] Those that are rudimentary and represent a small nubbin of tissue can be managed by ligating the base of the pedicle in the nursery. This will lead to necrosis of the nubbin, which will eventually fall off. The more developed type A digits require formal surgical ablation and may require reattachment of the ulnar collateral ligament at the metacarpophalangeal joint or the abductor digiti quinti tendon.

Preaxial polydactyly or thumb duplication occurs in eight in 100,000 births. Both the radial and ulnar duplicated thumbs show some degree of hypoplasia, although the radial duplicate is usually more affected. Wassel has categorized thumb duplication into seven types.[72] Type I is characterized by a bifid distal phalanx, whereas type II is a duplication at the level of the interphalangeal joint. Type III is a bifid proximal phalanx, and type IV, the most common, is a duplication at the level of the metacarpophalangeal joint. Type V is characterized by a bifid metacarpal and type VI is a duplication at the level of the carpometacarpal joint. Type VII describes thumb polydactyly with an associated triphalangeal thumb (Table 4 and Fig. 9).

Treatment of thumb polydactyly is based on the type of duplication. Types I and II can be treated with either resection of the radial duplication or central resection (Bilhaut operation) from each of the duplicated thumbs while preserving their outer portions.[73] Unbalanced thumbs are generally managed with resection of the radial duplication and balanced thumbs are managed with central resection. Treatment of duplication types III and IV must be individualized. In general, the best phalangeal portions of both thumbs are incorporated to create the best thumb.[74] The radial duplication is usually amputated, as it is less developed, followed by radial collateral ligament reconstruction of the metacarpophalangeal joint and reattachment of the thenar muscle insertion to the radial base of the proximal phalanx of the remaining thumb. Treatment of types V and VI involves amputation of the radial duplication along with intrinsic muscle reattachment and collateral ligament reconstruction if necessary. Osteotomies may occasionally be required to realign the metacarpal with the proximal phalanx.

Central polydactyly is a duplication involving the index, long, or ring finger. This is the least common type of polydactyly and may occur in isolation or as part of a syndrome.[75] Duplication of the ring finger is the most common, followed by the long and index fingers.[76] Central polydactyly may be hidden within a concomitant syndactyly, which is termed synpolydactyly. Treatment depends on the extent of involvement. A fully formed and functional central polydactyly does not necessarily require excision. Central polydactyly that is partially formed and/or has limited motion may require a ray resection. Given the potential spectrum of abnormality, the neurovascular bundles must be meticulously dissected so as not to compromise the remaining central digits.

Table 4. Wassel Classification of Thumb Duplication

Type	Description
I	Bifid distal phalanx
II	Duplication at the interphalangeal joint
III	Bifid proximal phalanx
IV	Duplication at the metacarpophalangeal joint
V	Bifid metacarpal
VI	Duplication at the carpometacarpal joint
VII	Triphalangeal thumb

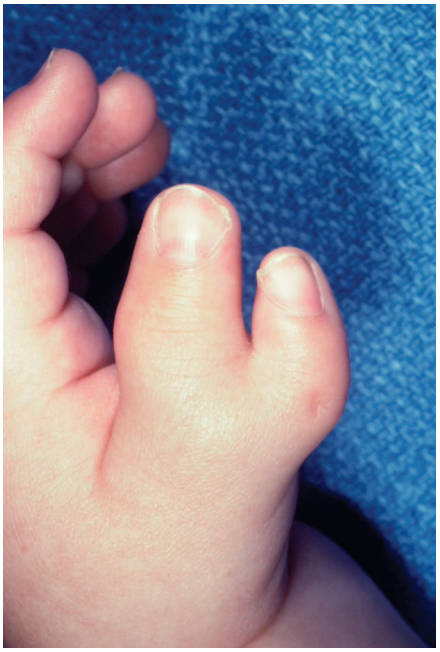

Fig. 9. Thumb polydactyly: type IV duplication at the metacarpophalangeal level.

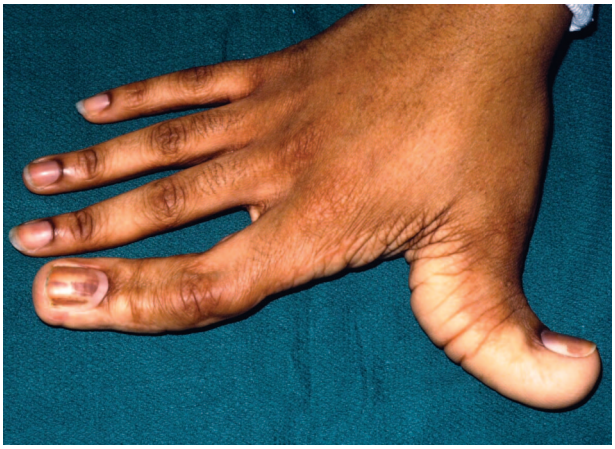

Fig. 10. Clinical appearance of thumb and index finger macrodactyly.

Overgrowth (Macrodactyly)

Macrodactyly, or gigantism, describes enlargement of all components of an affected digit and represents 1 percent of all congenital hand anomalies. Most cases are sporadic, without evidence of inheritance. Ninety percent of cases are unilateral, and the index finger is most commonly involved.[77] Several mechanisms have been proposed, including abnormal innervation leading to unimpeded growth, increased blood supply to the digit, and an abnormal humeral mechanism stimulating growth.[78–80] There seem to be two forms of macrodactyly: the static type, which is noted at birth and in which the affected digit grows at the same rate as the other digits; and the more common progressive type, in which the digit is large at birth and grows disproportionately. Macrodactyly may be associated with hypertrophy of the median, ulnar, or digital nerve, which may result in symptoms of a compression neuropathy that may require decompression.

Macrodactyly is extremely difficult to treat, but surgical intervention is often necessary, as the digit(s) lack function and may interfere with other normal digits. In addition, children with macrodactyly are subject to teasing and social embarrassment (Fig. 10). Surgical options include debulking the digit and/or disrupting further growth by obliterating the epiphyseal plates. Given the difficulty of treating this anomaly and the mediocre functional results, amputation should be strongly considered when only one or two digits are involved. If amputation is not warranted or if the parents refuse, staged debulking may be considered.

Congenital Constriction Ring Syndrome

Constriction Rings

Constriction rings may encircle a single digit or multiple digits or the entire limb of a newborn, causing varying degrees of vascular and lymphatic compromise. Constriction rings occur in one in 15,000 births.[81] The constrictions may be either circumferential or incomplete and may occur anywhere on the body, although they are most commonly seen around the limbs. The cause of this condition is not fully understood. According to the intrinsic mechanism, it is caused by a vascular disruption in the embryo.[82,83] According to the extrinsic mechanism, amniotic disruption causes release of amniotic bands that encircle and strangulate the limb or parts of a limb in utero.[84,85] Patterson classified constriction rings into four types.[86] Type 1 is a mild transverse or oblique digital groove. Type 2 is a deeper groove with an abnormal distal part. Type 3 is characterized by incomplete or complete syndactyly of the distal parts, which is termed acrosyndactyly. Type 4 is a complete amputation distal to the constriction. Treatment of a digit or limb threatened at birth by distal ischemia caused by a proximal constriction ring requires urgent release of the ring. Constriction rings may also affect the underlying nerves, necessitating decompression. In addition to releasing the constriction ring, the skin and subcutaneous tissues are rearranged with multiple Z- or

W-plasties. Some surgeons advocate release of constriction rings in two stages. Half of the circumference of the ring is excised at the first stage and the skin lengthened with multiple Z-plasties. The remaining 50 percent of the constriction ring is released in a similar manner at a second stage. However, it has been shown that constriction rings may be successfully released circumferentially around a limb or digit in a single stage.[87] If the constriction rings have transected extensor or flexor tendons, reconstruction with tendon grafts and/or tendon transfers may be necessary. Amputation may occasionally be required.

Flexion Deformities

Camptodactyly

Camptodactyly is a flexion deformity of the proximal interphalangeal joint that occurs most commonly in the small finger, although other fingers may be affected. The metacarpophalangeal and distal interphalangeal joints are not affected. This flexion deformity occurs in less than 1 percent of the population, and most patients are asymptomatic and do not seek treatment.[88] Two-thirds of the cases are bilateral, although the degree of flexion may not be symmetrical. The pathogenesis of this deformity remains unknown, although every structure surrounding the proximal interphalangeal joint has been implicated, including the skin and subcutaneous tissues, the ligaments (collateral, transverse, and oblique retinacular ligaments), the volar plate, the flexor tendons, the lumbricals, the interossei, and the extensor apparatus.[89] Camptodactyly has been divided into three types.[90] Type I deformities are the most common and are limited to the small finger. These become apparent during infancy and affect boys and girls equally. Type II deformities do not become apparent until preadolescence (ages 7 to 11) and affect girls more than they affect boys. Type II camptodactyly does not generally improve and may progress to a severe flexion deformity. Type III deformities are more severe, involving multiple digits of both extremities, and are generally associated with a variety of syndromes.

Treatment of camptodactyly depends on the severity of the deformity. Initially, physical therapy and splinting (static and dynamic) may be used to extend the finger. If the contracture progresses to greater than 60 degrees of flexion, surgery may be indicated. This includes exploration and release of any abnormal structure found limiting proximal interphalangeal joint extension, including skin, fascia, ligaments, and/or tendons. Transfer of the flexor digitorum superficialis to the extensor apparatus has been described to decrease proximal interphalangeal joint flexion and increase proximal interphalangeal joint extension.[91–93]

Congenital Clasped Thumb

Congenital clasped thumb (also known as isolated congenital thumb-palm deformity) represents a spectrum of thumb anomalies. It is more often bilateral and is seen in boys twice as often as in girls.[94] The mild form is caused by the absence or hypoplasia of the extensor mechanism. Moderate to severe forms are related to joint contractures, collateral ligament abnormalities, first web space contracture, and thenar muscle hypoplasia. A clasped thumb is commonly found in arthrogryposis or its associated syndromes.[95] In the classification system proposed by McCarroll and expanded by Mih, type I clasped thumb is flexible and has absence or hypoplasia of the extensor mechanism; type II clasped thumb is more complex, with additional findings of joint contracture, collateral ligament abnormality, first web space contracture, and thenar muscle abnormality; and type III clasped thumb is associated with arthrogryposis or its associated syndromes.[96,97]

The initial treatment of clasped thumb involves serial casting in extension and abduction for 3 to 6 months.[98] The goal of surgical management is to bring the thumb out of the palm and restore grasp by addressing any or all of the abnormalities of the thumb web space, intrinsic muscle contracture or deficiency, extensor tendon deficiencies, and joint stability.[96,99,100]

Arthrogryposis

Arthrogryposis (also known as arthrogryposis multiplex congenital) is a syndrome of nonprogressive joint contractures that is present at birth.[101] Multiple variants of arthrogryposis vary in presentation and severity, and the cause is unknown. This may affect all joints in all limbs. Commonly, the wrist and fingers are flexed and the thumb adducted and flexed into the palm.

Treatment should be individualized to achieve independent function. Manipulation of the deformities by a hand therapist shortly after birth may improve the range of motion and overall outcome.[102] If progress is not achieved by 6 months of age, surgical management should be considered. Delaying surgery until after 1 year of age makes improvement more difficult, as the contractures become more severe. Most surgeons advocate one-stage procedures that address the bone, joints, and soft tissue, as this gives the best results.[103]

CONCLUSIONS

Congenital differences of the upper limb represent a significant and unique challenge for the hand surgeon. In all cases, the ultimate goal is to provide a functional limb that can be integrated into the child's overall development. This goal may be met surgically or through specialized therapy and rehabilitation. Every case is unique and each patient (and parent) will have a different capacity to adapt. These differences should be taken into account before embarking on a long, often difficult reconstructive course. It should be made clear from the outset that the child will never have a "normal" hand. Once realistic expectations have been set, reconstruction and/or rehabilitation can commence.

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