Common Pediatric Congenital Conditions of the Hand

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Learning Objectives: After reading this article, the participant should be able to: 1. Accurately diagnose congenital differences that affect the hand. 2. Develop a surgical plan for syndactyly with regard to timing of intervention to prevent progressive deformity and principles to restore the commissure. 3. List the potential complications following polydactyly reconstruction. 4. State the indications for pollicization for thumb hypoplasia.

Summary: Congenital conditions of the hand are commonly encountered in a pediatric hand surgery practice. The physician must be comfortable and have a sound understanding of the diagnosis and treatment algorithm. Certain diagnoses are inheritable and require genetic testing and/or genetic counseling. Appropriate referral is necessary to facilitate education about the congenital difference and its effect on subsequent generations. Syndactyly, polydactyly, thumb hypoplasia, and cleft hand are particularly commonplace and are discussed in this article. The treatment principles and surgical techniques are emphasized to maximize hand function and aesthetic outcome. (*Plast. Reconstr. Surg.* 136: 241e, 2015.)

Syndactyly, polydactyly, thumb hypoplasia, and cleft hand are commonly encountered in a pediatric hand surgery practice. The conditions can be inheritable or are secondary to a spontaneous mutation. The classification scheme dictates the treatment algorithm. Accurate classification is necessary so that appropriate treatment can be recommended. This article discusses each entity with regard to classification and treatment principles. Surgical techniques are detailed to maximize hand function and aesthetic outcome.

SYNDACTYLY

Classification

The classification of syndactyly is dictated by the extent and degree of digital union.[1,2] The extent of syndactyly is defined as "incomplete" if the skin bridge does not extend the full length of the involved digits. Syndactyly is termed "complete" when the connection encompasses the entire length. Complete syndactyly can also result in a common fingernail called a synonychia, which complicates reconstruction (Fig. 1). The degree of syndactyly is defined as "simple" when the interconnection is soft tissue alone. If there is a bony connection, the syndactyly is "complex." Complex implies either fusion of adjacent phalanges or interposition of accessory bones (Fig. 2). Atypical forms of syndactyly are commonplace and these are characterized as "complicated." There can be elaborate soft-tissue interconnections or a hodgepodge of abnormal bones within the connected digits, especially in children with synpolydactyly (Fig. 3).

Surgical Timing

Most parents want to have their child's fingers separated as soon as possible. Complete syndactyly involving digits of unequal length (e.g., border digits) warrants early release, as the shorter digit tethers the longer digit during growth. Tethering

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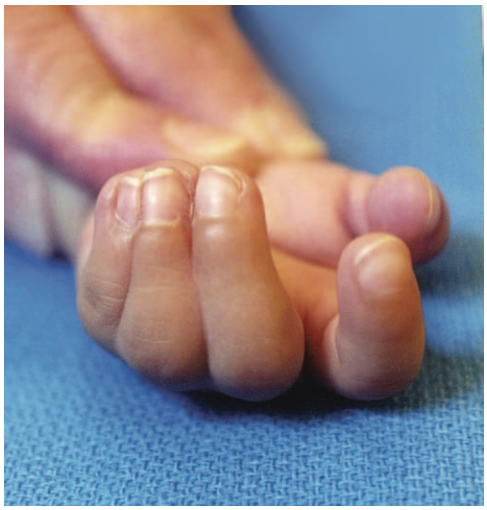

Fig. 1. Complete syndactyly with a synonychia across the conjoined digits. (Courtesy of Shriners Hospital for Children, Philadelphia.)

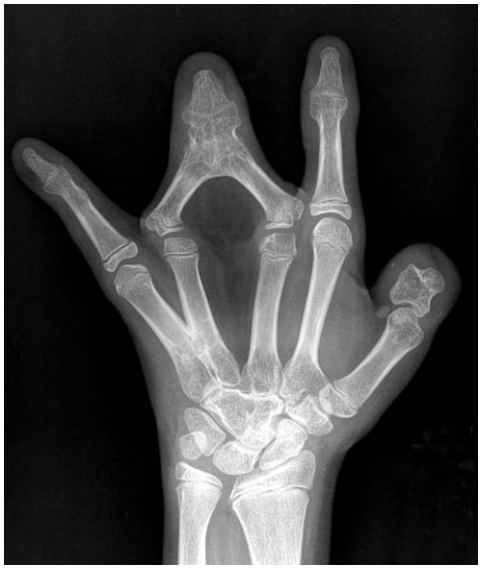

Fig. 2. Complex syndactyly with fusion of adjacent phalanges. (Courtesy of Shriners Hospital for Children, Philadelphia.)

leads to flexion contractures and rotational deformities of the longer digit. For these cases, we tend to perform surgery at 3 months of age as preferred by our anesthesia colleagues.

Syndactyly involving digits of relatively equal lengths (ring-long and long-index) is better treated later. Because the hand doubles in size in the first 2 years of life, delaying surgery results in a technically simpler operation with more predictable results.[3] We prefer to wait until the child is 18 to 24 months of age.

The numbers of web spaces involved influences surgical timing, as reconstruction should be performed only on one side of an affected

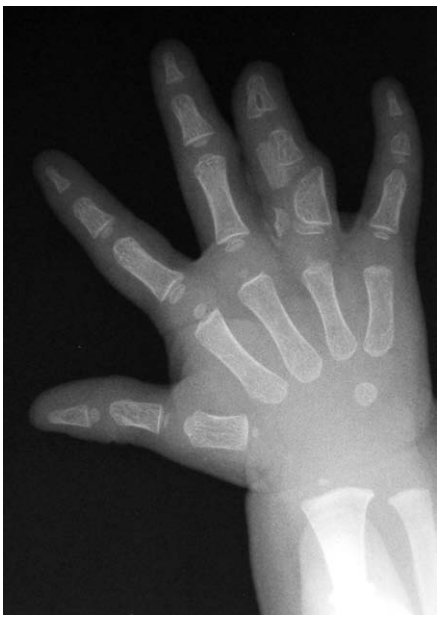

Fig. 3. Synpolydactyly, a hodgepodge of abnormal bones within the connected digits. (Courtesy of Shriners Hospital for Children, Philadelphia.)

digit at a time to avoid vascular compromise of the skin flaps or conjoined digits. Complete separation of three or more connected adjacent digits requires staged surgical procedures to comply with this principle. When syndactyly involves all the web spaces, the initial operation focuses on release of the first and third web spaces. The next procedure concentrates on release of the second and fourth web spaces. Thus, two procedures are able to achieve separation of all of the digits.

Incomplete syndactyly can undergo elective separation, as tethering is not an issue. In addition, many incomplete syndactyly separations can be closed completely by using a variety of local flaps based on Z-plasty principles.

Skin Grafting

Complete syndactyly release typically produces a skin deficiency that requires additional skin grafting. A skin graft is usually required because the circumference of two separated digits measured separately is 22 percent greater than the circumference of two conjoined digits (Fig. 4).[2] Full-thickness skin grafts are preferred over split-thickness skin grafts to lessen graft contracture. There are numerous donor sites; we prefer the distal wrist crease for color, consistency, and hairlessness (Fig. 5).

In complete syndactyly with a synonychia, coronal skin flaps across the tip of the conjoined

digits are necessary to reconstruct the nail fold. Long-term nail problems are endemic without an adequate nail fold.[1]

A technique to decrease digital volume by extensively defatting the full length of the fingers and the interdigital space can achieve complete closure with minimal tension (**Level of Evidence: Therapeutic, IV**).[4] We concur that the amount of skin graft required can be lessened by defatting of the fingers while preserving the dorsal venous system. However, aggressive defatting should be avoided because of the potential for iatrogenic vascular injury and the risk of producing a thin finger compounded by the involution of the fat of infancy when the child gets older.[5]

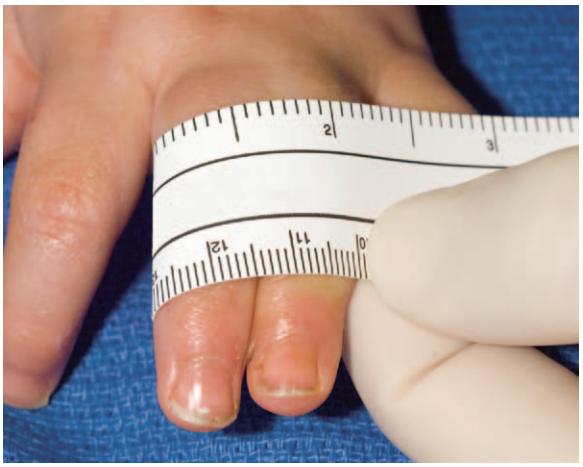

Fig. 4. A tape measure can be used to demonstrate that the circumference of two separated digits is greater than the circumference of two conjoined digits. (Courtesy of Shriners Hospital for Children, Philadelphia.)

Commissure Reconstruction

Supple skin is required within the commissure to facilitate proper digital movement in the coronal plane and flexion/extension in the sagittal plane. Skin graft within the commissure should be avoided. Instead, a local random flap should be used to resurface the commissure after digital separation.

Contraindications to Surgery

There are few contraindications to simple syndactyly separation. In contrast, complicated and/or complex syndactyly may defy surgical separation (Fig. 2). In some children, the connected digits are stable and function better compared with two unstable separate units. The decision not to operate requires a thorough examination and explanation to the parents that discusses that function trumps form. This decision is frequently more difficult to explain to parents compared with the decision to operate.

Surgical Techniques for Reconstruction

The most versatile technique for complete or nearly complete syndactyly reconstruction is a combination of flaps and grafts.[1–3,5] We prefer a modification of the flaps with a large trapezoidal dorsal flap to reconstruct the commissure and multiple small zigzag type flaps for the fingers. Full-thickness skin grafting is used to fill in the inevitable gaps (Figs. 6 through 12). Straight longitudinal incisions are tempting but should be avoided, as longitudinal scarring would lead to flexion contractures along the digits.

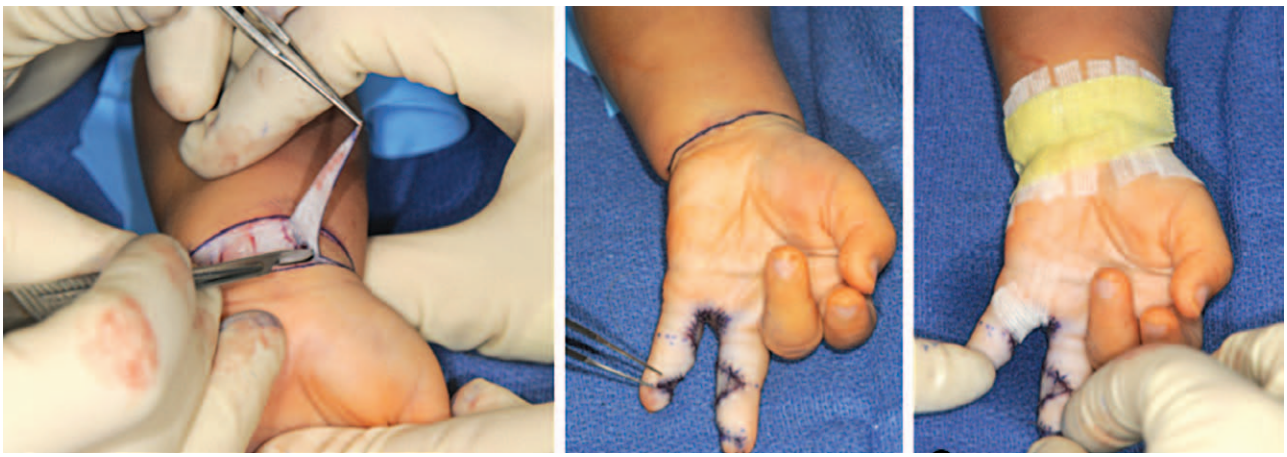

Fig. 5. Skin graft harvest and closure. (*Left*) Elliptical graft centered over the proximal wrist crease is harvested sharply superficial to the small cutaneous veins. (*Center*) If dissected at the right level, the graft does not have to be defatted and the bed does not bleed. Closure typically results in a thin scar concealed within the skin crease. (*Right*) Within the fingers, the graft recipient site is shielded from shear and compressed by petroleum dressing and gauze held down by noncircumferential adhesive strips. (Courtesy of Shriners Hospitals for Children, Philadelphia.)

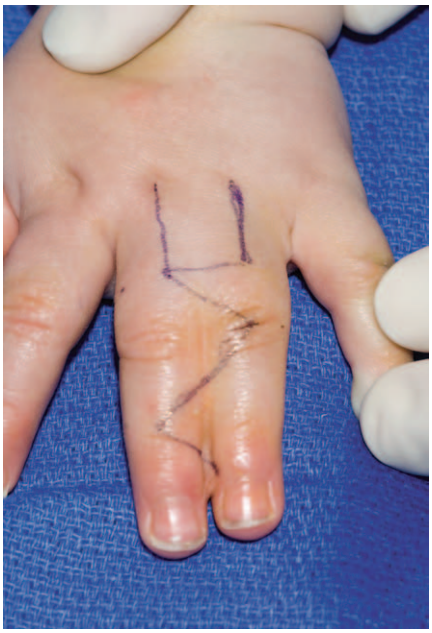

Fig. 6. Dorsal commissure flap begins at the level of the metacarpal heads and includes two-thirds the length of the proximal phalanx. (Courtesy of Shriners Hospitals for Children, Philadelphia.)

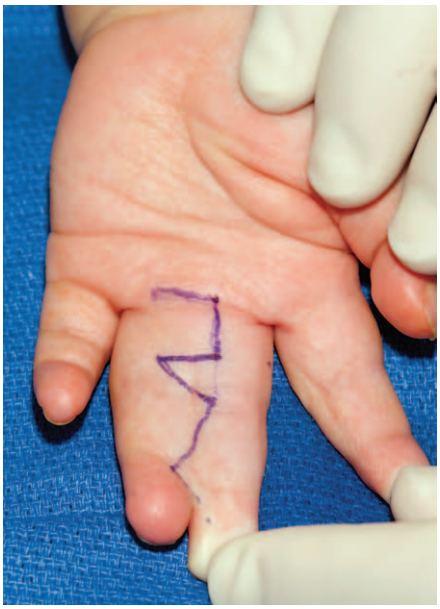

Fig. 7. A palmar rectangular flap is fashioned to resurface the proximal area of a digit adjacent to the commissure. (Courtesy of Shriners Hospitals for Children, Philadelphia.)

The postoperative dressing is crucial for limiting shear between the skin graft and underlying bed. Because the early oxygen and nutrient supply initially occurs by means of imbibition followed by inosculation, any hematoma or seroma can lead to early graft loss. After 3 weeks, secondary vascular ingrowth has begun and the graft is robust enough to survive outside the postoperative dressing.

Reconstruction of the Thumb-Index Web Space

Reconstruction of the thumb-index web space can be more challenging and requires a different set of surgical techniques. For mild web-space narrowing with no flexion contracture of the thumb, we prefer the classic four-flap Z-plasty (Fig. 13). The skin flaps allow excellent access to the adductor and the first dorsal interosseous muscles that may require partial release for adequate web-space breadth.

Patients with a tight thumb-index web space and a metacarpophalangeal joint flexion contracture of the thumb (clasp thumb) are ideal candidates for the index rotation flap (i.e., stiletto flap).[6] A stiletto-shaped flap is elevated from the radial aspect of the index finger and rotated into the area of the metacarpophalangeal joint crease. This expands the thumb-index web space and

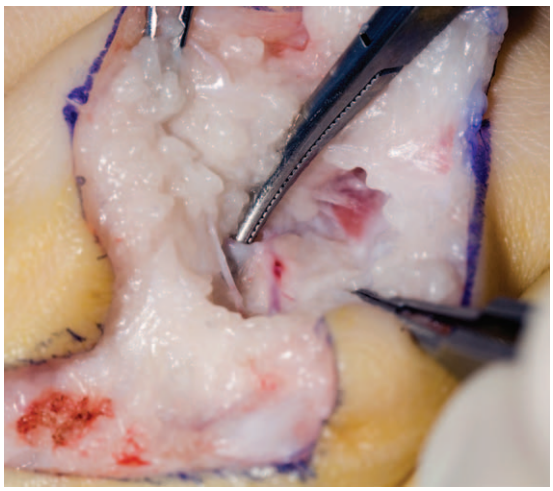

Fig. 8. From the dorsal side, the volar neurovascular structures are visualized. (Courtesy of Shriners Hospitals for Children, Philadelphia.)

resurfaces the volar thumb (Fig. 14). The donor defect can be closed.

If the thumb-index web-space syndactyly is complete or nearly complete, the preferred option is the modified dorsal rotation advancement flap described by Abdel Ghani (**Level of Evidence: Therapeutic, IV**).[7] Although this flap typically requires adjunctive skin grafting, it provides extensive deepening and widening of the web space. This flap has been a welcome addition to our therapies for thumb-index syndactyly, such as Apert syndrome (Fig. 15).

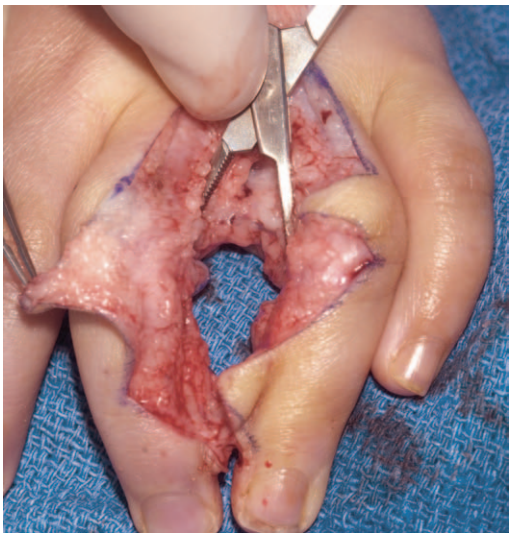

Fig. 9. Lateral spreading of the digits places the intervening tissue under tension and facilitates digital separation. (Courtesy of Shriners Hospitals for Children, Philadelphia.)

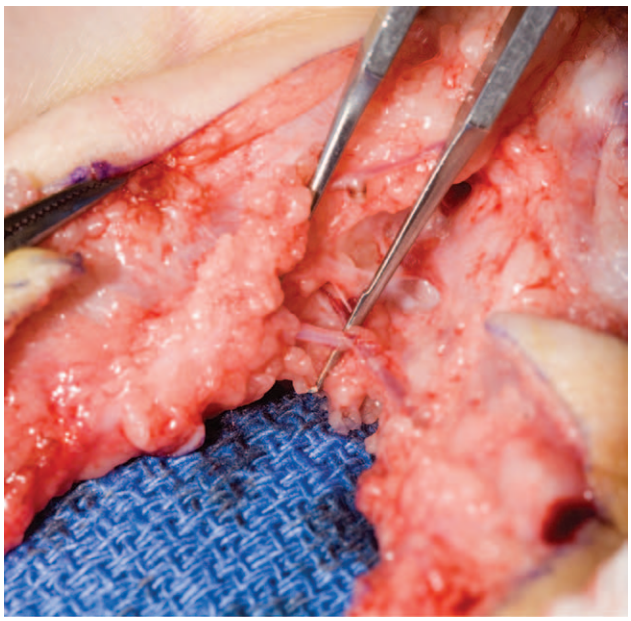

Fig. 10. The bifurcation between the common and proper neurovascular structures requires assessment. (Courtesy of Shriners Hospitals for Children, Philadelphia.)

Complications

Intraoperative complications are uncommon. Inadvertent nerve or artery injury requires immediate attention. Venous complications are more common and often related to too tight a closure. Releasing a few sutures often solves the problem. Delayed complications during growth are commonplace. Scar hypertrophy can frequently be managed by therapy consisting of massage and silicone elastomer. Extensive scar hypertrophy is associated with syndactyly combined with macrodactyly, which may require adjunctive medications that limit this intensive response.[8] Distal migration of the web over time (i.e., web creep) can occur during growth (Fig. 16).[9] Mild migration does not require treatment, whereas substantial creep requires revision surgery.

POLYDACTYLY

Polydactyly can occur along the ulnar border (postaxial), within the hand (central), or along the radial corridor (preaxial). Each form has a separate algorithm for treatment. Postaxial polydactyly is more prevalent in blacks and preaxial is more common in whites.[10]

Classification and Treatment

Postaxial Polydactyly

The postaxial polydactyly digit is either well-developed (type A) or rudimentary and pedunculated (type B).[11] A small nubbin (type B) can be safely removed by ligating the base in the nursery (Fig. 17). Suture or vessel clips can be secured to the base of the digit. The digit will turn gangrenous, die, and fall from the hand. The parents must be willing to accept this series of events. Another option is to remove the digit in the office. Local anesthesia is applied to the base of the digit and electrocautery used to delete the digit. The parents must be agreeable to this treatment as well. Unprepared parents may develop syncope and you may be treating two patients instead of one. A residual bump or nubbin is the most common complication of ligation. Simple excision under local anesthesia in the office may give a better long-term result.

A well-developed digit (type A) requires operative ablation (Fig. 18). The extra digit is removed, and any important functional parts (e.g., ulnar collateral ligament and abductor digiti quinti) are transferred to the adjacent finger. An elliptical incision is designed around the digit. The incision is extended in a proximal direction to allow ample exposure. The digital nerves to the extra digit are incised and the digital arteries are coagulated. If the ablated digit contains the abductor digiti quinti, the muscle and tendon are transferred to the adjacent digit to reestablish digital abduction. The skin is closed with absorbable suture and the limb immobilized in a long-arm cast for 3 weeks for protection.

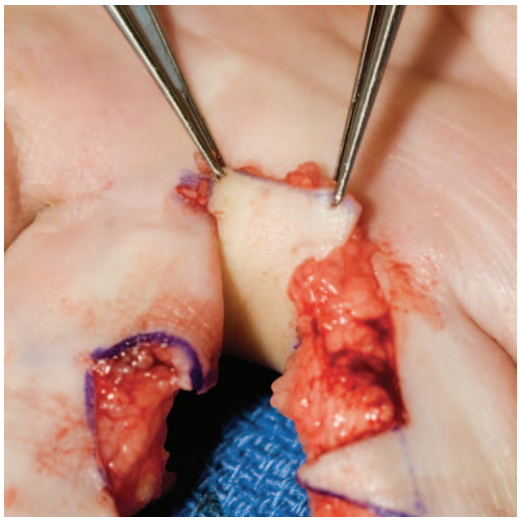

Fig. 11. The commissure flap is sutured first to assess position and shape of the web space. (Courtesy of Shriners Hospitals for Children, Philadelphia.)

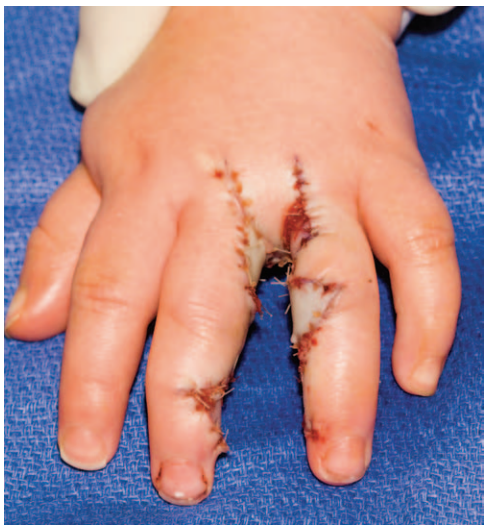

Fig. 12. Appearance after flap closure and full-thickness skin grafting. (Courtesy of Shriners Hospitals for Children, Philadelphia.)

Preaxial Polydactyly

Thumb duplication has been classified according to the degree of skeletal replication (Table 1).[12] Type IV is the most common type, with duplication of the proximal and distal phalanges that articulate with a bifid metacarpal head.

Dobyns and colleagues[13] defined the treatment principle of "spare parts" surgery for thumb reconstruction. The goal is to produce the best thumb possible using portions of each component. The ultimate goal is to achieve a thumb of sufficient size that is aligned along its longitudinal axis with stable joints and adequate motion. Reconstruction or preservation of the collateral ligaments is mandatory for duplications that arise from the joint surface (Wassel types II, IV, and VI).

Each degree of thumb duplication produces different obstacles to reconstruction. The surgical principles, however, are similar. For type IV duplication, the dominant thumb is selected and the lesser counterpart is ablated. The ulnar thumb is often dominant, which preserves the ulnar collateral ligament for pinch. The technique is performed in a stepwise fashion (Fig. 19). The soft tissue(s) from the radial thumb are used to enhance the ulnar thumb. The skin incision is designed to augment the soft tissue for the reconstructed thumb and to use any necessary pulp tissue and/or eponychial fold. This will increase the size of the thumb with the intent to generate a thumb similar in size to the contralateral side. Proximal exposure is achieved by a zigzag approach for full exposure. The collateral ligament is reconstructed using an osteoperiosteal sleeve from the deleted thumb (**Level of Evidence: Therapeutic, IV**).[14] The joint surface of the metacarpal must be visualized for the separate facet that articulates with the deleted radial component. Removal of this facet is necessary to correctly align the thumb and to prevent an angular deformity. Additional steps may be required to correctly align the thumb. For example, an angulated metacarpal may require a closing wedge osteotomy to align the metacarpophalangeal joint. Tendon rearrangement is also required to centralize the tendons and avoid any eccentric pull. If the abductor pollicis brevis inserts into the radial thumb, it must be included in the osteoperiosteal sleeve that contains the radial collateral ligament and reattached to the reconstructed thumb.

Stabilization of the thumb is obtained by means of a percutaneous Kirschner wire placed antegrade from the thumb tip or obliquely across the metacarpophalangeal joint. The incision is closed with absorbable sutures and a long-arm thumb spica cast applied for 4 to 5 weeks. The wire is removed in the office and a short-arm thumb spica splint is fabricated. Therapy is started for range of motion and the thumb gradually incorporated into daily use. The splint is discontinued 8 to 12 weeks after surgery, depending on the activity and fearlessness of the child.

The results following thumb reconstruction vary with the complexity of the thumb duplication. Reasonable results are readily obtainable in type I, II, and IV duplications.[15] Less pleasing results are common in type III, V, and VI

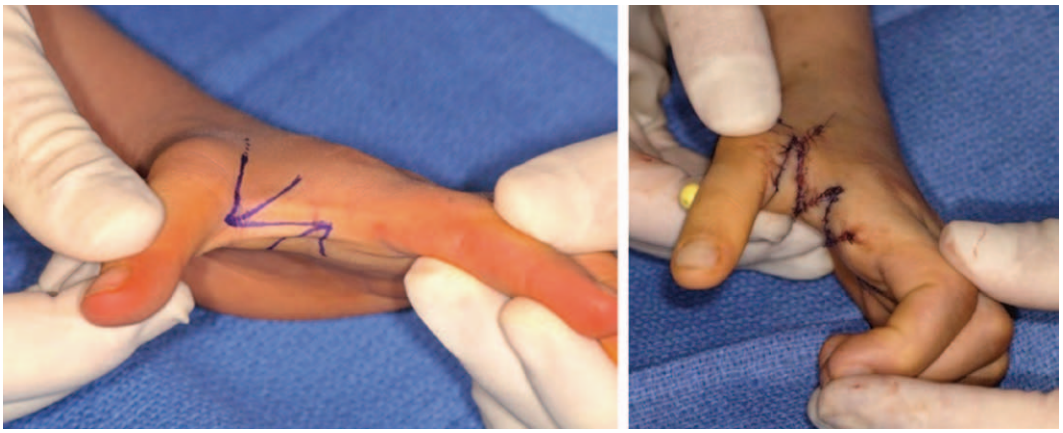

Fig. 13. Narrowed thumb-index web space treated with four-flap Z-plasty. (*Left*) Skin design for ample web deepening. (*Right*) After web widening and flap closure. (Courtesy of Shriners Hospitals for Children, Philadelphia.)

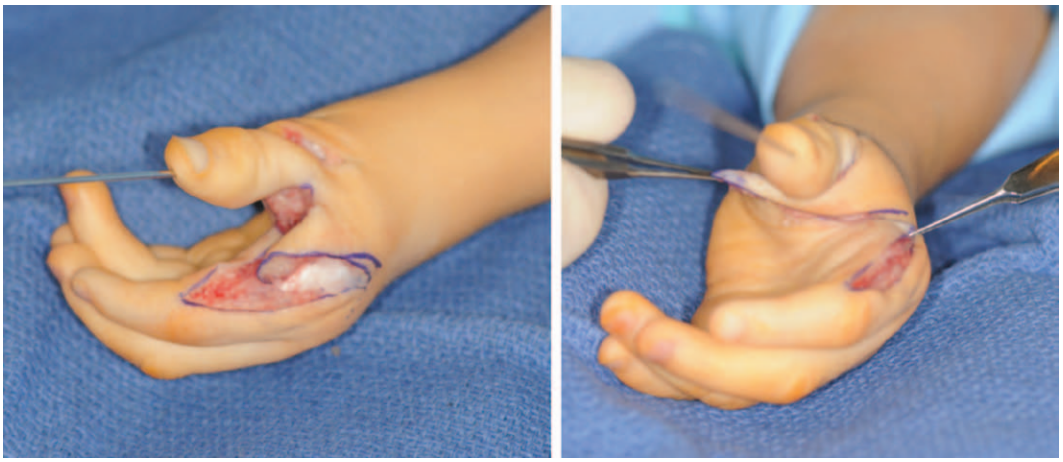

Fig. 14. Narrowed thumb-index web space in both the coronal and sagittal planes treated with a stiletto flap. (*Left*) Stiletto flap elevated from the radial side of the index finger. (*Right*) Flap mobilized to widen the web space and to cover the volar aspect of the thumb. (Courtesy of Shriners Hospitals for Children, Philadelphia.)

duplications and triphalangeal thumbs. Sources of dissatisfaction include smaller size, joint instability, stiffness, and angulation. Revision surgery can often improve outcome and may include osteotomy to improve alignment or arthrodesis to gain stability.

Central Polydactyly

Central polydactyly comes in various forms and diagnoses. Pure polydactyly contains five fingers, and the extra digit is often hard to define. The hand appears "perfect" except for five fingers. The parents may request treatment to avoid teasing. The ulnar digit is removed by ray resection using the tenets of spare parts surgery discussed above. Central polydactyly may be hidden within a syndactyly (i.e., synpolydactyly). In these cases, the extra digit can be removed at the time of syndactyly reconstruction. Central polydactyly of the ring finger combined with syndactyly is inheritable and has been linked to a gene mutation (*HOXD13* gene) on chromosome 2.[16] In these cases, the polydactyly can be bizarre, with multiple interconnections between the digits (Fig. 20). Surgical separation is very difficult and the results often disappointing. A wide surgical exposure is necessary, and intraoperative flexibility is required to restore the best functioning digit using parts of each digit.

THUMB HYPOPLASIA

Thumb hypoplasia can range from a slight decrease in thumb size to complete absence.

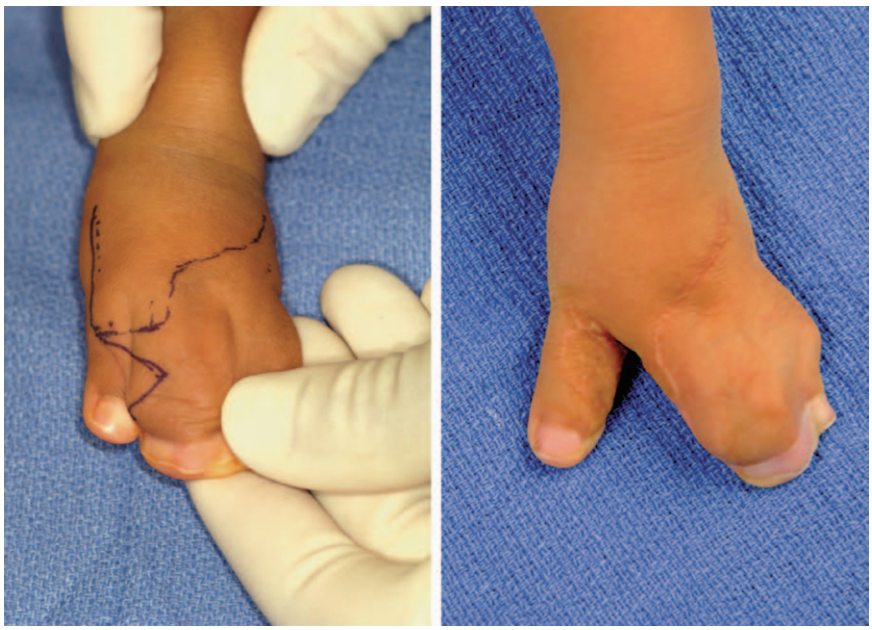

Fig. 15. A 1-year-old with nearly complete thumb-index web space syndactyly. (*Left*) Ghani flap drawn on the dorsum of the hand. (*Right*) Healed flap with ample first web space. (Courtesy of Shriners Hospitals for Children, Philadelphia.)

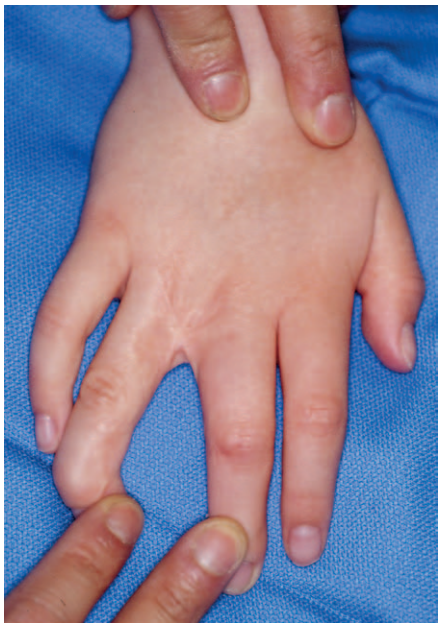

Fig. 16. Web creep and angulatory deformity years after syndactyly separation. (Courtesy of Shriners Hospitals for Children, Philadelphia.)

Fig. 17. A well-developed digit (type A) requires formal surgery with transfer of any collateral ligament and/or abductor digiti quinti to the adjacent finger. (Courtesy of Shriners Hospitals for Children, Philadelphia.)

Thumb hypoplasia is part of the radial longitudinal deficiency spectrum and therefore other organ systems may be affected. Workup must include evaluation for associations such as VACTERL (vertebral anomalies, anal atresia, tracheoesophageal fistula, renal abnormalities, and limb differences), and genetic conditions such as Fanconi anemia, Holt-Oram syndrome, and CHARGE syndrome (coloboma of the eye, heart defects, atresia of the nasal choanae, retardation of growth and/or development, genital and/or urinary abnormalities, and ear abnormalities and deafness) (Fig. 21). Appropriate workup is mandatory and can

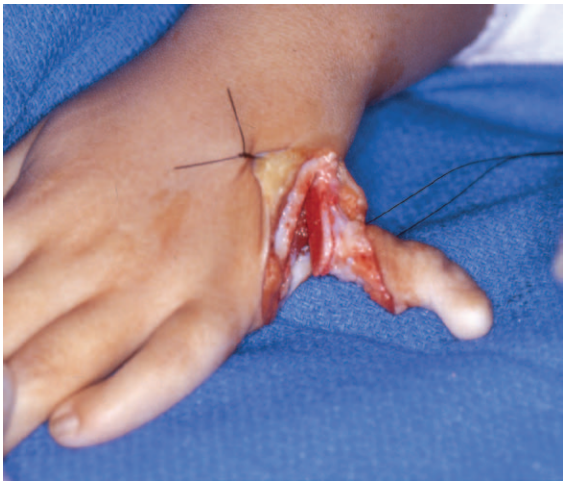

Fig. 18. Web creep and angulatory deformity years after syndactyly separation. (Courtesy of Shriners Hospitals for Children, Philadelphia.)

Table 1. Classification of Duplicated Thumbs

Type	Duplicated Elements
I	Bifid distal phalanx
II	Duplicated distal phalanx
III	Bifid proximal phalanx
IV	Duplicated proximal phalanx*
V	Bifid metacarpal phalanx
VI	Duplicated metacarpal phalanx
VII	Triphalangeal component

*Most common type.

be lifesaving to the child. Thrombocytopenia absent radius syndrome is also associated with thumb hypoplasia, but these children have distinct flat, broad thumbs (**Level of Evidence: Therapeutic, IV**).[17]

Classification

The modified Blauth classification is used to describe the degree of hypoplasia and guide treatment (Table 2).[18] A type I hypoplastic thumb is slightly smaller than a typical thumb but is stable, with intact intrinsic and extrinsic musculature. Type I thumbs have good overall function. A type II thumb has narrowing of the thumb-index web space, aplasia or hypoplasia of the thenar muscles, and instability of the thumb metacarpophalangeal joint. A type III thumb has all of the type II deficiencies plus extrinsic muscle aplasia or hypoplasia. Type III thumbs are further subclassified based on the stability of the carpometacarpal joint. A type A thumb has a stable carpometacarpal joint and a type B thumb has an unstable carpometacarpal joint. Type IV deficiency is a floating thumb or "pouce flottant," a rudimentary digit connected to the hand by only skin and a neurovascular bundle. Type V deficiency is defined as a complete absence of the thumb.

Treatment

Type I hypoplasia requires no treatment. Thumb reconstruction for types II and IIIA requires addressing each of the deficiencies to obtain a functioning thumb. Pollicization remains the procedure of choice for type IIIB, IV, and V hypoplasia. We prefer to perform surgery between 1 and 2 years of age, which corresponds with the development of radial-side pinch or prehension. The timing of intervention, however, remains controversial.

Thumb Reconstruction for Type II and IIIA Hypoplasia

The surgery should address the following: (1) narrowing of the first web space, (2) lack of opposition because of the absence or weakness of intrinsic muscles, and (3) metacarpophalangeal joint instability. Extrinsic muscle weakness is less commonly encountered and is more difficult to address. Subtle deficiencies in the flexor pollicis longus or extensor pollicis longus function are usually ignored.

The thumb-index web narrowing is usually mild and can be treated with a release of the skin and tight fascia. The skin is lengthened with a four-flap Z-plasty (Fig. 22). The thenar muscle absence is resolved by opposition transfer. We prefer the ring finger flexor digitorum superficialis tendon as the donor (Fig. 23).[19] This muscle tendon is synergistic, and the tendon can also be used to stabilize the metacarpophalangeal joint. If the flexor digitorum superficialis tendons are anomalous and unavailable, alternative donors include the abductor digiti minimi or the extensor indicis proprius.

The management of metacarpophalangeal joint instability depends on the direction and extent of instability. Unidirectional valgus instability is reconstructed at the same time as flexor digitorum superficialis opposition transfer by passing the tendon through the bone and using the terminal segment to reconstruct the ulnar collateral ligament (Fig. 24). Multidirectional metacarpophalangeal joint instability is treated with metacarpophalangeal joint chondrodesis, as reconstruction of both collateral ligaments has been unpredictable.

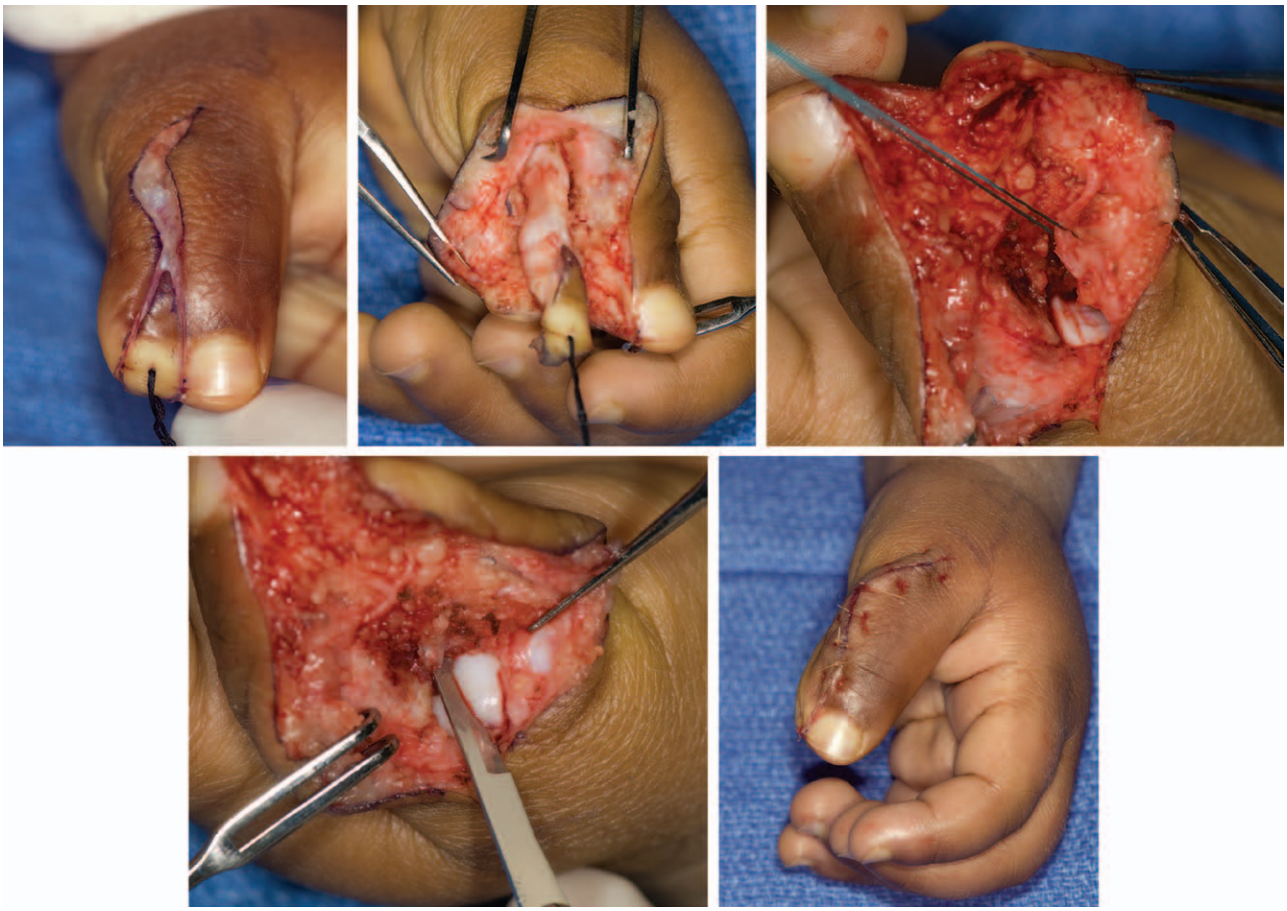

Fig. 19. A 1-year-old with Wassel type IV right thumb duplication. (*Above*, *left*) Skin incision for deletion of the radial component and reconstruction of the ulnar thumb. (*Above*, *center*) Radial component skeleton isolated with preservation of important soft-tissue structures. (*Above*, *right*) Preserved osteoperiosteal sleeve is raised from the deleted thumb with the collateral ligament and abductor pollicis brevis. (*Below*, *left*) Removal of the radial facet from the metacarpal head with a scalpel. (*Below*, *right*) Final reconstruction, with good alignment, stability, and bulk. (Courtesy of Shriners Hospital for Children, Philadelphia.)

Pollicization for Type IIIB to V Thumb Hypoplasia

Pollicization remains the criterion standard for reconstruction of these severely deficient thumbs. Despite advances in microsurgical techniques, the absence of a stable carpometacarpal joint precludes successful reconstruction. The decision to proceed with pollicization is difficult for parents, and they often benefit from seeing photographs of pollicizations, speaking with other parents, and meeting other children who have undergone pollicization. We use this multifaceted approach in our practice to help parents make the right decision for them.

Pollicization remains a challenging procedure that requires a stepwise approach (Table 3). (**See Video, Supplemental Digital Content 1**, which displays a detailed pollicization of the hand. This video is available in the "Related Videos" section of the full-text article on PRSJournal.com or at ***http://links.lww.com/PRS/B365***.) Some of the critical points are: (1) skin design to avoid a scar in the thumb-index web space, (2) transfer of the intrinsic muscles to provide thumb adduction and abduction, (3) epiphysiodesis of the metacarpal head to prevent excessive growth of the pollicized index finger, (4) extension of the metacarpophalangeal joint to avoid thumb hyperextension, and (5) fixation of the new thumb in the correct amount of abduction and rotation (Figs. 25 through 28).[20]

After completion of the procedure, the tourniquet is deflated and the "thumb" observed for 5 minutes to confirm adequate venous return. The postoperative dressings are crucial. A long-arm soft cast (3M Scotchcast Soft Cast Casting Tape; 3M,

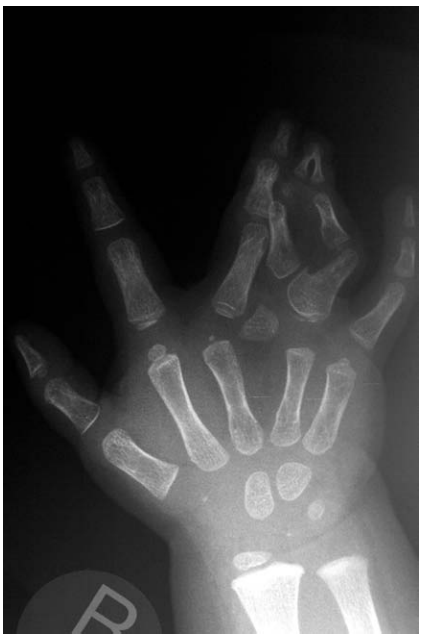

Fig. 20. Synpolydactyly with bizarre, multiple interconnections between the digits. (Courtesy of Shriners Hospitals for Children, Philadelphia.)

St. Paul, Minn.) is applied with the elbow flexed to greater than 100 degrees to decrease the chance of inadvertent removal. The child is admitted overnight and the arm is elevated to promote venous drainage.

Results of Thumb Reconstruction and Pollicization

As a general rule, results of thumb reconstruction for hypoplasia are consistently good compared with preoperative function. However, the surgical plan must address each deficient component of the thumb.[19] Comparison between studies and "thumbs" is difficult. There is considerable heterogeneity with regard to patient populations, severity of hypoplasia, and surgical techniques.

Video 1. Supplemental Digital Content 1, which displays a detailed pollicization of the hand, is available in the "Related Videos" section of the full-text article on PRSJournal.com or at ***http://links.lww.com/PRS/B365***.

The results following pollicization are dependent on the status of the index finger and its surrounding musculature. Pollicization of the index finger provides a better result in isolated thumb hypoplasia compared with patients with a hypoplastic or absent radius.[21] A mobile index finger transferred to the thumb position provides stability for grasp and mobility for fine pinch (Fig. 29). A stiff index finger, however, provides a stable thumb for gross grasp but will not be nimble enough to participate in pinch. The results of pollicization continue into adulthood, with long-term studies showing continued use.[22] Pollicization is a demanding procedure, and complications can occur even when it is performed by an experienced surgeon. Acute complications are less common than chronic problems. Technical errors can result in poor rotational position, carpometacarpal joint hyperextension, or cicatrix within the

Fig. 21. Coloboma of the eye associated with CHARGE syndrome. (Courtesy of Shriners Hospitals for Children, Philadelphia.)

Table 2. Thumb Deficiency Classification and Treatment Paradigm

Type	Findings	Treatment
I	Minor generalized hypoplasia	No treatment
II	Intrinsic thenar muscles hypoplasia	Opponensplasty
	First web space narrowing	First web release
	UCL insufficiency	UCL reconstruction
III	Similar findings as type II plus:	
	Extrinsic muscle and tendon abnormalities	
	Bone deficiency	
	A: Stable TMC joint	Reconstruction
	B: Unstable TMC joint	Pollicization
IV	"Pouce flottant" or floating thumb	Pollicization
V	Absent thumb	Pollicization

TMC, trapeziometacarpal; UCL, ulnar collateral ligament.

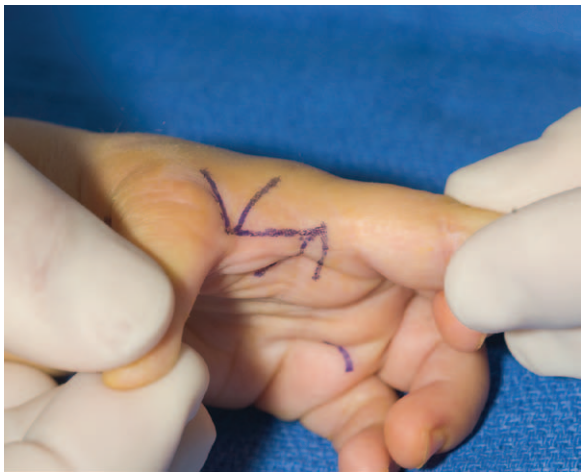

Fig. 22. The thumb-index web narrowing is usually lengthened with a four-flap Z-plasty. (Courtesy of Shriners Hospitals for Children, Philadelphia.)

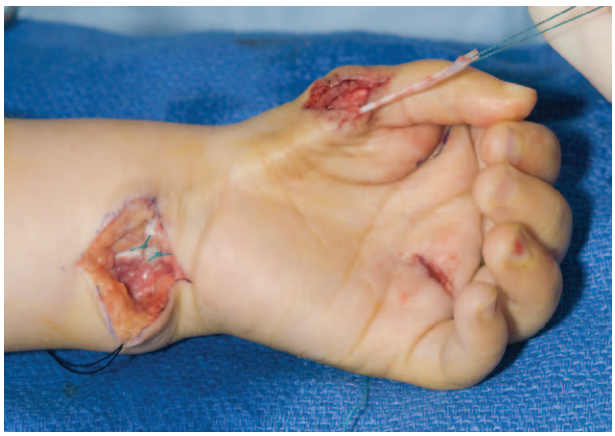

Fig. 23. Ring finger flexor digitorum superficialis tendon as the donor for opposition transfer. (Courtesy of Shriners Hospitals for Children, Philadelphia.)

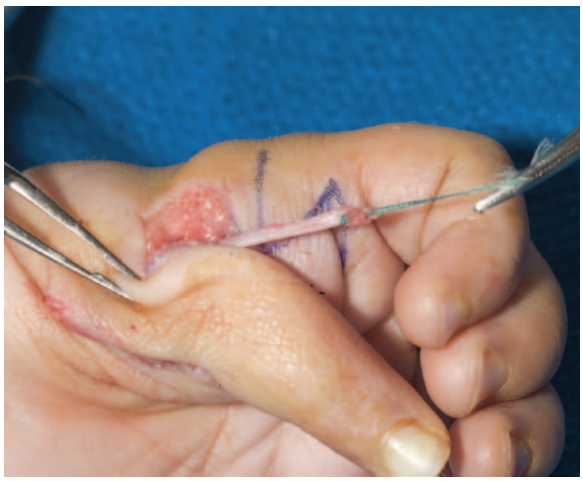

Fig. 24. Valgus instability corrected by passing the tendon through the bone and using the terminal flexor digitorum superficialis segment to reconstruct the ulnar collateral ligament. (Courtesy of Shriners Hospitals for Children, Philadelphia.)

thumb index web space. Secondary procedures, such as osteotomy, tendon transfer, or scar revision can improve the outcome.

CENTRAL HAND DEFICIENCIES (CLEFT HAND)

Central deficiency or cleft hand is characterized by a V-shaped cleft in the center of the hand, with or without missing digits. There are variable phenotypes, depending on the amount of digital suppression. The manifestations of cleft hand vary from a minor cutaneous cleft without absence of any fingers to a severe form with only the small finger present. Cleft hand can result from a spontaneous mutation or is inherited as an autosomal dominant trait.[23] Cleft hand is associated with a number of syndromes, most commonly split-hand/split-foot syndrome and ectrodactyly, ectodermal dysplasia, and cleft lip/palate syndrome. Syndactyly between the digits bordering the cleft and the thumb-index is common. The extent of syndactyly is one factor that drives the treatment paradigm. The other main feature is the status of the first web space (Table 4).[24]

Treatment

Many cleft hands do not require treatment. The presence of a thumb, thumb-index web space, and ulnar digit(s) results in outstanding function. The main issue becomes the social stigmata and teasing associated with differences. The

Table 3. Stepwise Approach to Pollicization

Step	Technique	Rationale
Exsanguination	Moderate	Vessel identification
Skin incision	Ezaki design	More glabrous skin along the palmar aspect of the thumb and excellent thumb-index web space
Isolation of palmar neurovascular bundles	Loupe magnification and meticulous dissection	Preserve sensibility and circulation to the index finger
Microdissection of common digital nerve	Intrafascicular dissection	Mobilize nerve for tension-free pollicization
Ligate proper digital artery to radial side of the long finger	Ligature clip	Allows constant visualization throughout the procedure
Release A1 pulley to the index finger		Prevent buckling of flexor tendons after pollicization
Incise intermetacarpal ligament		Allows repositioning of the index finger
Elevation of dorsal skin with preservation of dorsal veins		Delaying dorsal exposure allows veins to be filled with blood
Extensor tendons freed from adjacent		Confirm appropriate line of pull to index finger pollicization
Extensor and flexor tendons are not shortened		Adapt and shorten over time
Elevation of the first dorsal and palmar muscles from the index metacarpal and metacarpophalangeal joint with a strip of extensor hood	Sharp dissection	Muscles will be advanced to proximal interphalangeal joint and length is necessary
Identify and tag the radial and ulnar lateral bands about the proximal interphalangeal joint	Pull on lateral band until desired function is evident and tag band with suture	Before bony resection, identification is easier
Shorten the index finger by removing the majority of the metacarpal bone, including physis ablation	Fine-blade saw to cut metacarpal perpendicular to bone through its metaphyseal portion; distal cut directly through physis (epiphysiodesis)	Index too long for a thumb; physeal ablation prevents continued metacarpal growth
Reposition index metacarpophalangeal joint into hyperextension	Fixation of the index metacarpophalangeal joint into hyperextension using a nonabsorbable suture placed through the epiphysis and dorsal capsule	Prevents unwanted thumb carpometacarpal joint hyperextension
Kirschner wire is passed anterior to the metacarpal epiphysis, into the proximal phalanx, and out the proximal interphalangeal joint	Wire driver	This Kirchner wire is used as a joystick for index finger positioning and ultimate fixation
Align the index finger into the thumb position with 45 degrees of abduction and between 100 and 120 degrees of pronation	Metacarpal epiphysis is aligned anterior to its remaining base and Kirschner wire drilled retrograde across the metacarpal base to secure the position	Replicate thumb position
Tendon transfer to restore intrinsic function to the pollicization	First dorsal interosseous sutured into the radial lateral band and the first palmar interosseous sutured into the ulnar lateral band	Maximize function in grasp and pinch
Inset skin with absorbable suture	Skin inset advanced and inset along the palmar aspect of the "thumb"; inset web space skin; trim any excess skin	Index appearance similar to thumb; avoid suture line in thumb-index web space
Deflation of tourniquet and meticulous postoperative dressings	Bulky hand dressing with long-arm soft cast	Ensure circulation, protect pollicization, decrease chances of inadvertent dressing removal

main indications for treatment are syndactyly and the deficient first web space.

The management of finger syndactyly adjacent to the cleft is governed by the same principles highlighted earlier. The incidence of neurovascular abnormalities is increased in syndactyly associated with cleft hand. The thumb index web space must be assessed carefully for narrowing, which limits grasping of large objects. A narrow space requires the child to hold large objects within their cleft, thus further widening the separation. The management of the cleft is related directly to its degree of narrowing. A mild narrowing (type IIA) can be widened with release of the connective tissue and Z-plasty of the skin. A severely narrowed web space (type IIB) requires adjacent tissue transfer, usually a random flap from the radial side of the index or dorsum of the hand. When there is a syndactyly between the thumb

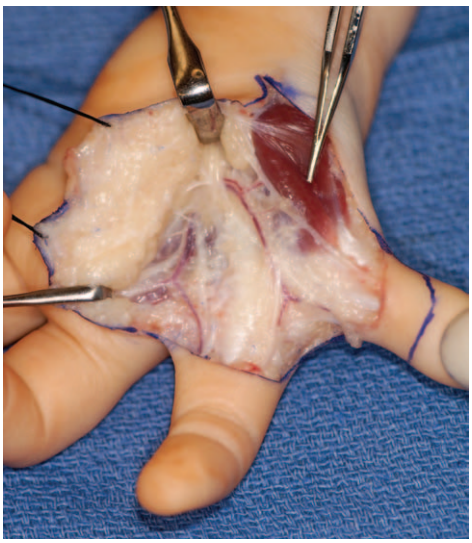

Fig. 25. Following gentle exsanguination, the palmar skin is incised and the neurovascular bundles are isolated. (Courtesy of Shriners Hospitals for Children, Philadelphia.)

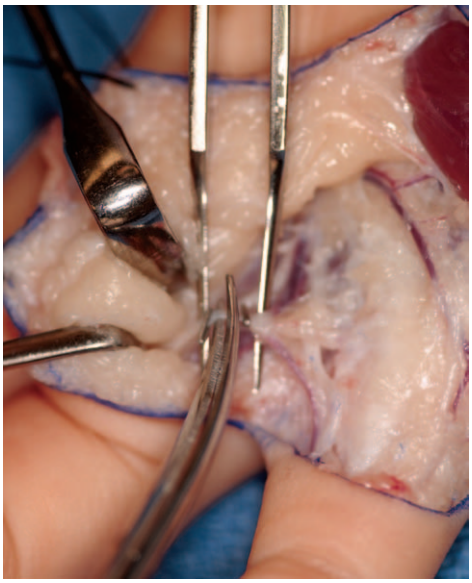

Fig. 26. Ligation of the proper digital artery to the long finger with ligature clips. (Courtesy of Shriners Hospitals for Children, Philadelphia.)

and index (type III), more creative treatment is necessary. The basic tenet is to close the cleft and to use the cleft skin to resurface the separated thumb and index. We prefer to use the technique described by Snow and Littler (Fig. 30).[25] (**See Video, Supplemental Digital Content 2**, which displays a cleft hand procedure. This video is available in the "Related Videos" section of the full-text article on PRS-Journal.com or at ***http://links.lww.com/PRS/B366***.) The cleft skin is raised as a palmar-based flap and transposed into the first web space. The initial viability of the palmar flap depends on careful preservation of its intrinsic blood supply by means of the subdermal plexus. On occasion, a digital artery is found that can be included with the flap to transform its blood supply from random to axial.

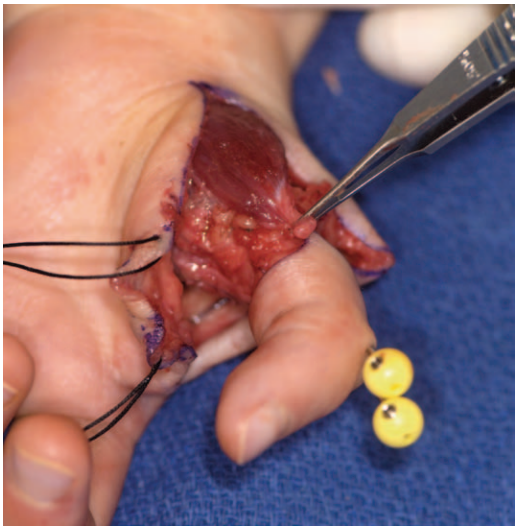

Fig. 27. Transfer of the first dorsal interosseous muscle for abductor function. (Courtesy of Shriners Hospitals for Children, Philadelphia.)

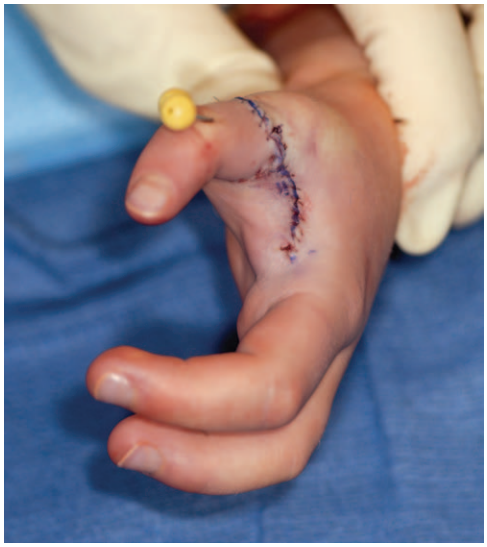

Fig. 28. Skin is carefully inset to avoid a suture line within the first web space. (Courtesy of Shriners Hospitals for Children, Philadelphia.)

The thumb-index syndactyly is separated using established techniques with a longitudinal incision beginning at the level of the thumb-index web space. To help narrow the cleft, the index

Fig. 29. A mobile index finger transferred to the thumb position is readily incorporated into fine pinch. (Courtesy of Shriners Hospitals for Children, Philadelphia.)

Video 2. Supplemental Digital Content 2, which displays a cleft hand procedure, is available in the "Related Videos" section of the full-text article on PRSJournal.com or at ***http://links.lww.com/PRS/B366***.

Table 4. Thumb-Index Classification of Central Deficiency

Type	Web Description	Characteristics
I	Normal	
IIA	Mild narrowing	Thumb web mild narrowing
IIB	Severe narrowing	Thumb web severely narrowing
III	Syndactyly	Thumb and index rays are syndactylized
IV	Merged	Index ray is suppressed, thumb web space is merged with the cleft
V	Absent	Thumb elements suppressed, ulnar rays remain, and thumb web space not present

finger can be transposed to the base of the third metacarpal. In cases without a third metacarpal, an osteotomy is performed at the base of the index to narrow the cleft. Fixation is usually with Kirschner wires or a small plate. Regardless of the technique, the surgeon must avoid digital scissoring secondary to rotation.

Once the cleft is narrowed, the palmar-based flap from the cleft is used to resurface the thumb-index web space. Any syndactyly flaps are also sewn into position. Any bare areas can be covered with full-thickness skin grafts.

CONCLUSIONS

Syndactyly, polydactyly, thumb hypoplasia, and cleft hand are congenital conditions amenable to surgical management. Each diagnosis has particular challenges with respect to preoperative assessment and operative indications. The surgeon must adhere to established principles regarding function and form to enhance hand function. The surgical techniques require meticulous detail to obtain the best outcome. Complications must be readily recognized and handled appropriately to prevent a degradation in hand function.

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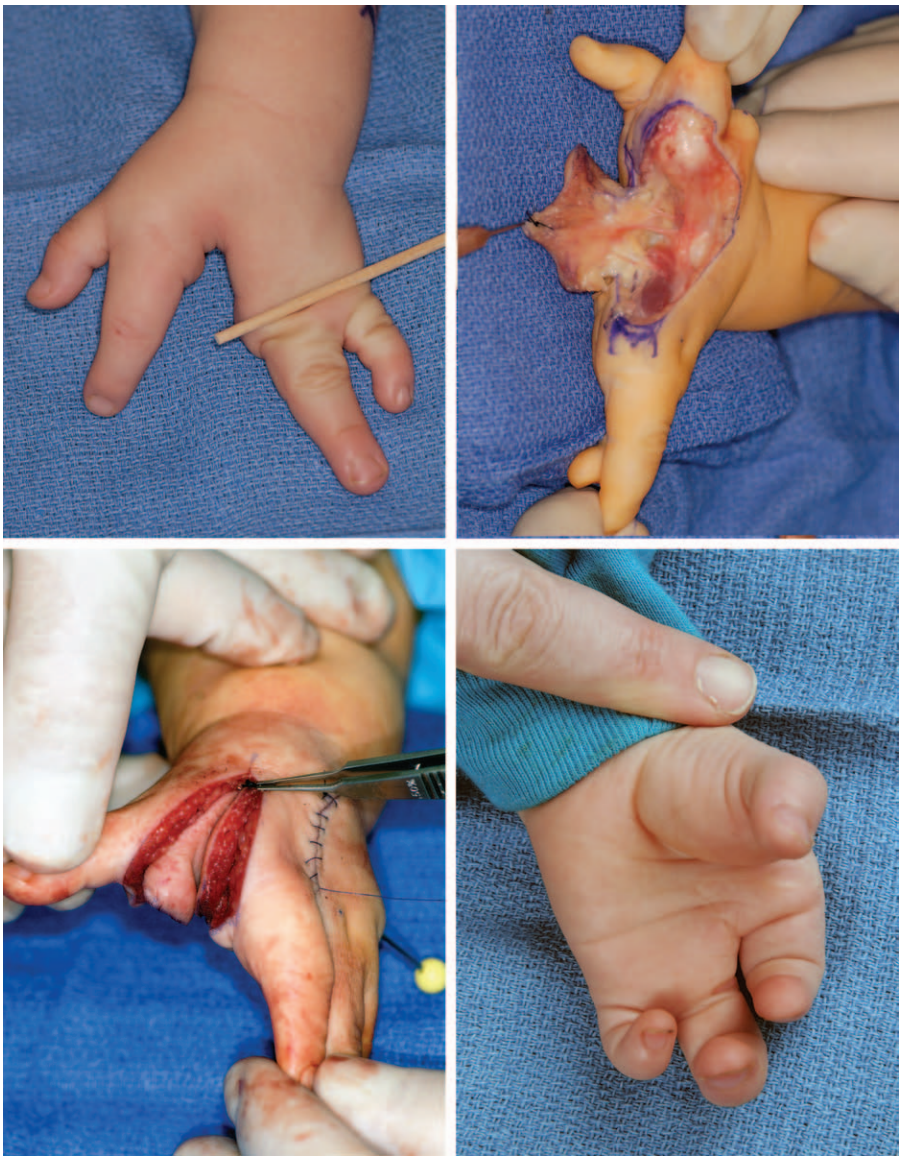

Fig. 30. A 16-month-old boy cleft hand and narrowed thumb-index web space. (*Above*, *left*) Preoperative dorsal view. (*Above*, *right*) Elevation of flap from cleft. (*Below*, *left*) Postoperative dorsal view. (*Below*, *right*) Postoperative view with good thumb-index web space and cleft closure activity. (Courtesy of Shriners Hospital for Children, Philadelphia.)

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