Congenital Hand Differences

40

Angelo B. Lipira, Joel S. Solomon,
Jourdan A. Carboy, and Juliana E. Hansen

Introduction

Congenital hand differences arise embryologically between the 4th and 8th weeks of gestation, with an incidence of 0.1–0.2% of births [1]. Disruption of signaling centers may be responsible for a wide spectrum of disorders which can present with varying degrees of severity (Table 40.1). This chapter will discuss terminology (Table 40.2) and general tips for dealing with the pediatric surgical patient. The most common of the congenital hand disorders and pediatric hand conditions requiring surgical referral will be reviewed, emphasizing tips and tricks for diagnosis and treatment.

General Evaluation of Pediatric Hand Patients

Hand examination for very young patients is challenging and requires some creativity. A good way to begin the examination is having the child seated on a parent's or caregiver's lap, playing with a toy. Observation of hand function is quite revealing, particularly with regard to thumb function and active motion of joints. Look for exclusions or substitutions of digits. Save direct "hands-on" examination for last, as the child may become frightened and uncooperative. Serial examinations over time are very helpful in determining existing function. These multiple visits are also helpful in preparing parents for their child's surgery.

A full history and exam are critical to investigate for associated abnormalities. This includes a thorough family history for relatives with similar hand differences or other congenital differences of any type. Consider a consultation to a geneticist if there is concern for possible syndromic features, or if the parents desire further genetic evaluation for heritability.

Counseling Parents

As the hand surgeon, you will likely be the first person parents have met with who has any true expertise regarding their child's condition. Parents may be quite anxious for their visit, as they want to know the impact their child's condition is going to have on his or her life. Conversely, in the age of the Internet, it is also common for the parents to have read a great deal of information (and misinformation) regarding their child's condition, as well as conditions not relevant to their child. It is the job of the hand surgeon to

A. B. Lipira · J. S. Solomon · J. A. Carboy
J. E. Hansen (✉)
Division of Plastic and Reconstructive Surgery,
Oregon Health and Science University,
Portland, OR, USA
e-mail: hansenju@ohsu.edu

S. R. Thaller, Z. J. Panthaki (eds.), *Tips and Tricks in Plastic Surgery*,
https://doi.org/10.1007/978-3-030-78028-9_40

Table 40.1 The International Federation for Societies for Surgery of the Hand (IFSSH) Classification [2]

I. Failure of part formation
A. Transverse deficiencies
B. Longitudinal deficiencies
1. Phocomelia
2. Radial (radial club hand)
3. Central (cleft hand)
4. Ulnar (ulnar club hand)
II. Failure of part differentiation
A. Synostosis
B. Radial head dislocation
C. Symphalangism
D. Syndactyly
E. Contracture
1. Soft tissue
(a) Arthrogryposis
(b) Pterygium
(c) Pediatric trigger thumb
(d) Absent extensor tendons
(e) Hypoplastic thumb
(f) Clasped thumb
(g) Retroflexible thumb
(h) Camptodactyly
(i) Windblown hand
2. Skeletal
(a) Clinodactyly
(b) Kirner deformity
(c) Delta phalanx
III. Duplication
A. Thumb
B. Triphalangism/hyperphalangism
C. Polydactyly
D. Mirror hand
IV. Overgrowth
A. Limb
B. Macrodactyly
V. Undergrowth (e.g., Brachydactyly, brachysyndactyly)
VI. Congenital constriction band syndrome
VII. Generalized skeletal abnormalities

The IFSSH classification is perhaps the most widely used schema of congenital hand differences. It attempts to organize diagnoses by embryological etiology

Table 40.2 Definitions of terms

Term	Definition
Syn-	Fused transversely
Sym-	Fused longitudinally
Clino-	Deviated in the coronal plane
Campto-	Flexed in the sagittal plane
Acro-	Peripheral, distal
Dactylos	Finger
-melos	Limb

provide them with clear and accurate information at the first visit, and it is important to discuss the expected outcome and function of their child as an adult. For the majority of congenital hand conditions, the child's expected *functional* outcome will be excellent. However, it is just as important to discuss with them what cannot be corrected and thus to identify future challenges for the child. This includes aesthetic as well as functional challenges.

The vast majority of congenital limb differences are the result of either spontaneous or inherited mutation. Environmental factors such as teratogens almost never play a role. This is important to understand when meeting parents of children with limb differences, as they often bear a great deal of unwarranted guilt. The surgeon treating congenital limb differences has the primary responsibility for educating parents, as well as following the child throughout development, often until they reach skeletal maturity and beyond. It is also the surgeon's responsibility to ensure that the patient has had appropriate evaluation for associated conditions (renal, cardiac, and neurologic) when warranted.

Finally, it is always important to consider the viewpoint of the parents. A hand surgeon may think their outcome looks good, while parents and others may be more likely to focus on the differences that remain. Instead of saying "it looks really good," say "this is how it is supposed to look." Showing parents postoperative pictures ahead of time is also very valuable.

General Timing Considerations

In most cases, surgery is delayed until around 12 months of age, when structures are larger and easier to manipulate. Also, general anesthesia may become safer as the child ages, due to maturation of the airway. Earlier intervention should be considered, however, if the condition affects limb development or interferes with developmental function. For example, border

digit syndactyly may interfere with growth due to length discrepancy of digits (see below for further discussion). An additional consideration regarding timing is that children start to experience peer pressure at around 4 years of age. Correction should ideally be planned before this time to facilitate social development and prevent children from being stigmatized.

Immobilization

As with hand surgery in adults, immobilization should only be used when necessary. Trigger finger releases and type B postaxial polydactyly excisions require no immobilization. For more complex procedures, some immobilization is often indicated. Recognize, however, that the active forces that can be generated by an infant or toddler are small and the "strength" of the needed immobilization is less than what we associate with similar procedures on adults. For these younger patients, excellent immobilization can often be achieved with cotton gauze alone. To create a cotton gauze splint, the hand should be surrounded by unfolded Kerlix or cotton fluffs. The cotton gauze is then saturated with saline and the dressing is compressed around the area to be immobilized. The dressing is then completed with additional dry cotton gauze, with or without a cast or splint. When the dressing is removed, one can see that the compressed cotton has been formed into an immobile mass that conforms to the contours of the hand.

Young children are escape artists and will try to get out of any cast or splint placed on them. There are many cases where immobilization or protection of the surgical site is critical for success, for example, syndactyly reconstruction, opponensplasty, or pollicization. A long-arm cast or splint can be made with fiberglass or plaster. To prevent inadvertent removal of the cast or splint, flex the elbow to 100° during application. Make certain that there is sufficient padding over the ulnar nerve at the medial elbow. An adult might not tolerate this position, but children tend to do fine. Cast material extends to the tips of the fingers, completely covering them. Great care must be taken when applying the cast to avoid any excessive pressure, which may result in pressure ulceration. Parents are encouraged to cover the dressing with a tube sock that can easily be exchanged whenever it gets dirty.

Specific Conditions

Syndactyly

Background

Syndactyly is fusion of two or more digits to each other. Syndactyly is one of the most common congenital hand conditions, with an estimated incidence between 1:1000 and 1:3000 live births [3]. Approximately half of cases are bilateral, and syndactyly is more common in males (2:1). Most are spontaneous, isolated mutations, but sometimes autosomal dominant inheritance with variable penetrance is seen (10–40% hereditary). Complex syndactylies are more often part of a syndrome such as Apert, Carpenter, or Saethre-Chotzen syndrome.

Syndactyly encompasses a spectrum of presentations and severities (Fig. 40.1). "Simple" syndactyly means that only the skin and soft tissues are fused, versus "complex" syndactyly in which fusion of bony elements is involved. "Complete" syndactyly means that the fingers are joined all the way to the fingertips, versus "incomplete" where the fusion does not extend to the fingertips. "Acrosyndactyly" refers to the situation where only the fingertips are fused, and the more proximal elements are not fused. Acrosyndactyly is classically a feature of Apert's syndrome.

Evaluation

Thorough past medical, family, and birth history should be obtained. Hand examination should determine what digits are involved and the extent of involvement. Look for fusion of the nails (synonychia), which is indicative of underlying bony fusion (complex syndactyly). When border digits are involved, there may be resulting deviation of the longer digit toward the shorter one (clinodactyly), as well as proximal interphalan-

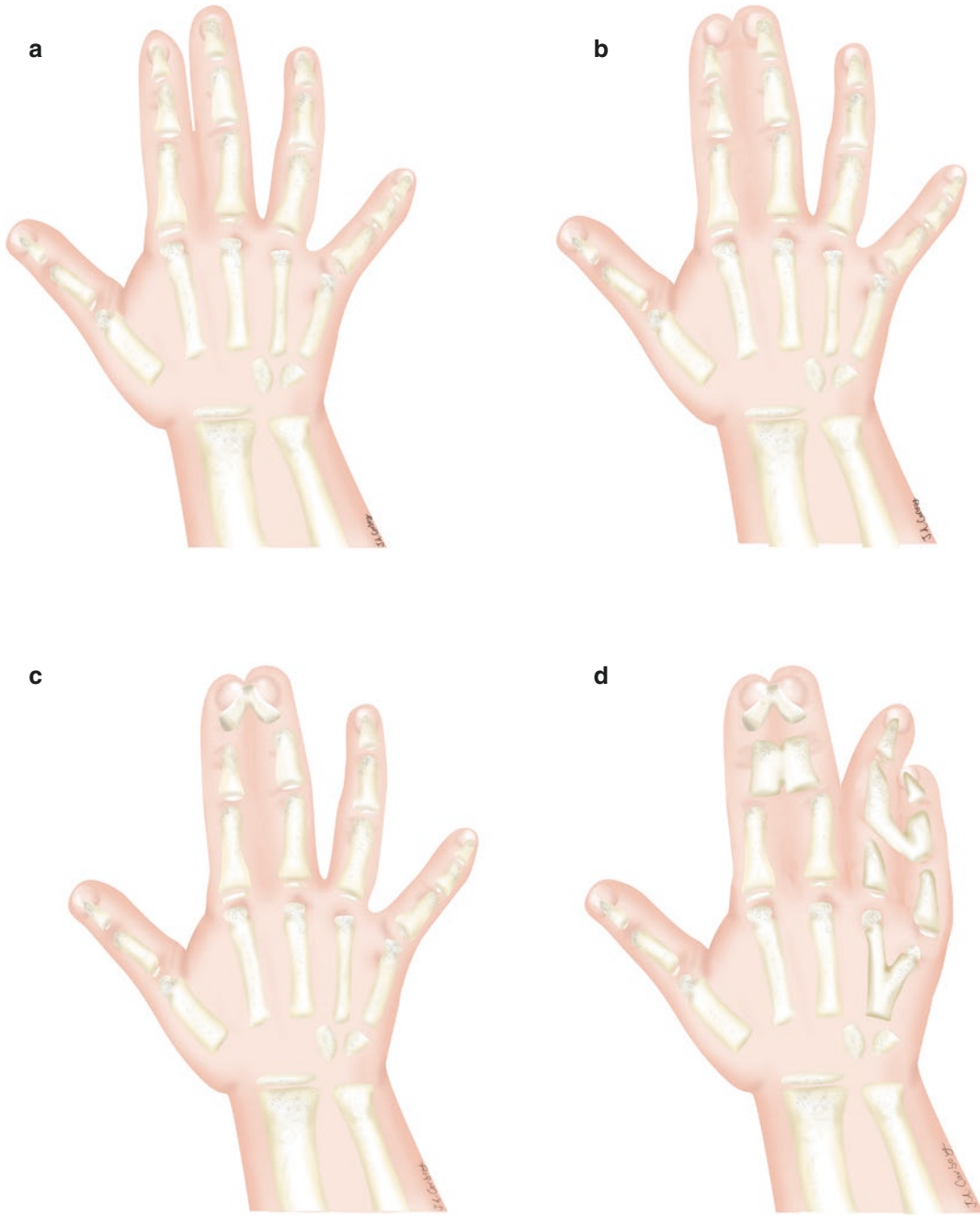

Fig. 40.1 Classification of syndactyly. (**a**) Simple incomplete. (**b**) Simple complete. (**c**) Complex. (**d**) Complicated. (Illustration by Jourdan A. Carboy)

geal (PIP) flexion contracture, which can be progressive with growth and is difficult to treat. Exam should include the entire involved and contralateral upper extremities, the chest wall (specifically looking for absence of pectoralis musculature and abnormalities of the bony thorax indicative of Poland syndrome), and the feet.

Plain radiographs of the hands should be obtained to evaluate for bony involvement.

Associated syndromes include Aperts, Crouzon, Pfeiffer, Saethre-Chotzen, and Poland syndrome. Syndromic syndactyly is more often complex and complicated, frequently with multi-digital involvement.

Treatment

Surgery is typically performed between 1 and 2 years of age, or earlier if border digits are involved or if there is other potential for progressive deformity of involved digits. If multiple web spaces are involved, do not separate two adjacent web spaces at the same time, as there is greater risk of ischemia of the central digit. The goals of syndactyly reconstruction can be broken down into (1) separation of the digits, (2) creation of a web space, and (3) soft tissue coverage.

Many different incision designs are described. The most important common feature is a dorsally based flap to create a web space. Avoid using skin grafts in the web space, as recurrence (i.e., "web creep") will be likely.

Though techniques have been described that avoid skin grafting altogether, these are demanding and rely on the mobilization of a large amount of dorsal hand skin. The authors routinely use full-thickness skin grafts, harvested from the volar wrist crease, volar elbow crease, or groin, depending on surgeon preference. All of these locations leave well-concealed donor sites.

- Always use full-thickness grafts, as split-thickness grafts undergo a greater degree of secondary contracture, provide less durable coverage, and have greater donor site morbidity.
- Separation of the digits, after elevation of flaps, involves dividing fascial interconnections between the two digits. To accomplish this safely, the neurovascular bundles of each finger must be identified and protected.

The authors' incision pattern for complete or incomplete syndactyly involves a dorsal rectangular flap for web space reconstruction and triangular flaps designed in a mirror-image fashion on the volar and dorsal surfaces of the fused digits (Fig. 40.2).The dorsal rectangular commissure flap should extend approximately 2/3 of the distance to the PIP joint. The distal edge of the flap extends to the dorsal midline of each digit. The lateral margins may be designed concave to match the curvature of the digits and decrease the need for skin grafts in this area. Draw the dorsal commissure flap first. Next draw a transverse line volarly. Keep in mind that the normal web space has a slope of approximately 45 degrees. The location of the volar extent of the web space can be designed by projecting its position from the adjacent uninvolved web spaces and is roughly half the distance between the palm crease and the PIP flexion crease.

The triangular flaps should also extend to the mid-point of each digit (although some advocate 1/3, saying the scars are better hidden). The size of the flaps can be varied, but in general we create one or two flaps per phalanx. Draw the dorsal triangular flaps before volar. After the pattern has been drawn dorsally, make marks on the lateral borders of the digits corresponding to the points of the triangles, as this will facilitate creating mirror-image flaps volarly and ensure the flaps interdigitate appropriately (Fig. 40.3). Every triangle tip on the dorsum should correspond the middle of a triangle base on the volar side. Every triangle base midpoint on the dorsum should correspond to a triangle tip on the volar side. When designing the flaps, keep in mind that there will be areas, typically at the base of the digit, which will require skin grafting. Attempt to design flaps so that the grafts will be located over the inner proximal phalanx, where they will be less visible and won't overly joints or involve the web-space.

In the case of synonychia (nail fusion), which is a sign of complex syndactyly, opposing narrow triangular flaps are designed distally as described by Buck-Gramcko (Figs. 40.4 and 40.5) [4]. These flaps serve to recreate the missing lateral nail fold once the digits are separated. Be careful, as these thin flaps are easy to accidentally damage or amputate.

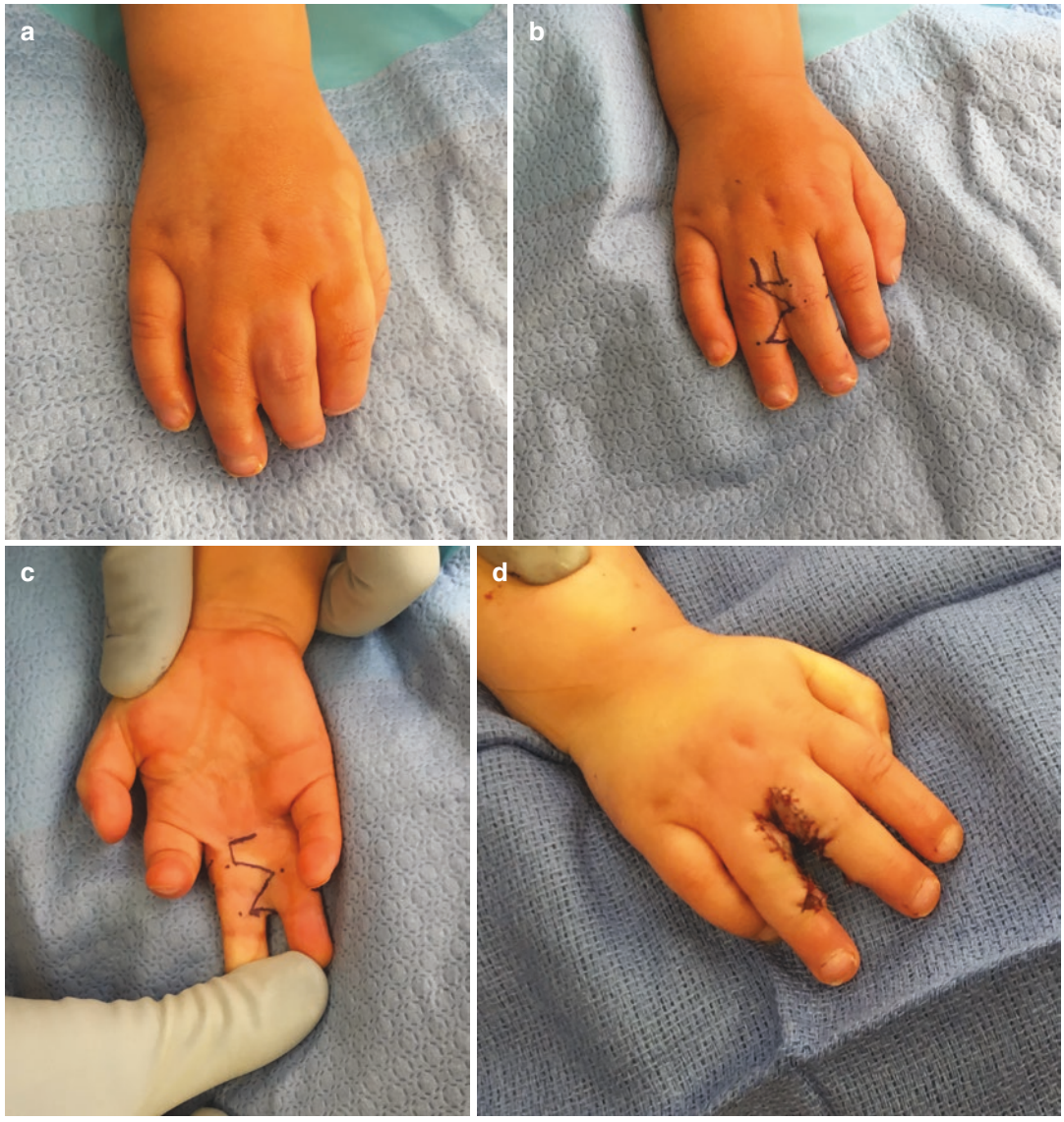

Fig. 40.2 (**a**) Incomplete simple syndactyly of the 3rd web space. (**b**, **c**) Incision markings. (**d**) After flap inset and skin grafting

Surgical Steps (Separation of Simple Syndactyly)

- Start by making all incisions through the dermis only. Then proceed with elevating flaps dorsally and then volarly. Flaps should be elevated to the dorsal midline of the two digits before proceeding. Make all incisions except for the volar proximal transverse incision: this will receive the tip of the dorsal commissure flap, and it is critical to ensure that the dorsal flap will reach before committing to this incision (after near complete separation of the digits, this can be checked and adjusted as needed).
- In simple syndactyly, there is a dense fibrous transverse band between the two digits. After elevating the flaps, this band is exposed dorsally and volarly through gentle scissor dissection, then divided in a distal-to-proximal fashion while steady retraction (separation

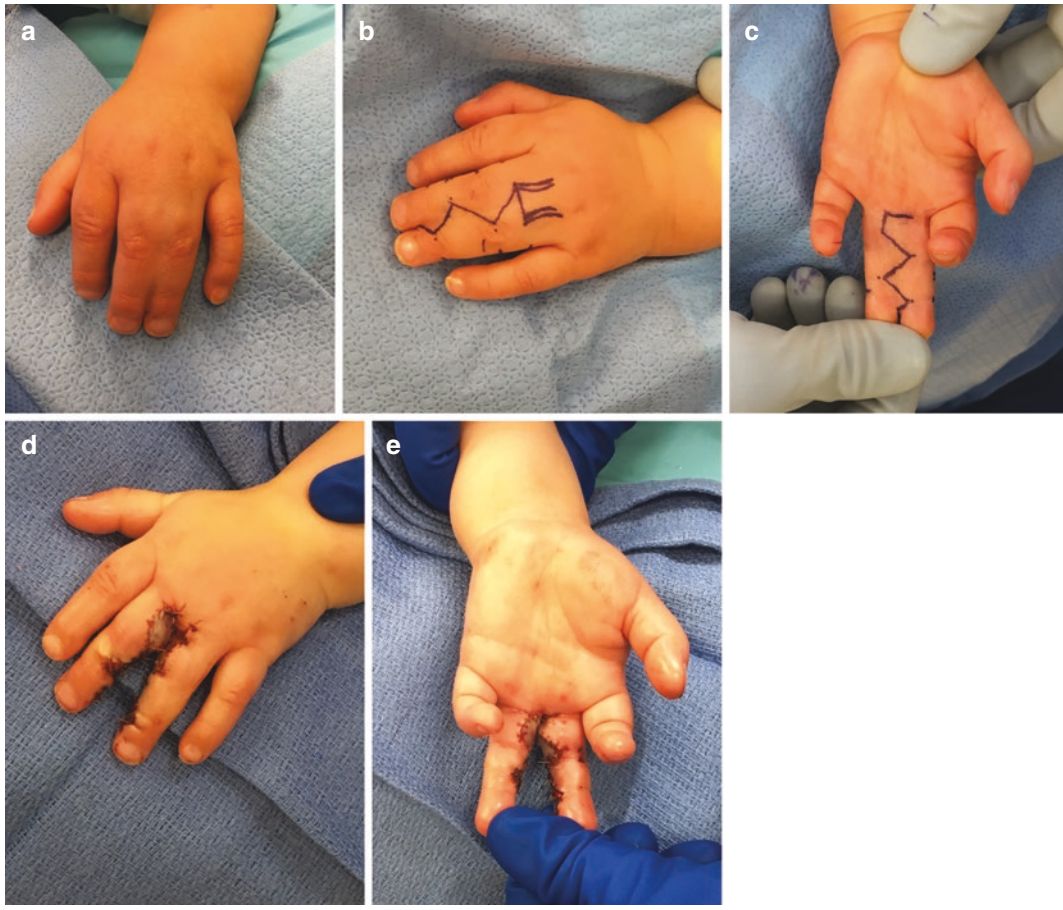

Fig. 40.3 (**a**) Complete simple syndactyly of the 3rd web space. (**b**, **c**) Incision markings. (**d**, **e**) After flap inset and skin grafting

pressure) is applied to the two digits. The surgeon should sit at the end of the hand table, looking down the digits and watching out for the neurovascular bundles—these typically are located clear of the midline and closer to their respective digits.

- Beware that a distal bifurcation of the common neurovascular trunk may exist. In this situation, the nerves may be neurolyzed to create a more proximal bifurcation, but one artery may need to be sacrificed. In this case, choose the smaller of the two arteries to sacrifice, as this generally implies that the remaining artery on that digit is dominant.
- After near full separation of the digits, check that the dorsal commissure flap will reach the desired volar insertion before making the volar proximal transverse incision. At this point, complete the elevation of the dorsal flap, getting thicker as you head proximally and attempting to preserve any veins going into the flap.
- Inset the dorsal flap first with sutures at the two corners and then midline.
- Inset the tips of the triangular flaps. During flap inset, finger contour can be improved by judicious fat removal from the digits.
- Use foil from a suture packet to make precise templates of the open areas. Mark the templates carefully to maintain their orientation, then trace the templates onto the planned donor site (volar wrist, elbow, or groin) such that an ellipse may be drawn encompassing the needed grafts. Test the ellipse using the

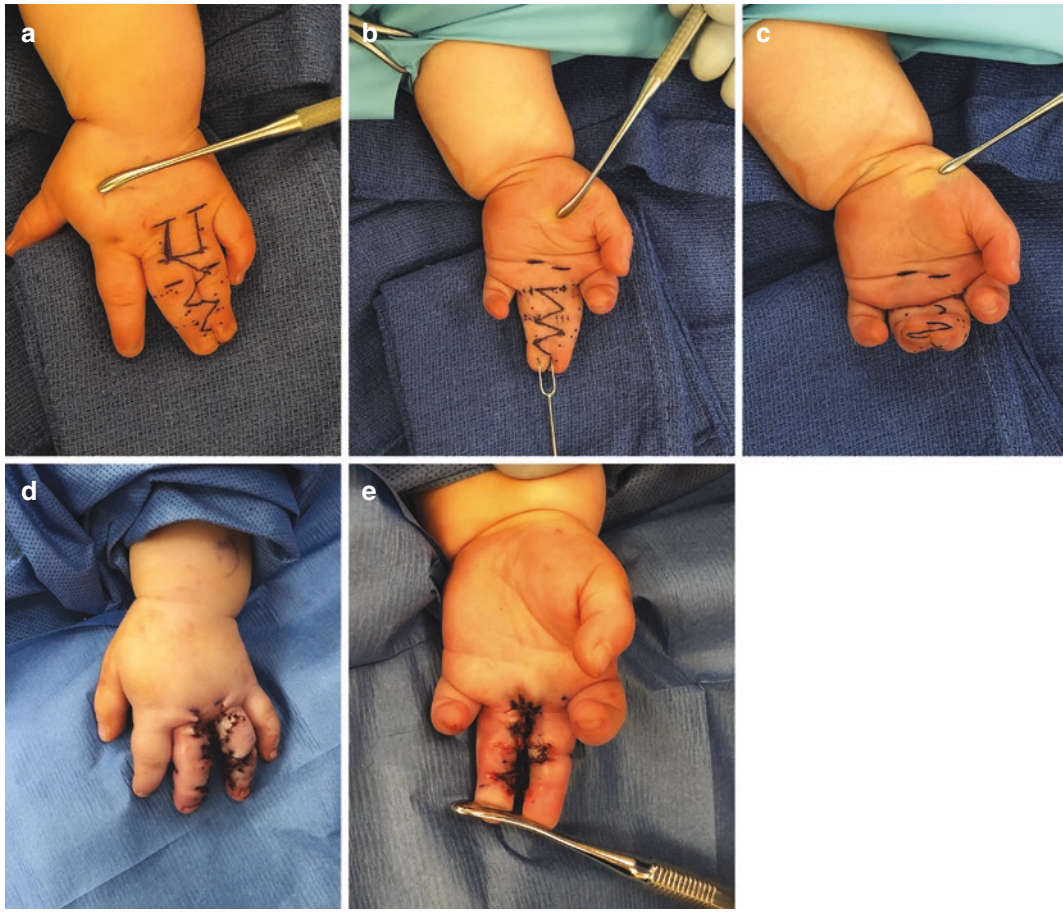

Fig. 40.4 (**a**–**e**) Complex syndactyly with synonychia. Note the markings made laterally on the digits to assist in creating mirror image volar and dorsal flaps. Incisional design incorporates Buck-Gramcko flaps for reconstruction of the nail folds

"pinch test" to ensure it can be closed safely. Using the templates and the pinch test, you can determine whether the wrist, elbow, or groin will supply the required amount of skin. Alternatively, determine the dimensions of each graft needed and excise an ellipse from the groin crease whose width will cover the widest defect and whose length is roughly the combined total length of all needed grafts.

- Inject epinephrine containing local anesthetic subcutaneously under the entire graft donor site for hemostasis/pain control.
- Harvest grafts using a 15-blade scalpel.
- Dressing and cast: xeroform over incisions. 4x4 cut into thirds, dipped in saline and wedged in between the digits to splint open the commissure and bolster the grafts. Over this, webril and long-arm cast or sugartong splint covering the fingers entirely. The elbow is flexed to 100 degrees to prevent cast removal.

Postoperative cast duration varies, but excellent results have been observed in our group with leaving the cast in place for either 1 or 2 weeks. The use of "wedge splints" is attempted by some surgeons postoperatively in hopes of lessening the risk of "web creep," but it can be difficult for parents and patients to comply.

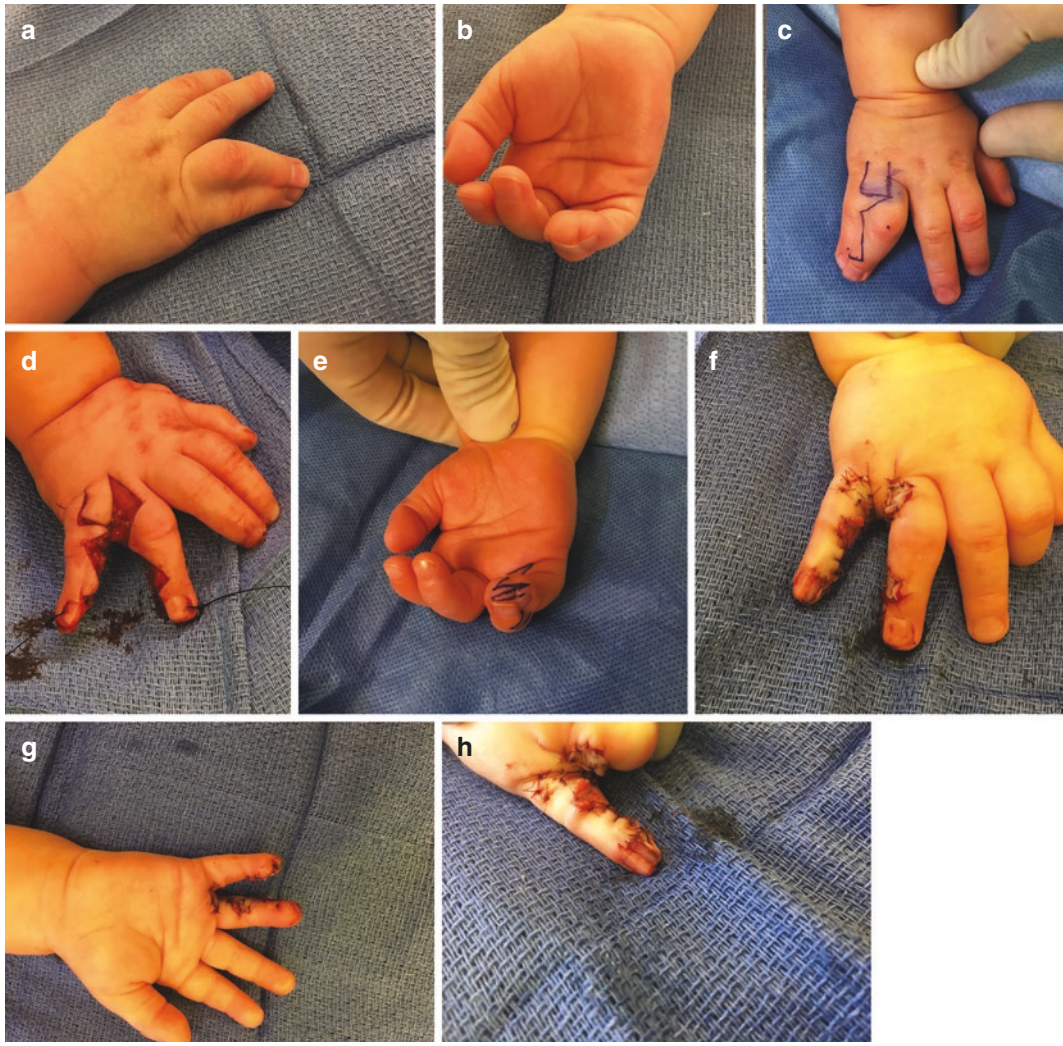

Fig. 40.5 (**a**–**h**) Complex syndactyly of the 4th web space. Note the tethering effect of the small on the ring finger. Buck-Gramcko flaps used for reconstruction of nail folds

Special Situations

Complex Syndactyly

- Defined by the presence of bony fusion.
- In addition to the steps outlined above, bony separation is required. This is done after triangular flaps have been elevated and dissection is proceeding from distal to proximal. A scalpel is usually sufficient to accomplish division of the bone. In complex syndactyly, there is often a more significant skin shortage, requiring more skin grafts than in simple syndactyly.

Minor Syndactyly, Shallow Web Spaces, and Web Creep

Patients may present with simple incomplete syndactyly that does not extend to the level of the PIP joints of the involved digits. This more minor syndactyly may be congenital or can occur following a previous syndactyly release (i.e., web space creep). In some congenital cases, there is a true excess of skin between the involved digits. Nevertheless, surgical techniques to deepen the web space by a dorsal rectangular flap or other means typically cannot make use of this skin and

often require the placement of additional skin graft (see Fig. 40.3). Skin-conserving methods, such as serial z-plasties, rarely provide sufficient deepening. An excellent technique for these shallow web spaces was published by Shinya [5] and is called the Dancing Girl flap. The original mathematical description can be difficult to interpret and implement. A simplified step-by-step method for constructing Dancing Girl flaps is illustrated in Fig. 40.6, and described below.

Designing the "Dancing Girl" Flap for Minor Syndactyly

Design begins on the volar side:

1. Draw volar web space line (a) along the distal border of the existing web
2. Draw line (b) extending proximally from one end of line (a)
   (a) Length of (b) = 0.7 × a
   (b) Angle ab = 120°
3. Draw line (c) proximally from the opposite end of line (a)
   (a) Length of (c) = 0.6 × (a)
   (b) Angle ac = 70°
4. Draw a guide line perpendicular to most proximal point of line (b)

   This guide line will mark the depth of the new web space.
5. Draw line (d) proximally from the intersection of lines (a) and (b)
   (a) Angle bd = 45°
   (b) Length of (d): to intersection with perpendicular from (b)
6. Repeat the pattern on the dorsum of the finger starting with a'
   (a) Length (a') = (a)
   (b) Parallel to (a)
   (c) Distance from (a) = 0.6 × (a)
7. Line (b') goes along opposite side of web to line (b)
8. Line (c') goes along opposite side of web to line (c)
9. Draw an oblique line (e) connecting lines (a) and (a')
   (a) Mark a point along (a) and (a') 1/3rd the length of (a) from the intersection of (a) and (b)
   (b) Connect these two points

Following flap transposition, the new web space is created by flaps (bd) and (b'd'). If the flap is employed for the treatment of web space creep, some effort can be made to align line (b) or line (d) on a preexisting scar; however, the presence of other scar lines does not preclude the use of this flap.

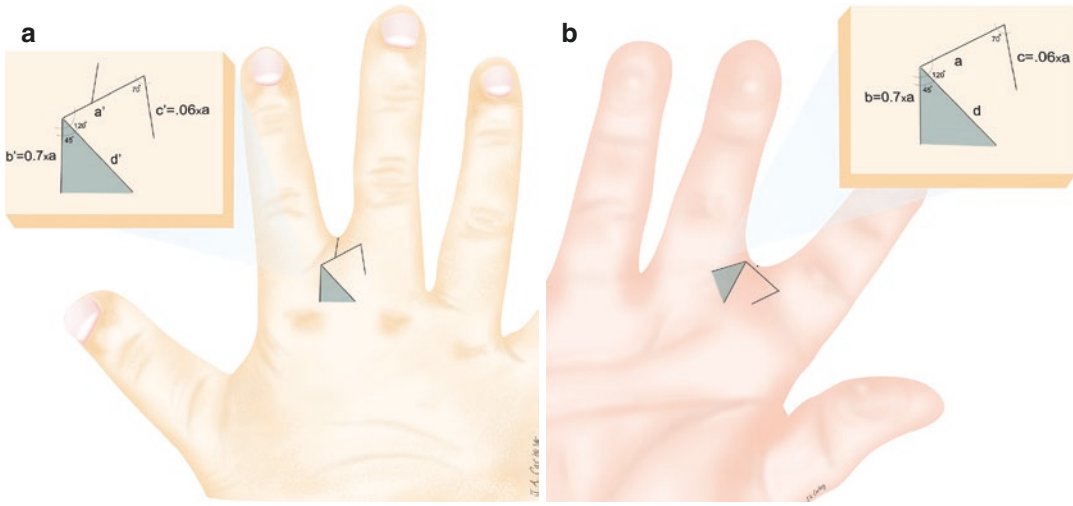

Fig. 40.6 (**a**, **b**) Flap design for the dancing girl flap. The shaded triangles represent the skin flaps that will form the new web space. The depth of the new web space is the proximal base of these two triangles. See text for detailed description. (Illustrations by Jourdan A. Carboy)

Multiple Digit Syndactyly

Multiple digit syndactyly most commonly arises in the syndromic setting, classically in Apert's syndrome. These tend to be complex and often complicated, with highly abnormal formation and sometimes absent bony elements. Achieving five separate digits is not always possible or advisable, and realistic expectations should be set early with parents. Joints are often abnormally formed and may exhibit little or no motion, which should be anticipated and explained to parents. The primary goal should be improvement in function of the hand.

When more than two digits are involved in a single fusion mass, separations should be staged to lessen the risk of devascularizing the central digit. It is recommended that surgeries be spaced 4–6 months apart.

Pediatric Trigger Thumb

Background

Pediatric trigger thumb, or stenosing tenosynovitis of flexor pollicis longus, has an incidence of around 3 in 1000 children [6]. Average age at presentation is 2 years, and approximately 25% are bilateral [7]. Trigger thumb most commonly presents as an inability to extend the thumb IP joint, with the child holding it in a position of fixed flexion. Unlike trigger digit in adults, pediatric trigger thumb is not an inflammatory process and usually not painful [8]. Although the term "congenital trigger thumb" is still widely used, multiple studies have documented that this condition is not present at birth and is, in fact, an acquired condition [9].

Evaluation

The affected thumb often displays a lack of full extension. A prominent nodule (Notta's node) is palpable over the volar metacarpophalangeal joint, representing a nodule in the FPL tendon as well as associated thickening of the A1 pulley (Fig. 40.7).

These children are often erroneously referred for a suspected trauma, with parents and pediatrician suspecting a dislocation of the IP joint despite no witnessed trauma. Trigger thumb must also be differentiated from congenital clasped thumb, which is usually due to a deficient extensor mechanism and has very different treatment considerations. Radiographs are not indicated, as the diagnosis can be confirmed through the history and physical examination.

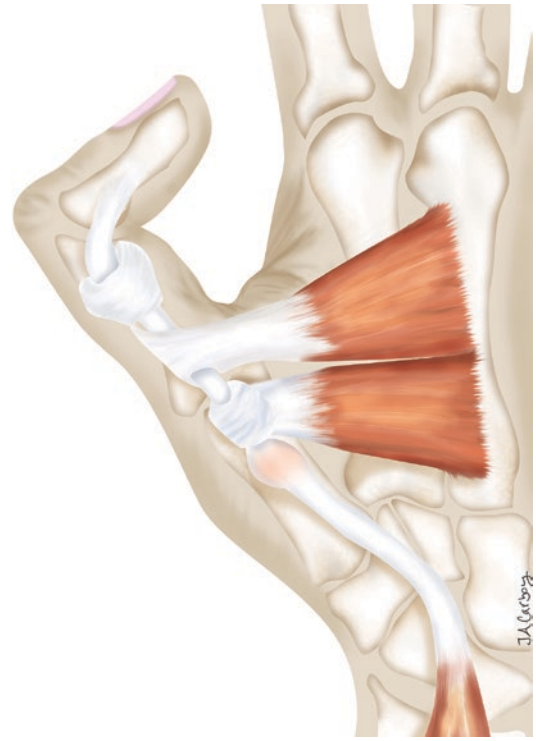

Fig. 40.7 Illustration of Trigger thumb pathoanatomy. Note nodular enlargement of flexor tendon obstructing passage through A1 pulley. The interphalangeal joint is held in a flexed position. Note the oblique pulley distal to A1 upon which adductor pollicis inserts. Division of the oblique pulley must be avoided. (Illustration by Jourdan A. Carboy)

Treatment

Surgery should be considered if trigger thumb is present for greater than 4 months, or if the child is 3 years of age or older. We wait until the child is at least 12 months old before surgery. About 1/3 will resolve spontaneously by 12 months of age.

Surgical Steps

- The surgery is performed under general anesthesia, positioned supine with the hand on the patient's chest or abdomen. The surgeon and assistant stand on opposite sides of the patient, with the surgeon on the contralateral side.
- A tourniquet is placed on the upper arm and set to 200 mmHg.
- Draping consists of a sterile towel with a clamp just below the tourniquet and a 3/4 sheet with a small hole cut in it.
- The assistant's critical job is to hold the thumb abducted and extended to create necessary tension on the tissues, while retracting for the surgeon.
- The incision is transverse and placed in the volar MP crease, directly over the palpable Notta's node. The incision is approximately 1 cm in length (Fig. 40.8).
- A 15-blade scalpel is held upside down so that the tip may be precisely pushed to make the planned incision through skin only. The digital nerves can be extremely close to the skin in children this age.
- After incising through the dermis, gentle longitudinal spreading over the mid-line of the pulley will provide safe exposure while avoiding harm to the neurovascular bundles. Attempt to identify any nerves in the surgical field, but do not search for them if not contained within the field.
- Once the pulley is adequately exposed, a Beaver or Weck blade is used to make a longitudinal "nick" in the center of the pulley through use of a pressing motion. The surgeon will feel the blade "pop" through the pulley without injuring the underlying tendon.
- An iris or similar small scissor is used to complete the pulley release proximally to the leading edge, and distally while preserving the distal oblique pulley; see Fig. 40.7).
- Do not slide the scissors. Rather, see the tips and only cut what can be directly visualized to avoid risk of dividing the oblique pulley.
- When adequately released, the two edges of the divided pulley should open like pages of a book, as opposed to appearing as a funnel when incompletely released.

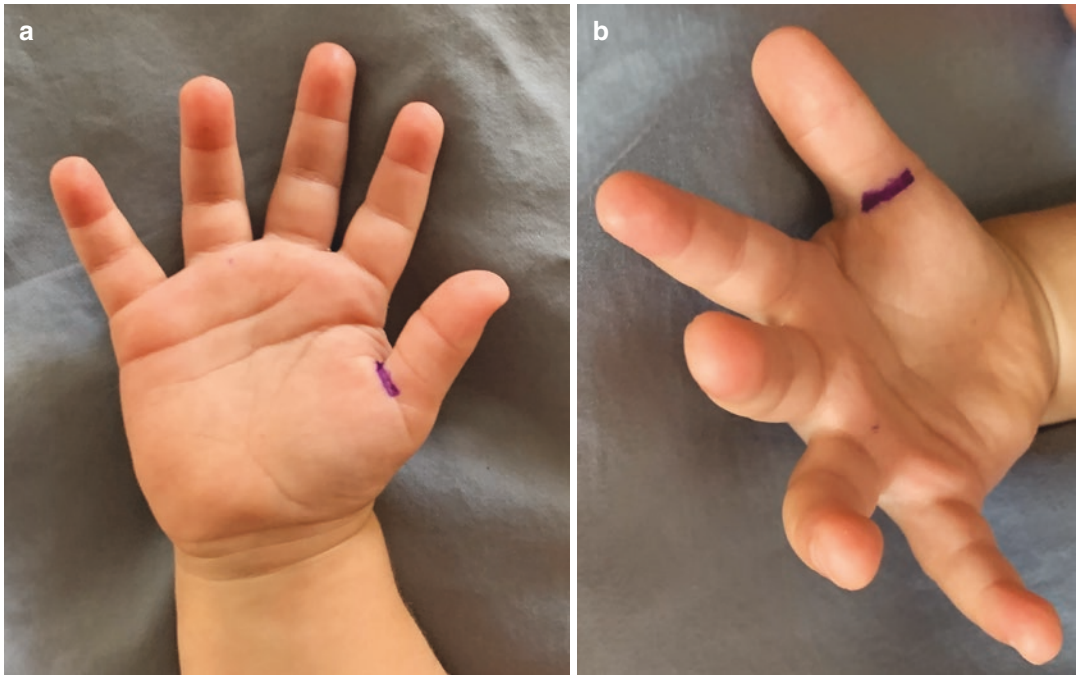

Fig. 40.8 (**a**, **b**) The incision for trigger thumb, placed in the volar joint crease of the metacarpophalangeal joint

- After release, the skin is closed with 5-0 plain gut suture.
- 0.25% bupivacaine is used to block the radial sensory nerve and median nerve at the wrist.
- Dressing is dermabond and a band-aid.
- Tylenol and ibuprofen are sufficient for pain control.

Polydactylies

The presence of extra digits at birth is the most common congenital hand difference. Digital duplications are commonly broken down into three groups: (1) postaxial polydactyly, involving a supernumerary ulnar digit, is the most common; (2) preaxial polydactyly is also referred to as thumb duplication or split thumb; and (3) central digit polydactyly, which is rare and will not be discussed further. Preaxial polydactyly and postaxial polydactyly tend to have different associated mutations and syndromes, different inheritance, and different treatments (Table 40.3 and Fig. 40.9).

Evaluation

Postaxial polydactyly commonly presents as a hypoplastic finger "nubbin" attached to the dominant small finger by a small soft tissue stalk, known as Temtamy and McKusick type B (Fig. 40.10) [10]. But it can also present with significantly formed duplicated small finger ray (Temtamy and McKusick type A; Fig. 40.11). An X-ray of the hand should be obtained prior to surgical revision of type A polydactyly, but is unnecessary in type B. While a referral to Genetics could be considered for any child with a congenital difference in order to rule out associated syndromes, this is particularly true for Caucasians with postaxial polydactyly, as the risk may be as high as 10%.

Preaxial Polydactyly

Preaxial polydactyly is now commonly referred to as "split thumb deformity" owing to the recognition that the thumb that will remain following surgery is always hypoplastic when compared to the opposite unaffected thumb. Treatment is predicated upon (1) the skeletal deformity as classified by the Wassel classification (see Fig. 40.9) [11], (2) the relative sizes of the two thumbs, (3) the motion of the two thumbs, and (4) the expected stability of the postoperative construct. X-rays should be obtained close to the planned surgery date. Wassel type 7 (triphalangeal thumb) is often associated with other abnormalities. Patients with Wassel 7 polydactyly should be referred for a full genetics evaluation.

Treatment

Postaxial Polydactyly

Patients with type B postaxial polydactyly typically present with a hypoplastic digit attached to the ulnar aspect of the small finger proximal phalanx by a tenuous skin bridge. Treatment is encouraged at the time of presentation, as a controlled removal in a clinic setting seems preferable to the predictable traumatic avulsion of the digit during the child's regular activities. Nurses in the newborn nursery commonly ligate the base of the accessory digit with a 4-0 silk suture. The digit undergoes ischemic necrosis and even-

Table 40.3 Polydactyly

Polydactyly type	Digit affected	Dominant digit	Accessory digit	Inheritance	Classification	Associated Syndromes
Preaxial	Thumb	Hypoplastic	Usually radial thumb	Usually sporadic mutation	Wassel 1–7	With Wassel Type 7
Postaxial	Small finger	Normal	Uniformly ulnar small finger	Usually autosomal dominant	Temtamy and Mckusick A or B	Rare, but more common in Caucasians

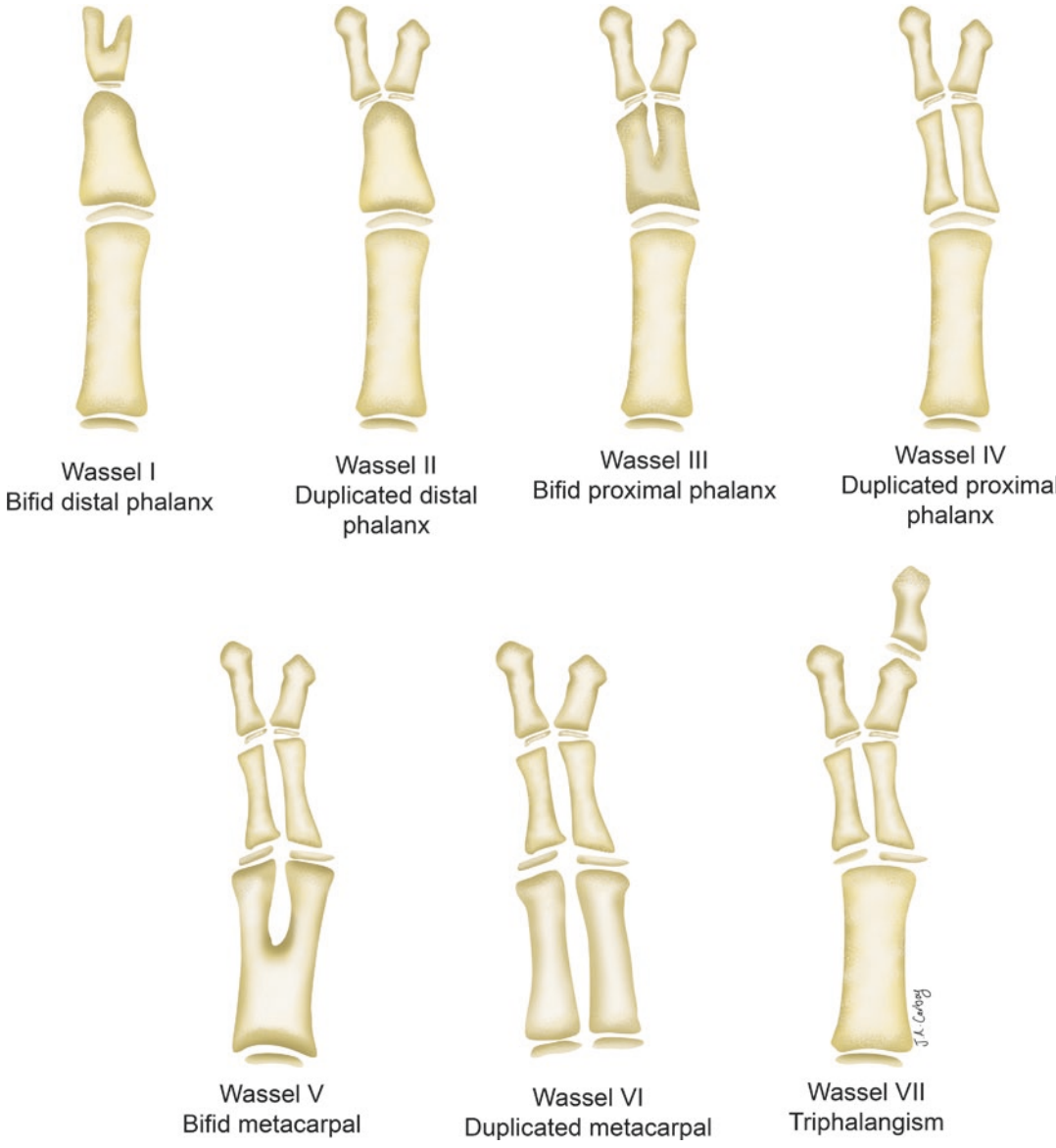

Fig. 40.9 Wassel classification of split thumb deformity (preaxial polydactyly). (Illustration by Jourdan A. Carboy)

tually autoamputates. This technique has some disadvantages:

1. Inadequate ligation may lead to tissue ischemia and pain that requires further treatment.
2. The accessory digit turns black but takes weeks to autoamputate.
3. Nearly half of patients will have residual contour deformity.
4. Many will later complain of a painful neuroma that requires future excision

A preferable alternative is to apply topical anesthetic and excise the accessory digit with a sterile scissors. There will be a genuine neurovascular bundle that can be treated with local pressure and a steri-strip, but our preference is to use a silver nitrate stick or bipolar electrocau-

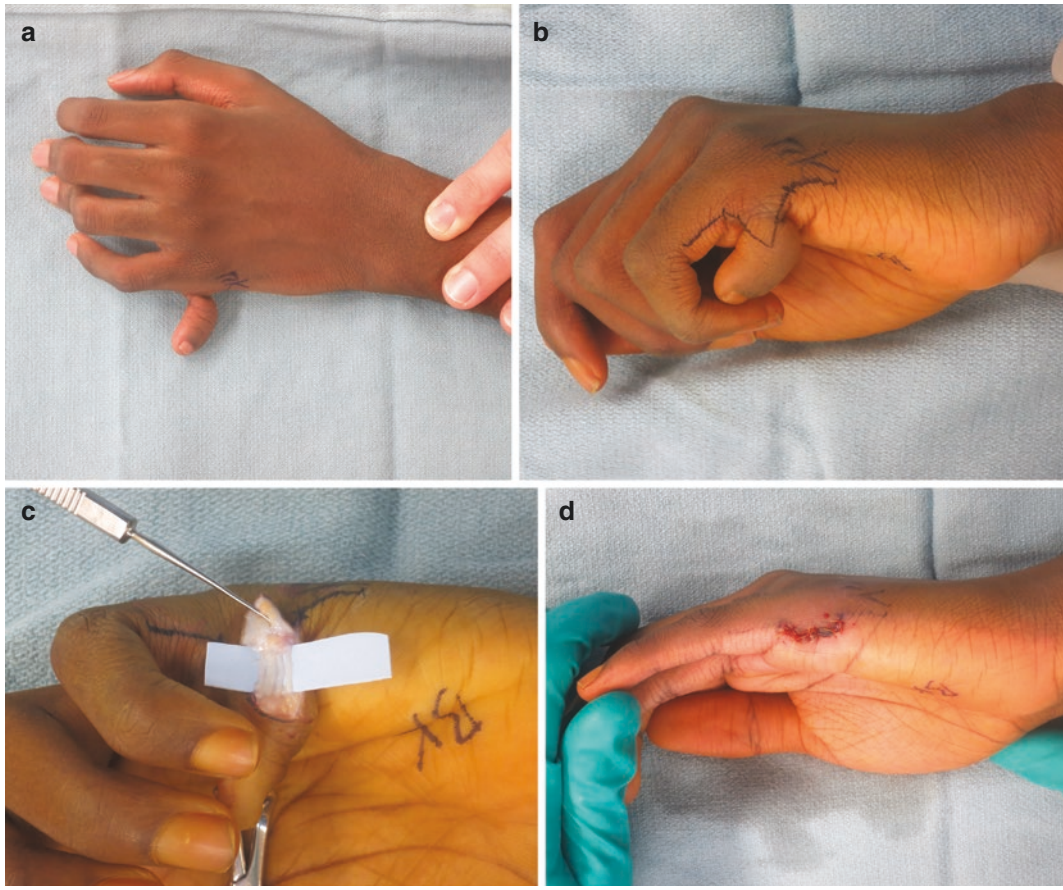

Fig. 40.10 Postaxial polydactyly, Temtamy, and McKusick type B. (**a**) The illustrated case is unusual, involving a previously untreated 15-year-old male. (**b**) Incision design incorporating flap for closure (**c**) Neurovascular structures are present in the digit. Children can develop symptomatic neuroma if nerves not adequately addressed. (**d**) Final closure

tery (Fig. 40.10 illustrates the neurovascular bundle).

For patients with type A postaxial polydactyly, surgery is typically performed under general anesthesia through a modified racquet-shaped incision (see Fig. 40.11). Excess skin from the accessory digit should be preserved. This skin can be trimmed at the end of the case to restore a normal contour to the hand, whereas a skin deficit can leave a soft tissue depression. As with the treatment of Wassel type 4 preaxial polydactyly described below, accessory digit excision at the level of the MP joint can lead to joint instability. If the ulnar collateral ligament or intrinsic muscle insertions are violated in the process of accessory digit excision, these should be reconstructed with suture. Abductor digiti minimi is often found to insert into the base of the accessory digit and should be elevated and reinserted into the ulnar aspect of the extensor mechanism of the small finger to be preserved. When ligament or tendon reattachments are required, a long-arm splint or cast is used for 2 weeks.

Preaxial Polydactyly

The treatment of split thumb is largely determined by the skeletal anatomy of the deformity as classified by Wassel (see Fig. 40.9) [11]. Although not always achievable, all reconstruc-

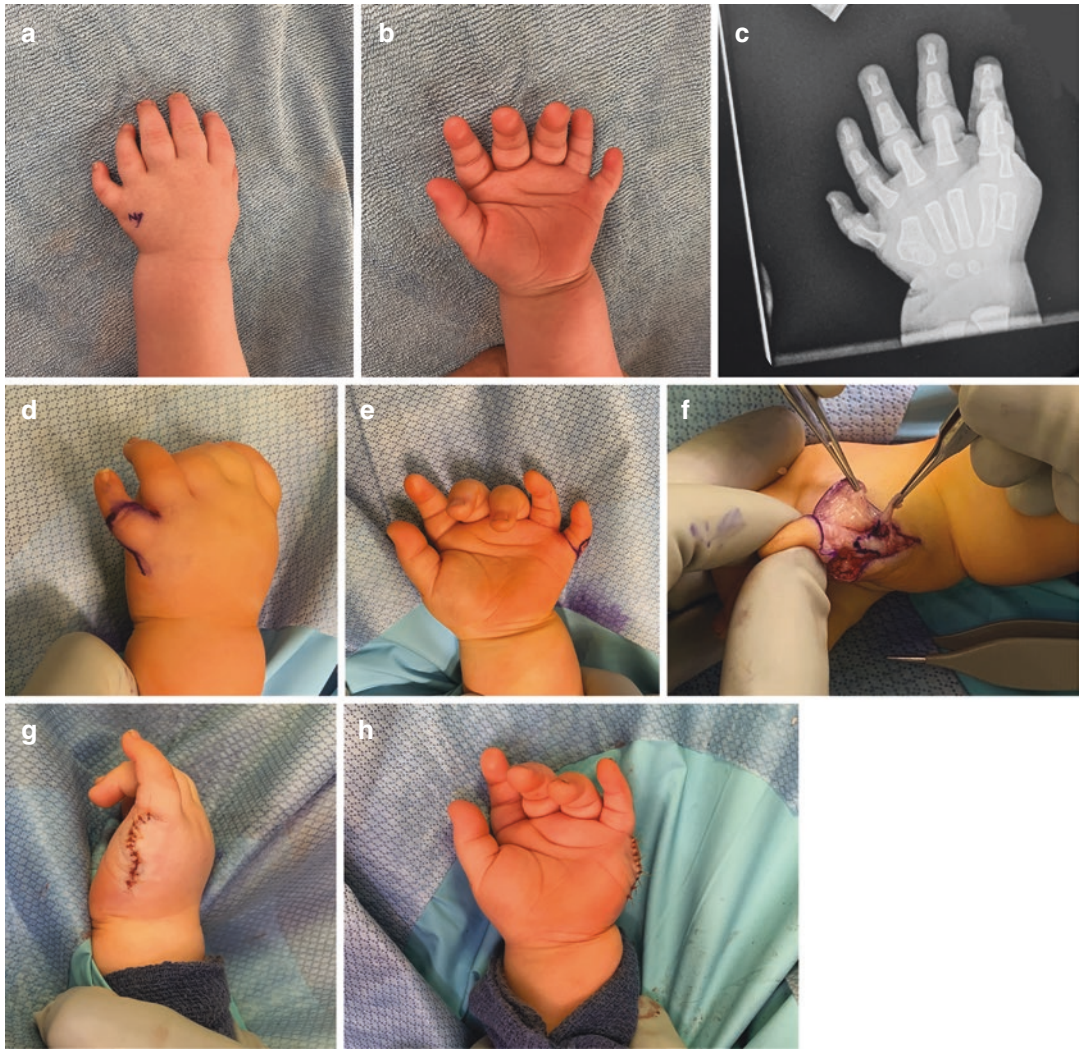

Fig. 40.11 Postaxial polydactyly, Temtamy and McKusick Type A in a 12-month-old male. Bilateral hands were involved (left illustrated). (**a**, **b**) Preoperative appearance. Patient had full mobility of supernumerary digit. (**c**) X-ray demonstrates abnormal 5th metacarpal with synostotic accessory articular element articulating with accessory digit. (**d**, **e**) Incisional markings with dorsal skin flap to be preserved for closure. (**f**) Extensor elements and abductor digiti minimi insertion preserved and reattached to small finger ulnar extensor mechanism. Note collateral ligament with periosteal attachment marked for transfer to proximal phalanx base. (**g**, **h**) After closure

tive procedures share the common goals of providing:

1. A near normal-sized, normal appearing thumb
2. A stable digit with normal MP and IP joint motion
3. Normal growth without developing angular deformity

Wassel type 1 and 2 thumbs typically present with a broad thumbnail or a true cleft separating two equal-sized thumb tips. If there is broad thumbnail that is aesthetically acceptable, no treatment is indicated. In the case of two well-formed "hemi-thumbs," ablation of one of the digital tips will leave an unacceptably small thumb tip. In such cases, the two "hemi-thumb"

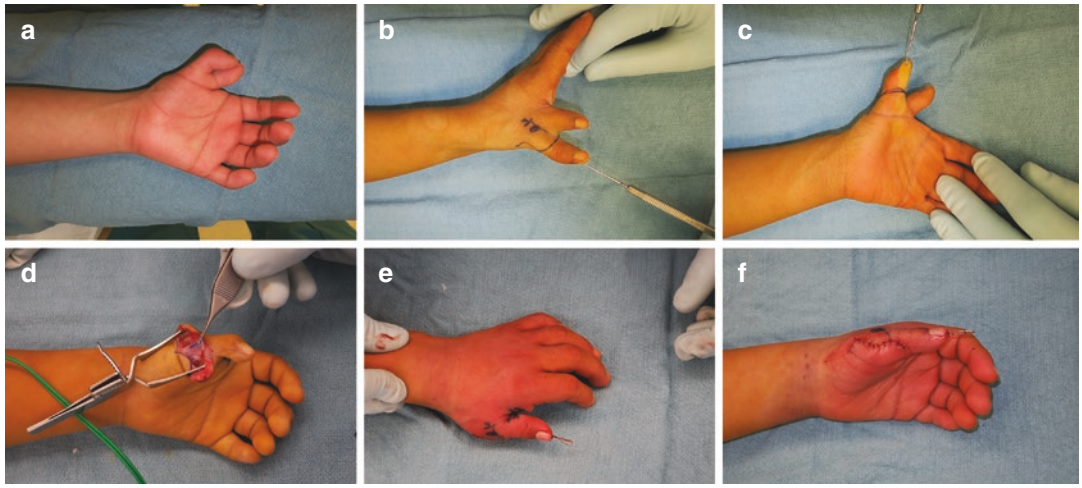

Fig. 40.12 (**a**–**f**) Preaxial polydactyly or "split-thumb" deformity. (**a**) Wassel IV thumb duplication. (**b**, **c**) Preop markings. (**d**) Preservation of tissue for reconstruction of radial collateral ligament. (**e**, **f**) Following closure with K-wire in place

tips are combined according to a modification of the Bilhaut-Cloquet procedure described by Baek et al. in 2007 [12]. In the Baek modification, asymmetric resections of skin, nailbed, and distal phalanx are carried out on the cleft between the two thumb tips and the residual tissues combined (Fig. 40.12).

For most Wassel 3 thumbs and virtually all Wassel 4, 5, and 6 thumbs, the goal is to ablate the smaller, less mobile, and more deformed thumb. The choice of which thumb remains is usually straightforward; however, some cases may require careful clinical and X-ray examination and a balancing of the objectives listed above in order to identify the preferred thumb. Thumb reconstruction proceeds as described for postaxial polydactyly (Fig. 40.13). A modified racquet incision is designed to maintain more skin than will be needed for closure so that the excess skin can be trimmed at the end of the case. Whereas joint instability is uncommon following excision of postaxial polydactyly, intraoperative thumb MP joint instability is common following excision of the accessory thumb. Wassel 4 split thumb is the most common subtype. In such cases, one MP joint collateral ligament will be attached to the resected thumb and must be preserved along with a flap of proximal phalanx perichondrium from the deleted thumb (Fig. 40.13d). If collateral ligament instability has been introduced by resection of the accessory thumb, the ligament must be reconstructed using suture. Gross instability is stabilized using a single 0.028 k-wire passed down the thumb tip and across the MP joint (Fig. 40.13f). The extremity is splinted in a sugartong splint or long-arm cast and the pin is maintained for 4–6 weeks. If minimal joint instability has been introduced, the collateral ligament should still be reconstructed, but sufficient immobilization may be achieved with the cotton gauze splint described above.

When the two thumbs are of similar caliber, motion, and stability, preference is given to maintaining the more ulnar thumb, as this maintains the more functionally important MP joint ulnar collateral ligament. Despite restoration of MP joint stability, the reconstructed thumb may still have residual angular deformity. Such deformity will only get worse as the child grows, and should be treated at the time of thumb ablation with a closing wedge osteotomy to the thumb metacarpal so that the metacarpal and remaining proximal phalanx are collinear. Accessory flexor and extensor tendons from the deleted thumb should be divided distally. If they have normal excursion, they can be transposed onto the retained

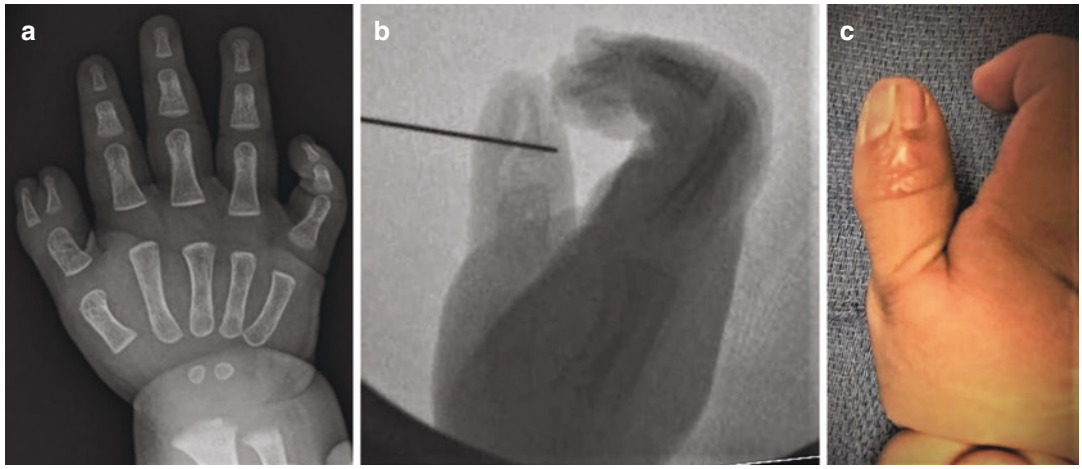

Fig. 40.13 (**a**) Wassel type 2 thumb duplication. (**b**) K-wire fixation during Bilhaut-Cloquet procedure. (**c**) Postoperative result from the Baek modification of the Bilhaut-Cloquet procedure. Note midline nail deformity and eponychial scar, which are downsides of this reconstruction. Benefits of this technique include a wider thumb and avoidance of instability at the interphalangeal joint by preserving native collateral ligament attachments

digit in order to centralize the pull of these tendons. If the tendons have no excursion, they can sometimes be useful tissue for restoring joint stability. Wassel type 7 split thumbs are rare (20% of all split thumbs) and are largely treated as described above; however, the extra joint may require special attention.

Thumb Hypoplasia

Background

Thumb hypoplasia encompasses a wide spectrum of thumb underdevelopment, ranging from subtly diminished size to complete absence. Intermediate grades are characterized by varying degrees of thenar muscle deficits, carpometacarpal (CMC) joint instability, and first web space contracture. The Blauth classification, as modified by Manske [13, 14] (Table 40.4), is most commonly used to describe hypoplastic thumbs. The single most critical factor in determining treatment is the presence of an unstable CMC joint (Blauth 3b or higher). The patient with an unstable CMC joint will require pollicization of the index finger, whereas the thumb with a stable CMC can be salvaged and reconstructed as detailed below. The stable CMC joint is the minimal required base upon which to reconstruct the thumb.

Thumb hypoplasia can be found in isolation or can be part of a larger radial deficiency. Radial deficiency has a very high (up to 80%) association with syndromes, many of which can have life-threatening features. These include VACTERL, Holt-Oram, thrombocytopenia-

Table 40.4 Blauth classification of hypoplastic thumb, as modified by Manske [13, 14]

Blauth type	Description	Treatment
Type 1	Slightly small, with all elements well formed	None
Type 2	Small thumb with: 1. Narrow 1st web 2. Unstable MPJ 3. Deficient thenar muscles	1. First web deepening 2. Opponensplasty 3. UCL repair/ plication
Type 3a	Type 2 plus: 1. Extrinsic tendon abnormalities 2. Metacarpal hypoplasia 3. Stable CMC joint	4. First web deepening 5. Opponensplasty UCL repair/ plication
Type 3b	Type 2 plus: 1. Extrinsic tendon abnormalities 2. CMC instability	Pollicization
Type 4	Floating thumb (pouce flottant)	Pollicization
Type 5	Absent thumb	Pollicization

absent radius (TAR) syndrome, and Fanconi anemia.

Evaluation

Key elements of the physical examination are assessing for the presence and function of the thenar musculature, first web space limitation, and MP and CMC joint stability. Observe the patient during play to determine if the child uses the thumb when grasping objects, versus excluding it from function and using the index finger in its place. This can help to differentiate 3a from 3b and thus predict which children are better served by pollicization. It is also important to assess for normal range of motion of the index finger if pollicization is being considered.

The entire upper extremity should be examined, especially looking for signs of radial deficiency.

Plain radiographs should be obtained and can help in classifying the degree of thumb hypoplasia. Type 3b hypoplasia classically demonstrates a tapered metacarpal without an apparent base. If concern for radial deficiency exists, forearm radiographs should be obtained. Genetics consultation should be sought if there is any evidence of radial deficiency due to the association with the syndromes mentioned above.

Treatment

The overall goal in the treatment of thumb hypoplasia is to achieve the most functional thumb possible for the patient. Basic elements of the functional thumb are the ability to oppose, length to allow prehension, and stability to act as a post for grasp. The specific procedures used are guided by the Blauth classification.

Type 1 thumb hypoplasia is defined by mild generalized decreased size of the thumb with normal function, and no surgical treatment is required.

Type 2 thumb hypoplasia is characterized by decreased overall size, underdevelopment of thenar musculature, narrowing of the first web space, and instability of the ulnar collateral ligament (UCL) of the MP joint. Useful reconstructive techniques include the following:

- First web space widening/deepening. When radial abduction of the thumb is less than 50 degrees, consideration of procedures that widen and deepen the web is warranted. Four and five flap Z-plasty techniques are most commonly employed. The authors' preferred technique is the four-flap Z-plasty (Fig. 40.14).

Technique

- With the thumb and index finger maximally abducted, the central limb of the Z-plasty is marked along the edge of the web (see Fig. 40.8a, b). The length of the central limb determines the length of all of the other limbs. Extension of this limb onto either the ulnar thumb or radial index finger surfaces should be avoided.
- A perpendicular limb of the same length is drawn, arising at the radial-most aspect of the central limb and running proximally, parallel to the first metacarpal. A second perpendicular limb is then drawn arising at 90 degrees from the ulnar-most aspect of the central limb, running volarly near the thenar crease. The two right angles are then bisected with 45 degree limbs, creating four equal triangular flaps. Two of these flaps are on the dorsum, and two are on the volar, glabrous surface of the hand. The volar flaps should be raised first, as it is easier to make small adjustments to the more mobile dorsal skin flaps.
- The volar flaps are incised and raised in the suprafascial plane using careful spreading dissection. The radial digital neurovascular bundle of the index finger and ulnar bundle of the thumb must be identified and protected. Next, the dorsal skin incisions are made, and the flaps are raised suprafascially while protecting the small branches of the radial sensory nerve and any longitudinal veins. If the fascia of the first web is constricting, it should be released as well (see Fig. 40.8b). A small amount of muscle can be resected to further deepen the web space.
- When well designed and completely dissected, the Z-plasty flaps should easily transpose to their new positions. Minor flap adjustments are typically required prior to

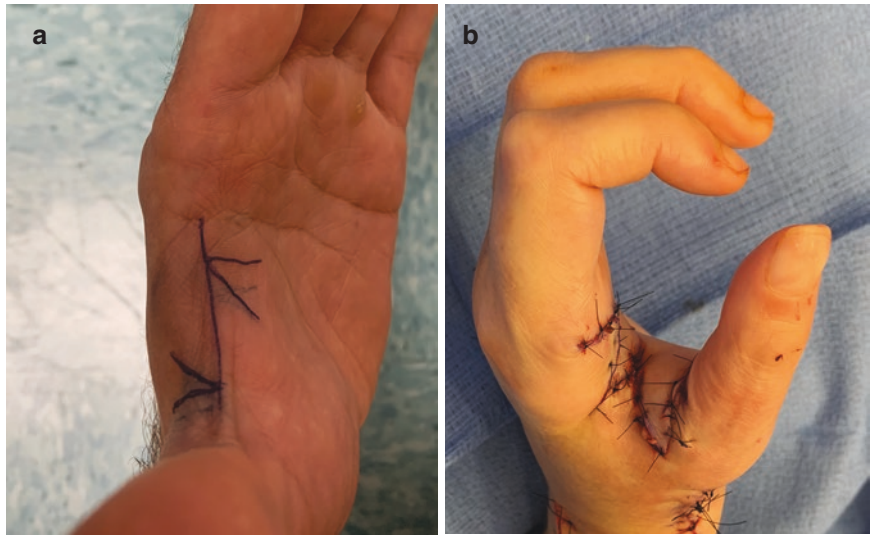

Fig. 40.14 (**a**, **b**) 4-flap Z-plasty for deepening first web space. See text for details

inset. In children, we typically use absorbable monofilament (5-0 fast absorbing gut) sutures. The four-flap design should create s natural concave contour in the first web space.

- Incisions are covered with a nonstick gauze, and a well-padded thumb spica splint or cast is applied with the thumb radially and palmarly abducted and the interphalangeal joint free. In children younger than school age, long-arm splints are less likely to be removed by the patient.
- Opponensplasty. The authors' preferred technique utilizes abductor digiti minimi (ADM; Huber opponensplasty). In addition to providing opposition of the thumb, ADM transfer adds bulk in the thenar region, improving the aesthetic appearance (Fig. 40.15). It is important to identify and protect the neurovascular pedicle to ADM, located proximally, deep to the muscle. The muscle is passed through a generous subcutaneous tunnel and the tendinous end is sewn into the radial aspect of the thumb MP joint, at the APB insertion with nonabsorbable braided suture.
- Repair or reconstruction of the UCL. If there is 20 degrees or more difference in laxity of the UCL as compared to the contralateral side, consider reinforcing the UCL with nonabsorbable braided suture.

Type 3 thumb hypoplasia is divided into 3a and 3b, which are differentiated by the presence or absence of a stable CMC joint. Type 3b or above requires pollicization, as described below. The type 3a thumb is characterized by the deficiencies of type 2 and additional deficiencies of the extrinsic musculotendinous units (EPL and FPL). They metacarpal may also be significantly hypoplastic.

- EPL deficiency may be addressed by performing tendon transfer of EIP to EPL.
- FPL deficiency may be more difficult to treat due to deficiency of the tendon sheath.
- In type 3 hypoplasia, there may be an abnormal connection between FPL and EPL known as pollex abductus. When present, this must be divided, as it interferes with IP joint motion and can weaken the UCL over time.

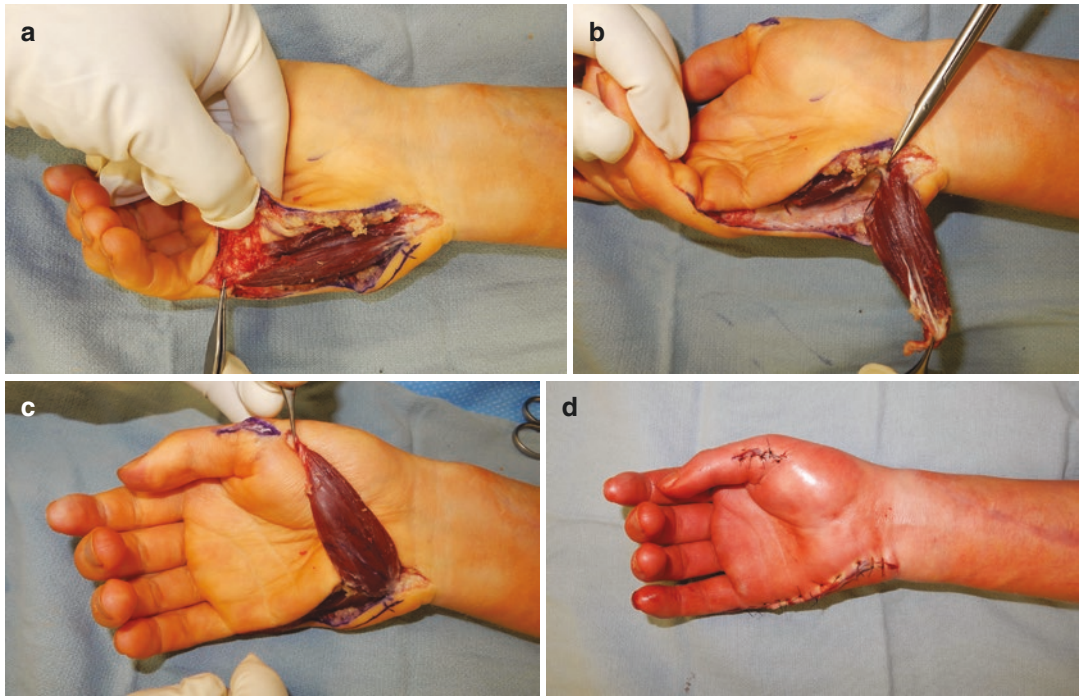

Fig. 40.15 (**a**–**d**) Huber opponensplasty

Pollicization of the index finger is the treatment of choice for type 3b, 4, and 5 thumb hypoplasia. *Index pollicization is a very complex and technically demanding procedure. An in-depth technical guide is beyond the scope of this chapter. What follows is a simplified list of surgical steps to help the reader conceptualize the procedure.*

Steps of Pollicization

1. Incisions are designed as a modified "fish-mouth," with the volar flap extending to the PIP joint and the dorsal flap extending to the mid-point of the proximal phalanx. A curvilinear incision extends along the radial border of the hand for the length of the metacarpal.
2. Identify neurovascular bundles and ligate proper (radial) digital artery to the middle finger.
3. Release A1 and A2 pulleys.
4. Isolate dorsal veins.
5. Divide the EIP and EDC (become EPL and APL).
6. Release the deep transverse inter-metacarpal ligament, preserving the collateral ligaments of the MPJ and a cuff of tissue.
7. Isolate the intrinsics (palmar interossei, 1st and 2nd dorsal interossei).
8. Metacarpal osteotomy through the physis.
9. Epiphysiodesis of metacarpal head (becomes new trapezium).
10. Hyperextend MPJ and pass K-wire (0.035 or 0.045).
11. Fix digit at 20° radial abduction, 35° palmar abduction, and 100° of pronation.
12. Advance interossei:
    (a) 1st dorsal interosseous advanced to base of middle phalanx (becoming abductor pollicis brevis)

(b) 2nd dorsal interosseous advanced to ulnar aspect of PIPJ (becoming adductor pollicis)

13. Plicate EIP (shorten with 3-0 or 4-0 ticron) to become EPL.
14. Shorten EDC (cat and suture to base of proximal phalanx extensor mechanism) to become abductor pollicis longus.
15. Release tourniquet and check perfusion.
    (a) Wrist block with 2% lidocaine.
    (b) 4% lidocaine pledgets to vessels.
16. Closure – be diligent to ensure not constricting perfusion or venous drainage.
17. Dressing – long-arm thumb-spica cast with elbow at 100 degrees of flexion.

Conclusion

This chapter has described practical considerations for some of the more common congenital pediatric hand conditions treated by hand specialists. Congenital hand surgery is an enormous and complex field, and while this chapter only scratches the surface, we hope the fundamental concepts will aid in caring for this patient population by providing practical details not covered in more comprehensive texts.

References

1. Kozin SH. Upper-extremity congenital anomalies. J Bone Joint Surg Am. 2003;85(8):1564–76. Review.
2. De Smet L; IFSSH, International Federation for Societies for Surgery of the Hand JSSH, Japanese Society for Surgery of the Hand. Classification for congenital anomalies of the hand: the IFSSH classification and the JSSH modification. Genet Couns. 2002;13(3):331–8.
3. Malik S. Syndactyly: phenotypes, genetics and current classification. Eur J Hum Genet. 2012;20(8):817–24.
4. Buck-Gramcko D. Congenital malformations. In: Nigst H, Buck-Gramcko D, Millesi H, Lister GD, editors. Hand surgery, vol. 1. New York: Thieme; 1988. p. 12.22–3.
5. Shinya K. Dancing girl flap: a new flap suitable for web release. Ann Plast Surg. 1999;43(6):618–24.
6. Kikuchi N, Ogino T. Incidence and development of trigger thumb in children. J Hand Surg Am. 2006;31(4):541–3. PubMed PMID 16632044.
7. Dinham JM, Meggitt BF. Trigger thumbs in children: a review of the natural history and indications for treatment in 105 patients. J Bone Joint Surg Br. 1974;56(1):153–5.
8. Buchman MT, Gibson TW, McCallum D, Cuda DD, Ramos AG Jr. Transmission electron microscopic pathoanatomy of congenital trigger thumb. J Pediatr Orthop. 1999;19(3):411–2.
9. Shah AS, Bae DS. Management of pediatric trigger thumb and trigger finger. J Am Acad Orthop Surg. 2012;20(4):206–13.
10. Temtamy SA, McKusick VA. The genetics of hand malformations. Birth Defects Orig Artic Ser. 1978;14(3):i–xviii, 1–619.
11. Wassel HD. The results of surgery for polydactyly of the thumb: a review. Clin Orthop Relat Res. 1969;64:175–93.
12. Baek GH, Gong HS, Chung MS, Oh JH, Lee YH, Lee SK. Modified Bilhaut-Cloquet procedure for Wassel type-II and III polydactyly of the thumb. J Bone Joint Surg Am. 2007;89(3):534–41.
13. Blauth W. The hypoplastic thumb. Arch Orthop Unfallchir. 1967;62:225–46. [Article in German].
14. Manske PR, McCarroll HR Jr, James M. Type III-A hypoplastic thumb. J Hand Surg Am. 1995;20:246–53.