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Congenital Hand Differences

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Congenital hand differences are frequently encountered by pediatric plastic surgeons. These anomalies may cause significant emotional and functional challenges for children. Pediatric plastic surgery nurses should have a basic understanding of common congenital hand differences and related treatment options to facilitate patient education and postoperative care. This article discusses clinical findings and management of 4 of the most common hand anomalies: syndactyly, polydactyly, thumb hypoplasia, and cleft hand. The goals of surgical treatment are to maximize hand function and aesthetics with minimal adverse outcomes.

Congenital hand differences are reported to occur in approximately 2 in every 1,000 live births (Ekblom, Laurell, & Arner, 2010; Lamb & Wynne-Davies, 1998). Syndactyly, polydactyly, thumb hypoplasia, and cleft hand are some of the most common differences encountered in a pediatric hand surgery practice. These anomalies often occur as an isolated malformation, although they have also been associated with at least 127 known syndromes (Rayan & Upton, 2014). Despite the mild to moderate functional deficits resulting from the aforementioned congenital hand anomalies, such diagnoses invariably cause significant psychological distress in both parents and children (Andersson, Gillberg, Fernell, Johansson, & Nachemson, 2011). Communication throughout the treatment process by both the hand surgeon and the nursing staff can improve outcomes by addressing all physical and emotional needs of both the patient and parents (Bradbury & Hewison, 1994; Bradbury, Kay, & Hewison, 1994).

HAND DEVELOPMENT

A child achieves a functional grasp by the age of 9 months, learns three-digit pinching by the age of 2 years, and establishes patterns of hand–eye coordination by the age of 3 years (Bates, Hansen, & Jones, 2009). Reconstruction of congenital hand differences should be complete by school age to minimize the impact on functional and psychological growth as well as social transitioning (Bates et al., 2009; Smith & Lipke, 1979). As the child's hand doubles in size in the first 2 years of life, these cases provide the surgical team with an exciting opportunity to monitor the function and appearance of surgical reconstruction throughout the patient's childhood (Flatt, 1994).

SYNDACTYLY

Syndactyly, or webbing of the fingers, is the most common congenital difference in the upper limb (Figure 1). It occurs in approximately 1 in 2,000 live births, most frequently in Caucasian males (Rayan & Upton, 2014). This condition is commonly inherited in an autosomal dominant fashion with variable expressivity and incomplete penetrance (Bosse et al., 2000; Kozin, 2001). Syndactyly is defined by the extent of fusion between adjacent digits (Kozin, 2001). The deformity is classified as *complete* when the interconnection extends through the entire length of the adjacent fingers, whereas *incomplete* syndactyly denotes an interconnection that ends proximal to the fingertip. *Simple* syndactyly involves only skin and fibrous tissue, whereas *complex* syndactyly denotes fusion of bones (Flatt, 1974).

The treatment goal of syndactyly reconstruction is to create a more natural webspace to improve the function and appearance of each finger (Kim & Chung, 2008). Surgery is indicated for all types of syndactyly, although the timing of reconstruction varies by the degree of complexity, the webspace involved, and surgeon preference. Ideally, syndactyly reconstruction should occur when the child is between 12 and 18 months of age. For border digit (thumb and small finger) involvement, surgery is recommended around 6 months of age. In these cases, surgery is indicated as soon as the child can safely undergo general anesthesia in an effort to limit permanent flexion contractures or rotational deformities that may occur if a smaller digit is fused to a longer one (Kozin, Zlotolow, & Ratner, 2014). Syndactyly involving more than one adjacent webspace must be staged to avoid operating on both sides of a single finger because this can cause vascular compromise. Surgical separation of the

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The authors report no conflicts of interest.

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DOI: 10.1097/PSN.0000000000000133

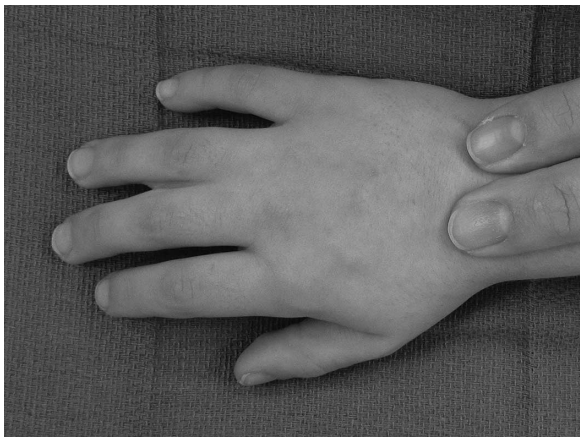

FIGURE 1. Syndactyly.

fingers involves some combination of adjacent skin flaps and full-thickness skin grafts harvested from the groin or extremity (Figure 2).

The separation of syndactylous digits generally improves appearance and motion, with a low chance for major complications. When complications occur, they are often related to skin necrosis at the edges of flaps, skin graft failure, and scar contracture (Oda, Pushman, & Chung, 2010). Web creep (longitudinal extension of the scar) is another complication that can occur as the child grows; this can be addressed with future surgery. Complex syndactyly, with inherent skeletal deformities, is often associated with more complications, a higher risk of contracture, and thus a greater loss of mobility (Toledo & Ger, 1979).

POLYDACTYLY

Polydactyly (an excess number of digits) is the second most common congenital hand anomaly. Preaxial (radial) polydactyly, also known as thumb duplication, is the most common form overall and is seen most frequently

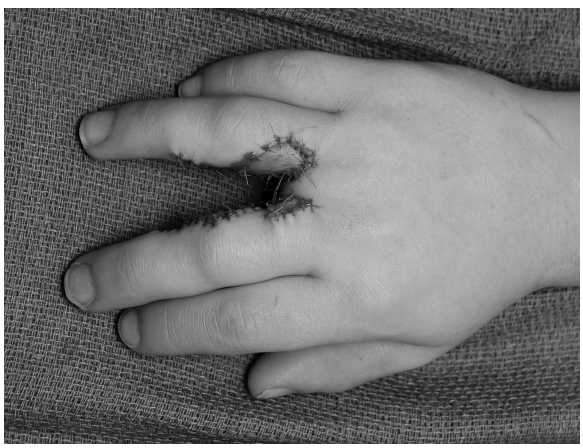

FIGURE 2. Syndactyly, surgical separation of the fingers.

in Caucasians and Asians (Ezaki, 1990; Graham & Ress, 1998; Wassel, 1969; Watson & Hennrikus, 1997). It occurs in approximately 8 in 100,000 births and is often sporadic, unilateral, and rarely associated with systemic conditions (Graham & Ress, 1998). Postaxial (ulnar) polydactyly involves duplication of the small finger and is more commonly seen in the Black population, with an estimated incidence as high as 1 in 300 live births (Oda et al., 2010; Watson & Hennrikus, 1997). It is commonly inherited in an autosomal dominant fashion with variable penetrance. Thumb duplication is defined by the anatomical level of duplication from distal to proximal. Postaxial polydactyly is subclassified as Type A when the extra small finger is well developed (Figure 3, *left*) versus Type B (Figure 3, *right*) when the digit is rudimentary and pedunculated (Temtamy & McKusick, 1978).

Surgical treatment of polydactyly requires careful consideration of the level of duplication, musculoskeletal components involved, developmental stage of the child, and cosmetic outcomes. The goal of any polydactyly correction is to achieve a single, stable, functional digit with a more natural appearance; surgery should never make function worse. Extra rudimentary digits seen in postaxial Type B polydactyly can often be managed with ligation at the base of the finger shortly after birth. This will cause distal necrosis of the digit, which leads to autoamputation (Kozin & Zlotolow, 2015). Alternatively, the extra digit can be surgically excised to improve the cosmetic outcome and prevent a future neuroma stump. Well-developed postaxial Type A digits always require surgical excision, proximal amputation of the neurovascular bundle, and primary closure, with tendon or ligament transfers, as needed (Kozin & Zlotolow, 2015).

Preaxial polydactyly (thumb duplication) poses a unique problem for treatment, as neither of the duplicated thumbs is anatomically normal, with both exhibiting some degree of hypoplasia. Surgery is recommended to optimize function and improve appearance. Typically, the radial duplicate is resected and the collateral ligament reconstructed (Watt & Chung, 2009; Figure 4). Persistent deviation of the reconstructed thumb may require osteotomy and/or tendon transfers. Treatment of more complicated cases requires individualized surgical planning, often incorporating the best components of each thumb into a single reconstructed digit (Kozin & Zlotolow, 2015).

Realistic expectations are important when discussing surgical outcomes in cases of polydactyly reconstruction. Thumb asymmetry, joint stiffness, growth arrest, and asymmetric growth are all potential complications following reconstruction (Tonkin & Bulstrode, 2007). Satisfactory outcomes are often obtained for postaxial polydactyly and simple or symmetrical preaxial polydactyly (Ogino, Ishii, Takahata, & Kato, 1996). However, certain complicated types of preaxial polydactyly are more

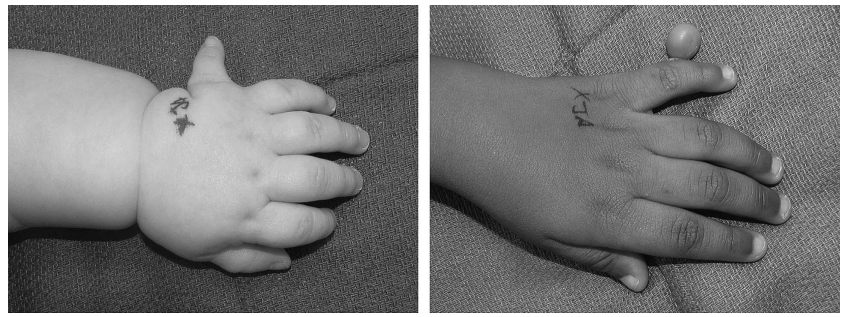

FIGURE 3. Postaxial polydactyly, Type A (left) and Type B (right).

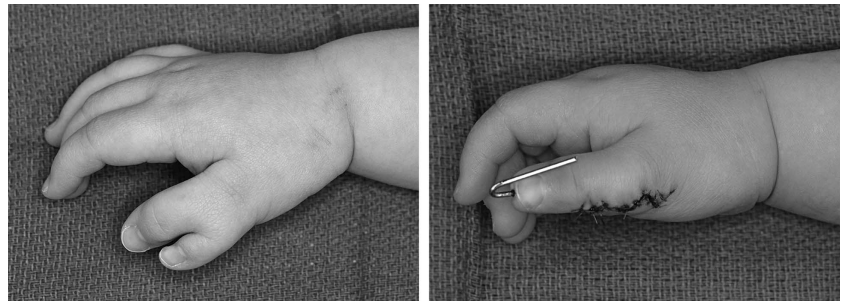

FIGURE 4. Preaxial polydactyly.

likely to yield unsatisfactory results related to difficulties of bony alignment and joint instability (Horii, Nakamura, Sakuma, & Miura, 1997).

THUMB HYPOPLASIA

Congenital thumb hypoplasia is defined as a short, underdeveloped thumb with deficient or absent intrinsic (in the hand) or extrinsic (not in the hand) musculoskeletal structures (Kozin, 2003; Figure 5). This condition occurs within a spectrum of radial deficiencies or hypoplasia along the radial side of the entire upper limb and is also associated with certain systemic syndromes (e.g., Holt–Oram syndrome, VATER anomalies, or Fanconi anemia; Abdel-Ghani & Amro, 2004; James, McCarroll, & Manske, 1996). Such radial longitudinal deficiencies can range from mild underdevelopment of the thumb to complete absence of the thumb and the radius (Little & Cornwall, 2016). As such, this diagnosis warrants a thorough musculoskeletal and systemic examination.

Surgical treatment of the hypoplastic thumb is guided by the presence or absence of a functional joint at the base of the thumb (carpometacarpal [CMC] joint). In children with a stable thumb CMC joint, the existing thumb is reconstructed using a combination of first (thumb–index) webspace deepening, thumb ligament reconstruction, and muscle–tendon transfer (i.e., opponensplasty). The muscle transfer provides critical thumb motion to allow for pinch and grasp activities. Conversely, an unstable CMC joint requires ablation of the digit and subsequent pollicization (Kozin et al., 2014).

Pollicization is a complex procedure in which a functioning digit (usually the index finger) is substituted for the absent/hypoplastic thumb (Figure 6). Surgical timing

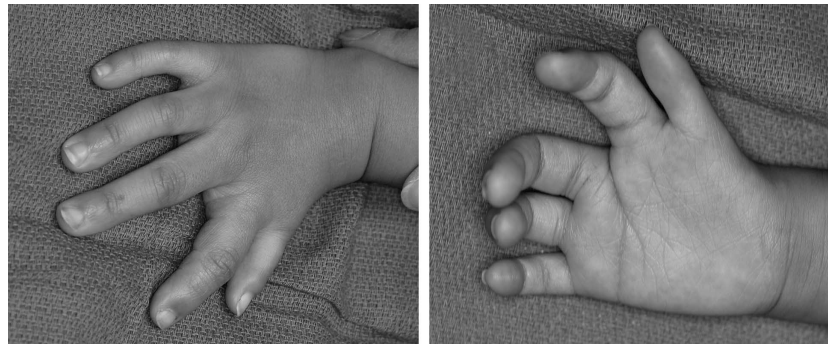

FIGURE 5. Thumb hypoplasia.

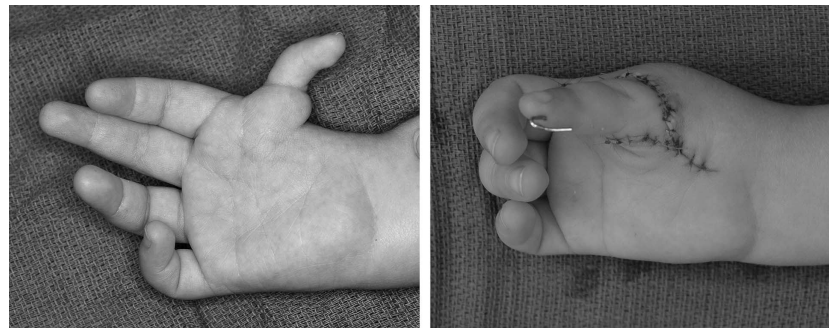

FIGURE 6. Pollicization of hypoplastic thumb.

depends on the child's motor learning associated with functional development of the thumb and generally occurs between the ages of 1 and 2 years (Little & Cornwall, 2016). The decision to proceed with pollicization is often difficult for parents and requires a thorough discussion of the procedure and outcomes. The parents often have difficulty understanding why the existing thumb is not reconstructible and must be amputated to improve function. The provision of photographs of similar procedures and coordinating conversations/meetings with other parents whose children have undergone reconstruction can be useful to guide the decision-making process (Kozin & Zlotolow, 2015).

Critical goals of pollicization include minimizing scar formation, transfer of muscles needed for thumb motion, growth plate obliteration to prevent excessive growth of the new thumb, avoidance of thumb hyperextension with pinch, and fixation of the new thumb in a natural position (Kozin & Zlotolow, 2015). Postoperative observation up to 24 hr is occasionally necessary to ensure that vascularity of the transposed digit remains intact (Little & Cornwall, 2016). Functional improvements provide the stability for grasp and the mobility for fine pinch that continue into adulthood (Clark, Chell, & Davis, 1998). Chronic problems due to poor positioning, hyperextension of the thumb, or webspace scarring may develop, although these outcomes can be improved with secondary procedures later in life (Kozin & Zlotolow, 2015).

CLEFT HAND

Cleft hand, also known as central ray deficiency or ectrodactyly, refers to a variety of deformities in which the central portion of the hand is missing (Figure 7). Cleft hand is characterized by variable expression in a wide range of clinical phenotypes (Rayan & Upton, 2014). This condition can occur due to a spontaneous mutation, autosomal dominant inheritance, or in the setting of many syndromes (e.g., split-hand/split-foot syndrome, ectodermal dysplasia, cleft-lip/cleft-palate syndrome; Ianakiev et al., 2000). Despite a noticeably poor appearance of the hand, function is actually quite satisfactory (Kozin & Zlotolow, 2015) and many of these children require no treatment. However, the abnormal appearance of the hand and social stigmata of a congenital difference lead many parents to seek surgical consultation.

Surgery is predominantly indicated to treat any associated syndactyly or abnormal first webspace that might negatively impact function (James et al., 1996). Typically, the cleft hand with a syndactylous first webspace is repaired by the Snow–Littler procedure, in which both abnormalities are corrected simultaneously (Snow & Littler, 1967). In this procedure, the skin covering the cleft is raised and transposed into the widened first webspace. If the digits are also affected by syndactyly, the fused digits can be released at the same time as cleft closure. Simpler cases without a narrow first webspace can be reconstructed by similar techniques, closing the cleft by transposition of the index finger to the middle finger position (Miura & Komada, 1979). In more complicated cases, any transverse bones must be removed from the cleft to prevent widening of the cleft as the abnormally oriented bones grow over time (Bates et al., 2009). Outcomes of cleft hand repair are generally acceptable and depend upon careful transposition of the index finger and restoration of adequate commissures in both the first webspace and within the cleft.

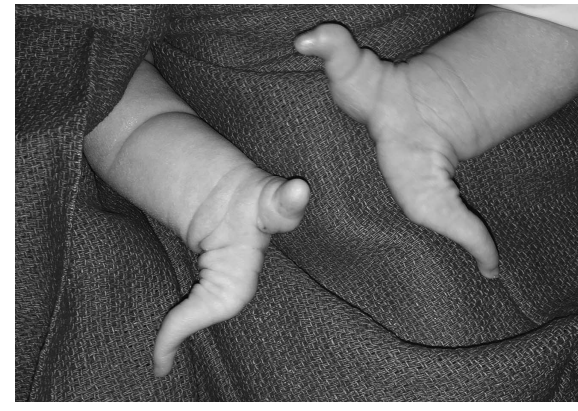

FIGURE 7. Cleft hand.

CONCLUSION

Embarking on surgical reconstruction of congenital hand differences provides the surgeon a unique opportunity to improve physical and psychosocial functions for a child. With a greater understanding of the common congenital hand differences and related treatment options, the surgical nurse is afforded an opportunity to ensure that each reconstructed hand is smoothly integrated into a more natural development for the child.

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