CONGENITAL HAND DIFFERENCES vary in their functional and aesthetic implications. This article explores recent developments for 6 common hand differences: polydactyly, syndactyly, camptodactyly, clinodactyly, trigger thumb, and cleft hand. Although there has been a great deal of progress in our understanding of upper limb development, the molecular mechanisms responsible for these abnormalities are only beginning to be appreciated. This progress in our understanding of the nature and role of various growth factors and signaling mechanisms is exciting and carries great promise for the future.

Although these 6 hand abnormalities all influence function in some way, affected children often compensate remarkably well. This ability to compensate is due to 3 basic factors: the early age at onset (birth or shortly thereafter) allows children to accommodate their hand difference; the opposite hand is often not involved, allowing either bimanual or 1-handed activities; and the specific deficiencies of this set of congenital hand differences do not substantially affect function. In contrast, hand appearance can be notably abnormal, with a corresponding social effect. There is a role for surgical intervention for each of these differences, but the timing, nature, and underlying need for any intervention varies based on the particular hand difference and its effect on the individual child.

ULNAR POLYDACTYLY

Ulnar polydactyly, particularly common in African American children with an autosomal dominant inheritance pattern, is most often treated with suture ligation or simple surgical excision. If the extra digit arises at the small finger metacarpophalangeal joint, reconstruction of the joint is performed to maintain stability of the preserved finger.

Radial polydactyly, in contrast, requires a more extensive reconstruction to maintain function and restore a more normal appearance. The Wassel classification, which uses the level of skeletal duplication to categorize the deformity, has been universally accepted, as it is an easy-to-apply system and has stood the test of time. Zuidam et al. noted that a major deficiency of the Wassel classification system was that the more complex deformities (such as the proximal extra digits, triphalangeal polydactyly, and triplicate thumbs) are not well described; their recent modification should assist in understanding and treating these especially complex presentations (Fig. 1).

Surgical reconstruction of radial polydactyly aims to produce a well-aligned, normally sized, and stable thumb; nail appearance and joint motion are less important to a successful outcome. Ideally, the smaller thumb is excised and the larger (typically ulnar) thumb

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<th>Level of Duplication</th>
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<td>I partial duplication distal phalanx</td>
<td>T Triplication</td>
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<tr>
<td>II complete duplication distal phalanx</td>
<td>Tph Triphalangism</td>
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<td>III partial duplication proximal phalanx</td>
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<td>VII partial duplication carpal bones</td>
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<td>VIII complete duplication carpal bones</td>
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The Zuidam classification system includes types I-VIII, each with specific anatomical features and associated deformities.
reconstructed to achieve these goals. Treatment of type IV radial polydactyly (the most common polydactyly, with a duplication at the level of the metacarpophalangeal joint), requires centralization of the preserved digit, collateral ligament reconstruction, narrowing of the metacarpal head if it is markedly wide, and osteotomy if bony deformity causes angulation.

Alternatively, a modification of the Bilhaut-Cloquet procedure might provide a satisfactory outcome for types II and III thumbs or types III, IV, and VII thumbs. In the modification for types II and III thumbs described by Baek, the majority of one thumb (including physis, joint, and nail elements) is combined with a smaller portion of the other thumb; by avoiding a central merging of the duplicated digits, the difficulties of the classic Bilhaut-Cloquet procedure (central nail ridge, joint stiffness, and growth arrest) are minimized while still achieving a satisfactory thumb size and stability. In a report of 7 thumbs, interphalangeal joint motion and growth were maintained and satisfaction with both appearance and function was high.

SYNDACTYLY

Syndactyly, one of the most common extremity differences, can be cutaneous or bony, complete or partial, or complicated (ie, syndromic); the most common presentation is a cutaneous, complete syndactyly involving the long and ring fingers (Fig. 2). Normally, the webbing between the digits in the developing hand regresses to a typical interdigital web, as signaled by fibroblast growth factors between 6 and 8 weeks of gestation. A failure of this signaling pathway leads to syndactyly with a variable effect on both function and appearance.

Function is affected by both location and the extent of the syndactyly; for example, thumb–index syndactyly is the most limiting, whereas a partial syndactyly between the long and ring fingers results in minimal impairment. Surgery is an appropriate consideration for any child with syndactyly, and surgeon preference usually dictates the timing of intervention. Early intervention at 4 to 6 months for border digit syndactyly might prevent angular growth and deviation of the longer digit.

Complete syndactyly can be reconstructed using a variety of techniques. Full-thickness skin grafts remain a satisfactory adjunct to obtain skin coverage following digital separation; the grafts are most commonly needed at the proximal aspect of each digit, adjacent to the commissural flap. Recently, several techniques have been reported that allow reconstruction without grafts due to the concerns of an additional scar from graft harvest, the propensity for the grafts to darken over time, and the possibility of hair growth on grafts harvested from the groin. Most graftless techniques use the dorsal metacarpal artery flap proposed by Sherif as the commissural flap, with zigzag interdigitating flaps along the dorsal and volar digits. Technical variations to this flap bring new skin to resurface the proximal interdigital space and avoid skin grafts. Defatting the digit and limited closure of the flaps are 2 additional techniques that can facilitate closure. In patients with a complete syndactyly and nail involvement, the flag and pennant technique, which uses skin from the pulp of each digit to reconstruct the lateral nail fold, usually provides an acceptable aesthetic outcome.

Partial syndactyly is treated with skin flaps alone, although skin grafts might occasionally be necessary. Several techniques have been reported with good success, including various island flaps and a 3-square flap; these allow resurfacing of the commissure and digital separation with satisfactory outcome.

CAMPTODACTYLY

Camptodactyly, a flexion contracture of the proximal interphalangeal joint most frequently involving the small finger, has been attributed to a multitude of causes, most notably tightness of the skin, underlying fascia, and flexor digitorum superficialis tendon. Many patients experience little functional impairment, especially with a mild contracture. In patients with functional limitations, nonsurgical care is the first line of
treatment, including serial casting and splinting. Patients with moderate to severe contracture (60° to 90°) and those not responding to splinting might benefit from surgical intervention. The lack of predictability of surgical outcome has led to caution in the recommendation for surgery (Fig. 3).

Foucher et al.\textsuperscript{16} used 155 fingers to evaluate a classification and treatment algorithm. Most of their patients were classified as early and stiff (5 or fewer years of contracture and without passive extension); the others were classified according to time interval (early or late) and mobility (stiff or mobile). The small finger was most commonly involved, and bony abnormality of the proximal interphalangeal joint was noted in 29% of all patients and in 58% of those with joint stiffness. The authors advocate a series of physical examination steps to assess the camptodactyly, including an evaluation of the volar skin, the tightness of the flexor digitorum superficialis, and laxity of the central slip. After specific deformities are identified, a step-by-step surgical plan for each component can be carried out in those patients who fail nonsurgical care. The authors report improved outcomes, including a lower failure rate, in patients treated according to this algorithm.

Smith and Grobbelaar\textsuperscript{17} also stressed the diffuse involvement of the digit, with skin, flexor digitorum superficialis tendon, fascia (retinaculum cutis), and lumbricals most commonly involved. Their recognition of the high surgical failure rate in these patients led to their recommendation of limiting surgery to those patients with functional issues that fail nonsurgical care with at least a 60° flexion contracture.

**CLINODACTYLY**

Clinodactyly most commonly presents as a radial deviation of the small finger due to an abnormal middle phalanx. Radiographs commonly demonstrate a bracketed epiphysis of the middle phalanx; the abnormal physis on the radial side of the phalanx tethers the growth of the middle phalanx. It is rarely painful or functionally limiting, even with severe angulation (Fig. 4). The uncommon patient with a functional limitation due to the deformity might benefit from surgery. Numerous surgical options can lead to functional (and aesthetic) improvement, including osteotomy (opening wedge, closing wedge, or reverse) and physiolysis. Recently, Ali et al.\textsuperscript{18} reported outcomes for 25 fingers with greater than 25° of angulation treated with a closing...
wedge osteotomy. At a mean of 6 years after surgery, the fingers maintained a corrected clinical position of 9° of angulation (33° before surgery). Range of motion was maintained, and patient satisfaction was high. Physiolysis can also be performed to allow restoration of symmetrical growth of the middle phalanx. In this simple surgery, first reported by Vickers,19 a small incision along the radial aspect of the middle phalanx exposes the phalanx and allows resection of a segment of the longitudinal physis; fat interposition prevents repeat tethering. Caouette-Laberge20 reported an average 11° of improvement after physiolysis in 35 children with at least 20° of preoperative angulation. A larger correction was noted in those patients with a severe deformity (≥40° before surgery) and those treated before 6 years of age (average of 20° and 18° of correction, respectively). There were 10 fingers treated with a repeat procedure to address insufficient correction; however, the repeat procedure did not result in any substantial improvement.

**TRIGGER THUMB**

Trigger thumb, one of the most common abnormalities of the pediatric hand, is straightforward to diagnose, but treatment is debated. The most appropriate name for this condition is pediatric trigger thumb rather than congenital trigger thumb, as several investigations have confirmed an absence of trigger thumb at birth. A recent study confirmed that none of 1,116 examined newborns were affected, but the incidence of trigger thumb at 1 year of age was 3.3 cases per 1000 live births.21

The child with a pediatric trigger thumb typically presents the thumb in fixed flexion at the interphalangeal joint, although it may also present as intermittent, painful catching of the interphalangeal joint. In addition to the flexed posture of the interphalangeal joint, there can be a palpable nodule (Notta’s node) at the A1 pulley that is felt to block passive thumb extension. A careful history and physical examination of both thumbs are sufficient to make the diagnosis.

A nonsurgical trial of management might be appropriate for the pediatric trigger thumb. To document the natural history of pediatric trigger thumb, Baek et al.22 prospectively observed 71 thumbs in 55 patients for at least 2 years. The authors considered neutral extension, even if the contralateral thumb hyperextended, as resolution. At an average 4-year follow-up, the average pretreatment 26° flexion posture resolved in 45 thumbs (63%) and improved in most of the rest. Several other studies also suggest a role for nonsurgical care, including stretching, with approximately 50% of children demonstrating resolution at 6 months.23,24 Many hand surgeons prefer surgical release of the A1 pulley to allow thumb extension, as it is a reliable procedure with minimal morbidity;23,24 long-term outcome is satisfactory, although extension may be limited even with surgery in 20%.25

**CLEFT HAND**

Cleft hand, also known as central longitudinal deficiency or split hand–foot malformation, can present as a component of a syndrome or can be an isolated abnormality. It is thought to result from abnormalities of the apical ectodermal ridge in the developing limb bud, but there is genetic heterogeneity, with at least 5 different genetic loci identified for the nonsyndromic cleft hand.26

The deficiency is characterized by absent or deficient central digits (Fig. 5) with a variable deficiency of the first web space and syndactyly between the ring and small finger. A deficient thumb web space is reconstructed to improve function, whereas reconstruction of the central deficiency is performed to correct the notable aesthetic difference. The Manske and Halikis27 classification categorizes central deficiency based on the first web space, thus highlighting the key functional
limitation of these hands; this classification can be used to guide treatment of web space reconstruction. Z-plasty can be performed for mild deficiency, whereas dorsal rotational flaps are necessary for more severe deficiencies.

The Snow-Littler procedure is used to reconstruct the thumb web space using volar rotational flaps and a bony transposition of the index ray in an ulnar direction. Rider et al. reported satisfactory results in 12 patients so treated. A recent study reported on 16 hands reconstructed for central deficiency using a soft tissue rearrangement alone or in combination with a transposition of the index ray to a more ulnar position. Results were satisfactory for most patients, with improved appearance and alignment. The divergence angle, measured radiographically between the index and ring finger metacarpals, improved from 33° to 12°. A functional limitation in some patients, potentially related to intrinsic muscle deficiency, was a flexed posture of the ring finger at the proximal interphalangeal joint.

Isolated congenital hand differences present several challenges. The surgeon must understand both the potential functional aspects and the social (ie, appearance) aspects of these abnormalities in order to provide ideal treatment for each child and family. Therapy or surgical correction plays a role for most of these children; however, some will adapt to their hand difference with no intervention.

REFERENCES