Polydactyly
A Review

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Abstract

Polydactyly of the hand is a difficult problem and poses a unique challenge for the hand surgeon. The embryology of limb development is complex, leading to a host of different phenotypes of polydactyly. Polydactyly can occur in any digit and is described as preaxial, postaxial, and central, based on location. Classification systems exist for each of these locations, which guide treatment options. Surgical treatment needs to address the aesthetic and functional aspect of hand reconstruction. Careful consideration and planning of surgical treatment individualized to each patient is required to obtain the best possible outcome.

Polydactyly is the most common congenital hand anomaly.[1] Characterized by the presence of extra digits beyond the normal complement of one thumb and four fingers, this simple explanation belies the complexity and the heterogeneity of polydactyly presentation.

The earliest recorded documentation of hand polydactyly dates back to 1670, although American Southwest rock art depictions of six fingered hands suggest an even earlier documentation.[2,3] Polydactyly is ubiquitous in nature and has been reported in other species, including cats, horses, pigs, and chickens.[4]

Polydactyly can present on the radial side (preaxial), ulnar side (postaxial) or involve non-border (central) digits. Each subset of polydactyly has its own characteristic ethnic and genetic predilections. Surgical treatment is directed toward creating a functional as well as an aesthetic hand.

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Classification

In 1964, Swanson first proposed a classification system for congenital malformations of the hand.[5] A modified form was adopted by the American Society for Surgery of the Hand (ASSH) and International Federation of Societies for Surgery of the Hand (IFSSH) in 1976. This classification system divides congenital anomalies by the embryologic mode of failure into seven main categories: failure of formation, failure of differentiation, duplication, overgrowth, undergrowth, constriction band syndrome, and generalized anomalies and syndromes. Each category is then further expanded into subcategories based on anatomic location, morphologic appearance, or tissue type. The benefit of this system is the use of simple terms to describe complex anomalies. However, this system is limited by the inability to describe multiple anomalies in the same limb, and not all anomalies are included in the classification.[6]

Embryology

The embryologic limb bud begins development between 26 to 28 days after fertilization. Upper limb digits are recognizable at 41 to 43 days and are fully separate at 53 days. It is within this period of rapid development where congenital hand malformations arise. By the eighth week of gestation, the scaffold of the upper limb is complete, and the limb continues to grow in size for the remainder of the gestational period.

On the cellular level, the differentiation of limb bud into separate digits occurs via a complicated cascade of molecular signaling and pathways. The limb bud originates from the migration of the mesoderm onto the overlying ectoderm on the ventrolateral aspect of the embryo. Lateral plate mesoderm cells are progenitors of bone, cartilage, and tendon,

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whereas somatic mesoderm becomes nerve, muscle, and vasculature. Three signaling pathways control the growth of the limb in a three dimensional axis: the apical ectodermal ridge (AER) controls proximal to distal orientation, the zone of polarizing activity (ZPA) controls anterior to posterior (radial to ulnar) orientation, and the wingless type (WNT-7a) pathway controls dorsal ventral orientation.

The AER is a transient, thickened ridge of ectoderm, which secretes fibroblast growth factors (FGFs) in a positive feedback loop, directing growth along the proximal to distal (PD) axis.[7] Removal of the AER results in truncated limbs, whereas transplantation results in the duplication of the limb in experimental animal models. The ZPA is a condensation of mesenchymal tissue along the posterior aspect of the limb, directing the anterior to posterior orientation of the limb. This tissue secretes Sonic Hedgehog (SHH) protein, a signaling molecule, which works in a concentration dependent method within ZPA. Areas with higher concentration of SHH developed into posterior or postaxial structures, whereas lower concentrations developed into more anterior or preaxial structures. In experimental animal models, transplantation of posterior limb tissue to the anterior aspect resulted in mirror duplication of the digits. This mimics the development of mirror hand in humans. SHH is also important in maintaining PD growth by inducing FGF expression in the AER.[8] Less is known about the signaling pathway controlling dorsal ventral orientation. Wnt-7a is expressed in the dorsal ectoderm and upregulates the production of LIM homeobox transcription factor 1, alpha (Lmx-1), which in turn causes dorsal differentiation of the limb. On the ventral surface, a complementary protein, engrailed-1 (En-1), blocks Wnt-7a production allowing for differentiation into a ventral surface.

Preaxial Polydactyly

Duplication of the thumb is the most common reported type of polydactyly. The incidence of preaxial polydactyly is reported to be as high as 1 in 3000 births.[9,10] More commonly seen in Caucasians compared to African Americans, preaxial polydactyly also has a high incidence in the American Indian and Asian populations. Within these groups, thumb duplications represent up to 90% of all polydactyly cases.[11] Preaxial polydactyly is usually seen as an isolated anomaly, and the etiology is thought to arise from spontaneous mutations. However, a triphalangeal thumb, with or without polydactyly, is linked to an autosomal dominant inheritance pattern.[12] This anomaly is also associated with a higher incidence of systemic syndromes, including Holt-Oram syndrome and Fanconi anemia.

Wassel proposed the most widely used and accepted classification system of preaxial polydactyly in 1969 (Fig. 1).[13] The seven groups are classified based on the level of the bifurcation. Starting from distal to proximal, types I, III, and V refer to bifid phalanges, and types II, IV, and VI refer to complete phalangeal duplications. Type I is a partial duplication of the distal phalanx with a common epiphysis. Type II is a complete duplication of the distal phalanx. Type III is a complete duplication of the distal phalanx on a bifid proximal phalanx. Type IV is a complete duplication of both the distal and proximal phalanges with one metacarpal. Type V is complete duplication of the phalanges on a bifid metacarpal. Type VI is the complete duplication of the phalanges and metacarpal. The exception to the numbering schematic is type VII in which one of the duplicated thumbs is triphalangeal (has three phalanges).

The complexity in the presentation of triphalangeal thumbs necessitated further additions to Wassel's original classification. Wood and Miura created a modification of Wassel type IV to include triphalangeal component into three subtypes.[12,14] Type IV A is complete duplication of triphalangeal thumbs on one metacarpal. Type IV B and C is a duplicated proximal phalanx with a triphalangeal thumb on the ulnar and radial side, respectively. Wood also further divided Wassel type VII duplications into four subtypes.[12] Type A is a triphalangeal ray with the proximal phalanx originating from the metacarpal on the ulnar side. Type B is a complete triphalangeal duplication at the level of the metacarpal. Type C has the triphalangeal component on the radial aspect of the metacarpal. Type D has a central triphalangeal ray with hypoplastic non-triphalangeal duplications on either side (triplication).

Surgical management is dependent on the extent of the bony duplication and soft tissue structures present. The general surgical principle is to remove the least functional component with reconstruction of the remaining parts. The reconstructive surgical algorithm is dependent on the level of the thumb duplication. The timing of the reconstructive process should ideally start before the development of prehension. Most recommend surgical intervention to start shortly after 12 months of age as earlier reconstructions allows for earlier cognitive training of hand functions and potential for remodeling of articular joint surfaces.[12,15-17] On the physiological level, the cardiac status is more stable, the liver is capable of clearing anesthetic agents better, and the persistent humeral artery that prevents a bloodless field under tourniquet has resorbed. Surgical options include ablation of the digit with collateral ligament reconstruction, central resection from each of the duplicated thumbs to recreate a single digit (Bilhaut-Cloquet) and the rarely indicated on-top plasty.

Ablation with Collateral Ligament Reconstruction

Ablation of the residual thumb is the treatment for the majority of thumb duplications. This technique is the preferred method for duplications with a size mismatch, the presence of a larger, dominant thumb, along with a smaller hypoplastic thumb. The principal of ablation is to preserve the dominant thumb, ablate the lesser counterpart, and reconstruct the surrounding soft tissue structures. Often the ulnar-sided thumb is bigger and more developed compared to the radial-sided

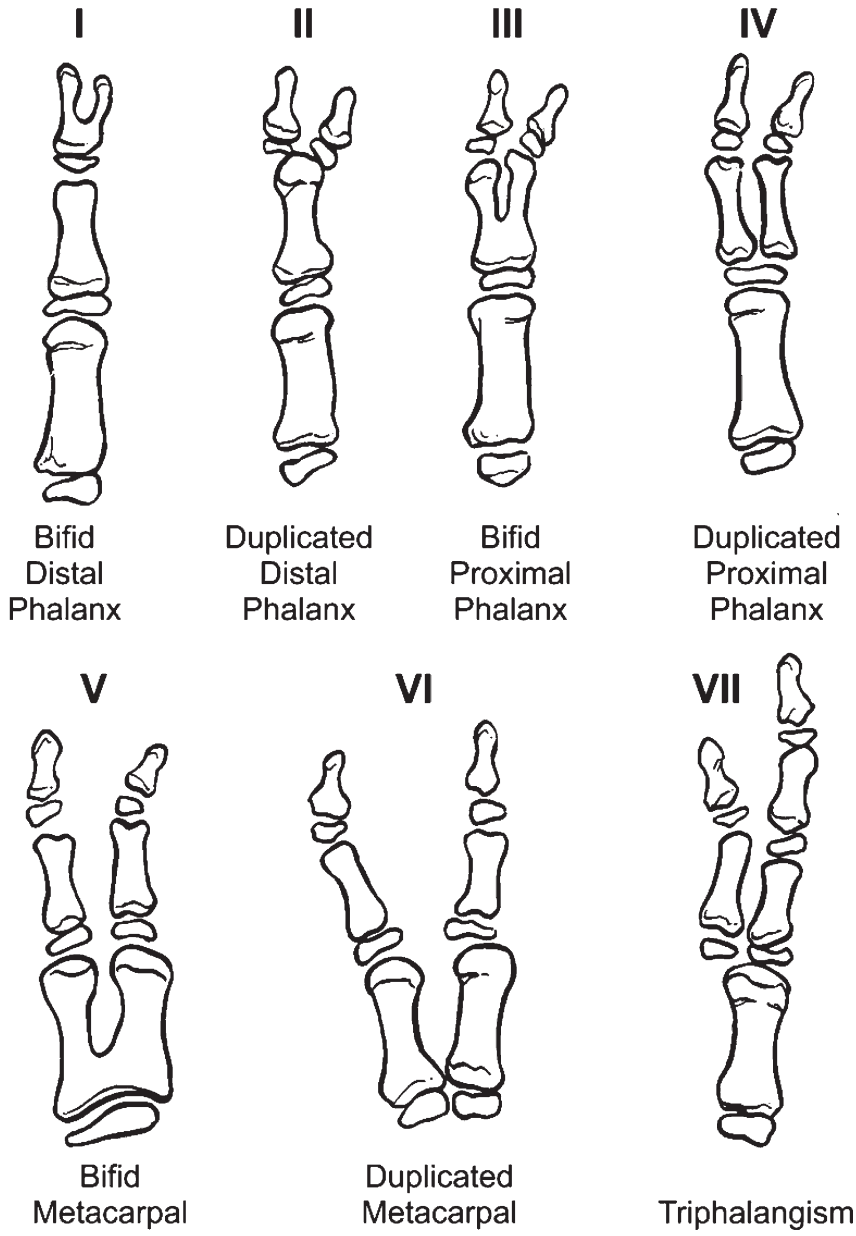

Figure 1 Wassel classification of preaxial polydactyly.

thumb. The preservation of the ulnar component also retains the ulnar collateral ligament in situ, which is important for stability and pinch.[17] Simple ablation of the duplicated digit without additional reconstructive procedures often leads to dysfunction and deformity necessitating revision surgery.[16,18-20]

An incision is made over the duplicated thumb to maximize soft tissue coverage over the retained thumb. The radial collateral ligament is elevated as a longitudinal osteoperiosteal sleeve off of the ablated thumb. This tissue sleeve is preserved and reattached to the radial aspect of the retained proximal phalanx. The adductor pollicis brevis (APB) and flexor pollicis brevis (FPB) are also elevated off its insertion sites and reattached with the radial collateral ligament tissue sleeve. Centralization of the flexor and extensor tendon may be needed to prevent angular deforming forces during range of motion of the digit. Angulation of the phalanx or metacarpal requires corrective wedge osteotomy for maintenance of the longitudinal axis. In type IV, the joint surface of metacarpal head needs to be examined closely for the presence of an additional articulating facet to the ablated digit. This dual articulation adds bulk to the digit. Partial resection is necessary for proper alignment of the phalanx onto the metacarpal head and prevention of growth deformities. The most radial digit is excised, and the remaining soft tissue is used to augment the remaining thumb.

Common complications after surgical ablations include angular deformities, joint instability, and stiffness. The reoperation rate for secondary deformities is as high as 20 to 25%.[17,21] The incidence of complications increases with

increasing complexity of the deformities being treated.[22] Angular deformities at the interphalangeal (IP) and metacarpophalangeal (MCP) can occur in isolation or as a Z-type deformity. The latter is characterized by ulnar deviation at the MP joint and radial deviation at the IP joint.[14] Aberrant anatomic insertion of the flexor pollicis longus has been postulated to contribute to the development of the Z-deformity.[19]

Tada and colleagues reported the largest longitudinal review of thumb duplications in 1983.[16] Tada created a scoring scale rating, which assigned points for the range of motion of the IP and MCP joint, the presence of instability, and mal-alignment. A total score greater than 4 was a good result, a score of 2 or 3 was fair, and a score of 1 or less was poor. Out of the 112 patients who underwent primary ablation with reconstruction, 75.7% had good outcomes, 20.2% had fair outcomes, and 4.3% had poor outcomes. He stressed the importance of appropriate initial surgery to prevent secondary deformity and poor outcomes. In the group of patients presented with residual deformity after previous surgical treatment, 74% (75 out of 105 hands) had been treated when the patients were less than 5 months old (avg. 2.8 months). Tada stresses if the surgery is undertaken too early, abnormal anatomical structures are hard to identify and appropriate reconstruction may not be achieved, resulting in postoperative deformity. Other subsequent studies have shown even with collateral ligament reconstructions and soft tissue balancing, residual deformity may still be present. Townsend and coworkers had 33 patients whom underwent ablation and reconstruction. Angulation or laxity of the IP or MCP joint, greater than 15° or 30°, respectively, and thumb girth less than one third of normal or length less than 80% normal was considered unacceptable results. Over 40% had unacceptable results, but out of the 14 patients, only 1 had significant enough deformity to proceed with a secondary surgery.[18] A significant part of Townsend's criteria is based on the post-surgical cosmesis of the thumb. Since the remaining ulnar duplication of the thumb is rarely of similar size and quality of a non-duplicated thumb, the overall function of the thumb may be a better determinant of successful reconstructive surgery. The most recent longitudinal study by Larsen and associates in 2005 reviewed 19 patients out of 74 treated with reconstruction. The average follow-up was 22 years. Using the Tada criteria, only 26% of the patients had good results. One-third of treated thumbs had a decrease of pinch strength up to 30% when compared to the normal contralateral side. Fifty-seven percent of thumbs also had instability at the joints even with collateral ligament reconstruction. But in subjective functional and cosmetic outcomes, 18 out of 19 were satisfied with their functional outcome, and over 60% were happy with the appearance of the thumb.[23]

Bilhaut-Cloquet

The Bilhaut-Cloquet procedure was first described by Bilhaut (1889) where he combined two equal parts of a Wassel type I duplication to reconstruct a thumb.[24] The concept is a central wedge resection of the duplication and coaptation of the remaining bone and soft tissue to create a new thumb. The theoretical advantage is the ability to create a thumb that is similar in size to the contralateral normal thumb. In reality, the thumb created can be of near-normal size, but it is impossible to create a normal thumb from the duplicated segments. Classically, this technique is used in more distal duplications, especially type I and II deformities. Adaptations have been made to expand the indications to include more proximal duplications (type III and IV).[25]

A zigzag incision is made over the volar and dorsal surface of the skin. If one thumb has a nail of greater than 70% of the contralateral side, the thumb should be used in its entirety.[26,27] Otherwise, the two nail beds should be split longitudinally and combined to a form a new nail of similar size compared to the contralateral thumb. In type I and II, the distal phalanx is split longitudinally and a central wedge is excised. In type III and IV, both the distal and proximal phalanxes undergo a longitudinal osteotomy, and the required width of bone is excised. If the physis is included in the osteotomy, when the bone pieces are matched together, careful attention must be made to ensure physeal matching as well. This step is necessary to diminish the possibility of growth disturbances from an irregular physis. The flexor and extensor tendon insertions must be maintained to prevent angular deformity. The osteotomies are held in place with Kirschner wires, which are removed after healing of the soft tissue and bone components.

The advantages of the Bilhaut-Cloquet procedure over ablation with reconstruction are two-fold. First, this procedure allows for the recreation of a thumb of near-normal size from two hypoplastic components since precise amount can be taken from each thumb for reconstruction. Second, since the collateral ligaments are not violated during the central wedge resection, stability of the IP and MCP joints is maintained, which is important in pinch and grip functions.

Bilhaut-Cloquet is technically difficult and has several disadvantages specific to this technique. Due to the longitudinal osteotomy of the phalanx, one must align the physis, articular surface, and nail matrix simultaneously. It is rare that the two components are identical and mismatch often occurs. Nail plate deformity and ridging arises from the mismatch during the coaptation of the two halves. Therefore, reconstruction using a nail in its entirety if it is greater than 70% of normal has been advocated. However, this still results in a smaller nail plate and is not applicable to every reconstruction. Mismatch of the physis may cause a tethering physeal bar, and this iatrogenic epiphysiodesis can stop longitudinal growth, resulting in a shortened thumb. Physeal mismatch can result in asymmetric growth and angulation of the thumb. Tonkin and coworkers in a series of five patients found postoperative IP joint flexion was significantly limited to 13° compared to 68° on the non-operative side.[27] The MCP flexion was also moderately limited, 55° compared to the

75° on the non-operative side. Samson and colleagues noted in 11 patients the postoperative range of motion of the IP averaged 30° to 45°.[28] Stiffness of the digit, however, may be more preferable than joint instability and laxity.

On-top Plasty

On-top plasty has a very limited role in the treatment of preaxial polydactyly. Triphalangeal thumbs are best indicated for this procedure. This procedure involves the transfer of a suitable distal portion of a thumb to the metacarpal of another thumb, which has a good carpometacarpal joint.[9] This allows the reconstruction of a thumb as close to normal length and size as the contralateral thumb. This procedure is technically very difficult and rarely indicated in the treatment of thumb duplication.

Postaxial Polydactyly

Postaxial polydactyly are duplications of the small finger of the hand. The presentation spans from the well-recognized skin tag and nubbin along the ulnar border of the hand to a fully duplicated digit. The true prevalence may be underestimated because the common nubbin is often treated by ligation without the intervention of a hand surgeon.

Postaxial polydactyly is 10 times more common in the African and African American population compared to their Caucasian counterparts. The frequency is reported to be one in 143 live births in African Americans, compared to one in 1,339 live births in Caucasians.[29] Isolated presentation of postaxial polydactyly presents with an autosomal dominant inheritance pattern with variable expression. When present in Caucasians, post-axial polydactyly is associated with an autosomal recessive transmission and syndromic conditions such as Ellis-van Creveld and chondroectodermal dysplasia.[30] Bilateral duplication is most common, with unilateral presentation occurring most often in the left hand.[31]

Two separate classifications have been proposed to describe post-axial polydactyly. Stelling and Turek subdivide into three groups based on the morphological appearance. Type I is a soft tissue mass attached with a skin bridge, type II is a partial duplication, and type III is a complete duplication of the metacarpal and phalanges.[32,33] The most commonly used classification system by Temtamy and McKusik was derived from study of pedigrees, which showed two distinct phenotypical and genotypical varieties.[31] Type A is a well-developed digit that has an articulation with the fifth metacarpal. Type B is the more common skin tag or a pedunculated soft tissue devoid of any bone, tendon, or nail.[34]

Surgical treatment is based on the subtype of the involved digit. The pedunculated soft tissue remnant in type B polydactyly can be treated with surgical excision or with suture ligation. The latter procedure is often performed in the immediate post-natal period by obstetricians or pediatricians. Suture ligation disrupts the vascular supply to the soft tissue remnant. Dry gangrene develops, and auto-amputation of the rudimentary digit completes the process. Even with successful ablation of the soft tissue, a residual bump may remain at the base requiring revision amputation. Watson and associates reviewed 28 fingers treated with suture ligation and found 40% had a residual bump with an average diameter of 2 mm.[29] No complications of infection or bleeding occurred. In another case series, Rayan and coworkers found 16% had residual or tender bumps, 6% had an infection, and 1% had a bleeding complication.[31] Suture ligation is best performed on rudimentary digits with a thin stalk and with attention to the placement of the suture at the base of the digit. If the rudimentary digit has a broad base, surgical excision may be the better treatment option. With appropriate indications, the ligation technique is safe with minimal complications. If revision surgery or primary excision is necessary, elective excision should be performed after 6 to 12 months of age.[29] From the medical standpoint, the patient is able to tolerate the physiologic stress of surgery and anesthesia.

Type A postaxial polydactyly is more analogous to preaxial polydactyly with regards to surgical technique. The goal is to maintain maximal functional capability after resection of duplicated soft tissue and bony elements. Surgical treatment depends on the level of duplication and the quality of the soft tissues. Usually the ulnar-most digit is the least developed and is excised. For partial duplications, the ulnar collateral ligament (UCL) along with the insertion of the abductor digiti minimi (ADM) should be elevated off the base of the proximal phalanx of the rudimentary digit and reattached to remaining digit. Partial resection of the metacarpal head may be necessary to contour the remaining proximal phalanx. In complete duplications, if the extra digit is functional, surgical intervention may be unnecessary. Otherwise, during the excision of the accessory metacarpal, intrinsic muscle transfers should be performed along with the UCL and ADM reconstruction. To our knowledge, there is no long-term follow-up for type A postaxial polydactyly post-surgery. This may be due to the limited impact a duplicated small finger has on the overall functionality of the hand.

Central Polydactyly

Central polydactyly is a duplication of a non-border digit in the hand. This anomaly is less prevalent than radial or ulnar polydactyly. The most common duplication is seen in the ring finger, followed by the middle finger, and is rarely seen in the index finger. Although complete duplication of an entire digit can occur, central polydactyly is often hidden within a syndactyly and referred to as synpolydactyly. Hence, radiographic evaluations of patients with syndactyly are important in identifying these variations of central polydactyly.

Wood and coworkers reviewed 144 patients with polydactyly and found 22 (15%) patients with duplication of the middle and ring finger.[35] Of the 22 patients, 20 (91%) had associated syndactyly. The review also noted bilateral involvement in 59% of cases, along with 36% incidence

of foot deformities and 55% incidence of family history, suggesting an autosomal dominant transmission. Although Wood noted a high incidence of hereditary predilection, Tada and associates in a review of 12 cases had no patients with any family history.

Stelling and Turek proposed a classification system based on the extent of the soft tissue and bony duplication.[32,33] Type I is a soft tissue mass duplication with no osseous connection to the hand and is devoid of bone, cartilage, or tendon. Type II is a duplication of a digit with normal components and articulates with a bifid or broad-based metacarpal or phalanx. Type III consists of a complete, well-formed duplicated digit with its own metacarpal and soft tissues. Further modification of this classification system by Tada and associates divides type II based on the presence (type IIa) or absence (type IIb) of syndactyly.[36] The most common presentation is the type II deformity.

Surgical treatment is based on the principles of creating a cosmetically pleasing extremity while maintaining maximal function of the hand. The goals are individualized to each hand and its deformity. Intervention is best performed starting at 6 to 12 months of age, prior to the development of angular deformity and contractures due to tethering of structures during growth. Reconstruction is further complicated by the presence of hypoplastic joints and soft tissue changes, which predispose the digits to contractures as well as angular deformities.[36]

Wood and coworkers reported on the largest series of central polydactyly cases.[35] Out of 22 patients, 20 patients (91%) had type II central polydactyly. Of the 15 type II patients whom underwent surgical reconstruction, a total of 90 surgical procedures were performed. Outcome ratings were based on subjective functional assessment and residual scarring. Only six (40%) of patients had a good outcome. The remaining nine (60%) patients had fair or poor outcomes. These patients had functional impairment along with web space and flexion contractures. No patient had an excellent outcome. Poor surgical outcome was also seen in the 10 patients who underwent reconstruction in the series by Tada and associates.[36] Due to the importance of a five-fingered hand in Japanese culture, all subjects underwent procedures to create five digits. The majority of the patients had worsening of the deviation and contractures of the affected digits after surgical intervention. Adjacent fingers had no improvement in appearance or function. Recreation of a five-digit hand requires a multiplicity of surgical procedures resulting in variable and often poor functional results. In order to create a functional hand, both Wood and Tada advocate the ablation of the hypoplastic digit, creation of three good digits with an opposable thumb as the preferable route in the reconstructive surgery algorithm.[35,36]

Ulnar Dimelia

Ulnar dimelia, is an exceedingly rare congenital anomaly of the upper limb. It is characterized by the symmetric duplication of the ulna and the ulnar hand elements around a midline axis producing seven or eight digits in mirrored symmetry. Radial elements, including the radius, thumb, scaphoid, and trapezium, are absent. Neurovascular structures are also duplicated, with variable presence soft tissue anatomy.[37] Clinically, the wrist is radially deviated and flexed because the pre-axial ulna is often shorter along with a lack of wrist and finger extensor tendons. Loss of normal joint articulations due to the presence of two ulnas causes significant stiffness and limited range of motion at the wrist, forearm, and elbow.

Although less than 100 cases have been reported in the literature, many variants of ulnar dimelia have been described. Quattan and Al-Thunayan proposed classification system based on the concept ulnar dimelia is part of a spectrum of duplication disorders of the forearm.[38] It is stratified based on the type of forearm bones present and whether other congenital anomalies are present. Type 1 is the classically described ulna dimelia. Type 2 has two ulnas and one radius. Type 3 has the normal complement of one radius and ulna. Type 4 consists of the syndromic form, which is characterized by bilateral mirror hand, mirror feet, and nasal defects. Type 5 has a normal forearm with multiple hands. The most common variation is type 1. All other types are exceedingly rare and usually encountered as case reports.[39,40]

The embryology of hand development helps explain the etiology of this anomaly. At the distal end of the nascent limb, the apical ectodermal ridge (AER) overlies mesenchymal tissue called the progress zone. The zone of polarizing activity (ZPA) is located at the posterior aspect of the progress zone and secretes Sonic hedgehog (SHH). Radioulnar orientation and differentiation is SHH gradient dependent with ulnar structures developing in areas of high SHH concentration. Duplication of the limb is seen when the ZPA is transplanted to the anterior aspect of the progress zone.[41] Abnormalities within this pathway are thought to be the etiology of mirror hand.

Treatment is directed towards achieving a functional limb with an acceptable cosmesis. Surgical goals include thumb reconstruction and selective ablation of supranumery digits. During thumb reconstruction, balancing of intrinsic muscles and tendon transfers are necessary for preserving thumb function. Adequate first web space reconstruction is critical to allow for excursion of the new thumb. Principles of pollicization are used during the one-stage thumb reconstruction. Deformities at the wrist and elbow may need to be addressed with further surgical procedures, including excision of the proximal end of the ulna.[39]

Summary

Polydactyly of the hand is a complex problem and a unique challenge for the hand surgeon. Understanding of the molecular basis of limb formation underscores the complexity of the molecular signaling cascade needed for the proper orientation and development of the limb. Surgical treatment

needs to address the aesthetic and functional aspect of hand reconstruction. Careful consideration and planning of surgical treatment individualized to each patient is required to obtain the best possible outcome.

Disclosure Statement

None of the authors have a financial or proprietary interest in the subject matter or materials discussed, including, but not limited to, employment, consultancies, stock ownership, honoraria, and paid expert testimony.

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