Review

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Symbrachydactyly

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Abstract

Background: Symbrachydactyly is a unilateral congenital hand malformation characterized by failure of formation of fingers and the presence of rudimentary digit nubbins. The management is variable and are investigated in this review. **Methods:** A detailed review of the literature was compiled into succinct clinically relevant categories. **Results:** Etiology, classification, non-surgical management, surgical intervention, and patient oriented outcomes are discussed. **Conclusions:** All interventions should prioritize realistic, evidence-supported appearance and functional gains. Studies of the baseline function and quality of life of children with symbrachydactyly would allow surgeons to better understand functional changes associated with various interventions and would help surgeons and parents to make the best treatment decisions.

Keywords: congenital hand, symbrachydactyly, Poland syndrome

Introduction

Symbrachydactyly is a unilateral hand malformation characterized by failure of formation of fingers and presence of rudimentary nubbins that include elements of nail plate, bone, and cartilage. Typically, the central digits are absent and the border digits are relatively spared, and syndactyly may be present[27] (Figure 1).

The incidence of symbrachydactyly is approximately 0.6/10 000 live births. It is usually isolated, but can be associated with Poland syndrome, in which hypoplasia or absence of the pectoralis major occurs with additional variable abnormalities[13] (Figure 2). In fact, Poland first described symbrachydactyly in 1841.[51]

Etiology

The etiology of symbrachydactyly is unknown, but vascular dysgenesis during fetal development ("subclavian artery supply disruption sequence") is a leading hypothesis.[2] Based on this hypothesis, isolated transverse terminal limb deficiencies are associated with interruption of the subclavian artery distal to the internal thoracic artery before a gestational age of 42 days, leading to a failure of outward limb growth and interdigital tissue degeneration. In support of this theory, a study of 8 patients with Poland syndrome showed decreased blood flow velocity in affected limbs, likely from a subclavian malformation.[5]

Based on the current understanding of upper limb development, symbrachydactyly likely arises through disruption of the apical ectodermal ridge (AER) of the developing limb bud. The AER, a thickening of ectodermal cells at the distal end of the limb bud, directs proximal-distal limb development through a complex cascade of growth factors and genetic signaling, while controlling aspects of mesenchyme cell differentiation.[15] In animal models, disruption of the AER and its signaling pathways causes transverse deficiencies, including symbrachydactyly.[60,66]

Although limb development occurs in a proximal-distal direction, there may be some regenerative capacity of distal limb elements after a partial or complete insult to the AER that may result in the characteristic "nubbins" or rudimentary digits seen in symbrachydactyly.[23]

Clinical Presentation and Differential Diagnosis

The term *symbrachydactyly* has been used to describe hand malformations that overlap with transverse deficiency, central deficiency, brachymetacarpia, brachyphalangism, and hypodactyly.[38] Symbrachydactyly was previously called "atypical cleft hand" due to morphological similarities with central deficiency,[16] but this terminology has been abandoned[41]; central deficiency is an autosomal dominant condition in which the central rays are absent; it is usually bilateral and often associated with cleft feet. Other conditions in the differential

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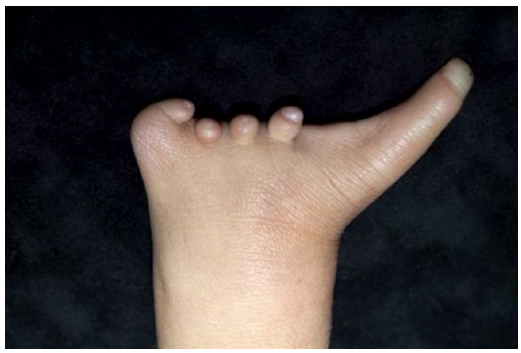

Figure 1. A typical hand with symbrachydactyly.

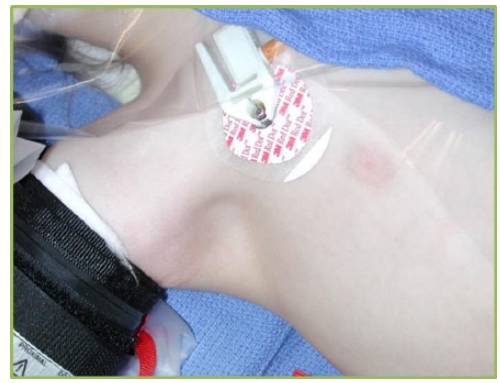

Figure 2. The chest in Poland syndrome, with hypoplasia of the pectoralis major muscle.

Table 1. Differential Diagnosis of Symbrachydactyly.

	Symbrachydactyly	Apert syndrome	Amniotic constriction band	Central deficiency	Ulnar longitudinal deficiency	Hypodactyly
Origin	Sporadic	Mutation, FGFR2	Sporadic	Heritable, multiple loci	Generally sporadic, occasionally syndromic	Sporadic
Upper extremities	Unilateral	Bilateral, complex syndactyly	Usually bilateral, unilateral is rare	Generally bilateral	Unilateral	Unilateral
Lower extremities	Not affected	Affected	Affected (constriction bands and talipes equinovarus)	Affected	Not affected	Not affected
Thumb involvement	Least likely digit to be involved (peromelic and monodactylous forms)	Commonly involved in complex syndactyly	Equally likely to be involved as other digits	First web space commonly shallow/ narrow. Thumb rarely suppressed	Least likely digit to be involved, but can be affected	Least likely digit to be involved, but can be affected
Shape of hand defect	U-shaped cleft	Cup like	Amputations common distal to bands	V-shaped cleft	Missing ulnar-sided structures, smooth contours on hand	Missing terminal elements only, deficiency may be more severe at ulnar digits
Unique features	Rudimentary nubbins with ectodermal tissue. Metacarpals present	Facial features (craniosynostosis, acrocephaly)	Visible scarring/ depression from amniotic band	Absent metacarpals	Proximal structures (ie, elbow) more likely affected than wrist	Multiple shortened digits without terminal ectodermal elements

diagnosis of symbrachydactyly include Apert syndrome, amniotic constriction bands, ulnar longitudinal deficiency, and hypodactyly[38] (Table 1).

Confusion surrounding the definition of symbrachydactyly is due to variability in clinic presentation, including the amount of hypoplasia of the central digits, affected hand size, and the function of border digits. We will use "symbrachydactyly" to describe the unilateral, sporadic presence of shortened or absent central digits with relative sparing of the border digits.

Classification

The International Federation of Societies for Surgery of the Hand (IFSSH) has recently adopted the Oberg, Manske, and Tonkin (OMT) classification system,[30] which incorporates

Table 2. Foucher's Classification.

Type	Features	Thumb	Ulnar digit	Interventions
I	All bones and digits present, brachydactyly and syndactyly	Normal	Bones present, brachydactyly or syndactyly	Syndactyly release
IIA	≥2 fingers. Normal thumb, hypoplastic fingers	Normal	Hypoplastic, syndactyly	Nonvascularized toe phalanx transfers, ablation, or stabilization
IIB	Functional border digits, variable central nubbins	Normal	Present, variable hypoplasia and stability	Surgery rarely indicated
IIC	"Spoon hand," thumb conjoined with hypoplastic ulnar digits	Present (± stability)	Hypoplastic, clinodactyly	Variable
IIIA	Monodactyly	Normal	Absent	Vascularized toe-to-hand transfer
IIIB	Monodactyly	Hypoplastic and/or unstable	Absent	Variable, vascularized toe-to-hand transfer, thumb stabilization, thumb lengthening
IVA	Peromelic, wrist mobility	Absent	Absent	
IVB	Peromelic, no wrist mobility	Absent	Absent	Surgery not indicated

current understanding of embryology and molecular biology. Under the previous IFSSH system, symbrachydactyly was categorized as an undergrowth[61]; in the OMT system, symbrachydactyly is categorized as failure of formation of the proximal-distal axis, which can involve the entire upper limb or the hand plate.[63]

Several classifications of symbrachydactyly have been described. Blauth[4] refined Müller's original concepts[47] into a classification system for symbrachydactyly that included 4 phenotypes:

- Short finger type
- Cleft hand type
- Monodactyly type: absence of digits 2 to 5, with the thumb present
- Peromelic type: adactyly with rudimentary nubbins.

Yamauchi and Tanabu[68] described a more elaborate classification of 7 types based on the morphological and radiographic bony deficiency, allowing for precise description of skeletal elements of the affected hand and extremity, but not providing guidance for treatment. Foucher[19] modified the Blauth classification to make it more useful in this regard, based on a series of 117 patients. The presence of a thumb, the stability of joints, and the patient-specific needs were used to recommend surgical treatment (see Table 2 and Figure 3).

Treatment Goals

The goals of treating congenital hand differences are to maximize function, normalize appearance, and help the child and family accept the difference to the extent that it cannot be "normalized." Selection of a specific treatment for symbrachydactyly depends on the clinical and radiological findings, the ability of the child to meet developmental milestones and perform activities of daily living, and the expectations of the child and family regarding the appearance of the hand. Treatment is grounded in the framework of realistic functional goals, with a reasonable expectation that the treatment can achieve that goal.

Nonoperative Management

Function is classified according to the World Health Organization International Classification of Functioning and Disability (http://www.who.int/classifications/icf/en/).[67] Activity, participation, and quality of life may be very close to normal for children with unilateral hand absence.[32] Often the biggest challenge to the child and family is the psychological burden of appearing different to others.[1]

Nonoperative interventions, such as therapy, prostheses, and orthotics, have been used to treat children with symbrachydactyly. An occupational therapist can help children with unilateral malformations master activities of daily life while increasing self-esteem and gaining independence.[39]

Because they cannot provide sensation, prostheses have limited applications for children with unilateral conditions, especially when the affected side has wrist motion and/or at least one sensate digit that can assist the contralateral hand with bimanual activities. Opposition paddles or partial hand prostheses may be helpful for patients with a stable monodactyly because they provide a surface to pinch against (Figure 4). Customized passive hand prostheses may provide relief from unwelcome questions and comments. As technology improves so that components can be made smaller, less expensive, and more durable, myoelectric partial hand prostheses may become useful to children with symbrachydactyly.

Peer groups and hand camps may improve the child's quality of life more than medical or surgical treatment.[44,59] Camps exist for children with various chronic illnesses and

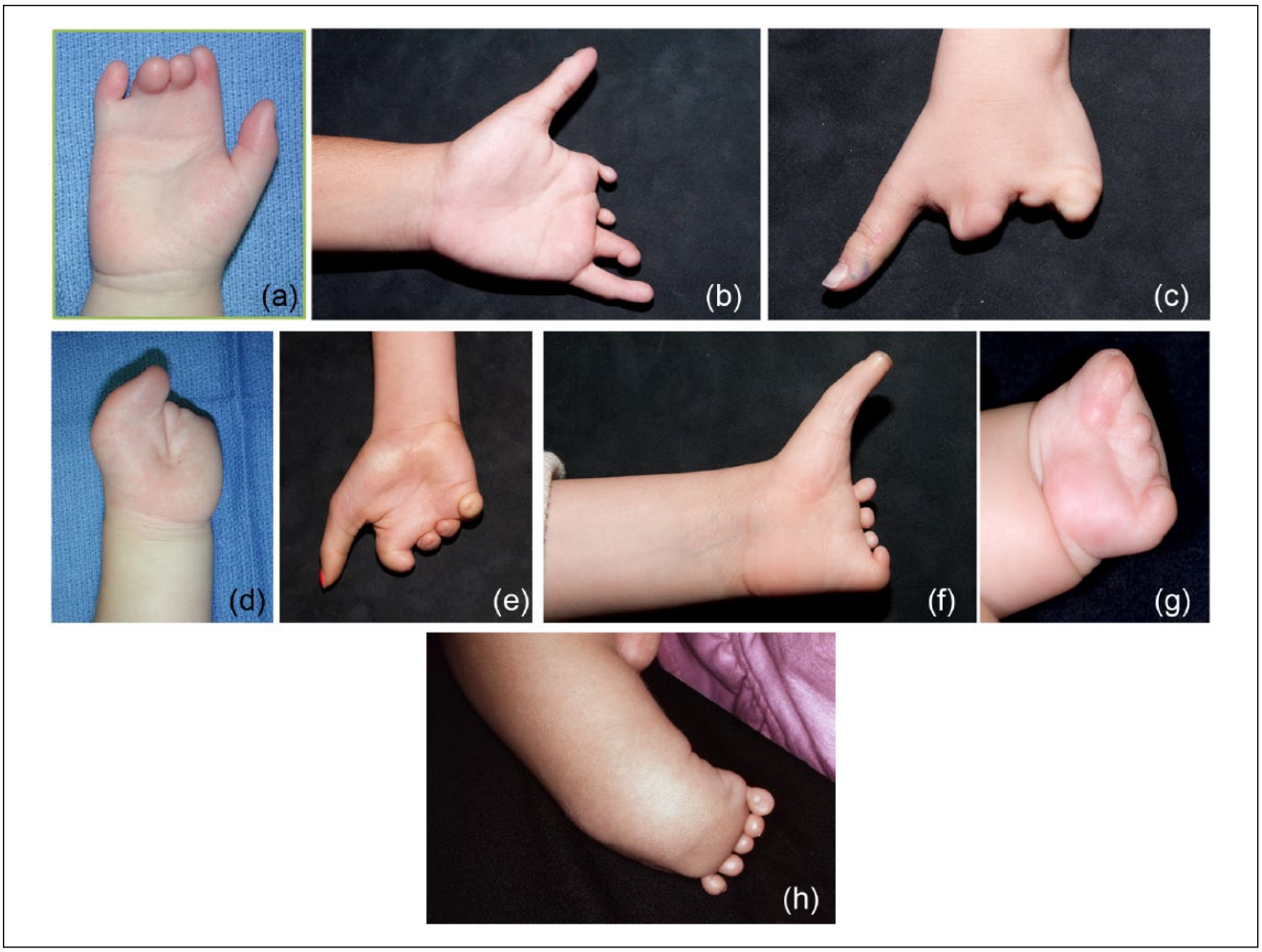

Figure 3. The spectrum of symbrachydactyly as classified by Foucher: (a) type I, (b) type IIA, (c) type IIB, (d) type IIC, (e) type IIIA, (f) type IIIB, (g) type IVA, and (h) type IVB.
Note. A description of each type is listed in Table 2.

congenital conditions; these have been shown to provide short-term psychosocial benefits, including improved social interaction and acceptance.[46,49] Camps for children with congenital hand differences include Camp Winning Hands in Livermore, California (http://www.shrinersinternational.org/Press/Camp_Winning_Hands.aspx) and Hand Camp in Meridian, Texas (http://www.tsrhc.org/camp).

Operative Treatment

Surgical treatments are categorized by the specific aspect of symbrachydactyly that the treatment addresses: syndactyly and web contracture, brachydactyly and digit instability, and lack of opposition.

Syndactyly and Web Contracture

Syndactyly and web contractures are treated to improve independent digital function, grasp span, and appearance. For incomplete simple syndactyly of the digits, 2-fold or 4-fold Z-plasty is usually sufficient[26] (Figure 5). Options for deepening the first web space include multiple Z-plasties, local rotational

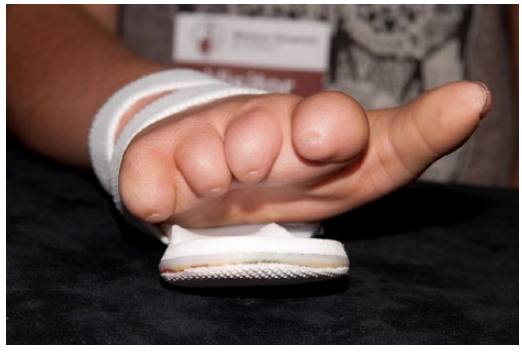

Figure 4. A patient with symbrachydactyly wearing an opposition paddle.

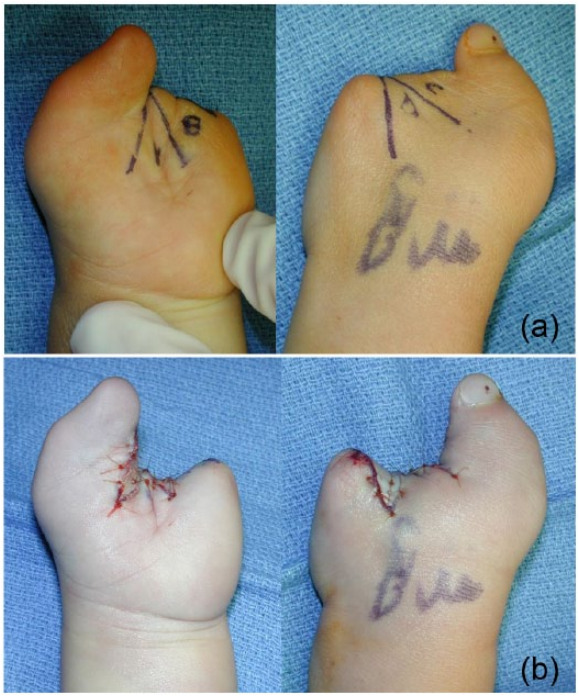

Figure 5. Fourfold Z-plasty to open the web space of the patient with type IIC symbrachydactyly. (a) Preoperative planning of the flaps. (b) Postoperative appearance.

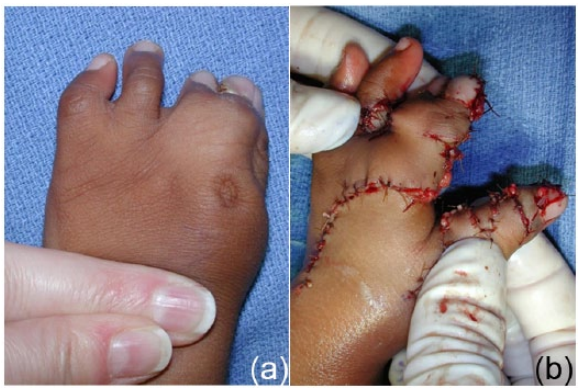

Figure 6. A dorsal rotational flap for more severe deficiency of the first web space. (a) Preoperative and (b) Postoperative.

flaps, and more complex advancement techniques[6,8,10-12,17,21,65] (Figure 6). Release of the first web space in symbrachydactyly can be more challenging than similar releases done for other diagnoses due to a lack of local skin available.[21]

Brachydactyly and Digit Instability

Nonvascularized free toe phalanx transfers. This operation is intended to augment the length and stability of the fingers to improve prehension and appearance. It has been advocated for short metacarpals or phalanges with an adequate distal soft-tissue envelope to receive the transferred phalanx; toe proximal phalanges must be available for transfer[50] (Figure 7). The literature reveals variable results, with longer term follow-up showing more disappointing outcomes.

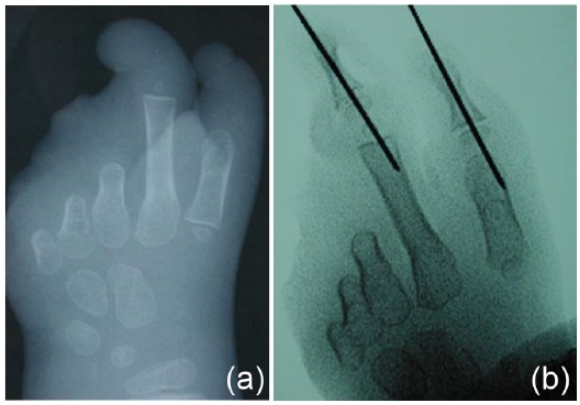

Figure 7. (a) Preoperative and (b) postoperative radiographs of nonvascularized toe phalanx transfers.

The technique includes preserving the volar plate, collateral ligaments, physis, and periosteum of the transferred phalanx, as described by Goldberg.[25] In that series of 15 patients, the authors found that physes were more likely to remain radiographically open in children who were younger at the time of the transfer.[25]

Buck-Gramcko reported similar outcomes in 97 extraperiosteal nonvascularized proximal toe phalanx transfers in 57 children,[7] and Radocha[52] reported that the preservation of the periosteum contributed to the growth of the transferred segment in children younger than 12 months at the time of surgery.

More recently, however, Cavallo[9] studied 64 nonvascularized toe phalanx transfers in 22 children, 18 with symbrachydactyly, and concluded that little longitudinal growth occurred but transverse growth did occur, and contributed stability to the digits. In 11 cases, the transfer was unstable; this complication was most common in patients with symbrachydactyly.

Tonkin[62] reported on 10 children treated with nonvascularized toe phalanx transfers, with a mean follow-up of 7 years. Function testing showed that 5 could use their digit for complex activities, 2 for simple tasks, and 3 for assisting the other hand. Parents reported satisfaction with appearance of the hand and feet, but felt there was no improvement in the hand's function. Most recently, Garagnani[22] studied 40 children with a mean follow-up of 10 years, and found ubiquitous donor site morbidity that increased with growth, along with a high rate of emotional problems with foot appearance and functional problems with footwear.

None of these studies followed children to skeletal maturity and few offered thorough assessments of the function and appearance of the hand following surgery, making evaluation of this procedure difficult. Complete resorption of the transferred phalanx has been observed, as might be expected with a terminal bone graft, and claims of advantages of performing this operation early have not held up. Furthermore, claims of growth of the transferred phalanx are often based on radiographic patency of the physis, but animal studies have shown that radiolucency does not necessarily indicate that a physis is growing.[55] If surgery is postponed until the child is older they can participate in the decision making, the phalanx is larger, and there is less growth potential to lose.

This operation may be useful in type IIA symbrachydactyly, when the base of the proximal phalanx is present along with a generous soft-tissue envelope. The cartilage at the base of the transferred phalanx can be debrided allowing bony healing between the ossific nucleus and the phalanx, reducing the likelihood of instability. When considering this operation, the surgeon should counsel parents and patients to adjust their expectations for cosmetic and functional outcomes, including foot appearance.[33]

Distraction lengthening. Lengthening a bone by distraction osteogenesis with or without secondary intercalary bone grafting is potentially useful for the treatment of short fingers in symbrachydactyly, but the indications for lengthening are unclear.[42] This treatment rarely normalizes appearance and is fraught with complications. The literature shows mixed results with little information to indicate whether this procedure improves function and appearance.

Hulsbergen-Kruger's series of 3 patients with symbrachydactyly indicated that attempts to lengthen hypoplastic bones, including transferred toe proximal phalanges, resulted in pseudarthroses, infections, and resorption with no reported improvement in function.[29] Foucher[20] reviewed results of distraction lengthening in 41 patients (22 with symbrachydactyly) and reported an average gain of 2.3 cm over 4 months. Complications included infection, nonunion, or fracture in 32%. Miyawaki[45] reported 4 successful cases of metacarpal lengthening in patients with types IIA, IIB, and IIIA, noting improved pinch strength with no major complications; others[43] have reported angulation of the lengthened bones, with unsatisfactory appearance. Heo[28] reported a series of 24 metacarpal and 27 phalangeal lengthening procedures with a 31% complication rate, including nonunion, fracture, premature consolidation, angulation, and hardware failure.

Seitz[57] has reported a large series reflecting his long-term experience with distraction lengthening in the arm, forearm, and hand for children with a wide range of conditions. He acknowledges that this treatment is complex and arduous and has a high complication rate (50% minor, 9% major) but reports that in most cases increased length is achieved and the family and child are satisfied.

Given the high rates of complications reported for distraction lengthening and the paucity of evidence to support significant functional gains, we rarely perform this procedure and do not advocate it for symbrachydactyly.

Free vascularized toe-to-hand transfer. Although toe-to-hand transfer is well accepted for the treatment of traumatic amputations in adults and children, its indications for reconstruction of congenital malformations remain more nuanced.

In 1988, Lister[40] described 12 toe-to-hand transfers in children with various congenital hand differences, including 3 cases of symbrachydactyly, and noted unique neurovascular anatomic variations in each patient. Others have reinforced that there is a wide variation in the neurovascular structures in symbrachydactyly.[35,40,53] In 2001, Foucher[18] reported on 51 toe transfers in 45 patients with symbrachydactyly. Transfers to types IIIA and IIIB were the most common, followed by type IVA. In children with type IIIB, a combination of toe transfers to finger positions with vascularized epiphysis and nonvascularized toe phalanx transfer to the thumb was done. They found no functional problems with donor feet. At 5-year follow-up, range of motion (ROM) was adequate, and most participants reported that they used the affected hand in daily activities.

In 2004, Richardson[53] described the results of 18 free toe transfers in 13 patients with symbrachydactyly. The results of a bimanual hand function questionnaire indicated that 61% could lift a cup, 54% could button, 38% could cut paper, and 30% could use a knife and fork; 85% of parents were happy with the appearance of the hand and 77% were happy with the function of the hand. Because of diverse vascular anatomy, the authors recommended obtaining an angiogram preoperatively.

Schenker[56] evaluated grip function after free toe transfer in children with hypoplastic digits, finding that the participants could use the transferred digit to lift a small object with a precision grip, but only one-third modulated their grip in proportion to the load being grasped. They found increased forces on the fingertips of transplanted digits during grasp, and concluded that this was due to misalignment of the finger during the grasp.

Bellew[3] reported 10-year follow-up of 33 children (21 with symbrachydactyly) who had toe-to-hand transfers focusing on psychological outcomes. Children and their parents reported psychological well-being, satisfaction with the appearance of the transferred digit and the donor site, and positive reactions from others, and felt that the transferred digit served a functional role in daily activities.

Kaplan[36] administered the Pediatric Outcome Data Collection Instrument (PODCI) to 15 children who had toe-to-hand transfers and found that scores for self-reports of upper extremity function and transfer/mobility, and parent

reports of global function, upper extremity function, and sports/physical function, were lower than normative scores (they did not compare postoperative with preoperative scores). They also found that parents underestimated sports/physical function and happiness compared with the patient reports; this has been shown for other congenital conditions as well.[58]

Anatomical variation in vascular structures engenders concern about potential effect of these anomalies on the viability of the vascular anastomoses and survival of the transferred toe. A review of toe-to-hand transfers in congenital hand differences, however, found an average transplant survival rate of more than 96%.[34] Other issues include unpredictable range of motion in the transferred digit; as some surgeons have noted, the vascularized toe transfer often results in a sensate post for prehension by another digit.[24,48,64]

Jones[35] proposed a morphologic framework of indications for vascularized toe transfer for thumb and finger reconstruction in congenital conditions, including "complete absence of the thumb and all four fingers" and finger reconstruction when there is "absence of all four fingers but with normal thumb function," which correspond to types IIIA and IVA symbrachydactyly. However, the indications for toe-to-hand transfers are still being established for unilateral symbrachydactyly. Creating opportunities for pinch and grip are important but should be weighed against the risks of surgery, and parental expectations must be aligned with realistic outcomes. Further research is needed to determine whether this operation improves function from baseline and to distinguish any postoperative functional changes from normal child development.

Future Directions

The advent of composite tissue transplantation has led to the reality of hand transplantation for adults with traumatic hand loss. There are many risks to this operation, including the long-term need for immunosuppressive medication; in addition, psychological counseling is imperative beforehand.[14,37,54] These challenges, along with the uncertainty of growth potential, contraindicate this operation in children with unilateral hand conditions. Tissue engineering has the potential to address the challenge of limited tissue availability for reconstructing the congenitally malformed hand. Engineering de novo tendons or augmenting tendon regeneration has the potential to overcome the lack of suitable donor tendons for use in the hand.[69] The creation of composite tissues could someday mean that patient-derived digits could be grown ex vivo and transplanted.[31]

Conclusion

Symbrachydactyly is an uncommon unilateral congenital malformation of the hand in which the fingers are variably hypoplastic. There is no well-accepted classification for symbrachydactyly that guides treatment and provides prognosis, although Foucher's classification may prove useful. The clinical evaluation of the patient with symbrachydactyly should include a multidimensional analysis of the child's functional abilities as well as the appearance of the hand. Thumb function, including prehension (pinch and grasp), is a priority for assessment and reconstruction.

Currently available surgical treatments of symbrachydactyly address hand structure by treating the individual components of the condition: web space contracture, unstable digits, short digits, and inability to pinch. Some surgeons have expanded the indications of free toe-to-hand transfer to provide children with symbrachydactyly with an additional sensate and variably mobile digit. However, given the high level of functioning of children with a unilateral congenital hand difference who have not undergone surgery, and the improved abilities that accrue with normal development, it has not yet been determined whether surgical treatment of this condition improves function and quality of life for the child.

Parents of infants with symbrachydactyly may underestimate their child's potential and request complex procedures such as lengthening and toe-to-hand transfer because they hope that these will make their child's hand "normal." Surgery for this condition cannot achieve normal appearance; the pediatric hand surgeon must help parents reconcile their desires and expectations with their child's potential, by discussing goals and expectations of surgery. Early functional assessments by a pediatric occupational therapist, and peer contacts with older children with similar conditions can help the parents accept their child's condition and make informed decisions about surgery.

All interventions should prioritize realistic, evidence-supported appearance and functional gains. Studies of the baseline function and quality of life of children with symbrachydactyly would allow surgeons to better understand functional changes associated with various interventions and would help surgeons and parents to make the best treatment decisions.

Authors' Note

The content is solely the responsibility of the authors and does not necessarily represent the official views of the National Institutes of Health (NIH).

Ethical Approval

Ethical approval for this study was waived by our institutional review board. There was no study of patients associated with this article.

Statement of Human and Animal Rights

This article does not contain any studies with human or animal subjects.

Statement of Informed Consent

Informed consent was obtained when necessary.

Declaration of Conflicting Interests

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