

The Impact of Microsurgery on Congenital Hand Anomalies Associated with Amniotic Band Syndrome

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Background: Amniotic Band Syndrome is a clinical constellation of congenital anomalies characterized by constricting rings, tissue synechiae and amputation of body parts distal to the constriction bands. Involvement of the hand with loss of multiple digits not only leads to devastating deformities but also loss of functionality.

Methods: In this series, utilizing microvascular transfer of the second toe from both feet, along with local tissue reconfiguration, a tetra-digital hand with simile of normal cascade was reconstructed. A consecutive series of eight children with Amniotic Band Syndrome, younger than two years in age operated on by single surgeon over a twenty five year interval was reviewed.

Results: There was no flap loss. The hands were sensate with effective simple prehensile function.

Conclusion: Application of Microvascular toe-to-hand transfer for well selected, albeit severe hand deformity in Amniotic Band Syndrome is a valid surgical concept. (*Plast Reconstr Surg Glob Open* 2018;6:e1657; doi: 10.1097/GOX.0000000000001657; Published online 4 April 2018.)

EPIDEMIOLOGY AND ETIOLOGY

Amniotic band syndrome (ABS) carries an incidence ranging from 1/1,200 to 1/15,000 live births.¹ Despite its rarity, the American Society for Surgery of the Hand provides ABS its own category for congenital disorders. Hippocrates provided the earliest account of ABS referring to a syndrome of amputated extremities by fetal membranes.^{2,3} Numerous anomalies have been associated with ABS including cleft palate, imperforate anus, equinovarus, and body wall defects.⁴

The etiology of ABS remains controversial.⁵ The intrinsic defect theory purports that inherent abnormalities in the subcutaneous germplasm leads to mesenchymal hypoplasia and scarring; the extrinsic theory espouses that amniotic tissue entangles the fetal parts causing constriction.^{6,7} Despite the debate over the etiology, the timing of injury to the fetus is relatively certain.⁸

EMBRYOLOGY

The upper limb develops between weeks 4 through 8 with the hand plate evident at day 33.⁹ The skeletal maturi-

ty occurs at week 5 with rays of the digit appearing shortly afterwards. Cell apoptosis that follows engenders fingers with deepening of the web spaces. The structures that protrude remain more susceptible to entrapment evident by the more prominently protruding middle digits being the most commonly affected.

CLINICAL MANIFESTATION

The fetal constriction varies in each ABS patient producing a spectrum of clinical manifestations from skin dimpling to digital amputation.^{1,2} Similarly, the severity of neurovascular injury can run the gamut from peripheral nerve palsies, lymphedema, and arterial insufficiency. In general, the presence of syndactyly is common, but it should be noted that acrosyndactyly is pathognomonic of ABS.

CLASSIFICATION

The Patterson classification system endeavors to structure the variability by presenting 4 subtypes based on severity.¹⁰ The first subtype involves a simple constriction ring. The second subtype has a constriction ring that involves

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digit distal to the ring, with or without lymphedema. The third subtype consists of constriction rings with acrosyndactyly. This group is further subdivided into 3 types: type I conjoined fingertips with appropriate web space depth; type II the tips of the digit are joined, but the web space is not adequately developed; type III joined finger tips, sinus tracts between the digits, but no web spaces. The fourth subtype is characterized by amputation at any level.

In the senior author's (D.T.W.C.) opinion, the variability of the ABS hand manifestations can make a rigid classification cumbersome. To be sure, the presence of a constriction ring is 1 of the pathognomic signs of ABS. Yet, the constriction ring may present as a simple, circular indentation of the digit, or in a more complex form producing obstruction of lymphatic flow distal to the ring manifesting as distal lymphedema. In even more severe cases, vascular compromise may be present mandating emergent surgery.

Furthermore, acrosyndactyly is commonly encountered in ABS. Acrosyndactyly by definition is the fusion of the distal part of the digit in the presence of a formed web space proximal to this fusion. The level of fusion varies ranging from distal to the proximal phalanx. The more proximal the level of the fusion, the more difficult it is to identify the presence of a formed web space. For example, when the fusion is at the level of the proximal phalanx, the only evidence of web space is a tiny pit, that barely allows the passage of a safety pin. When acrosyndactyly is present, distal amputation of the adjacent digits has most likely occurred.

In the series of patients presented, all had the following characteristics: (1) multiple constriction rings in the body; (2) in the involved ABS hand, all had acrosyndactyly and conjoinment of multiple digits; and (3) conjoined digits have distal amputation.

SURGICAL MANAGEMENT

The extent of the disease and skeletal growth influence the timing of surgical intervention. Patients with severe edema of the extremity should have release of constriction bands within a few days after birth in an effort to circumvent fibrosis or necrosis. Those with acrosyndactyly should be addressed as soon as the child is cardiovascularly stable, usually between 3 and 6 months. Early intervention permits appropriate bone growth and minimizes secondary deformation.

Imaging modalities can prove beneficial in certain patients. These can include radiographs, ultrasound study, or MRI. Other helpful studies include nerve conduction studies and vascular studies.

SURGICAL TECHNIQUE

The overarching goal in the surgical correction of ABS is to restore the prehensile function of the hand. This implies the establishment of a pinching and grasping unit, inclusive of opposable thumb and fingers, which lie in an anatomic cascade.¹¹⁻¹⁵ This is a multistage undertaking, often requiring 4 stages. The prolonged reconstruction course should be fully conveyed to the patient and family before embarking on this surgical journey.

Stage 1

The first stage consists of release of the constriction bands and separation of the digits having acrosyndactyly. For the acrosyndactyly release, we build upon the principles by Adrian Flatt, namely closing the radial and ulnar sides of the fingers using zig-zag local flaps, covering raw surfaces with full-thickness skin grafts, and releasing a single side of 1 finger at each stage.¹⁶⁻¹⁸ In addition, to minimize web space creep and contracture, the senior author has developed a technique utilizing an acute triangular, dorsal flap that interdigitates with a slightly smaller, isosceles, volar flap to establish a new web space without a scar along the web margin.

Stage 2

The second stage relates to reconstruction of the thumb that ranges from hypoplastic to absent. Regardless, the hand surgeon must be adept to the full spectrum of thumb reconstruction techniques ranging from index pollicization to toe-to-hand transfers. It remains paramount that a normal cascade exists to optimize pinch and grip, the primary prehensile mechanism. The various methods that can be utilized for creation of thumbs using toe transfers are beyond the scope of this review.¹⁹⁻²⁵

Stage 3

The third stage pertains to establishment of a cascade. The characteristic cascade of a human hand consists of a relatively short, but stout thumb opposable to a set of fingers, which are longest in the middle. The distance between the thumb and the adjacent digits is about 4 times of that between each of the fingers.

Albeit, being the shortest, the thumb is the most mobile of the digits, and an adequate lever arm is critical. The thumb, with the combined flexibility of the joints and the length of the phalanges is endowed with a sphere of a reach encompassing the tip of each fingers and the distal one third of the palm. When a thumb is missing and the index ray is partially amputated, an efficacious reconstruction is to transpose the index stump radially to restore the requirement of a joint and critical length for a useful thumb.

If the other fingers are also partially amputated, the missing fingers could be replaced with second to-to-hand transfer. Two fingers could be reconstructed in this manner utilizing the second toe from both feet. When proper care is given to repair the donor-site defect, harvesting of the second toe should incur minimal disfigurement. The senior author (D.T.W.C.) introduced the concept of pollicization of the index stump for thumb reconstruction and utilizing the second toe for recreation of the cascade in ABS. The first such case was recorded and televised in The Learning Channel.

Stage 4

After these 3 stages, most patients need surgical resculpting as they mature. Such revisional procedures include web space deepening, scar revision, and correctional osteotomies for improvement of finger projection.

In short, our surgical technique mandates the creation of a functional, opposable hand unit that allows pinch, and grip in aesthetically appealing manner derived largely from a normal digital cascade.

RESULTS

We retrospectively reviewed the first author's charts from 1992 to 2015 identifying 8 patients who had ABS and complex hand anomalies characterized by multiple digit deficiency and acrosyndactyly. All these patients underwent 2 second toe-to-hand transfers. All who underwent the first 3 stages of reconstruction had this occur before 24 months in age. Stage 4 was completed by skeletal maturity.

Microsurgical technique was used to transfer in total 16 toes to restore prehensile function of the hand in these 8 patients with ABS. There was no loss of the 16 toe transfers with all patients having restoration of the prehensile function of the hand. Each of the patients regained the ability to effectively grasp and pinch with simile of a cascade of a hand.

In terms of motion, the MP joints had full extension and active range of motion of flexion ranging from 40 to 70 degrees. While the reconstructed PIP joints had a PIP flexion contracture ranging from 5 to 15 degrees, active range of motion of flexion ranged from 30 to 60 degrees. The DIP joint was not measured; however, all patients had a stable DIP joint, albeit with an inherent flexion attitude. Furthermore, the ability to pinch restored including 3-point chuck pinch.

We present 2 cases to illustrate our management of congenital hand anomalies associated with ABS contingent on microsurgical techniques. Of note, in 1992, case 1 represents the youngest patient to receive a second toe transfer for the reconstruction of a digit in ABS.

CASE PRESENTATIONS

Case 1

A female who had ABS (Patterson fourth subtype) of her left hand, characterized by amputations of the thumb through ring fingers at the level of the proximal phalanx and syndactyly of the index and long fingers, presented to our clinic. She underwent surgical correction in accordance with the sequence aforementioned. At 7 months, the first stage consisting of syndactyly release between the left long and ring fingers was performed using the principles as discussed above [see **Supplemental Digital Content 1**, which displays Patient 1 (A) Preoperative syndactyly of left long and ring fingers. (B) Postoperative release of syndactyly of left long and ring fingers, <http://links.lww.com/PRSGO/A725>].

The second stage began at 13 months with partial pollicization of the left index finger stump to the left thumb proximal phalanx enabling its lengthening and deepening of the first web space using a z-plasty [see **Supplemental Digital Content 2**, which displays Patient 1 (A) Preoperative marking for pollicization of index. (B) Index pollicization to left thumb for lengthening and web

space deepening with z-plasty, <http://links.lww.com/PRSGO/A726>]. The third stage began at 14 months when the left second toe was transferred using microvascular techniques to the proximal phalanx of the left ring finger [see **Supplemental Digital Content 3**, which displays Patient 1 (A) Schematic demonstrating second toe transfers for recreation of digital cascade. (B) Preoperative marking of left second toe. (C) Second toe harvest with labeling of artery, vein, and tendons. (D) Closure of second toe donor site on foot, <http://links.lww.com/PRSGO/A727>]. The contralateral second toe was harvested at 17 months and transferred to the proximal phalanx of the left middle finger using microvascular techniques [see **Supplemental Digital Content 4**, which displays Patient 1 (A and B) Microanatomical transfer of left second toe to left proximal phalanx of ring finger, <http://links.lww.com/PRSGO/A728>].

As the patient grew, she underwent a revisional surgery from 21 to 41 months to resculpture the left hand including z-plasty of the web space between the small and ring fingers and osteotomies of the index and middle finger at the level of the proximal phalanx to correct improve her digital cascade.

Case 2

A male who had ABS (Patterson fourth subtype) of his left hand, characterized by absence of his index, ring and little fingers and constriction rings of the thumb and long fingers. The first stage commenced at 14 months, where constriction rings of the left thumb and long fingers were released. For the thumb, the dissection was carried out dorsally to the periosteal plane revealing a divided extensor tendon complex over the proximal phalanx. This divided extensor tendon complex was repaired primarily using #4-0 Ti-Cron sutures. Z-plasty flaps were raised based on the radial and ulnar side of the thumb IP joint and transposed to provide skin lengthening sutured in placed with 6-0 plain catgut sutures. Similarly, the left long finger constriction ring was released at the proximal phalanx and closed using z-plasty flaps (dorsal and distally based; and volar and proximally based).

In this case, stage 2 as defined by our algorithm was unnecessary, given the intact thumb. Thus, stage 3 as outlined consisted of utilizing staged second toe transfers of the left and right feet to reconstruct the left little and index fingers, respectively, using microsurgical techniques. The little finger was reconstructed at 17 months and the index finger at 24 months. Of note, psychological assessment of this patient demonstrated full acceptance of the reconstructed hand [see **Supplemental Digital Content 5**, which displays patient 1 (A) pinch and (B) grip at 24 years of age, <http://links.lww.com/PRSGO/A729>]. It should be noted that in both these toe transfers, the usual dorsally based arterial system was so diminutive it coerced reliance on the volar system. It is paramount that surgeon employing toe-transfer understand the volar anatomy of the toe to ensure surgical success.

DISCUSSION

One of the guidelines for reconstructive surgeons is to restore form and function. In terms of form, ABS patients have severe deficiencies, particularly in the type IV class, that can only be enhanced by adding tissue. Microsurgical, heterotopic, osteogenic transplantation to the hand is a logical step to address these deficits.

To enhance this form, we must rely on much of our surgical armamentarium from skeletal fixation to microvascular anastomoses. We have to add tissue in the correct anatomical location to restore prehensile function of the hand. Nothing suits better than a unit part that is dispensable such as the second toe to the hand. Furthermore, it can be motorized to restore function.

Although the etiology of the ABS remains yet to be fully deciphered, we do know that the anatomy proximal to the constriction ring remains retrievable. This is a critical distinction from a hypoplastic hand. Specifically, in ABS, tendons and motor nerves remain available, whereas in hypoplastic hand the presence of these structures is not certain. Therefore, congenital hand anomalies in ABS is amenable for reconstruction by a relatively, dispensable, second toe.

Operating at children at this early age, as the senior author expected, there is no difficulty in acceptance of the reconstructed hand. Each of the patients returned to a productive life with no handicap from the reconstructed, albeit less than normal, hand. All patients accepted the reconstructed hand as measured by psychological assessment in their school systems. This justifies undertaking such a reconstruction in children before the age of 2 years, as the plasticity of the brain assists with the acceptance of the hand, restoring both form and function.

Second toe transfers have minimal deformity and no observable functional deficit, even in the pediatric population. One second toe from each foot can be harvested and leave minimum donor-site morbidity. The senior author has never employed a great toe transfer in ABS patients as the functional deficit can be significant. Thus, this is the rationale behind our formulation of harvesting second toe from each foot for hand reconstruction of ABS patients.

CONCLUSIONS

ABS presents in varying degree of severity that demands a hand surgeon comfortable with skeletal fixation, soft-tissue manipulation, and microsurgical proficiency to restore these patients with a functionally, aesthetically appealing, opposable hand unit.

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