Amniotic Constriction Bands: Secondary Deformities and Their Treatments

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Abstract

Background: The purpose of this study was to report the surgical treatment experience of patients with amniotic constriction bands (ACB) over a 35-year interval and detail consequential limb deformities with emphasis on hands and upper extremities, along with the nature and frequency of their surgical treatment methods. **Methods:** Fifty-one patients were identified; 26 were males and 25 females. The total number of deformities was listed. The total number of operations, individual procedures, and operations plus procedures that were done for each patient and their frequency were recorded. **Results:** The total number of operations was 117, and total number of procedures was 341. More procedures were performed on the upper extremity (85%) than the lower extremity (15%). Including the primary deformity ACB, 16 different hand deformities secondary to ACB were encountered. Sixteen different surgical methods for the upper extremity were utilized; a primary procedure for ACB and secondary reconstructions for all secondary deformities. Average age at the time of the first procedure was 9.3 months. The most common procedures performed, in order of frequency, were excision of ACB plus Z-plasty, release of partial syndactyly, release of fenestrated syndactyly, full-thickness skin grafts, resection of digital bony overgrowth from amputation stumps, and deepening of first and other digital web spaces. **Conclusions:** Many hand and upper extremity deformities secondary to ACB are encountered. Children with ACB may require more than one operation including multiple procedures. Numerous surgical methods of reconstruction for these children's secondary deformities are necessary in addition to the customary primary procedure of excision of ACB and Z-plasty.

Keywords: amniotic, constriction, bands, in utero amputation, deformities

Introduction

Amniotic constriction band (ACB), also known as Streeter dysplasia and over 34 other descriptive terms in the literature,¹² is a congenital nongenetic condition characterized by the development of skin indentations or grooves that vary in depth and circumference. The condition often affects the limbs, but torso, face, and viscera may also be involved. Amniotic constriction bands have a wide variety of presentations at birth.

The first description of ACB was by Montgomery in 1832, but over the following century, no clear consensus was reached about its etiology.⁸ In 1930, Streeter¹⁶ attributed the etiology to a defect in the germ plasma or dysplasia resulting in sheets of epidermis that encircle the limb, resulting in the characteristic ring constriction deformity. The term “Streeter dysplasia” remained in use until the 1960s, when newer evidence supported the etiology to be disruption of the amnion as the source of the embryologic tissue that creates the deformity.^{4,20} Torpin, in 1965, proposed a plausible extrinsic theory, in which failure of obliteration of the outer chorionic space results in rupture of the

amniotic sac and subsequent formation of mesodermal bands of amnion remnants that wrap around and constrict the extremities.²⁰ Patterson¹¹ proposed a classification system for ACB, dividing them into 4 distinct types: Type 1, simple ring constrictions; Type 2, ring constrictions with distal deformity with or without associated lymphedema; Type 3, ring constrictions accompanied by fusion of distal parts; and Type 4, intrauterine amputations. Because of its unique nature, in the 1970s the condition was classified in a category of its own by the International Federation of Societies for Surgery of the Hand.¹⁷ Later, the Oberg, Manske, and Tontin classification divided congenital hand deformities into 3 main categories: malformations, deformations,

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Figure 1. (a) Amniotic constriction bands of the middle 3 fingers with associated distal lymphedema and (b) amniotic constriction bands of the middle 2 fingers.

and dystrophies. ACB were classified as deformations.¹⁹ Due to the wide variety of its clinical presentations, ACB subclassification remains difficult, and most authors continue to describe the lesions by their individual clinical appearance.

There are no reports detailing deformities associated with ACB and their sequelae and the surgical procedures necessary for these secondary deformities. The purpose of this study was to report the surgical experience of patients with ACB treated by the senior author over a 35-year interval and detail secondary limb deformities with emphasis on hand and upper extremity along with the nature and frequency of their surgical treatment.

Materials and Methods

We reviewed the total registry of operative cases for all surgical procedures performed for ACB children and related sequelae performed by the senior author at two institutions. These procedures were done to treat the primary condition, which is the ACB and all its secondary limb deformities spanning a period of 35 years from 1981 to 2016. This included review of recent electronic medical records and operative records from paper charts, scheduling books, and a registry of operative reports. The surgical procedures for ACB were all recorded. Demographic information was collected, including the patient's gender, age at the time of each operation, limb and side affected, and digits involved.

The total number of operations and individual procedures on each patient were also recorded. An operation was defined as a single operative setting, during which one or multiple procedures were performed. A procedure was defined as the individual surgical correction of each specific deformity during a given operation. The total number of operations, total number of individual procedures, and total number of operations and procedures that were done for each patient and their frequency were recorded.

Results

A total of 51 patients were identified to have had upper extremity procedures for correction of deformities caused by ACB. There were 26 males and 25 females. The total number of operations was 117, averaging 2.3 (range, 1-7) per patient. During these 117 operations, 341 individual procedures were performed, averaging 6.7 procedures per patient, with 185 done for the right extremities (upper and lower) and 156 for left. Of the 341 procedures, 290 (85%) were performed on the upper extremity, and 51 (15%) were performed on lower extremities. There were 9 patients who had prior operative procedures at other facilities before their presentation for treatment in our institutions. Twelve patients had only one procedure, while 7 patients had 11 or more procedures (range, 11-19). In the group of patients having their index procedure in our facilities, the average age of patients at the time of the first procedure was 9.3 months. This timing for surgery is earlier than the usual for most congenital hand differences because fenestrated syndactyly, ie, acrosyndactyly procedures were performed in the neonatal period or shortly after.

A total of 16 different deformities of ACB were encountered in the hand, including the primary ACB plus 15 secondary deformities requiring surgery, and these encompassed the following: (1) partial constriction band; (2) complete constriction band (Figures 1a and 1b); (3) in utero amputation (Figures 2a and 2b); (4) fenestrated acrosyndactyly; (5) partial syndactyly; (6) bony overgrowth (Figure 3) of in utero digital amputations; (7) web space deficiency thumb-index and digital (Figure 4 and Supplemental Figure S1); (8) lymphedema (Supplemental Figure S2); (9) remnant of digit/nubbins; (10) ectopic implantation of in utero amputated digits into other parts of the body (Supplemental Figure S3); (11) clinodactyly (Supplemental Figure S4); (12) proximal interphalangeal (PIP) joint contracture (Supplemental Figure S5); (13) nerve constriction (Supplemental Figure S6); (14) complete

Figure 2. (a) and (b) Fenestrated syndactyly and in utero amputation of central 3 digits.

Figure 3. Digital bony overgrowth.

Figure 4. Deficient first web space.

nerve disruption from severe deep band; (15) nail deformity (Supplemental Figure S7); and (16) inclusion cysts adjacent to skin grafted area.

There were 16 different surgical methods of reconstruction done (Table 1). They include, in order of descending frequency, the following:

1. Excision of ACB and Z-plasty: 103 procedures (50 on right and 53 on left extremities). Of these, 73 were performed for the upper extremity; 5 procedures were done at the forearm or wrist level and the remainders were for the hand or digits. Of the total procedures, 30 were performed on the lower extremity; 18 at the leg or ankle level and the remainder in the foot or toes.
2. Release of syndactyly: 82 procedures (43 on right and 39 on left). Of these, 30 were done for fenestrated syndactyly (acrosyndactyly); 27 on fingers and 3 on toes. In addition, 52 procedures were done for partial syndactyly; 46 on hands and 6 on feet. There were no procedures done for a complete syndactyly.
3. Full-thickness skin grafts from the groin: 53 procedures, all performed on the hands primarily for skin shortage after syndactyly release or deepening of web spaces. Full-thickness skin grafts from the groin as the donor site were used exclusively to hand and were utilized in 29 (25%) of operations.
4. Resection of digital bony overgrowth: 32 procedures for in utero amputation stumps of fingertips (17 on right and 15 on left).
5. Deepening of the first web space with flaps and grafts: 21 procedures (12 on right and 9 on left). These were done for thumb-index web space deficiency. Eleven (52%) were done with a pedicle first dorsal metacarpal artery flap, and two utilized a 4-flap Z-plasty. This is in addition to the use of full-thickness skin grafts for the remaining web spaces.
6. Deepening of other digital web spaces: 21 procedures (15 on right and 6 on left). These were done for partial syndactyly. Seven (33%) required a butterfly flap, and 2 required a triangular flap. The remainder were treated with full-thickness skin grafts.
7. Debulking/lymphedema excision: 10 procedures (8 on right and 2 on left).
8. Excision of nubbins: 5 procedures (4 on right and 1 on left hand).

Table 1. Surgical Methods of Reconstruction.

Deformity	Procedure	Number
Amniotic constriction band (ABC) (Figures 1a and 1b)	Excision of ABC/Z-plasty	103
Syndactyly (Figures 2a and 2b)	1. Release partial syndactyly	52
Digital bony overgrowth (Figure 3)	2. Release acrosyndactyly	30
Deficient first web space (Figure 4)	Resection of digital bony overgrowth	32
Deficient digital web space (Supplemental Figure S1)	Deepening of first web space	21
Distal lymphedema (Supplemental Figure S2)	Deepening of digital web space	21
Digital remnant/nubbin	Debulking procedure	10
Ectopic implantation of amputated parts (Supplemental Figure S3)	Excision of nubbins	5
Clinodactyly (Supplemental Figure S4)	Removal of ectopic implantation	3
Deformed nonfunctional digit	Correction of clinodactyly	3
Proximal interphalangeal joint contracture (Supplemental Figure S5)	Amputation of deformed digit	2
Nerve laceration from ACB	Release proximal interphalangeal joint contracture	2
Nerve constriction from ACB (Supplemental Figure S6)	Sural nerve graft	2
Nail deformity (Supplemental Figure S7)	Neurolysis	1
Epidermal inclusion cyst	Nail ablation	2
	Excision of epidermal inclusion cyst	2

9. Removal of ectopic implantation of digits: 3 procedures.¹³ These were done for in utero amputated digits that were implanted in other parts of the body such as the buttocks and legs.
10. Correction of clinodactyly: 3 procedures (all on right hand).
11. Amputation of painful deformed fingertips: 2 procedures (1 on the right, 1 on the left).
12. Release PIP joint contracture: 2 procedures (both on left hands).
13. Sural nerve graft: 2 procedures, 1 for median and 1 ulnar nerve defects (both on left).
14. Nail ablation: 2 procedures (both on the left hand) for fingernail deformities.
15. Excision of inclusion cyst: 2 procedures (both on right hands). These were done for cysts forming adjacent to prior skin grafts.
16. Neurolysis: 1 procedure (on left)¹ for median nerve constriction at the wrist.

Discussion

Amniotic constriction bands can present to the treating surgeon with a variety of secondary hand deformities that need to be addressed. There is no single surgical procedure that

can be offered for all associated secondary deformities to achieve a desired outcome. Prior reports of ACB surgical treatment have focused primarily on management of the main pathologic lesion, namely, the constriction bands caused by the amniotic tissue that encircles the anatomic structures and create strangulation, lymphedematous changes, or in utero amputation.^{5,15,21} However, our study has shown that the primary ACB condition presents with a broad range of secondary deformities, and a great variety of surgical techniques are necessary to address the full spectrum of pathologies encountered and for achieving the best functional outcome for the child.

We encountered 16 different deformities in a variety of combinations, and these required 16 different surgical procedures, a primary for the ACB plus reconstructive for all secondary deformities. As expected, the most common procedure performed in our series was excision of ACB and Z-Plasty, representing 36% of the procedures performed.

Stevenson¹⁵ and Davis,³ in 1946, described the treatment of constriction bands excision and Z-plasty. Upton and Tan²¹ modified this technique to include reconstruction with simple Z-plasty, excising the skin associated with the band and debulking of the surrounding excessive tissues to establish a smooth contour between the proximal and distal aspects of the constricted portion of the affected part. We

utilized the Upton technique of debulking in all of our cases that had associated distal lymphedema, with 10 such procedures. Upton and Tan²¹ also suggested that attention should be given to the full excision of the deep portions farther proximal to the digital bands to reduce the risk of recurrent lymphedema. Tada et al¹⁸ noted that limbs with associated lymphedema may also have cellulitis, but we did not observe any cellulitis in our series.

Moses et al¹⁰ reported 45 patients with ACB and over 15 years follow-up, with emphasis on neural deficits, digital temperature gradients proximal and distal to the constriction bands, and the presence or absence of associated abnormalities. These authors utilized 4 different surgical procedures for the affected limbs, with a total of 126 in 42 patients. Of these, 32 patients underwent staged Z-plasty as their only procedure. In our series, a much broader distribution of associated deformities is described, as well as an expanded listing of the surgical procedures necessary to address them. We did not assess digital temperature gradients in our series.

Miura⁷ reviewed 56 ACB patients and reported 2 procedures performed for treatment of in utero amputations and syndactyly. He did not include specific information regarding the frequency of surgical procedures for each child.

Foulkes and Reinker⁵ described the demographics of 71 patients with ACB treated at their institution over a 70-year period. They reviewed gestational history, associated abnormalities, family history, birth incidence, and number of surgeries, without specifically addressing each deformity and its treatment.

Moses et al¹⁰ noted that after ring constriction deformities, syndactyly was the second most common sequelae noted in their review of 45 patients. Our series has a similar pattern, with 28% of the procedures done to correct syndactyly, and 36% of those procedures specifically done to address a fenestrated acrosyndactyly.

Moore⁹ noted that the syndactyly patterns are often associated with ACB, such as fenestrated syndactyly. Walsh²² noted in his series of 27 patients with acrosyndactyly that when a fenestration is present, often the base of the commissure is located more distally than in the normal hand. In our series, fenestrated acrosyndactyly was treated shortly after birth to allow normal longitudinal growth of the fingers and prevent their angular deformities. We treated the anatomical variation of the web spaces by reconstruction to maximize function and increase the length of the digits.

Bony overgrowth of in utero amputations as the child grows is a result of the normal physal anatomy proximal to the constriction band and restricted soft tissue growth distal to it. This is in contrast to the congenital amputated digit, ie, failure of formation, in which the bone continues to grow proportionally and does not exponentially gain length. Both Miura⁷ and Burgess² confirmed that the metacarpal bone in the involved digit maintains its normal length and shape.

However, Satake et al¹⁴ noted that mild metacarpal shortening can occur in cases of ACB that involve the proximal phalanx, but not the middle phalanx. This finding challenges traditional thinking that metacarpal hypoplasia was only seen in congenital amputations as a result of failure of formation, and not in ACB. Burgess² noted that the proximal phalanx is often shorter than normal proximal to a band or in utero amputation.

This factor, combined with the distal placement of the web seen in associated syndactyly, often necessitates a web space deepening to offer the child increased functional length of the remaining digit. Because of differences in surgical technique necessary to perform web space deepening in digital spaces 2-4 versus the first web space, we counted these separately in our study. Of the procedures in our study, 15% were done for web space deepening, equally divided between the first web space and digital web spaces. Full-thickness skin graft, most often harvested from the groin, was necessary in all cases to surface skin shortage after syndactyly release.

Kessler et al⁶ reported 11 cases of digital lengthening in congenital hand deformities other than ACB and noted that this is a viable option for increasing digital length. Miura⁷ reported 2 patients in his study who underwent thumb lengthening by transfer of a shortened index ray to the thumb. No patients in our study underwent digital lengthening because these patients achieve apparent lengthening offered by deepening of web spaces. Digital lengthening however can be added to the potential list of procedures for ACB.

One of the weaknesses of our study was the variety of medical records used that existed in the decades prior to full implementation of electronic medical records. The retention of long-term medical records seen in countries with nationalized health care systems, along with the ability of the chart to follow children as they move geographically with parents, lends itself to better retrieval of demographic information. In the years following implementation of electronic record keeping, the use of key words by the surgeon in the operative report became essential for maximizing the search capabilities, and we recommend that careful documentation be made of the etiology of congenital hand deformities in the operative report.

Foulkes and Reinker⁵ calculated an average whole body impairment rating of 20% and a mean upper extremity impairment rating of 16% in their patients with ACB, based on their initial anatomic deformity and not on their adult ultimate functional outcome. With the introduction of electronic medical records and the long-term follow-up they provide, the postoperative functional capacity level of these patients is an area that bears recording and further investigation.

Our study emphasizes the importance of recognition by the treating physician of the broad spectrum of the deformities

encountered in ACB and the need for multiple reconstructive procedures to address them. In addition, family counseling is necessary about expectations relevant to the potential development of future secondary deformities and the fact that excision of the ACB may be the first in a multiple staged procedures.

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Ethical Approval

This study was approved by our institutional review board.

Statement of Human and Animal Rights

All procedures followed were in accordance with the ethical standards of the responsible committee on human experimentation (institutional and national) and with the Helsinki Declaration of 1975, as revised in 2008.

Statement of Informed Consent

Informed consent was obtained from all individual participants included in the study.

Declaration of Conflicting Interests

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