CHAPTER **3**

STREETER DYSPLASIA: Review and Case History

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INTRODUCTION

Streeter Dysplasia is a term that represents a complex congenital fetal syndrome characterized mainly by constricting rings about appendages, prenatal amputations of extremities, and acrosyndactyly (1). Although there is a plethora of names for this syndrome throughout the literature (Table 1), it is commonly referred to as amniotic band syndrome (ABS), constriction band syndrome (CRS) or, more recently, the more inclusive congenital constriction ring syndrome (CCRS).

By far the majority of the associated manifestations involve the distal extremities including the digits (Figure 1). The rings may encircle the limb partially or completely and be horizontal or spiral. They can be superficial or deep with the latter causing more lymphatic and vascular embarrassment. As the limb grows, severe constrictures can result in spontaneous intrauterine amputation of the limb or digit (2-7) (Figure 2). Postnatally, the tighter constrictures can lead to distal gangrene and amputation. Less severe bands may cause gross distal swelling and deformity, or distal contractures, ulceration and syndactyly, all of which call for urgent treatment (Figure 3). This article will review the anomalies affecting the extremities, although it is emphasized that the child should also be evaluated for possible associated deformities or other pathologies.

ASSOCIATED ANOMALIES

Besides the asymmetric constriction rings, other associated extremity anomalies include distal atrophy, limb-length discrepancy, anterolateral tibial bowing, hemihypertrophy, tibial pseudarthrosis (8,9), and resistant teratologic clubfoot (10).

Malformations of the upper extremity are two-times as common as lower extremity deformities (11) with incidences reported as high as 90% in ABS cases. (9) Non-limb anomalies include craniofacial clefts from bands about the head and face (12), cardiothoracic and internal organ defects from bands across the body or those swallowed, and renal abnormalities (37% of cases). The bands can also cause scaring on the surface of the skin to which they can attach (Figure 4).

Table 1

TERMINOLOGY FOR STREETER DYSPLASIA

Streeter Dysplasia
Streeter's Dysplasia
Streeter's Syndrome
Constriction Band Syndrome
Annular Band Syndrome
Annular Constrictive Rings
Pseudoainhum
Amniotic Band Sequence
Amniotic Band Syndrome (ABS)
Congenital Constriction Ring Syndrome (CCRS)

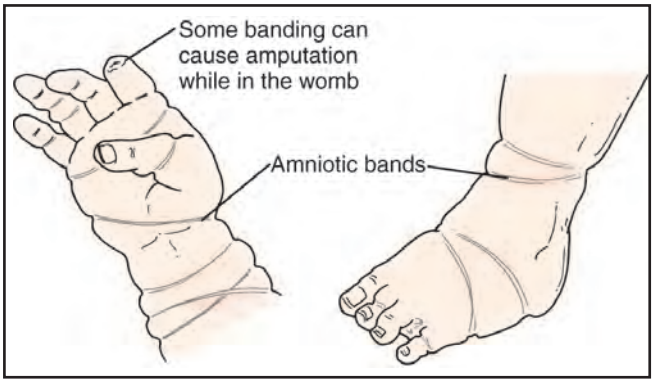

Figure 1. Illustration demonstrating amniotic bands around appendages.

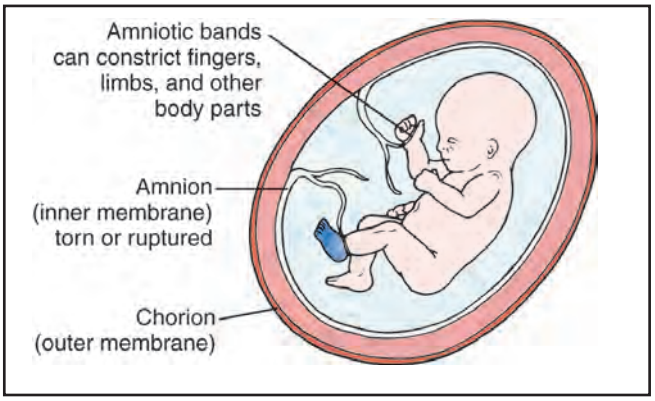

Figure 2. Artist rendition of intrauterine gangrene caused by ABS.

Figure 3. Limb in infant with amniotic band around ankle. Notice absent hallux due to intrauterine autoamputation.

Figure 4. Skin scars on plantar foot from attachment of amniotic bands.

INCIDENCE

Reports range from 1 case per 1,200 to 1 per 15,000 live births. A more accurate epidemiologic study from Atlanta cites the frequency as 1.16 per 10,000 population (13), while the birth defect registry for Western Australia records a 1.15 per 10,000 incidence. Interestingly, approximately 17% of stillbirths are attributed to ABS. There is no sex predilection for ABS and no familial tendencies have been reported (1).

As a stricture disorder, ABS must be differentiated from ainhum (dactylolysis spontanea), an inherited digital stricture syndrome that predominantly affects the fifth toe in African adults between 20 and 50 years of age. In 75% of the cases both feet are affected. Although there is no sex preference, affected women are younger than affected men. The histologic appearance of the constriction ring is very similar to the band tissue in ABS. These ainhum digital bands often ulcerate and can lead to spontaneous amputation of the toe (14-16) (Figure 5).

Figure 5. Classic example of ainhum involving the fifth toe.

ETIOLOGY

ABS was recognized by Hippocrates as early as 300 BC when he suggested that extrinsic pressures caused by a ruptured amniotic membrane lead to the formation of congenital bands or digital amputations. J. B. van Helmont in 1652 attributed intrauterine amputations to pregnant mothers who had seen maimed soldiers. Later, Montgomery in 1832 (17) and Simpson in 1836 (18) evaluated stricture deformities and discussed the differences between agenesis- and amniotic band-induced amputations.

In 1930, George Streeter introduced the intrinsic germ plasm defect theory (a disruptive event during blastogenesis) to explain the etiology, particularly when there are associated anomalies other than the bands (19). Even though this theory faded from support, Streeter stuck to it throughout his illustrious career in embryology and anatomy and was thus rewarded by having the syndrome carry his name. Patterson supported the intrinsic explanation in 1960 when he performed a histologic study of the bands, concluding that they were simply abnormal skin creases that resulted from the same lack of mesodermal development in the area of the strictures (20). His more sustaining contribution was his classification of the extremity deformities in ABS.

Type I – involves simple ring constriction

Type II – involves ring constriction accompanied by fusion of the distal boney parts

Type III – involves ring constrictions accompanied by fusion of soft tissue parts

Type IV – demonstrates intrauterine amputation

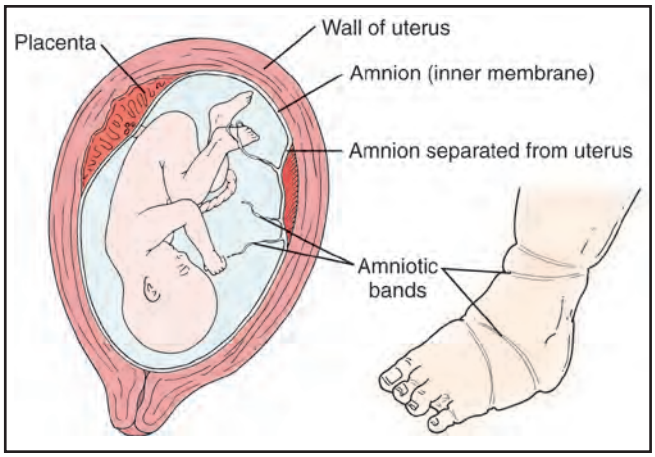

Figure 6. Illustration of ruptured amnion and the release of fiber-like bands that can lead to the constrictions.

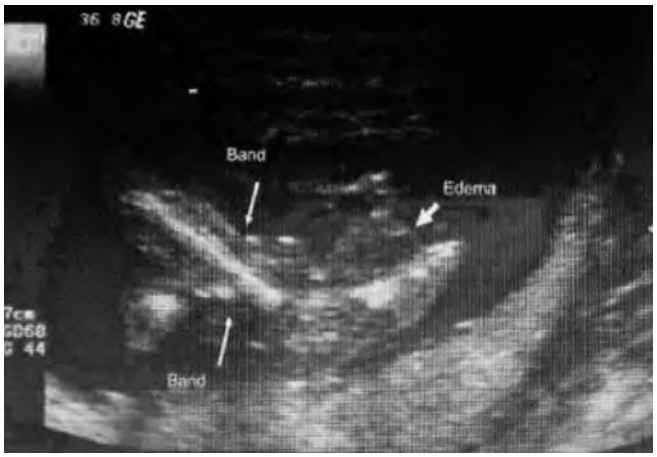

Figure 7. Ultrasound image showing low tibial band and distal edema.

Torpin in 1965 reintroduced the extrinsic concept originally expressed by Hippocrates of maternal trauma to explain the anomalies caused by constricting bands (21). After studying many placentae from newborns affected with ABS and observing the lack of a complete amniotic lining, he proposed that external trauma to the mother during fetal development leads to rupture of the amniotic membrane, forming bands that can encircle digits, limbs, head, neck or even umbilical cord. Bands can also be swallowed causing internal pathology. The delivery of fully developed amputated limbs with these neonates further validates the extrinsic explanation. This is the most accepted theory today.

The pathophysiology involves the interaction between the two membranes that form the cavity surrounding the developing embryo: the amnion (more internal) and the chorion. Up until about the 12th week of gestation there is an extracoelomic space between these membranous structures that is obliterated about this time by the amnion adhering to the chorion to support it. Incomplete obliteration of the extracoelomic space produces a fragile amnion vulnerable to spontaneous or traumatic rupture. The ruptured amnion breaks up into floating bands (6, 22) (Figure 6). Trauma or excessive uterine muscle contractions during pregnancy (such as the mother working) as a cause of ABS was proven by Kino's study in 1975 (23) and Roswell's in 1988 (24).

The rupture also produces a transient oligohydramnios due to the extravasation of amniotic fluid, causing the chorion to close in on the developing fetus, leaving little room within which to move or develop. This may explain the severity and stiffness of the clubfoot deformities associated with ABS.

DIAGNOSIS AND PRENATAL HISTORY

Streeter Dysplasia has been shown to be associated with elevated levels of maternal serum alpha-fetoprotein (MSAFP) but this is also elevated in other prenatal defects so it is by itself not diagnostic of ABS. Serial ultrasounds can help delineate the malformations (Figure 7) or intrauterine amputations while 3D and 4D ultrasound studies can characterize the finer defects in anatomy such as nerves and muscles distal to the extremities (26-28).

Vascular ultrasound studies and magnetic resonance angiography can be utilized to analyze the viability of distal vascular structures and supply. Amniocentesis is contraindicated as it has been associated with needle placement rupture of the amniotic membrane.

Approximately 60% of the mothers delivering ABS babies report having a history of abnormal pregnancies. Besides trauma to the uterus, other prenatal risk factors include low birth weight (<2,500 g), maternal illness during pregnancy, maternal drug exposure (e.g., cocaine, mifepristone), maternal hemorrhage, and attempted abortion in the first trimester (28). In one series of ABS newborns average gestation was 37.5 weeks and average weight was 3.0 kg (11).

Besides the already discussed gross extremity pathology, distal neurological deficits are common secondary to the band compression on the peripheral nerves, especially the peroneal nerve. Vascular embarrassment up to and including impending gangrene or gross distal swelling may be observed, all of which are indications for urgent surgical treatment.

CLUBFOOT ASSOCIATED WITH ABS

The incidence of congenital clubfoot deformity with ABS is 12-58%, and 30% occur bilaterally (10,23,28-33). Two types of clubfoot are recognized: rigid and paralytic. Gomez reported that 80% of the ABS neonates born with clubfoot had a congenital annular constricting ring on the ipsilateral extremity (28). Often, there is band compression on the peroneal nerve causing distal neuropathy that can lead to the paralytic clubfoot due to damage to the evertor muscles (10,32). Thus, the more distal the ipsilateral band along with neurologic deficit, the more severe the clubfoot compared to those without neurological deficit (10).

Generally, these clubfeet respond very poorly to casting. Gomez obtained only 6% success with casting and all but 2% of those clubfeet in his study (n = 43) required surgery (28). These clubfeet are very rigid and challenging to correct. As indicated by their severity, the annular bands are excised prior to the foot surgery. Clubfeet with neurologic deficits had poorer surgical outcomes than those without neurological deficits and the more flexible/reducible the clubfoot, the better the outcome (10).

TREATMENT

No known medical treatment is available for ABS. Surgical treatment for ABS depends on the neurovascular status, amount of distal swelling, and severity of the strictures. Digit or limb amputation should be reserved for situations of impending gangrene; however, it is still a standard of care treatment in many countries. Severe threatening gangrene requires urgent surgery and has been healed by stricture releases (8, 34, 35).

Mild bands with minimal constriction can be simply released but more severe constricting bands have been released in one, two or even three stages (36). These procedures begin with excision of all or part of the band and the implementation of a method of closure. Although the most popular method of closure has been utilizing multiple Z-or W-plasties (37-39), other methods have included direct primary closure (40) saw-tooth approximation (7,41-43), and using triangular flaps (44). Endoscopic in utero release of amniotic bands has been successfully performed on human fetuses (26,45,46) after experimental surgery on in utero lamb fetuses (47).

CASE HISTORY

During a humanitarian medical mission to the Late Atitlan area of Guatemala, two infants with ABS were referred to the volunteer surgical team for evaluation and treatment. Prior to the scheduled screening day, one of the infants was taken to Guatemala City for another opinion and the affected limb was amputated. We did not examine this child.

The second child was a 9-month-old infant girl of Mayan descent from the Lake Atitlan region of Guatemala. This child was brought to our screening clinic with her mother by an American volunteer to see if there was an alternative to amputation. She presented with a classic mid to low left leg annular band constricting the skin and underlying subcutaneous tissue down to the deep fascia (Figure 8). There was no distal edema; the tissues for the length of the leg were found to be pink, warm, and supple with very mobile skin except for the constricting band. Both the dorsalis pedis and posterior tibial pulses were easily palpable.

The first and second toes on the left foot were absent with the foot supple and mobile in a normal position; all the toes on the right foot were absent. The right foot was otherwise a stiff clubfoot in equinovarus deformity (Figure 9).

Figure 8. Left leg in 9 month old infant showing low amniotic band stricture. Note that toes one and two are absent due to intrauterine amputation.

Figure 9. Right clubfoot.

Figure 10. Scars on plantar skin due to amniotic band adhesions.

Figure 11. Hypoplastic fingers 4 and 5.

There were linear and serpentine scars on the sole of the right foot with no history of trauma, obviously the result of amniotic band attachments and resultant scarring (Figure 10).

When placed and held upright with the feet on the supporting surface, the child responded with the extension reflex. There was no evidence of spina bifida or other neurological disorder. No facial defects were evident. Fingers 4 and 5 bilateral were hypoplastic (Figure 11). Her cardiovascular and physical examinations by our team family physician and anesthesiologists were unremarkable.

The diagnosis was made of Streeter Dysplasia or ABS and a surgical plan devised to excise the annular ring and bring together and close the circumferential tissue defect with plastic surgery techniques designed to restore the continuity of the limb without compromising the circulation. A one-stage excision with triangular flap repair was executed without complication (Figure 12). The contour of the leg was restored (Figure 13).

The sutures were removed after three weeks. (Figure 14) At six months follow-up the wounds had healed with little scarification and no distal edema. There was no hourglass residual deformity (Figure 15). The right clubfoot was treated using the Ponseti casting technique and surgical correction, if necessary planned for 12-18 months into the future (Figure 16).

DISCUSSION

When surgically treating amniotic bands on extremities or digits, whether using one or more stages, it is very important to excise the complete width of the stricture. Otherwise, there is a high incidence of recurrence. Several techniques have been reported for repair of the defect. As already noted, Z-plasty or W-plasty have probably been the most common methods in the literature with progression from two or three stage repair to single-stage repair as long as the distal circulation will support the healing (36,48-52). The trouble with using these plastic flap techniques is that they tend to heal with considerable local scar contracture and/or a saw tooth disfigurement (44).

Choulakian and Williams described a two-stage excision with direct approximation at their institution used for decades without complication and a much more cosmetically acceptable scar (40). They did not mobilize or debulk the local subcutaneous tissue as recommended by others (37,49), allowing it to reform naturally and found their patients to heal without any hourglass deformity. Unfortunately, they presented no cohort of patients studied and no statistics.

The procedure chosen for the annular ring excision and repair for our case history patient was via the use of triangular flaps closed primarily in a single stage. Tan and Chiang reported on the success of this procedure in 2011 in eight patients with a mean follow-up of 43 months (40). Besides leaving a minimal scar, another advantage of this procedure is that it addresses the difference in circumference on either side of the stricture due to the conical shape of the leg. For every centimeter difference between the proximal and distal the circumference, a one centimeter based triangular flap is created on the largest side. This can be reversed in cases where there is excessive swelling distally. Only minimal subcutaneous tissue was necessary to maintain contour (37, 40).

In conclusion, the pathophysiology and anomalies associated with Streeter dysplasia, or ABS, have been presented. The majority of the pathology clearly occurs in the extremities and is largely consistent with extrinsic theory of pathogenesis. Surgical treatment is the mainstay of care, sometimes with considerable urgency, and it is evolving towards one-stage repair of the constricting bands and

Figure 12A. Preoperative lateral view.

Figure 12B. Preoperative medial view.

Figure 12C. Marking band and triangle.

Figure 12D. Excising band.

aggressive reconstructions of the associated, rigid clubfeet. One surgeon has reported successful repair of both the ipsilateral stricture and the clubfoot at the same operation (36). The use of a relatively new and simplified single-stage approach using triangular flaps and primary approximation is described with a successful case history illustration.

Figure 12E. Approximation of triangle.

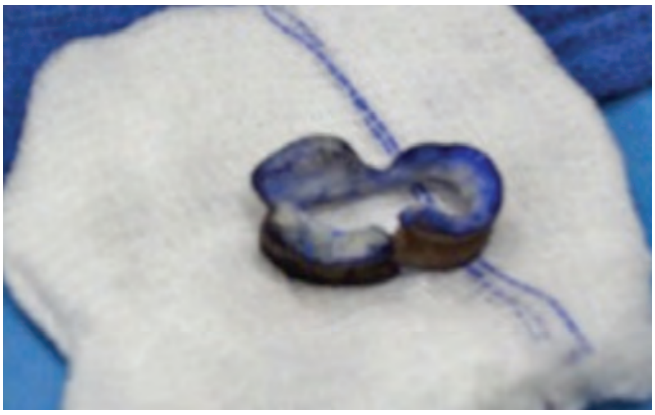

Figure 12F. Excised amniotic band stricture.

Figure 13A. Preoperative congenital stricture.

Figure 13B. Postoperative view after excision and repair of stricture.

Figure 14. Wound at 3 weeks, sutures removed.

Figure 15A. Lateral view at 6 months postoperative.

Figure 15B. Medial view at 6 months postoperative.

Figure 16. Ponseti cast on right clubfoot.

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