

Education-Family Physician Corner

Single Stage Release of Bilateral Amniotic Band Syndrome

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We present a case of amniotic band syndrome, a rare entity, affecting both legs of a newborn. It is the first case managed at our institution. Our approach is a single stage release. At birth, the baby had deep constriction rings around both legs with distal lymphedema which was severe on the right side involving the distal portion of the ring; the vascularity was maintained bilaterally. The right tibia was fractured due to the tight band. Release of both bands was performed on the first day of life with primary closure. At 2 weeks of life, the surgical wounds had healed. At 6 months, it was noticed that the swelling of the right foot increased and the band recurred. Repeat excision was performed and intra-medullary nailing of the tibia was inserted. Amniotic band syndrome has wide implications on the quality of the life of the affected infant. In-utero diagnosis of the condition is important for the management; however, in-utero release is associated with a high incidence of premature birth. A staged release of the band is the most commonly used surgical method though single staged procedures have yielded favorable results.

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Amniotic band syndrome, also known as streeter dysplasia, are complex disorders ranging from constriction bands around the extremities to pre-natal extremity amputation and acrosyndactyly¹⁻⁴. Nearly 50% of the cases have associated anomalies involving the lower limbs more than the upper limb; associated defects include distal atrophy, limb-length discrepancy, hemihypertrophy, tibia pseudoarthrosis and resistant club foot^{2,3}. Though the true incidence of amniotic band syndrome is unknown, it is estimated to affect 1 in 12,000 to 1 in 15,000 live births⁴. Most of the cases are sporadic and no sex predilection or familial tendencies have been observed⁵.

The aim of this report is to present a case of a newborn with amniotic band syndrome affecting both lower limbs, the first case to be reported in the Kingdom of Bahrain. A single stage release of the band was performed.

THE CASE

A routine prenatal ultrasonographic (US) screening was performed on our patient. At 6 months of age, US revealed bilateral lower limbs constriction bands. A discussion regarding the treatment options included the intra-uterine intervention and the risks associated with it. A decision was made to intervene after birth.

The baby was born full term with no other medical illness. He had amniotic bands affecting the lower limbs, the left side was deep reaching to the muscle fascia whereas the constricting ring affecting the right lower limb was deep and tight enough to cause fracture of the tibia in-utero, also causing lymphatic obstruction of the limb distally with the lymphedema involving the distal part of the ring itself, see figure 1. According to Patterson classification, the constriction band was classified as type I for the left leg and type III for the right leg. After discussion with plastic surgery, a decision for single stage



Figure 1: Bilateral Constriction Rings Affecting Both Lower Limbs



Figure 2: Type III Constriction Ring of Right Lower Limb

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release was made due to poor lymphatic drainage at the distal part of the ring on the right side.

Incisions were performed on either side of the band including 2mm of the normal skin to release the fibrous band including the muscle fascia. A similar procedure was performed on the other side, the proximal and distal parts were then approximated using simple absorbable sutures, see figures 2-6.

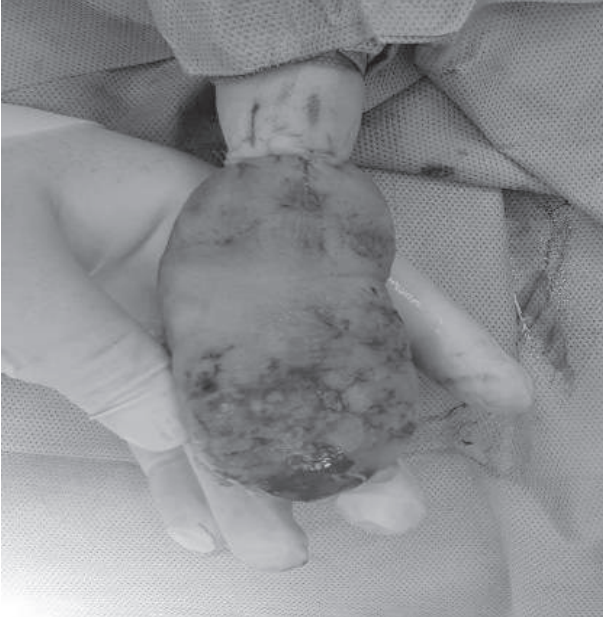


Figure 3: Post-Release of the Right Constriction Ring



Figure 4: Left-Sided Constriction Ring Type I



Figure 5: Intra-Operative Release of Left-Sided Constriction Ring



Figure 6: Post-Release of Left Foot Constriction Ring

The wounds were completely healed in 2 weeks, see figure 7. After 3 months, the left lower limb had completely healed and looked normal; the wounds over the right lower limb had healed and the swelling was persistent over the dorsum of the right foot, but significantly reduced, see figure 8. However, the fracture still had not healed and there was evidence of pseudoarthrosis of the tibia. At 6 months of age, an increase in swelling of the right foot was noticed with deepening of the ring with pseudoarthrosis of the right tibia and fibula. The pseudoarthrosis was excised and intramedullary nailing with revision and excision of the ring. At 7 months follow-up, the wounds bilaterally have healed and the swelling of the right foot considerably reduced, see figure 9. Plain radiographs at 7 months follow-up revealed fracture healing of the right tibia, see figure 10.



Figure 7: Two Weeks Follow-Up Showing Completely Healed Wound Post-Left CCB Release



Figure 8: Three-Month Follow-Up Post-Release of Right CCB



Figure 9 (A)



Figure 9 (B)

Figure 9 (A-B): Seven-Month Follow-Up Image of the Right Foot



Figure 10 (A)



Figure 10 (B)

Figure 10 (A-B): Seven-Months Follow-Up Plain Radiograph of the Right Tibia

DISCUSSION

Amniotic band syndrome is a synonym with congenital constriction band syndrome, annular band⁶. The effects of the band could be broadly classified into limb anomalies and non-limb anomalies¹. Extremity band syndrome involves the lower limb more than the upper limb; these intra-uterine bands may encircle the limb partially or completely. The latter may lead to intra-uterine amputation due to mechanical effects exerted by the band. The constriction bands may either be superficial or deep, depending on the depth of the band; the depth of the band is an important contributory factor that dictates the prognosis⁴. The deep ring causes vascular and lymphatic injury, which might lead to severe lymphedema. On the other hand, superficial rings occasionally might not require any treatment¹.

Patterson classified the constriction bands into four subtypes depending on the depth and the clinical effects caused by the bands¹. Type I refers to a ring constriction that might be superficial or deep. Type II includes constriction band with distal deformity such as lymphedema. Type III includes the presence of acrosyndactyly. Type IV includes complete intrauterine amputation.

Our case was type III for the right leg and type I for the left leg. Classification does not account for various effects that the constriction bands exert dynamically as well as prognosis.

Amniotic band syndrome is responsible for 7-14% of still births⁷. Though the abnormality has been recognized from the time of Hippocrates, the exact etiology of amniotic band syndrome is not known. Various hypotheses have been suggested; the most acceptable was by Torpin in 1965, which was based upon extrinsic concept expressed by Hippocrates¹. He suggested that any uterine trauma to the mother during fetal development may lead to rupture of the amniotic membrane leading to the formation of free bands encircling the various parts of the fetus. Oligohydramnios is one of the associated factors with the rupture of the amniotic membrane¹. The other is the presence of atretic segments in major blood vessels or absence of blood vessels as a cause of constriction band syndrome. Various risk factors have lately been found to be associated with an increase in the incidence of constriction band syndrome, such as maternal smoking, aspirin or living at high altitudes.

Amniotic band syndrome can be recognized in-utero by ultrasound and its early detection is of prime importance for counseling the mother and for possible endoscopic in-utero

treatment of the constriction band^{5,6-9}. There have been reports suggesting successful salvage limbs by timely intervention in-utero. One of the prime indications of endoscopic amniotic band release in-utero is a limb at risk of amputation. In-utero release of the band results in immediate improvement of the distal blood flow resulting in the arrest of strangulation. Any residual constriction may require to be released post-natally. Adverse effect of in-utero release of amniotic band syndrome is premature rupture of membranes, the incidence of which is reported to be 6-10% and has its own implications.

The surgical management of amniotic band has evolved over time but multiple stages Z-plasty or W-plasty are still being used⁸. Other methods include direct primary closure, saw-tooth approximation and the use of triangular flaps¹. Single-staged procedure avoids tethering of the skin, faster rehabilitation and avoids anesthetic complications^{6,9}.

It is important for surgical treatment to aim at complete excision of the band to avoid recurrence. Additional removal of 1-2 mm of normal skin around the indentation may improve the circulation in addition to avoiding recurrence.

CONCLUSION

ABS is rare disorder. Timely recognition and intervention is of prime importance to avoid associated complications. A single-stage treatment provides additional benefits as compared to multi-stage release whilst yielding comparable results.

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