



Case Report

## Amniotic Band Syndrome- A Rare Case Presentation

Kshirsagar Ashok, Vekariya Mayank, Nagur Basavraj, Pednekar Akshay, Gupta Vaibhav, Patankar Ritvij,  
Mahna Abhishek

Krishna Institute of Medical Sciences, Karad, Maharashtra, India 415110.

Corresponding Author: Kshirsagar Ashok

Received: 05/08/2014

Revised: 21/08/2014

Accepted: 22/08/2014

### ABSTRACT

**Introduction:** Amniotic band syndrome is a rare condition caused by strands of the amniotic sac that separate and entangle digits, limbs, or other parts of the fetus. It is believed that amniotic band syndrome occurs when the inner membrane (amnion) ruptures, or tears, without injury to the outer membrane (chorion).

**Case Report:** A 13-year-old male child was presented with constriction rings in the left upper and lower extremity since birth. For upper limb constriction band he was operated previously. Now came for lower limb. On examination there was a constriction ring about 3 cm distal to the left knee joint, involving total circumference, skin of the limb distal to constriction ring is dry, edematous and toes were absorbed due to auto amputation. Patient underwent excision of constriction band and Z-plasty in the left lower limb.

**Discussion:** The Amniotic bands may be single or multiple, superficial or deep and can occur anywhere in the upper or lower extremity resulting in intrauterine gangrene and fetal amputations. Musculoskeletal disorders commonly associated with bands include club foot, syndactyly, hypoplastic nails or fingers, distal lymphoedema, cleft lip and palate and umbilical hernia.

**Conclusion:** Amniotic constriction band presents at birth and should be treated as early as possible, because it becomes more pronounced as time passes and leads to various deformity of the limbs and ends up with auto amputation. Excision of constriction band and Z-plasty is the treatment of choice.

**Key words:** Amniotic band, Z-plasty, Autoamputation.

### INTRODUCTION

Amniotic band syndrome is a rare condition caused by strands of the amniotic sac that separate and entangle digits, limbs, or other parts of the fetus. It is believed that amniotic band syndrome occurs when the inner membrane (amnion) ruptures, or tears, without injury to the outer membrane (chorion). The developing fetus is still floating in the fluid but is exposed to the floating tissue (bands) from the ruptured

amnion. This floating tissue can become entangled around the fetus.

The incidence of amniotic band syndrome is 1 in 1200 to 1 in 15000 live birth.

Each case is unique multiple strands may be entangled around the fetus, and the severity can range from mild to life-threatening depending on where the bands are constricting and how tightly they are bound.

Exact etiology of this syndrome is not known but it may be caused by prenatal environmental factors and it appears to be result of excessive contraction of the uterine muscles and hemorrhages from the marginal blood sinus.

### CASE PRESENTATION

A 13-year-old male child was presented with constriction rings in the left upper and lower extremity since birth. For upper limb constriction band he was operated previously. Now came for lower limb. On examination there was a constriction ring about 3 cm distal to the left knee joint, involving total circumference (Figure 1), skin of the limb distal to constriction ring is dry, oedematous and toes were absorbed due to autoamputation. Distal pulsation could not be felt at the constriction ring site in involved limb. Sensations were present and equal in both limbs distal to constriction ring. Patient underwent excision of constriction band and Z-plasty (Figure 2) in the left lower limb. Post operative period was uneventful. On follow-up distal pulsation present in left lower limb.



Figure 1. Showing constriction ring of left lower extremity about 3 cm distal to knee joint.

### DISCUSSION

Amniotic band syndrome is set of congenital malformation arising from minor

constriction ring to complex multiple congenital anomalies that are attributed to amniotic bands that entangle and disrupt fetal parts.



Figure 2. Postoperative photograph of Z-plasty over left lower leg constriction ring.

Amniotic bands, also called constriction bands, congenital rings, Streeter dysplasia and annular defects are anomalous bands that encircle either partially or completely, a digit or an extremity. [1] It occurs in 1 in 1, 200 to 1 in 15,000 live births. Most cases present with multiple congenital anomalies that are incompatible with life. [2] Constriction bands are usually present since birth and may become more severe as the age of the child advances so it is necessary to manage them at the earliest to prevent complication.

Although several theories have been proposed to explain the genesis of ABS (Amniotic Band Syndrome), the most widely accepted view is that early rupture of the amnion results in mesodermic bands that emanate from the chorionic side of the amnion and insert on the foetal body, leading to constrictions, amputation and post natal deformities secondary to immobilization. [3]

Higginbottom et al [4] found that early insult (before 45 days gestation) produced severe facial clefts and severe

defects of the brain and calvarium, while late insult (after 45 days gestation) results in limb involvement with no facial clefting or central nervous system involvement. The index cases probably resulted from late insult as they had only limb involvement.

According to Pattersons [5] four diagnostic criteria for ABS, there should be presence of at least one of the following: 1) simple ring constrictions, 2) ring constrictions with distal deformity plus or minus lymph oedema, 3) ring constrictions accompanied by syndactyly or acrosyndactyly, 4) amputation.

Usually the constriction bands are confined to the skin and the soft tissues, but sometimes they are deep enough to cut off the normal vascular and lymph return resulting in chronic edema of the limb. [6] Various degrees of the anomaly can occur, from a partial constriction ring to complete amputation of a limb. [7]

The bands may be single or multiple, superficial or deep and can occur anywhere in the upper or lower extremity resulting in intrauterine gangrene and fetal amputations. [7] Musculoskeletal disorders commonly associated with ABS include club foot, syndactyly or acrosyndactyly, hypoplastic nails or fingers, pseudoarthrosis of underlying bones, peripheral nerve defects, distal lymph oedema, intra-uterine amputations, cleft lip and palate and umbilical hernia. [8]

Early detection of ABS in utero is not only important for counseling but also for possible intra-uterine surgical repair. [9] In utero, with the use of obstetric ultra sound scan, amniotic bands can be seen as linear echoes floating in the amniotic fluid and connected to the fetal body. [10] Three dimensional and four dimensional ultrasound and MRI(Magnetic Resonance Imaging) contributes to more sensitive prenatal diagnostics of amniotic band syndrome. Factors which affect accuracy of

Ultrasonography include: a) timing and duration of examination, b) expertise of the operator, c) technical difficulties such as oligohydramnios, maternal obesity, multiple gestation and multiple malformations. [11]

The accepted modality of treatment for amniotic band syndrome in utero is by foetoscopic laser surgery before the band compress fetal parts. [12] Plastic and reconstructive surgery to treat any resulting deformity has been performed after birth. Physical and occupational therapy for long term rehabilitation must be considered. Recently constriction ring can successfully released by a minimally invasive endoscopic surgical technique avoiding severe limb dysfunction or foot amputation. [2]

## CONCLUSION

Amniotic constriction band presents at birth and should be treated as early as possible, because it becomes more pronounced as time passes and leads to various deformity of the limbs and ends up with auto amputation. Excision of constriction band and Z-plasty is the treatment of choice.

## ACKNOWLEDGEMENT

We are thankful to Miss Rupali Salunkhe from Department of Surgery for her secretarial help.

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How to cite this article: Ashok K, Mayank V, Basavraj N et. al. Amniotic band syndrome- a rare case presentation. *Int J Health Sci Res.* 2014;4(9):386-389.

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