

Case Report

Amniotic band syndrome: a case series

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ABSTRACT

Amniotic band syndrome (ABS) is a rare congenital disorder which involves fetal entrapment in strands of amniotic tissue, which can manifest as constriction rings, limb and digital amputations or complex congenital anomalies and sometimes results in stillbirth. Etiopathogenesis is varied and ABS can be associated with syndromes. Antenatal diagnosis is made by ultrasonography in most cases. Postnatal examination reveals constrictions, amputations and craniofacial or abdominal wall defects. Treatment is case based and surgical intervention is required to release constriction rings. Authors report here six cases (one stillbirth and five live births) of amniotic band syndrome that were encountered over the last one and half years.

Keywords: Amniotic band syndrome, Constriction rings, Streeter's dysplasia

INTRODUCTION

Amniotic band sequence is a heterogeneous condition with broad spectrum anomalies, most commonly limb and digital amputations and constriction rings, with association of fibrous bands.¹

Cutaneous and visceral abnormalities may also be associated. Amniotic deformity, adhesion and mutilation (ADAM) sequence, amniotic band sequence, Streeter's dysplasia, congenital constriction bands, and pseudoainhum are the other names used for this condition.

Most cases are sporadic with no recurrence in siblings of affected patients. The prevalence of deformities secondary to partial or complete constriction ring is 1 in 10000-45000.² Males and females are equally affected.

Authors present six neonates with varied deformities and/or amputations with constriction rings, syndactyly and were diagnosed as amniotic band syndrome over last 1.5 years in our tertiary care NICU were analysed.

CASE REPORT

In this series of six, all were born to non-consanguineous marriage with no family history of amniotic band syndrome. Amongst them four were primigravida and two were multigravida mothers. In case one, 1st trimester Ultrasonography was suggestive of Dichorionic Diamniotic twins with single umbilical artery in twin A and oligohydramnios in twin B. The common anomaly in all the six neonates was the typical constriction ring on the lower limb (Figure 1 and 7) with one neonate having constriction ring around the fingers, one had fused malformed fingers (Figure 2) and other with fused toes (Figure 1). Case five had complete amputation of the upper arm (Figure 6). Case 3 (Figure 3 and 4) and 6 (Figure 7) had amputations of fingers and toes respectively, case one had syndactyly (Figure 1). Case 4 (Figure 5) had deformed foot. Amongst six cases, five survived and one died due to prematurity. Routine investigations were done to look for systemic anomalies and were managed conservatively with routine follow-up after discharge for further surgical intervention and rehabilitation.

Table 1: Cases with defect details and intervention done.

	Case 1	Case 2	Case 3	Case 4	Case 5	Case 6
Gender	Male	Female	Male	Male	Female	Male
Birth weight (grams)	2110	3218	900	2800	1700	2100
Defects	Constriction ring at lower limb, fused malformed toes, left hand syndactyly	Constriction ring at lower limb, Fused malformed fingers	Constriction ring at lower limb, partially amputated fingers.	Constriction ring at lower limb, deformed foot	Constriction ring at lower limb, complete amputation at the upper arm	Constriction ring at lower limb, constriction ring at the finger.
Interventions	Conservative management	Conservative management	Conservative management	Conservative management	Conservative management	Surgical release of the constriction ring.

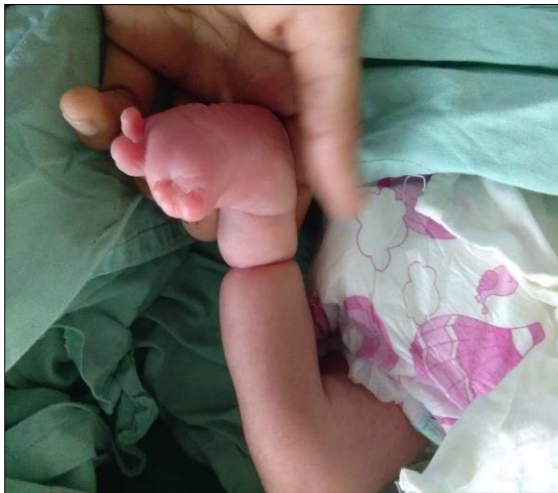


Figure 1: Constriction ring and fused malformed toes (Case 1).



Figure 3: X-ray malaligned phalanges and metacarpals (Case 2).



Figure 2: Completely fused malformed fingers (Case 2).



Figure 4: Partially amputated index finger (Case 3).



Figure 5: Deformed foot (Case 4).



Figure 6: Complete amputation at the upper arm (Case 5).



Figure 7: Tight constriction ring with necrosis and swelling limb distal to the ring (Case 6).



Figure 8: Constriction at the fingers (Case 6).

DISCUSSION

Amniotic band syndrome is a set of congenital birth defects caused by entrapment of fetal parts due to fibrous amniotic bands while in utero.

The exact pathogenesis is unknown. However, two theories have been put forth, the intrinsic and the extrinsic pathway. The extrinsic model by Tropin's model of 1965 suggested that the malformations are due to rupture of amnion in early pregnancy, and subsequent formation of amniotic bands with loss of amniotic fluid, causing extrusion of fetal parts in chorionic cavity.³ These bands entrap the fetal parts and limbs subjecting it to compression and necrosis.⁴ The intrinsic model was proposed by Streeter in 1930.⁵ He suggested that there was a common origin for fibrous bands and anomalies, caused by perturbation of developing germinal disc of the embryo. Most common are limb deformities. The broad fusion of disrupted fetal tissue and intact amniotic membrane lead to formation of adhesive bands. These bands cause abnormalities of craniofacial region such as encephalocele and facial clefts.² Whereas visceral defects such as renal agenesis³, microtia, clubfoot and rarely septo-optic dysplasia are also known to occur.⁶⁻⁸

The vascular disruptions cause limb body wall complex affecting embryonic structures, which includes 2 of the following 3 features: exencephaly or encephalocele with facial clefts, thoracoschisis and/or abdominoschisis, and limb defects.² The risk factors have been commonly implicated are maternal age, trauma, prematurity, amniocentesis, chorionic villous sampling and drugs. The lack of family history or predictable recurrences in families of children born with ABS negates the theory of an inherent or genetic component to the condition. The defect depends on the intensity of the constriction. It can be deep causing lymphatic obstruction leading to edema and vascular compromise that requires immediate release

of the band or it could be minimal as a merely cosmetic band.

The wide spectrum of clinical manifestations ranges from abnormalities such as hemihypertrophy, leg-length discrepancy, pseudarthrosis, resistant teratologic clubfeet and anterolateral bowing. The extension of the constriction bands to various body parts can lead to deformities such as facial clefts can be caused by constriction bands across the head and face. Encephaloceles can be seen secondary to extension of the cleft into the cranium. Chest deformities (thoracoschisis or extrathoracic heart) or abdominal deformities (gastroschisis) are seen when bands cross the body.⁹⁻¹¹

The rupture of the amnion occurs mainly before 12th weeks of gestation.¹² Diagnosis by fetal ultrasonography in early gestational age and in second trimester can detect visible amniotic bands, constriction rings on extremities, amputations of fingers and /or toes with a terminal syndactyly.¹³ 3D MRI and Ultrasonography aid in prenatal diagnosis of amniotic band syndrome.^{14,15} Treatment is case based and involves conservative and surgical management depending on the area of the body affected, severity of the constriction bands and neurovascular status. Most bands are superficial involving only the skin and subcutaneous tissue. However, there may be displaced neurovascular bundle around areas of banding. Amputation should be considered in cases of impending gangrene.^{16,17} Reconstructive plastic surgery along with Physiotherapy and occupational therapy may be necessary to correct or repair the associated anomalies and to ensure optimal use of affected fingers, toes and limbs.

CONCLUSION

Amniotic band syndrome is not very often seen and is a heterogeneous disease with broad spectrum anomalies. The basis for post-natal diagnosis is physical examination of the newborn with additional investigations to screen for internal organ malformations. Timely planned surgical intervention and close follow up due to the complexity of amniotic band syndrome is necessitated.

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