

## Limb hypoplasia and gangrene due to amniotic band syndrome – A case report and literature review

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### Abstract

Amniotic band syndrome (ABS) is a rare condition associated with vascular constrictions from fibrous bands entanglement of fetal body parts. We report a case of a neonate with amniotic band syndrome diagnosed at birth. The neonate presented with hypoplasia, vestigial, and gangrenous distal third of the right leg. Below the knee- amputation was done 48 hours after delivery, with a satisfactory outcome. This report captures a literature review on the pathogenesis, clinical presentations and treatment of ABS.

**Keywords:** Amniotic band, limb amputation, hypoplasia, limb gangrene

### Introduction

Amniotic band syndrome (ABS) is a notable cause of congenital anomaly [1]. The fetus develops in the amniotic sac of two layers called the chorion and amnion. The outer chorion is in contact with the uterus, and the inner amnion is with the fetus [2]. These two layers are fused to constitute the fetal (amnio-chorionic) membrane. Membrane integrity is usually maintained throughout gestation, mediated by well-balanced homeostasis involving matrix-degrading collagenolytic enzymes and their inhibitors [3, 4]. The amniotic membrane's epithelium contributes to amniotic fluid secretion. This fluid bathes and buoys the fetus, acting as a shock absorber [2]. The amnion is believed to be the source of the amniotic band [2].

ABS goes by other names such as constriction ring syndrome, annular ring defects, constrictive band syndrome, Streeter anomaly/bands, ADAM (amniotic deformities/adhesions/mutilations) Complex, amniotic band sequence, amniotic disruption complex, amnio-chorionic mesoblastic fibrous strings, congenital amputation, congenital constriction bands, tissue bands, e.t.c [5]. Most of these terms are more depictive of the pathology [6]. Amniotic bands are thought to be an accidental event and do not appear to have any genetic or hereditary background, and so have a remote likelihood of recurring in another pregnancy. However, according to Allen and colleagues, there have been reports of familial amniotic bands and an association with connective tissue disorders [7].

Whatever the name they are called, they are a complex kind of lesion, seen in various forms, grades and parts of the body, making their classification quite challenging. Patterson first put forward a classification system based on severity [8]. It is the most widely accepted classification because of its usefulness in treating different types of this condition. Amendments to the Patterson classification have separated the depth of the ring into mild, moderate, severe, and amputation and further defined the presence or absence of lymphedema or soft tissue loss distal to the ring [8].

Amniotic Band can entangle any part of a developing fetus. Most commonly affected are the limbs, manifesting as indentations, constriction rings, syndactyly, club foot, hypoplastic limb and, in rare occasions, amputation and resorption of parts of the digit or limb [9]. Craniofacial, umbilical and other extra-skeletal affectations are more lethal and are significant causes of intrauterine fetal death. The reported incidence of ABS varies from 1/1200 to 1/15,000 live births, while its prevalence has no sex predilection. It is seen in 1 in 70 spontaneous abortions [10]. The low incidence of this condition informed our desire to report. This report is the first of the syndrome in our tertiary health facility, which has been operating for over 30 years. We report this rare case of ABS and review the literature on its pathogenesis and clinical management.

### Case presentation

We present a male neonate, the first of a set of dizygotic twins, delivered through a cesarean section at 38 weeks of gestation. The baby was noticed at birth to have a vestigial hypoplastic and non-viable distal third of the right leg. No other abnormality was seen in the neonate.

The mother is a 34-year-old Para four, five, alive. The co-twin is healthy and has regular physical features. The mother had routine antenatal care, which was uneventful. Antenatal ultrasounds were reported to be normal. She had no trauma, prenatal instrumentation or febrile illness during pregnancy. She had no exposure to known teratogens or trauma. Her last confinement was also a twin gestation, one alive and intrauterine fetal death (IUFD) of a co-twin at 27th-week gestation.

The neonate had a constriction band at the junction of the proximal two-thirds and distal one-third of the right leg, creating an obvious demarcation at that leg level. The well-developed proximal two-thirds had normal muscle tone, skin colour and motor-sensory responses. The knee joint was typical, with a full range of motion. Part of the leg

distal to the constriction is poorly developed, including the ankle, foot, and toes. These parts were hypoplastic, vestigial, insensate, and showed no movements (fig 1).



**Fig 1:** Hypoplastic and gangrenous right leg, immediate post-delivery

The skin of these parts had a greyish hue and had turned dark and desiccated within hours of delivery. Systemic examination of the neonate showed no other abnormalities. Our diagnosis was amniotic band constriction in the distal third of the right leg with a gangrenous hypoplastic distal third of the leg, ankle and foot.

The baby was evaluated for surgery and had formal below-the-knee amputation under general anaesthesia on the third day of life. The constriction band was excised, and an appropriate skin and muscle flap was raised. We transected the tibia and the fibula 1.0 cm shorter and fashioned below the knee amputation stump (fig 2).



**Fig 2:** Right leg, immediate post below knee amputation

On the ninth day post-surgery, the stump had healed completely (Fig 3), and the patient was discharged for follow-up and prosthesis consultation. The patient was seen in the clinic two weeks post-discharge and is doing well.



**Fig 3:** Right leg, 10 days post below knee amputation

### Discussion

ABS is regarded as a rare condition that can bring sadness to an expected joyous condition of the arrival of a baby. It encompasses a range of congenital abnormalities that stretch from minor constriction rings and lymphoedema of the digits to complex and bizarre multiple anomalies, all linkable to entanglements by amniotic bands [7]. The implication of this is that ABS may result in fetal anomalies that vary in severity from minor to lethal, determined by different degrees of constriction, structure being constricted and gestational time at its formation [11].

### Etiopathogenesis

There is no consensus on how amniotic bands come about [3]. Some authors opine that they are generally of unknown aetiology and may be associated with other malformations like club foot, cleft lip and palate. [10]. However, some other authors propounded that the origin of malformations is multifactorial, involving multiple pathological processes. Different authors have put forward three theories to explain how these bands are formed – the Intrinsic theory [5], the extrinsic theory [6] and the vascular hypothesis [12].

The amnion, the side of the amniotic membrane in contact with the fetus, is believed to be the source of amniotic bands. Extrinsic, or the 1965 Torpin theory, is widely accepted [2]. It suggests that a tear in the amnion layer forms fibrous strands or cords of the amniotic band in its healing process. This fibrous band then entangles the affected fetal part; as the fetus grows, the band remain inelastic and does not expand. They become constricting and lead to the symptom complex of amniotic band syndrome [8]. The Intrinsic theory proposed by Streeter in 1930 implicates an anomaly in germplasm as the cause of ABS. This less accepted theory attributes the constriction bands to a localized imperfectly formed tissue at the constricted area due to a defective germplasm [5]. The theory is supported by the association of these rings with other internal visceral and systematic anomalies [9]. The vascular hypothesis links ABS to postulated vascular abnormalities [12]. Such defects could lead to malformation due to interruption of normal morphogenesis or deformation due to distortion of established structures and disruption of structures already formed.

Whatever may be the mechanism of development of the bands, it is commonly agreed that the effect of the amniotic band is due to mechanical obstruction of vascular supply distal to points of constriction and consequent ischemic injury. Our patient showed clear evidence of arrest of limb growth and development; hence, the distal third of the leg, ankle, and foot remained hypoplastic and vestigial due to poor blood perfusion. It turned gangrene when the constriction became strangulating. It has been reported that such gangrenous leg occurring early *in utero* could undergo auto-amputation and resorption<sup>[13]</sup>. The index case, though gangrenous, did not auto-amputate.

The mother of this neonate did not demonstrate any risk factor associated with amniotic band syndrome, such as age less than 25 years, primiparity and use of progesterone drugs<sup>[10, 14]</sup>. She did not have fetal surgery, amniocentesis or any form of abdominal trauma<sup>[15]</sup>. She, however, had a bad obstetric history of an intra-uterine fetal death (IUFD) of a co-twin in a previous twin pregnancy at 27th weeks gestation. The cause of the IUFD was not investigated, and the placenta was not examined. Although, the dead twin was noted to be stunted when compared with the surviving co-twin. Umbilical cord ABS may be responsible for the stunting and subsequent intrauterine fetal death. There is no ruling out a coincidence as ABS is reputed to be a sporadic event, with no evidence of familial occurrence<sup>[14]</sup>.

### Clinical Presentation

Amniotic bands usually occur within the first and second trimesters<sup>[1]</sup>. They can entangle any part of a developing fetus<sup>[9]</sup>. However, about 80% of cases involve the limbs, as seen in the index case, where the right leg was affected. The presenting symptoms vary depending on the gestational time the band is formed, the region or organ being constricted, and the degree of constriction.

The presentation could be mild if the constriction is not strangulating and may appear as an indentation or abnormal creases and grooves<sup>[10]</sup>. It could also be severe and lead to fatality in extreme cases. When constriction is severe and strangulating, it cuts off the blood supply and leads to gangrene, as in our case. Limb gangrene or amputation is a less common presentation of ABS. The involvement of digits is more often seen, ranging from digital ring constriction, digital atrophy, hypoplasia, and acrosyndactyly<sup>[7]</sup>.

When bands form in the first trimester of pregnancy, it often results in craniofacial and visceral constrictions, with a large number leading to intrauterine deaths or severe malformation. Early bands (those occurring in the first and early second trimesters) produce more devastating consequences due to the arrest of development following vascular compromise. Our patient showed clear evidence of arrest of growth and development of the affected limb, as a result of which the distal third of the leg, ankle and foot remained vestigial or hypoplastic before it subsequently became gangrene. The retardation in development and growth made us consider it as an early band formation.

The potential of an amniotic band to entangle any region or part of the body gives it its varied manifestations<sup>[9]</sup>. Entanglement and constriction of the upper and lower limb manifest as indentations, constriction rings, hypoplasia or complete absence of digits or limbs, club foot and, in rare occasions, lymphedema, amputation and resorption of the distal part of the digits, or limb gangrene. Most of these

deformities are compatible with life, as seen in the case presented. Cranio-fascial amniotic band constriction manifests as encephalocele, facial cleft or cleft lips and palate. The most lethal form of ABS is seen in umbilical cord entanglement and constriction. It is less reported but justifies and accounts for the higher incidence of ABS in abortions. It is important to note that not all umbilical ABS constrictions would lead to intrauterine fetal death or stillbirth<sup>[16]</sup>.

### Diagnosis

It is challenging to detect amniotic band syndrome before birth. The condition is usually diagnosed after birth following post-delivery examination, as was the case here. Sometimes, intrauterine diagnosis can be made with an ultrasonography scan, usually at the end of the first trimester or the beginning of the second trimester<sup>[17]</sup>. Because misdiagnosis is common with ultrasonography, 3D ultrasound and MRI are recommended for a more detailed and accurate diagnosis of bands and the resulting damage/danger to the fetus<sup>[10]</sup>.

Advances in the intrauterine diagnosis of ABS have raised hope for salvaging an ABS-threatened limb *in utero*<sup>[18]</sup>. Decisions can be made on the affected pregnancy and the severity of the deformity. However, These high-tech diagnostic procedures are currently unavailable to us in our institution and in many other developing economies. So, we rely primarily on postnatal diagnoses of fetal malformations.

### Treatment

The treatment options for amniotic band syndrome include in-utero fetal surgery and postnatal repair of the resulting abnormalities. The most appropriate choice is determined based on a detailed assessment involving high-resolution 3-D ultrasound, examination of the blood supply of the affected part, and magnetic resonance imaging (MRI). Following prenatal diagnosis, fetal surgery can be performed through an operative fetoscope to release the constriction caused by the amniotic band before it causes further damage<sup>[19]</sup>.

In cases where intra-uterine diagnosis and surgery are not possible, postnatal repair would be the option. Postnatal treatment consists of excision of the band and/or plastic and reconstructive surgery, followed by specialized physical and occupational therapy/rehabilitation. The observed defect would determine the extent and type of surgery. Sometimes, amputation and use of prosthesis may become necessary<sup>[20]</sup>. Our patient had a below-knee amputation as the only viable option available to us. Apart from the gangrenous limb, the vestigial or hypoplastic limb was not salvageable. The amputation stump was fashioned entirely with normal tissues above the constriction point. The patient has done well after surgery.

### Conclusion

ABS is uncommon in our practice but is well-documented in the literature. While its aetiology is still debated, researchers universally agree that variations in the degree of vascular constriction and strangulation account for its diverse presentations. These can range from simple indentations and grooves to more severe outcomes such as hypoplasia, gangrene, auto-amputation, and even fetal death, depending on the affected fetal part. Hypoplasia and gangrene of a limb are rare presentations. However, early intrauterine diagnosis

and fetoscopic surgeries provide hope for salvaging an affected limb. Following delivery, the only feasible option for limbs that are hypoplastic or gangrenous is amputation to create an acceptable stump suitable for rehabilitation, as demonstrated in the index case.

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