Case Report

Amniotic band syndrome; a case report

Osama T. Abu-Salah
Neonatal Unit, King Hussein Medical Center, Amman, Jordan

ABSTRACT
We describe a Jordanian infant with congenital malformations with multiple bands of various organs. (Rawal Med J 2011;36:159-162).

Key words
Amniotic bands, cleft lip, cleft palate, hypertelorism.

INTRODUCTION
Amniotic band syndrome is a sporadic constellation of congenital malformations with varying clinical presentation that ranges from mild localized limb lymphedema to severe lethal malformations such as anencephaly. Between these extremes, various organs can be affected, most commonly the fingers and toes. Amniotic bands attach to body parts and cause disruption in their formation and development. We report a case of a Jordanian neonate with cleft lip and palate as well as amputations in the fingers and toes.

CASE PRESENTATION
The baby was a male infant born at 34 weeks gestation. His mother was a healthy 20 years old primigravida and pregnancy was uncomplicated. There was no radiation or drug exposure and or an infection during pregnancy. The family history was negative to similar conditions and there was no consanguinity. Routine antenatal investigations were normal.

Fig 1. Cleft lip and palate with low set ears and deformed nose.
Examination showed a weight of 2200 grams and head circumference of 33cm and length of 48 cm. He had cleft lip and palate, abnormal nose, low set ears as well as hypertelorism (Fig 1).

**Fig 2. Left hand showing amputations and amniotic band is visible encircling the distal part of the ring finger. There is localized lymph edema of the index finger.**

![Left hand showing amputations and amniotic band](image1)

The right hand had normal thumb but digit II had constriction band at the base and lymph edema distally. Digit III was amputated from the level of the proximal interphalangeal joint. Digit IV shows a tourniquet like amniotic band wrapped around the finger at the level of the distal interphalangeal joint with distal gangrenous discoloration and swelling. The fifth digit was unaffected (Fig 2).

**Fig 3. Both hands showing visible amniotic bands, swelling, edema and ischemic changes.**

![Both hands showing visible amniotic bands](image2)

The left hand showed that digits III, IV and V were affected. There was an amniotic band visible, encircling digit III with distal rudimentary phalanx. The other digits show amputation distally (Fig 3).

**Fig 4. Both feet showing amputations of various toes of varying degree.**

![Both feet showing amputations](image3)
In both feet, there are amputations affecting the toes. In the right foot, all toes are affected, again with amputation, visible amniotic band and lymph edema of the unamputated first toe. The left first toe was completely amputated. There are amputations in the distal toes II and III. Toes IV and V are not affected (Fig 4).

**DISCUSSION**

The amniotic band syndrome (ABS) has a reported incidence of 1 in 10000 to 450000 births, with incidence among abortions of as high as 178:10.000. The visualization of the amniotic band is considered diagnostic of ABS. This precludes the need for further etiological investigations, such as karyotyping and genetic studies. There is negative family history of similar conditions in this case. This is consistent with previous report of the sporadic nature of this condition in most cases. However, association with family history of Ehlers Danlos and epidermilosis bullosa was noted in previous reports. Rare reports of autosomal familial ABS cases are found in the literature.

There are various theories attempting to explain the pathogenesis of ABS. The most popular is the “amniotic disruption” (exogenous) theory proposed by Torpin in 1965. in this theory, Tropin suggested that once the amnion is ruptured, the fetus lies outside the amniotic cavity and bands extending from the chorionic side of the cavity entrap various parts of the fetus and disturb normal development. The severity depends upon the period of gestation during which the bands develop, especially when accompanied with oligohydramnios.

The second popular theory is the “embryonic dysplasia”(endogenous) theory which was proposed in 1930 by Streeter. He suggested that abnormal histogenesis causes fetal disruption leading to defective tissue which later sloughs and its healing resulting in constricting rings. Patterson study of skin creases made him to suggest that amniotic bands are abnormal creases because of the histological similarities between creases and amniotic bands.

In the “vascular disruption theory,” proposed by van Allen, congenital malformations are a result of vascular insults.

The most frequent organs involved in ABS are the fingers and toes, with or without association with cleft lip and palate. Feet abnormalities such as club feet and fingers abnormalities such as syndactyly, cranial, cardiac, abdominal wall defect and abdominal organs extrophy, chest wall defect with heart extrophy are reported. Etiological associations have been reported with abdominal trauma, intrauterine devices, cerclage
and chorionic villous sampling.\textsuperscript{5,16} However, none is confirmed. It is possible to diagnose ABS antenatally with antenatal ultrasounds but minor defects are unlikely to be diagnosed.\textsuperscript{7} In conclusion, ABS is sporadic condition without agreeable etiology and pathogenesis. It has a wide spectrum of clinical most are mild and involving the limbs but severe abnormalities may be present and can be lethal.

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REFERENCES
