

## Amniotic band syndrome - A Case Report

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### Abstract:

*Amniotic band syndrome (ABS) is a set of congenital malformations attributed to amniotic bands that entangle fetal parts during intrauterine life, which results in a broad spectrum of anatomic disturbances - ranging from minor constriction rings and lymphedema of the digits to complex, bizarre multiple congenital anomalies incompatible with life. ABS is not seen very often, but should be considered in every newborn with congenital anomalies, especially defects of extremities and/or body walls. ABS can be diagnosed prenatally by ultrasound; otherwise, the defects are seen after birth. A team of specialists should be included in the treatment and follow-up of children with ABS, according to individual needs of every single patient. Earlier surgical intervention is must for proper growth & development of child. The aim of this paper is to point out diagnostic and therapeutic approaches in child with ABS through this case report.*

**Key Words:** *Amniotic band syndrome, Congenital band syndrome, Amniotic band disruption complex, Congenital annular defects.*

### Introduction

Amniotic band syndrome (ABS) is a set of congenital malformations attributed to amniotic bands that entangle fetal parts during intrauterine life, which results in a broad spectrum of anatomic disturbances - ranging from minor constriction rings and lymphedema of the digits to complex, bizarre multiple congenital anomalies incompatible with life (1,2). There is no genetic predisposition. Both genders are equally affected. Presentation can be varied from superficial circumferential grooves in skin to digital or whole limb amputation. There are various theories put forward to explain this syndrome. The most accepted one is of premature rupture of membrane with amniotic band formation causing compression and ischemia of fetal parts, Incidence of ABC is estimated in wide range of 1:

1200 – 1: 15000 live births (1, 3, 4-6) in 1: 70 in stillborns (7) and among abortuses as high as 178: 10000 (2). Among total of 3% major congenital malformations in general population (8), ABS is responsible for 1-2% (9). Surgery is not needed for shallow constriction bands that are not circumferential and without distal swelling. Distal edema or impairment of neurovascular function requires staged constriction band excision, Z-plasty, or W-plasty. Multiple plastic surgical procedures are required for corrections of the complex craniofacial abnormalities.

### Case report

A 7 month old male child 1<sup>st</sup> born of non-consanguineous marriage was brought to pediatric outpatient department by parents with complaint of right upper and both lower limb deformity present since birth. Parent also noticed shortening of right leg compare to left leg. Antenatal ultrasonography done in 3<sup>rd</sup> trimester showed constriction bands on both feet with edema. On examination child was active. Anthropometric measurements were appropriate for age. There was

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acrosyndactyly of index and middle finger and partial amputation of index finger of right hand. (Figure 1) Left hand was normal. Both feet were edematous. A deep circumferential groove was present 4 cm above both ankle joint. (Figure 2) Right leg's length was short compare to left leg by 2 cms. No other craniofacial or spine malformations were noted. On clinical basis diagnosis of amniotic band syndrome was present. Ultrasounds, echocardiography, X-ray were normal Reference of plastic surgeon was taken.

### Surgical management

Transverse incisions were marked out by outlining one side of the deep groove with a marking pen. Simple compression of the soft tissues leaves a corresponding mark on the other side. Under tourniquet control, incisions were made proximally and distally. All skin within the side wall was excised in two layers – first the skin and superficial fat; then the deep fat and areolar tissue. All scarred tissue was excised until the extensor tendons and flexor fascia palmarly was exposed.

Z plasty flaps were marked on both the lateral and medial sides of the limbs and raised with the adipose and soft tissue.

Soft and adipose tissue was mobilised advanced and approximated to correct the groove deformity. These flaps were approximated and closed with 4-0 Prolene sutures. The skin margins were approximated with subcuticular sutures.

### Post operative progress

All skin sutures were removed on the seventh day post surgery. Wounds healed well and three weeks post surgery the distal limb swelling had started decreasing.

### Clinical Photographs of the Patient:



**Fig. no 1:** Right hand showing acrosyndactyly of index and middle finger and partial amputation of middle finger



**Figure 2:** Constriction groove above both ankle joints



**Figure 3:** Post-operative picture: Z- plasty with excision of the constriction band

### Discussion:

ABS etiopathogenesis is still unknown, but there are two main theories (10-13). The widely accepted “extrinsic model”, proposed by Torpin and Faulkner in 1966 explains defects genesis by rupture of the amnion in early pregnancy, with forming of amniotic bands and amniotic liquid loss, followed by extrusion of all or parts of the fetus into the chorionic cavity. Bands entrap the parts of the growing fetus, and fetus’ limbs and other body parts become entangled and are subjected to compression. This compromises fetal circulation and also his growth and development with consecutive disturbances of functions and anatomy. The” intrinsic model” was proposed by Streeter in 1930 and suggests that the anomalies and the fibrous bands have a common origin, caused by a perturbation of developing germinal disc of the early embryo. Beside ABS etiopathogenesis, risk factors which start such sequence of events are also poorly known. Most cases of ABS are not of genetic origin, and there is no recurrence in siblings or children of affected adults. However, there are some reports of amniotic band syndrome among families with collagen disorders, more specifically Ehler-Danlos syndrome (2,6), in other diseases that involve connective tissue, e.g. in osteogenesis imperfecta , and in one case of

epidermolysis bullosa congenital (14,15). Some other possible etiologic factors besides inheritance were explored in several studies. Some studies found connection between ABS and mother's age (especially primiparas under the age of 25 (12,15), prematurity (16), abdominal trauma (2,15), unsuccessful abortion (17), intrauterine contraception (2), cerclage (18), chorionic villus sampling (17-20), amniocentesis (2,17-20), malformations of the uterus (15), some drugs like ergotamine (21), acetaminophen (12), misoprostol (21). However, there is no firm evidence of definite causality for any of these factors.

Amniotic band syndrome has very polymorphic clinical findings. Early amniotic rupture, during first 45 days, leads to the most severe cranio-facial and visceral malformations (5). Every part of the fetal body can be damaged, but most often extremities, especially upper extremities are affected. Most often there are minor defects, such as constriction rings or digit amputations (4). Abnormalities of the extremities can be expressed in several ways: constriction rings of the soft tissue accompanied by distal edema, shortening of the limb or intrauterine limb amputation, amputation of the digits (most often II, III and IV finger) and toes, syndactyly, hypoplasia of the digits, foot deformities, pseudoarthrosis, peripheral nerve palsy (1,21). If bands compress the fetal head or face, different cranio-facial disturbances appear – asymmetric face clefts, orbital defects (anophthalmos, microphthalmos, enophthalmos), corneal abnormalities, central nervous system malformations (anencephaly, encephalocoele, asymmetric meningocele), calvaria defect. Amniotic bands can also cause abdominal wall defect and abdominal organs extrophy (1), chest wall defect with heart extrophy (6), umbilical cord strangulation with often lethal outcome (1). Amniotic rupture and consecutive oligoamnion can, by mechanical pressure on the fetus, cause deformities such as metatarsovarum, scoliosis (5) or hip dislocation (1). Because of such a wide spectrum of possible anomalies and many combinations of their simultaneous appearance, there are no two identical cases of ABS (1). Beside all previously mentioned malformations caused by amniotic bands itself, a subset of cases manifest additional findings that are not consistent with that mechanism, such as congenital heart

defects, renal anomalies, hemangiomas, imperforate anus, polydactyly, septo-optic dysplasia, typical cleft lip and palate (10).

ABS can be diagnosed prenatally by ultrasound, which can sometimes show amniotic bands, but more often malformations consistent with ABS, as well as oligoamnion and reduction of foetal movements (22). The most important ultrasound diagnostic criteria are visible amniotic bands, constriction rings on extremities and irregular amputations of fingers and/or toes with terminal syndactyly. Mild defects, however, are less likely to be diagnosed prenatally, in which case defects are seen after birth (23). Latest ultrasound techniques – three-dimensional and four-dimensional ultrasound contribute to more sensitive prenatal diagnostics of ABS, and in complicated cases foetal magnetic resonance can be helpful (3). Placenta and amnion examination after the delivery should be obligatory part of the newborns health evaluation because it can show presence of amniotic bands, among other things (3,17,19). Physical examination is the main stay of postnatal diagnosis of ABS. However use of additional investigations like Ultrasound, echocardiography, X-ray are important in order to establish potential malformations of different organs and body parts.. ABS must be considered in differential diagnosis of all complex or asymmetric malformations, especially those on extremities, face and body walls. ABS should be differentiated from the whole spectra of symmetric fusion defects of middle body line (19).

Therapy of ABS is mostly surgical, with an individual approach to every single case. Interdisciplinary consulting and work is very often needed (plastic surgeon, orthopedic surgeon, orthodontist, ophthalmologist, neurosurgeon...) (1). Lately, there have been some attempts of prenatal ABS treatment - foetoscopic laser cutting of amniotic bands, before their compression on the fetus makes malformations (25). In cases when foetal anomalies incompatible with life are prenatally seen, pregnancy termination is advised (2).

### Conclusion

ABS is not seen very often, but should be considered in every newborn with congenital anomalies, especially defects of extremities and/or body walls. The basis for

postnatal diagnosis is physical examination of the newborn, with additional investigations to rule out other potential internal organs malformations. Because of ABS complexity, the treatment and follow-up of these children requires a team of specialists, according to special needs of every single patient.

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