

Amniotic band syndrome

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ABSTRACT

Amniotic band syndrome (ABS) is a well-described clinical entity, which includes several congenital deformities. Misdiagnosis is common for this entity. Incidence being 1 in 12000 births. Hand malformations and limb defects are frequent, constriction rings, acrosyndactylies and amniotic amputations, anencephaly, encephalocele, clefting, face (asymmetric), body wall and internal organs deformities, are associated with it.

Keywords: Amniotic bands, incidence, hand malformations

INTRODUCTION

Amniotic Band Syndrome is a rare disorder that comprises of a spectrum of defects including disruptions, deformations and malformations.¹Misdiagnosis is common. It is recommended that the clinician involved in the delivery of any infant with these defects seek specialized consultation for an accurate diagnosis and appropriate genetic counseling. Diagnosis earliest can be at 12 weeks^{2,3}.

CASE REPORT

A primi (24years) with 22weeks gestation referred from a private hospital with intrauterine growth restriction and an ultrasound, reporting amniotic bands and AFI-5cms. She came in active labour. On Per abdomen examination- fundal height was 18 weeks, 3 contractions lasting for 40seconds in 10 minutes. On obstetric ultrasound – Amniotic fluid index was 5cms, fetal heart rate was 84/min, a band of membranes constricting the fetal upper limbs, skull, neck which was reported as amniotic band. Per/Vaginum :cervix 20% effaced, 1 finger lose. As the patient came in labour, augmentation with misoprostol, oxytocin was done. Male stillborn child of weight 300gms was delivered. A band tightly constricting the neck, skull, upperlimbs was present [Fig 1] and some show constriction of fingers [Fig 2].



Figure 1: Shows the neonate delivered after augmentation of labour.



Figure 2: Shows constriction of fingers due to bands.

DISCUSSION

Synonyms of Amniotic Band Syndrome-ABS, amnion rupture sequence, amniotic bands, amniotic band sequence amniotic deformity, adhesions, mutilations (ADAM) complex, congenital constriction rings, constriction band syndrome, limb body wall complex, Streeter anomaly, Streeter bands, Streeter dysplasia. Amniotic band syndrome is a well-known condition potentially associated with a variety of different birth defects. It is important to note that no two cases of amniotic band syndrome are exactly alike and that the associated symptoms are highly variable.

Supposed risk factors are populations living at high altitude, multiparous women young maternal age, low maternal education, young paternal age, unplanned pregnancy, and non-white/non-Hispanic race/ethnicity. Cigarette smoking during early pregnancy is vasoconstrictive and has been related to gastroschisis, which is thought to arise from vascular disruption. The American study observed that use of acetaminophen in early pregnancy was associated with

increased risks Several different patterns have been identified with amniotic band syndrome. The three most common patterns are amniotic band syndrome characterized by one or more limbs being affected; the limb-body-wall complex; and amniotic band syndrome characterized by abnormalities of the head and face (craniofacial abnormalities), certain birth defects of the brain and spinal cord (neural tube defects) and serious malformation of the arms and legs. Adams-Oliver syndrome (AOS) is an extremely rare inherited disorder characterized by defects of the scalp and abnormalities of the fingers, toes, arms, and/or legs. The severity of amniotic band syndrome can range from a single, isolated finding to multiple, disfiguring complications. The arms and legs are most often affected.

The birth prevalence rate of ABS is reported to be 0.89 per 10, 000 births. Pathogenesis can be explained by several theories^{4,5,6} which include amniotic disruption, vascular disruption, embryonic dysplasia theory, Genetic Disorganization. Diagnosis antenatally is not possible in all cases, as individual bands are hard to see on USG. Misdiagnosis is common. 3D USG and MRI are more useful for accurate diagnosis. Usually treatment starts after birth. Other treatment modalities include realignment surgery for affected bones, compression garments to control swelling, Tools that helps the child do things on their own (adaptive equipment) and prosthetics to replace missing body parts^{5,6}.

Orioli et al⁷. have reported an increased incidence of ABS in the population living at a high altitude, in primipara, in women with a history of febrile illness in the antenatal period, and in women with a history of vaginal bleeding in the first trimester.

CONCLUSION

The phenotypic manifestations of amniotic band syndrome cannot be explained in totality by any one of the above mechanisms. The association of limb reduction defects and malformations involving different organs and different devices, supports the “intrinsic model”. According to this theory, in fact, the ABS would be the result of a vascular insult, which occurs very early in the embryogenesis. No less important, however, appear to be acquired yearly factors, such as the use of drugs, tobacco, diabetes, known for their action on the vascular system or even iatrogenic factors like the sting from amniocentesis which are an insult to the amniotic membranes. The finding, reported in the literature of iatrogenic cases of amniotic band syndrome, which occurred after amniocentesis or amnioreduction could support instead a different pathogenetic mechanism, which is closer to the extrinsic model. Presumably, both mechanisms operate, alone or in combination, to determine the syndrome, which is given

by a combination of genetic and environmental factors. All cases cannot be diagnosed antenatally as the bands are not visible on ultrasound always. The diagnosis is confirmed after birth of the neonate by visualizing the deformities and treatment is based upon the necessity⁸.

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