

AMNIOTIC BAND SYNDROME/STREETER'S DYSPLASIA: A CASE REPORT

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INTRODUCTION

It is a set of congenital birth defects believed to be caused by entrapment of fetal parts (usually a limb or digits) in fibrous amniotic bands while in utero. Prevalence-1 in 1200-15,000 live births.^[1] No gender or ethnic predispositions. About 80% of the cases involve the hands & fingers. The distal portion of the extremities is most often involved esp. the long central fingers of hand. In the feet, constricting bands most commonly involves the big toe.

Pathophysiology

It is not genetic & extremely unlikely that it will affect a future pregnancy.

The theories are- Extrinsic Theory- It occurs when the inner layer of amniotic sac tears exposing the fetus to strands of fibrous tissue that may float freely in the amniotic fluid or remain partially attached to the amniotic sac. These bands can disrupt the normal development of a fetus. It can entangle around the fingers, toes, arms, legs or any other parts of body.^[2]

Intrinsic Theory- In areas where there is impaired blood-flow, injury occurs to the blood-vessel walls of the fetus, causing bleeding & tissue-loss in the affected areas. The constriction bands develop secondary to the impaired blood-flow & subsequent damage to the fetus. The third theory postulates the involvement of intrauterine trauma like-amniocentesis.

Prenatal risk factors

- Prematurity
- Low birth weight
- Oligohydramnios
- Maternal illness during pregnancy
- Maternal drug exposure
- Maternal hemorrhage/trauma

Features

- Restriction of the growth around the digits, arms & legs.

- Constriction rings around the digits, arms & legs causing a deep groove in the baby's skin.^[3]
- Swelling of the extremities distal to the point of constriction(congenital lymphedema).
- Amputation of digits, arms & legs (congenital amputation).
- It may also cause-cleft lip or palate, encephalocele, omphalocele, scoliosis or kyphotic lordosis.

CASE REPORT

A 27years old G2P0010 was referred to hospital as a case of 36 weeks with Pre-eclampsia (not on drugs) with IUGR with oligohydramnios with Rh negative pregnancy in labour. She was diagnosed with raised B.P at 35wks of gestation. There was no significant past or family history. She had regular menstrual cycles & no dysmenorrhea. On examination, her general condition was fair, pulse rate was 90bpm, BP was 138/90mm Hg in right arm supine position. Her cardiovascular & respiratory system were within normal limits. On per abdomen examination, uterus was corresponding to 30 weeks gestation with cephalic presentation, contraction, with fetal heart rate 186bpm On per-speculum examination, thick meconium stained liquor present. On per-vaginumexamination, cervix was soft, central, 2.5-3cm dilated, 3-40% effaced, membranes absent, liquor was thick meconium stained, vertex at -2 station, pelvis average gynecoid. She was posted for caesarean section in view of Meconium stained liquor with fetal tachycardia, under spinal anesthesia. A live, preterm, male child of 2.1kg was delivered & handed over to pediatrician.



CONCLUSION

- In our case report, the baby had gangrenous left hand and distal forearm (peeling of skin on left hand and distal forearm present).
- No other gross congenital anomalies seen.
- Amniotic band syndrome can be difficult to diagnose. USG does not usually reveal it. 3D ultrasound and MRI can be used for more detailed & accurate diagnosis.^[4]
- The presentation of the patient with a history of amniocentesis must be monitored closely.
- Treatment usually occurs after birth and where plastic and reconstructive surgery is considered to treat the deformity.
- If diagnosed in utero, fetoscopic laser release of amniotic bands for threatened limb amputation.

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