

CONGENITAL CONSTRICTION BAND SYNDROME- A RARE CIRCULAR ABDOMINAL CONSTRICTION

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ABSTRACT

Aim: We describe a case of Congenital constriction band syndrome in a male baby of gestational age 39 weeks at the time of delivery. **Material And Methods** We hereby discuss a case of male baby born by vaginal delivery at full term with congenital abdominal constriction band along with isolated lower limb toe involvement. **Result:** Multidisciplinary approach taken which varies case wise according to the extent of deformity present in the baby. **Conclusion:** Congenital constriction band syndrome is a rare condition wherein the newborn presents with gross anomaly seen at one or more location present since intra uterine life due to encircling of amniotic band. The defect can range from limbs to abdominal wall to craniofacial anomaly or visceral defect. It can be detected antenatally by ultrasonography. Good prognosis with timely detection and treatment.

KEYWORDS : Amniotic band, constriction, abdominal, defect

INTRODUCTION

Congenital constriction band syndrome also known as amniotic band syndrome or Streeter's dysplasia refers to continuous spectrum of manifestations occurring due to intra uterine rupture of amnion.

It is a rare condition characterised by simple soft tissue constriction bands to amputation of digits or more severely of whole of the limbs.^[1]

The incidence is estimated to be 1 in 15000 to 1 in 10000.^[2] There is no sex predilection.^[3] The occurrence is sporadic. Prevalence is higher in foetus of gestational age between 9 to 20 weeks.^[4]

We describe a case of Congenital constriction band syndrome in a male baby of gestational age 39 weeks at the time of delivery. A rare form of constriction / amniotic band was seen in the baby which was present in the abdomen circumferentially. Also there was distal digits amputation seen as described further

Case Report

Male baby of gestational age 39 weeks was born out of non consanguineous marriage by vaginal delivery to a primiparous mother.

The antenatal period was uneventful. There was no history of any abdominal trauma or contraceptive use or any invasive antenatal investigation like amniocentesis by mother. Mother took iron supplementation, folic acid and two tetanus toxoid prophylaxis.

Antenatal ultrasonography scans were done. The 14th week ultrasonography scan was suggestive of amniotic band in the form of septum in the uterine cavity.

The delivery was spontaneous with vertex being the presenting part. Baby cried immediately after birth. Baby did not require any resuscitation or NICU admission. APGAR score at 1 minute after birth was 8 and at 5 minutes after birth was 9. Birth weight was 2.7 kg (between 3rd and 50th percentile).

A circular abdominal constriction band was seen just above the umbilicus [figure 1].



Figure 1

There was a constriction band involving left great toe and right 4th toe of lower limb leading to digital amputation [figure 2].



Figure 2

Right lower limb 2nd and 3rd toe syndactyly also seen. No other congenital anomaly seen. No other symptom like difficulty in breathing or any neurological pressure symptom seen. On day 1 of life abdominal and pelvic ultrasonography was done, which under normal limits Diagnosis can be done prenatally in early pregnancy, as early as 14 weeks of gestation, by antenatal ultrasonography. In later half of the pregnancy ultrasonography may even show gross deformities in the form of syndactyly, amputation etc.^[5] Parents were counselled and asked for regular follow up. On 8th month follow up the abdominal groove was noted to be involving skin, subcutaneous tissue, superficial and deep fascia and

underlying muscle [figure 3]. On abdominal radiography no bony involvement was found. Reconstructive surgery in future for amputated digits has been suggested to the parents.



Figure 3

DISCUSSION

The pathogenesis of amniotic band syndrome involves two theories namely; 'intrinsic model' and 'extrinsic model'.^[6] Streeter proposed intrinsic theory in 1930 and suggested that anomaly and fibrous band have a common origin, caused by a perturbation of developing germinal disc of developing embryo.^[7] In 1965 Tropin came up with extrinsic theory that said birth defects are caused by the action of fibrous amniotic bands with the sequence rupture of the amnion followed by loss of amniotic fluid and extrusion of all or parts of foetus into the chorionic cavity.^[8] It is an idiopathic syndrome however familial association with connective tissue disorder like Ehler Danlos syndrome has been found.^[9] Risk factors involving Amniotic band syndrome are mothers less than 25 years of age^[6], abdominal trauma^[10], any invasive investigation like amniocentesis, abortions that were not successful, drugs such as ergotamine, mesoprostol, acetaminophen, cocaine.^[6,11] Clinical manifestations include distal ring constriction, limb deformity and amputation.^[12] In few cases it may also manifest as syndactyly, clubfoot, cleft lip and palate, craniofacial synostosis.^[13] There has been cases reported where amniotic strangulation of umbilical cord has lead to foetal deaths.^[14] Constriction band syndrome can be diagnosed by antenatal ultrasonography. Three dimensional ultrasound has been done these days in few centers which can detect the condition as late as third trimester.^[15] Bands that are superficial may not need any intervention but deeper bands involving neurovascular bundle or lymphatic system may require urgent multispeciality intervention depending upon the case and degree of involvement. The closure of skin may be done by Z plasty or W plasty technique.

CONCLUSION

Constriction band syndrome or amniotic band syndrome is a rare disorder which can be detected even in antenatal period. Prognosis is fair if timely detection and management is done. As the involvement can be vast and varied, the treatment will require a team of specialist to look after each and every need of patient

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Conflict of interest

There is no conflict of interest.

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