

Case Report

Congenital constriction band syndrome: a case report

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ABSTRACT

Congenital constriction bands are anomalous bands formed during antenatal period that encircle a digit or an extremity in a fetus leading to a wide spectrum of manifestations, ranging from shallow grooves to acrosyndactyly and amputations. It is also associated with skeletal and other birth defects like craniofacial abnormalities, spinal dysraphism, porencephaly, hydrancephaly and visceral body wall malformations. We hereby present one such case which presented to us with below knee amputation of the lower limb and syndactyly.

Keywords: Amniotic band syndrome, Amputation, Congenital constriction band syndrome, Syndactyly

INTRODUCTION

Congenital constriction band syndrome (CCBS) also known as annular groove, amniotic band, Streeter's dysplasia, Amniotic deformity, adhesion, and mutilation (ADAM) sequence and ring constriction syndrome, is an uncommon condition with an incidence varying from 1 in 1200 to 1 in 15,000 live births.¹

Congenital constriction bands are anomalous bands formed during antenatal period that encircle a digit or an extremity in a fetus leading to compression effect and vascular compromise resulting in wide spectrum of manifestations, ranging from shallow grooves to acrosyndactyly and amputations. While most of limb anomalies are due to embryonic malformations, CCBS is a non-genetic, non-familial cause of limb anomalies causing post-conceptual deformity and disruption of developing limb during intrauterine period.²

It is also associated with skeletal and other birth defects like pseudoarthrosis, metatarsus adductus, peripheral nerve palsy, craniofacial abnormalities, spinal

dysraphism, porencephaly, hydrancephaly and visceral body wall malformations.^{3,4}

We hereby present one such case of a newborn which presented to us with below knee amputation of the left lower limb and syndactyly of 2nd and 3rd fingers of right hand.

CASE REPORT

A female newborn was born to a primigravida at 32-week gestation and 1.5kg birth weight via normal vaginal delivery. The baby cried immediately after birth. The baby had major limb anomalies at birth. There was amputation of the left lower limb 3.5cm distal to the knee (Figure 1) along with syndactyly of second and third digits of right hand (Figure 3). Radiological skeletal survey was done to search for No additional skeletal abnormalities were found in radiological skeletal survey (Figure 2 and 4). There was no history of any consanguinity or congenital malformation in other siblings or in the family. There was no history of maternal drug intake, radiation exposure, trauma or any

history of infection during pregnancy (like fever, rash or lymphadenopathy). Antennal ultrasound was done only in first trimester which was normal. A detailed clinical and systemic examination showed no other abnormalities. Baby's neurological examination was normal for gestation age showing normal tone and limb movements. Ultrasound whole abdomen, cranial ultrasound and echocardiography were all normal.



Figure 1: Below knee amputation of left lower limb.



Figure 2: Radiograph showing below knee amputation of left lower limb.

The baby had mild respiratory distress at birth (Transient tachypnea of newborn), which settled within 48 hours of birth. Baby was started on spoon feeds and appropriate nutritional supplements were added. The baby was discharged on the 10th day of life in fair condition. Baby showed normal growth and development pattern on follow up at 1 and 3 months. The baby was further

attached to orthopedic department for prosthetic management of limb anomaly and rehabilitation.



Figure 3: Syndactyly of 2nd and 3rd digit of right hand.



Figure 4: Radiograph showing syndactyly of 2nd and 3rd digits of right hand.

DISCUSSION

CCBS presents with a wide spectrum of severity ranging from superficial circumferential grooves in skin to digital or whole limb amputation. Some defects associated with it are pseudoarthrosis, metatarsus adductus, peripheral nerve palsy, craniofacial abnormalities, spinal dysraphism, porencephaly, hydrancephaly and visceral body wall malformations.^{3,4} CCBS shows no sex predilection or genetic predisposition.⁴ The condition is usually idiopathic, although association has been reported with advanced maternal age unplanned pregnancies and non-white, Hispanic races, but condition is largely idiopathic.⁵ Some other associations reported are

maternal trauma, oophorectomy in pregnancy, intrauterine contraceptive device, and amniocentesis.^{6,7}

Among the theories put forward to explain the etiology of the syndrome, the most accepted one is of amniotic band formation due to premature rupture of membranes leading to disruption of foetal parts due to compression effect.^{2,8} Possible mechanism is transient oligohydramnios due to loss of amniotic fluid through initially permeable chorion. The foetus passes from amniotic to extra-embryonic coelom through defect and comes in contact with sticky mesoderm on chorionic surface of mesoderm.⁹ Fibrous cords or amniotic bands are formed due to decidual reaction. Compression by these amniotic bands leads to a wide variety of birth defects like distal atrophy, digital or limb amputation, pseudo-syndactyly, lymphedema, club foot, in addition to severe craniofacial, visceral and trunk deformities.^{10,11} Prognosis depends on severity of associated anomalies. Patient with minor constriction rings and lymphedema have good prognosis while interventions like reconstructive surgery/ prosthesis are required in patients with amputation. The treatment of CCVS is therefore aimed at improvement of function and cosmetic appearance.¹² Consequently, the treatment plan for each patient must be individualized to maximize the outcome of surgery. Superficial grooves may be left as they are, as long as they do not interfere with lymph drainage and are not circumferential. The customary techniques described in many references to close the skin are mainly Z-plasty or W-plasty.¹³

CONCLUSION

CCBS is a rare but interesting syndrome requiring early recognition and investigation to rule out associated anomalies for their timely diagnosis and management. Appropriate counselling of the parents and timely referral to pediatric orthopedic unit for prosthesis and surgical management is essential for good prognosis.

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REFERENCES

1. Morovic CG, Berwart F, Varas J. Craniofacial anomalies of amniotic band syndrome in serial

- clinical cases. *Plast Reconstr Surg.* 2004;113:1556-62.
2. Light TR, Ogden JA. Congenital constriction band syndrome. Pathophysiology and treatment. *Yale J Biol Med.* 1993;66:143-55.
3. Allington NJ, Kumar SJ, Guille JT. Clubfeet associated with congenital constriction bands of ipsilateral lower extremity. *J Pediatr Orthop.* 1995;15:599-603.
4. Seeds JW, Cefalo RC, Herbert WN. Amniotic band syndrome. *Am J Obstet Gynecol.* 1982;144:243-8.
5. Werler MM, Lovik C, Mitchell AA. Epidemiologic analysis of maternal factors and amniotic band defects. *Birth Defects Res A Clin Mol Teratol.* 2003;67:68-72.
6. Strauss A, Hasbargen U, Paek B, Bauerfeind I, Hepp H. Intra-uterine fetal demise caused by amniotic band syndrome after standard amniocentesis. Intra-uterine fetal demise caused by amniotic band syndrome after standard amniocentesis. *Fetal Diagn Ther.* 2000;15(1):4-7.
7. Tanaka O, Koh T, Otani H. Amniogenic band anomalies in a fifth-month fetus and in a newborn from maternal oophorectomy during early pregnancy. *Teratology.* 1986;33:187-93.
8. Kawamura K, Chung KC. Constriction band syndrome. *Hand Clin.* 2009;25:257-64.
9. Torpin R. Fetal malformations caused by amnion rupture during gestation. *Bull Am Coll Nurse Midwives.* 1971;16:108-111.
10. Waltes JH Jr, Goss LR, Lazzara AT. Amniotic band syndrome. *J Foot Ankle Surg.* 1998;37:325-33.
11. O'Driscoll M, Peckham C, Kerr B. Four limb syndactyly, constriction rings and skin tags; amniotic bands or disorganization-like syndrome. *Clin Dysmorphol.* 2008;17:255-8.
12. Williams HB. Local gigantism of the hand and congenital constriction band syndrome in children. In: Kernahan D, Thomson H, eds. *Pediatric Plastic Surgery.* St Louis: CV Mosby Company;1981:423-8.
13. Wiedrich TA. Congenital constriction band syndrome. *Hand Clin.* 1998;14:31-7.

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