

Congenital constriction rings

S Rabah, S Salati, S Wani

Citation

S Rabah, S Salati, S Wani. *Congenital constriction rings*. The Internet Journal of Plastic Surgery. 2008 Volume 6 Number 1.

Abstract

Constriction band syndrome is set of congenital birth defects caused by entrapment of fetal parts usually a limb or digits in fibrous amniotic bands while in the uterus. It is also known as amniotic band sequence, streeter dysplasia, congenital bands or rings and amniotic adhesions mutilations

CASE PRESENTATION

Our patient is a 6 days old male baby, first in birth order and product of a non consanguineous marriage .The patient was born with congenitally abnormal right hand. On thorough examination, the patient has bilateral talipes equinovarus in addition to abnormal right hand. In the right hand, the thumb is normal, the index finger has amputated distal phalanx, the middle and the ring finger each has amputated distal phalanx and each bears a constriction ring at the level of middle phalanx and the little finger has amputated middle and distal phalanx. The patient has been attached to outpatients department for follow-up of hand defect .The worried parents were counseled and reassured that the disease has no proven familial link and that the abnormality in their first child does not increase the possibility of constriction/auto amputations in future offspring.

Figure 1

Figure 1: Constriction rings right hand (dorsal view)



Figure 2

Figure 2: Constriction ring Right hand (palmar view)



DISCUSSION

Constriction ring syndrome is a rare congenital anomaly ¹² Incidence of this condition is around 0.01% ³. Male and female patients in equal ratio and belonging to all ethnic groups are involved ¹³. Clinical presentations are varied from simple constriction rings to in-utero amputation or gangrene requiring amputation after birth. The part distal to the constriction may be normal or may be edematous (congenital lymph edema). Additional musculoskeletal disorders that may be present include talipes equinovarus, syndactyly, acrosyndactyly, hypoplastic nails, hypoplastic fingers, pseudoarthrosis of underlying bones, and absence of bones, peripheral nerve defects, cleft lip, cleft palate, umbilical hernia, microphthalmia, hydrocephalus, microphthalmia, thoracochisis and gastroschisis.⁴

It is believed to occur due to partial rupture of amniotic sac.

Only amnion is ruptured leaving chorion intact. The fibrous bands of the ruptured amnion float in the amniotic fluid which encircle and trap some part of fetus (extrinsic theory) leading to congenital abnormalities.³ There are however some supporters of intrinsic theory who defend that the constriction ring represents a defect of the embryological development resulting from abnormal differentiation of the germinative plasma.¹⁵

The constriction band can be superficial or deep. Superficial bands usually do not cause neurovascular damage or distal lymphedema and the medical attention is usually sought for cosmetic reasons. Deep bands, on the other hand, can result in neurovascular damage of variable intensity, requiring at times, urgent surgical intervention due to intensification of the progressive distal edema and vascular compromise.⁴⁶⁷⁸ Peripheral nerve suffer from lesion (axontomesis, neurotmesis) at the level of constriction band and the nervous tissue, proximal and distal to the band have been shown in literature to have normal at the microscopic level.⁹ Early diagnosis of the congenital band is possible using ultrasound after the first trimester of pregnancy.¹⁰

Treatment in the patients when surgical intervention is required includes Z-plasty, W-plasty or excision of the band with flap rotation of subcutaneous fat and closure of skin.

The procedure can be carried out in a single stage¹¹ as per some workers or as per other school of thought, half of the band is excised in first stage and rest in the second stage after an interval of six to twelve weeks.⁵

References

1. Askins G, Error G. Congenital constriction ring J Ped Orthop 1988;8;461-66
2. Kulkarni M L, Gopal P V. Amniotic band Syndrome J Indian Acad Ped 1990;27;471-76
3. Walter, J.H., Goss, L.R. & Lazarra, A.T. Amniotic band syndrome. J. Foot Ankle Surg. 1998; 37; 325-33
4. Pardini G Jr, Santos M A, Frietas A D. Congenital constriction bands. Acta Ortop Bras 2001;9(2); 3-10
5. Wiedrich TA. Congenital constriction band. Hand Clin 1998;14(1);20-38
6. Upton J, Tan C. Correction of constriction ring J Hand Surg (Am) 1991;16;947-953
7. Visuthikosol, V. & Hompuem, T.: Constriction band syndrome. Ann Plast. Surg. 21: 489-495, 1988.
8. Zych, GA, Ballard, A. Constriction band causing pseudarthrosis and impending gangrene of the leg. A case report with successful treatment. J Bone Joint Surg 1983;65A: 410-12.
9. Uchida, Y. & Sugioka, Y.: Peripheral nerve palsy associated with congenital constriction band syndrome. J. Hand Surg. (Br) 1991; 16:109-112
10. Quintero, R.A., Morales, W.J., Kalter, C.S. et al: In utero lysis of amniotic bands. Ultrasound Obstet. Gynecol 1997; 10; 316-320
11. Muguti, G.I.: The amniotic band syndrome: Single-stage correction Brit. J. Plast. Surg. 1990;43;706-708

Author Information

Sari Rabah, MBBS, FRCS(C)

Consultant, Department of Plastic & Reconstructive Surgery, King Fahad Medical City

Sajad Ahmad Salati, MBBS, MS, MRCS (Glasgow)

Assistant Consultant, Department of Plastic & Reconstructive Surgery, King Fahad Medical City

Shabeer Ahmed Wani, MBBS, MS, Mch

Assistant Consultant, Department of Plastic & Reconstructive Surgery, King Fahad Medical City