

Research Article

Presentation and treatment of congenital constriction ring syndrome: case series of 12 patients

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ABSTRACT

Background: We intend to present our experience of varied presentation and treatment of 12 cases of congenital constriction ring syndrome.

Methods: A prospective study of patients presenting with the characteristics of the congenital constriction ring syndrome was undertaken. Twelve patients made up of 7 males and 5 females were seen. The age at presentation ranged from nine days to 5 years with a mean age of 12.9 months. Nineteen limbs were affected, made up of four right upper limbs, six left upper limbs, six right lower limbs and three left lower limbs. In the upper limb malformations involved total 36 digits, arm in one case and forearm in one case; in the lower limb malformations involved total 27 toes, foot in one and leg in four cases. Types of lesions which were found: constriction rings, intrauterine amputations, simple syndactyly.

Results: Treatment options ranged from observation to multi-staged operations. Surgical correction of the constriction ring was done by excision and w-plasty to prevent or alleviate lymphoedema, multiple release incisions over distal limb to reduce the edema.

Conclusion: Congenital constriction ring syndrome is of uncertain aetiology and could cause morbidity in the newborn. The syndrome and its complications are amenable to corrective surgery with good results. Early intervention is desirable for a successful outcome.

Keywords: Constriction ring syndrome, Amniotic band syndrome, Streeter's dysplasia, W-PLASTY

INTRODUCTION

Numerous names have been suggested for constriction ring syndrome. These include annular groove, amniotic bands, Streeter's dysplasia, and ring constriction syndrome.

Amniotic band syndrome is an uncommon clinical entity that has been recognized for centuries.^{1,2} Amniotic band syndrome (ABS) is a set of congenital malformations attributed to amniotic bands that entangle fetal parts during intrauterine life, which results in a broad spectrum of anatomic disturbances - ranging from minor constriction rings and lymphedema of the digits to

complex, bizarre multiple congenital anomalies incompatible with life.^{3,4}

Congenital constriction band syndrome is estimated to occur in approximately one of each 15,000 live births.⁵

The term congenital constriction band syndrome is most appropriate in view of current understanding of the pathophysiology of this condition.

METHODS

During the period from August 2009 to November 2015, we studied 12 patients which presented with features of

congenital constriction band syndrome. The patients were examined clinically and the findings recorded. An x-ray and clinical photograph of the affected limbs was taken. Twelve patients made up of 7 males and 5 females were seen. The age at presentation ranged from nine days to 5 years with a mean age of 12.9 months. Nineteen limbs were affected, made up of four right upper limbs (Figure 1,2; Figure 7,8) six left upper limbs, six right lower limbs and three left lower limbs(Figure 5,6). In the upper limb malformations involved 36 digits (Figure 3 and 4) one arm and one forearm; in the lower limb malformations involved 27 toes (Figure 4), one foot and four legs. In one case involving lower leg with distal lymphedema, underlying tibia was fractured during the delivery (Figure 5 and 6). Types of lesions which were found: constriction rings with distal lymphedema, intrauterine amputations, simple syndactyly.



Figure 1: Constriction ring at distal forearm.



Figure 2: Circumferential involvement of the arm.



Figure 3: Involvement of fingers in Amniotic band syndrome.



Figure 4: Involvement of fingers and toes.



Figure 5: Distal lymphedema in a case of constriction ring at left leg.



Figure 6: Decompression by release incisions and W plasty.



Figure 7: Pre-op image of constriction ring at right arm.

Treatment options ranged from observation to several staged operations. In bands, where the depth was mild and moderate (no vascular compromise or limb contour deformity), observation and conservative measures were

undertaken. Surgical correction of the constriction ring was done by excision and W-plasty (hemi-circumferential) to prevent or alleviate lymphoedema, multiple release incisions over distal limb to reduce the edema.



Figure 8: Pre-op image of the patient (in figure 7) after w plasty.

RESULTS

Of the 12 patients who were subjected to the surgical correction, 3 had distal lymphedema in lower leg. Such cases were treated with release incision and/or hemi circumferential band excision and W plasty in first stage. The excision was then completed in next stage. This resulted in significant improvement in limb contour deformity and distal circulation (Figure 5,6). In rest of the cases the bands were excised and W-plasty was done.

DISCUSSION

At the time of birth, effects of the process are noted, but causative constricting bands are generally not seen. The lack of direct evidence led to the development of 2 main theories of etiology. The intrinsic theory holds that germ cell deficiencies result in malformations of the affected parts.^{6,7,8} The extrinsic theory supports the role of bands of ruptured amnion in the creation of extrinsic compression that results in constriction rings and other deformities of the developing fetus.^{1,9}

Although ABS etio pathogenesis is still unknown, there are two main theories¹⁰⁻¹³ as already mentioned. Widely accepted extrinsic model, proposed by Torpin and Faulkner in 1966 explains defects genesis by rupture of the amnion in early pregnancy, with forming of amniotic bands and amniotic liquid loss, followed by extrusion of all or parts of the fetus into the chorionic cavity. Bands entrap the parts of the growing fetus, and fetus' limbs and other body parts become entangled and subjected to compression, which compromises fetal circulation and also his growth and development with consecutive disturbances of functions and anatomy. The intrinsic model was proposed by Streeter in 1930 and suggests that the anomalies and the fibrous bands have a common origin, caused by a perturbation of developing germinal disc of the early embryo.

Amniotic band syndrome has very polymorphic clinical findings, because type of deformities depends on the time of amniotic rupture during pregnancy and part of the fetal body which is entangled in amniotic bands. Early amniotic rupture, during first 45 days, leads to the most severe cranio-facial and visceral malformations.¹⁴ Every part of the fetal body can be damaged, but most often extremities, especially upper extremities. Most often there are minor defects, such as constriction rings or digit amputations; but, even minor defects are multiple in 77% of cases.¹⁵ Abnormalities of the extremities can be expressed in several ways: constriction rings of the soft tissue accompanied by distal edema, shortenage of the limb or intrauterine limb amputation, amputation of the digits (most often II, III and IV finger) and toes, syndactyly, hypoplasia of the digits, foot deformities, pseudoarthrosis, peripheral nerve palsy Amniotic bands can also cause abdominal wall defect and abdominal organs extrophy, chest wall defect with heart extrophy, umbilical cord strangulation with often lethal outcome.^{3,16,17}

Because of such a wide spectrum of possible anomalies and many combinations of their simultaneous appearance, there are no two identical cases of ABS. The severity of the condition is decided by depth of the constriction bands. It is classified by separating the depth of the ring into mild, moderate, severe and amputation and by further defining the presence or absence of lymphedema or soft tissue loss distal to the ring.¹⁸⁻²¹

ABS can be diagnosed prenatally by ultrasound, which can sometimes show amniotic bands, but more often malformations consistent with ABS, as well as oligoamnion and reduction of foetal movements.²² ABS can be diagnosed as early as 12 gestational weeks.²³ In the second trimester of gestation most of ABS defects could be seen during routine ultrasound examinations.²² The most important ultrasound diagnostic criteria are visible amniotic bands, constriction rings on extremities and irregular amputations of fingers and/or toes with terminal syndactyly.

Therapy of ABS is mostly surgical, with an individual approach to every single case. Interdisciplinary consulting and work is very often needed (plastic surgeon, orthopedic surgeon, orthodontist, ophthalmologist, neurosurgeon and others).³

Surgical interventions include triangular flaps, Z or W plasty or simple approximation of the excision margins.

We found that decompression of the distal edematous limb by multiple release incisions reduces the tension on the skin and it becomes more pliable to be used for W-plasty.

CONCLUSION

ABS is not very often, but should be considered in every newborn with congenital anomalies, especially defect of extremities and/or body walls. The basis for postnatal diagnosis is physical examination of the newborn, with additional examinations after potential internal organs malformations. Child's chromosomal karyotype analyses in order to exclude wrong diagnosis and consecutive incorrect information about recurrence risk for the parents is of utmost importance.

Because of ABS complexity, the treatment and follow-up of these children requires a team of specialist, according to special needs of every single patient.

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