

Complete circumferential congenital constriction of the trunk – operation procedures and results

Vollständige zirkumferentielle Schnürfurche des Rumpfes – Operationstechniken und Ergebnisse

Abstract

Constriction ring syndrome is an uncommon deformity with unknown etiology and multiple manifestations. The most common change occurs at the lower extremities. A complete circular amniotic band syndrome of the trunk is an extremely rare condition. There are less than ten other reported cases in the literature. We present a new case of this congenital abnormality, the operation procedure and the results.

Keywords: congenital constriction, operation technique, plastic surgery, surgical flap, trunk

Zusammenfassung

Eine kongenitale Schnürfurche ist eine seltene Deformität bei unklarer Ätiologie und mit zahlreichen möglichen Manifestationen. In den meisten Fällen findet man derartige Veränderungen im Bereich der Extremitäten. Eine vollständige zirkumferentielle Einschnürung des Rumpfes ist eine sehr seltene Veränderung. In der Literatur sind weniger als 10 Fälle beschrieben. Wir stellen einen neuen Fall mit einer derartigen kongenitalen Deformität vor und präsentieren die Operationstechnik und die klinischen Ergebnisse.

Schlüsselwörter: Schnürfurche, Operationstechnik, plastische Chirurgie, Rumpf

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Introduction

Constriction ring syndrome is an uncommon deformity with unknown etiology and multiple manifestations. The incidence varied from 1 in 3,000 to 1 in 15,000 live births [1]. Numerous different clinical deformities are familiar with the amniotic band syndrome, ranging from simple ring constrictions to complex craniofacial or abdominal defects.

Clinical report

A 16-week-old girl was referred for advice by her mother. The reported prenatal follow-up as well as the delivery was uneventful. The child presented a constricting ring of the abdominal wall in total circumference involving the umbilicus (Figure 1). Moreover the left lower extremity about 3 cm proximal to ankle joint and the II-IV toes of both feet were affected. The child showed no functional problems. The infant was born maturely by normal vaginal delivery. There was no family history of congenital deformities and no other associated abnormality in body. We

managed our case by excising the abdominal constriction band and multiple z-plasties (Figure 2). The postoperative course was uneventful. The suture material was removed in the 15th postoperative day. The 6-month follow-up showed good cosmetic results with no signs for hypertrophic scar formation (Figure 3). At this time the plastic surgical corrections of the left leg by multiple z-plasties were performed.

Discussion

Constriction ring syndrome is an uncommon deformity with unknown etiology and multiple manifestations. The most common change occurs at the lower extremities. Digital ring constrictions, digital atrophy, congenital intrauterine amputations, acrosyndactyly, lymphoedema and clubfeet can occur.

The constriction ring or band causes soft tissue depressions, encircling digits, extremities or limbs, sometimes the neck, thorax or abdomen. A complete circular amniotic band of the trunk is an extremely rare condition.

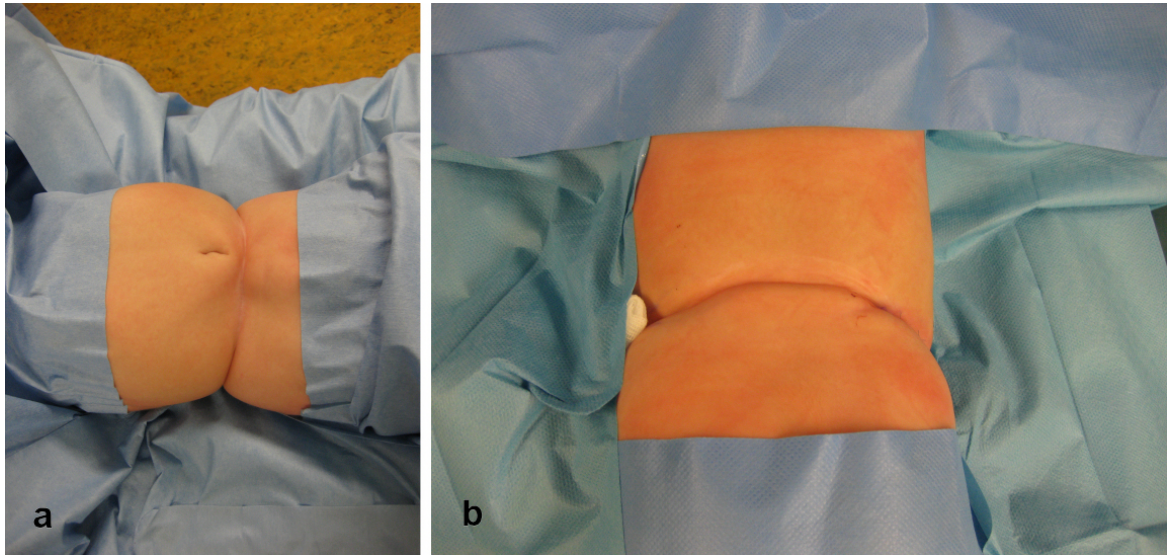


Figure 1: Examination findings before operation. Abdominal wall frontside (a) and backside (b)

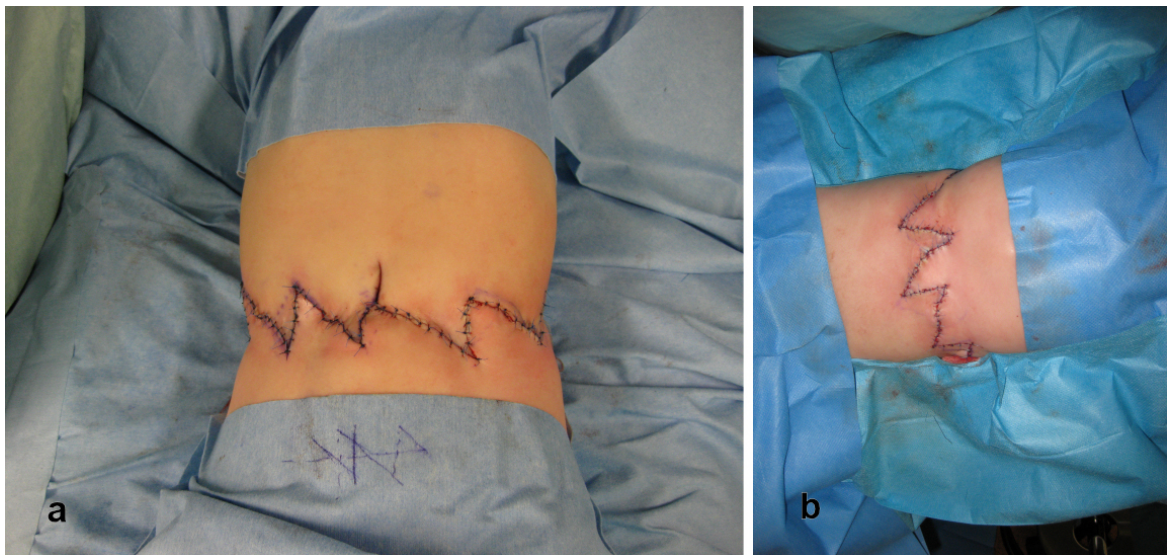


Figure 2: Postoperative findings following multiple z-plasties. Abdominal wall frontside (a) and backside (b)

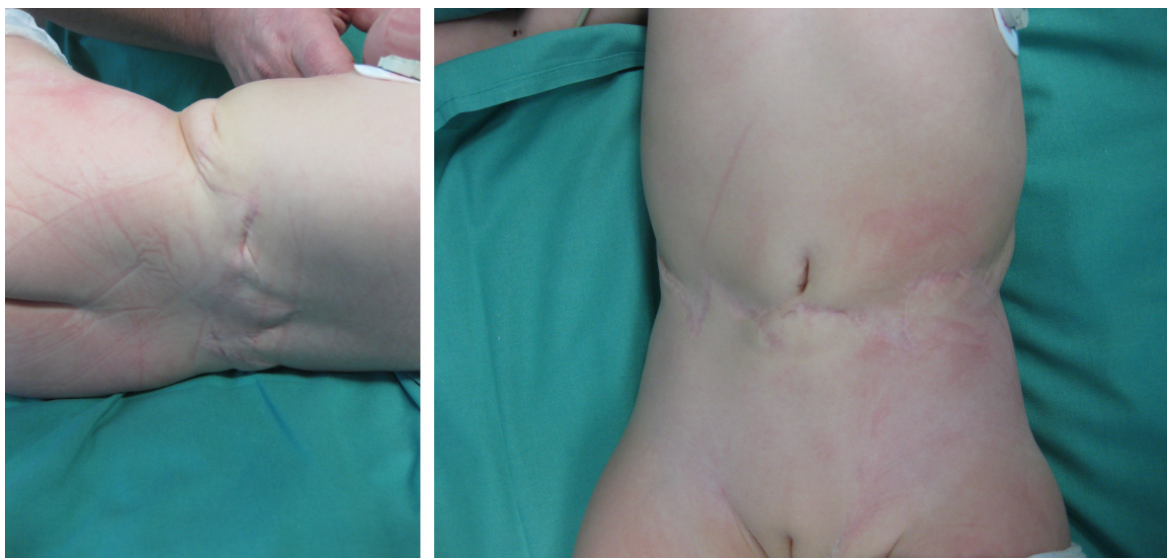


Figure 3: Postoperative findings after six months. Abdominal wall frontside (a) and backside (b)

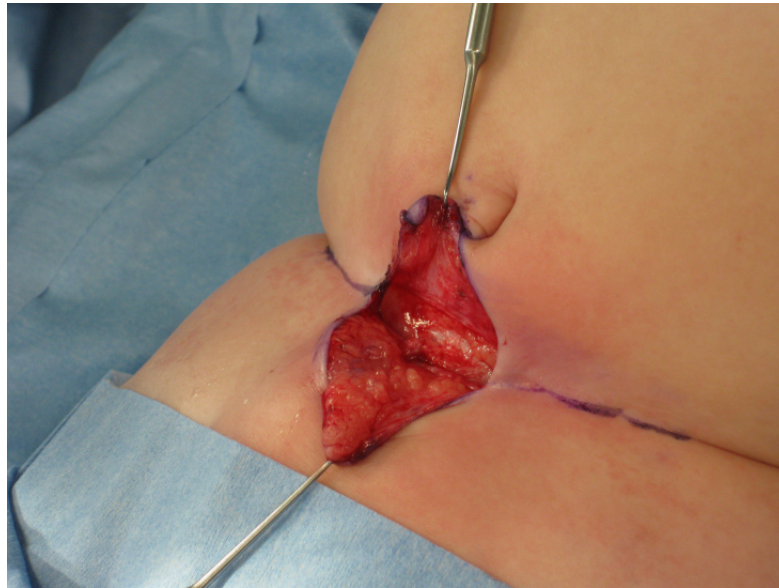


Figure 4: Preparation in the umbilical region

There are less than 10 reported cases in the literature [2], [3], [4].

The extent of the constrictions were first described by Patterson in 1961 [1]:

- Simple constriction rings without distal deformity
- Constriction rings with distal soft tissue deformities including or excluding lymphedema
- Constriction rings with distal deformities including distal bony fusions from mild to severe acrosyndactyly
- Intrauterine amputations

Two conflicting theories concerning the pathogenesis:

1. Amniotic band theory

Amniotic bands occurred as a result of premature rupture of the amniotic sac. Bands and fragments of the rapid amniotic in combination with mesodermic fibrous strings enclose different parts especially at the extremities. Amniotic bands are unelastic and produce ligature effects. The time of the amniotic sac rupture correlates with the severity of deformities. An early rupture results in multiple severe deformities, late rupture (after day 45 of gestation) in limb constrictions [5].

2. Failure of development theory

Limb ring constrictions were due to localised areas of imperfectly formed tissue caused by defective germplasm. Supporters of this theory consider that the amniotic bands arise as an integral part of the same localised failure of development that gives rises to the limb constriction rings [1], [6].

The preferred method of treatment for constriction ring syndrome in general is serial excision followed by multiple z-plasties, a common flap-surgery procedure [7] (Figure 2). This procedure will often produce excellent cosmetic results, but the surgical correction should be carried out carefully [2], [3], [4], [8]. Special considerations are required in the umbilical region. It is necessary to avoid a

deviation of the umbilical region out of the midline (Figure 4). Deeper layers of the abdominal wall are in general not involved so that further reconstructions i.e. of the abdominal fascia are not necessary. Hypertrophic scar formations were described in other reported cases. Early protective treatment by local silicone sheets could be favorable.

Conclusions

A complete circular amniotic band syndrome of the trunk is an extremely rare condition. Early plastic surgery operation procedure results in satisfactory functional and aesthetic results.

Notes

Competing interests

The authors declare that they have no competing interests.

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