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## CASE REPORT: CONGENITAL AMNIOTIC BAND SYNDROME – A RARE CASE OF TOTAL CIRCUMFERENTIAL ABDOMINAL CONSTRICTION AND ITS REPAIR

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### ABSTRACT

Amniotic band syndrome is a rare and complex condition often associated with severe complications in fetal development. We present the case of a 3-year-and-4-month-old female patient with total circumferential abdominal constriction due to an amniotic band. Referred to the General Hospital of Fortaleza, the patient exhibited a constricting ring in the abdominal wall, with imaging studies indicating the presence of fat and possible intestinal involvement in the hernia. A reparative surgical procedure was performed, involving complete resection of the band and the application of the W-plasty technique to enhance tissue mobility. The procedure presented significant challenges due to the pediatric nature of the patient and the deep location of the band, necessitating multiple incisions and complete removal of the fibrous band, with particular emphasis on postoperative healing to minimize the risk of dehiscence and flap necrosis. The primary objective of the surgery was to reduce intra-abdominal pressure and restore skin elasticity. Postoperatively, the recovery was closely monitored, as follow-up was essential to prevent complications given the patient's age. Initial outcomes demonstrate improvements in intra-abdominal pressure and skin elasticity, positively contributing to the patient's quality of life and promoting her adequate growth.

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## INTRODUCTION

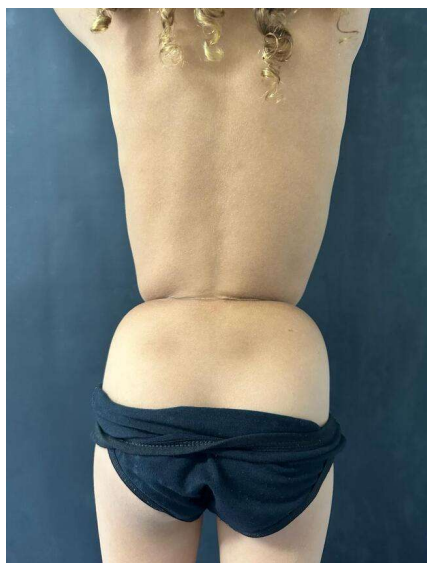
The amnion, a vital component of the amniotic sac, plays a crucial role in safeguarding and promoting fetal development during gestation. Premature ruptures of the amnion can lead to the formation of amniotic bands, which are fibrous tissue strands encircling parts of the fetus. Although the etiology of these amniotic bands remains uncertain, potential causes include premature rupture of membranes and vascular anomalies (Askins & Error, 1988; Burgens *et al.*, 1991). The formation of these bands can result in amniotic band syndrome, a rare congenital anomaly causing various fetal developmental complications, most commonly limb constrictions that may lead to deformities, amputations, or tissue loss (Kulkarni & Gopal, 1990).

Less frequently, these bands can affect facial structures and thoracoabdominal organs, potentially impacting internal organs such as the heart, lungs, and intestines. Furthermore, it is important to highlight that abdominal constriction bands are associated with an increased risk of miscarriage or stillbirth, particularly when there is direct involvement of the umbilical cord. These bands can be located in various regions of the abdominal area, displaying variable depths and extensions (Levy, 1998; Muguti, 1990). Typical features include a more pronounced band located posteriorly, which may be positioned above or below the iliac crest, resulting in constriction and consequently elevated intra-abdominal pressure, along with restricted growth. The diagnosis is typically achieved through prenatal ultrasound examinations, where structural anomalies in the fetus can be observed (Quintero *et al.*, 1997). However, in some cases,

diagnosis may be challenging due to the complex and variable nature of the condition. Treatment often requires immediate surgical intervention after birth to correct deformities and restore the function of affected organs, with the prognosis depending on the extent and severity of associated complications, as well as the timeliness of diagnosis and appropriate treatment (Uchida & Sugioka, 1991; Upton & Tan, 1991).

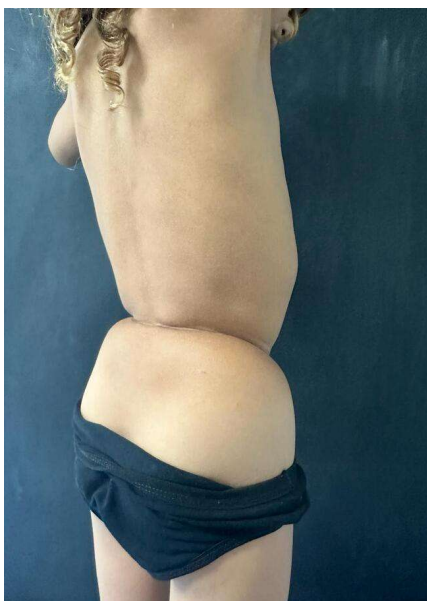
### Case Report

A 3-year and 4-month-old female preschooler from Fortaleza, Ceará, was referred to the Hospital Geral de Fortaleza for evaluation by the plastic surgery team. The patient presented with a deep constricting ring completely encircling the abdominal wall, located at the level of the posterior-superior iliac crest (Figures 1 and 2) and inferior to the umbilicus (Figure 3).



Source: Authors' Collection

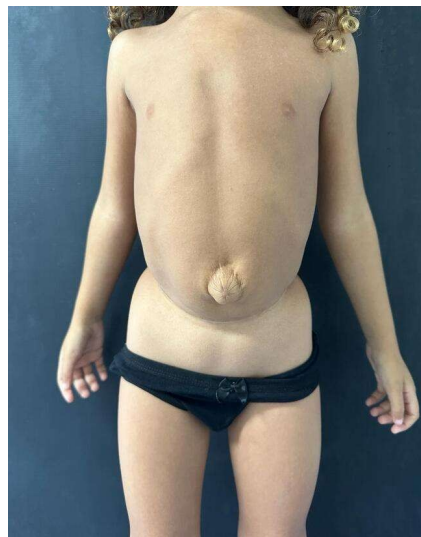
**Figure 1. Constricting ring on the dorsal wall posterior-superior to the iliac crest**



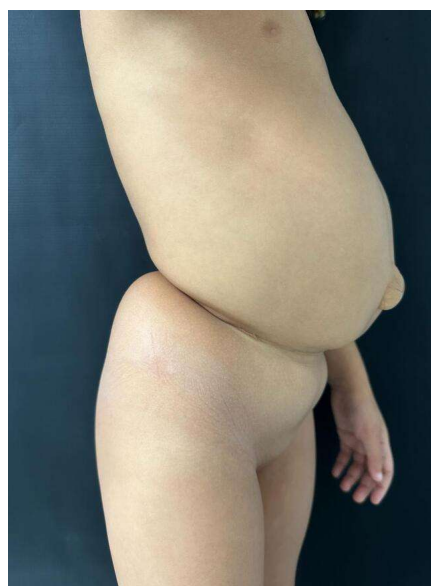
**Figure 2. Constricting ring on the lateral abdominal wall**

At the time of evaluation, the patient demonstrated normal development, except for complications arising from increased intra-abdominal pressure, which resulted in an umbilical hernia (Figure 4). A review of the patient's medical records, photographic documentation of the lesion, and details of the surgical procedure characterized this study as observational and descriptive. The patient

underwent magnetic resonance imaging (MRI), blood tests, and chest radiography. An abdominal MRI performed in August 2023 revealed that the hernia contents consisted of fat, with possible involvement of intestinal loops, measuring 1.5 cm in diameter and without evidence of acute inflammatory processes. Additionally, the MRI noted circumferentially reduced skin and subcutaneous tissue around the lower abdominal region, likely corresponding to the fibrous ring secondary to amniotic band syndrome.



**Figure 3. Constricting ring on the inferior abdominal wall**

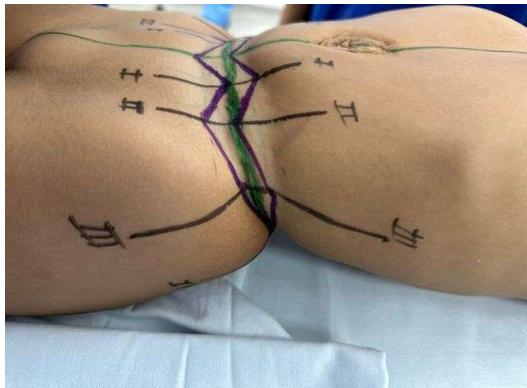


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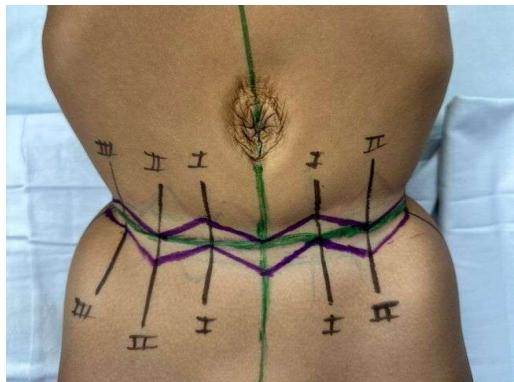
**Figure 4. Umbilical hernia**

Blood tests, including a complete blood count, creatinine, urea, glucose, prothrombin time (PT), and activated partial thromboplastin time (aPTT), showed no abnormalities. Finally, chest X-rays in anteroposterior (AP) and lateral views also showed no structural alterations. The surgical plan initially involved the incision and excision of the fibrous band, followed by multiple Z-plasties or W-plasties. A distinctive aspect of this procedure was the patient's age (4 years and 5 months) and the deep, significantly constricting location of the band at the iliac crest. Despite the MRI not revealing any intracavitary abnormalities, the pronounced constriction increased the risk of difficulties in simultaneously addressing the anterior and posterior components of the procedure due to the need for patient repositioning. The proposed surgical approach included the complete excision of the band through a circumferential incision starting anteriorly and subsequently released near the umbilicus (Figure 5), aiming to remove the "belt-like" band while preserving tissue integrity

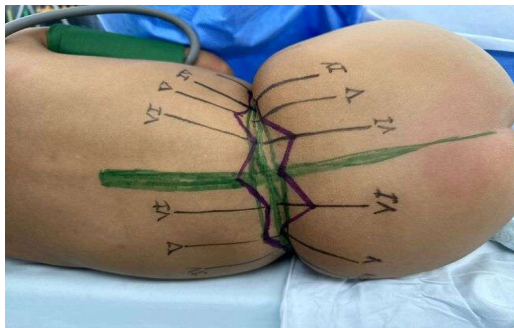
in a single surgical session. The surgical intervention then proceeded with Z-plasty due to the efficiency of the procedure (Figure 6), after which the posterior portion of the band removal was initiated (Figure 7).



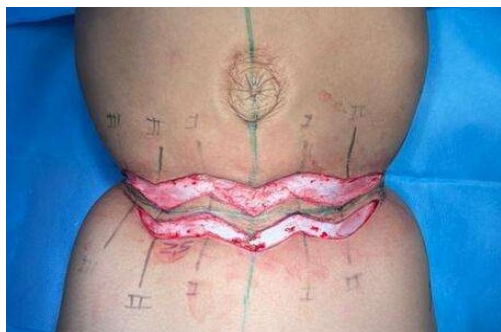
**Figure 5. Preoperative markings, lateral view**



**Figure 6. Preoperative markings, anterior view**



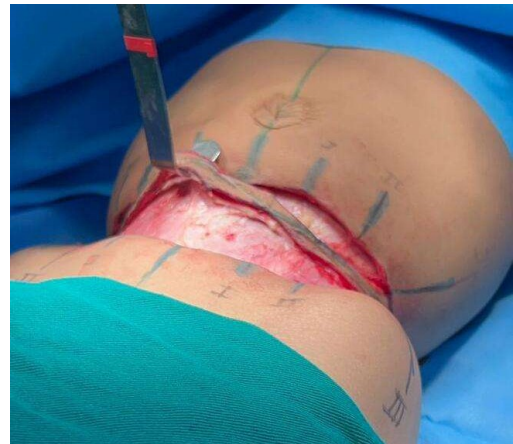
**Figure 7. Preoperative markings, posterior view**



**Figure 8. W-plasty excision of tissue around the band**

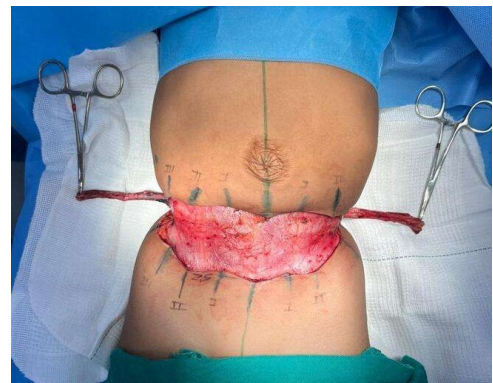
During the surgical intervention, a W-plasty was performed due to the dimensions of the band encircling the entire abdomen (Figure 8), consisting of interconnected multiple Z-plasties. This procedure was conducted in a single surgical session. Once the anterior band was released (Figures 9, 10, and 11), tissue closure was completed, addressing three layers commonly treated in abdominoplasty: subcutaneous, subdermal, and intradermal layers (Figure 12). The

primary goal of this intervention was to reduce tension in the superficial layer, improve mobility, and facilitate adequate tissue growth in this pediatric patient (Figures 13, 14, 15, and 16).



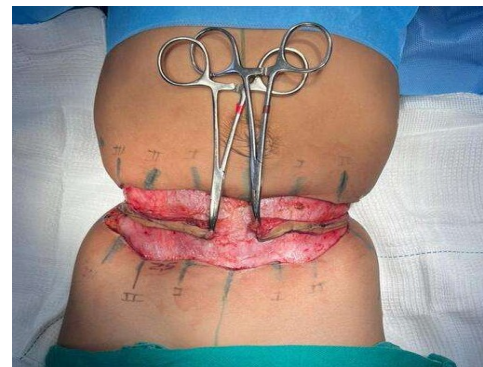
Source: Authors' Collection

**Figure 9. Complete release of the anterior band**



Source: Authors' Collection

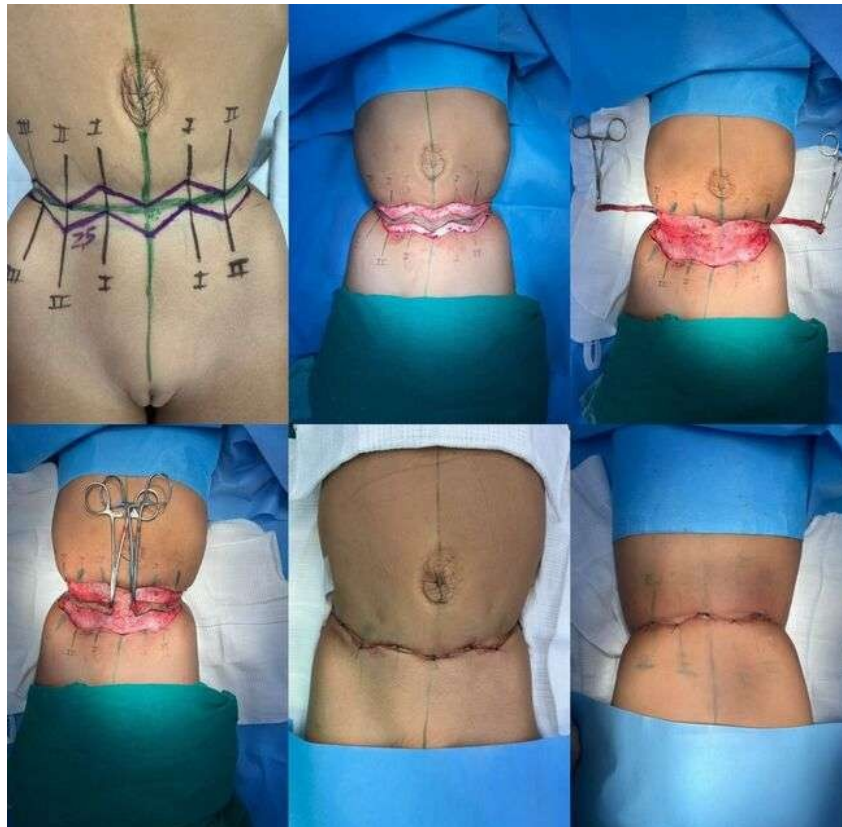
**Figure 10. Separation of the anterior band to facilitate its removal**



**Figure 11.**

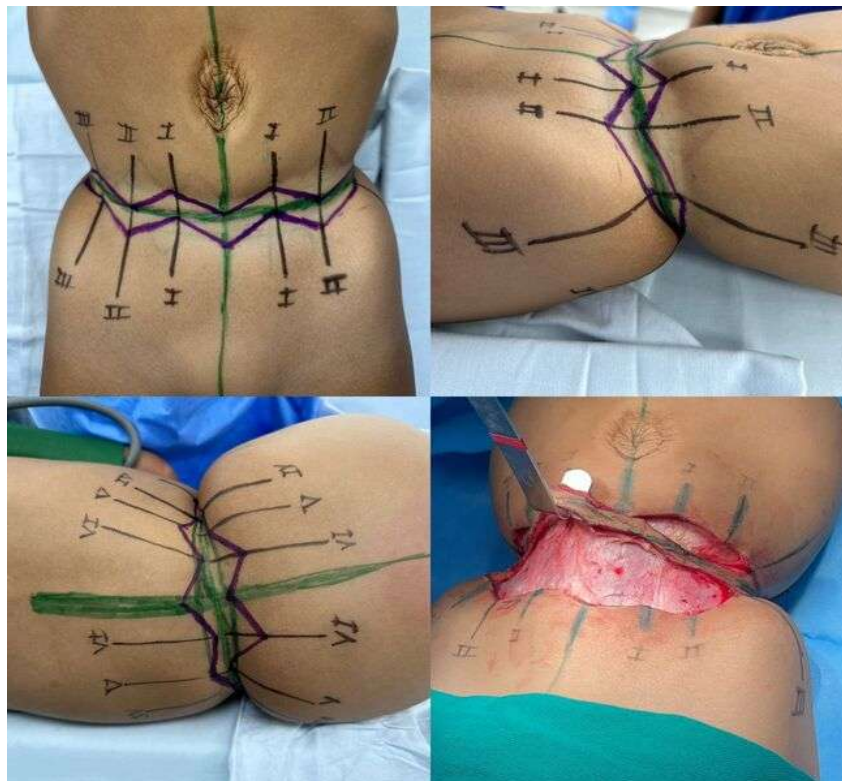


**Figure 12. Immediate results after closure of the edges**



Source: Authors' Collection

**Figure 13. Step-by-step surgical procedure**



Source: Authors' Collection

**Figure 14. Demonstration of constriction caused by the band**

Postoperative follow-up is essential, particularly given the pediatric age of the patient, to prevent complications such as dehiscence and necrosis of the flaps. The primary expected outcome of the surgery is the reduction of intra-abdominal pressure and restoration of normal skin elasticity, aiming to prevent further growth impairment, which has already been observed.

Planning for a subsequent surgical procedure, including umbilical reconstruction and scar refinement, is also underway.

## DISCUSSION

The formation of congenital constriction bands is a rare pathology with an unclear etiology.



Source: Authors' Collection

**Figure 15. Comparison between preoperative and postoperative stages**



Source: Authors' Collection

It presents a wide range of clinical manifestations, including involvement of limbs, face, or abdomen, often leading to outcomes such as amputation, acrosyndactyly, syndactyly, and impairment of lower limbs. Trunk involvement is uncommon, and the presence of a total circumferential abdominal amniotic band is even rarer compared to other affected regions. This condition was first reported by Brown *et al.* in 1957, with approximately 20 cases documented in the literature to date. Regarding the surgical treatment of this pathology, several approaches can be considered, ranging from the excision of the band with deep tissue dissection and release to the use of protective techniques. There is ongoing debate about whether the dissection should be performed in a single or multiple stages. Earlier studies advocate for releasing the fibrous ring in two or more stages. In this approach, the dorsal half of the ring is released initially to improve venous and lymphatic drainage, with the other half being addressed 12 weeks later (Askins & Error, 1988; Upton & Tan, 1991; Walter *et al.*, 1998; Wiedrich, 1998; Choulakian & Williams, 2008). Conversely, more recent publications suggest that the procedure can be safely completed in a single surgical stage (Muguti, 1990; Visuthikosol & Hompuem, 1988; Pardini *et al.*, 2001; Costa *et al.*, 1996). However, there is no substantial evidence favoring one approach over the other. The most critical aspects remain the safety of the procedure and the restoration of anatomical and functional integrity.

## CONCLUSION

In summary, after a thorough clinical evaluation and meticulous documentation of exams and the patient's medical history in their medical records, it was determined that surgical correction was the most appropriate treatment. Amniotic band syndrome is rare, and its approach presents unique challenges, especially in a pediatric context, as surgery is the only effective treatment.

The chosen technique involved isolating the fibrous ring of the band, followed by W-plasty, which proved to be an excellent option for closing abdominal wall defects. This approach aligns with the outcomes of the few similar cases reported, enabling the restoration of mobility and the growth of constricted tissue, particularly in the pediatric context of this patient. The entire procedure was conducted in a single surgical stage, addressing three tissue layers: subcutaneous, subdermal, and intradermal. Subsequently, the surgical procedure was carried out in accordance with standard and safe practices, without complications. Postoperative follow-up included evaluating the patient's overall condition and monitoring the surgical wounds, focusing on the healing process and identifying potential complications resulting from the intervention.

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