

Surgical correction of congenital constriction band syndrome in children: Replacing Z-plasty with direct closure

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Congenital constriction band syndrome is a sporadic condition that may also be present in association with other congenital anomalies. It has an incidence varying from one in 1200 to one in 15,000 live births. There is a significant predilection for the upper extremities and distal limbs. The two main objectives for the treatment of congenital constriction band syndrome are improvement of function and improvement of cosmetic appearance. Different surgical techniques, such as Z-plasty, have been described and used for decades; however, direct closure after the excision of the constricting band seems to be the simplest and most appropriate, allowing the fatty tissue to naturally reposition itself under the skin. This technique is used in a two-stage approach to avoid affecting distal circulation to the limb.

Key Words: Amniotic bands; Congenital constriction band syndrome; Direct closure; Z-plasty

Congenital constriction band syndrome (CCBS) is a sporadic condition that may also be present in association with other congenital anomalies such as musculoskeletal, craniofacial and thoraco-abdominal disorders (1,2). It has an incidence of one in 1200 (3) to one in 15,000 live births (4) and affects both sexes at a ratio of 1:1. There is a significant predilection for the upper extremities and an increased frequency in distal limbs (1), and longer digits are significantly more involved than shorter ones (5,6). CCBS is categorized as a class VI congenital limb malformation (7). Other names used for this clinical entity include Streeter's dysplasia (8) and amniotic bands (9).

The deformities in CCBS vary widely, ranging from shallow grooves to complete amputations; they may also be partial or completely circumferential (10). In 1961, Patterson used a classification that is still widely used today (4). The classifications are as follows:

- simple ring constrictions;
- ring constrictions accompanied by deformity of the distal part with or without lymphedema;

Correction chirurgicale du syndrome des brides amniotiques chez les enfants : Remplacement de la plastie en Z par une fermeture directe

Le syndrome des brides amniotiques est une maladie sporadique qui peut également se manifester en association avec d'autres anomalies congénitales. Son incidence varie de 1 pour 1 200 à 1 pour 15 000 naissances vivantes. La maladie semble affecter significativement plus les membres supérieurs et les extrémités. Le principal objectif du traitement du syndrome des brides amniotiques est l'amélioration à la fois fonctionnelle et esthétique. Différentes techniques chirurgicales, comme la plastie en Z, ont été décrites et sont utilisées depuis des décennies. Toutefois, la fermeture directe après excision des brides constrictives semble plus simple et plus appropriée, car elle permet une répartition plus naturelle des tissus adipeux sous la peau. Cette approche s'applique en deux étapes pour éviter d'affecter la circulation distale dans les membres.

- ring constrictions accompanied by fusion of distal parts ranging from mild to severe acrosyndactyly; and
- intrauterine amputations

The exact cause of CCBS remains unknown, although two main theories cause controversy when attempting to explain its etiology. The intrinsic theory, described by Streeter (8) in 1930, suggests that the primary defect occurs before the natural differentiation of the embryo, and arises from the level of the subcutaneous germplasm. This could therefore explain the craniofacial and internal organ abnormalities associated with CCBS (11). On the other side, the extrinsic theory, described by Torpin (12) in 1965, suggests that early rupture of the amnion, leading to oligohydramnios, could cause proliferation of mesenchymal bands or amniotic bands. These bands could then entangle the limb, causing the constriction. This later theory is consequently supported by orthopedic findings, such as acrosyndactyly (4), amputation (13) and club feet (10,12).

Concerning the treatment of CCBS, most references recommend the use of Z-plasty or W-plasty after the excision of

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Figure 1 A A two-year-old boy with a deep circumferential constriction band in the distal one-third of the leg. Both legs were involved with identical bands, and distal edema of the feet was evident. B The same patient following a two-stage bilateral excision of the constriction bands. The right side was closed with Z-plasty, and the left side was closed directly. Distal foot edema was relieved after the first-stage excision. Note the improved scar over the left leg

the constriction band, in a one- or two-stage approach (10,14-19). In the present study, we describe how the use of direct closure yields improved scarring and superior cosmetic results, without compromising the limb. This technique has been used by the division of Pediatric Plastic Surgery at the Montreal Children's Hospital (Montreal, Quebec) for several decades.

SURGICAL TECHNIQUE

Once the patient is under general anesthesia and a tourniquet has been applied to the limb, the band is outlined with a surgical marking pen. The constricted area must be meticulously dissected to avoid damaging the underlying neurovasculature. The constriction band is excised and direct closure is completed. This allows the fatty tissue to naturally reposition itself under the skin. For wound closure, a 3-0 Monocryl suture (Ethicon Inc, USA) is used in the deep layer and a 4-0 Monocryl suture is used in the subcuticular layer.

With complete circumferential constriction bands, it is recommended that a two-stage correction approach be used. At the first operation, one-half of the circumference is excised and the other one-half can be excised after three to six months. This will avoid any problems to the distal circulation in the limb, which may already be compromised. Lymphedema, when present, will significantly improve within a few weeks of the first surgery.

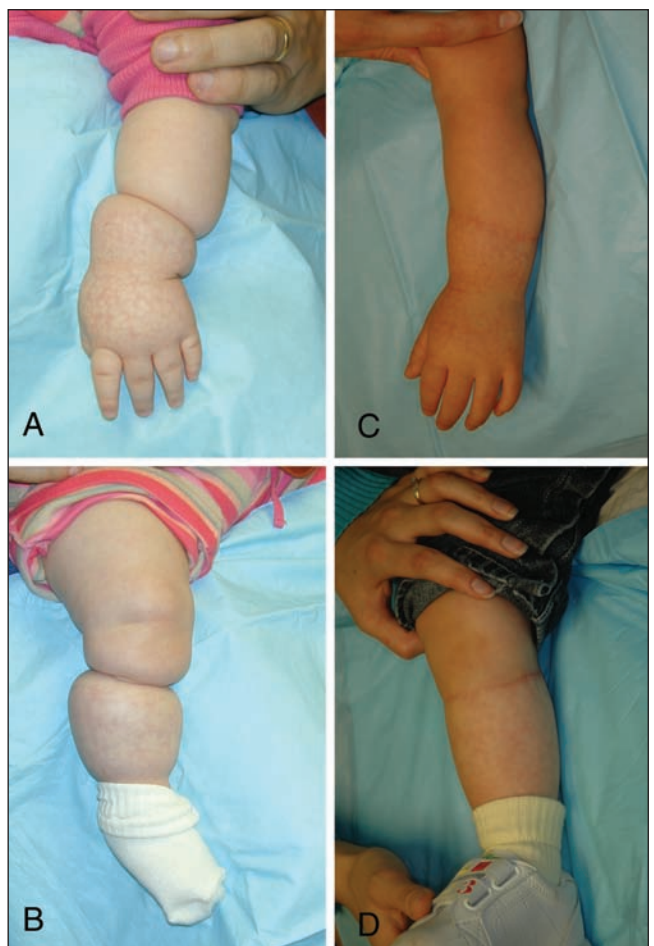


Figure 2 Arm (A) and foot (B) with deep circumferential constriction bands. C,D The same limbs following two-stage excision of constriction bands with direct closure

DISCUSSION

CCBS not only causes aesthetic deformity in the affected limb, but may also cause vascular compromise, which can lead to lymphedema and/or amputation. The treatment of CCBS is therefore aimed at improvement of function and improvement of cosmetic appearance (20). Consequently, the treatment plan for each patient must be individualized to maximize the outcome of surgery (10). 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 (10,14-19). In 2006, Mutaf and Sunay (21) suggested the use of a rectangular-plasty technique, aligning the major rectangular limbs in parallel with the relaxed skin tension lines (21). However, we believe that the superficial scar resulting from the surgery is much more aesthetic with direct closure (Figures 1 and 2). Stevenson (22) acknowledged that this technique works, but thought that the circular scar could contract, thus reproducing a constriction band. Nevertheless, in our clinical experience, none of our patient's follow-up visits revealed the presence of a contracted scar.

Excision of the dermis and subcutaneous tissue may result in a deficiency under the constriction band, which could cause a 'sand glass deformity' (21). This phenomenon has

been described when closure is done with Z- or W-plasty and the subcutaneous tissue is not mobilized. In 1991, Upton and Cissy (18) recommended excising the abundant subcutaneous fat beside the ring, then debulking dorsally the remaining adipose tissue and advancing it over the tissue-deficient site. Subcutaneous fat advancement flaps such as these could therefore prevent the recurrence of a contour deformity. Mutaf and Sunay (21), on the other hand, used rectangular dermofat flaps turned over in alternation. Although we strongly agree with these two procedures, we do not believe

there is a need to mobilize the underlying fat with our technique. Our clinical experience has demonstrated that with simple excision and direct closure, the fat naturally falls into place over time, thus eliminating the possibility of an underlying deformity.

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