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CASE REPORT

Urgent release of constriction band syndrome of the leg with single-stage Z-plasty procedure in neonates: A case series

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Abstract

The congenital constriction band syndrome (CCBS) is a group of fetal abnormalities with myriad of clinical presentation that is characterized by the presence of a fibrous band that encircles fetal tissues in utero. It is a rare disease with a reported incidence of 1/15.000 live births. The disease is treated with release of the constricting band with Z-plasty, where it is classically done as a two-stage procedure. Single stage z-plasty is a relatively new approach that still needs more reports regarding to its result. Our cases described successful release with a single stage Z-plasty reconstruction in the setting of an urgent surgery in neonates aged less than 48 hours.

Keywords: congenital constriction band syndrome, contracture, Z-plasty

Introduction

Congenital constriction band syndrome (CCBS) is a rare entity in which entanglement of body parts occurs in the amniotic membrane, that leads to deformation, malformation, and amputation.[1] It is also referred to as amniotic band syndrome or congenital annular/ring constrictions. The formation of intrauterine fibrous bands can partially or completely encircle a limb or other body part by the mechanical forces exerted. The incidence of CCBS is estimated to be 1 in 1,200 to 15,000 live births. This syndrome affects male and female similarly, with 1:1 ratio.[2,3]

There is no universal agreement on the etiology of CCBS, however there are two main theories that are to explain the development. The intrinsic theory, which Streeter first proposed in 1930, postulated that the amniotic band is created as an improper development of the germinal sac in embryo. As stated by Torpin in 1965, the extrinsic theory contends that an early rupture of the amniotic sac results in the formation of fibrous amniotic bands, which can wrap and entrap a portion of the developing fetus as they float in the amniotic fluid. These act as limiting bands that, as the fetus develops, decrease blood flow and, as a result, can result in auto-amputation of a finger or limb in utero. Sometimes it causes necrosis, which needs to be surgically amputated after birth.[4]

Congenital constriction band syndrome with its various clinical manifestations, most commonly affect the extremities, with a predilection for distal digits. The manifestations CBS include acrosyndactyly, neurovascular compromise, neuropathy, lymphedema, congenital amputation, and other deformities of the hands and feet. Although less common, it can also present with multiple craniofacial and truncal abnormalities.[4]

Patterson developed a useful classification that consists of 4 types for describing the extent of banding: (1) simple constriction ring, (2) constriction ring with deformity of the distal part, (3) constriction with fusion of distal parts (acrosyndactyly), and (4) complete intrauterine amputation.[5]

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Mild cases of CCBS that do not result in neurovascular impairment typically does not need urgent surgical intervention. Surgical intervention is typically postponed until after six months of age to prevent anesthesia-related problems, unless the constricting band induces significant lymphedema, severe hypoperfusion, or functional impairment. The most common indication for urgent surgical intervention during the neonatal period is vascular compromise or progressive lymphedema of limb. This is treated by 1- or 2-stage excision of the constriction band with tissue rearrangement and closure with Z-plasty. The timing of surgery remains a controversial discussion, as it is not considered the optimal treatment of CCBS. Various evidence depict the pros and cons of both early and delayed surgery with early surgery generally shows better functional outcome but comes along with the theoretical downside of the lower operative tolerance in neonates.[2,4–7] The purpose of this case series was to report two cases of constriction band syndrome in neonates treated with urgent contracture release of the leg with multiple Z-plasty procedures.

Case Report

Patient 1, an 8-day-old male baby presented with main complaint of narrowing on both calves. There was no history of trauma. This condition has been observed since birth. The baby exhibited decreased movement in the affected limbs but otherwise showed normal activity and feeding. The patient is the second of twin babies and was delivered by sectio caesarea with the indication of oligohydramnion. The patient was born at 34 weeks gestational age on the first pregnancy, with 2100 g birth weight and 49 cm birth weight. Prenatal care was uneventful, with no significant findings on ultrasounds or maternal history of infections or trauma. The family history was unremarkable for congenital anomalies or genetic conditions. He was treated at the NICU and was on continuous positive airway pressure, then was referred to the Orthopaedic Department and Perinatology Care at our hospital for further treatment.

Physical examination revealed:
Right leg: A constriction band across the distal third of the shaft, swelling (-), necrotic tissue (+), the great toes are necrotic, the ankle joint is limited, and the toes are all constrained by PROM. Skin loss of about 0.5 cm with week pulses and prolonged capillary refill time.

Left leg: A constriction band at distal third of the shaft, edema (+) and ischemia (+) at the distal side of the constriction, absence of 1-4 toes, limited PROM of the ankle joint, and limited PROM of all toes. Skin loss of about 0.5 cm with week pulses and prolonged capillary refill time.

Figure 1. Physical examination of patient 1

Patient 1 was diagnosed with: 1) CCBS on the distal right leg; 2) CCBS on right 1st, 2nd, 3rd, 4th toes Patterson classification type 4; 3) CCBS on the distal left leg; and 4) CCBS on left 1st, 2nd, 3rd, 4th toes Patterson classification type 4

The patient was planned to undergo urgent multiple Z-plasty procedures for relieving structural issues on both bilateral legs. Pre-operative X-ray assessment was done for both hands and both foots. Abdominal ultrasound ruled out any other congenital anomalies. The result of the X-ray showed in Figure 2. The treatment to release bilateral contracture with multiple Z-plasty procedure is depicted in Figure 3.

Figure 2. Pre-operative X-Ray of patient 1

Figure 3. During surgery

Prior to surgery, the patient was anesthesized generally. Marking for the excision of the band was done and the flaps were planned. The angle was planned to be as closed as 60˚ to provide optimal balance for vascular supply of the flip of flap and the mobility of the base of the flap. The length was attempted ≤2 times the width of the base of the flap. Once the constricting band has been excised and the tissue flaps were mobilised, they are then transposed.

After surgery, the patient was observed and treated with standard post-operative procedure. Within days, the patient has recovered. Patient was then allowed to go home and continue treatment as outpatient clinic. Nine months after surgery, surgical site wound has healed with some remaining scar. The patient has also begun to walk (Figure 4).

Figure 4. Clinical picture of patient's extremities during follow-up visit

Patient 2 is a 2-day-old male baby presented with a swollen and cyanosed right leg. Complaints were experienced since birth; the patient is the first child delivered through caesarian section owing to breeching position. There was no unusual history during the mother's pregnancy. After being born, the patient immediately began to cry loudly and move around, but his right leg began to swell and became bluish. As the result, he was referred to another hospital for surgery.

On physical examination, constriction band was observed on distal third right leg with swelling and ischemic sign on the distal side of the constriction (Figure 5). Patient was then diagnosed with constriction band syndrome on the distal right leg Patterson classification type 2.

Surgical treatment to release contracture on bilateral legs with multiple Z-plasty procedure (Figure 6). The procedure was done with the same steps as the above.

Several days after surgery, the patient began to show imporovement. The patient was then allowed to go home. Eight months later, the wound has healed uneventfully with minor scar (Figure 7). The constriction bands were effectively released, and the transposed flaps healed appropriately without indications of infection, necrosis, or vascular impairment. Distal limb perfusion remained intact, exhibiting enhanced range of motion in the afflicted joints and clearance of distal edema. The patient demonstrated age-appropriate limb function, exhibiting no restrictions in movement or activity. The surgery site exhibited well-aligned scars with low stress, and both parents expressed satisfaction with the esthetic outcomes.

Figure 5. Physical examination of patient 2

Figure 6. Intra-operative condition of patient 2

Figure 7. Eight months after surgery

Discussion

Orthopaedic care of CCBS begins after birth and is focused on enhancing limb function and aesthetic appearance while decompressing the muscles, arteries, and nerves. As long as normal deep veins can be released by removing the band, the release procedure is generally considered to be safe. Constriction band syndrome does not cause an expansion of the oedema after birth.[5,8,9]

Z-plasty is a frequently employed surgical method for alleviating constriction bands in individuals with CCBS. The method operates by elongating the constricted tissue and transferring tension, so enhancing both functionality and aesthetics. Unlike other surgical techniques, Z- plas-

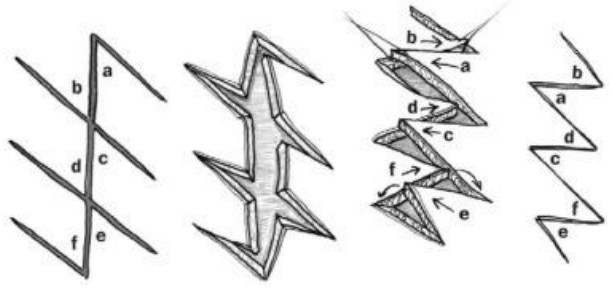

Figure 8. Illustration of Multiple Z-plasty[10]

ty is versatile, efficient, and can be performed in a single stage, making it ideal for emergencies.[10]

It remains in debate about the ideal time for Z-plasty and type of Z-plasty that are best for CCBS. Depending on the clinical manifestation, CCBS can be limb or life threatening. Therefore, the goal and urgency of the surgery will also depend on the patient's condition. Current evidence suggest that early surgical intervention resulted in generally better short- and long-term results. However, must be considered in doing an early surgical interference is that the patient will need an extra care because of possible lower surgical tolerance in the younger patients or neonates. For this reason, although the latest evidence suggested the benefits of early intervention, the approach of "faster is better" is still a topic of debate.[11,12]

In this report, we described two cases of very early surgical intervention of CCBS because of the limb threatening conditions. Urgent release must be done in these cases regardless their extremely young age (in our cases under 48 hours after birth). In our patients, early intervention was not associated with any comorbidities and still produced desirable results.

Another issue about z-plasty that is still arguable is regarding the stages of the surgery, whether single- or two-stage z-plasty procedure. The classic technique is the two-stage z-plasty procedure where release of the fibrotic band is done gradually. However, single-stage process is now preferred due to less exposure to anesthesia.

A review by Prasetyono et al confirmed that 1-stage circumferential ring-constriction release with skin recontouring using multiple Z-plasties is considered as a safe treatment for the correction of both circumferential and semi-circumferential congenital constriction ring. Range of patients' age was from 12 hours of life to 16 years old when they had circumferential ring constriction releasing surgery, either in a single stage or numerous stages. Three out of 12 patients had distal lymphedema in the lower leg. According to the review, while the authors did not specify any parameters for assessing limb function progression, they indicated that the limb exhibited normal development, appeared excellent, healed satisfactorily, experienced no functional impairment, recovered, and displayed full functionality following the release of 1-stage circumferential ring constriction.[13]

In their case report, Belani et al stated that in terms of wound healing, surgical complications, scar quality, and limb function, single-stage surgery with skin rebuilding by Z-plasty provides favourable results. In their patient, the distal ring was scheduled for excision after a suitable interval. This was to allow the proximal wound to heal and the leg edema to subside. Simultaneous excision of both rings are not performed as they were uncertain about the safety of this approach. Additionally, a single-stage operation does not require extra procedure as well as anaesthesia. They also emphasized on excellent outcome observed in a patient 17 after the surgery.[14]

Based on our report, the early and single stage Z-plasty approach showed to be successful, in which wound healed uneventfully with overall good function. Similar result has been reported by Hendra et al, that concluded that Z-plasty resulted in good outcome and could avoid unnecessary morbidity[3]. We also realised that although early constriction release is considered to have a better prognosis, rigorous preoperative evaluation is required to reduce the risk of operation, especially in neonates with underlying medical issues. However, evidence regarding the clinical outcomes of the Z plasty surgical method is still very limited. Additional research is required to compare the overall prognosis and establish criteria to guide management and support decision-making.

Conclusion

We reported two cases of an extremely early surgical intervention in an emergency setting of CCBS. Both patients were treated with single stage z-plasty procedure and evaluation 8 months after surgery showed no notable negative outcome and overall satisfying restults. Although long term monitoring is definitely needed, according to our findings in our patients, a single stage z-plasty maneuver have benefits that outweigh the risks that can be used for other similar cases.

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