SINGLE STAGE RELEASE SURGERY FOR CONGENITAL CONSTRICTION BAND IN A CLUBFOOT PATIENT MANAGED AT A TEACHING HOSPITAL IN UGANDA: A CASE REPORT

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ABSTRACT

Congenital constriction band or amniotic band syndrome is a rare condition with a prevalence of 1:11200. It is characterized by presence of strictures around a body part, commonly around the distal part of the extremities. These bands can be treated with a single or staged approach. This study presents the case of a 3 month old infant who presented with a type III constriction band localized on the right leg and surgery was indicated. A single stage multiple Z-plasty was performed. The postoperative course was uneventful and the outcome was satisfactory at 10 months of follow-up. A single-stage constriction band release approach provided satisfactory results; both functional and aesthetic results and is feasible in our setting.

Key words: Constriction bands, Single stage release, Stricture

INTRODUCTION

Congenital constriction band or commonly referred to as amniotic band syndrome or Streeter dysplasia is a rare condition characterized by congenital strictures that can be partial or circumferential. It is generally believed that there is no genetic predisposition of this condition (1) though the opposite has been implied in some literature (2).

The prevalence of constriction band is said to be about 1:11200 births (3). These bands are most common on the extremities especially the distal parts particularly the lower extremities (4), though have also been found on parts like the abdomen and chest (5-7).

The main intentions for treatment of congenital constriction band syndrome are improvement of function and aesthetics and this can be achieved by single release or a staged approach (1). Some studies recommend the correction of amniotic band syndrome be done in two or three stages of releasing the constricting ring because of the good outcome and since this technique is a safe way to eluding lymphatic and neurovascular embarrassment (8).

There is a plethora of literature that describes the release of congenital constriction band through the single stage (5,9-12) though most of it is from the western world which has a completely different structure of the health care system (13) from low income countries. Therefore the reason for sharing this case is to show the good outcome following single stage release of an amniotic band in a limited resource setting.

CASE REPORT

We present a case of a 3 month old boy that was seen in the Paediatric Orthopaedic Clinic with chief complaint of worsening right leg deformity since birth (Figure 1). The mother reported that the child had been born after a full-term pregnancy and normal delivery. This was the mother's fifth child and there was no history of illness or drug use during pregnancy. No one else in the family was similarly affected. At the age of one month, the child had been initiated onto correction for the associated clubfoot using the Ponsetti method but defaulted after two visits due to financial constraints.

Physical examination revealed a syndromic looking baby with the following abnormalities;

(i) Low set ears.
(ii) Absent ring finger on right with acrosyndactyly.
(iii) Constriction band syndrome involving the digits of the left hand.
(iv) Bilateral congenital talipes equinovarus.
(v) Circumferential constriction band on the right leg Dorsalis pedis and posterior tibial artery pulses were both palpable.

Figure 1

Child with constriction band of the right leg with associated clubfoot deformity

A diagnosis of type III congenital constriction band according to Patterson was made from both history and physical examination. Preoperatively, baseline blood investigations were done which included full

blood count and blood grouping tests. No radiological investigations were done (Figure 2).

Consent was sought from the mother. The mother was counseled about the need for surgery to release the constriction band on the right leg plus the complications associated with the surgery; to which she consented. .

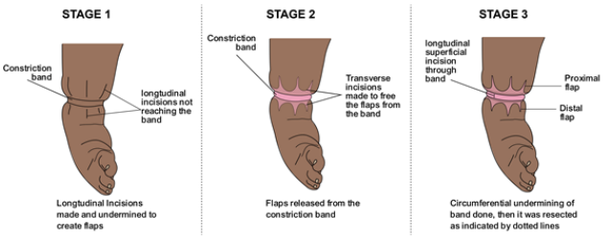
On the day of the operation, the surgical site was marked, aseptic technique was observed and a sterile tourniquet was applied mid-thigh. The surgery started with making of 1cm long incisions proximally and distally that were equidistant 1 cm apart. The incision extended just close to the constriction. This was followed by undermining of the subcutaneous tissues to free the flaps and release them from the band. Transverse incisions were then made to create independent flaps. This was followed by dissection of the constriction band using a dissecting scissor to undermine the underlying fascia taking care not to injure the traversing nerves and blood vessels underneath the deep fascia. The band was finally resected away and the flaps were repaired in a z-plasty fashion using a vicryl 2-0 suture then wound dressing was applied. Post operatively the child was managed with oral antibiotics plus analgesics and foot was elevated on a pillow. The swelling of the foot resolved after 2 days. No postoperative complications were faced and the patient was discharged on the seventh day post operation.

Figure 2

2a) Intraoperatively after resection of the band.
2b) Illustration describing the procedure

2a

2b



The first review date was scheduled 2 weeks from the discharge date. The patient returned for review in the Paediatric Orthopaedic Clinic and we found the wound was healed, the lymphedema had not recurred and distal neurovascular status was intact. There was no attempt at stricture reformation.

The child was then referred to the Club Foot Clinic for correction of the clubfoot deformity and also requested to return for monthly reviews in the Paediatric Orthopaedic Clinic for the first 2 months. The patient missed all the appointments due to lack of transport fare to the hospital but communication with the parents about the state of the child continued via phone. Child was seen at 10 months after surgery with a healed scar and no attempt at stricture reformation (Figure 3). The child is carrying on with club foot management at a nearby hospital.

Figure 3

3a) at 2 weeks post op 3b) at 10 months post op

a b

DISCUSSION

Constriction bands can be in form of superficial or deep bands (9). The severity of the bands is characterized in grades with grade 1 being subcutaneous, grade 2 is at the level of the fascia but doesn't compromise vascular supply of the distal part of the limb, grade 3 is at the level of the fascia and compromises lymphatic and vascular supply to the distal part. Grade 4 includes all congenital amputations (4).

The strength of this study is that we prospectively followed up the child for ten months which is a fairly long time. The limitation however was our inability to observe the outcome following management of the clubfoot since the parents were constrained for finances. As indicated above these bands can sometimes be deep enough to cut off the normal vascular and lymph return of the extremity therefore resulting in chronic edema or ischaemia of the limb hence the need to fix them as early as possible (14).

The theory most attached to the genesis of amniotic bands is that by Torpin (15) where he describes that rupture of amnion permits the fetus (or part of) to enter into the chorionic cavity with tethering of extremities and the resulting oligohydramnios produce club feet and other positional anomalies so the ruptured amnion act as a band around the limb leading to constriction band in the limb or amputation in uterus (9).

Patterson classified the constriction bands into four subtypes depending on the depth and the clinical effects caused by the bands (16).

(i) Type I refers to a ring constriction that might be superficial or deep.

(ii) Type II includes constriction band with distal deformity such as lymphedema.

(iii) Type III includes the presence of acrosyndactyly.

(iv) Type IV includes complete intrauterine amputation.

The child in this case had a type III constriction band on the right leg which tallies with the presentation listed in the case description.

Surgery to the constriction bands can be via a single or staged approach that is two or three stages. The authors who advocate for a staged approach reason that it reduces the likelihood of lymphedema and vascular complications in distal parts (8,17). Proponents for single stage approach base on the studies that show its success but also explain that single stage approach protects the patient from double exposure to anaesthesia and complications associated with surgery (9, 11).

In Uganda, some patients will not comply with further appointments for serial excision therefore single-stage correction may well be the procedure of choice to reduce the costs patients have to incur from multiple trips to the hospital; case in point, is the patient presented in this report who due to financial constraints failed to return for multiple appointments.

In this study, single stage surgery was done and patient had a good outcome with a good scar, with no attempt at reformation of the constriction band and no swelling of the part distal to the released band.

In conclusion single stage release surgery can successfully address constriction band syndrome even in limited resource setting if done well.

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Conflicts of interest: None.

REFERENCES

1. Adu, E.J. and Annan, C. Congenital constriction ring syndrome of the limbs: A prospective study of 16 cases. *Afr J Paediat Surg.* 2008; **5**(2):79.
2. Blyth, M. and Lachlan, K. Amniotic bands in paternal half-siblings. *Clin Dysmorphology.* 2010; **19**(2): 62-64.
3. Cignini, P., Giorlandino, C., Padula, F., Dugo, N., Cafà, E.V. and Spata, A. Epidemiology and risk factors of amniotic band syndrome, or ADAM sequence. *J Prenatal Med.* 2012; **6**(4):59.
4. Ozkan, K., Unay, K., Goksan, B., Akan, K., Aydemir, N. and Ozkan, N.K. Congenital constriction ring syndrome with foot deformity: two case reports. *Cases J.* 2009; **2**(1): 6696.
5. Prasetyono, T.O. and Sitorus, A.S. A review on the safety of one-stage circumferential ring constriction release. *Intern Surg.* 2015; **100**(2):341-349.
6. Jones, S. and Sinclair, L. Amniotic bands: an unusual chest lesion. *J Royal Society Med.* 1988; **81**(3): 168-170.
7. Fawzy, M., Goon, P. and Logan, A. Abdominal constriction bands are a rare complication of the amniotic band syndrome. *J Plastic, Reconstructive Aesthetic Surg:* 2009; **62**(3):416.
8. Stevenson, T.W. Release of circular constricting scar by z flaps. *Plastic Reconstructive Surg.* 1946; **1**(1):39-42.
9. Das, S.P., Sahoo, P., Mohanty, R. and Das, S. One-stage release of congenital constriction band in lower limb from new born to 3 years. *Indian J Orthop.* 2010; **44**(2):198.
10. Sanjaya, I.G.P.H., Hamid, A.R.R.H. and Samsarga, G.W. One stage release of circumferential constriction band in upper limb with multiple Z-plasty technique: a case report. *Indonesia J Biomed Sci.* 2019; **13**(2): 63-66
11. Greene, W.B. One-stage release of congenital circumferential constriction bands. *J Bone Joint Surg.* 1993; **75**(5): 650-655.
12. Visuthikosol, V. and Hompuem, T. Constriction band syndrome. *Annals Plastic Surg.* 1988; **21**(5):489-495.
13. Ronderos-Dumit, D., Briceno, F., Navarro, H. and Sanchez, N. Endoscopic release of limb constriction rings in utero. *Fetal Diagnosis Therapy.* 2006; **21**(3):255-258.
14. Chaturvedi, V., Agarwal, G., Narula, I.M. and Singh, H. Congenital circular constriction band syndrome. *Indian J Pediat.* 1971; **38**(2):74-75.
15. Torpin, R. Amniochorionic mesoblastic fibrous strings and amnionic bands: associated constricting fetal malformations or fetal death. *Amer J Obstet Gynecol.* 1965; **91**(1):65-75.
16. Basheer, S.M., Karashi, A.R. and Abdulbasith, M. Single stage release of bilateral amniotic band syndrome. *Bahrain Med Bull.* 2019; **41**(1):38-41.
17. Choulakian, M.Y.and Williams, H.B. Surgical correction of congenital constriction band syndrome in children: Replacing Z-plasty with direct closure. *Canadian J Plastic Surg.* 2008; **16**(4): 221-223.