

# Constriction Band Syndrome of the Left Leg with Late Presentation: A Case Report

Abdullahi Galdima Bello<sup>1</sup>; Dr. Altine Aliyu Nuradeen<sup>2\*</sup>;  
Lukman Olalekan Ajiboye<sup>3</sup>; Abdulazeez Mukhtar<sup>4</sup>; Aminu Hassan<sup>5</sup>;  
Abdulrahman Hassan Muhammad<sup>6</sup>

<sup>1;2;3;4;5;6</sup>Department of Orthopaedics and Trauma, Usmanu Danfodiyo University Teaching Hospital, Sokoto, Nigeria

Corresponding Author: Dr. Altine Aliyu Nuradeen<sup>2\*</sup>

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**Abstract:** Congenital constriction band is uncommon and usually requires no treatment for the milder form. The surgical release of the band through Z plasty is promising in those requiring intervention to avoid vascular complications and loss of the affected limb. Early presentation is therefore crucial. The purpose of this study is to report a male baby with late presentation who had amputation. The 1-week-old male baby presented with a swollen, dark, and painful left leg with CBS in the middle of the left leg that warranted amputation with a subsequent uneventful postoperative period. This case has highlighted the need for early presentation of such cases for assessment and prompt treatment to avoid complications, including loss of the affected limb.

**Keywords:** Constriction Band Syndrome, Leg Constriction Band, Amputation, Syndactyly.

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## I. INTRODUCTION

Constriction band syndrome (CBS) is an uncommon pathology in newborns with a still unclear etiology [1]. In literature, different terminologies can be used to refer to this condition, like Streeter dysplasia, amniotic constriction bands, annular defects, and anomalous bands. It has varying clinical presentations from simple constriction rings of the digits, syndactyly, acrosyndactyly, to more severe malformations of the head, face, and trunk with anencephalia, encephalocele, microphthalmia, cleft lip, hydrocephalus, thoracoschisis, gastroschisis, and extra-thoracic heart [2]. The limb lesion is usually asymmetric, and the incidence is 1:2000 to 1:15000 live births with an equal male to female ratio [3]. There is a high incidence among the premature and low weighted babies [4]. Treatment with constriction release through Z plasty is the common surgical procedure for early presentation [5]. We reported this case because of its rarity and the late presentation, which may complicate management.

## II. CASE REPORT

A week-old male neonate presented with circumscribed narrowing of the left leg and deformed fingers since birth. No hospital treatment was sought, and the left leg gradually started swelling and becoming dark, which prompted the parents to present to the hospital. There was deformity of the right hand with fusion and flexion deformity of fingers bilaterally. No other associated anomaly was noticed. The antenatal period and labour were uneventful. No history of similar condition in the family. The examination findings were a healthy-looking neonate, Pinkish, febrile, anicteric, acyanosed. Heart Rate=110b/m and Heart sound=1 & 2, no murmur. Chest, Abdomen, urological, and neurological examinations were essentially normal.

Musculoskeletal system: Circumferential constriction around the middle of the left leg with swollen, darkened, and non-sensory part of the leg distal to the constriction (Figure 1A). There was a presence of bilateral upper limb acrosyndactyly (Figure 2).

A diagnosis of CBS was made, and investigations to prepare the patient for operation and to rule out other congenital anomalies, which include abdominal USS, Echocardiography, and Haematological profiles, were done. Diagnosis of a congenital constriction band of the left leg was made. The patient was optimized, and below-knee

amputation was done (Figure 1B & 1C). Management of syndactyly was deferred for a later time in conjunction with the plastic and reconstructive surgery specialty. The patient was discharged 5 days after an uneventful postoperative period and was to be followed up on periodic clinic visits.



Fig 1 [A] Left Leg CCB with Imminent Gangrene; [B] and [C] After Amputation



Fig 2 Right Upper Limb Syndactly/Acrosyndactly

### III. DISCUSSION

The congenital constriction band syndrome (CBS) is rare, and its etiology is not completely known and it mainly presents sporadically [1,4,6]. Although the literature mentioned equal gender occurrence, our study has a male baby presenting. Presentation is usually at birth and varies with the degree of constriction, location, and associated deformities. The time of presentation is also important in the management, as late presentation may risk limb loss, as seen in our patient who presented 1 week after birth. However, the degree and severity of the constriction may be more important because a study by Jaiman R et al. reported satisfactory results after Z-plasty constriction band release in a 4 year old child who presented with Congenital CBS [7]. Another study showed one-stage 24 constriction bands release in 19 patients with excellent results [8]. Our patient developed imminent gangrene after late presentation, probably due to the severity of the constriction band. The CBS varies in its type and level of soft tissue affection, ranging from only the skin, subcutaneous tissues to a deeper

location affecting vascular supply and lymphatic channels, resulting in chronic limb oedema [9]. The CBS could be a partial or complete ring, single or multiple, superficial or deep, and can occur anywhere in the limb, resulting in intrauterine limb gangrene and amputation [8,10]. The treatment options for the CBS include observation in superficial ring that is confined to the skin to Z-plasty in deep rings with risk of vascular compromise [3,11]. To minimize recurrence and risk of further vascular injury, complete excision of the constrictive tissue with a 1 to 2 mm skin margin and soft tissue with removal of dense fibrous tissue is advised.

### IV. CONCLUSION

Congenital constriction band syndrome of the limb may present either in mild form or severe form requiring intervention which usually results in good outcome when presented early. The late presentation particularly with associated vascular compromise may threaten loss of limb by gangrene and subsequent amputation.

#### ➤ Conflict of Interest

The author declares no conflict of interest

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