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Clinico-pathological Spectrum of Congenital Constriction Ring Syndrome and It's Management

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ABSTRACT

Objective: To describe in detail, the spectrum of congenital Constriction Ring Syndrome (CRS), or amniotic band syndrome and its unique presentations. Since it is a relatively rare condition, and we have compared with existing literature, the knowledge of such a vast spectrum will help the clinicians in diagnosis and treatment.

Method: The study was done over a period of 3-years. Data regarding patient's demographic profile, surgical procedure and outcomes were collected from previous records from January 2020 to April 2023. Demographic characters, limb involvement, distal deformities, surgical procedure etc were evaluated.

Result: Total of 48 rings in 33 patients were analysed. minimum and maximum reported age range was newborn to 16-years, mostly within 1-year, with female preponderance. Lower limb was affected more than upper limb, with a right sided predominance. Lymphoedema was most common complications, and all patients were managed surgically.

Conclusion: CRS is a rare entity. Timely and adequate management must prevent lifelong deformity. Detailed knowledge of spectrum and treatment protocol will help the clinician in satisfactory management of such patients.

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INTRODUCTION

Constriction Ring Syndrome (CRS) was first described by Van Helmont in 1689, as intrauterine amputation and Montgomery in 1832 as free strands of tissue. Estimated incidence is approximately, 1 in 10,000 live births with equal sex distribution. Cutaneous constriction ring or CRS or Amniotic Band Syndrome is a rare congenital anomaly, occurs equally in male and females, with an incidence of 1 in 1200 to 1in 15000 live births. Occurs in intrauterine period of the foetus, with unknown aetiology. Van Helmont was first to describe CRS as intra-uterine amputation, in 1689. However, Montgomery described CRS as free tissue strands. Although, Constriction ring can occur at any part of the body of developing foetus, but it is commonly found over the upper or lower limbs. Depending on the extent, severity and clinical features CRS can be grouped in various categories.

Patterson classification is most widely used classification for CRS.[1]

Type 1. Simple constricting ring
Type 2. Constricting ring with deformity of distal part
Type 3. Constricting with fusion of distal parts
Type 4. Complete intrauterine amputation

Constriction ring develops as a fibrotic band from amniotic membrane which gradually encircles the effected part. As a result of encircling, the band exerts significant amount of pressure which can cause lymphatic obstruction, arterial obstruction, venous drainage obstruction, ischaemic necrosis,

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under-development of limb, intra-uterine amputation, limb gangrene etc.

CRS may present as isolated or syndromic deformity. Craniofacial clefts, club hand or foot, CTEVS, cutis aplasia etc are common congenital anomalies associated with CRS.[2,3]

Farrukh M *et al.*, reported a case of CRS, of a 25-days-old neonate. Child presented with bilateral hand acro-syndactyly and in-utero, partially amputated deformed foot with congenital constriction ring on leg. The child was managed with release of constriction ring by multiple Z-platy procedure.[4]

Another study by Korhan O *et al.* They reported cases of 6-months and 8-months-old babies who presented with unilateral club foot and congenital constriction ring band. Both patients were managed by surgical release of the ring and cast application for the club foot.[5]

Cowell and Hensinger published 14 cases of constriction ring syndrome. Patients had different locations of the constriction ring, like thigh, leg, ankle and foot. Most of the patients presented with foot deformity, which was managed by Ponseti method of serial casting. None of their patient had CRS above the thigh. According to them incidence of club foot deformity is between 12–56% in patients presenting with congenital CRS. The association of club foot and congenital CRS demands thorough examination, screening and management.[6,7]

Another case series published by Wahegaonkar *et al.,* showed study of 12 patients, with age range of 9 days to 5-years. Patients had CRS in upper limb as well as lower limb. Upper limb involved fingers, forearm, and arm whereas, lower limb involved toes, foot and leg. All patients were managed by surgical release of ring.[8]

Masmoudi K *et al.,* reported a case of a neonate, born with recent fractures of both leg bones with an amniotic band encircling the limb. Child did not undergo surgical intervention. Non-operative treatment with splintage was successful.[9]

Tadmor *et al.* reported a case of a new-born with spontaneous limb amputation in utero due to a constriction ring encircling both lower limbs.[10]

Zych *et al.*, in 1983, reported a case of congenital CRS over fingers associated with pseudoarthrosis with posterior tibial nerve entrapment of leg. The patient was managed with fasciectomy and nerve release.[11]

Martinot-Duquen Published case of CRS of foot with bilateral amputation of great toe, along with acute ischaemia of distal part. Patient was managed with debridement and external fixation.[12]

Tanguy, 1995 also reported a case of CRS with bilateral amputation of hallux. Agashe, 2011 published case of CRS with acro-syn-dactyly of both hands. The vascular compromise and significant lymphoedema resulted in amputation of distal part.[13,14]

Ho Christine, 2014 presented a case of pseudoarthrosis forearm along with CRS and acute ischaemia. The patient was treated by decortication and immobilisation.[15]

Firdous K *et al.,* Conducted a retrospective study on 27 patients of congenital CRS. Male female ratio was 11:16, with age range of 1 to 18-years. Upper and lower limbs were equally affected. Commonly associated anomalies were lymphoedema, acro-syndactyly, hypoplastic hand and foot etc.[16]

METHODOLOGY

The study includes both prospective as well as retrospective data, over a period of three years. We had total 33 patients, who visited for consultation in the department of plastic surgery of our institute.

We had operated all 33 cases of CRS in last three years. The demography and other features of clinical profile are shown in table 1.

OBSERVATION

Age- The age range of the patients was one day to 16-years. The minimum age at which, we operated was one day (just within few hours after birth), such an early and urgent intervention is not reported till now.

Sex- Among our 33 patients, 20 were female and 13 were male. We noted female preponderance in our study.

Age of presentation- Most common presentation was within one year (n = 14, 42.42%), followed by 1 to 3 years (n = 13, 39.39%) followed by more than 3-year (n=6, 18%). (Figure 1).

Limb involvement- Out of total 33 patients, 23 patients had lower limb involvement. Hence our study

Table 1: The demography and other features.

S.No.	age	sex	diagnosis	Distal changes	treatment	Post-op complications
1	7 mth	F	Right leg	with toe atrophy	Surgical release	Nil
2	7 Y	M	B/L leg	foot hypoplasia	Surgical release	Nil
3	9 mth	M	Right arm and index finger	Hypoplasia digits	Surgical release	Nil
4	1-mth	M	B/L foot	Edema	Surgical release	Nil
5	8-mth	M	L foot	Toes hypoplasia	Surgical release	Nil
6	1Y	M	Right foot	Nil	Surgical release	Nil
7	1Y	F	B/L foot	Edema R>L	Surgical release	Nil
8	1.5Y	F	Left hand	Acrosyndactyly	Surgical release	Nil
9	16Y	F	Left hand residual ring	Scarring	Surgical release	Nil
10	3Y	F	Right foot	hypoplasia	Surgical release	Nil
11	7Y	F	B/L foot	Hypoplasia toes	Surgical release	Nil
12	6Y	F	B/L forearm	Syndactyly with hypoplasis	Surgical release	Nil
13	2Y	F	L leg	Hypoplasia toes	Surgical release	Nil
14	16Y	F	R leg	Nil	Surgical release	Nil
15	7 mth	M	B/L leg+ Left hand	Foot hypoplasia	Surgical release	Nil
16	1Y	F	Right leg with Left hand	Acrosyndactyly left hand with hypoplasia	Surgical release	Nil
17	3 mth	M	Right leg B/L CTEV	Hypoplasia foot B/L CTEV	Surgical release	Nil
18	5 mth	F	Left middle finger	Near total amputation	Surgical release	Nil
19	12Y	F	R forearm With ulnar nerve compression	Nerve compression	Surgical release	Nil
20	2 mth	M	B/L leg	hypoplasia	Surgical release	Nil
21	1 mth	M	Right Leg	Edema	Surgical release	Nil
22	1Y	F	Right hand	Acrosyndactyly	Surgical release	Nil
23	1.8Y	F	Right Leg acrosyndactyly left hand	edema	Surgical release	Nil
24	1Y	M	B/L hand with syndactyly	syndactyly	Surgical release	Nil
25	9 mth	F	Left leg	Edema	Surgical release	Nil
26	1 Y	F	Rt forearm	Hypoplasia fingers	Surgical release	Nil
27	1.5 Y	M	B/l foot	Hypoplasia	Surgical release	Nil
28	3 mth	M	Right hand	Nil	Surgical release	Nil
29	1 yr	M	Right thigh abdomen	lymhoedema	Surgical release	Nil
30	2 y	F	Left leg	Hypoplasia	Surgical release	Nil
31	1 day	F	B/L foot	Impending gangrene	Surgical release	Nil
32	4 mth	F	B/L leg	Edema	Surgical release	Nil
33	10 mth	F	Right leg Right hand	nil	Surgical release	Nil

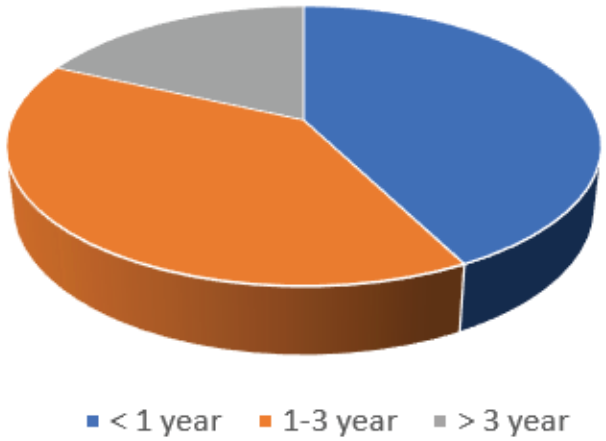

Figure 1: Showing age of presentation.

contradicts the study of Firdous K *et al.*, as both upper and lower limb were equally affected in their study.

On further categorisation of lower limb in to right, left and bilateral, 10 patients (43.47%) had right sided lower limb involvement, 9 patients (39.13%) had bilateral involvement and only 4 patients (17.39%) had left lower limb involvement. Hence lower limb of right side was found to be more affected than left side. Unlike lower limb, both left and right sides were equally affected in upper limb. 6 patients (42.85%) had right hand involvement, 6 patients (42.85%) had left upper limb involvement, and 2 patients (14.24%) had bilateral upper limb involvement (Figure 2).

In lower limb, most common involvement was leg (n = 14, 60.86%), followed by foot (n = 8, 34.78%), followed by thigh (n = 1, 4.34%). In upper limb, most common site of involvement was hand, (n = 12, 85.71%), followed by forearm (n = 3, 21.42%), and only one patient (7.14%) had CRS over arm.

Distal deformities- Distal deformities were categorised into 5 major groups, hypoplasia or atrophy or complete amputation (group 1), lymphoedema, (group 2), grossly deformed hand or foot (group 3), (Acro syndactyly or CTEV) and neurovascular deficit (group 4), and no distal deformity (group 5). Among all patients, 16 patients (48.48 %) were found in group 1, followed by 7 patients (21.21%) in group 2, 3 patients in group 3 (9.09%), 2 patients in group 4 (6.06%) and 6 patients in group 5 (18.18%). Hence, most common distal deformity was growth related, which followed by lymphoedema (Figure 3). Familial tendency, known risk factor, or consanguity was not observed in our study.

All patients in our study were managed by surgical intervention, none of them underwent conservative management like splinting, cast etc. unlike study of Masaudi *et al.,* where they managed the child conservatively. Post-operative period was uneventful and none of the patients developed complications like stich line infection, flap necrosis, neurovascular damage etc.

Along-with the data of last three years we also want to highlight our two cases which were rare, unreported and unique in their presentation. A full term born female child brought after few hours of birth with complaints of deformed and shrivelled right foot along with black patches over the dorsum of foot (Figure 4). And a tight band on the left leg. On examination of right foot, Foot was showing features of gangrene. Foot was cold and a tight, full thickness, bone deep, circumferential constricting band was found on the distal part of the leg. Skin

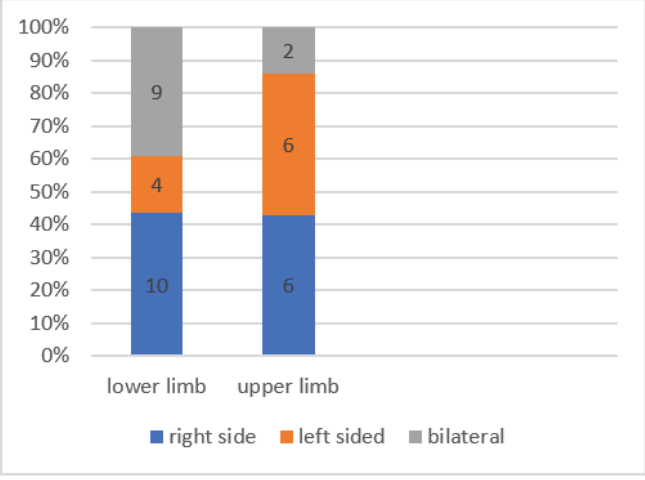

Figure 2: Showing sides of upper and lower limb involvement.

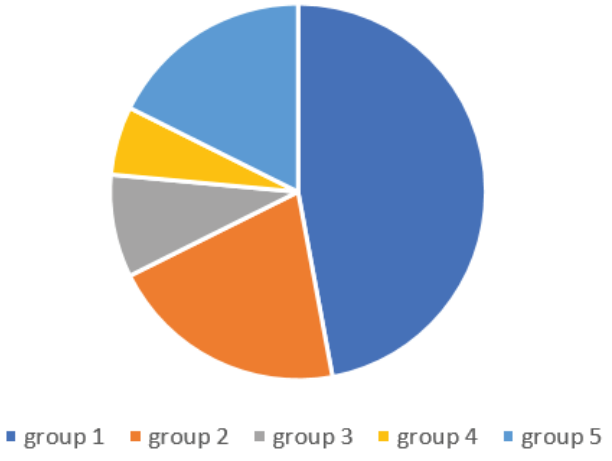

Figure 3: Showing percentage of distal changes.

Figure 4: Cutaneous Constriction ring on bilateral foot.

necrosis was present on the dorsum of foot. Foot was oedematous and shrivelled. Capillary refilling on the toes was significantly delayed, >6 seconds. On examination of left leg also a tight, full thickness, bone deep, circumferential constricting band was found on the distal part of the leg. Foot was cyanosed and capillary refilling was delayed. There were not any congenital anomalies (Figure 5).

Child was immediately investigated for fitness of anaesthesia and surgery was planned under general anaesthesia. Parents were explained about the procedure and its urgency. Informed consent was taken for surgery. Surgical release was done under general anaesthesia on the same day by multiple Z-plasty on both the legs in single sitting (Figure 6). Surgery was performed by two team approach to reduce anaesthesia time as much as possible. Surgery of both legs was completed within 30 minutes, and child was safely extubated. Post-operatively gangrenous changes and compressions changes of

Figure 5: Constriction ring showing the changes.

Figure 6: CRS release by multiple Z-plasty.

foot were found to be drastically reverted. Long term follow up was excellent and bilateral foot regained full growth without and neurological compromise (Figure 7).

We could not find any report mentioning newborn with bilateral foot CRS along with impending gangrene of both feet. The impact of surgical release was so effective that immediately the vascular compromise reversed, and blood circulation established. We could appreciate the reversal of cyanosis in both feet. We were able to salvage both limbs by early surgical release. Hence clinician should have high suspicion for potential limb loss while dealing such cases and should perform urgent intervention, without further delay.

Our second case (double CRS with entrapped lymphoedema) is also a unique presentation. Patient was a 1-year-old male child referred to us with diffuse swelling in inguino- scrotal area. There were no complaints of foot deformity or difficulty in walking. Parents were also unaware of existence

Figure 7: Post-operative image showing full growth of bilateral foot and well-developed foot fat pads.

Figure 8a: Inguinoscrotal and thigh lymphoedema.

Figure 8b: Tight band over lower trunk and thigh entrapped lymphoedema.

Figure 8c: Thigh band released.

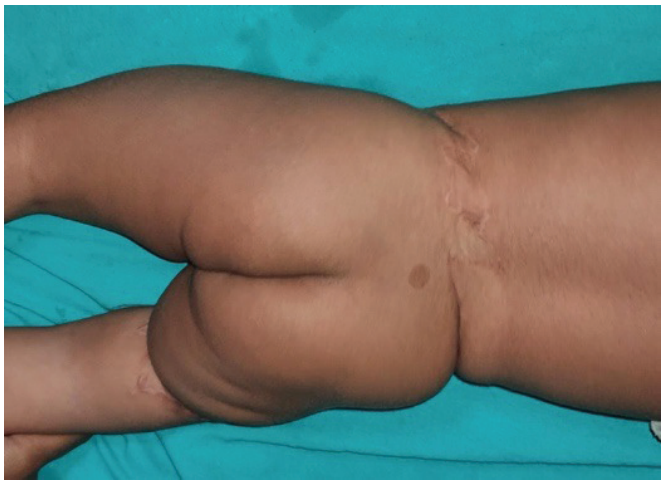

Figure 8d: Trunk band released.

of any unusual band or ring on the child's. body. Child was thoroughly examined and evaluated. On exposure a diffuse swelling was found on right inguino-scrotal area and right buttock area. A constriction ring was found on the right leg, which was more prominent and deeper on posterior aspect. Another tight, constriction ring band was noticed in suprapubic region which was continued posteriorly in lumbar region. The diffuse swelling was found to be associated with poor lymphatic drainage. There was no sensory, motor or vascular deficit distally. Child was investigated for anaesthesia fitness as well as to

rule out other internal deformities of bone as well as soft tissue. External genitalia were found to be normal, except the swelling. X-ray bilateral lower limb, x-ray spine, ultrasound abdomen and pelvis were done, and found to be normal. Child was manged by surgical release of both the rings under general anaesthesia. Ring of right thigh was released completely whereas supra-public ring was released partially. Post-operatively child recoverd well, swelling and edema reduced and is waiting for release of remaining part of the ring in next sitting (Figures 8 (a to d)).

Frank S *et al.* also reported a case of CRS over trunk, managed by z-plasty procedure. But it was an isolated ring without distal compression effect. However, our case is unique in its presentation. Double CRS over thigh and abdomen with entrapped lymphoedema is not yet reported.[17]

DISCUSSION

Exact cause of CRS is not known due to lack of supportive theories. However, two theories are considered as a potential cause. According to intrinsic theory deficiency of germ cell lead to such deformity and malformation. Whereas according to extrinsic theory combination of amniotic bands and external compression leads to CRS.[18]

Extrinsic theory is most widely accepted and is further supported by Tropin *et al.,* who stated that combination of early rupture of amnion, formation of fibrous bands, reduced amniotic fluid, and excrusion of foetal parts especially limbs are associated with formation of CRS.[19]

According to Streeter, 1930, bands are formed due to macerated sheets of epidermal tissues, which encircles are forms band like structure.[20] Intra-uterine trauma theory was given by Kinol *et al.*, according to them, it is intra-uterine trauma which causes cessation of blood supply to the marginal sinuses of limb spaces which in turn leads to acrosyndactyly, hypoplasia, constriction, amputation, developmental arrest etc.[21]

CRS is a condition which almost always needs surgical intervention, depending on the severity of the deformity. Incomplete or loose rings without any distal lympho-vascular compromise can be delt on elective basis for cosmetic correction. However complete, tight rings, associated with limb deformity or distal lympho-vascular compromise require urgent intervention to treat the existing deformity and to minimise the potential deformity, which may occur in presence of continuous pressure exerted by the band. In presence of continuous and unrelieved pressure Foot or hand may become oedematous, atrophic, deformed, gangrenous or even amputated.[22]

Indication for surgery in CRS- CRS is associated with both cosmetic as well as physiological changes. Hence indications of surgery include both cosmetic and physiological conditions. Unsightly scar band and unnatural appearance are cosmetic indications for surgery CRS. However, Distal deformity, hypoplastic body parts, lympho-neurovascular compromise, potential limb deformity etc. are various physiological indications for surgery of CRS.[23]

Aim of surgery- ring should be released sufficiently and up to adequate depth/ skin fold thickness, so as to achieve following

- Relieve pressure effect completely
- Retain lympho-vascular flow
- To achieve optimum nerve conduction distally

Techniques of surgery- Multiple Z-plasty or W-plasty are most commonly used techniques for release of CRS because of their ability to lengthen the constricting band and to re-orient the scar.

However, other techniques include rectangular plasty, excision and debulking, subcutaneous fat advancement flap, dermo-fat flap etc. CRS once diagnosed, shoud be treated as early as possibe, especially those involving complete or almost full circumference of limb.[24]

CONCLUSION

Constriction band syndrome are rare conditions and occur due to unknown etiology and present as various forms. Commonly involved body parts are limbs, and trunk, occasionally. It may or may not be associated with various syndromes. Constriction ring syndrome is a rare condition which has multiple presentations, ranging from simple and uncomplicated to complex and complicated. Neonates presenting with CRS should be thoroughly examined, especially to rule out any neuro vascular or lymphatic obstruction. And if found, urgent release should be priority to prevent permanent limb deformity and limb loss. CRS are

known to cause significant lifelong morbidity and deformity hence all children presenting with CRS should be treated with priority.

Declaration of Patient Consent

The authors certify that they have obtained all appropriate patient consent forms from the patients or parents. Consent for surgery, image, publication etc were taken.

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Conflicts of interest

Authors declare no conflicts of interest.

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