Congenital constriction ring syndrome of the limbs: A prospective study of 16 cases

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ABSTRACT

Background: The congenital constriction ring syndrome is characterised by fibrous bands that encircle, strangle and even amputate parts of the foetus. It is a common condition amongst Ghanaian patients, but data on it is quite scanty. **Materials and Methods:** A prospective study of patients presenting at a plastic surgical clinic in Ghana with the characteristics of the congenital constriction ring syndrome was undertaken. The patients were examined clinically and the findings recorded. An x-ray and clinical photograph of the affected limbs was taken. Treatment required several staged operations. Surgical correction of the constriction ring was done by excision and Z-plasty to prevent or alleviate lymphoedema, separation of distally fused digits and skin grafting of defect. **Results:** Sixteen patients made up of 10 males and six females were seen. The age at presentation ranged from nine days to 12 years with a mean age of 14.6 months. Twenty-two limbs were affected, made up of four right upper limbs, six left upper limbs, seven right lower limbs and five left lower limbs. In the upper limb malformations involved 42 digits; in the lower limb malformations involved 33 toes, one foot and five legs. Four main types of lesions were found: constriction rings, intrauterine amputations, acrosyndactyly, and simple syndactyly. **Conclusion:** Congenital constriction ring syndrome is of uncertain aetiology and could cause morbidity in the newborn. The syndrome and its complications are amenable to corrective surgery with good results. Early intervention is desirable for a successful outcome.

Key words: Acrosyndactyly, amputations, congenital constriction rings

INTRODUCTION

The congenital constriction ring syndrome embraces a group of abnormalities which occur in a variety of combinations, mainly affecting the limbs and rarely the trunk and head. It is associated with fibrous bands that encircle, strangle and even amputate some parts of the foetus.[1] Patterson[2] classified the varieties as follows: (a) simple constriction rings (b) constriction rings accompanied by deformity of the distal part, with or without lymphoedema, (c) constriction rings accompanied by fusion of distal parts, ranging from mild to gross acrosyndactyly, and (d) intrauterine amputations.

According to Patterson,[2] to diagnose this syndrome a patient must have two or more of the above. The deformities are the result of a cascade of events that follow an intrauterine disruption.[3] The aetiology of the syndrome is not known; three main theories attempt to explain the specific cause of the disruption: the intrinsic theory,[4] the extrinsic theory[5] and the intrauterine trauma theory.[6] The condition is not uncommon in Ghanaians but data on it is quite scanty.

MATERIALS AND METHODS

Patients presenting with two or more of the characteristics of the congenital constriction ring syndrome were entered into the study. Patients with congenital amputations were entered into the study if they were transverse; ray amputations were excluded from the study unless they also had other features of the syndrome.

The study began in June 2003 at the Reconstructive Plastic Surgery and Burns Unit of Korle Bu Teaching Hospital, Accra where two patients were seen, and continued at the Reconstructive Plastic Surgery Unit of Komfo Anokye Teaching Hospital in Kumasi, from November 2004 to June 2007, where 14 patients were seen.

The patients were examined clinically. A complete family history, thorough pregnancy history including information about medical illnesses, medications, alcohol and substance abuse, onset and vigour of foetal movements, delivery and neonatal adaptations were

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taken. The site of the malformation and any associated internal or external anomalies were recorded. X-rays of the affected limbs in AP and lateral positions were also taken.

All patients had pre-operative and post-operative photographs taken. Surgical procedures performed included excision of constriction rings followed by Z-plasty usually done before the first year of life. Patients with acrosyndactyly had their fingers separated; the site of tissue loss was either grafted or allowed to heal secondarily. This was done within the first year. Most of these procedures were staged with intervals of three months.

One patient with massive swelling distal to a circumferential constriction ring to the second toe [Figure 1] had immediate decompression of the fluid-filled cyst with release of the constriction and resurfacing of the excised scarred tissue with the extra skin. This was done in the neonatal period.

RESULTS

There were 16 patients with 22 affected limbs; 10 were males and six females. The age at presentation ranged from nine days to 12 years with a mean age of 14.6 months. The affected limbs were right upper limbs (4), left upper limbs (6), right lower limbs (7) and left lower limbs (5) [Table 1]: there was no family history of constriction ring syndrome in any patient.

In the upper limb 42 digits were involved; the hand, forearm and upper arm were never involved. In the lower limb 33 toes were involved; the foot was involved once, the leg five times [Figure 2] and the thigh once [Figure 3].

Constriction rings involved 12 sites in the lower limbs and five sites in the upper limb [Figures 4, 5]. In the upper limb only the digits were involved namely, the thumb (1), index finger (1), middle finger (1) and, the ring finger (2). The little finger was never involved. In the lower limb constriction rings were found on the thigh (1), the distal third of the leg (5), the foot (1), and the toes (5).

Table 1: Sites affected by constriction ring syndrome

Upper limb		Lower limb	
Fingers	**42**	Thigh	1
Index	10	Leg	5
Ring	10	Foot	1
Middle	10		
Thumb	8	**Toes**	**33**
Little	4	I (Great toe)	7
Forearm	0	II (Second)	10
Hand	0	III (Third)	8
Upper Arm	0	IV (Fourth)	5
		V (Little Toe)	3



Figure 1: Lymphoedema of second toe and amputation of first toe



Figure 2: Constriction ring of the leg



Figure 3: Constriction ring on thigh

Intrauterine amputations involved the digits of both the upper and the lower limbs [Figure 7]. In the upper limb, there were 15 amputations involving the thumbs (4), index fingers (3), middle fingers (3), ring fingers (3) and little fingers (2). There were 13 amputations in the lower limbs, which included three each of the great toes, second toes and little toes, and two each of the third and fourth toes.

There were 10 cases of acrosyndactyly involving six hands and four feet. In the hand [Figure 8] digital involvement in acrosyndactyly were as follows: three thumbs, six index fingers, six middle fingers, five ring fingers and three little fingers, while in the foot the involved digits were the great toes (3), second toe (4), third toe (3) and fourth toe (1). The fifth toe was never involved in acrosyndactyly. Two cases of simple syndactyly were found involving the index, middle and ring fingers of one hand, and the second, third and fourth toes of one foot.

Four associated anomalies were noted in four patients, and these were lymphoedema of the foot and ankle distal to a constriction ring in two patients, and one case each of cleft lip and palate, and isolated cleft palate.

Three patients with five affected limbs did not require surgery for the limbs (except that one of these patients had cleft palate repair and another had cleft lip and palate repair).

Thirteen patients with 17 affected limbs had a total of 24 procedures. The procedures included excision of constriction rings and Z-plasty (13), release of acrosyndactyly (8), release of acrosyndactyly and skin grafting of defect (3) [Figure 6a and b].

DISCUSSION

The incidence of constriction ring syndrome varies from 1 in 1500 to 15,000 live births. There is no known

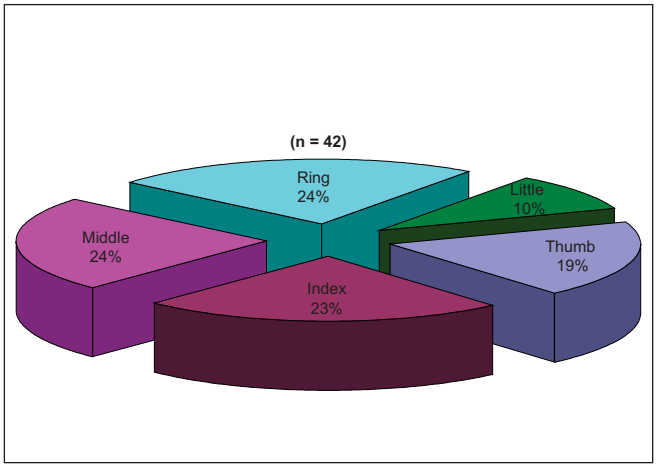


Figure 4: Sites of Malformation in the digits in C. R. S

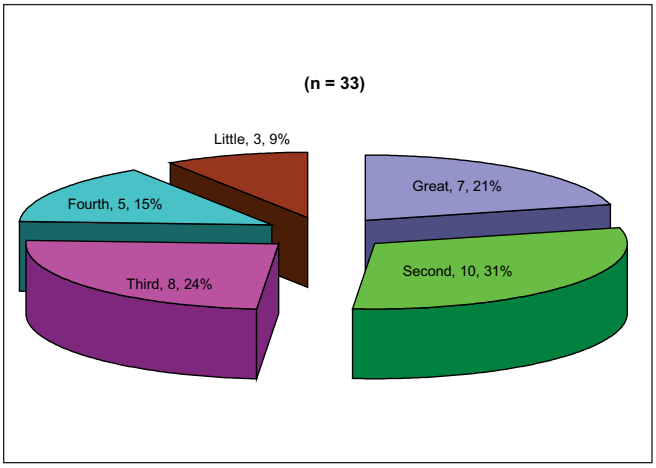


Figure 5: Sites of Malformation in the Toes in C. R. S



Figure 6a: Excision and Z-plasty



Figure 6b: Lymphoedema is subsiding



Figure 7: Constriction rings and amputations of toes



Figure 8: Acrosyndactyly of Rt. hand

racial predilection, and the male preponderance in this study is at variance with the equal sex affectation in other reports.[7] Familial occurrence is rare, and it is believed that there is no genetic predisposition to this condition.[3]

Congenital constriction ring or congenital constriction band syndrome occurs when deep cutaneous creases encircle a limb as if a string were tightly tied around the part. Its frequent association with congenital amputations and acrosyndactyly led to this malformation's designation as a syndrome. Other terms used in the literature include annular band, Streeter dysplasia, intrauterine or congenital amputations, acrosyndactyly and fenestrated syndactyly.[8]

In 1832 Montgomery described a five-month aborted foetus with fibrous bands passing from both hands to the legs and wound around deep grooves in the limbs.[9] Whether the intrauterine amputations and ring constrictions are due to amniotic bands and similar circumferential constricting agents or due to interference with development remains controversial.

The pathogenesis of this disorder is not known for certain, but a number of theories attempt to suggest the aetiology of congenital constriction ring syndrome. The intrinsic theory[4] is the earliest, and it states that these deformities were the result of a "defective germ plasm", within the embryo. Streeter[4] believed that the bands represented macerated sheets of epidermis and the residual of defective local tissue. This theory is supported by localised areas of involvement within the limb and the presence of systemic and internal visceral anomalies.

The second (extrinsic) theory was first described by Torpin.[5] In this theory, the lesions are caused by the strangulating action of the mesodermic bands which occur due to an early rupture of the amnion. After rupturing, the amniotic sac stops growing normally and separates itself from the chorion. The amniotic fluid escapes, causing oligo-hydramnios. The foetus leaves the amniotic sac and lies next to the chorion. Multiple mesodermic bands issuing from the chorionic face of the amnion strangulate the fingers, the limbs and the cranium, inducing the typical lesions. Lack of familial incidence, the transverse disposition of the lesions, the exclusive limitation of the lesions to long digits/limb, the delivery of amputated parts, the presence of engrafted amputated parts on different sites of the body and the absence of associated internal malformations, all support this theory.[5]

The third (intrauterine trauma) theory postulated by Kino[6] believed that congenital constrictions, amputations and acrosyndactyly are caused by intrauterine trauma during pregnancy, which disrupts blood supply to the marginal sinuses of the limb plate.

The number of associated anomalies in this report was lower than in other studies.[1,2,10,11] Other known malformations associated with constriction ring syndrome but not in our series include clubfoot, leg length discrepancies and bone abnormalities.[1]

Surgical correction of a constriction ring should aim at preventing or alleviating distal lymphoedema, separation of an associated distal fusion and removal of an unsightly groove for the sake of cosmesis. Z-plasty was effective and yielded a cosmetic good result in the present series. Where intrauterine amputation of digits has occurred there may be opportunities to improve function by transfer of finger stumps from

one position to another, deepening of web spaces or free-toe transfer.[12]

In conclusion, congenital constriction ring syndrome is of uncertain aetiology and could cause morbidity in the newborn. The syndrome and its complications are amenable to corrective surgery with good results. Early intervention is desirable for a successful outcome.

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