CASE REPORT

Two-stage release in Streeter's dysplasia

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SUMMARY

Streeter's dysplasia is a rare disorder of congenital constricting bands, occurring in 1–10 000 birth, with no sex predilection. In severe cases, it is associated with autoamputation of distal extremities such as fingers and toes. Recent advances have made it possible, in developed countries, to recognise this condition in the prenatal period and to initiate prompt intrauterine management, however, these, being uncommon procedures, need technical advances and experienced hands. In developing countries, recognition of the condition with timely management of primary and associated abnormalities required. We present a case of a 4-month-old child with type IV Streeter's dysplasia associated with right flexible clubfoot, who underwent successful two-stage release of symptomatic constricting bands from bilateral legs.

BACKGROUND

Streeter's dysplasia is a disorder characterised by constricting bands, associated with acrosyndactyly or amputation of extremities in neonates, it is also known as amniotic band disease or constriction bands. The condition was first recognised as early as 300BC. This condition is now widely accepted and supported.¹ Its incidence is 1–10 000–15 000, with no sex predilection.² According to Hippocrates, rupture of amniotic membrane pressure leads to band formation, and in severe cases may even cause autoamputation of extremities. Known associated anomalies include clubfeet, facial cleft palates,³ acrosyndactyly or intrauterine amputation, renal and cardiac abnormalities, tibial pseudoarthrosis, bowing on tibia, etc.^{4 5} Several theories of its pathophysiology have been offered but it is still unclear. So as the treatment option and the right time of treatment.

CASE PRESENTATION

We present a case of a 3-month-old child with Streeter's dysplasia type IV who was born of a full-term, normal vaginal delivery with no prenatal, natal or postnatal issues. He had constricting bands around his legs bilaterally (as shown in figure 1), right flexible clubfoot and an autoamputated right little finger (as shown in figure 2). An X-ray showed the right tibia with anterolateral bowing at the point where a constricting band was located, as shown in figure 3. An arterial Doppler study showed mild compromise of distal flow to the feet. The patient was also evaluated for possible associated abnormalities, as mentioned in the Discussion section, but none other than the right clubfoot were found. In our case, indication of surgical intervention was the development of tibial

Figure 1 Arrow representing constricting bands on lower limbs.

Figure 2 Arrow showing autoamputated left hand little finger.

anterolateral bowing secondary to the constricting bands, distal limb oedema and diminished distal limb arterial flow. The patient underwent successful two-stage release of the constricting bands; pre-operatively they were found to be fascial bands,

Figure 3 Arrow showing right tibia anterolateral bowing.

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Figure 4 (A) Arrow showing first-stage release of constricting bands on (B) right leg and (C) left leg.

constricting the area. Half of the constricting bands were released with multiple z-plasty incisions, closed with absorbable sutures in both lower limbs, as shown in [figure 4](#). The second stage of the release procedure was performed after 1 month of the first release. On follow-up, the wounds had healed completely, as shown in [figure 5](#). Clinically, the peripheral oedema subsided gradually and the distal vascularity status improved, with non-progression of tibial bowing. The right-sided flexible clubfoot is being treated via serial ponsity casting.

OUTCOME AND FOLLOW-UP

At follow-up, the patient showed good healing of the released area, with no postoperative infection. Follow-up will continue until spontaneous correction of the bowing tibia, and management of the right-sided flexible clubfoot with serial ponsity casting and gradual decreasing Pirani score have been achieved.

DISCUSSION

Streeter's dysplasia was first described in 1930 by George Streeter, who stated that the possible aetiology was germ plasma defects, which was accepted at the time.⁶ Later, Patterson hypothesised that lack of mesodermal development occurs in the area of the bands, making them crease abnormally, which he showed histologically.⁷ Richard disagreed with Patterson and reintroduced Hippocrates theory of amniotic band rupture, which is accepted today. A wide spectrum of clinical deformities

is known to be associated, ranging from simple ring constrictions to major craniofacial and visceral defects. Known associated anomalies include clubfeet, facial cleft palates, acrosyndactyly or intrauterine amputation, renal and cardiac abnormalities, tibial pseudoarthrosis, bowing on tibia, etc. Extremity deformities in ABS are classified as Patterson's types,⁸ which are as follows:

- ▶ Type I involves simple ring constriction.
- ▶ Type II involves ring constriction accompanied by fusion of the distal bony parts, with or without lymphoedema.
- ▶ Type III involves ring constrictions accompanied by fusion of soft tissue parts.
- ▶ Type IV demonstrates intrauterine amputations.

The diagnosis is obvious, but treatment timing and addressing of associated abnormalities are demanding. Recent advances now make it possible, in developed countries, to recognise this condition in the prenatal period, allowing prompt intrauterine management; however, these are uncommon procedures and need technical advances and experience hands; however, in some cases, in utero spontaneous resolution of amniotic bands have also been reported.^{9 10} Improvements in prenatal diagnosis will eventually allow treatment of amniotic band syndrome in utero. Several options for treatment of in utero lysis of constricting bands to one-stage release after birth or two-stage release of amniotic bands have been offered. We opted for two-stage release with uneventful recovery.

Figure 5 (A) Second-stage release of bilateral lower limb (B) right leg completely healed scar and (C) left leg completely healed scar.

Learning points

- Recognition of the condition, surgical planning and timely repair are important in improving functional outcome.
- Patients with distal limb constricting bands resulting in peripheral oedema, vascular compromise or developing bony abnormalities require timely repair for a better prognosis.

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Contributors ZGN performed the literature search and wrote the case report. TY and WA performed the literature search. IR supervised the work.

Competing interests None declared.

Patient consent Obtained.

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