A passive constriction phenomenon has been proposed to explain the development of congenital pseudoarthrosis of the tibia. We report a case of congenital pseudoarthrosis of the tibia in a child with congenital constriction band syndrome, successfully treated with soft tissue release (cutaneous and deep) only, restoring normal periosteal blood supply and enabling gradual correction of the angular osseous deformity and reformation of the intramedullary canal.

Keywords: congenital pseudoarthrosis of the tibia; congenital constriction band.

INTRODUCTION

Congenital constriction band syndrome (CCBS) is known to cause a variety of congenital abnormalities with diverse clinical presentations. Growth disturbance of long bones and congenital pseudoarthrosis of the tibia is described throughout the CCBS literature (2, 3, 4, 6, 7, 8, 10). However, we found few reports of successful treatment of congenital pseudoarthrosis of the tibia with spontaneous resolution of the osseous deformity following release of the offending constriction band in the CCBS literature (7, 8). Wright et al. produced a rabbit model of a pseudoarthrosis of the tibia by passively constricting the tibial diaphysis of growing rabbits (9). They proposed a “passive constriction phenomenon” as the major contributor to the development of congenital pseudoarthrosis of the tibia. We report a case of congenital pseudoarthrosis of the tibia in a child with CCBS, which provides clinical support for their hypothesis, and the successful treatment of this deformity with soft tissue release only.

CASE REPORT

The patient, a 2.9-kilogram newborn female, was the product of a normal pregnancy and delivery. She was born with findings consistent with CCBS of both lower extremities. On the left lower extremity there was near amputation of the left lower leg at the distal region of the tibia and...
fibula with a remnant of the foot remaining. There were no bony structures in the left foot. The patient’s right lower extremity had marked anterolateral bowing of the distal two-thirds of the tibia and an associated equinovarus deformity.

Radiographs (figs 1 & 2) revealed increased sclerosis of the intramedullary canal, similar in appearance to a sclerotic sub-type of congenital pseudoarthrosis of the tibia secondary to the congenital constriction band (1). The patient was treated initially at two weeks of age with completion of the left below-knee amputation and partial release of the congenital constriction band of the right lower extremity. A running W-plasty was performed to release the constriction band about the medial and anterior portion of the right lower extremity. The dissection was carried down to expose the tendons, bone, and the neurovascular bundle. The remaining constriction scar was excised and the subcutaneous layers were re-approximated. She was subsequently treated with serial casting for her right clubfoot.

At five months of age, the staged excision of the remaining posterior constriction band with W-plasty closure was performed. Again, the dissection was carried down to expose the tendons, bone, and the neurovascular bundle. The remaining constriction scar was excised and the subcutaneous layers were re-approximated. The Achilles tendon, exposed during the release of the posterior CCB was lengthened at the same time.

Fig. 1. — Initial clinical appearance of lower extremities demonstrating marked anterolateral bowing of the right tibia and equinovarus deformity of the foot, and left residual limb.

Figs. 2a-b. — AP and lateral radiographs of right lower extremity at one month of age showing the sclerosis of the intramedullary canal and anterolateral bowing deformity.

A residual equinovarus of the right foot remained at six months of age despite serial casting and the tendo Achillis lengthening. She was fitted with an ankle foot orthosis (AFO) in maximum dorsiflexion and continued with regular stretching exercises. The anterior tibial bowing had improved significantly by this time, but there was minimal improvement of the lateral tibial deformity. She was fitted with a left below knee prosthesis at eight months of age.

At twenty months of age a right circumferential clubfoot release and Z-plasty of the posterior constriction band was performed without complications. The patient began walking independently at twenty-three months of age. We have observed steady continued improvement of the anterolateral tibial bowing, and formation of the intramedullary canal (fig 3). At her most recent follow-up at 3 + 6 years of age, the tibial bowing had improved to 13° of lateral apex angulation and complete correction of the previous anterior apex angulation.

DISCUSSION

Although congenital pseudoarthrosis of the tibia (CPT) caused by CCBS has been described in the
CONGENITAL CONSTRICTION BAND WITH PSEUDOARTHROSIS OF THE TIBIA

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REFERENCES