Case Report

Congenital Pseudarthrosis of the Fibula

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Congenital pseudarthrosis of the fibula is rare with only 15 cases reported in the literature.1-4 Dooley et al2 described four gradations in the severity of congenital pseudarthrosis of the fibula: 1) fibular bowing without fibular pseudarthrosis, 2) fibular pseudarthrosis without ankle deformity, 3) fibular pseudarthrosis with ankle deformity but without late development of the tibia, and 4) fibular pseudarthrosis with late development of pseudarthrosis of the tibia.

Fibular pseudarthrosis with late development of pseudarthrosis of the tibia overlaps with Boyd and Anderson’s classification for congenital pseudarthrosis of the tibia type V in which there is an associated dysplastic fibula.4,6 Neurofibromatosis is associated with congenital pseudarthrosis of the fibula.1,4,7,9 The tibia demonstrates a characteristic anterolateral bow with cortical thickening without constriction, which with orthotic protection is thought to remodel over time.1,4 Ankle valgus is commonly associated and may present at an early or late age.1,4,9

This article reports a case of congenital pseudarthrosis of the fibula with a 16-year follow-up.

Figure 1: The lower limbs at 6 months of age show deformity over the right leg. Note the cream-colored patches.

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A 6-month-old girl was referred to Newington Children’s Hospital in 1982 for a right leg deformity. An anterolateral bow with a varus deformity in the distal third of the right leg that had been present since birth was noted. The birth history was uneventful. Cream-colored spots were noted over the trunk, back, and gluteal region. The left lower limb, along with the spine, hips, and knees were normal. There was no history of neurofibromatosis in the family.

Radiographs revealed a thin hypoplastic fibula with cysts present in the distal third of the diaphysis. There was anterolateral tibial bowing with lateral angulation of 45° at the distal third of the diaphysis. A narrow medullary canal without cysts was seen in the tibia (Figures 1 and 2). Neurofibromatosis type I with a Boyd type V congenital pseudarthrosis of the tibia and fibula was diagnosed, and a

Figure 2: AP radiograph of both legs at 6 months shows lateral bowing of the distal third of both tibia and fibular diaphysis on the right side (A). Note the thin hypoplastic fibula with cysts. Lateral radiograph of the right leg at 6 months of age shows anterior bowing of the distal third of the fibular diaphysis (B). Note the medullary canal of the tibia is narrow but maintains continuity.
Surgical repair of the fibular pseudarthrosis is not recommended by Dooley et al.\(^1\) and the authors of these 15 reported cases favor orthotic support. The corrective distal tibial osteotomy performed in this case was dictated by pain on weight bearing due to malalignment of the ankle plafond and should always be deferred until skeletal maturity.\(^1\)

This case presented as a spontaneous mutation of neurofibromatosis I, which according to Morrissey and Weinstein\(^4\) is thought to be more frequently associated with congenital pseudarthrosis of the fibula rather than the tibia. Congenital pseudarthrosis of the tibia remains a difficult condition to treat. Nevertheless, this rare and relatively benign congenital pseudarthrosis of the fibula should be listed as a distinct separate entity. Bracing at an early age and continuing until skeletal maturity affords the best opportunity for tibia remodeling during growth and may delay the appearance of progressive values deformity of the ankle.

**REFERENCES**