—**Report on Experiments and Clinical Cases**—

# A Case of Congenital Pseudarthrosis of the Tibia Treated with Pulsing Electromagnetic Fields

## 17-Year Follow-up

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**Abstract**

Congenital pseudarthrosis of the tibia presents surgeons with one of the most challenging of all orthopedic problems. Various surgical treatments have succeeded only rarely. We report long-term follow-up of a patient with congenital pseudarthrosis of the tibia treated with pulsed electromagnetic fields (PEMF) and bone grafting. In this severe case, Bassett type III and Boyd type II, encouraging results were achieved with Boyd's dual onlay grafts and PEMF. Seven years after surgery, skeletal maturity was complete and an unacceptable degree of leg shortening had been avoided. (J Nippon Med Sch 2000; 67: 198—201)

**Key words**: congenital pseudarthrosis, tibia, pulsing electromagnetic fields, bone grafting

## Introduction

First described early in the 18th century, congenital pseudarthrosis of the tibia is a rare orthopedic condition characterized by false joint formation at a site of nonunion that first is evident at or near the time of birth. This event is often associated with neurofibromatosis involving the long bones. The exact cause of congenital pseudarthrosis of the tibia is unknown. Tachdjian[1] listed birth fracture, metabolic disturbance, and vascular malformation as possible causes while noting that these theories had been discarded. McFaland[2] considered five etiologies: focal neoplasia, e. g.osteoclastoma; severe tibial angulation with associated muscle splinting and increased osseous resorption; separate centers of ossification; calcaneal foot position in utero resulting in pressure from the foot upon the tibia; and fatigue fracture occurring in a congenitally defective tibia.

To the present day, congenital pseudarthrosis of the tibia represents a major therapeutic challenge[3,4]. We describe 17-year follow-up results in a patient with congenital pseudarthrosis treated with pulsed electromagnetic fields (PEMF) and bone grafting.

## Patient Presentation

A 7-month-old male infant was brought to the orthopaedic department of Nippon Medical School, where a radiograph (**Fig. 1a**) led to a diagnosis of congenital pseudarthrosis of the tibia. His mother had first noted excessive bending of the right lower leg 4 months after birth. About 18 months later, at the age of 2 years, the boy was brought to our department again after failure of treatment at another hospital. Transtarsal intramedullary fixation and corticocancellous bone grafting had been attempted at an age of about 20 months, but union was not accomplished. The patient's neurovascular status was normal and no lesions

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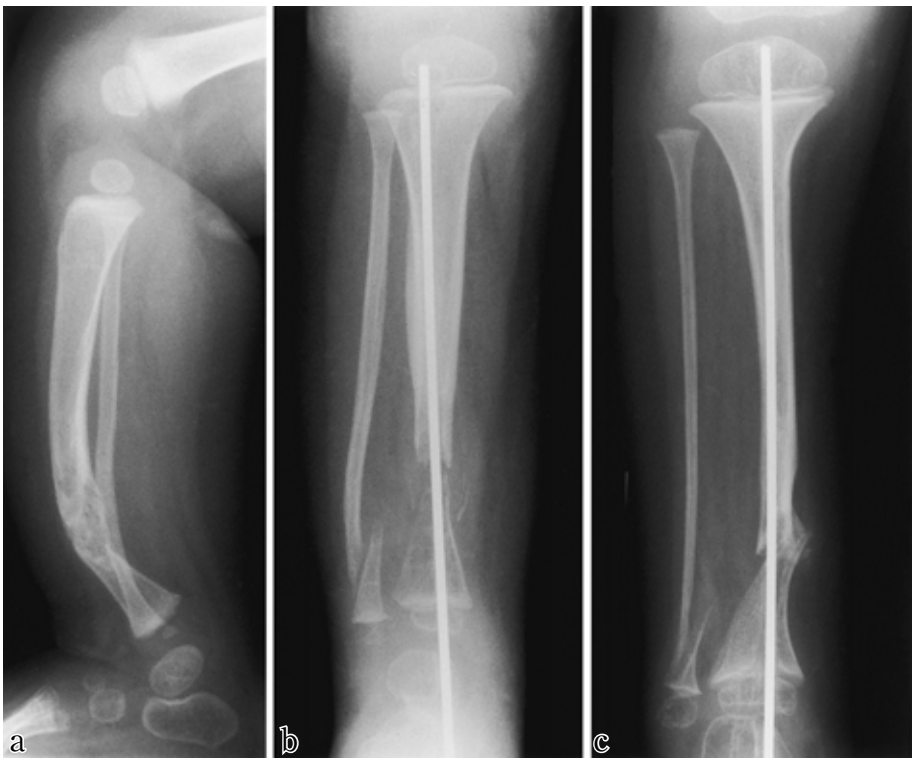


Fig. 1 Sequential radiographs during early childhood in a patient with congenital pseudarthrosis of the tibia. a: a lateral radiograph obtained in September 1979 (7 months after birth), shows severe posterior bending of the leg. b: anteroposterior radiograph, February 1981 (age, 2 years). Pulsing electromagnetic fields (PEMF) stimulation has been initiated with a plaster cast. c: anteroposterior radiograph, April 1983 (age, 4 years). PEMF stimulation has been administered for 2 years.

suggestive of neurofibromatosis were present. With fixation in a long leg cast, pulsed electromagnetic field (PEMF) treatment was administered for 10 hours per day (**Fig. 1 b**). After 2 years of stimulation, radiographic bone union had not been obtained (**Fig. 1 c**). Using a lower leg brace for fixation, PEMF was continued for another year. At outpatient follow-up at age 10, radiographic bone union had not occurred, and the distal fragment of the tibia was bent, tapered, and sclerotic (**Fig. 2 a**). In congenital pseudarthrosis was classified as Boyd's type II and Bassett's type III. The right lower leg was about 10 cm shorter than the left.

Boyd's dual onlay bone graft operation was performed. Within a week PEMF stimulation was resumed for 10 hours per day, using a plaster cast for fixation. Bone union was obtained 8 months after the operation (**Fig. 2 b**). A radiograph of 7 years after operation indicated rigid bone union and remodeling of the grafted tibial span (**Fig. 2 c**). Shortening of the affected lower leg resulted in a right-left difference of about 15 cm, and the right foot showed continuous atrophic changes.

## Discussion

Congenital pseudarthrosis of the tibia is a rare condition that has proven taxing for successive generations of orthopedic surgeons. Until 1930, amputation was considered the standard treatment of choice. Fortunately, clinical advances in orthopedics have rendered this desperate measure virtually obsolete. Use of direct current stimulation[5,6] pulsing electromagnetic fields[7–10], or free vascularized bone grafts[11,12] for treatment of this lesion has been described.

Various classifications of congenital pseudarthrosis of the tibia have been proposed by different authors including Boyd[13], Bassett[7], and others[14]. Boyd's[13] six-type classification, type II is the most common and carries the poorest prognosis. The underlying pathologic change is an aggressive osteolytic fibromatosis. Treatment failures in these patients result from recurrence of osteolytic fibromatosis, a process that

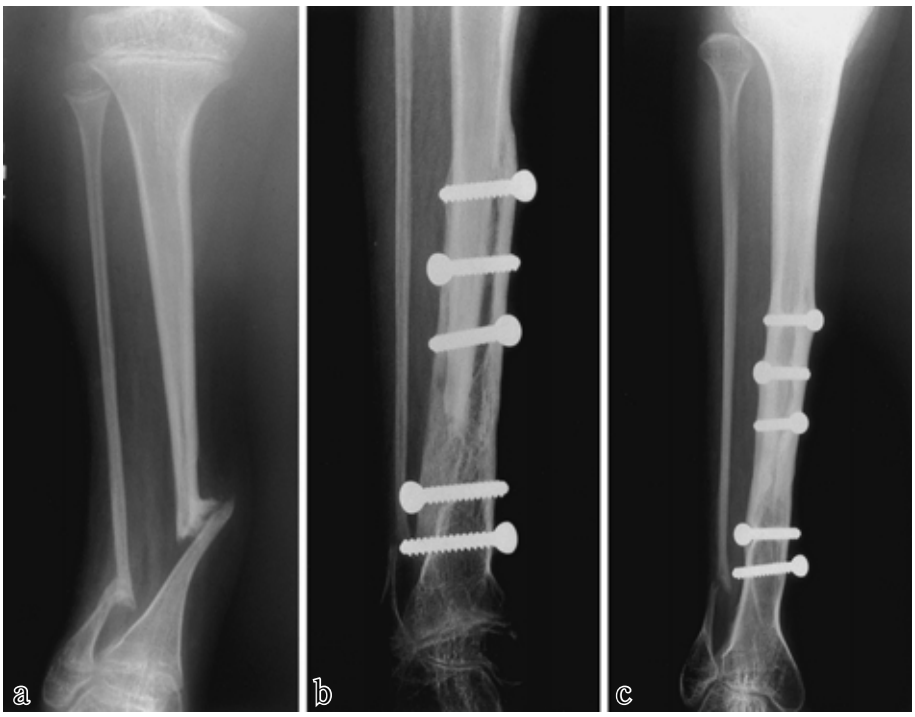


Fig. 2 Sequential radiographs in later childhood for the patient in Figure 1. a: anteroposterior radiograph, July 1989 (age, 10 years). Bony union has not been achieved. The distal tibial fragment is bent and tapered. b: anteroposterior radiograph, August 1990 (age, 11 and a half years). Boyd's onlay graft operation had been performed 8 months earlier, followed by Pulsing electromagnetic fields (PEMF). c: anteroposterior radiograph, December 1997 (age, 17 years old). Seven years following operation, the grafted bone shows remodeling.

can destroy living bone or dead bone grafts. The three-type classification of Bassett[7] is based on radiographic morphology. Type I lesions are characterized by anteromedial or anterolateral bowing with significant sclerosis extending across the medullary canal. In type II lesions, a fracture occurs through a cystic or lytic lesion in the medullary space. Type III lesions show a gap exceeding 5 mm as well as atrophic, spindled bone ends. Our case was classified as Boyd type II and Bassett type III, representing very severe disease with the worst prognosis.

Bassett[15] reported an overall success rate of 54% for treating congenital pseudarthrosis with pulsed electromagnetic fields. In type I and II cases, union was attained in 43 of 60 (72%), while only 19% of type III lesions united (6 of 31). Only one type III case did not require surgery. In patients requiring surgical realignment and in type III lesions with wide gaps, Boyd's dual onlay graft provides the best realignment as well as rigid internal fixation during the relatively long period required to achieve union. Boyd's technique initially used in conjunction with PEMF for the most challenging cases, is recommended as a salvage approach in the treatment of all types of lesions before an amputation is considered. Generally, grafts are placed medially and laterally and fixed with AO cortical screws to produce a compression clamping effect on the proximal and distal fragments of the tibia. The gap is filled with fresh autogenous cancellous chips from the patient's ilium. Postoperatively the treated extremity is protected in a plaster spica cast, with coils added to the cast within 1 week following operation.

Shortening has been recognized as a problem in congenital pseudarthrosis of the tibia, for a long time. Van Ness[16] maintained that shortening resulted from premature fusion of the distal tibial epiphysis, and occurred no matter what type of fixation or grafting was used. He suggested that growth retardation in the distal tibial epiphysis had the same etiology as segmental dysplasia of the distal part of the diaphysis. Anderson[14] concluded that greater atrophy of the bone and a smaller distal fragment worsened the prognosis, also emphasizing problems with limitation

of motion at the ankle, pain in the ankle, and atrophy of the leg and foot. Therefore, for patients with congenital pseudarthrosis of the tibia, the degree of shortening is an important prognostic factor, and close attention should be given to the growth of the distal epiphysis.

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