Successful Treatment of Congenital Pseudarthrosis of the Tibia with Long Segment Fibular Allografting in a Young Child

Yu-Ping Su,1,2 Matthew N.H. Wang,3* Wei-Ning Chang2,4

Congenital pseudarthrosis of the tibia (CPT) is one of the most challenging orthopedic diseases. The pseudarthrosis is usually not present at birth (and therefore is not truly congenital) but occurs during the first decade of life. The etiology is unknown, with neurofibromatosis playing a role in approximately 50% of patients.1,2 The prognosis is poor with an amputation rate of 10%, and more than 60% of patients are unable to participate in sport activity in the European Pediatric Orthopaedic Society (EPOS) multicenter study.1,2 Fusion could be achieved in more than 70% of cases with the Ilizarov technique and vascularized fibula graft,1,3–9 but additional problems such as leg length discrepancy, refracture, axial deformity, progressive malalignment, and ankle valgus usually influenced the final outcome. In patients younger than 3 years, combining procedures such as resection and shortening of the tibial diaphysis through the pseudarthrosis site, internal fixation with intramedullary rods, and autogenous bone grafting is a common initial approach with the advantages of good patient tolerance and a less labor-intensive technique.10–13 However, in some children younger than 5 years treated with bracing, disuse atrophy and deformity inevitably progress, especially for those with a dysplastic fibula. Once they are surgically treated, the large bone defect will be difficult to reconstruct. In this situation, structural allografting might be a practical alternative in such cases but data are limited. We report a girl 1 year and 10 months of age with CPT successfully treated with en bloc resection of the recurrent extensive pseudarthrosis, intramedullary rod placement, and long-segment intercalary allografting with supplemental autograft of the bony defect. At 6 years of follow-up, radiographs showed equal leg lengths, with good incorporation into the host bone. This method is technically simple and less time-consuming. It may be considered as an alternative method to the other more complicated procedures, such as microvascular fibula graft or the Ilizarov technique, in such a young child. [J Formos Med Assoc 2007;106(3 Suppl):S44–S49]

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1Department of Orthopedics and Traumatology, Taipei Veterans General Hospital, 2Department of Surgery, National Yang-Ming University School of Medicine, Taipei, 3Department of Orthopedic Surgery, Kuang-Tien General Hospital and Hung Kuang University, Taichung, and 4Department of Orthopedics and Traumatology, Kaohsiung Veterans General Hospital, Kaohsiung, Taiwan.

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*Correspondence to: Dr Matthew N.H. Wang, Department of Orthopedic Surgery, Kuang-Tien General Hospital, 321 Chin-Kuo Road, Tachia, Taichung 437, Taiwan.
E-mail: admin_tachia@ktgh.com.tw
alternative to autogenous bone grafting for reconstruction. But reported descriptions of such cases are limited. Here, we report a patient with CPT treated with long-segment fibula allograft, supplementary autograft, and intramedullary rod fixation at the age of 1 year 10 months.

Case Report

The girl was born with anterolateral bowing of her left leg. Radiographs taken at birth showed Boyd type V CPT with a sclerotic segment of the medullary canal and a dysplastic fibula. She had no family history or evidence of cutaneous features of neurofibromatosis. A spontaneous fracture at the point of greatest curvature in her left tibia occurred at the age of 5 months despite preventive bracing. The fracture united after conservative treatment with a cast. However, procurvatum of the leg progressed and prohibited the use of a functional brace. In addition, concomitant pseudarthrosis of the fibula developed (Figure 1).

The patient's first operation was performed at the age of 1 year 6 months. The procedure included excision of the lesion and dense thickening rubber-like tissue surrounding the pseudarthrosis, additional proximal osteotomy to accommodate the axial alignment, intramedullary rod fixation, and autogenous bone grafting from the proximal osteotomy. Four months later, recurrent lesions developed in the entire middle segment treated with osteotomy, both periosteally and endosteally, and pseudarthrosis developed at the distal osteotomy site (Figure 2). At the age of 1 year 10 months, the patient underwent en bloc resection of the defective tibia, which was reconstructed with a 6-cm long deep frozen intercalary fibular allograft from our bone bank. Supplemental autogenous cancellous bone grafts from the left anterosuperior iliac crest were placed at the junctions between the allograft and host bone, and the tibia was internally fixed with an intramedullary rod that crossed the ankle and the subtalar joints (Figure 3). The operated limb was placed in a long leg cast for 6 weeks followed by bracing. Union was achieved in 8 months. During that period, the patient received k-wire augmentation to prevent dislodgment of the intramedullary rod. At 6 years of follow-up, radiographs showed mild genu valgum.

![Figure 1](A) Radiograph at the age of 3 months shows Boyd type V congenital pseudarthrosis of the left tibia associated with a dysplastic fibula. (B) At the age of 5 months, the patient had a spontaneous fracture (arrow) at the point of greatest curvature in the left tibia. (C) The fracture is united, with marked procurvatum after 6 months of bracing. Note the formation of pseudarthrosis of the fibula (arrowhead).
of the left lower limb (femoral tibial angle: +8°; normal right femoral tibial angle: +4°) with equal lower limb lengths (Figure 4). Her ankle motion was free and painless, though 12° valgus deformity through the tibia diaphysis was noted.

Discussion
Optimal timing for the surgical treatment of CPT is debatable. In a multicenter study, the EPOS analyzed a 340 case series and found that the best results were obtained in patients older than 5 years and the optimal age for surgery was after 3 years. A multicenter study in Japan also concluded that the Ilizarov method and vascularized fibular graft were the most acceptable methods of treatment. However, Joseph et al. noted that the surgical results in patients younger than 3 years were not only comparable but also better than the results in older groups, in terms of the union rate and remaining abnormalities. The conflicting
findings between the EPOS study and Joseph et al may reflect differences in the surgical procedures chosen by the investigators: the Ilizarov technique in the former study and intramedullary rod placement and dual-onlay cortical bone grafting in the latter. If a patient can be treated when the diseased bone is short and the deformity is limited, good results may be achieved using a less invasive procedure such as resection and shortening of the tibia with intramedullary rod fixation and autogenous bone grafting that is well tolerated by the patient. In contrast, if a patient presents at an older age, more complicated procedures such as microsurgical or the Ilizarov technique may be needed to obtain union and address the deformities, which will progressively worsen over time.

The use of autograft is superior and preferred to allograft for the treatment of CPT because of its better osteogenicity, absence of immunologic conflict, and ease of incorporation with the host bone. However, it will also subject the patient to additional wound, risk of donor site morbidity, and is limited by the supply of donor tissue with suitable bone shape.

Deep frozen allograft has been used for the reconstruction of large bone defects after tumor resection, spinal fusion, and other entities with 50–89% good results. Disadvantages of allograft include slow incorporation with the host bone by creeping substitution, immunologic reactions, risk of infection, nonunion, disease transmission from the donor, and late fracture. Our review of the literature found only one paper reporting two cases of CPT in children treated with intercalary allograft reconstruction. These children were aged 6 and 8 years, and only one (8-year-old) achieved union. DiGiovanni and Ehrlich reported an 11-year-old boy with congenital pseudarthrosis of the fibula treated with intercalary allograft who had near normal growth of the affected leg during 6 years of postoperative follow-up. In our patient, the first operation failed to achieve union. Inadequate excision of lesion is the major cause of early recurrence. Recurrent lesions typically appear extensively both endosteally and periosteally. Ippolito et al suggested that CPT is a pathologic process like “osteofibromatosis”, with clinically diverse types which should be treated as a benign, aggressive bone tumor with complete extraperiosteal excision of the pseudarthrosis to prevent recurrence. The bone defect resulting from resection requires adequate bone grafting to achieve union. Short segment bone loss can usually be treated with immediate shortening with planning for future leg length equalization procedures. Large bone defect usually requires vascularized free fibular graft or a lengthening through the Ilizarov apparatus to guarantee a higher union rate. Intercalary allografting is technically simple, quantitatively ample, and less time-consuming than other procedures. In addition, experience with
microvascular techniques is not needed. Also, microvascular or Ilizarov techniques are not suitable for children younger than 5 years of age. The allograft can be fashioned according to the defect and the deformity can be corrected at the same time. In this case, the junctions between the allograft and the host bone were augmented with cancellous autograft to enhance union.

The refracture rate is lower in patients treated with intramedullary rod placement and autogenous bone grafting than in those treated with free vascular fibular grafting as well as the Ilizarov technique.\textsuperscript{13,20,21} Retaining an intramedullary rod as an internal splint until skeletal maturity is advocated to minimize the risk of refracture. In our patient, the rod was removed because of distal dislodgment. At the time of rod removal, there was a $12^\circ$ valgus deformity through the diaphysis of the tibia, which did not progress during the last 4 years of follow-up. The presence of pseudarthrosis of the fibula might have contributed to the deformity in this patient. Several authors have suggested that pseudarthrosis of the fibula is a risk factor for worse functional results at the end of skeletal maturity.\textsuperscript{22–24} Excision of the fibular pseudarthrosis and interosseous fusion to the tibia might be needed if rapid progression of the deformity occurs before skeletal maturity is achieved. The risk of fracture in the long-segment allograft was likely increased in our patient, not only because of its slow incorporation and adaptation with the host environment but also because of the limited increment in diameter, as the surrounding periosteum was radically excised. Therefore, long-term bracing, at least until skeletal maturation, is recommended.

In conclusion, this case illustrates that tibia length discrepancy in a young child with CPT can be restored with long-segment intercalary allograft, and bone union can be achieved with supplemental autograft, solid intramedullary rod, and long leg cast fixation. The lack of recurrence in this case during 6 years of follow-up was attributed to thorough resection of the diseased bone and periosteum. This simpler approach might be a more practical alternative than the other more complicated procedures, such as microsurgical or the Ilizarov technique, especially in settings where these methods are not readily available. Prospective study and long-term follow-up are needed to evaluate the role of long-segment fibular allografting in young children, either as a primary or as a salvage procedure.

References


