



Free vascularized fibular graft for tibial pseudarthrosis in neurofibromatosis

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Four patients with neurofibromatosis and a dysplastic type of congenital pseudarthrosis of the tibia were treated by resection of the lesion and reconstruction of the extremity with a free vascularized fibular graft. Solid union of the graft was achieved within 3 months in 3 children. However, valgus malalignment of the tibia progressed in all 3. One child was treated with resection of a lateral fibrotic band and another with distal tibial epiphysal distraction and realignment. In the fourth patient, who was skeletally mature, a nonunion developed proximally.

Free vascular bone transfer has been reported in congenital pseudarthrosis of the tibia (Weiland and Daniel 1980, Pho et al. 1985). We have used this technique in 4 patients with congenital pseudarthrosis associated with neurofibromatosis. We wish to emphasize the progressive malalignment problems that we experienced in 3 children.

Patients and methods

The records of 4 patients with neurofibromatosis who underwent a fibular transfer for reconstruction of a bony defect after radical resection of a dysplastic type congenital tibia pseudarthrosis were reviewed. The mean follow-up in this study was 2.5 (1-4) years. There were 3 children aged 7 to 8 years and 1 skeletally mature 17-year-old patient.

Despite an average of four previous operations, no union had been obtained, and all 4 patients

walked with the aid of external support, and they experienced pain.

The technique described by Gilbert (1979) and Weiland (1984) was used in all the patients with an orthopedic and a plastic surgeon operating together.

After wide resection of the pseudarthrosis with surrounding fibrotic envelope, reconstruction was performed with a free vascularized fibular graft (average length 17 cm) from the contralateral leg. The circulation in the graft was restored by vascular anastomosis. Stability was maintained with internal screw fixation proximally and/or distally supported by external fixation. Unequivocal radiographic evidence of bone healing at both ends of the graft was required for confirmation that the bone segment had united. Finally, all major complications and necessary secondary treatment were recorded.

Results

In 3 patients, bone scans, performed one week postoperatively, were positive in the region of the vascularized bone transfer, indicating good graft vitality. Postoperative angiography in the fourth patient showed patency of the anastomosis. In the children, union was achieved within 3 months; and all of them walked without external support within 1 year. At that time, there was marked

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hypertrophy of the graft with a well-formed medullary canal.

An increasing valgus malalignment of the tibia developed in these children (Table 1). In the first child (Figure 1), now 12 years old and 4 years after reconstructive surgery, the valgus position of 15° at the ankle had become stationary and did not require further surgery.

The second child in whom reconstruction took

place at the age of 7 had a progressive valgus position, which over 2 years had increased to 37°. Because the malalignment was located at the ankle, we elected to correct this with distraction of the distal tibial epiphysis. With the use of external fixation, this valgus malalignment could progressively be corrected. A residual valgus of 16° remains (Figure 2).

The third child (Figure 3) had a valgus malalign-

Table 1. Results of treatment of congenital tibial pseudoarthrosis in neurofibromatosis with free vascularized fibular graft

Case	Sex	Age (yr)	Follow-up (yr)	Previous operations ^a	Preop. valgus	Length of graft (cm)	Result bone scan	Time to union (mo)	Immediate postop valgus	Progressive postop valgus tilt	Secondary surgery ^b	Valgus at follow-up
1	M	8	4	1 H	20°	16.5	+	3	0°	15°	—	15°
2	M	7	2.5	1 A	0°	12.5	not done	2	5°	37°	D	16°
3	F	7	2	4 AF	0°	19	+	2	5°	25°	E	0°
4	M	17	1.5	9 AHF	5°	21	+	proximal nonunion	5°	—	G	5°

^a Number and type:

H homologous bone graft.

A autologous bone graft.

F intramedullary pin fixation.

^b Secondary surgery:

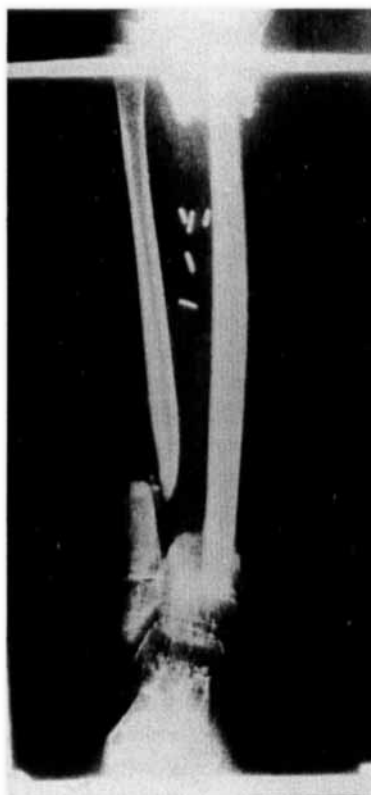
D distal epiphyseal distraction.

E excision lateral fibrotic band.

G debridement, gentamicin-beads implantation.



A



B



C

Figure 1 The first patient

A Before reconstruction

B Postoperatively

C At 4 years follow-up, there was marked hypertrophy of the graft

ment at the proximal junction of the graft that had progressed over 18 months to 25°, and after a fall the patient sustained a fracture on that side. Correction of this valgus deformity could only be obtained after partial excision of a lateral fibrotic band that included the fibula affected by neurofibromatosis. Radiographs 10 weeks after this fracture revealed union and progressive hypertrophy of the graft with good alignment.

In the fourth case (Figure 4), a 17-year-old

patient, now 18 months after reconstructive surgery, there was still nonunion proximally. Six weeks after reconstructive surgery, a pin-tract infection with *Staphylococcus aureus* progressed proximally from the external fixator. Local debridement and serial gentamicin-beads implantation combined with systemic antibiotics cleared the infection. The patient has practically been without signs of infection for a year, and has recently been stabilized with internal fixation.



Figure 2. The second patient.

A. Four months postoperatively.

B. Two years' postoperatively, there was a valgus of 37° in the ankle.

C. After epiphyseal distraction, there was a residual valgus of 16°.



Figure 3. The third patient.

A. At age 7, after four previous operations. There is still a pseudarthrosis with loosening of the pin.

B. Immediately, C. 12 months and D. 18 months postoperatively. There is a progressive valgus malalignment with a fracture of the graft just below the screw at 18 months.

E. After correction of the valgus deformity with partial excision of a lateral fibrotic band (arrow).

F. 10 weeks after correction the fracture had united.

Discussion

Treatment of the dysplastic type of congenital pseudarthrosis of the tibia remains one of the most challenging orthopedic problems. Notwithstanding multiple operations, union can be expected in only 20 percent of these cases, and in as many as 50 percent end in amputation (Morrissy 1982). After excision of a pseudarthrosis together with

its fibrotic envelope and reconstruction utilizing a vascularized fibular graft, union can be expected. Our results confirm the union rate reported by Gilbert (1982) and Pho et al. (1985). Gilbert discussed 50 patients with a congenital tibia pseudoarthrosis from six different centers around the world treated with a vascularized bone graft. In 32 of these patients, neurofibromatosis was



Figure 4. The fourth patient.
A. Before reconstruction.
B. Proximal infected nonunion treated with gentamicin beads.
C. 15 months postoperatively, there was a hypertrophic nonunion proximally.

present. The primary union rate was 57 percent. The overall union rate after secondary surgery was 90 percent. Pho et al. (1985), in 5 patients, reported one proximal nonunion that united after secondary surgery.

An increasing valgus deformity of the tibia was found in the 3 not yet skeletally mature patients. This is in agreement with Pho et al. (1985), who reported an increase in valgus deformity in 4 out of 5 patients. This valgus deformity could be the combined effect of overgrowth of the tibia and

bowstring action of a lateral fibrotic band including the neurofibromatotic fibula.

It seems reasonable to prevent this angular deformity in the skeletally immature patient by excising the lateral fibrotic band, which can be done primarily or after reconstruction as soon as a progressive valgus position at the proximal junction presents. However, creation of a synostosis distally through the residual fibula and tibia may, in the future, decrease the valgus deformity at the ankle.

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