

## CONGENITAL PSEUDARTHROSIS OF THE FOREARM, TREATED WITH A VASCULARIZED FIBULA

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**Congenital pseudarthrosis of the forearm is extremely rare. Only a few cases have been reported, all in association with neurofibromatosis. The condition is similar to congenital tibial pseudarthrosis and is difficult to treat. We report a case of successful healing with a vascularized fibular transplant.**

### CASE REPORT

A 13-year-old boy was seen with a deformity of his right forearm consisting of shortening and radial deviation in the wrist joint. The wrist was unstable, preventing all hand function. He was the oldest boy of 2 non-related parents (maternal age: 25, paternal age : 30). His mother showed evidence of neurofibromatosis with dermal neurofibromas and café-au-lait spots.

The boy also demonstrated café-au-lait spots, axillary freckling and one Lisch nodule. All these findings were suggestive for neurofibromatosis. The younger sister had opisthomological alteration, also related to neurofibromatosis. At the age of 11 years his right forearm began to deform (fig. 1). This bowing progressed (fig. 2) to end up one year later in a frank pseudarthrosis of the radius (fig. 3). Previous attempts with autogenic and allogenic bone grafts and a bone transport with Ilizarov's device had failed when the boy was seen at our clinic. At the age of 13 years, the pseudarthrosis was largely debrided and a vascularized fibular graft from the ipsilateral leg was transplanted into the defect and was fixed with two screws (fig. 4).



*Fig. 1.* — Bowing of the radius at the age of 11 years (August, 1994).

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Fig. 2. — Progressive bowing (November, 1994).



Fig. 3. — Pseudarthrosis of the radius (October, 1994).

Uneventful healing occurred at the distal epiphysis but a conventional bone grafting 6 months later combined with solid fixation with plate and screws was required to obtain union proximally. Three years postoperatively, the radius remained solid and was still growing, but there was some residual shortening. Wrist motion was limited (extension 20°, flexion 50°, prosupination impossible) (fig. 5).

### DISCUSSION

Congenital pseudarthrosis of one or both bones of the forearm is very rare. The cases reported since

1979 are summarized in table 1. In this particular case, the pseudarthrosis developed only at the age of 12 years ; similar to tibial pseudarthrosis, bowing was present previously. Free vascularized fibula transplantation was described in 1975 by Taylor *et al.* (10). In most of the cases traumatic bone defects (usually of the tibia) were treated. The procedure proved to be reliable but never gained great popularity because it is technically demanding and time consuming (4).

Iizarov's technique and his philosophy on bone regeneration made this procedure obsolete. In 1981 Allieu *et al.* presented 2 cases of congenital pseudarthrosis of the forearm successfully treated with



*Fig. 4.* — Vascularized fibula graft fixed with minimal fixation (March, 1996).



*Fig. 5.* — Healed pseudarthrosis and incorporation of the fibular graft (October, 1998).

Table I. — Summary of the published cases.

Author	Year	N	Localization	treatment
Allien <i>et al.</i>	1981	2	2R+U	FVF
Hadlow	1979	1	U	1BFA
Ostrowski <i>et al.</i>	1985	2	U	1BFA
Bell	1989	6	1R, 4U, 1R+U	2 x FVF
Young and Arford	1991	1	U	?
Masterson <i>et al.</i>	1993	1	U	FVF
Mathoulin <i>et al.</i>	1993	6	1R, 1U, 4R+U	2x1BFA 4xFVF
Cheng <i>et al.</i>	1994	2	2U	1x1BFA 1x1FVF

FVF : free vascularized fibula ; U : ulna ; R : radius ; 1BFA = one-bone-forearm.

a vascularized fibular graft. Mathoulin *et al.* have performed this procedure in 6 patients (7). Other occasional reports have been published (Table I).

Another therapeutic option in this rare condition is the creation of a one-bone forearm, which also seems to have achieved satisfactory results.

Ilizarov's technique – resection of the pseudarthrosis and bone transport – has been used with success in congenital pseudarthrosis of the tibia, but as far as we know, it has not been reported in the forearm.

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## SAMENVATTING

*L. DE SMET : Congenitale pseudarthrose van de onderarm, behandeld met een vrije gevasculariseerde fibula-ent.*

Congenitale pseudarthrose van een onderarmbeen is bijzonder zeldzaam en quasi altijd in associatie met neurofibromatose. De behandeling is, net zoals de congenitale tibia pseudarthrose zeer moeizaam. Wij beschrijven een casus, die met een gevasculariseerde fibula ent succesvol kon worden behandeld.

## RÉSUMÉ

*L. DE SMET : Pseudarthrose congénitale du radius traitée par greffe du péroné vascularisé.*

La pseudarthrose congénitale est extrêmement rare au niveau de l'avant-bras. Quelques cas seulement ont été rapportés, tous associés à une neurofibromatose. La lésion est similaire à la pseudarthrose congénitale du tibia; elle est de traitement difficile. L'auteur rapporte un cas traité avec succès par une greffe de péroné vascularisé.