FROM THE CHILDREN'S HOSPITAL, UNIVERSITY OF HELSINKI, FINLAND

CONGENITAL PSEUDARTHROSIS OF THE TIBIA

By

M. SULAMAA and P. VILKKI

Congenital pseudarthrosis of the tibia is a rare clinical entity resistant to all forms of therapy during infancy and early childhood. *Knight* (1) classifies the cases into three types: 1. defect of the mid-tibia at birth, 2. fracture associated with a congenital cyst of the tibia, and 3. fracture associated with congenital bowing of the tibia. The third group is probably the rarest and forms a curious entity for which no underlying cause of the weakness of the tibia can be discovered. A minor trauma or corrective osteotomy for bowing results in pseudarthrosis in which union is most difficult to secure—even more difficult than in the other forms of congenital pseudarthrosis (2). The published data (1, 2, 3, 4) show that union, if obtained in this condition, is often accompanied by considerable deformity and shortening of the leg. Although rare, the disease merits the attention of clinicians, since the consequences to the child, if treatment is not adequate, are most serious, amputation of the leg being by no means infrequently the end-result (2).

The purpose of this paper is to describe those cases of congenital pseudarthrosis of the tibia treated in this hospital, in which the weakness of the bone is really "idiopathic" in nature and not due to a tumour or cyst or to a macropscopic defect. In all our three cases the tibia was too weak to allow normal weight-bearing from the beginning, and the pseudarthrosis became manifest when the patient first began to walk. The history of bowing of the leg was apparent from birth. In one case the pseudarthrosis was not present when the patient was first seen in this hospital but resulted from a corrective osteotomy for severe bowing of the tibia.

CASE REPORTS

Case 1. R.S., a boy with a curved left leg took his first steps at the age of 14 months. A very slight trauma resulted in swelling and ten-

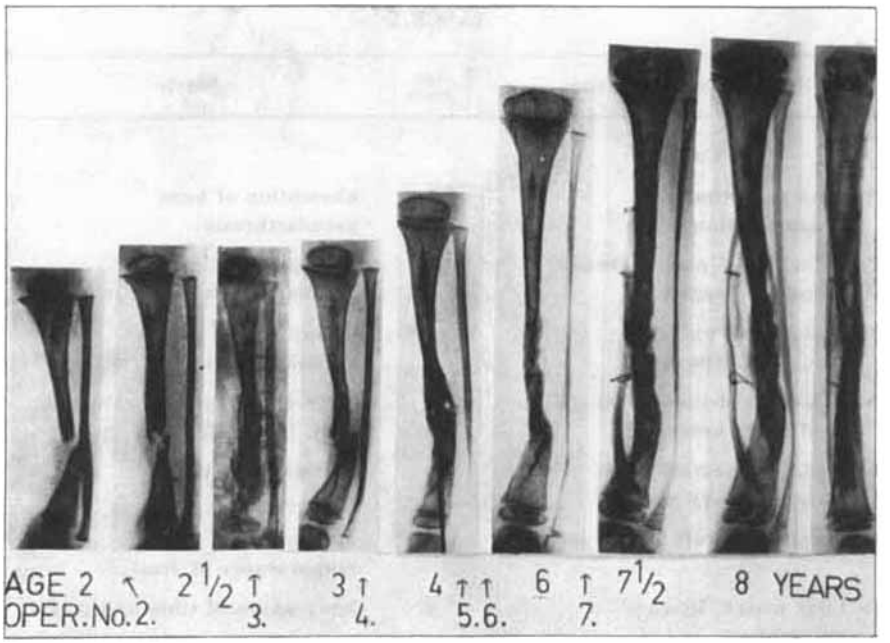

Fig. 1.

Case 1. Age of the patient and operations performed are indicated below the X-ray pictures. Operation numbers relate to table 1.

derness of the leg, and the patient was admitted to the local hospital. There an "old fracture" of the left tibia was diagnosed. The bowing at the site of the fracture was corrected by open reduction, and the limb immobilized in a plaster cast. Despite long and effective immobilization no union occurred.

The boy was first seen at this hospital at the age of two years. X-ray examination (Fig. 1) showed a typical picture of pseudarthrosis in the left tibia between the middle and lower thirds of the bone. At operation (Oper. 2, Table 1) the fibrous pseudarthrosis was removed, and the contralateral fibula transfixed like an intramedullary nail across the defect. Biopsy showed fibrous connective tissue with islets of hyaline cartilage, there being no sign of normal bone formation. The first bone grafting failed but five grafting operations over the next six years gradually resulted in bony union (Table 1). The growth of the leg was not arrested by the disease or by the grafting operations (Table 2). The boy is now walking with the aid of a light leather brace.

TABLE 1

No. and type of operation	Age years	Result
Case 1.		
1. Open reduction Fragmentation	1	absorption of bone pseudarthrosis
2. Resection of pseudarthrosis Fibular autograft	2	absorption of graft pseudarthrosis
3. Costal dual onlay auto- and homograft	2½	absorption of grafts pseudarthrosis
4. Resection of pseudarthrosis Ileal crista autograft	3	bony union through callus at first, later refracture
5. Costal homograft with intramedullary nailing	4	partial bony union
6. Costal autograft	5½	absorption of graft reappearance of fracture
7. Long costal "by-pass" autograft	7	bony union of tibia and grafts
Case 2.		
1. Closed reduction Plaster cast	1	pseudarthrosis
2. Fibular autograft	1½	partial bony union, reappearance of fracture at mobilization
3. Resection of pseudoarthrosis Fibular autograft	2½	callus formation, later fracture through callus
4. Costal homograft	4	partial bony union bowing of tibia
5. Long costal "by-pass" homograft	7	partial bony union of tibia, fracture line through graft
Case 3.		
1. Tibial osteotomy, dual onlay costal homograft	1	absorption of grafts pseudarthrosis
2. Long costal auto- and homografts, "by-pass"	1½	first incorporation, later fracture of grafts
3. Long costal homografts "by-pass"	2	bony union of grafts defect of tibia

Case 2. J.H., a boy, aged 14 months. Shortly after learning to walk he fell from a low footstool, hurting his left leg. He was immediately admitted to this hospital. X-ray (Fig. 2) showed an "old fracture" of the left tibia with bowing below the site of the fracture. Closed reduction

TABLE 2
Length of both tibiae in cm.

Age years	Case 1		Case 2		Case 3	
	dx	*sin*	dx	*sin*	*dx*	sin
1	13.9	13.7	13.9	13.7	14.5	14.2
2	14.0	13.5	15.7	15.8	17.4	17.2
3	17.3	16.5	18.1	18.2	...	...
5	22.4	22.5	23.2	23.3	...	...
7	24.3	24.2	27.6	27.6	...	...
difference in length	—0.1		0		+0.2	

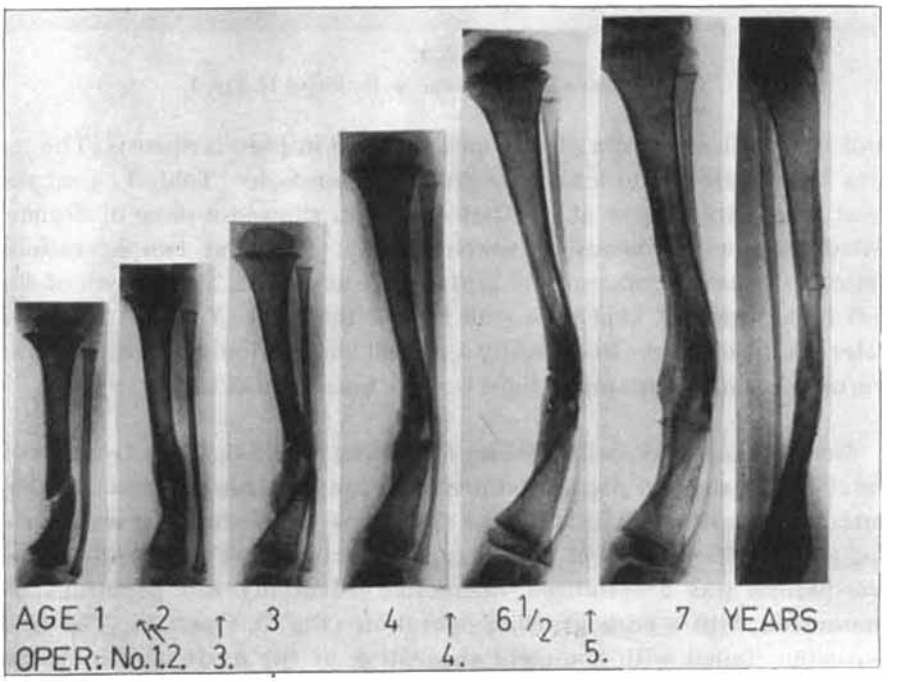

Fig. 2.
Case 2. Age and operations as indicated in Fig. 1.

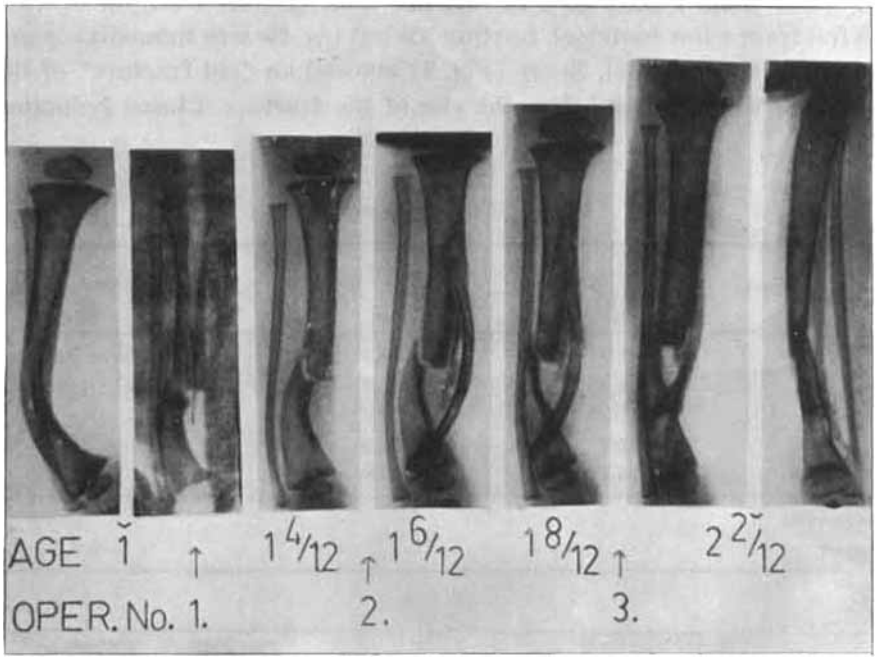

Fig. 3.
Case 3. Age and operations as indicated in Fig. 1.

and immobilization in a plaster cast resulted in pseudarthrosis. The leg was then subjected to four bone grafting operations (Table 1) over the next five years. Biopsy at the first operation showed a piece of degenerated bone with fibrous connective tissue. The first two operations failed to secure union, and the grafts were absorbed. The growth of the left tibia, however, kept pace with that of the right (Table 2). The two later operations were followed by a partial bony union of the tibia. Some varus deformity remains. A light leather brace is still worn.

Case 3. S.L., a girl, had a history of bowing of her right leg from birth. In addition, she had pigmented areas of skin, regarded as possible stigmata of neurofibromatosis. At the age of one year, when she was learning to walk, the bowing of the leg rapidly increased (Figs. 3 and 4), and the patient was hospitalized. Corrective osteotomy was performed in connexion with a bone grafting operation (Fig. 3, Oper. 1). The first operation failed with complete absorption of the graft, and a typical pseudarthrosis formed. The second operation was more successful, and after the third operation a stable leg of normal length resulted. In the

biopsy specimen from the second operation small pieces of bone embedded in scar-like connective tissue were seen. To-day a bony union of the grafts is apparent. There is, however, a cystic defect of the tibia proper, and she is still wearing a walking plaster cast.

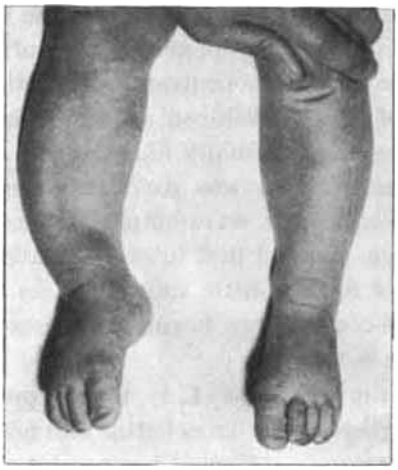

Fig. 4
Photograph of the legs of case 3 before the first operation

DISCUSSION

The three cases of congenital tibial pseudarthrosis described required multiple bone grafting operations before a bony union was achieved. In spite of the multiple operations—or perhaps thanks to them—a gross deformity of the bone was avoided, and no arrest of the normal growth of the leg occurred. As bony union was secured at the age of 7 in two of the cases and at the age of 2 in the third, there will be ample time for the normal growth of the bone to correct the residual minor deformities. The great tendency to recurrence of childhood pseudarthrosis is well known. Therefore, a light brace to protect the leg from external traumata is still continued in all cases. Despite the brace, the children are able to undertake fairly normal physical activities.

In our cases the source of the bone used in the grafting operations seems to have been of minor importance. Fibula, ileal crista, and ribs from the patients themselves were used as autografts. Frozen ribs from the bone-bank as well as fresh homografts were also used. Long "by-pass" grafts bridging far beyond the pseudarthrosis on each side seem to be incorporated more readily than short onlay grafts or intramedul-

lary grafts. Long "by-pass" grafts of *McFarland* (4) type were preferred at the latest operations. No attempt was made to remove the fibrous pseudarthrosis tissue, since this might have jeopardized the endeavour to maintain the normal length of the bone. Although well incorporated at the ends, the "by-pass" grafts also showed a great tendency to fracture at the original site of the pseudarthrosis. The tendency to fracture at this "magic" level even in the opposite leg is curiously demonstrated by the two patients in whom the contralateral fibula was used as grafting material. Both of these developed a pseudarthrosis at the exactly same level of the otherwise normally regenerating contralateral fibula.

Intramedullary nail fixation was used only once in our cases. No other internal fixation devices were employed, except suturing of the onlay grafts with wire. We feel that internal fixation, as recommended by some authors (2, 4, 5), has little value in these cases, provided that efficient grafting procedures are begun early enough to prevent the development of gross deformity.

As is known from the literature (1, 2), the prognosis of bone-grafting in congenital pseudarthrosis becomes better and better with the growth of the child. If a more or less normal leg can be maintained until puberty, a good result is the rule. Unfortunately, serious deformity and shortening of the leg is often so evident even at preschool age that amputation has to be resorted to. We feel that our active attitude to regrafting as soon as the previous graft shows signs of failure is of the utmost importance in preventing the deformity and shortening. Also by frequent regrafting advantage is taken of the earliest possible moment for the bone consolidation as conditions gradually become favourable for union. The age of possible consolidation must vary from patient to patient; otherwise the unpredictable results of the same grafting procedure at various ages and in different patients are incomprehensible.

These cases of ours, like other series in the literature, show clearly enough that a leg with congenital pseudarthrosis can be restored to normal function and length by careful surgical management. The failures, too often encountered, may be largely avoided if the disease is diagnosed early, and the treatment planned with all the thoroughness demanded by these very difficult cases.

SUMMARY

Three cases of congenital pseudarthrosis of the tibia treated with multiple bone-grafting operations are described. The result of the treat-

ment in all three cases is a stable leg of normal length. None of the cases has yet reached skeletal maturity.

RESUME

Trois cas de pseudarthose congénitale du tibia traités par des opérations de multiples greffes osseuses sont décrits. Dans les trois cas, le résultat du traitement a été une jambe stable le longueur normale. Dans aucun des cas la maturité squelettique n'est encore atteinte.

ZUSAMMENFASSUNG

Drei Fälle von angeborener Pseudarthrose der Tibia, die mittels wiederholten Knochensprangenoperationen behandelt wurden, werden beschrieben. Das Ergebnis der Behandlung ist in allen drei Fällen ein stabiler Unterschenkel von normaler Länge. Keiner der Fälle hat noch Skelettvollreife erreicht.

REFERENCES

1. *Knight, R. A.:* Congenital anomalies. Campbell's Operative Orthopedics. St. Louis, 1956.
2. *Boyd, H. B. & Sage, F. P.:* Congenital Pseudarthrosis of the Tibia. J. Bone & Joint Surg., *40-A*: 1245, 1958.
3. *Evans, E. B. & Eggers, G. W. N.:* Nonright Fixation in the Treatment of Childhood Pseudarthrosis of the Tibia. The American Surgeon *28*: 510, 1962.
4. *McFarland, B.:* Pseudarthrosis of the Tibia in Childhood. J. Bone & Joint Surg., *33-B*: 36, 1951.
5. *Cathro, A. J. M.:* A Case of Congenital Pseudarthrosis of the Tibia Treated by Kuntscher Nailing. East African Med. J., *38*: 581, 1961.