INFANTILE PSEUDARTHROSIS OF THE TIBIA

Three Cases Treated Successfully by Delayed Autogenous By-pass Graft, with Some Comments on the Causative Lesion


This paper is conceived as a corrective to a somewhat widespread pessimism in relation to infantile pseudarthrosis of the tibia, a pessimism shown in the advocacy of delay in surgery, in the acceptance of damage to the ankle and subtalar joints by a nail introduced from the sole of the foot, and even in the concept that amputation is a reasonable alternative to reconstructive procedures. Experience in Bristol appears to have been quite different, and we wish to report the result of four consecutive cases with satisfactory union, although in the first child the leg was eventually amputated. The series is small, but the condition is now rare in this country and the consistent results appear to justify this report. We wish to underline the title of the condition as “infantile pseudarthrosis of the tibia”. It is not a congenital pseudarthrosis. We will later discuss the lesion, often congenital, in which the pseudarthrosis develops.

The four children described in this paper were treated by a by-pass graft as described by McFarland in 1940 and 1951. We believe this to be a fundamentally sound procedure, from which one should confidently expect good results. Five important points are as follows. 1) There is no mobilisation of the pseudarthrosis and the deformity is accepted. 2) There is no devitalising effect on the bone ends by extensive stripping of periosteum or the introduction of foreign bodies. 3) The graft is embedded in healthy bone above and below, and well away from the tissue of the lesion. 4) The graft is stable and under compression as it lies vertically between the knee and the ankle. 5) The operation can be done at any age, no tibia being too young for the procedure.

The experience of the first child in this series was very educational but cost the child her limb. The first graft united but was too puny and fractured. As is frequent in a child of under three years, the donor tibia was small and the cortex thin, but on repeating the procedure we learnt the first lesson that a delayed graft, lifted and replaced and transferred to the recipient leg at least eight weeks later, carries abundant callus, increasing the chances of success manyfold. This is particularly so in younger children because the graft is more rigid and easier to handle. The technique of the delayed graft is no new procedure; it has been advocated and employed over many decades for a number of conditions, and even for pseudarthrosis of the tibia by Moore (1949), although not as a by-pass graft.

The second lesson learnt from this child was that the grafts need support until wholly revascularised as shown by the appearance of a medullary cavity in the radiograph. This may take up to five years. This second lesson was learnt when the graft fractured a year and a half after sound incorporation at each end. This point has been stressed by others, notably Boyd and Sage (1958), but unfortunately had to be learnt at great cost in this case. We failed to obtain union, the fracture having occurred through the centre of the graft.

The third lesson learnt from this first child came when we suggested amputation but indicated that the parents might wish to seek the advice of Professor McFarland himself. An enthusiast cannot accept the failure of his operation in another man’s hands, and it was five years before the shortened limb, still ununited, was accepted as correctly treated by amputation. We have not repeated any of these mistakes in subsequent cases and the following plan of action has thrice proved successful at the first venture.
PROGRAMME ADVOCATED IN TREATING INFANTILE PSEUDARTHROSIS OF TIBIA

The graft should be lifted and replaced, and an interval of eight weeks should elapse before the graft is again lifted with its accretion of callus on all surfaces and inserted as a by-pass graft vertically across the ununited segment between the healthy bone above and below, into which it is slotted without any periosteal stripping and with minimal soft-tissue dissection (Fig. 1). The leg is placed in plaster, which should be retained initially for eight weeks. The child is then allowed to walk for a further eight weeks in a walking plaster—a total of four months from the insertion of the graft. We may then expect radiological incorporation of the graft at both ends, although some patients may need a further period in a walking plaster.

Finally, we continue support for a total of five years in a polythene splint or until vascularisation of the graft is complete, as shown by thickening of the bone and the formation of a medullary cavity. At this stage it is not uncommon to find that the pseudarthrosis has united as well. The polythene splint leaves the knee free and fits in a larger shoe; it does not hamper the child in normal activities, scholastic or otherwise. The leg may be left free at night.

CASE REPORTS

Case 1—The first child, a girl, presented at the age of twenty-three months with the dysplastic process seen in Figure 2 but no fracture. A month later she returned with a fracture (Fig. 3). At the age of two years and nine months a by-pass graft was inserted without straightening the tibia. Radiographs four weeks after the operation showed early incorporation of the lower end of the graft though not at the upper end (Fig. 4). Five months later incorporation had taken place at both ends but fracture of the graft had occurred (Fig. 5). The graft was too slender and had been fractured in an attempt to straighten the leg—a manoeuvre that is never required owing to natural corrective moulding in these young subjects.

A second operation was performed at three and a quarter years, six months after the original graft, and so impressive was the thickening of the tibia at the donor site from the previous surgery that we have always since then used a delayed graft. Three months after the
second operation the graft was well incorporated above but not below. Two months later there was sound incorporation at both ends of the graft (Fig. 6).

Progress—A year and a half after the second operation the graft fractured through its centre, which was still largely dead bone, vascularisation occurring much more slowly in a by-pass graft, with limited contact with the host bone. All efforts to re-establish union failed and five years later the stunted leg was amputated. There is a time for amputation, which should not be delayed, and shortness of the limb is at least as strong an indication as repeated failure to obtain union.

Case 2—The second child, a girl, presented at the age of nine months with bowing of the lower end of the tibia (Fig. 7). Because of the attenuated bone, sclerosis and a rarefied...
Case 2. Figure 7—Aged 9 months, there is a congenital pseudarthrosis of tibia with typical sclerosis and bowing anteriorly and laterally. Figure 8—At the age of 3 years 9 months there is non-union. It is now twelve months since the fracture.

Case 2. Figure 9—Aged 5 years and seven months after a delayed by-pass graft had been done. There is good incorporation at each end. Figure 10—Aged 6½ years, eighteen months after the radiograph in Figure 9. The by-pass graft dominates the scene and the pseudarthrosis has united.
area in the middle of the lesion we decided that it was unwise to straighten this tibia and risk a pseudarthrosis.

The child was therefore kept under observation and it was not until the age of two and three-quarter years that she was admitted with a pathological fracture (Fig. 8). As was expected, we failed to promote union by simple immobilisation. A year and a half later, when a pseudarthrosis had developed at the age of four and a quarter years, a reinforced by-pass graft was introduced and at the same time a block was removed from the lower end of the lesional tissue to ascertain the pathology. Seven months later the graft was well incorporated at each end (Fig. 9). A year and a half later still the grafted bone was much thickened and the attenuated remnants of the original bone had united (Fig. 10). Fifteen years after the grafting operation, at the age of nineteen years, the tibia was straight and there was no shortening (Fig. 11).

**Case 3**—The third child, a girl, presented at the age of two years with established infantile pseudarthrosis (Fig. 12). The bone had fractured when the child was two months old. Reinforced by-pass grafting was undertaken, and a biopsy specimen removed at the same time showed the lesion to be fibrous dysplasia. Subperiosteal new bone and the general hyperaemia and thickening of the graft are well shown. It is to be noted that the by-pass graft was inserted without any attempt to correct the deformity. Nevertheless the tibia gradually straightened out over the years (Figs. 13 to 16). Although the pseudarthrosis united, the original lesion was later found to be enlarging and to be eroding at the lower end of the by-pass graft (Fig. 16). It was evident that further operation was necessary for the removal of the dysplastic lesion. Block excision of the original tibia with its periosteum over the involved area was therefore undertaken, and where the lesion had extended into the by-pass graft
it was carefully curetted out of the depression. This by-pass graft and the fibula retained good stability but were reinforced by the introduction of a further graft from the right tibia in the line of the excised segment (Fig. 17).

**Case 4**—The fourth child, a girl, presented at the age of three years with an oblique fracture of the tibia (Fig. 18). The bone at the site of fracture was sclerotic, and showed an area of rarefaction in the medial cortex, suggestive of non-osteogenic fibroma. A year later a pseudarthrosis had developed (Fig. 19). The child was then four years old and a delayed by-pass graft was performed. Seven months later the graft was soundly incorporated above but imperfectly at the lower end (Fig. 20). Though its full incorporation was delayed the appearance of the graft two and a half years after operation was reassuring (Fig. 21). Nevertheless we considered that external support was still required, until we could feel that this by-pass graft was fully vascularised.

**COMMENT**

In this series the four by-pass grafts were carried out between the ages of two and three-quarter years and four and a half years, with an average of three years and seven months. Union was obtained in each case. We would stress the advice to proceed to a by-pass graft without delay. Even in the younger children, good graft material can be obtained by the delayed graft technique, and the recipient site does not benefit by delay and an extended period of limited use. All four children were girls, perhaps no more than a coincidence.

We commend this sound biological operation of the late Professor McFarland to renewed attention. We have complete confidence in the operation provided the details of treatment after operation are understood and rigorously applied. It is to be noted that the two more mature patients have fully overcome any shortening and have developed normal straight tibiae. The grafting in deformity which assists in stabilising the by-pass graft has no disadvantage, and the straightening to be seen in the subsequent years is of course no surprise to anyone familiar with the moulding capacity of young growing bone.

In spite of our advocacy of the delayed by-pass graft as the standard method of treatment for infantile tibial pseudarthrosis, we agree that there are occasional children with so short a
FIG. 18

FIGS. 18 AND 19
Case 4. Figure 18—At 3 years of age. There is a fracture through a rarefied area in the tibia, possibly a non-osteogenic fibroma. Figure 19—Twelve months later the pseudarthrosis is well established.

FIG. 19

FIG. 20

FIGS. 20 AND 21
Case 4. Figure 20—Aged 4½ years, seven months after the introduction of a delayed by-pass graft which is well incorporated above but incompletely below. Figure 21—Aged 6½ years. The graft is now well incorporated at both ends.

FIG. 21
lower tibial metaphysial segment that the use of a Kuntscher nail inserted through the subtalar and ankle joints has to be condoned.

THE BACKGROUND OF INFANTILE PSEUDARTHROSIS OF TIBIA

The main histological features of tissues removed at the time of grafting are summarised in Table I.

<table>
<thead>
<tr>
<th>Case number</th>
<th>Sex</th>
<th>Age (years)</th>
<th>Dead bone</th>
<th>Reactive bone</th>
<th>Fibrosis</th>
<th>Specific tissue in the lesion</th>
</tr>
</thead>
<tbody>
<tr>
<td>1</td>
<td>Female</td>
<td>3</td>
<td>+</td>
<td>+</td>
<td>+</td>
<td>None</td>
</tr>
<tr>
<td>2</td>
<td>Female</td>
<td>4</td>
<td>-</td>
<td>+</td>
<td>+</td>
<td>None</td>
</tr>
<tr>
<td>3</td>
<td>Female</td>
<td>2</td>
<td>-</td>
<td>-</td>
<td>+</td>
<td>Fibrous dysplasia</td>
</tr>
<tr>
<td>4</td>
<td>Female</td>
<td>4</td>
<td>+</td>
<td>Slight</td>
<td>Periosteum</td>
<td>None</td>
</tr>
</tbody>
</table>

In Case 3 pre-existing fibrous dysplasia of bone was present. This has also been observed in a newly born infant briefly described below, but not included in the group of treated cases here presented. More recently another girl, one year old, has been seen with pseudarthrosis of a tibia affected by neurofibromatosis. In the Bristol Bone Tumour Registry's series of seventy-two examples of fibrous dysplasia of bone there are twelve involving the tibia. Most of the patients sustained some degree of fracture, but in only one (Case 3) did a pseudarthrosis develop. Most of the other eleven patients presented for treatment at the ages eight to fourteen years when the stouter tibial cortex may, when fractured, more readily undergo bony union, with a diminished chance of pseudarthrosis. In the congenital lesion mentioned above, however, the tibial cortex was in places less than a millimetre thick and displayed simultaneous active endosteal osteoclasis and subperiosteal osteogenesis as a result of the expanding medullary lesion. Although this thinned cortex had not yet fractured, cartilaginous metaplasia was already present among the subperiosteal new bone. This of itself may be a factor in the evolution of a false joint.

In the material studied from this small group of four children, the striking histological features have been the presence of more or less cellular fibrous tissue (perhaps scar tissue) ensheathing, sometimes separating, the fragments of a fractured bone, and the considerable activity of osteoblasts, in some instances laying down new bone on dead fragments or on the surface of an autograft.

As is well known, movement at a fracture site often induces cartilaginous metaplasia in related fibrous tissue; in fact, the latter is probably indicative of the former and denotes the "plasticity of the mesenchyme", a term attributable to Willis (1948) which most aptly describes the ability of connective tissue to adapt structure to modified function.

In our series of four children, one patient (Case 1) showed much necrosis of bone microscopically, but whether this preceded or followed the pseudarthrosis is uncertain. In the second child no evidence was found of any predisposing condition. The third case was associated with fibrous dysplasia involving the tibia. In the fourth child there may possibly have been a previous fibrous cortical defect.

Fibrous lesions of the tibia, if extensive in a young child, may well predispose to pseudarthrosis, but there appears also to be a large group of cases in which no such disease is demonstrable. In these, the radiographic appearance of a slender sclerotic deformed tibia (Fig. 7) suggests some other unrecognised skeletal abiotrophy of obscure type.
FIG. 22

FIGS. 22 AND 23
Figure 22—An infant of 2 weeks with extensive fibrous dysplasia. Figure 23—The lesion was evacuated at age of 3 weeks and the appearance at 15 months is shown here.

FIG. 23

FIG. 24

FIGS. 24 AND 25
Same case. Figure 24—The appearance at 21 months. Figure 25—The appearance at 3½ years.

FIG. 25

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In connection with causation, it may be of interest to illustrate a case of fibrous dysplasia seen in a baby of two weeks, in which infantile pseudarthrosis might well have developed had we not evacuated the lesion, which extended well above the main defect, almost to the superior metaphyseal region (Fig. 22). The evacuation of this fibrous dysplastic lesion took place at the age of three weeks. At fifteen months the outcome was somewhat doubtful (Fig. 23), but at twenty-one months we were more sanguine (Fig. 24), and at three and three-quarter years really quite hopeful (Fig. 25). There are sclerosis and the rarefaction of a fibrous dysplastic lesion in this tibia, which emphasise the importance of this diagnosis in infantile pseudarthrosis.

**SUMMARY AND CONCLUSIONS**

1. Three cases of infantile pseudarthrosis of the tibia treated successfully by delayed autogenous by-pass graft are reported.
2. The delayed autogenous graft is stouter, stronger and more easily handled and has enhanced osteogenic properties than a graft transferred immediately.
3. The by-pass graft commends itself, firstly, because it does not disturb the pseudarthrosis, which in consequence helps the immobilisation of the graft; secondly, because it is well embedded in healthy bone above and below, well away from the abnormal bone; thirdly, because it lies under compression and, ideally, is vertically disposed between the knee and the ankle; and fourthly, because there is no devitalising stripping of periosteum or introduction of foreign bodies.
4. Support to the grafted leg is needed for at least five years, but only by a polythene splint after four to six months.
5. With early grafting the deformity straightens out and shortening is overcome, as there is early return to normal use of the limb.
6. Prolonged follow-up is called for lest the basic lesion in the tibia should extend.
7. Fibrous dysplasia and similar fibrous lesions of bone account for many cases of infantile pseudarthrosis of the tibia. Many of these lesions are congenital and subsequently lead to fracture.
8. Postponement of surgery should not be countenanced.

**REFERENCES**


