EXTENDING INTRAMEDULLARY RODS IN CONGENITAL PSEUDARTHROSIS OF THE TIBIA

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Five patients with Boyd type II congenital pseudarthrosis of the tibia underwent excision of the pseudarthrosis and double onlay bone grafting. Stability was maintained by extending intramedullary rods. Clinical union was achieved in all cases at a mean of 8.6 months (range six to 11). The rods extended by 15.7% (range 2% to 31.4%) as growth occurred. One rod was removed because of infection and a vascularised free fibular graft was subsequently performed. The extending rods provided stability while union occurred and did not require revision as the legs grew. The rods can be removed easily and have not jeopardised further surgical options.

Boyd (1982) described six types of congenital pseudarthrosis, each with its own pathology, prognosis and natural history. Type II cases are the most common and have the most dismal prognosis. The children are born with anterior bowing and an hour-glass constriction of the tibial medullary canal. Fractures occur spontaneously or following minor trauma and produce tapered sclerotic bone ends. There is an association with neurofibromatosis in about half the cases.

Numerous methods to promote bony union have been described, most using some form of bone graft and stabilisation of the fracture. In 1941, Boyd described the double onlay cortical bone graft combined with cancellous bone chips. The cortical grafts seemed to prevent recurrence of the osteolytic fibromatosis associated with type II disease, and the cancellous bone promoted union. Charnley (1956) and Van Nes (1966) supported the idea of massive corticocancellous grafting in combination with rigid internal fixation, both favouring an intramedullary device.

Bitan et al (1987) reported primary union in 17 of 18 cases, achieved in a mean time of 6.5 months by intramedullary stabilisation and bone grafting. Five of the intramedullary rods became too short because of bone growth, and six revision operations were required in three other cases. In seven cases they used Bailey-Dubow

extending intramedullary rods and observed elongation of the rods with bone growth in four.

We report our experience of double onlay corticocancellous bone grafting with intramedullary stabilisation using four Bailey-Dubow and one Sheffield extending rods.

PATIENTS AND METHODS

Five boys with type II congenital pseudarthrosis of the tibia, associated with neurofibromatosis (see Fig. 1), presented at Sheffield Children's Hospital between 1983 and 1987.

Four had fractured spontaneously at an average age of 17 months (range 6 to 27). One presented at 10 months with nonunion after osteotomy to correct a congenitally bowed tibia. Two children had undergone one previous bone grafting procedure and two children had had two previous bone grafts. Electromagnetic stimulation had been tried without success in two cases. Three children required operations to correct cavus deformities of their feet before intramedullary fixation.

Bone grafting, with stabilisation by an extending rod, was performed at an average age of 6.5 years (range 3.1 to 12.5).

OPERATIVE TECHNIQUE

Pre-operative measurement allows selection of a rod approximately 2 cm shorter than the tibia and a little narrower than the medullary canal.

The pseudarthrosis is exposed via an anterior incision. The abnormal bone around the pseudarthrosis is excised back to normal bone. The knee is exposed through a medial parapatellar incision, and a medial arthrotomy is performed at the ankle. The rod is inserted

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Anteroposterior and lateral radiographs of a case of congenital pseudarthrosis of the tibia associated with neurofibromatosis. Figure 1 - Before operation. Figure 2a - After insertion of a Bailey-Dubow rod. Figure 2b - Showing elongation of the rod with tibial growth.

as described by Marafioti and Westin (1977) and Lang-Stevenson and Sharrard (1984). Cortical bone graft is harvested from the opposite tibia using the standard Boyd technique (1941).

The grafts were simply laid over the site of the pseudarthrosis in four cases, and were held in position with K-wires in one. The wounds were closed over suction drains which were removed after 48 hours.

Postoperatively, a long-leg plaster cast was applied and retained for three months, after which a plastic tibial brace was used to allow protected weight-bearing. Splintage has been continued throughout the follow-up period (mean 4.5 years; range two to six). Two patients had postoperative electromagnetic stimulation while in plaster.

Clinical and radiological follow-up has continued over a mean period of 4.5 years (range two to six).

RESULTS

The affected legs were 2.4 cm (range 1.5 to five) shorter than the contralateral normal legs. Bony union, based on clinical and radiological assessment, was achieved in all at an average of 8.6 months (range six to 11).

Rod extension (Figs 2a and 2b) was measured as a percentage of the known length of the outer sleeve of the rod. The intramedullary rods extended with growth by 15.2% (range two to 31.4). One rod extended 2% and then became fixed, subsequently migrating distally with bone growth. Excluding this case, the average extension of the remaining rods was 4.7 cm (range 1.5 to 6.4) or 19.1% (range 4.5% to 31.4%).

One patient refractured 15 months after surgery, bending the intramedullary rod. A closed manipulation straightened the rod, but the pseudarthrosis failed to reunite despite further electromagnetic stimulation. The following year, the rod was removed because of infection. Subsequently, a vascularised free fibular graft was successful and the fracture remains united after three years.

One patient fell from a height of four feet and fractured his tibia below the united pseudarthrosis (Fig. 3). The leg was manipulated into a satisfactory position, thus straightening the rod. The fracture united and the rod continued to extend with further growth (Fig. 4).

DISCUSSION

Congenital pseudarthrosis of the tibia is not a distinct clinical entity. The six different types now recognised contribute to the 'confusion and ambiguity' in regard to its treatment (Boyd 1982). Type II disease has the poorest prognosis but, if union can be achieved, the incidence of refracture decreases as skeletal maturity is approached. Intramedullary stabilisation combined with massive bone grafting is favoured by many authors (Van Nes 1966; Anderson 1976), but with growth of the bone the standard intramedullary rod becomes too short. Our experience with extending intramedullary rods (Bailey and Dubow 1963) in the treatment of osteogenesis imperfecta was encouraging (Lang-Stevenson and Sharrard 1984) and suggested their use in the management of congenital pseudarthroses. This view has since been supported by Bitan et al (1987).

Figure 3 – Tibial fracture below the united pseudarthrosis, showing the rod bent at the junction of the outer sleeve and the inner rod.
Figure 4 – Anteroposterior and lateral radiographs two years after manipulation to show continued elongation of the rod with tibial growth.

Fig. 3

Fig. 4

The 19.1% (range 4.5% to 31.4%) elongation of the rods with growth in this series is to be compared with the 16% (range 3% to 57%) elongation reported by Stockley, Bell and Sharrard (1989) in the treatment of osteogenesis imperfecta.

We agree with Boyd and Fox (1948) that an adequate brace should be worn until the child is past puberty, as the result at skeletal maturity gives a reliable indicator of long-term outcome (Crossett et al 1989). In our series, two legs refractured during unprotected weight-bearing, bending the solid element of the rod at or near the level of the pseudarthrosis. If the standard technique of rod insertion were reversed and the outer sleeve inserted distally, the stronger section of the rod would lie at the level of the pseudarthrosis, possibly reducing the chance of refracture.

Despite the enthusiasm for the use of electromagnetic stimulation expressed by Sutcliffe and Goldberg (1982), no benefit was found in this series.

The average leg-length discrepancy of only 2.4 cm compares favourably with the series reported by Bitan et al (1987).

CONCLUSIONS

In the treatment of type II congenital pseudarthrosis of the tibia, extending intramedullary rods provide stability while fusion following bone grafting is awaited.

The age of the patient at the time of operation did not influence the results and elongation of the rods occurred with growth, reducing the need for revision.

We suggest that the rod be inserted with the outer sleeve positioned distally to provide maximal resistance to bending at the site of the pseudarthrosis, and we

endorse Boyd's view that external splintage should be maintained until skeletal maturity is reached.

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