THE ROLE OF EXPANDING INTRAMEDULLARY RODS IN OSTEOGENESIS IMPERFECTA

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We report the results of using 83 expanding intramedullary rods in 24 children with osteogenesis imperfecta after a mean follow-up of five years three months. In all, 62% of the rods have expanded after one primary operation. Thirty-four additional operations were necessary; 11 for the correction of rotation or angulation deformities and 23 for revision of the rod or T-piece. All these revisions were successful. Complications were more frequent in children who required very small rods. Problems with Bailey-Dubow rods led to the development of the Sheffield rod system; 17 bones treated with these rods are included in the series.

Before surgery only eight of the 24 children were able to walk but at review 20 children were walking, 15 without walking aids. Elongating intramedullary rods should be available to all children with osteogenesis imperfecta as they improve walking capability, reduce the number of fractures, prevent deformity and allow integration of the child into society.

Multiple osteotomies and intramedullary rodding for the surgical treatment of osteogenesis imperfecta was first described 30 years ago (Sofield and Millar 1959). Problems arise with rods of fixed length when the child outgrows the rod; angulation or fracture may occur at the levels of unsupported bone.

In 1963, Bailey and Dubow produced an extensible intramedullary device for use in children with osteogenesis imperfecta and several clinical studies have been published (Williams 1965; Marafioti and Westin 1977; Albright 1981; Bailey and Dubow 1981; Lang-Stevenson and Sharrard 1984). The use of expanding rods has reduced the number of further operations as compared with fixed length rods (Marafioti and Westin 1977).

Lang-Stevenson and Sharrard reported the initial results from this centre in 1984; we now present our

further experience, which has included the development of modifications to the Bailey-Dubow system. The Sheffield rod has been used in six patients.

MATERIALS AND METHODS

Twenty-four children who had undergone intramedullary rodding with expanding rods were reviewed. Mean follow-up was five years three months (range one to eight years). There were 11 girls and 13 boys. The mean age at insertion of the first rod was eight years (range two to 14 years).

All the children were severely affected and had had multiple fractures either at birth or during infancy. The indication for surgery in all patients was the correction of deformities and stabilisation of the bones to promote mobility. Before surgery, most of the children were confined to a wheelchair.

The surgical technique used to insert the Bailey-Dubow rods was much the same as that described by Lang-Stevenson and Sharrard (1984). A major modification relates to the corrective osteotomies made during rod insertion; in the past, we produced osteotomies so that a segment could be rotated and threaded onto the rod to correct a deformity. We now cut wedges in the bone to correct angulation so as to retain maximum soft tissue attachments.

Complications due to loosening of the T-piece led to the development of the Sheffield rod system (Fig. 1). In the standard Bailey-Dubow rods, detachable drill points

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Fig. 1

The Sheffield rods and instruments.

Table I. Corrective operations for deformity in five children

Supracondylar medial rotation osteotomy	4
Stapling distal femoral epiphysis	3
Stapling proximal tibial epiphysis	2
Closing wedge upper tibial osteotomy	1
Medial rotation osteotomy tibia	1
Total	11

Table II. Summary of 23 revision operations

For migration of the rod	8
For detached T-piece	3
For cutting-out through bone	1
Rod removal for infection	4
Rod replacement after infection	3
Bone outgrown rod	1
Bent rod	2
AO plating for non-union of osteotomy	1

are screwed into the tubular sleeve of the rod to ream the medullary cavity. These drill bits tended to become loose whilst reaming, though, fortunately, none became detached inside the bone. The Sheffield rod system has a fixed and slightly larger T-piece, and separate hand drills of varying sizes are used to ream the medullary cavity. When there have been multiple fractures in a bone, the medullary cavity may not have reformed, in which case reaming is difficult and power reamers are necessary.

RESULTS

In 24 children, 117 operations on 83 bones were performed, including the 28 roddings previously reported (Lang-Stevenson and Sharrard 1984). Forty-two femora and 39 tibiae have been treated with 64 Bailey-Dubow rods and 17 Sheffield rods. Two humeri were also rodded in one patient. Of the other 34 operations, 11 were osteotomies to correct rotation and angulation deformities (Table I) and 23 were for the complications of rodding (Table II).

In all, 18 patients have required intramedullary rods for all four lower-limb bones, three patients for two bones and the remaining three patients for one bone only. The mean number of operations per patient was 4.9, the maximum number on a single patient being 10. Eleven patients had two bones rodded at the same operative session, seven for ipsilateral femur and tibia and four for

Fig. 2

Fig. 3

Fig. 4

Fig. 5

Figure 2 – Serial radiographs of a tibia to show elongation of a Bailey-Dubow rod. Figure 3 – A rod has expanded with growth and there is now a gap between the pin and the sleeve. A fracture subsequently occurred at this level. Figure 4 – Proximal migration of a femoral rod. Figure 5 – A T-piece has become unscrewed and the rod has failed to expand.

bilateral tibiae. Ten children (42%) required revision of either the rod or T-piece in 23 bones, a revision rate of 28%.

Before the surgery, 12 children had never walked, four had walked at sometime in their life but were unable to do so just before the operation, and eight were walking with walking aids. At review, 20 children were walking on dry land, five of them without the aid of rollators or crutches. Three children were walking in the hydrotherapy pool and the remaining child who had a severe kyphoscoliosis was confined to a wheelchair.

Relative elongation had occurred in 62 of the rods (89%) (Fig. 2), the mean increase in length being 16% of the original length (range 3 to 57%). No assessment could be made of 10 rods because the radiographs were inadequate for measurements. One patient outgrew one of his rods and a fracture occurred where the bone was unsupported in the gap between the two components of the rod (Fig. 3). After revision the fracture united.

Eleven rods failed to elongate. In four cases, this resulted from poor fixation in the epiphysis (Fig. 4) and in one case the T-piece became unscrewed from the rod (Fig. 5). Three patients had fused epiphyses at the time of surgery, and one child with a leg length discrepancy had undergone stapling of a lower femoral epiphysis. No

Fig. 6

The obturator pins are not inside the sleeves.

cause could be found in the other two cases for the failure of the rods to extend.

No radiographic evidence has been seen of any growth disturbance secondary to the crossing of the epiphyses by the rods, nor has there been premature closure of an epiphysis. Only one osteotomy failed to unite. This occurred in a child who was 16 years old at operation and has been successfully treated by AO compression plating and cancellous bone grafting.

Seven children in our care had previously undergone intramedullary nailing of 13 limbs in other centres, using non-expanding rods such as Steinmann pins, Rush nails or Kuntscher nails. All failed with time, as part of the growing bone became unsupported, and this led to deformity and fracture. In one patient from another centre, two Bailey-Dubow rods had been inserted incorrectly (the obturator pin never engaged the sleeve) and both bones became infected (Fig. 6). All these 15 rods were replaced by expanding intramedullary rods with satisfactory results to date.

Table III. Walking ability related to range of hip rotation

Number of patients	Range of rotation	Walking ability
4	External = internal	Good, no aids
5	External > internal	Good, one with no aids
3	Full external rotation, internal rotation to neutral only	Only with aids
7	Fixed external rotation deformity	3 with aids 3 in hydrotherapy pool only 1 wheelchair
5	Not recorded	Only with aids

The range of rotation at the hip was measured in 19 children and related to their walking capacity (Table III). It seems that children with good symmetrical ranges of rotation were excellent walkers, but those with rotary abnormalities were less able. External rotation deformities were so gross in five femora that medial derotation osteotomies were needed, but despite this corrective surgery, the deformity recurred in all five limbs.

The incidence of fractures following rodding has been considerably reduced in the new series. There was clinical and/or radiological evidence of 26 lower limb fractures in 12 children since the rods were inserted. Five children had sustained their fractures in falls while walking. Twenty of the fractures were treated conservatively, either by simple bed rest or immobilisation with wool and crêpe bandages or plaster. Only four fractures were secondary to lack of support by a short rod, either

because the rod had failed to expand or because the patient outgrew the rod. These were all successfully revised. In the other two fractures, the rod had been bent excessively at the time of the injury; revision was required although the original fracture had united. No rods have broken but two tibial and two femoral rods have bent. In three cases, there were associated fractures of the bone and in two revision was necessary.

Six children have been treated with the Sheffield system. Seventeen rods, 10 femoral and seven tibial have been used with a mean operative follow-up of one year and five months (range one to two years). To date only one complication has arisen, proximal migration from the distal femoral epiphysis. This was minimal and no revision surgery is planned. In comparison, six of the first 17 Bailey-Dubow rods had required revision within two years of the initial operations.

Pelvic radiographs before operation and at review were of sufficient quality to measure the neck-shaft angle in 30 of the 42 femora rodded. In 10 bones there had been no change in this angle but in 17 the neck had become more varus by 5 to 20° (mean 10°). In several of these femora, the rods had been inserted somewhat laterally rather than through the base of the greater trochanter. In consequence the proximal femur was unsupported medially and the neck drifted into progressive varus. In the other three femora, the neck had become more valgus by an average of 27° (range 5 to 50°). In two cases, valgus osteotomies were made at the time of rod insertion to correct pre-operative varus angulation.

Most problems with the elongating rods were in small or young children: 70% of the rods which failed to expand, bent, or migrated from their epiphyseal insertions were small in both length and diameter. They had been inserted in children who were under five years old at the time of surgery or in older children who were very small for their age.

COMPLICATIONS

Infection. Four rods became infected. One child had had an infection with a previous fixed length rod; after revision to the Bailey-Dubow system, infection recurred despite antibiotic therapy. The rod has since been removed and the infection has resolved. Two infected rods were in one patient, 24 and 33 months after primary insertion. The fourth case presented with a discharging sinus 13 months after tibial rodding. These last three cases were all re-rodded three to six months after removal of the rod, following antibiotic therapy. After a minimum follow-up of five years there have been no sequelae.

Migration of the rod. Sixteen rods appeared to have migrated from their epiphyseal insertions, 10 in the femur and six in the tibia. Eight femoral rods had migrated proximally from the distal epiphysis (one from the metaphysis into which it had been wrongly placed)

and two migrated distally from the proximal end of the femur; one of these had a detached T-piece (see Fig. 5). Four tibial rods had migrated proximally and one had migrated distally (Fig. 7).

Six of the femoral rods and two tibial rods have required revision, the commonest indication being the development of a fracture or threat of a fracture at the junction of the end of the rod with unsupported bone. All eight revisions are satisfactory after follow-up of 18 months to six years.

T-piece. Despite careful tightening, the T-piece became unscrewed in eight cases and in three of these it became completely detached. Two of the three have been successfully replaced and the rods have continued to lengthen, but in the third, the T-piece could not be found at operation and the rod subsequently failed to lengthen. The other five T-pieces, although loose, have not become detached and their position has been accepted.

Intra-operative complications. During placement of the proximal end of one rod, the anterior femoral cortex was broken and the rod failed to extend. This was revised satisfactorily.

There have been no vascular, neurological or wound healing problems despite the extensive skin incisions in many of the patients. Although four rods have needed revision because of infection there have been no joint infections in the series.

DISCUSSION

Our results provide more confirmation of the reported benefits of extensible intramedullary rods in children suffering from osteogenesis imperfecta (Marafioti and Westin 1977; Bailey 1981; Niemann 1981; Lang-Stevenson and Sharrard 1984). In our series 50% of the children have had no further lower limb fractures since surgery. The improvement in these children's lifestyle persists into adult life; Moorefield and Miller (1980) have shown how they integrate with society.

The main question asked by some authors (Middleton and Frost 1987) is whether elongating rods are really necessary: they require major surgery, and considerable complications have been reported (Lang-Stevenson and Sharrard 1984). Frost, Middleton and Hillier (1986) have designed a stereotactic device to allow closed retrieval and interchange of intramedullary rods. This relatively atraumatic procedure allows more frequent rod exchanges, but such exchanges require many hospital admissions and many anaesthetics during childhood. After multiple fractures, bones often show very little medullary cavity; it would be difficult to achieve stereotactic placement within this hard bone. Even where the bone looks wide on radiographs, Lang-Stevenson and Sharrard (1984) have shown that it often resembles a rib, with only a very small area available for the rod. Indeed, Middleton and Frost (1987) failed to re-

insert the rods by their closed method in one-third of their cases.

No bones were found to be too small for the rod system; we strongly disagree with Ryöppy, Alberty and Kaitila (1987) who state that telescoping nails cannot be used in infants because of the small calibre and the fragility of the bones, though we agree that more problems arise in children less than five years old who require small rods.

Of the 83 rods inserted, 51 (62%) have elongated after one primary operation without T-piece loosening or migration, and only one bone has outgrown the rods. Most complications should be avoidable by inserting the

Fig. 7

The T-piece was not rotated under the articular cartilage to fix the rod; as a consequence there was distal migration of the rod.

rods in the right orientation with careful technique, and ensuring that a wide range of sizes of rod are available.

When the patient is supine, the lower limbs tend to lie in external rotation, and after intramedullary rodding (despite placing the limbs in internal rotation in a hip spica) this deformity may recur. This may be due to microfractures at the osteotomy sites; rotational forces cannot be neutralised by elongating intramedullary rods.

We have no experience in the use of pneumatic orthoses. Ross et al. (1986) have shown that 58% of children treated with them were walking at review as against 10% at the start of their study. No lower limb fractures occurred whilst their patients were wearing an

orthosis, but on average they were worn for only three hours a day. Patient selection was important and treatment was only offered to 19 of the 30 referred patients. A disadvantage of the orthosis is that it is cumbersome and this may compromise the patient's improved mobility. Rodded patients have the potential to walk at all times and not only when they are wearing an orthosis.

The insertion of elongating rods is associated with a learning curve; in our series 10 different surgeons of varying experience performed the operations. Since the initial report from this centre (Lang-Stevenson and Sharrard 1984), the incidence of revision surgery has decreased slightly (32% to 25%). The revision operations have been successful and no further surgery has been necessary over a follow-up in several cases greater than five years. Rod exchange, when required, is not difficult; we can see no place for the expensive and cumbersome

stereotactic device advocated by Middleton and Frost (1987).

Continual use of the Sheffield rod system may further reduce the number of complications, since the T-piece is fixed and cannot become loose. The problems of relatively high cost and difficult availability of the Bailey-Dubow rods (Lang-Stevenson and Sharrard 1984) have been overcome by the less expensive and more readily available Sheffield rods.

We believe that expanding intramedullary rods should be available to all children with osteogenesis imperfecta, because they reduce the incidence of fractures, prevent deformity, and improve walking capability. This allows better integration into society and a better quality of life.

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