INTRAMEDULLARY RODDING WITH BAILEY–DUBOW EXTENSIBLE RODS IN OSTEOMEGENESIS IMPERFECTA

AN INTERIM REPORT OF RESULTS AND complications

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The results and complications of the use of Bailey–Dubow extensible rods in 28 lower limb bones of 10 patients suffering from osteogenesis imperfecta are reviewed. Twenty-eight operations were for the primary insertion of the rods into the femur or tibia; a further nine operations were needed for the treatment of complications. These complications included 10 instances of proximal migration of the distal end of the rod, one of incorrect placement in the proximal femur, four instances of loosening of a T-piece and three of infection about a rod, two of these being in one child. Most complications arose from technical faults at insertion. The details of technique which have evolved from experience are described. Only one fracture has occurred in a bone after correct placement of a rod. Of the 10 patients, seven of whom had never walked before, seven were able to walk and two others had achieved walking, but were under treatment for complications at the time of review. There was no evidence of damage to growth epiphyses. The greater technical complexity of insertion of Bailey–Dubow rods is well justified by the results obtained when they are correctly applied.

Multiple osteotomy and internal fixation with an intramedullary rod from metaphysis to metaphysis was first described by Sofield and Millar in 1959. This method has been extensively used in the treatment of osteogenesis imperfecta (Marafioti and Westin 1977; Bailey 1981; Niemann 1981). In children non-extensible rods are outgrown by the bone with the development of angulation or fracture in the unsupported bone distal to the tip of the rod; frequently the tip of the rod penetrates the cortex (Sharrard 1979; Bailey 1981). Most authors agree that non-extensible rods in children need to be revised every two to two and a half years (Williams 1965; Marafioti and Westin 1977; Albright 1981; Bailey and Dubow 1981), with the possibility of a revision operation almost every six months (Bailey 1981).

In 1963 Bailey and Dubow introduced extensible rods (Fig. 1) comprising a hollow sleeve with an internal obturator. With these rods the number and frequency of operations during growth could be considerably reduced (Marafioti and Westin 1977; Bailey 1981; Niemann 1981). Extensible rods are more difficult to insert and they have their own complications. This paper reviews experience of insertion of extensible rods since October 1979 and the complications encountered, in the treatment of 10 children suffering from osteogenesis imperfecta.

MATERIAL

The records and radiographs of eight girls and two boys suffering from osteogenesis imperfecta were reviewed; their mean age was eight years and eight months with a range of four years and seven months to 14 years. All the children were severely affected and had suffered multiple lower limb fractures at birth or during infancy. The Bailey–Dubow rods had been inserted into the deformed femora and tibiae at the Children's Hospital or King Edward VII Orthopaedic Hospital, Sheffield. The mean age at the insertion of the first rod was six years and six months with a range of two years to 12 years. The mean
time of follow-up was two years and two months. The rods were inserted because of increasing deformity which was impairing function or making fractures more likely, or because of recurrent fractures.

METHODS
Radiographs in anteroposterior and lateral planes were used to plan the exposure needed and the approximate sites for osteotomies. In general, more osteotomies were needed than initial radiographs suggested because medial or lateral bowing as seen in the anteroposterior radiograph was often at a different level from anterior or posterior bowing seen on the lateral radiograph.

Exposure. There was usually need to expose the shaft of the bone along its whole length. The deformed sections of bones were exposed subperiostally. This was done in short segments at a time to minimise blood loss, since children with osteogenesis imperfecta bleed easily and are small for their age, with a relatively small blood volume. In a few patients with a single level of deformity, only a short length of shaft required exposure and separate incisions were used to insert the rods proximally and distally.

For the femur, the exposure was posterolateral with forward mobilisation of the quadriceps. The quality of bone in osteogenesis imperfecta varies considerably, some bones being almost normal, while others have the soft quality of balsa wood. Mobilisation of the periosteum by levering against the bony surface with an elevator was avoided. Exposure of the greater trochanter proximally and of the knee distally was left until last, the knee being opened through an anterolateral continuation of the shaft incision or through a separate anteromedial incision.

The tibia and knee were exposed through an anteromedial incision. The ankle was exposed through a medial approach with division of the deltoid ligament. Osteotomy and rod insertion. A rod of the largest diameter which would fit through the narrowest diameter of the shaft was selected. The length of the rod chosen was such as to allow a little collapse as the osteotomised segments settled together. The suggestion by Marafioti and Westin (1977) that the rod should be two centimetres less than the calculated length measured clinically before operation was found to be acceptable. Osteotomies were made so that, if appropriate, a segment could be rotated and threaded onto the rod to correct a deformity. Wherever possible the periosteum was left attached to the bone. A short section of bone was sometimes removed if soft tissue tension so indicated, or if the diameter of the section was too narrow for the rod. A small wedge was removed if necessary to improve apposition of the bone ends. The technique of insertion of the rod was similar to that described by Marafioti and Westin (1977).

In the femur, the first osteotomy was just above the apex of the most proximal bowing (Fig. 2-A). The medullary canal was drilled out towards the greater trochanter using a detachable drill point screwed into the tubular sleeve of the rod (Fig. 2-B) and the sleeve was advanced until its distal end was nearly flush with the osteotomy. The drill was then attached to a separate sleeve of the same diameter, introduced through the distal side of the osteotomy and advanced along the medullary cavity until it impinged on the cortex at the next part of the curve (Fig. 2-C). The next osteotomy was made at this site (Fig. 2-D), the second drill removed and the definitive sleeve advanced across the osteotomy site into the reamed medullary cavity of the next segment (Fig. 2-E). This procedure was repeated until all curvature had been corrected, the whole sleeve introduced and the medullary cavity drilled down to, but not beyond, the level of the distal metaphysis. If the segments required impaction a sleeve of large enough internal diameter was placed over the definitive sleeve and tapped lightly against the cortex to push the segments together.

The distal obturator was then tapped through the articular cartilage of the knee at the anterior part of the intercondylar notch without drilling. It entered the distal metaphysis and engaged the sleeve. The passage of the obturator in the correct direction, so that it passed into the medullary canal to engage the sleeve was critical, and often difficult. A second obturator inserted from the distal osteotomy site was sometimes used as a guide and also to provide a narrow track along which the definitive...
obturator could be passed (Fig. 2-F). Excessive reaming of the distal metaphysis or multiple puncturing of the distal epiphysis was avoided to prevent later proximal migration of the obturator. The sleeve and obturator were then engaged fully by driving the sleeve downwards and the obturator upwards (Fig. 2-G).

Finally, the proximal free T-piece was screwed firmly into the proximal end of the sleeve and placed in the digital sulcus of the upper end of the femur and the T-piece of the obturator placed just beneath the articular cartilage of the knee (Fig. 2-H). The placing of the obturator T-piece is a delicate manoeuvre if over-penetration is to be avoided and was therefore left until last.

Insertion of tibial rods was done in the same way with the sleeve proximally and the obturator distally. Earlier in the series, some femoral rods were put in with the sleeve distally and the obturator proximally. It was soon found that this produced a much greater instance of proximal migration of the distal end of the rod and this mode of insertion is not now recommended.

**Choice of rod.** Differences in the lengths of femoral and tibial rods needed for children of various ages required that a set of rods varying between 10 centimetres (4 inches) and 25 centimetres (10 inches) was needed. Although it was possible to make a clinical and radiological estimate of the length of the rod that would be appropriate, the correct diameter was much more difficult to forecast. Some bones that had appeared to be quite wide radiologically were frequently found on section at operation to be shaped like a rib (Fig. 3) with only a small portion capable of accommodating a rod. Five possible diameters were available. Thus, the possible permutations of the length and diameter that might be needed required that a choice of at least 35 rods had to be available if a rod of the correct diameter and within 2.5 centimetres of the correct length was to be used. It was not thought advisable to cut the sleeve at the time of operation since this could produce distortion of the tube and loss of the sliding mechanism.

**Rehabilitation.** Mobilisation of the limbs and establishment of walking took a considerable time, often as long as six months. Some children had never walked and those who had were frightened of weight-bearing because of previous experience of repeated fractures. Muscular weakness seemed to be a feature of the disease. Graduated exercises, hydrotherapy and the judicious use of light external support were needed to achieve walking on dry land, usually with the help of a walking aid. Excessive stress on the upper limbs by trying to progress too rapidly to elbow crutches or sticks was liable to cause fractures in the upper limb.

**RESULTS**

Thirty-seven operations on 28 bones (femora and tibiae) were performed in 10 patients (Table I). Twenty-eight operations were for the primary insertion of Bailey–Dubow rods, 17 in the femur (Figs 4 and 5) and 11 in the tibia (Fig. 6). The remaining nine were for complications, one of which occurred at the time of operation and eight after previous rodding. The mean number of operations per patient was 3.7. The maximum number of operations on a single patient was nine.

At review of the 10 patients, one was walking unaided, and six did so using a walking aid. Three patients were not walking; two of these had used a walking aid after rodding but at review were unable to walk because of late infection which necessitated removal of the rod. Since then, the infection has been eliminated and new rods have been inserted with the prospect of walking once more. One patient was in a plaster spica after a recent operation, but since review he has regained walking ability. Seven of these patients had not walked previously and nine had had multiple fractures of the lower limb bones before Bailey–Dubow rodding.

The period in hospital ranged from three weeks to two years and four months. The long period of two years and four months related to one patient with difficulties in parental management at home and also the need for prolonged hospital care because of infection. If this exceptional case is discounted, the average length of stay in hospital, including that for physiotherapy, was 17 weeks per long bone. In five instances, the stay was less than 1.5 weeks per long bone.
Relative elongation of the rod had occurred in 17 out of 26 of the rods inserted for the first time (Figs 4 and 5). In two patients the operation had been too recent to allow assessment. No patients have outgrown their rods since the method was started three years ago. No premature closure of an epiphysis has been seen nor has any deformity arisen from damage to part of an epiphysis. None of the osteotomies has failed to unite.

Only one patient in this series had previously undergone intramedullary nailing with conventional non-expanding rods, either Steinmann pins or Rush nails, before Bailey-Dubow rods became available. Over a six year period she had 12 rods inserted, five in one tibia, two in the other tibia, two in one femur and three in the other femur. All these bones had suffered at least one fracture while a rod was in situ and some had multiple fractures. Since she had Bailey-Dubow rods inserted in October 1979 none has required revision. She had not previously been able to walk. She is now 12 years 6 months old and is able to walk with a walking aid.

Only one patient suffered a fracture when a rod had been satisfactorily placed, and had elongated with growth so that the ends had remained in the epiphyses. This patient fell and bent the obturator at its junction with the sleeve (Fig. 7). Union in an angulated position was accepted since the patient could walk and was symptom free; an elective revision will probably be needed later. Two other patients have each sustained a fracture; in each, there had been a technical fault. In one patient the rod was very close to the anterior cortex and in the other the distal end of the rod migrated proximally leaving unsupported bone which fractured under the stresses of weight-bearing. No adverse effects were seen from arthrotomy of the ankle and satisfactory stability and
normal movement were regained after suture of the delto-oid ligament. Some limitation of flexion of the knee was observed in patients who had required tibial and femoral rodding, but all had flexion to a right angle or more. When the knee has been opened on a second occasion six months or more after the primary rodding, the presence of cartilage covering the T-piece has been noted, confirming the findings of Marafioti and Westin (1977).

COMPPLICATIONS

Migration of the end of the rod. There were 10 cases in which the distal end of the rod migrated proximally, six in the femur and four in the tibia. Five of the six femoral rods which showed proximal migration of the T-piece at the knee had been inserted with the sleeve placed distally (Fig. 8). In these five knees, the distal part of the metaphysis and the distal epiphysis of the femur had been drilled to accommodate the diameter of the sleeve (Finidori et al. 1979) and it seems likely that, for this reason, the distal T-piece migrated proximally instead of remaining in the epiphysis. Four tibial rods showed proximal migration of the distal end, and of these three had been inserted with the sleeve distally and one with the obturator pin distally. There was no case of downward migration of the T-piece from the upper tibial epiphysis or from the digital sulcus of the greater trochanter.

So far, three of the femoral rods that had migrated proximally have required revision. One became loose and then septic, requiring removal of the rod. The second was replaced but three months later also developed sepsis and required removal (see below under infection). The third patient developed a fracture at the junction of rod and unsupported bone. A satisfactory revision has been carried out.

Anterior placement of the rod. Early in the series, a femoral rod failed to elongate because of incorrect placement of the proximal end, which breached the anterior cortex of the femoral metaphysis. The fixation was satisfactory in that walking was possible with a walking aid. After 14 months, the threat of continued cutting out through the cortex led to a successful revision operation.

Unscrewing of the T-piece from the sleeve. The T-piece became unscrewed (Fig. 9) in four cases and as a result one rod failed to lengthen. Two T-pieces have been replaced and the third will need revision. The fourth has been accepted since bone growth is almost complete. Unscrewing occurred in spite of apparently adequate tightening of the T-piece into the thread of the sleeve. Since these cases presented, the thread of the T-piece has been scored before insertion to produce a tighter fit and the problem has not recurred.

Infection. Two patients developed infection, one in relation to two rods and the other in relation to one rod. In the first patient both right and left femora had had satisfactory rods in place for 12 months and 11 months respectively. An attempt to place a rod in the right tibia was aborted because of turbid fluid in the knee. Twelve months later the right femoral rod was revised because its distal end had migrated proximally. Three months after this the revised rod was removed because a sinus had developed. The initial rod had lasted 24 months and the revision only three months. The left femoral rod had been in place for 11 months when turbid fluid was found in the other knee. The rod appeared to be satisfactory until, 33 months after insertion, pus was found around the distal end when revision for proximal migration was attempted. The rod was removed. Rods have since been reinserted into both femora after three months of plaster and antibiotic treatment and there has been no recurrence of infection.

In the second patient a sinus developed down to a tibial rod, which had been satisfactory for 13 months after operation. The sinus dried up during treatment with flucoxacinil but recurred when antibiotics were stopped. The rod was eventually removed seven months later. The sinus has since remained healed and the rod has now been replaced.

In neither of these two cases was any growth of bacteria obtained on culture, although Gram-positive cocci and Gram-negative rods were seen on microscopy in one. Both patients were given antibiotics before and after operation. It seems probable that haematogenous spread of infection was the cause in the case with bilateral infection. No nidus of infection was identifiable in the other.

Excessive rod length. One femoral rod was 2 centimetres too long and failed to lengthen with bone growth.

DISCUSSION

Bailey–Dubow rods have been successful in correcting deformity, in allowing bones to resist the stresses of weight-bearing and in preventing recurrent fractures and deformity. Ability to walk, previously not possible in severely affected patients, has been achieved in all patients when satisfactory rods have been placed. Two disadvantages of Bailey–Dubow rods are their relatively high cost and the considerable delays that have been experienced in obtaining replacements for rods that have been used.

The technique of insertion of these rods was more difficult than that of non-telescopic rods, but even if a rod failed to elongate or was badly placed, it required revision later than a conventional rod, since it initially
tranversed both epiphyses whereas the latter was inserted from metaphysis to metaphysis. As originally described by Sofield and Millar (1959), multiple osteotomies did not give rise to non-union nor did any epiphysial injury occur by passing the rod through it (Siffert 1956; Bailey and Dubow 1963). The complication of infection necessitated removal of the affected rods, but after this symptoms and signs resolved quite rapidly, and were paralleled by a fall in the raised erythrocyte sedimentation rate. No organism was firmly identified in our two cases, though staphylococcal infection was strongly suspected in both, since the sinus in one case became quiescent on flucloxacillin therapy, and a Gram-positive coccus was identified in the second case. Millar (1981) suggested that children suffering from osteogenesis imperfecta were resistant to osteomyelitis and he, too, found that removal of the affected rod was followed by resolution of infection. Marafioti and Westin (1977) in their series allowed a period of 18 months quiescence after removal of infected rods but infection recurred in the same bone 18 months after rods were re-inserted. They also had a patient like one reported above in which haematogenous spread of the same bacterium occurred from one infected rod to other rods.

Bailey-Dubow rods are considered to be the treatment of choice in growing children suffering from deformity or multiple fractures due to osteogenesis imperfecta. Most of the problems associated with them are avoidable with careful technique, although infection does remain a significant complication.

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REFERENCES