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Osteogenesis imperfecta in Zimbabwe: a comparison between treatment with intramedullary rods of fixed-length and self-expanding rods

W.J. HARRISON AND K. C. RANKIN
Mpilo Central Hospital and King George VI Rehabilitation Centre, Bulawayo, Zimbabwe

The results of surgical treatment of 15 children with osteogenesis imperfecta in Bulawayo, Zimbabwe are reviewed. A total of 23 self-expanding and 27 fixed-length rods were used. Outcome was measured in terms of mobility status, growth, incidence of refracture, need for reoperation, and complications. Eight of the children improved their mobility status over the course of treatment. Self-expanding rods appeared to confer more benefit to growth than fixed-length rods. Refracture was more common in bones splinted with fixed-length rods and more often necessitated revision surgery in these bones. The complication rate was high in all cases, but the complications associated with outgrown fixed-length rods were a particular problem. The 15 children benefited from surgical treatment. The self-expanding rods performed better than fixed-length rods in reducing the number of surgical interventions. They also appear to facilitate growth. The self-expanding rods may be used to good effect in appropriate centres in the developing world.

Keywords: osteogenesis imperfecta, internal bone fixation.

The brittle bone disease, osteogenesis imperfecta, is found in all races, and the incidence in the developing world is thought to be similar to that in Western Europe. However, geographical prevalence is recognized, and in the Southern half of Zimbabwe this condition presents with great regularity.

Osteogenesis imperfecta represents a group of genetic disorders with impaired synthesis of type 1 collagen. There is currently no biological solution to this condition and the mainstay of treatment in the severe forms is surgery. Surgical intervention aims to correct and prevent the deformity which results from multiple fractures (Figure 1).

Figure 1 Pre-operative rotational and angular deformity of the tibia in severe osteogenesis imperfecta.

MATERIALS AND METHODS

Patients

During the period 1982—94 a total of 4O children with osteogenesis imperfecta received treatment in Bulawayo, of these 15 were treated using Bailey Dubow (1963)[1] self-expanding intramedullary rods. These 15 patients (10 females and 5 males) form the focus of this study. Most were admitted to the rehabilitation centre at between 5 and 7 years of age. The diagnosis was made at this time and surgical treatment commenced. Average age at commencement of surgery was 5 years 3 months, and at the time of review was 13 years 6 months (range 7—18 years).

Bailey Dubow rods were limited in availability, so each of the 15 children had extensible rods in some bones and fixed-length rods (usually Rush nails or Kuntscher nails) in other bones. A total of 23 Bailey Dubow rods were used

and 27 fixed-length rods. The mean time since insertion of the extensible rods was 82 months (range 33—109 months).

All extensible rods and most of the fixed-length rods were inserted by the same surgeon using the methods of fragmentation realignment as described by Sofield and Millar (1954).[2] The use of this technique with extensible rods was that described by Marafiotti and Westin (1977).[3] This technique entails exposure of the whole length of the bone and multiple osteotomies. In the case of the Bailey Dubow nails the joints above and below the affected bone must also be opened to allow insertion of the T-pieces across the growth plates. Most of the rod insertions were carried out as primary treatment but a few were done as revision procedures.

A summary of patient details is provided in Table 1. All the children in the study had severe forms of osteogenesis imperfecta (Sillence types 3 and 4). Child 15 lived in Zanzibar and was unavailable for review at the time of this study. Child 7 died of asthma during the study period but after the data had been collected.

Table 1 Summary of patient details

Patient number	Age	R. Femur	L. Femur	R. Tibia	L. Tibia
1	16	SER	FLR	SER	FLR
2	14	FLR	FLR	SER	SER
3	16	FLR	FLR	SER	FLR
4	8	FLR	SER	FLR	FLR
5	15	FLR	SER	NIL	NIL
6	15	FLR	SER	NIL	NIL
7	16	FLR	SER	FLR	SER
8	18	SER	SER	NIL	NIL
9	11	FLR	SER	NIL	NIL
10	16	FLR	FLR	SER	SER
11	18	SER	FLR	BKA	FLR
12	7	SER	SER	NIL	FLR
13	13	SER	SER	FLR	FLR
14	11	SER	FLR	FLR	SER
15	8	FLR	SER	FLR	FLR

(SER= self-expanding rod, FLR= fixed-length rod, BKA = below knee amputation)

Assessment

A retrospective review was carried out by the first author, who was not involved with any of the surgery. The review was based on clinical examination and limb length measurement, review of case notes, radiographic assessment including a fresh radiographic examination of each femur and tibia and review of previous radio-graphs from the time of initial surgery.

Assessment of the benefit of therapy was made in terms of mobility status, growth, and incidence of refracture. Complications were also documented and evaluated.

Mobility status at the time of review was compared with that at presentation to the rehabilitation centre. Each child was allocated to one of four categories as follows: (a) unable to walk independently; (b) walks with aids but house-bound; (c) walks with aids in the community; (d) walks unaided.

Limb length was measured clinically. Femora were measured from the anterior superior iliac spine to the adductor tubercle. Tibiae were measured from the adductor tuberele to the tip of the medial malleolus. Radiographic measurements of length were also taken but were deemed less accurate than clinical measurements because joint contractures resulted in projection errors.

In assessing the results, particular comparison was made in the 12 cases (eight femora, four tibiae) where an individual had a self-expanding rod in one side and a fixed-length rod in the equivalent bone on the contralateral side (Figure 2). This allowed comparison to be made between the performance of self-expanding and fixed engths rods in terms of effect on growth and complication rate.

Figure 2 Radiograph allowing comparison between a self-expanding rod in the right femur and a fixed-length rod in the left femur. The right femur is clearly longer.

RESULTS

Mobility status

Four children were unable to walk independently, three children walked with aids but were house-bound, six children walked with aids in the community, and one child walked unaided (Table 2).

Table 2 Summary of patient mobility status

Mobility Status	Pre-treatment	Review
A non walker (wheelchair-bound)	9	4
B household walker	3	3
C community walker	2	6
D walks unaided	0	1

Some children had a mobility at review below their usual status because of a recent complication. Overall, one child had regressed one category in mobility status, five children went up one category, and three children progressed by two mobility categories. The four children who were wheelchair-bound had good sitting balance and were able to use their wheelchairs to good advantage.

Growth

The length comparison measurements in the 12 cases that allowed cross comparison between fixed-length and self-expanding rods are shown in Table 3. Only a difference of 1 cm or greater was considered significant. Eight children had greater length on the side with the self-expanding rod, three children had greater length on the side with a fixed-length rod and one showed no difference. These results were subjected to statistical analysis employing simple binomial and normal distribution methods. This analysis gives a significance level of 0.19 and 0.147, respectively. Furthermore, this analysis is based on the premise that leg lengths were equal at the beginning of surgical treatment. The data is not available to substantiate or refine this premise. In either case, a possible benefit to growth conferred by self-expanding rods is not statistically significant.

Only two children outgrew the self-expanding rods whereas 14 children outgrew fixed-length rods.

Table 3 Comparative limb lengths in patients with self-expanding rod (SER) in one long bone and a fixed-length rod *(FLR)* in his/her contralateral bone. Difference is marked negative where the side with the FLR is longer. Patient number refers back to Table 1

Patient number	Bone	SER	*FLR*	Difference
1	Femur	33.0	32.0	1.0
4	Femur	32.5	29.0	3.5
5	Femur	32.0	34.0	-2.0
6	Femur	34.0	34.0	0.0
7	Femur	37.0	34.0	3.0
9	Femur	30.0	29.0	1.0
11	Femur	*26.0*	28.0	-2.0
14	Femur	26.0	23.0	3.0
1	Tibia	34.0	33.0	1.0
3	Tibia	29.0	33.0	-4.0
7	Tibia	35.0	32.0	3.0
14	Tibia	*29.0*	27.0	2.0

Refracture

Four refractures occurred in the presence of self-expanding rods and eight occurred in the presence of fixed-length rods. In none of the four cases with the self-expanding rods was revision surgery precipitated by fracture although one resulted in a bent rod (Figure 3). In this case surgery was deferred and the fracture healed without revision being undertaken. The other fractures were effectively splinted by the self-expanding rods. Fractures in the bones with fixed-length rods commonly occurred at the ends of the outgrown rods and, thus, required revision surgery.

Figure 3 Radiograph showing several complications in a femur with a self expanding rod namely: Fracture, rod bending, T-piece detachment, and rod migration.

Reoperation

In the time of the study one self-expanding rod was revised and nine fixed-length rods were revised. Other roddings in both groups would benefit from revision but service commitments dictated that revision was normally precipitated only by fracture in outgrown rods.

Complications

The complications recorded over the time of the study are shown in Table 4. Complications such as failed expansion and T-piece detachment are clearly only applicable to self-expanding rods. All complications are shown whether or not they led to clinical detriment.

In total, there were 23 complications in 14 (64%) of the 23 self-expanding rods and 40 complications in 25 (93%) of the 27 fixed-length rods. In both groups, this represents a high complication rate and comparison is rather artificial. The self-expanding rods are more prone to implant failure whilst the fixed-length rods bear the complications relating to outgrown rods. Even the self-expanding rods that failed to expand took much longer to outgrow because at insertion they spanned epiphysis to epiphysis, compared with metaphysis to metaphysis in the case of the fixed-length rods.

In no cases did a self-expanding rod cause premature closure of a growth plate. Only one self-expanding rod (5%) was complicated by infection.

Table 4 Summary of complications

Complication	SER	FLR
Failed expansion	7	N/A
Cortex penetration	3	13
Repeat fracture	4	8
Infection	1	0
Rod migration	2	5
T-piece detachment	3	N/A
Rod bending	1	0
Outgrown rod	2	14

(N/A = not applicable)

DISCUSSION

For some time now the benefits of intramedullary rodding of long bones in severe forms of osteogenesis imperfecta has been widely accepted. Self-expanding Bailey Dubow rods have been available for 30 years and several reports of their use in the developed world have been published. Most of these reports have been favourable[4-7] but one recent report described no benefit of self-expanding rods over fixed-length rods.[8]

This present report describes successful use of self-expanding rods in the developing world. They are effective in promoting mobility by splinting bones in a good functional position. The self-expanding rods are considerably more expensive than their fixed-length counterparts but this difference is offset by reducing the frequency of surgical interventions.

It is our impression (although we accept that the design of this study did not provide statistical evidence to substantiate this) that the self-expanding rods confer advantage in growth, when compared with fixed-length rods. This impression is gained from the 12 cases of cross comparison available in our study. The benefit to growth may result from: fewer operative insults; fewer fractures, or better splinted fractures; less deformity; or improved splinting in the growth regions at the epiphyses. Certainly, there was no evidence for self-expanding rods impairing growth as might be feared in an implant that crosses the growth plate.

Our complication rate was high in both the self-expanding and fixed-length rods. The figure of 64% of the self-expanding rods suffering complications is similar (69—72%) to that in several previous reports.[4,5~9] Concerns that the setting for the surgery might be associated with a high infection rate were not borne out by this study, with only one deep infection following surgical intervention.

This is the first published report of the surgical treatment of osteogenesis imperfecta from Central and Southern Africa. We believe that the condition can be managed with good effect and that the self expanding rods are most beneficial in reducing the number of fractures and operations for each child and promoting improved quality of life.

CONCLUSION

Patients with severe forms of osteogenesis imperfecta responded well to intramedullary rodding, and self-expanding rods proved better that fixed-length rods.

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Correspondence: Mr W J Harrison, 23 Harwood Drive, Simonside Park, Killingworth, Newcastle upon Tyne NE12 OFQ UK.

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