Modified Sofield-Millar operation: less invasive surgery of lower limbs in osteogenesis imperfecta

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Abstract We present the results of intramedullary rodding of long bones of the lower limbs in children with osteogenesis imperfecta using a modified Sofield-Millar operation. Fourteen patients (mean age at primary operation was 5 years 11 months) were treated with a modified Sofield-Millar operation which allows minimal bone exposure, preservation of the periosteum and keeping the number of osteotomies to the minimum. Union was achieved in all cases within 7 weeks. Of the 14 patients (29 bones) treated with nonelongating rods, rod revisions were needed in 13 patients (26 bones). We found no statistically significant difference between the width of the bone immediately postoperatively and at the final follow-up. The walking ability was improved in four patients. Advantages of less invasive surgery in osteogenesis imperfecta are rapid bone union, no bone atrophy or nonunion, better postoperative mobility and small scars.

Résumé L'objectif de cette étude est d'évaluer les résultats du brochage intra médullaire des os longs des membres inférieurs chez les enfants présentant une ostéogénèse imparfaite en utilisant la technique modifiée de Sofield-Millar. Matériel et méthode : 14 patients (âge moyen à la première intervention 5 ans) ont été traités avec cette méthode de Sofield-Millar modifiée qui permet, à partir d'un abord minimal de préserver le périoste; de réduire au

minimum le nombre d'ostéotomies. Résultats, une consolidation a été obtenue dans tous les cas au bout de sept semaines. 14 patients (29 segments osseux) ont été traités avec un clou ne permettant pas la croissance, la révision du clou a été nécessaire chez 13 patients (26 segments osseux). Nous n'avons pas trouvé de différences significatives sur la qualité osseuse en post-op immédiat et au suivi final. La marche a été améliorée chez 4 patients. En conclusion : cette technique moins invasive est avantageuse dans le traitement de l'ostéogénèse imparfaite et permet une consolidation rapide sans atrophie osseuse et sans pseudarthrose avec une bonne mobilité post-opératoire et des cicatrices minimales.

Introduction

Osteogenesis imperfecta (OI) is a syndrome with congenital brittle bones secondary to mutations in the genes that codify for type I procollagen. Deformities of long bones and multiple fractures are common in patients with OI and can affect their ability to walk [14].

The value of multiple osteotomies and intramedullary rodding in OI is well established since Sofield and Millar [18] reported their technique of bone fragmentation and subsequent fixation with nonextensible rods. Several innovations of the design of the device and/or modifications of the original operative technique have been developed [2, 8, 21].

Li et al. [10] described a modified technique of the Sofield-Millar operation to minimise damage to the periosteum and preserve the blood supply to the bone. Minimally invasive surgery using a Bailey-Dubow rod was developed by Fassier and Duval [6]. Despite these innovations and modifications, intramedullary rodding in

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osteogenesis imperfecta is not without complications and some of these complications necessitate further surgery [3, 7, 8, 15, 23].

This study was undertaken to report our results of intramedullary rodding of long bones of the lower limbs in children with OI using a modified Sofield-Millar operation as described by Li et al. [10]. In particular, we tried to evaluate its effect on bone width, its complications, the frequency of revision surgeries and its effect upon ambulation.

Patients and methods

Between 2001 and 2006, we treated 14 patients (8 boys and 6 girls) with OI at Mansoura University Hospital, Mansoura, Egypt. According to the Sillence et al. classification [17], six patients were type I, four patients were type III and four were type IV.

In our study, a modified Sofield-Millar operation of the lower limbs was used as described by Li et al. [10]. These patients had 17 femur and 12 tibia treated with nonelongating rods. Their mean age at primary operation was 5 years 11 months (range: 3 years 7 months to 8 years 6 months).

According to the size of the medullary canal, Rush nails or Kirschner wires were used. Rods of varying lengths and thickness were available during the operation. Bilateral femoral or tibial procedures were sometimes performed concomitantly.

Operative technique of femoral rodding

The patient was positioned on the operating room table to allow use of a C-arm image intensifier. The site of the femoral fracture or the apex of the femoral deformity was approached through a small incision, about 3 cm in length, either directly or through the anatomical planes of the standard lateral approach (this in contrast to Li et al. [10] who exposed the tip of the greater trochanter and reamed as for closed insertion of an intramedullary nail). The bone was exposed subperiosteally and with the aid of an oscillating saw, a small lateral or anterolateral wedge of bone was removed to correct the angulation. The awl for the Rush rod (or the drill appropriate for the rod to be used) was passed retrogradely into the proximal fragment to exit just medial to the greater trochanter. The distal fragment was prepared by antegrade drilling of the intramedullary canal down to the metaphysis.

A rod of appropriate size was passed through the proximal fragment. Then, the deformity was reduced and the nail advanced distally. Usually a second block then occurred and the same procedure was repeated. Normally, two osteotomies were sufficient to correct the deformity. Occasionally, a third osteotomy was necessary. Ideally, the

end of the rod should be in the middle of the femoral condyles. The periosteum was sutured if possible, the wound closed and a spica cast was applied.

Operative technique of tibial rodding

For the tibia, the entry site was just medial to the patellar tendon. The awl for the Rush rod (or the drill appropriate for the rod to be used) was passed antegradely through the proximal fragment. The progress of the reamer stopped at the first site of angulation of the tibia which was confirmed using the image intensifier. Here, a small incision was made and a small wedge of bone was removed to correct the angulation. From then on, the operation was similar to that for the femur and a long-leg cast was applied.

Postoperative plaster immobilisation was maintained for 6 weeks. If any delay in union of any osteotomy site was seen on the radiographs, a further period of plaster immobilisation was used for an additional 3 to 4 weeks. Ambulation with the aid of crutches or a walker was encouraged once radiographic and clinical union was achieved. Clinical and radiographic assessments were done every 6 months.

Radiological measurements

In order to determine the effect of the operation on bone width, we measured the width of the bone immediately postoperatively and at the final follow-up. It was measured as a ratio to the width of the intramedullary rod (the width of the rod/the width of the bone; a/b ratio, Fig. 1) as described by Li et al. [10].

Statistical analysis

Data were analysed using SPSS (Statistical Package for the Social Sciences) version 10. Numerical data were presented as mean $\pm$ standard deviation (SD). The statistical difference of the width of the bone immediately postoperatively and at the final follow-up was evaluated by the paired *t*-test. A value of $P < 0.05$ was considered to be statistically significant.

Results

Union was achieved in all cases within 7 weeks (range: 6–10 weeks, Fig. 2). Follow-up was continued for almost 5 years (mean $\pm$ standard deviation: 4.5 ± 0.7 years). Rush rods were the type of rods most frequently used (49 Rush rods versus 6 Kirschner wires). Of the 14 patients (29 bones) treated with nonelongating rods, 13 patients needed rod revisions in 26 bones (Table 1).

The femoral revision rate was 88.2% and the tibial revision rate was 91.7%. The mean interval between

Fig. 1 Measurement of the a/b ratio. **a** Width of the rod. **b** Width of the narrow straight portion of the bone at the diaphysis

primary rodding and revision was 2 years (range: 1 year 4 months to 2 years 11 months). The mean interval between primary femoral rodding and revision was 1 years 9 months (range: 1 year 4 months to 2 years 7 months). The mean interval between primary tibial rodding and revision was 2 years 3 months (range: 1 year 6 months to 2 years 11 months).

The causes for revision in our series included 14 rod cortical extrusion (Fig. 3), 7 recurrences of deformity and 3 refractures [1 of them with rod bending (Fig. 4) and two with rod migration]. Infections, neurovascular complications, hyperplastic callus or nonunion did not occur in any patient. The walking ability of our patients preoperatively versus postoperatively is shown in Table 2.

We found no statistically significant difference between the width of the bone immediately postoperatively and at the final follow-up (mean a/b ratio was 0.322 immediately postoperatively versus 0.320 at the final follow-up, $P=0.17$).

Discussion

Until there is a no specific treatment for the genetic defect in OI, the goal of treatment should be directed to maximise the affected patient's function and to prevent deformity and disability resulting from fractures. Correction of any pre-existing deformities of long bones and implantation of

intramedullary rods is one option currently available for achieving this aim [13, 19]. Some disadvantages of nonextensible rods were overcome with the introduction of the Bailey-Dubow extensible intramedullary rod [2]. The problem of T-part disconnection frequently seen with the Bailey-Dubow rod was solved with the Sheffield rod [21]. In a recent study, Joseph et al. [8] preferred to use double Rush rods in the femur.

We preferred to use Rush rods as they are easily inserted and revised, inexpensive, multiple rods of all sizes can be stocked easily and their length can be adjusted by cutting the end of the rod. We followed the technique described by Li et al. [10]. The advantages of this technique are that the

Fig. 2 **a, b** Anteroposterior and lateral radiographs of case 2 showing bowing of the right tibia. **c, d** Radiograph taken at 6 months after placing the Rush rod demonstrating anatomic alignment of the tibia and union of the osteotomy site

Table 1 Details of patients with osteogenesis imperfecta (OI)

Case	Gender	OI type	Age at primary rodding and at revision (years+months)			
			R femur	L femur	R tibia	L tibia
1	M	IV	3+7 4+11	3+7 4+11	-	-
2	F	I	-	-	8	-
3	M	III	5+1 6+10	5+1 6+10	-	-
4	M	I	-	8+2 9+0	-	-
5	F	I	-	-	7+3, 9+5	7+3, 9+5
6	F	IV	-	-	8+6, 11+5	-
7	F	IV	3+10 6+5	6+5	-	-
8	M	I	-	-	4+5 5+11	4+5 5+11
9	M	III	8+1	6+5 8+1	-	-
10	F	III	4+3 6+6	4+3 6+6	5+2 7+1	5+2 7+1
11	M	I	-	6+3 7+11	6+3 7+11	-
12	F	III	7+4 8+10	7+3 8+10	-	-
13	M	IV	-	3+10 5+8	3+10 6+6	-
14	M	I	6+2 8+1	6+2 8+1	7+5 10+3	7+5 10+3

R right, L left, M male, F female

exposure is minimal with less blood loss and small scars, the periosteum is preserved and the number of osteotomies is kept to the minimum allowing the bone to heal rapidly (all of our cases healed within an average of 7 weeks).

Tiley and Albright [20] advised delaying intramedullary rodding (nonelongating and elongating) until the child begins to stand. In their series, intramedullary rodding was used in children from the age of 4 years. However, early surgical intervention may be indicated to stabilise the long bones in order to optimise functional ability and walking capacity [5]. Ryöppy et al. [16] mentioned that early stabilisation of the involved bones was indicated to prevent an arrest in motor development caused by a cycle of fractures, immobilisation, osteoporosis, refractures and diminished motor activity due to fear of refracturing. Satisfactory results were reported by several authors [3, 11, 12, 16] who recommended operative intervention in infancy with “percutaneous” and “semiclosed” techniques for nonelongating rods in the treatment of the severely affected patients. These authors believe that the benefits of straightening and stabilisation outweigh the risks of additional procedures.

In our patients, surgical revisions were performed at an average of 2 years following primary rodding because our sample included patients who had been operated when their mean age was 5 years 11 months and had, therefore, a large potential for residual growth. With bone growth, the rod had become relatively smaller, allowing the metaphyseal bone segment that was not protected and supported to deform, with a tendency to fracture or rod extrusion. In our study sample (29 bones), rod cortical extrusion without rod migration developed in 14 patients, recurrence of deformity developed in 7 patients, refractures developed in 3 patients and 2 patients had rod migration. Review of the literature shows the average lifespan of a nonelongating rod to be 2–2.5 years [2, 3, 6, 7, 9, 11–13, 16, 18, 20, 21] making multiple revisions inevitable in children who have significant long bone growth remaining.

Nonunion in OI is not a rare phenomenon [1, 7]. Agarwal and Joseph [1] encountered nonunion of long bones in 18% of children with OI. Nonunion can develop at the osteotomy sites that are required to correct the

Fig. 3 **a** Intraoperative picture of the right leg of case 6 taken 26 months after a Rush rod was inserted showing skin penetration. **b** Lateral radiographs of the same patient showing rod cortical extrusion and anterior angulation at the distal end of the tibia. **c, d** Final follow-up after reoperation using a modified Sofield-Millar operation

Fig. 4 **a** Left femur fracture in an 8-year-old boy with a history of osteogenesis imperfecta (case 4). **b, c** Three weeks after fixation of the fracture and placing of a Rush rod: early healing is present. **d** Ten months following fixation: bending of the rod and fracture of the upper end the femur has occurred. **e** Another rod had been inserted and final follow-up showing correction of the deformity and union of the multiple osteotomies carried out

deformities of the long bones in OI. Two or more osteotomies on a bone can jeopardise the blood supply of the intercalary segment, especially if, while realigning the fragments, soft tissue attachments and periosteum of the intercalary segment are stripped circumferentially [1]. As our exposure is minimal and the periosteum is preserved and the number of osteotomies is kept to a minimum, nonunion did not occur in any of our cases. Bone atrophy was also not noted in our study.

The improvement of possibilities for ambulation after intramedullary rodding of the lower extremities remains

questionable because no randomised clinical trials have been performed for ethical reasons. Several authors [5, 8, 15, 22] have stated that intramedullary rodding of the legs improved the possibility for ambulation, whereas others [4] found no differences in subsequent ability to walk between those patients who received intramedullary rodding when they were young (at the age of first achieving the major motor milestones) and those who did not. Engelbert et al. [5] mentioned that after intramedullary rodding of the legs was performed, functional ability, especially in the pre-standing milestones, improved in patients with types III and

Table 2 Preoperative and postoperative walking ability of patients with osteogenesis imperfecta (OI)

OI type	No. of patients	Walking ability					
		Ambulators		Assisted ambulators		Confined to wheelchair	
		Preoperative	Postoperative	Preoperative	Postoperative	Preoperative	Postoperative
I	6	4	6	2	0	0	0
III	4	0	0	0	0	4	4
IV	4	1	3	3	1	0	0
Total	14	5	9	5	1	4	4

IV, whereas in patients with type I, walking ability improved.

In our study, the walking ability was not worsened in any patient. On the contrary, the walking ability was improved in four patients (two of them were OI type I and two were OI type IV). It has been reported that in types III and IV, even when ambulation is achieved, walking is frequently lost in the second decade of life because of progressive spinal deformity, decreased motivation in physical therapy and the increasing use of a wheelchair [5]. We do not know whether the children in our study will be able to walk in the same way as they did at the time of the study, especially when they reach adolescence.

Based on our results, the advantages of less invasive surgery of the lower limbs in OI are rapid bone union, no bone atrophy or nonunion, better postoperative mobility and small scars.

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