SURGICAL TREATMENT OF DEFORMITIES AND FRACTURES ON LOWER LIMBS WITH OSTEOGENESIS IMPERFECTA

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ABSTRACT

Objective: To provide a review of patients with Osteogenesis Imperfecta by analyzing the deformities, fractures and results of surgical treatment on lower limbs in order to determine the efficiency of the use of non-elongating rods (non extensible). Materials and Method: Medical records, preoperative and postoperative X-ray images of all the patients who had imperfect osteogenesis and treated at the Alfred I duPont Institute (USA) between 1965 and 1999 have been reviewed. Results: Fourteen patients (five boys and nine girls) were submitted to the non-elongating rods on their lower limbs, totaling 37 procedures. Conclusion: The procedure of intramedullary fixation with non-elongating rods to treat fractures and deformities on lower limb in Osteogenesis Imperfecta was proven to be a low morbidity method without interfering with the ambulatory status of these patients.

Keywords: *Osteogenesis imperfecta. Bone fractures. Lower extremity.*

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INTRODUCTION

Osteogenesis imperfecta, according to Melebranche, in 1684, Ekman, in 1788, Lobstein, in 1834, Vrolick, in 1845, and Porak-Durante, in 1905 apud Correa et al[1] was described as a syndrome caused by a change on the connective tissue involving type-I collagen, which is the organic component of the bone. However, with the recent DNA studies, it was proven that many patients with Osteogenesis Imperfecta did not show changes on the genes codifying collagen production.[2] Today, the concept has been broaden, being defined as a syndrome caused by a genetic change and with variable complexity levels. Diagnosis is pretty much provided by clinical examination and X-ray tests. The classification by Sillence[3], published in 1979, has been employed for grouping these patients.

The orthopaedic treatment, both for fractures and for deformities correction, which consists of corrective osteotomy and stabilization of those bones, using several kinds of implant materials for osteosynthesis has been used by a number of authors.[4-9] Osteosynthesis can be made with fixed or non-extensible nails, which may present complications due to bone growth, such as nail protrusion through cortical, with recurrence of deformity, and extensible nails with different fixation techniques on the epiphyseal region of the long bones, targeting nail stretching as the bone grows, thus avoiding complications and reducing the required time between procedures.

Sofield and Millar[4] described a procedure for treating Osteogenesis Imperfecta, which consisted of multiple osteotomies and bone realignment using an intramedullary nail extending between both metaphyses of a same long bone. This procedure using fixed (non-extensible) nails allows long bones growth to "create" bone segments without nail protection, which occasionally presents fractures or deformities requiring new surgeries. Despite of this complication, this has been accepted as a preventive treatment for fractures and deformities.

The objective of this study is to review patients diagnosed with Osteogenesis Imperfecta treated by using the Sofield technique, assessing surgical treatment of fractures and deformities on lower limbs in order to determine the effectiveness of the technique using fixed (non-extensible) nails.

CASE SERIES AND METHOD

Medical files and X-ray images of all patients with Osteogenesis Imperfecta submitted to multiple osteotomies and bone realignment with fixed (non-extensible) intramedullary nails on lower limbs were reviewed at the Alfred I du Pont Institute, Wilmington, Delaware (USA), between 1965 and 1999. Twenty-seven patients were treated according to this procedure. Of these, 13 were excluded due to a postoperative follow-up inferior to four years.

The 14 remaining patients (five boys and nine girls) were classified according to the criteria described by Sillence.[3] Type IA (7), Type IB (1); Type II (0); Type III (4); Type IVA (0); Type IVB (2).(Table 1)

Type I is the most prevalent form, including the milder bone weakness forms, with few fractures, no significant deformities and with

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Table 1 – *Patients' characteristics for gender and according to the classification by Sillence*

case	gender	Classification
1	M	IVB
2	M	IA
3	F	IB
4	F	IVB
5	F	IA
6	M	III
7	F	IA
8	F	III
9	F	III
10	M	III
11	F	IA
12	F	IA
13	M	IA
14	F	IA

Source: Alfred I duPont Institute – Hospital for Children – year 1999
M – male; F – female

normal height. We had eight patients, seven of which in IA, with normal dentinogenesis, and one patient in IB com dentinogenesis imperfecta. Type II includes the most life-threatening kind, with a significant incidence of death at birth, with serious bone weakness, in which a number of intrauterine and delivery fractures can occur. Death usually occurs during delivery or on the first days after birth. We had no patients included in this group. Type III includes classic cases showing significant bone weakness and deformity; the patients are usually short and show dentinogenesis imperfecta. We had four patients included in this group. Type IV includes patients with bone weakness, normal sclera, and skeletal deformity with short heights, which is subdivided into IVA with normal dentinogenesis and IVB, with dentinogenesis imperfecta. The two patients we had were included in the IVB type group.

On the 14 patients, a total of 37 nails were inserted on lower limbs. The procedures were carried out on 18 femurs (51% - nine right and nine left femurs), and on 19 tibiae (49% - 11 left and eight right tibiae). In this case series, one patient was submitted to tibial and femoral surgery simultaneously (Figure 1 and 2 - case 8, and Table 2).

Figure 1 – *Deformity on the four segments (case 8)*

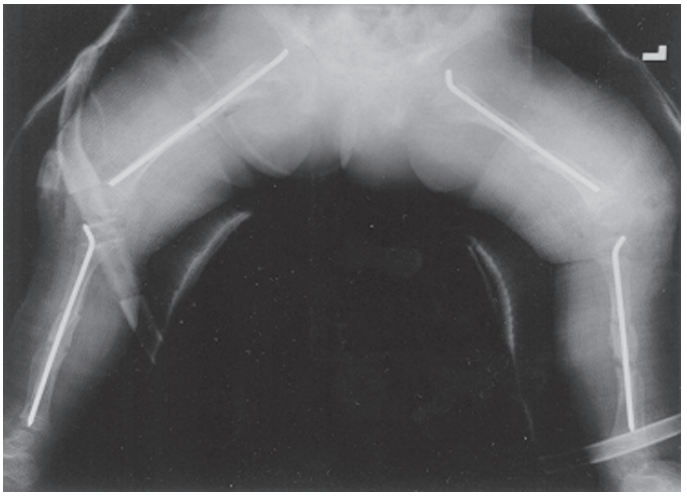

Figure 2 – *Result after the procedure*

Table 2 – *Procedures performed on the lower limbs.*

Case	Type	LT	RT	TOTAL TIBIA	LF	RF	TOTAL FEMUR	Total NAILS
1	IVB	3	0	3	0	0	0	3
2	IA	0	1	1	1	1	2	3
3	IB	0	0	0	0	1	1	1
4	IVB	2	2	4	0	0	0	4
5	IA	0	0	0	1	1	2	2
6	III	0	0	0	1	0	1	1
7	IA	0	0	0	1	0	1	1
8*	III	1	1	2	1	1	2	4
9	III	1	1	2	0	0	0	2
10	III	2	1	3	0	0	0	3
11	IA	1	0	1	1	0	1	2
12	IA	1	1	2	2	4	6	8
13	IA	0	0	0	1	1	2	2
14	IA	0	1	1	0	0	0	1
Total		11	8	19	9	9	18	37

Source: Alfred I duPont Institute – Hospital for Children – year 1999
LT-Left tibia; RT – Right tibia; RF – Right femur; LF – Left femur.

* This patient (case 8) was simultaneously submitted to four fixation procedures.

Thirty-seven fixed nails were employed, of which 27 were Steinmann's wires; one Rush's nail; three Nancy's nails; two Luque's nails, and four Kuntscher's nails.

The preoperative and postoperative gait ability was assessed by dividing the patients into two groups of ambulating (8) and non-ambulating (6).

The surgical procedure was carried out by using the same technique as described by Sofield and Millar[4], which consists of exposing long bones and preserving periosteum, performing multiple bone osteotomies between proximal and distal metaphyses using an electric saw, achieving bone straightening. Prophylactic antibiotic therapy with cephalosporin (50 mg/Kg/day) was routinely used on anesthetic induction and on the first postoperative day. All

patients had their operated limbs immobilized by plastered devices and/or removable immobilizers with a mean union time of 42 days (ranging from 40 to 51 days).
For statistical analysis, the InStat 3.1 software for Windows was employed, with the Mann-Whitney's test with a significance level of 0.05.

RESULTS

The surgical procedures are presented on Table 3. The 14 patients with a total of 37 procedures had six nails reviewed due to deformity complications and three nails were reviewed for fractures.
Indications for the primary surgery included: fracture recurrence (15) and deformity (22). The mean age at the primary procedure for nail insertion was seven years and seven months, ranging from one year and eight months to 13 years and seven months. The mean follow-up time for the first review was three years and one month (ranging from one year and five months to four years and seven months). The mean time for the second review was two years and seven months (ranging from one year and eight months to three years and six months), and only one patient (Figures 3 and 4 - case nr. 12) was submitted to a third review after one years and three months. Four patients (28%), in a total of 9 (24%) nails required review procedures. (Table 4)
The reasons for these reviews were: fractures (3 cases) and recurrent deformities with nails exceeding bone limits (6 cases).
The incidence of nails complications requiring review procedures is represented on Table 5.
The pre- and postoperative results of patients' ambulation status are shown on Table 6.
The eight patients unable to ambulate prior to surgery remained as non-ambulating. The only exception was case nr. 3, in which the patient was one year and eight months old and unable to ambulate at the time of the first procedure, but at the age of four, he started to walk, after being submitted to surgery on the right femur. The six patients who were able to ambulate prior to surgical procedures remained so after surgeries.

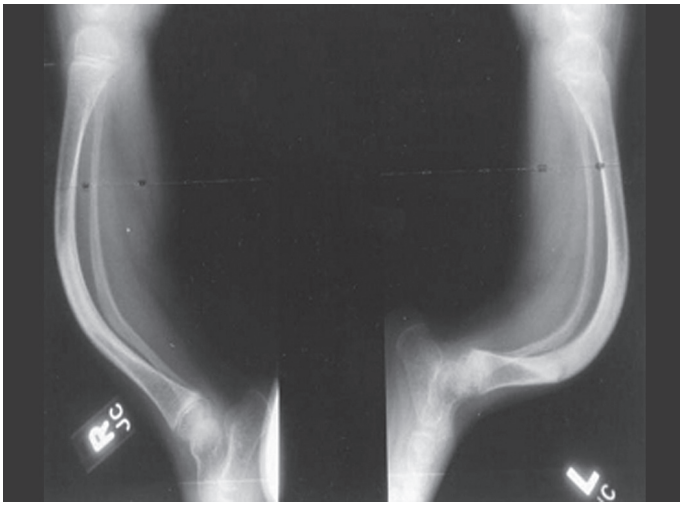

Figure 3 – *Typical deformity of tibial antecurvatum (case 12).*

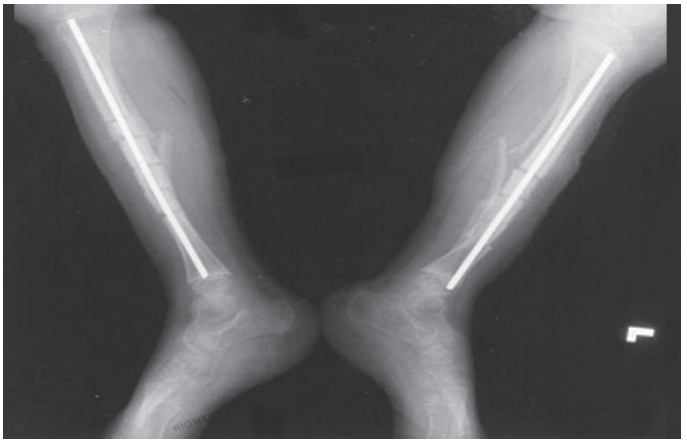

Figure 4 – *Result after review with non-extensible nails.*

Table 3 – *Summary of treatment with non-extensible nails*

Case	Classification	Gender	Age at 1st procedure	Nr. of procedures	Review for deformity	Review for fracture	Preoperative ambulation	Postoperative ambulation
1	IVB	M	2+5	3	1	1	No	No
2	IA	M	7+4	3	0	0	No	No
3	IB	F	1+8	1	0	0	No	Yes
4	IVB	F	9+6	4	2	0	No	No
5	IA	F	9+5	2	0	0	Yes	Yes
6	III	M	5+4	1	0	0	Yes	Yes
7	IA	F	6+11	1	0	0	Yes	Yes
8	III	F	4+6	4	0	0	No	No
9	III	F	7+5	2	0	0	Yes	Yes
10	III	M	4+4	3	1	0	Yes	Yes
11	IA	F	10+7	2	0	0	Yes	Yes
12	IA	F	7+7	8	2	2	No	No
13	IA	M	13+7	2	0	0	No	No
14	IA	F	11+4	1	0	0	No	No
Total				37	6	3		

Source: Alfred I duPont Institute – Hospital for Children - year 1999
M – male; F - female

Table 4 – *Mean age (months and years) and follow-up after surgery*

case	age at 1st surgery	I	F at 2nd surgery	I	F at 3rd surgery	I	F at 4th surgery	I	
Site									
1	2+5	D	4+7	F	3+6	D			LT
2	7+4	D							RF
2	7+8	D							RT
2	7+5	D							LF
3	1+8	F							RF
4	9+6	D	3+5	D					RT
4	9+6	D	3+5	D					LT
5	9+5	F							RF
5	10+4	F							LF
6	5+4	D							LF
7	6+11	D							LF
8	4+6	D							LF
8	4+6	D							RF
8	4+6	D							LT
8	4+6	D							RT
9	7+5	F							LT
9	8+6	F							RT
10	4+4	D	3+1	D					LT
10	7+5	D							RT
11	10+7	D							LF
11	10+7	D							LT
12	7+7	D							LT
12	7+7	D							RT
12	8+8	F	2+3	D	1+8	F	1+3	F	RF
12	9+6	F	1+5	D					LF
13	13+7	F							RF
13	13+7	D							LF

Source: Alfred I duPont Institute – Hospital for Children – year 1999
D - deformity; F – fracture; LT – Left tibia; RT – Right tibia; LF – Left femur; RF – Right femur; I – indication; F – follow-up

Table 5 – *Nail complications requiring review surgeries*

Type	Nr. of nails	Complications requiring review surgery	Percentage (%)
I	20	4	20%
III	10	1	10%
IV	7	4	58%

Source: Alfred I duPont Institute – Hospital for Children – year 1999

Concerning osteotomy complications with the use of non-extensible nails: none of the patients was submitted to blood transfusion. Osteotomies showed an union rate of 100%. In the present study, the procedure has been shown not to interfere with physis, because we had no patient with physeal changes. We didn't find growth disorders inherent to the adopted procedure. No postoperative infection case was found. A total of nine cases with nails showed complications, i.e., 24% of the limbs had to be submitted to a new surgery. The complications found included deformity (67%) and fracture (33%).

Table 6 – *Patients' characteristics for gait*

case	gender	Classification	Preoperative ambulation	Postoperative ambulation
1	M	IVB	No	No
2	M	IA	No	No
3	F	IB	No	Yes
4	F	IVB	No	No
5	F	IA	Yes	Yes
6	M	III	Yes	Yes
7	F	IA	Yes	Yes
8	F	III	No	No
9	F	III	Yes	Yes
10	M	III	Yes	Yes
11	F	IA	Yes	Yes
12	F	IA	No	No
13	M	IA	No	No
14	F	IA	No	No

Source: Alfred I duPont Institute – Hospital for Children – year 1999
M - male; F - female
Note: Case nr. 3 - achieved ambulating status after surgery.

DISCUSSION

Our patients were classified according to the Classification by Sillence[3], which, in spite of not presenting clearness between groups, was the way we found to differentiate and evaluate our patients. Our series encompassed types I, III and IV of the classification by Sillence.[3] Type I had the highest number of patients (57%), Type III (29%), Type IV (14%) And Type II, for being the most life-threatening one showing the highest incidence of death at birth, included no patient. These data are consistent to the study by Ryoppy et al.[10] Concerning the complications requiring reoperation, Type IV showed a higher prevalence (58%), Type III (10%) and Type I (20%).

The procedure of conducting multiple osteotomies fixated with intramedullary nails in order to fix deformities and prevent recurrent fractures on patients with Osteogenesis Imperfecta has been accepted since 1959, when Sofield and Millar[4] described this procedure. It has been modified by a number of surgeons[6,9,11-13], but its principle, which consists of multiple osteotomies, realignment and fixation with intramedullary nail on long bones, remains the same.

This procedure improves the quality of life for these patients, although complications are commonly seen, such as bone segments deprived of nail protection due to bone growth, enabling the occurrence of fractures or recurrent deformities, nail migration, pseudoarthrosis and union delay.[4,12] Indeed, with a continuous growth, bone segments exceeding the nail can be deprived of protection, favoring deformities and nail protrusion through the cortical. The bone can either fracture or deform at this level, requiring nail replacement by a longer one and a new realignment of the limb. In our study, nine nails required review, six as a result of deformities and three as a result of fractures. The mean time for nails review was two years and eight months. The result we found was consistent to other authors'[4,12,13] who used non-extensible nails with a mean time for review of two years.

Several authors state that the time for reviews can be extended with the use of extensible nails.[9,12,14,15] However, these extensible nails lead to complications requiring reoperations. By comparing the present study to these authors', we can see that non-extensible nails do present a reduced mean time to review compared to extensible nails, but the mean complication rate requiring reoperation, which, in our study, was 24%, shows a similar value.

Although the authors[9,10,11,14,16] indicate extensible nails, they recommend non-extensible nails for children, when tibial and femoral spinal canal is too narrow for extensible nails to pass through.

The procedure is associated to a low morbidity rate. Osteotomies showed union in all patients, no blood transfusion was required, and no infection or anesthetic complications cases were found.

Regarding the best time for surgery, literature does not indicate a consistent concept about the best age to fix deformities or the best time to prevent recurrent fractures. Ryoppy et al.[10] recommend interventions in children using non-extensible nails, emphasizing that the early realignment and stabilization of the lower limbs improves motor development, with no minimum age for surgery. Williams et al.[13] concluded that the best time to start inserting nails is when deformities are detected. Tiley and Albright[8] reported that the optimal age for starting lower limbs correction is when the patient keeps orthostatism. This study did not determine the best time for surgical procedure. The question is: what is best for the patient: to provide an early treatment before a deformity or fracture occurs, or to wait and treat the complications?

We believe that a surgical treatment using non-extensible nails is indicated when complications such as fractures and/ or deformities are present, based on the fact that these nails do not follow bone growth, if early implanted, complications will eventually occur, resulting in a larger number of review surgeries.

In our study assessing pre- and postoperative gait, we divided the patients as ambulating and non-ambulating. All six patients able to remain at a standing position ad walk prior to the first surgery remained with this ambulation status. The remaining eight patients who were unable to walk prior to surgery remained as so after the procedure, except for one patient (case nr. 3) who was one year and eight months old when the primary surgery was performed, subsequently becoming a walker. We believe that such ability was due to the fact that the procedure was early performed, and that ambulation status was acquired with the patient's own development. The authors[7,9,12,17-19] also concluded that the procedure allows patients to keep their ambulatory ability.

By comparing the time for non-extensible nails review in the present study to the results of the use of extensible nails reported on literature[9,12,14,15] using the Mann-Whitney's test, with literature values reporting 3.7 ± 0.6 compared to 2.8 ±1.1 in the present study, evidencing no significant difference, although the longevity of the non-extensible nails is shorter.

In summary, the surgical treatment of lower limbs deformities and fractures on Osteogenesis Imperfecta shows the following advantages: gait status is kept, does not present physeal injury sequel, brief hospitalization time (mean of four days), and deformity fixation with improved limb function. One disadvantage would be bone growth enabling the emergence of areas without protection of the internal fixation, which might cause recurrence of deformities, fractures and wire migration. These complications usually require review surgeries.

CONCLUSION

The procedure of intramedullary fixation of non-extensible nails for treating lower limbs fractures and deformities on Osteogenesis Imperfecta was shown to be a low-morbidity method, able to maintain or even improve the ambulatory status of these patients.

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