Acta Orthop. Belg., 2019, **85**, 1-11

ORIGINAL STUDY

Use of sliding transphyseal flexible intramedullary nailing in pediatric osteogenesis imperfecta patients

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In our country, the sliding Flexible Intramedullary Nailing is used alone or in combination with Ilizarov frame in children with osteogenesis imperfecta. The study assesses the results of sliding intramedullary nailing in deformity correction in severe types of osteogenesis imperfecta.

We retrospectively reviewed 17 consecutive cases (mean age 5.2 y.o.) of types III, IV and VII of osteogenesis imperfecta. In group I (9 patients) the transphyseal FIN was performed using titanium nails. Sliding flexible intramedullary nailing was associated with Ilizarov frame in group II in 8 children. Patients in group I had overall complication rate of 88.9%: proximal nail migration (3), early secondary torsional displacement (4), non-telescoping (12), angular deformity (2), delayed or non-union (2). The reoperation rate was 100%. In group II we observed complications in 6 patients: nail migration (2), bowing of femur (2), non-telescoping (3). The reoperation rate was 87.5%. Flexible intramedullary nailing allows realignment and good functional outcomes. Its major disadvantage is an important complication rate and related reoperation rate. The use of Ilizarov frame provides additional stability and allows early weight-bearing.

Keywords : osteogenesis imperfecta ; flexible intramedullary nailing ; osteoinduction.

INTRODUCTION

Osteogenesis imperfecta (OI) is a group of genetic disorders with wide phenotypic and molecular heterogeneity. The major orthopaedic features are bone fragility, osteopenia, progressive bone deformity and varying degree of short stature *(8,15)*. It is a rare genetic disease with an incidence of 1 in 10.000 to 1 in 20.000 births *(8,15,29,38)*.

The classification of Sillence is the most accepted and based on modes of inheritance, radiological and clinical findings and includes OI types I (mild non-deforming), II (perinatal lethal), III (severe), IV (moderate-to-severe) *(34)*. The addition of types V, VI, VII became necessary to distinguish individuals who present clinical diagnosis of OI but are negative for collagen type I mutations *(29)*. The Sillence classification with additional type V (calcification of the intraosseous membrane and hypertrophic callus) is used as the prototypic and universal way to classify the degree of severity in OI being phenotypically rather than molecularly based *(6,8)*. Genetic classification of OI is not currently used in clinical practice, and the phenotype classification is still the most widely accepted *(8,35,38)*.

The multidisciplinary approach including medical treatment with bisphosphonates, orthopaedic treatment and rehabilitation for muscular

The author reports no conflict of interest concerning the materials or methods used in this study or the findings specified in this paper.

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strengthening and walking strategy has as a goal amelioration of mobility, self-care, functional independence and better quality of life *(3,24,)*. The goal of orthopaedic surgery implies the correction of long bone bowing, rotational malalignment, angular deformity and prevention or reduction of the fracture incidence *(12,30,31,36,40)*.

The telescopic rodding and nailing have been developed in order to obtain a long lasting osteosynthesis in a growing long bone, thus, reducing the need of replacement *(9,18,22,39)*. The single entry telescopic rod system – the Fassier Duval Telescopic IM System allows to decrease the complication rate and reoperation rate, and to reduce the blood loss *(13,19,40)* in comparison to other telescoping systems. The Fassier-Duval rod has advantage of avoiding knee and ankle arthrotomy in femur and tibia surgery *(31)*. Unfortunately, this outstanding telescopic system isn't in widespread use in pediatric orthopaedic centres around the world. For example, the Fassier-Duval rod has not been approved by authorities of our country yet. In such situation, the sliding transphyseal Flexible Intramedullary Nailing (FIN) has to be used *(13,19,21)*.

But the major and still unsolved drawback of any telescopic rod/nail design is the lack of rotational stability *(5,36,37)*. And when this problem is associated with insufficient longitudinal bony stability (considering the severity of underlying conditions) and/or diminished healing capacity, the result might be unfavorable. Furthermore, all telescopic systems don't allow an immediate weight-bearing. Only in case of radiological evidence of callus formation an active physiotherapy and weight bearing are started, at least, 3 weeks after intramedullary rod placement *(5,25,31)*. But minimization of the period of immobilization should be obligatory in any surgical treatment. It prevents secondary bone mass reduction, disuse osteoporosis and further fractures *(18,35,36,37)*. However, the combination of the Fassier-Duval rod or FIN with an external fixator in patients with severe underlying bone pathology proved its advantages from the point of view of stability and early weight-bearing and rehabilitation *(5,20,28)*.

The aim of this retrospective observational study was to assess the results of sliding FIN in deformity correction in children with severe and moderate-to-severe types of OI and evaluate whether the use of FIN in combination with Ilizarov frame had difference to the postoperative period.

MATERIAL AND METHODS

We retrospectively reviewed the patients records and radiographs of our 17 consecutive cases with a diagnosis of OI who had undergone surgery with telescopic FIN between May 2012 and June 2015. The mean follow-up was 1.9 years (range, 1.0-3.2 years). Approval from the Local Human Ethics Committee was obtained to conduct this retrospective study.

The age of children ranged between 1 year and 8 months to 15 years 9 months (mean: 5 years and 4 months) at the time of initial flexible intramedullary nailing. Six patients had type III, 11 type IV and one type VII. We had no patients with type V, VI. All patients had experienced multiple long bone fractures, bone deformity over 20° or torsional deformity causing functional impairment. Before admission in our clinic three patients underwent attempts of deformity correction with locked plates or nailing but without positive results.

The patients with type I were excluded from the study because there was no indication for FIN.

Only in seven patients the deformity correction was performed after the beginning of pamidronate therapy. In all cases bisphosphonate therapy was arrested, at least, 3 months before surgery. In post-operative period pamidronate therapy was initiated 4 to 6 months later in all children.

Depending on the applied technique, the patients were divided in two groups. In group I (9 patients, 34 segments) the conventional transphyseal sliding bipolar flexible intramedullary nailing was done using titanium nails of 1.5 to 3 mm diameter in 28 segments or hydroxyapatite (HA)-coated titanium nails (6 segments) aiming to avoid nail migration in long term follow-up (fig 1 and 2). The precurved nails were inserted through epiphyseal or apophyseal areas (depending on the affected bone), one proximally and another distally. Moderate

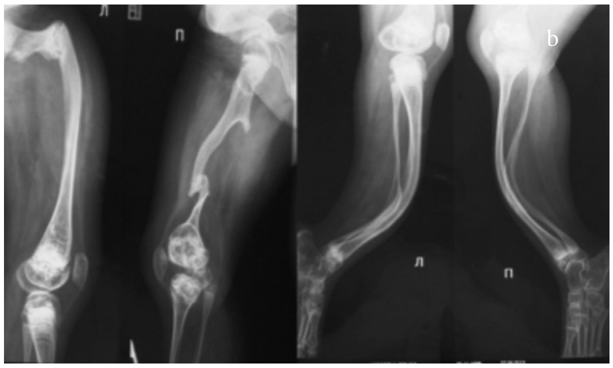


Fig. 1. — Girl 13 y.o. with OI type III: A – images of the patient before treatment; B – radiographs of lower limbs before treatment; C – HA-coated FIN performed, patient standing with walker; D – image of patient and radiographs of lower limbs in 2 years after surgery

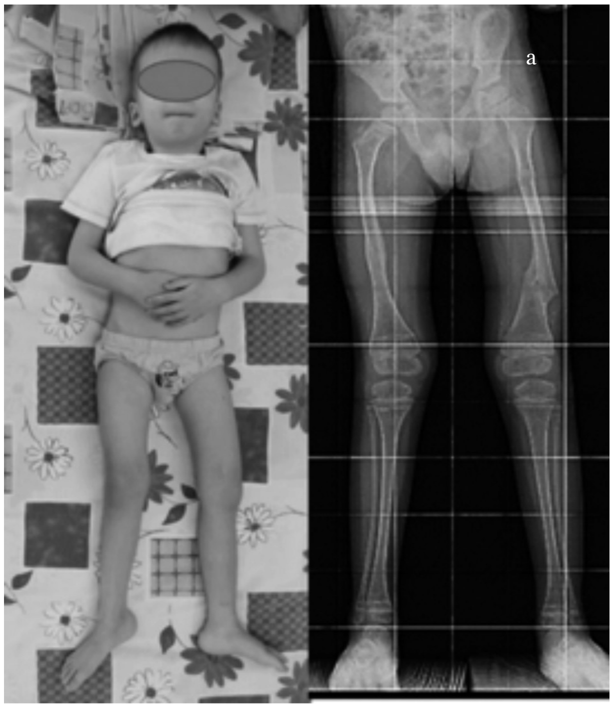
contouring of each nail is used to make two opposite curves into medullary canal. The nails were advanced through the medullary canal to the opposite metaphysis without prior reaming in cases of preventive nailing. In deformity correction, the antegrade and retrograde nails were introduced until bowing level, then, the necessary osteotomies have been done (percutaneously or in open technique using oscillating power saw) and nails were advanced to the opposite metaphysis under

Fig. 2. — Girl 1 y.10 m. o. with OI type IV: A – before treatment (radiographs and clinical aspect), child was not either walking or standing due to frequent fractures; B – radiographs and clinical aspect 3 years later after the initial surgery, good alignment of lower limbs and correct orientation of feet.

visual and X-ray control. If necessary, especially at the bowing level, where the obliteration of medullary canal was often observed, it was rearmed antegradely and/or retrogradely through osteotomy to the appropriate size. The curvatures of nails were reoriented in opposite direction to the eventual residual angular deformity.

In situation of a very small external diameter of some part of diaphysis (diameter inferior to 3 mm), only one nail was inserted throughout the bone. The second nail was initially introduced through epiphysis but placed between periosteum and bone at the level of small diameter part of diaphysis. Then its leading end was again inserted into the bone under visual control, usually, in opposite metaphysis.

Fibula osteoclasis was performed by applying direct force to the bone before completion of the tibial osteotomy. Care was taken to limit surgical approaches and bone exposure to minimum to ensure the best possible biological environment for bone consolidation. At the final phase of the



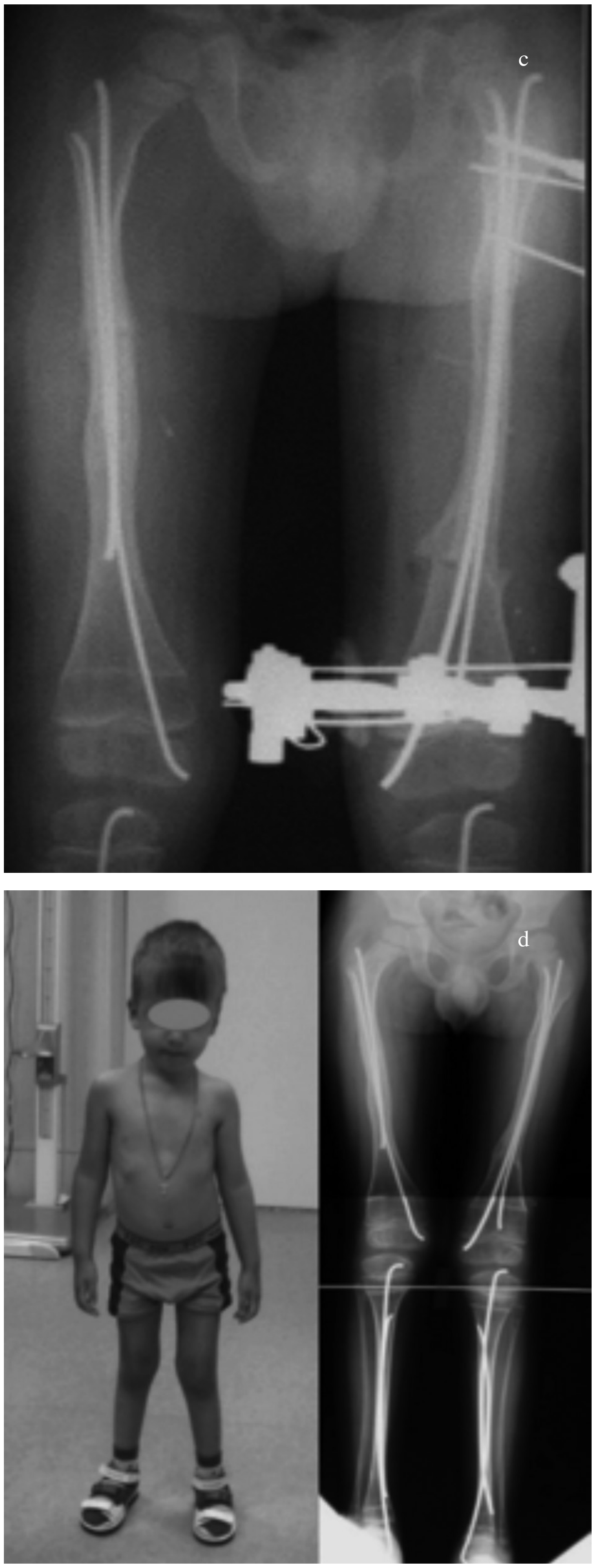


Fig. 3. — Boy, 4 y.9 m.o. with OI type IV: A – clinical aspect (external torsion of the right leg, limb length discrepancy) and radiographs before treatment; B – initial surgery on the right leg: detorsion osteotomy of femur, sliding FIN of femur and tibia, Ilizarov frame at femur, patient stays with total weight-bearing at 7th day after surgery; C – the next intervention has been done 3 weeks later the initial surgery: correction-shortening osteotomy of the left femur, FIN of left femur and tibia, frame removal at right femur; D –1 year after treatment: good realignment of lower limbs, telescoping of nails, patient walks with total weight-bearing.

procedure, the trailing ends of nails were bent and anchored in the epiphysis. External immobilization was applied for 6 weeks with partial weight-bearing started 3-5 weeks after intramedullary nail placement. Usually the surgery for the contralateral leg was done in 5 to 8 weeks.

The sliding FIN was associated with Ilizarov frame in group II (fig.3) in 8 patients (28 segments) aiming to achieve rotational and longitudinal stability as well as to provide early weight-bearing. Briefly, the steps of the procedure were as follows: insertion of the first nail until deformity level, osteotomy, nail advancing and realignment (this step should be done depending on the number of planned osteotomy levels), insertion and advancing of the second nail, Ilizarov frame application, final orientation of nail curvatures and slight final impaction in metaphysis bone, application of moderate compression forces done in Ilizarov frame, trailing ends of nails bent and cut.

In comparison to technique in group I, the surgical approaches for nail insertion and osteotomies didn't change except the final slight impaction of the nails. It was performed only if the definitive position of bone fragments was achieved and Ilizarov frame was applied in order to preserve even minimal stability of nail tip in weak bone tissue. The feature of frame assembly was the use only two rings in tibia or short arc and ring in femur placed at metaphysis levels and perpendicular to the anatomic axe of femur or tibia. We never performed acute or progressive correction of deformities or bone lengthening by means of external fixator. The configuration of external fixator was definitive by the end of surgical procedure. As elements of fixation, half-pins of 3.5-4.5 mm diameter and wires and half-wires of 1.5-1.8 mm diameter were used. Usually, three elements were placed per ring or arc. In 3 patients we used cast (tibia+foot) instead of Ilizarov frame with in-cast-intergrated half-ring only at tibial level attached by threated rods to the the Ilizarov frame at femur (fig.4). In group II external fixation lasted 3-8 weeks but progressive weight bearing started from the 3rd week and became total by the 10th-14th day of postoperative period. Legs were operated one by one (in two sessions) within 3 to 4 weeks' period. In group II intramedullary titanium nails were applied in 22 segments and HA-coated nails - in 6 segments.

All patients were advised to maintain an adequate calcium intake. After cast and frame removal they underwent physiotherapy and kinesiotherapy, including exercises and design of special devices for transportation, walking and sitting.

During post-operative period the patients were reviewed every 6 months by one of authors of this study for clinical and radiological examinations. Anteroposterior radiographs of the lower extremities with the patella centered forward, and lateral radiographs of each segment with adjacent joints were taken. Torsional deformations were assessed by clinical and radiological examination in all patients.

Criteria of assessment were as follows: consolidation rate, number of intraoperative and postoperative complications (including nail migration, limited telescoping, joint intrusion, secondary deformities, deformity recurrence) and their outcomes, reoperation rate, functional outcome.

Assessment of ambulation skills were performed using the Gillette Functional Assessment Questionnaire Ambulation Scale (FAQ) *(26)*. Postoperative changes from baseline to 6 months, 1 year, 18 months and 2 years postoperatively were noted depending on the duration of follow-up period for each individual.

The statistical values described the mean and standard deviation. The FAQ score in both groups was compared by using the Student t-test and the Wilcoxon rank-sum test for independent samples. All tests were two-tailed with a 0.05 level of significance. StatPlus software was used for the statistical analyses.

RESULTS

In group I, the 17 sliding flexible intramedullary naiing procedures (34 segments) with informative follow-up more than 1 year (2 years and 1 months, in average) were done in 9 children (mean age 5 years 7 months). The nails were simultaneously inserted in femur and tibia of one side. The contralateral leg was operated on average in 46.4÷6.8 days later, only if the bone callus formation was observed at osteotomy levels of the first leg. There were no cases of bilateral femur and tibia procedures.

The patients of group I had an overall 88.9% complication rate (fig.5): 8 out of 9 patients developed complications. Proximal nail migration irritating soft tissues of proximal femur or knee joint did occur in 3 patients (5 segments – 3 femurs and 2 tibias). We observed early (during the first month) secondary torsional deformities of segments (in 5 femurs and 4 tibias) in 4 cases. Later, such vicious positions caused functional impairment of gait and compromised comfortable sitting position, thus, parents payed attention to it. Correction of torsional deformity was done in 1 patient simultaneously with realignment procedure for angular deformity over than 20° developed n 9-12 months after the initial surgery. In another patient the problem of torsional deformity due to retroversion of femoral necks was simultaneously fixed during surgical correction of coxa vara (fig.2). Two more patients had only angular deformity of femur due to non-telescoping on nails. In these cases, the nails were changed. There was one case of non-union of proximal femur with angulation more than 25° that required resection of pseudarthrosis zone and change nails. In one case of delayed union the realignment remained good with satisfactory function, that is why, reoperation was not indicated. Two patients had fractures with nails in situ but without secondary deformity requiring intervention. In three patients younger than 10 y.o. the non-telescoping of nails without secondary angular deformity was observed in 8 segments (4 segments had nails with hydroxyapatite coating). The risk of fractures or deformity at the level of bone without internal reinforcement was evaluated high and additional transphyseal nails were inserted. In 4 cases a revisional surgery was scheduled; and related to patient's natural growth. There were no cases of infection in group I.

In total in group I the patients underwent 3.6 surgeries, in average. The reoperation rate for the period of observation was 100% (1.7 operations per case, in average). Unscheduled interventions were performed in 8 patients (in 88.9% cases or 1.22 unscheduled surgeries per patient).

The pre-operative mean FAQ score in group I was 2.4 (range, 1-3) demonstrating that the patients were able to make some steps only with permanent assistance or only during of rehabilitation sessions. At 1 year post-operatively, the mean FAQ score raised to 5.1. Six from nine patients started to walk independently with a walker and/or orthotic devices at home. But two patients did not advance their walking capacity. The mean FAQ score didn't change at 1 year 6 months and 2 years after reconstructive surgery in comparison to 1-year-point of follow-up.

In group II 15 sliding FIN procedures (28 segments) with the mean follow-up of 1 year 8 months were performed in 8 children (the mean age of 5 years 1 month at the moment of nailing). After the first surgery on one leg the contralateral limb was operated on in 25.7÷8.1 days in average. Simultaneously, the frame of the first operated leg was removed under the same general anesthesia.

The total number of complications in group II was significantly lower. We observed 9 complications in 6 patients, that is in 75% of cases. But only 6 cases required reintervention. Migration of external proximal and distal nail irritating soft tissues occured in 2 patients (2 segments – 1 femur and 1 tibia) when the surgical revision (reinsertion or cut of the displaced nail) became necessary. One patient developed bilateral non-telescoping in femurs and bowing of both segments at the level of middle and distal metaphysis – the zone without sufficient reinforcement. This patient needed simultaneous correction of deformity and replacement of intramedullary nails. In three children younger than 10 y.o. non-telescoping of nails caused their migration into the medullary canal in 4 segments (2 segments had nails with hydroxyapatite coating) but there was no secondary angular deformity. In these cases, additional transphyseal nails were inserted to prevent a secondary fracture or deformity at the level of new formed bone. There were two fractures with FIN in situ so patients didn't need surgical management (fig.5). We didn't observe any case of infection in group II. And there were no cases of external fixator-related complications. A probable explanation is a short duration of external fixation without any fixator-related manipulation, like distraction or progressive correction. Surgeries related to the growth of patients were performed in 2 cases. There were no cases of delayed union or non-union in this group.

Thus, in total in group II the patients underwent 23 surgeries (2.88 interventions, in average). The reoperation rate for the period of observation was 87.5% (1.0 reoperation per case). Unscheduled interventions were performed in 6 patients (in 75% cases or 0.75 unscheduled surgeries per patient).

The pre-operative mean FAQ score in group II was 2.2 (range, 1-3) demonstrating that the patients needed permanent assistance to make several steps. In 1 year post-operatively, the mean FAQ score became 5.3. There was not significant difference between the groups for the 1-year-point of follow-up. But in 6 months after surgery in group II the mean FAQ score was significantly higher than in group I of conventional FIN (3.1 in group I and 4,25 in group II). At the latest control seven from eight patients were starting to walk independently at home and using the walk as the main method to move. Orthotic devices remained obligatory for all patients. Only one child did not advance in his ability to walk. As well as in group I, the mean FAQ score continued stable in 1 year 6 months and 2 years after reconstructive surgery in group II.

Regarding the problem of non-telescoping nails with HA-coating, it was observed in all segments reinforced with HA-nails in patients younger than 12 years. In older children, a limited residual growth of long bones explains that non-telescoping was not observed. On the other hand, there were no cases of external migration of HA-nails.

DISCUSSION

A better quality of life including improvement of mobility, self-care, functional skills and functional independence are the main goals of treatment for children with OI *(8,10,11,23,24,31)*. These therapeutic plans are based on long-term multidisciplinary approach, which includes medical treatment with biphosphonates (bone-remodelling drug therapy), orthopaedic treatment (conservative and surgical) for fractures and deformity correction and stabilization, rehabilitation (muscular strengthening and amelioration of range of motion) *(10,11,16,17,23,27,30)*.

The benefits of telescoping intramedullary rodding in fractures and deformity of long bones in children with OI are well known. These systems allow multilevel realignment, possibility to achieve walking with weight-bearing and, to prevent or decrease the rate of fractures and to ameliorate self-care and mobility *(22,36,39,40)*. The telescopic character of modern implants permits to overcome inconvenience of regular rods related to the bone overgrowing *(5,9,14,31,33)*.

The Fassier-Duval telescopic intramedullary rod is acknowledged and recognized intramedullary telescoping device *(2,4,13,19)*. The advantage of Fassier-Duval rod over the Bailey-Dubow or Sheffield rods or sliding FIN is the only one proximal entry point excluding knee arthrotomy in femoral surgery as well as ankle arthrotomy for tibial rodding. Implantation of Fassier-Duval rod is associated with fewer surgical scars, reduced blood loss, decreased time of operation *(2,5,13,31,19)*. Thus, it allows to perform intervention on multiple bones during the same procedures.

The major and unsolved inconvenience of any telescopic system design is the lack of rotational stability *(5,36,37)*. In children with OI this problem can be associated with insufficient longitudinal bone stability (in case of shortening of a segment at surgery or due to weakness of bone tissue) *(7,37)* and diminished healing capacity *(25)*. Furthermore, all telescoping systems don't allow an immediate weight-bearing. Only in case of radiological evidence of callus formation an active physiotherapy and weight bearing with the aid of orthoses can be started *(5,25,31)*. But the period of immobilization must be reduced in patients with OI in order to prevent secondary bone mass reduction and disuse osteoporosis and to allow early rehabilitation *(18,35)*. In young preschool children with severe forms of OI the long bones are narrow, and not suitable for telescoping rods *(12,35)*. In such a situation, regular or elastic nailing is able to provide good middle-term results from the point of view of realignment and prevention of refracture *(1,7,36,37)*. Finally, the sliding FIN becomes the only method to perform if no telescopic rod is approved by authorities.

The reported reoperation rate in severe and moderate-to-severe forms of OI with Fassier-Duval rod varies from 13% to 53% depending on the average follow-up and operated segment *(4,5,13,19,31)*. Revision was necessary in 9% in 2 years' follow-up and 28% in 3 years in use of Dyna-Locking Telescopic Rod *(9)*. The use of sliding FIN in children with OI made the reoperation rate of 75% for 8 years' follow-up *(7)*. But in the mentioned study there were 6 patients with type I of OI and 8 patients with type III.

Complications of all types of rods are not rare. Non-telescoping, joint intrusion, migration of the parts of rod, fractures, delayed or nonunion, epiphysiodesis are among them: 35%-40%-complication rate with the Fassier-Duval rod was reported *(5,31)*. There were 55% of complications for the Bailey-Dubow rod *(13)*. Boutaud reports 25% rate of complication in use of elastic nailing but in series with 42.9% patients with Type I of OI *(7)*.

Infections are not common in OI *(1,4,5,7,31)*. Mechanical complications remain the main problem in all published series *(5,14,7,31)*. The rate of nonunion and delayed union varies from 0% to 14.5% *(4,5)*. Boutard et Laville report one case of pseudarthrosis (femur) for a series of 14 patients *(7)*. Munns et al. observed delayed bone healing after 103 of the 200 interventions *(25)*. And this complication was more frequent in patients receiving pamidronate, in children with OI type IV and in osteotomy of the tibia.

As for results and outcomes of our series, it is difficult to compare it with thoes of telescopic rods. The complication rate and reoperation rate in group I are evidently higher that in series based on experience with Fassier-Duval rod. But the number of surgeries per case is close to the results of series with sliding FIN: 2.5 operations per case with 1.5 reintervention per patient in series of Boutaud et al. *(7)* and 3.6 surgeries and 1.7 reinterventions per case in our study. In our group I, the complication rate is higher because the actual study included only severe forms of OI diagnosed as types III and IV.

In group II the combined technique uniting sliding FIN and external fixation showed several advantages in comparison to conventional sliding FIN in group I. We did not observe any secondary torsional deformity in post-operative period. The early weight-bearing was possible since first postoperative days. This feature presents an advantage in comparison to our group I and advanced telescopic rods as well. Increased stability of bone fragments in immediate post-operative period and early full weight-bearing allowed to avoid delayed and non-union at all in group II. The total of complications was reduced in children treated by combined technique. But the average intervention rate (2.88) and average reoperation rate (1.0) stay higher than in published series with Fassier-Duval rod.

The idea to use external frame as additional stabilization in treatment of orthopedic problems in metabolic bone disorders is not new *(5,20,28,32)*. Birke et al. supposed the use of Ilizarov frame to be beneficial as additional stabilization in Fassier-Duval rodding in patients with severe underlying bone pathology with insufficient torsional or longitudinal stability *(5)*. Authors performed that combination in children with hypophosphatemic rickets and achieved good results, but they had no experience with the combined technique in OI. Kong et Sabharwal used FIN and monolateral external fixation in treatment of femoral shaft fractures in children with OI *(20)*. The external fixator provided angular and torsional stability at the fracture site and permitted to avoid inconvenient supplemental casting. Our own experience with Ilizarov frame and HA-coated FIN in surgical treatment of children with hypophosphatemic rickets was encouraging *(28)*.

Regarding HA-coated FIN, the use of bioactive implants is justified in metabolic bone disorders because it provides more stable elastic osteosynthesis and prevents external migration of nails *(28)*. In this study the HA-coated nails remained in situ in both groups. But those implants caused non-telescoping of FIN in children under 10 years old. Thus, we suggest the use of HA-coated FIN in OI only in non-telescoping system or in children at the end of active growing, i.e., at age over 12 y.o.

Functional outcomes in the groups assessed with the Gillette Functional Assessment Questionnaire Ambulation Scale in our series were comparable in 1 and 2 years of follow-up. Changes from baseline to 1 year post-operatively on the FAQ were statistically significant. The difference between the groups was observed in 6 months postoperatively when the patients of group II showed significantly better performance in mobility and ambulation. Furthermore, at 1-year and 2-years point time functional outcomes were close to results achieved with Fassier-Duval rod *(31)*. On the other hand, we recognized that similar functional results were achieved at sake of greater number of complications and higher reoperation rate.

The mean follow-up in our study is relatively short, only 2 years. It's one of limitations of the study. But the patients continue to be followed at our institution every 6 months for clinical and radiological examinations and we hope to be able to evaluate outcomes in long-term follow-up exceeding 3-4 years. Ruck et al. *(31)* reported that obtained gains of rodding persist up to 4 years but the score of FAQ showed slight decrease at the latest time point.

CONCLUSION

Transphyseal sliding flexible intramedullary nailing allows to achieve realignment of limb segments and outcomes similar to results obtained with the Fassier-Duval telescopic rod. But an important disadvantage of FIN is much higher complication rate and related to it reoperation rate. If both devices are available, the single entry point intramedullary rodding should be preferred.

Use of Ilizarov frame with FIN provides additional rotational and angular stability in early post-operative period that prevents secondary displacement of bone fragments and allows the early weight-bearing. Furthermore, additional short stabilization with external fixator is associated with faster improvement of functional ability in short-term follow-up.

HA-coated nails should not be applied in sliding intramedullary nailing in order to avoid their blocking and non-telescoping due osteoinductive features. However, bioactive coating provides stabilization of nails and prevents their migration. We advise to use nails with HA-coating in osteogenesis imperfecta patients older than 12 years old.

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