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Assessment of telescoping nail in the management of long bones fractures and deformities in patients with osteogenesis imperfecta

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***Abstract*---**Background: Osteogenesis imperfecta (OI) is a genetic disorder characterized by increased bone fragility and multiple fractures. Approximately 90% are caused by mutation in COL1A1 or COL1A2 genes, the disease is transmitted either autosomal dominantly or recessively or appears as a spontaneous mutation. Protein product of these two genes is type I collagen, the major structural protein in the bone and skin, which is affected in quality, quantity or both. The most distinct feature of this disease is repeated fractures and deformities which are very challenging to treat. Making the bone harder by bisphosphonate treatment and by Intramedullary splinting of long bones is the standard of care in most pediatric orthopedic centers. The problem of treating young child with a fixed length intramedullary rod, is overgrowth of the bone beyond the edge

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of the nail, creating a segment of bone which is unsupported and very liable to fractures and deformities. The need for a device that elongates with bone growth (telescoping intramedullary nail) was necessary to overcome this problem. There were different types of telescoping Intramedullary nails (baily-dubow, Sheffield, interlocking telescoping nail that has been used for the treatment of Osteogenesis imperfecta. The last version of telescoping intramedullary rod to appear is the fassier-duval type telescoping intramedullary nail that has been used in this current study. This Current study indicates that Fassier-duval type telescoping IM system resulted in improvement of BAMF in 60% of patients at 2 year follow up, with good telescoping of nail(in 60% of patients) and has the advantage of eliminating the need for knee and ankle joints arthrotomies (that was needed for insertion of previously used telescopic nails). N.B; The Fassier-duval type telescoping nail which have been studied in this study is the one of Carefix company of shanghahay in China.

***Keywords*---**Osteogenesis imperfect, Telescoping nail, Brief assessment of motor function score.

Introduction

Osteogenesis imperfecta is a genetically determined disease that implies a bone fragility with a great variability of symptoms between patients. With characteristic early onset of fractures after a low energy trauma. The two main genes that are involved are COL1A1 and COL1A2 and the mutation in these 2 genes is transmitted recessively, dominantly or appears as a new mutation. (Burtsev et al., 2019)

Type I collagen is a triple helix, consisting of two pro-α1 chains and one pro-α2 chain, which are synthesized from the COL1A1 and COL1A2 genes, respectively. Mutations slow down the modified α-chain folding. Which leads to longer interaction between the enzymes involved in post translation modification and the α-chain, disrupting its structure. Which leads to impairment of exocytosis and cross-linking of collagen molecules into fibrils, which can lead to apoptosis activation. The architectonics of bone tissue is violated due to decease in quality or quantity of type I collagen or both (Zeitlin et al., 2003).

Classification of Osteogenesis imperfecta

In 1849,W.Vrolick coined the term "Osteogensis imperfecta" (Womack, 2014). In 1979, Sillence et al conducted an epidemiological and genetical study of OI in Victoria, Australia. On the basis of their findings, they classified OI into "4 distinct syndromes" based on radiographic findings, clinical characteristics, and mode of inheritance, calling this the "Sillence Classification of Osteogenesis Imperfecta. (Womack, 2014). This classification has become the gold standard for classifying OI. In 2004,Rauch and Glorieux expanded the classification by adding 3 additional types(V, VI, and VII), which had the clinical symptoms of OI, but had defective genes other than COL1A1 or COL1A2 (Rauch and Glorieux

2004). In 2007, recessive inheritance type VIII was added by Van Dijk et al. (2010). Since then, types IX, X, and XI have been described in the latest literature(Van Dijk et al. 2010).

The purpose of deformities correction is to reduce the frequency of fractures and the problem of recurrent fractures and deformity with osteogenesis imperfecta has led to the use of intramedullary fixation devices ensure the proper growth of the bone and to make the long bones straight to allow the child to walk (Wilkinson et al., 1998). Protection with plastic braces can be used temporarily in children with thin bones until the bone diameter permits rodding. Children with mild forms of OI, who have straight legs and low fracture rate, do not need rodding.(Wilkinson et al., 1998)

After the use of Bisphosphonat (BP), the possibilities of surgical treatment have significantly expanded. El Sobky et al. showed that children who received pamidronate preoperatively showed better results than those who underwent only surgical treatment (El-Sobky et al., 2006). The combined multidisciplinary approach has significantly improved the treatment results.(Montpetit et al., 2015; Ruck et al., 2011).

The literature describes using the following structures for fixing bone fragments:
Non-telescoping, Rushnail, Kuntscher's pin, titanium elastic nails (TEN), Kirschner pins, unreamed humeral nails and External fixation devices. Telescoping internal fixation devices (Bailey–Dubow, Sheffield, Fassier–Duval rods), (Burtsev et al., 2019).

First described the technique of multiple osteotomies (Sofield and Millar, 1959) followed by an intramedullary nail insertion (Fig.14). Sofield and Millar (1959) were the first to develop an intra- medullary rod system for the treatment of osteogenesis imperfecta, but it was solid and unable to accommodate for axial skeletal growth this leads to recurrence of deformity at the tip of the nail and high risk of fracture in the un-splinted distal segment. Requiring repeated surgery for nail exchange and deformity correction, also the low stability of the nails leads to loosening and slippage when the bone increases in length and diameter. (Sofield and Millar, 1959; Santili et al., (2004)

This led to the development of an extensible rod by Bailey and Dubow, However, the complication rate of the Bailey-Dubow rod exceeded that of the non-elongating rod which it was designed to replace(Wilkinson et al., 1998). In 1989, a modification of Baily-Dubow rod was done (the Sheffield rod). The T shaped piece was permanently fixed to the rod ends and each T piece was notched to allow for controlled rotation when buried into the epiphysis. The sleeve T piece has a hole at its center to facilitate exchange rodding during axial growth(Wilkinson et al., (1998)

Fig.(1):Deformity of the femur under the tip of non-telescoping nail(Sofield and Millar, 1959)

Comparison between results of telescopic (Baily dubow and sheffield)versus non-telescopic rodding have shown that the rate of re-rodding is 50% for simple and between 20 and 40% for telescopic rods at 5 years follow up (Porat et al., 1991; Zeitlin et al., 2003).

A modification of the Sheffield rod was removal of the T piece from the distal rod and placement of locking hole to facilitate its placement and removal (interlocking telescoping rod), The rod was inserted without need for arthrotomy for the distal joint with better anchoring to the distal epiphysis but it is Technically demanding to insert a very small locking wire through a small slot, with high exposure to image intensifier. Both insertion and removal are much less invasive than removal and insertion of telescoping nail with a T piece end.(Cho et al., 2007)

In 2003, the Fassier-Duval telescoping nail was first introduced in Canada, it has the advantages, over the older telescoping nail, of having single proximal entry point, its insertion doesn't require joint arthrotomy either proximally or distally, so decreases the damage of articular cartilage and allows early rehabilitation. It has threaded epiphyseal portion that allows the rod to settle into the epiphyseal portion both proximally and distally and gives the nail some stability. The reports show good stability and function of the implant(Noe, 2011; Biggin Munns, 2017).This design of telescoping nail achieves the basic goal of telescoping mechanism and this makes the growing bone internally splinted along its entire length as the nail elongates when the bone grows(Noe, 2011).

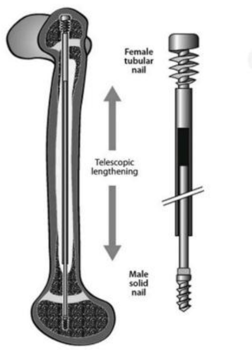


Fig.(2):Diagram showing design of Fassier –Duval rod(Makhdom et al., 2015).

The components of fassier-Duval nail

1) Outer tubular sleeve with proximal threaded portion to be anchored to the proximal epiphysis
2) Inner solid rod with threaded distal part to be anchored to the distal epiphysis.

It has the same design for the femur, tibia and humerus.

Surgical technique of fixation by Fassier-duval telescoping nail

The standard Surgical technique for nail insertion is the open osteotomy technique, The first step is to do an open osteotomy at the most proximal part of diaphysis at the first center of rotation and angulation (1st CORA), then gradual reaming of the proximal diaphysis is done reaching the entry point at the epiphysis. Then reaming of the distal diaphysis reaching the second center of rotation and angulation (2nd CORA), if present, and finally reaching the distal physeal growth plate. This is followed by insertion of a male rod of appropriate length and diameter from proximal epiphysis through the entire length of the bone to the distal epiphysis. The next step is insertion of the suitable size female hollow tubular part from the proximal epiphysis to be stopped shortly before the threaded part of the male rod and above the physeal growth plate(Fassier, 2017) Fig.(3)

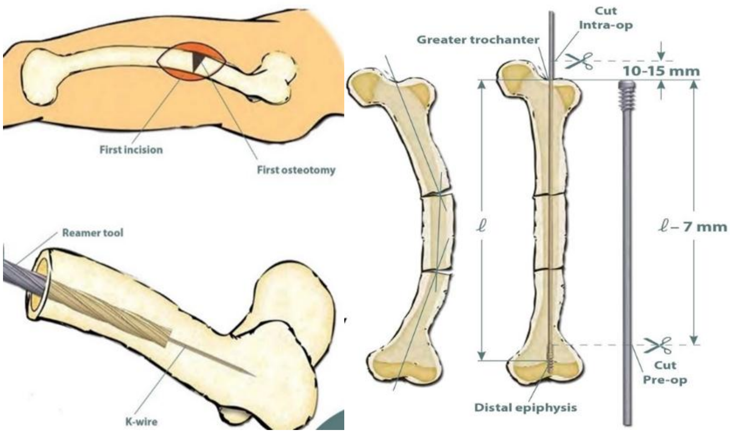


Fig.(3) Diagram showing technique of fassier -Duval nail for the femur. (Fassier, 2017)

Patients and Methods

This prospective case series study was conducted on 70 lower extremity bones with deformities or fracture in 48 patients suffering osteogenesis imperfecta. They were treated using Fassier-Duval telescopic nail (growing nail) in kasr Al Ainy school of medicine (Abu-Elresh new children Hospital). From September 2018 till June 2021 with 2 year follow-up. Kasr Al Ainy School of medicine Ethical Committee have approved this study.

Sample size:
70 lower extremities with deformities or fractures in 48 patients with OI

Inclusion criteria are;
1- Clinically confirmed diagnosis of Osteogenesis imperfecta.
2- Presence of Long bone fractures or deformities
3- Age between 2.5 years and 10 years in female and 12 years in female to make sure that the child still have potential to grow, as most OI patients suffers premature physeal closure because of microfractures that affect the growth physeal plate(Van Dijk et al., 2010).

Exclusion criteria are:
1- OI patients near skeletal maturity.
2- Infected previous operations.
3- Narrow medullary cavity that doesn't permit the smallest nail diameter (3.5 mm nail).

A full workup was performed for every patient; this includes patient assessment, Clinical evaluation (history, general and local examination) and patient counselling.

Follow up protocol:
Suture in children is always an absorbable suture, so no need for suture removal. Care of the wound is done by repeated dressing if the wound is soaked, cast is always mandatory to control the Rotation of the bone fragments as this type of telescoping nail is rotationally unstable, the cast should be removed if wound infection is suspected.

X ray follow up is done
1. Immediate post operative.
2. At 6 weeks before removing the cast.
3. At 3 months.
4. And every 6 months.

Follow up parameters
1- Fracture union
2- Refracture
3- Lengthening of the nail
4- Improvement of life style and the ability of walking

Possible Risk
- Infection
- Residual deformity
- Hardware complication (migration, loosening)
- Non-union

Primary outcomes
- Correction of the deformity.
- Union of the fracture.
- Ongoing lengthening of the nail.

- Range of motion in the proximal and distal joint.
- Motor function outcome using Brief Assessment of Motor Function (BAMF)score.

Secondary outcome parameters

- Operative time.
- Blood loss.
- Stability of the fracture site.
- Imaging time.
- Infection

Results

From September 2018 to June 2021, 48 patients (70 operated lower extremity) (70 segments) with Osteogenesis Imperfecta, met the inclusion criteria, operated upon in their lower extremities using telescoping Intramedullary nails. The 48 patients (70 operated lower extremities) were followed up for 2 years.

Demographic data:
Age:
The age of the patients was ranging from 2.5 to 13 years with the mean age is 5.6 year and SD 2.49 (5.6 ± 2.49) years.

Gender:
58.6% of the operated patients were male, while (41.4%) of the operated patients were female.

Parent consanguinity
37.1% of patients were have consanguineous parents, while(62.9%) of patients were have non-consanguineous parent. .

Classification of the studied Patients according to Sillence and Danks classification for Osteogenesis Imperfecta:
In 50 operated lower extremities the patients were type I (71.4%), 26 of them were type IA (37.1%) and 24 were type IB (34.3%).
In 15 operated lower extremities the patients were type III (21.4 %).
In 5 operated lower extremities the patients were type IV (7.4 %).

Although different types, the same treatment modalities were applied.
Indications for surgery in the studied patient.

1. Only in 13 operated lower extremities (18.57 %) the indication for surgery was fracture. because their parents are familiar with dealing with fracture because fractures are very common.
2. In 57 operated lower extremities (81.43 %) the indication was deformity correction.

In both deformity correction and acute fracture management intramedullary splinting is a must.

Operated side:
36 Rt lower extremities (51.4%) and 34 Lt lower extremities were operated upon (48.6%).

Preoperative Bisphosphonate treatment.

All patient with OI once diagnosed are referred to assessment for Bisphosphonate therapy. In 65 operated lower extremities the patient were on Bisphosphonate which was stopped 2 months pre-operative for long acting I.V zoledronic acid and 2 weeks for Oral Alendronate (92.8%). And in 5 operated lower extremities the patients weren't on preoperative Bisphosphonate (7.2%) because they presented with Acute fracture and they were not on bisphosphonate.

Intra operative blood loss
Is variable according to the disease severity, deformities severity, operative time, site of operation (femur or tibia because of the possibility of the use of torniquet). In tibial telescoping nails the blood loss was variable from 40- 90 cc with Mean blood loss 69.5 cc. thanks to the use of torniquet with no need for intra or post-operative blood transfusion. In femoral telescoping nails, the blood loss was variable from 100-300 cc with Mean blood loss 217.5 cc

Blood transfusion
According to pre-operative Hb level, intra operative and post operative hemodynamic stability of the patient and the amount of blood loss. According to these parameters blood transfusion was needed in **60%** of patients underwent unilateral femoral telescoping nail and **100%** of patients underwent Bilateral femoral telescoping nail but it wasn't needed in patients underwent telescoping nail tibia.

Intra operative image intensifier time.
The mean image intensifier time was 269.39 ± 89.31 seconds. It is variable according to surgeon experience and the severity of the deformity.

Post-operative data.
1. Infection:
Post operative wound infection occurred in two patients(segments) with femoral telescoping nail out of 70 telescoping nail operations(segments) (2.9 %) and turned into chronic infection with repeated remissions and exacerbations for them revision with exchange nailing has been done.

2. Joint Intrusion:
Joint Intrusion by male rod is not a frequent complication and usually occurs due to inappropriate technique in nail insertion. This complication occurred in 5 operations out of 70 operations (7.42%).
In the tibia 3 out of 16 tibial telescoping nail operations (18.75 %), in the femur 2 out of 54 femoral telescoping nail operations (3.7 %).
If it was detected intraoperatively, it could be managed by changing the site where the threaded part of male rod is anchored, because withdrawing the nail up and keeping it in place will result in joint intrusion again.

Fig.(4): Radiography shows joint intrusion by the nail

1. Proximal migration of the female rod

It occurred only in the femur due to inappropriate insertion of threaded part of female rod, above the ossified nucleus of greater trochanter. It occurred in 8 patients (segments) out of 70 (11.42 %).

Fig (5): Radiography shows proximal migration of female rod

4- Over growth of greater trochanter or tibial epiphysis above the threaded part of female rod.

It occurred in only 1 patient with telescoping nail tibia out of 16(6.25 %) and in 13 patients with telescoping nail femur out of 54 (24.07%).

This makes the removal of the female threaded part is extremely difficult) when needed).

Fig(6): Radiography shows over growth of greater trochanter(Rt side) and proximal migration of female rod over greater trochanter (lt side).

5-Dislodgment of the threaded part of the male rod from distal epiphysis:
Occurred in 13 telescoping nails out of 70(18.57%), and resulted in failure of telescoping or limitation of telescoping, this occurred either secondary to jamming of both component which was the cause in 7 telescoping nails femur out of 54(12.9%) or without jamming which occurred in the tibia.

In the tibia, it occurred either early by jamming of the nail components in 1 telescoping nail tibia out of 16 (6.25 %) or occurred later during follow up after 1 year due to the use of male rod that have long threaded part which was fixed to the distal tibial metaphysis in addition to the distal tibial epiphysis. Normally the metaphysis moves upward with longitudinal bone growth, and it gradually pulls the threaded part of male rod upward if it is attached to it. This occurred in 5 telescoping nail tibia out of 16(31.25%). this complication could be avoided by appropriate insertion of threaded part of male rod (central in the distal epiphysis in both Ap and lateral views) and by the use of male rod that have short threaded part (the threads should be completely inserted inside the epiphysis) and making sure that the distal tibial epiphysis will admit three full complete threads without crossing the distal physeal growth plate or the subchondral bone.

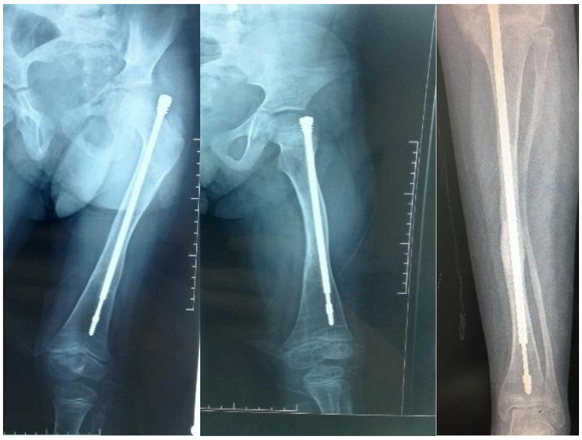
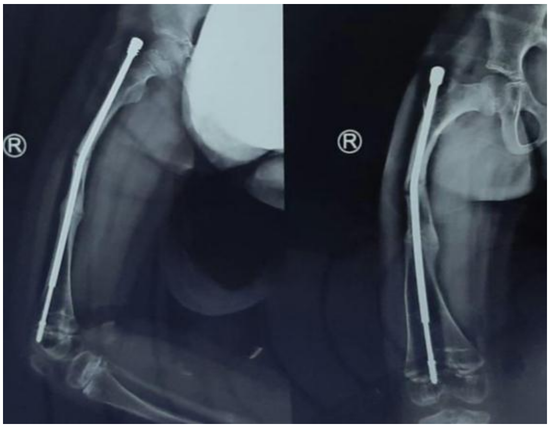


Fig (7): Radiography shows dislodgement of threaded part of male rod from distal epiphysis

6-Bending of the telescoping nail:

It Occurred in 2 cases out of 70(2.85%). it occurred after 1 year follow up after expansion of two parts of the nail from each other's (mostly due to sustained trauma and incomplete fracture of bone).



Fig(8):Radiography shows bending of telescoping nail.

7-Failure of expansion or limited expansion:

It occurred in 29 telescoping nail operations out of 70(41.42 %) Bending, jamming, dislodgment of male, proximal migration of female and joint intrusion all are causes of failure of expansion, also early closure of physeal growth plates (which is a manifestation of the disease) will results in limited expansion.

8-Fracture after operation:

It occurred in 3 cases out of 70(4.28%). In two cases of them the fracture occurred and the nail has been only bent (2.85%) while in the third case the fracture occurred on the tip of the nail(due to failure of telescoping and the bone overgrewed the nail tip)(1.14%).

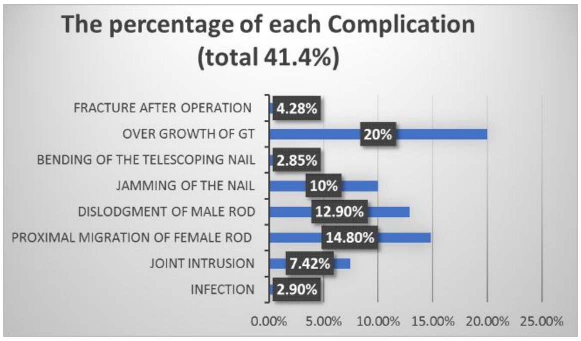


Fig.(9):Distribution of incidence of complications in studied patients.

Because of the overlap of complications in one patient, the total incidence of complications was (41.4 %).

10-Revision of operation

In 16 telescoping nail out of 70(22.8 %) revision was done for different causes:

1. Chronic infection in 2 cases out of 70(2.8 %) required Debridement, culture and sensivity and exchange nailing.
2. Joint penetration in 5 telescoping nail out of 70(7.14%) required not only readjustment of nail length but also change the site of distal epiphysis insertion.
3. Proximal migration of female rod in 8 telescoping nails out of 70 (11.42 %) all of them in the femur where re-insertion of female rod in the appropriate site has been done.
4. Failure of telescoping of nail with bone overgrowth the nail and fracture on nail tip in 1 telescoping nail out of 70(1.42%).

Functional outcome:

Despite having straighter and stronger bones following rodding and bisphosphonate treatment, children with OI must have adequate muscle strength in order to ensure good functional outcome (Biggin and Munns, 2017).

According to the Brief Assessment of Motor Function (BAMF) classification, the improvement occurred in standing, walking and running in 29 patients out of 48(60%). this improvement occurred in:

1) Patients with less severe form of the disease and all of them were receiving Bisphosphonate pre operatively and post operatively, 5 patients were type IV and 24 patients were type I
2) The age of improved patients was ranging from 2.5-4.9 years at time of surgery. (the Younger the age of the patient at which correction of deformity has been done, the better the functional outcome).
3) Patients who didn't sustain any fracture post operatively and started a good physical therapy program.

In theses 29 patients the BAMF was (5-9) pre-operative and improved to (7-10) at 2 years follow up, this improvement is due to improved mechanical environment by I.M fixation, biological environment by bisphosphonate and muscle power by physiotherapy program.

The other group of patients which doesn't show significant improvement in the BAMF was representing 40% of the studied patients (19 patients out of 48 patients). They were older age, their age was (5-8) years at time of operation, higher weight patients, with more severe form of the disease and more severe deformity, The results from the multivariate linear regression indicate that older age (P = 0.0045) and higher weight (P = 0.0164) are significantly associated with lower scores. Although they showed no improvement in their BAMF at 2 years follow-up, they showed improvement in mobilization, feeding, disfigurement and pain.

Our findings indicate that telescopic I.M nails resulted in beneficial effect in ambulation, gross motor function, self-care, feeding and mobility for children with OI. Telescopic I.M nail is a procedure which can improve the overall mobility in children with OI with significant deformities and repeated fractures.

Discussion

Ostogenesis imperfecta is a hereditary bone disorder that results in quantitative and/or qualitative defect in type 1 collagen synthesis, type 1 collagen is a major structural protein of bone and connective tissue. This defect manifests clinically by bone that is weak and liable to deformity and fractures. The severity of deformities and frequency of fractures depend strongly on the type of Osteogenesis imperfecta, they are variable from type to type and within patients of the same type of the disease.

Because OI is a genetic disorder, so its definitive management is to correct this genetic disorder at the genetic and molecular level by stem cell and gene therapy. Till now this modality of treatment is under experimental level and not well established as the standard of management. And till now our aim in treatment patients with OI is to decrease the incidence and frequency of fractures by bisphosphonate therapy and by intramedullary splinting of long bones which are the standard of management of OI patients.(Burtsev et al., 2019)

Sofield and Millar (1959) are described the first technique of multiple osteotomies and intramedullary fixation for the management of long bone deformities in OI patients and it is the standard till now. non-telescopic intramedullary implants were first used and it was documented to have a re-operation rate of 73 % of treated patient also the low stability of the nails leads to loosening and slippage when the bone increases in length and diameter.

The problem of treating young children with residual bone growth is bone overgrows the nail and these results in unsupported distal end of bones with secondary deformity and fracture at the tip of the nail, this was the cause of high reoperation rate. The telescopic intramedullary devices were developed to provide a good mechanical stability of bone and to elongate with bone growth to overcome this problem.

The first introduced telescoping intramedullary device was the Baily-Dubow telescoping nail, However, it was associated with a high incidence of complications, particularly related to the T piece .(Gamble et al., 1988)· A modification of this nail was The Sheffield telescopic rod system with the T piece permanently fixed to the outer sleeve overcame T piece related complications but it shared the complications of joint arthrotomy, traumatic insertion and rod migration.(Luhmann et al., 1998)

In hopes of optimizing the function and stability during periods of bone growth, A single entry telescopic IM system that provides implant elongation with longitudinal bone growth (Fassier-Duval telescoping intramedullary system) has been developed as the latest modality of treatment of OI patients and it has a widespread acceptance in several pediatric orthopedic centers around the world. but it is known to have several complications like joint penetration, rod migration, Bending of nail and failure of telescoping. (Spahn, et al., 2019)

The current study we investigated the functional outcome of the patients after telescoping IM nail, nail elongation with longitudinal bone growth, and the decrease in the re-operation rate secondary to this elongation. Our findings support the good result of telescoping IM nails in OI pediatric patients based on higher implant survival among telescoping rods during the first 24 month's surgery as compared with static intramedullary devices or other telescoping IM devices.

Non telescopic Sofield rods were documented to have a revision rate as high as 73% (Spahn, et al., 2019). The telescopic Bailey and Dubow rods were reported to have reoperation rates ranging from 21% to 32% and the Sheffield rods had a reoperation rate of 20%. Nicholas et al. (1990) reported a re-operation rate in 50 %of patients with Baily-Dubow rod in mean follow up of 4.8 years. Zionts et al. (1998) reported a re-operation rate in 37%of patients with Baily-Dubow rod in mean follow up of 7.5 years. Fassier (2021) and Fassier et al. (2006) reported a reoperation rate of 14% after Fassier-Duval femoral rodding. In another study, Fassier et al. (2010), reported a same reoperation rate in a study of FD tibial rodding. Brike et al. (2011) and Suresh et al., 2021), reported a reoperation rate of 13% in tibial and femoral FD rodding.

In this study the reoperation rate was 20 % in the first 2 years follow up that could be decreased by being meticulous and avoiding pitfalls during surgical technique. One of the most significant complications is intra operative or post-operative joint penetration (intrusion) by threaded tip of male rod, this occurred due to overscrewing of male rod inside the distal epiphysis or pushing of male by female rod during its insertion.

If it is identified intra-operative, it will be managed by change the site of insertion of the male rod in the distal epiphysis by slight extension of distal fragment because if it is screwed out without changing its insertion site, it will re-advance again through this defect in the articular cartilage and if it is not identified intra-operative it will necessitate revision. Another cause for joint penetration is pushing of the male rod by the female rod if there were any difficulties during its insertion; this could be avoided by adequate correction of bone deformities and insertion of appropriate size FD nail.

In this study the incidence of joint intrusion was 7.42%. Holmes et al. (2020), reported joint intrusion in 3 patient out of 13 patients (23.07%) with FD nail. Azzam et al. (2018) reported an incidence of 1 patient out of 58 patients with FD nail (1.7%). Brike et al. (2011) and Suresh et al., 2021, reported incidence of 14.6 % of joint intrusion in a study of 15 patients with FD nail. As regards Baily-Dubow nail, Nicholas et al. (1990), reported a joint penetration rate in 4 %of patients with Baily-Dubow rod in a mean follow up of 4.8 years. Zionts et al. (1998), reported a joint pentation rate in 12%of patients with Baily-Dubow rod in mean follow up of 7.5 years.

Proximal migration of female rod occurred due to Loss of fixation of its threaded part in the proximal epiphysis or ossified nucleus of GT secondary to inappropriate insertion, this occurred more in younger age patients due to small ossified nucleus of GT as this depends on tactile feedback of proper purchase in the apophysis which is different from bone. This complication is a cause for reoperation (necessitates reinsertion again).

In this study, the incidence of proximal migration of female rod was 14.8%. Brike et al. (2011) and Suresh et al., 2021, reported 13% incidence of proximal rod migration in FD nail. Azzam et al. (2018) and Liebs (2018), reported a 16% incidence of proximal rod migration in FD nail. Cox et al. (2020), reported a 7% incidence of proximal rod migration in FD nail. Dislodgement of threaded part of male rod from distal epiphysis results in failure of telescoping or limited telescoping. it occurres more commonly in the tibia rather than the femur.

In our study, it occurred in 13 FD telescoping nails out of 70(18.57%). In the femur it occurred due to twisted tip of male rod which made the female jammed to the male rod (function as an ordinary nail) with secondary overgrows of bone distally and migration of the nail upward.

This occurred in 7 telescoping nails femur out of 54(12.9%) (Who suffered failure of expansion). In the tibia, it occurred either early by this previously mentioned cause (jammed nail)in 1 telescoping nail tibia out of 16 (6.25 %). or occurred later during follow up due to fixation threaded part of male rod in the distal tibial metaphysis(long threaded part of male rod transfix the distal epiphysis with the metaphysis). Normally the metaphysis moves upward with longitudinal bone growth, and it gradually pull the threaded part of male rod upward if it is attached to it.

This occurred in 5 telescoping nail tibia out of 16(31.25%). This complication could be avoided by appropriate insertion of threaded part of male rod (central in the distal epiphysis in both Ap and lateral views) and avoid transfixing the distal tibial metaphysis by long threaded part of male rod (the threads should be completely inserted inside the epiphysis) the distal tibial epiphysis should admit three full complete threads without crossing the distal physeal growth plate or the subchondral bone in order to be a stable fixation. Otherwise, proximal migration is anticipated.

Holmes et al. (2020), reported an incidence of pull out of threaded male alone in 3 cases out of 15(20%) and pull out with failure to telescope in 2 out of

15(13.3%).Brike et al. (2011), reported an incidence of 16,6% of pull out of male rod in a study of 15 patients with OI treated with FD nail.

FD rod placement centrally within the distal tibial epiphysis improves rod survival in patients with OI. The greater the rod positioning deviated from midline of the distal epiphysis, primarily lateral deviation, the greater the risk of the rod failing. All efforts must be made for positioning the distal tip of the male rod centrally within the epiphysis on both the AP and lateral planes, even if it requires an extra osteotomy to accomplish that(Holmes et al.. (2020).

Failure of telescoping occurs when the friction between male and female rods exceeds the pullout strength of threaded part of male and female rods, commonly due to bending of female or male rods during their cutting or their insertion. In this study 29 FD nails failed to telescope out of 70 FD telescoping nail operaton (24.07 %). Holmes et al. (2020)., reported an incidence of failure of telescope in 2 cases with FD nails out of 15 cases(13.3%). Brike et al. (2011), reported failure of telescoping in 5 out of 24 cases(20.83%)·(27)

The incidence of surgical site infection is very low in OI patients in either telescoping or non-telescoping nail operations. Gamble et al. (1988), Nicholas et al. (1990), Porat et al. (1991), Zionts et al. (1998), Boutaud et al. (2004), Grossman et al. (2018), Spahn et al. (2019), Brike et al. (2011), didn't report any infection in their studies.

In our study, 2 patients with telescoping nail femur out of 70 telescoping nail operations got infected (2.9 %) and turned into chronic infection with repeated remissions and exacerbations. That was managed by exchange nailing after failure of rebated debridement only. It is considered a high rate if compared with other studies in the literature that didn't report any infection.

Non-union after repeated fractures is not uncommon complication in OI patients, in this study Nonunion occurred in 2 cases out of 70 (2.85%), and delayed union for 1 year in 2 other cases out of 70(2.85%). Which is less than incidence of non-union reported by Agarwal and Joseph, (2005), reported a non-union in (20 %) of patients after non-telescoping nail operations. and Azzam et al (2018), reported a non-union in (14 %) of patients after FD nail operation.

Bending of the telescoping nail occurs when the patient sustains trauma, usually unicortical fracture occurs accompanied by bending of the nail. In this study, It Occurred in 2 cases out of 70(2.85%). And this occurred 2 years post -operative in one patient and 1.5 year post-operative in another patient, as the two parts of the nail became weaker after expansion of two parts from each other and its liability for bending increased. When bending occurs, the nail loses its capability to expand and function as an ordinary nail.

Azzam et al. (2018) reported an incidence of bending in 5.5 % of patients after FD nail operation (10 cases out of 179). Although Crossing of the Physeal growth plate by the threaded part of male rod theoretically carries the potential risk of physeal bar formation and subsequently growth disturbance and mal-alignment of the limb, Fortunately this complication hasn't been reported neither in our study nor in the literature (Brike et al., 2011; Holmes et al., 2020; Rodriguez Celin et al., 2020).

Conclusion

This Current study indicates that the telescoping IM system has good results in 60% of the studied patients with good telescoping of nail while complications occurred in 40% of patients. The improvement occurred in self-care and mobility for children with OI and they showed an improvement in BAMF in 60% of operated patient at 2 years follow-up.

The newly design telescopic nail (Fassier- Duval type) has the advantage of eliminating the need for knee and ankle joints arthrotomies (that was needed for insertion of previously used telescopic nails), and this reduces the rehabilitation time required. The improvement in function outcome in OI patients following telescopic nail operations needs good muscle strength together with straighter and stronger bones following bisphosphonate treatment and intramedullary splinting.

The FD telescoping nail procedure does involve meticulous technique and experience to ensure proper correction of all deformities and appropriate placement of the rods to achieve lengthening of the nail with bone growth. The high incidence of complications in telescoping nail operations will decrease with increase learning curve for this procedure and this emphasizes the role of specialized center and well qualified surgeon.

Fixation of threaded part of female rod in the ossified nucleus of GT is very difficult in younger children with not well ossified nucleus of greater trochanter and this results in higher incidence of proximal migration of female rod in patients with non ossified nucleus of greater trochanter. Also, fixation of distal threaded part of male rod in the distal tibial epiphysis is very difficult in younger children with a small distal tibial epiphysis and it should admit three complete threads without penetrating the ankle joint or transfixing the distal tibial metaphysis as this resulted in proximal migration of male rod.

We should put in mind that telescoping nail insertion is a very difficult and meticulous procedure, in a very delicate and difficult patient that needs very high learning curve and very good selection of patients. As regards lack of rotational stability of FD type telescoping nail, cast should be done for a period of 6 weeks to control the rotation. With no significant complications detected for the cast during follow-up.

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