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Case Series

Treatment of Osteogenesis Imperfecta with Corkscrew Tipped Telescopic Nail

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ABSTRACT

Introduction and Aim: Osteogenesis imperfecta (OI) cause health problems in all tissues contain type-1 collagen. The patient is complaining from easily fragile bones due to loss endochondrial normal ossification. Different treatment techniques are in use. Telescopic nails gain wide acceptance to treat OI since its introduction. However, it newly introduced in our institution. The aim of this work was to assess the results of surgical intervention of osteogenesis imperfecta with corkscrew tipped telescopic nail

Patients and methods: This prospective case series included 10 cases of OI, and treated surgically with corkscrew tipped telescopic nail and followed up for six months. The study completed during the period from January to August 2020. It was ethically accepted and an informed consent signed by the patient or his/her guardians. Each included patient was subjected to full history, physical examination and laboratory examinations. In addition, an X-ray had been performed. The orthopedic repair was performed and every patient had a cast after surgery. The collected data included operative time, duration of hospital stay. Postoperatively, patient ambulation was encouraged.

Results: Patients were 7 males (70.0%) and 3 females (30.0%). Their age ranged between 3 and 10 years. The operation was for tibia in 40% and femur in 100%. The postoperative complications were infection in 10%, and migration of the nail in 10%. No union or fracture after nailing was reported.

Conclusion: Telescopic nailing is an effective and relatively safe technique in the management of osteogenesis imperfecta.

Keywords: [Osteogenesis Imperfecta](#); [Telescopic Nailing](#); [Corkscrew](#); [Complications](#).



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INTRODUCTION

Osteogenesis imperfecta (OI) is a genetic connective tissue disease, which inherited in either autosomal dominant or recessive nature. It is due to Type-I collagen production deficiency of alpha-1 and/or alpha-2 chain. All tissues containing type I collagen are prone to problems of the disease. The main bony defect is the loss of normal ossification, especially of the endochondrial bone. Also, the disease exerts systemic problems (e.g., otosclerosis, blue sclerotic, cardiac disorders, joints' elasticity and thinning of the fascia. Bone fragility is a dominant compliant⁽¹⁾.

Although associated recurrent fractures heal spontaneously, they usually lead to high rate of deformities⁽²⁾. In OI, the aim of treatment, regardless its type, is the induction and maintenance of osteosynthesis. It could be accomplished by the usage of intramedullary nailing, which considered as the cornerstone in surgical treatment especially in patients with bone deformity. Intramedullary nailing can correct deformity and prevent recurrent fractures. It should be performed in a way that assure that the growth of children will not be hindered and should decrease the need for recurrent surgeries as the child grows⁽³⁾.

The types of intramedullary nailing are limited in number which specifically designed for treatment of OI and research is continued for the standard nail. Elastic telescopic nails with extendable features are introduced and tested. It showed promising results. However, the elastic nails had increased risk of migration over time, and telescopic nails are associated with the need to open the joint (arthrotomy) during insertion and fixation to prevent migration, which represented another challenge⁽⁴⁾.

Accordingly, we started to introduce telescopic nails in treatment of OI, and here we presented our clinical results of the first ten patients.

The aim of this work was to evaluate the results of surgical therapy of OI with Corkscrew Tipped Telescopic Nail.

PATIENTS AND METHODS

This prospective case series conducted on 10 cases of OI, selected from Al-Azhar University Hospital (Damietta) by surgical treatment with corkscrew tipped telescopic nail and followed up for six months. The study completed during the period from

January to August 2020. Damietta Faculty of Medicine Al-Azhar university ethical committee had approved this study. In addition, all participated patients or their guardians signed an informed consent.

The inclusion criteria were: patients with OI, age group between 3 and 15 years, of both genders. On the other hand, exclusion criteria were: pathological conditions like tumors and infection, and the age group below 3 or more than 15. Each included patient was subjected to full history, physical examination and laboratory examinations. In addition, an X-ray had been performed.

The orthopedic repair was performed as described by Sarikaya *et al.*⁽⁴⁾. Briefly, general anesthesia was used for all patients who complete operation in the supine position. The surgical incision was performed proximal to the bone to be nailed. Bone osteotomy was introduced to the sites where the bone deformities were found by insertion of the thin nails. The advancement of thin nails was continued till it reached the epiphysis line. Then, 1-2 pieces of Kirchner wires and the distal end of the nail were secured. The entry site was secured by placing the thick over the thin nail after gaining the proper length and cutting it in the bone. All cases underwent casting after the end of the nailing surgery.

The collected data included operative time, duration of hospital stay. Postoperatively, patient ambulation was encouraged and ambulatory patients could bear weight after their casts were removed (started after the sixth week).

RESULTS

The current work included 10 patients, 7 were males (70.0%) and 3 were females (30.0%). Their age ranged between 3 and 10 years; the mean age was 6.3±2.9 years (Table 1).

The site of operation was the tibia in 40% and femur in 100%. The indication for surgery was un-union in 10%, acute fracture in 50.0% and deformity problem in 70.0%. The postoperative complications were infection in 10%, and migration of the nail in 10%. No union or fracture after roding was reported in the current study (Table 2).

Figures 1 through 5, represented two of the studied cases (preoperative and postoperative results) with good bone alignment and correction of deformity.

Table (1): Demographic Character of Studied patients

		Statistics
Gender	Male	7 (70%)
	Female	3 (30%)
Age (years)	Mean ±SD	6.3±2.9
	Range	3 - 10

Table (2): Operative details among the studied patients

		N	%
Site of operation	Tibia	4	40
	Femur	10	100
Indication	Un-union	1	10
	Acute Fracture	5	50
	Deformity problem	7	70
Postoperative Complications	Infection	1	10%
	Un-union	0	0
	Fracture after Roding	0	0
	Migration of nail	1	10%



Figure (1): Preoperative plain X-ray, Anteroposterior and lateral views of a four years' female patient, showed C-shaped right femur and left corrected femur by telescopic nail



Figure (2): Intraoperative picture of the same patient, showing entrance of male part of telescopic nail.



Figure (3): Immediate postoperative, plain X-ray, anteroposterior and lateral views of the same patient showing correction of right femur deformity.



Figure (4): Preoperative plain X-ray, anteroposterior and lateral views of a male patient, 7 years old, showing deformity of both femur and tibia.



Figure (5): Immediate postoperative plain X-ray, anteroposterior and lateral views of the previous patient showing correction of deformed femur and tibia by telescopic nail.

DISCUSSION

In this study we were used corkscrew tipped telescopic nail in treatment of 10 cases of OI, at Al-Azhar University Hospital, New Damietta. Children with OI experience repeated fractures and associated severe deformities which demanded orthopedic surgery. The overall outcome was excellent. The rate of postoperative complications was 20% (two patients; one of infection and one of nail migration). Intramedullary nailing become the gold standard of treatment as recommended by Rosemberg *et al.*⁽⁵⁾. However, the characteristics of the applied nail must be individualized for each child due to continuous growth. In addition, osteogenesis imperfecta is a characteristic situation, as the bone is of low quality which need specific nail with narrow channel diameter to reduce the expected epiphyseal damage and permits nail extension with growing of bone. Also, the use of nail without extension needs frequent surgical intervention with its complications. Thus, telescopic nails are used in patients with osteogenesis imperfecta⁽⁴⁾. However, the narrow diameter of the nail is a disadvantages (making the nail weak)⁽⁶⁾.

Tibial nailing was reported in only 40% of patients in the current work, compared to 100% of femur. These results are in line with Rodriguez *et al.*⁽⁷⁾ who reported that, femoral rodding was done more frequently tibial rodding. In addition, femoral rodding was initially performed followed by the tibial one when both bones were rodded in successive manner.

Results of the current work as regard overall rate of

complications (20%) are higher than those reported by Bhaskar AR, Khurana⁽⁸⁾ who reported complications in 2 out of 21 (about 9.5%) (both were refracture). But their study is in line with the current as regard the final conclusion. They finally reach to the conclusion that, intramedullary nailing for pediatric osteogenesis imperfecta (OI) improves their potential of mobility. It also prevents muscle wasting, repeated fractures and osteopenia that affects bone quality.

The complications rate in the current work is lower relatively low when compared to previous literature. The overall complications rate of all types of nails in previous studies ranged between 35 and 40% and includes nail migration, refracture, delayed union or even nonunion, bent nail and others⁽⁹⁻¹¹⁾.

In addition, Azzam *et al.*⁽¹²⁾ reported a 14.5% (26 out of 179 cases) rate of incomplete non-union in their study. In the current one, no patients developed non or delayed union.

Sterian *et al.*⁽¹³⁾ report that telescoping rods are linked to a long-lasting osteosynthesis. However, potential joint stiffness is a potential unwanted complication. This complication usually developed as a result of arthrotomies and passage of nail insertion through joint cartilage during its insertion. They concluded that telescoping nailing is a good alternative intervention to avoid these problems.

In addition, Georgescu *et al.*⁽¹⁴⁾ reported that, in people with osteogenesis imperfecta of moderate to severe deformity of

their long bone, the surgical treatment is the sole chance to walk again.

Chiarello *et al.* ⁽¹⁵⁾ carried out 245 procedures, compared conservative treatment (with different surgical treatment modalities (e.g., pinning, intramedullary nailing, plating) and reported non-significant difference regarding the complications rate between both groups. However, they concluded that in OI, the intramedullary devices appeared to be better.

Behera *et al.* ⁽¹⁶⁾ concluded that, telescopic nails have improved the lives of osteogenesis imperfect patients by permitting greater mobility and reduced the number of future required surgeries.

Of great interest, Sa-Ngasoongsong *et al.* ⁽¹⁷⁾ reported that, pre-operative planning and individualization of each OI patient regarding appropriate treatment and best implant for patients is very important and represents a challenge. Many factors are work together (e.g., patient age, deformity and its severity, available instruments, its advantages and disadvantages, the surgeon experience, and the rate of post-operative complications are all factors that should be considered to achieve the best selection for each patient.

In short, results of the current case series added to existing evidence. It confirmed the effectiveness of telescopic nailing in management of osteogenesis imperfecta. In addition, the procedure is relatively safe with lower rate of complications.

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