



EARLY TELESCOPIC ROD OSTEOSYNTHESIS IN OSTEOGENESIS IMPERFECTA PATIENTS

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ABSTRACT Osteogenesis imperfecta is a genetically inherited condition with abnormal or deficient collagen resulting in generalized osteoporosis and early onset of multiple fractures even with low energy trauma or bowing of the long bones. The most commonly used classification in the literature is by Silience. The study group included 10 patients aged from 2 years to 12 years, both males and females. All the patients presented to the hospital are of walking age with multiple fractures. They underwent treatment with prolonged cast immobilization resulting in rapid bone demineralization, axial deformities, increased fractures and loss of ambulation. On follow-up, patients (Lobstein Disease) treated by open bone alignment and telescopic rod osteosynthesis showed better results. It has the advantages of rapid ambulation and decreased soft tissue damage.

KEYWORDS : Osteogenesis Imperfecta, Lobstein disease, Telescopic rod osteosynthesis

INTRODUCTION -

Osteogenesis imperfecta is a genetically inherited condition with abnormal or deficient collagen resulting in generalized osteoporosis with early onset of multiple fractures after low energy trauma or bowing of the long bones [1]. Clinically the diagnostic triad includes the blue sclera, dentinogenesis imperfecta and generalized osteoporosis in patients with multiple bone fractures and bowing of the long bones of which two characteristics present in all the patients are fractures and osteoporosis. COL1A1 and COL1A2 are the two main genes involved in pathogenesis. The mutations show autosomal recessive, autosomal dominant or spontaneous inheritance resulting in the synthesis of the defective collagen either in quantity or quality [2]. Silience classification initially divided the disease into four groups and added to type VII [3,4].

Table 1: Silience DO, Danks DM : The Differentiation Of Genetically Distinct Varieties Of Osteogenesis Imperfecta In The Newborn Period, Clin Res 26:178, 1978

Type	Inheritance	Bone fragility	Deformity of long bones	Growth retardation	Spine	Incidence
I	Autosomal dominant	Variable, less severe than mostly	Moderate	Short stature	Scoliosis and kyphosis in 20%	1/30.000
II	Autosomal recessive	Very extreme	Crumbled bone	Unknwn	-	1/62.000
III	Autosomal recessive	severe	Progressive bowing of the long bone	Severe, smallest of all patients	kyphosis	Very rare
IV	Autosomal dominant	Moderate	Moderate	Short stature	kyphosis	unknown

MATERIALS AND METHOD

10 patients, including male and female in a ratio of 1:2 ranging from 2-12 years, were taken under the study. All the patients presented with fractures of the long bones, most commonly involving femur at the age of walking. The mean number of long bone fractures including both upper and lower extremities was 3. According to Silience classification, the patients were grouped under type IV, type III and type I of 5, 3 and 2 children respectively who underwent conservative fracture treatment before presenting to us.

They underwent treatment with prolonged cast immobilization

resulting in rapid bone demineralization, axial deformities, increased fractures and loss of ambulation. The diagnosed patients were started on bisphosphonate protocol before the surgery. The treatment available for these cases was surgery proposed by Sofield-Miller corrective osteotomies and telescopic nail osteosynthesis. A 7-day course of bisphosphonates was given before the surgery. The rapidly evolving anterior bowing necessitates such an aggressive surgical treatment (sometimes associated with rotation) and the increasing incidence of fractures after cast removal. Choosing Sheffield-Millard osteotomies is based on the angulation of more than 20° and reaming because of the narrow medullary canals. In severe cases with marked bowing, a shortening of the segment is done to reduce tension on the soft tissue after the alignment of the bone.

Previously Kirschner wire [5], Rush nails [6], Kuntscher nails [7], Ender nail and recently, elastic nailing [8] were used in treating the fractures. However, these techniques provide short term results with bone alignment and prevent the refracture of the bone. But, the disadvantage with these implants with ageing of the patients were secondary bowing, refractures, increased distance between the rod and the growth plate, and the bone outgrowing the nail. Also, with fracture healing and growth of the bone, low stability of nails was seen with loosening and slippage. This led to the development of the first telescopic rod by Bailey and Dubow, later improved by Sheffield of the UK (9-11). It requires joint arthrotomies to insert the nail through the joint cartilage (12-14).

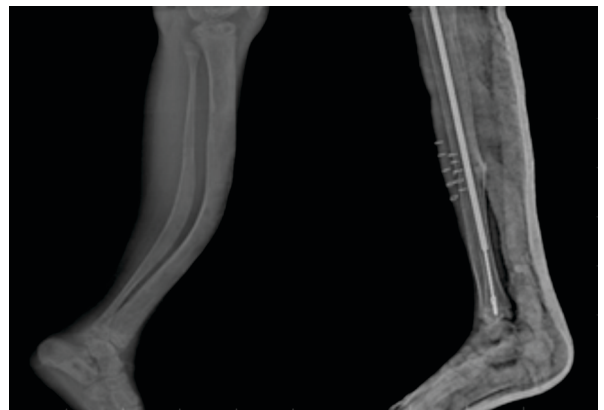


Fig 1: Lateral view showing Preop and postop xray of a 10y old child with OI with anterior bowing of both tibia treated with Osteotomy and telescopic nailing



Fig 2 : AP view showing Preop and postop xray of a 10y old child with OI with anterior bowing of both tibia treated with Osteotomy and telescopic nailing



Fig 3 : Preop and postop xray of a 7y old child with OI and fracture shaft of right femur treated with telescopic nailing

These new systems achieved the basic goal of telescopic nailing to obtain a long-lasting osteosynthesis in a growing bone and decrease the risk of secondary fractures, bowing and cannot be outgrown by the bone.

The disadvantages are the lack of rotational stability, so postoperative immobilization in a cast for at least 3weeks or till the radiographic callus appearance is needed.

CONCLUSION

On follow-up, patients (Lobstein Disease) treated by open bone alignment and telescopic osteosynthesis showed better results. It has the advantages of rapid ambulation, continued bone growth in length without bowing, preventing refractures and decreased soft tissue damage. Also, the rigid fixation and fracture healing gives the child an opportunity to lead a

normal life and decrease the secondary fractures that may damage the nail and eliminate the need for repeated surgeries. The goal of the telescopic rod is to provide a perfect solution for the OI patient to have a normal life without the fear of having a fracture and prolonged periods of cast immobilization or surgical interventions with uncertain results.

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