Telescopic Nail in Osteogenesis Imperfecta Early Experience in Nitor

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Abstract
Paediatric long bone lesions often need to be fixed or stabilized as a part of the treatment. But difficulties arise due to the bone is growing on. Osteogenesis Imperfecta is a genetic disorder that mainly affects the bone. It results in bones that break easily & deformed. It has no cure. The incidence ratio of OI is 1: 20000 to 1:50000 live births. For many years, Surgical Correction of deformities, physiotherapy and the use of orthotic support devices were the primary treatment. Intramedullary Fixation with the use of Telescopic Rod is one of the Standard Methods for long bone Stabilization in Growing children with Osteogenesis Imperfecta. Surgical Technique, Preoperative Radiographs are thoroughly evaluated to assess the deformity & to estimate the length and diameter of the Rods to be used. However, for severely angulated limb segments, we determined the Rod length intra-operatively, after performing multiple Osteotomies. From June 2016 to June 2018 in NITOR, we managed 9 Tibiae 6 Femur by Telescopic Nail at the age of 2 to 12 years.

Keywords: Telescopic Nail, Osteogenesis Imperfecta, Surgical Technique, Preoperative Radiographs.

Introduction
Osteogenesis imperfecta is an autosomal dominant or recessive connective tissue disorder caused by the deficiency of Type I collagen production associated with the deficiency of collagen Type I alpha 1 chain and collagen Type I alpha 2 chains. This disease causes problems altogether tissues that contain Type 1 collagen. In addition to several systemic problems like blue sclerosis, otosclerosis, cardiac diseases, elasticity within the joint and thinning of the fascia, it also causes the loss of the normal ossification of the endochondral bone.1 This results in easily fragile bones. Osteogenesis imperfecta may be a genetically determined pathology that suggests bone variability and osteoporosis with early onset of fractures after low energy trauma.2 The 2 main genes involved are COL1A1 and COL1A2 and
therefore the mutations are transmitted recessively, dominant or appear spontaneously and affect the synthesis of type one collagen during a quantitative, qualitative, or mixed way.\textsuperscript{3} For a far better understanding of the clinical problems, Sillence and Danks created a classification that originally had 4 groups but continued to feature more and more subclasses because the genetic study evolved progressing up to type XI.\textsuperscript{4,5} Today, telescopic nails with elastic or extendable features frequently resort. The elastic telescopic nails work as a drag of retraction over time, while the matter encountered the necessity to open the joint during fixation to prevent migration, which posed another challenge.\textsuperscript{6-8}

### Results

#### Table 1: Patients Diagnosis History & Status at a Glance

<table>
<thead>
<tr>
<th>Type</th>
<th>Inheritance</th>
<th>Bone fragility</th>
<th>Deformity of long bones</th>
<th>Growth retardation</th>
<th>Spine</th>
<th>Incidence</th>
</tr>
</thead>
<tbody>
<tr>
<td>IA</td>
<td>Autosomal dominant</td>
<td>Flexible, less severe</td>
<td>Functional</td>
<td>Short build</td>
<td>Scoliosis and kyphosis in 20%</td>
<td>1/30.000</td>
</tr>
<tr>
<td></td>
<td></td>
<td>than typically</td>
<td></td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>IB</td>
<td>Autosomal dominant</td>
<td>Flexible, less severe</td>
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</tr>
<tr>
<td></td>
<td></td>
<td>than typically</td>
<td></td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>II</td>
<td>Autosomal recessive</td>
<td>Very extreme</td>
<td>Crumbled bone</td>
<td>Unknown</td>
<td></td>
<td>1/62.000</td>
</tr>
<tr>
<td>III</td>
<td>Autosomal recessive</td>
<td>severe</td>
<td>Progressive bowing of the long bone</td>
<td>Severe, smallest of all patients</td>
<td>kyphoscoliosis</td>
<td>Very rare</td>
</tr>
<tr>
<td>IVA</td>
<td>Autosomal dominant</td>
<td>Moderate</td>
<td>Functional</td>
<td>Short build</td>
<td>kyphoscoliosis</td>
<td>unknown</td>
</tr>
<tr>
<td>IVB</td>
<td>Autosomal dominant</td>
<td>Moderate</td>
<td>Functional</td>
<td>Short build</td>
<td>kyphoscoliosis</td>
<td>unknown</td>
</tr>
</tbody>
</table>

Due to several fractures, rapid bone demineralization causes of a patient. Axial deformations of bones increased number of fractures and loss of ambulation. The only treatment for these cases was Surgery. Field–miller corrective osteotomies and Fassier–Duval are two telescopic nail osteosynthesis methods. Prior to the surgical intervention, patients had a bisphosphonate treatment. The necessity for such aggressive surgical treatment is given by the rapidly evolving anterior bowing. Sheffield-Millard osteotomies are approximately 200 angulations narrow medullar channel, which makes solution for passing the rod. Sometimes, in very severe cases, when the distance between the epiphysis gets smaller as the bone gets more and...
more bowed. It is imperative to do a shorting of the segment so that the soft tissues are not put in excessive tension after alignment.

**Figure 1:** 3-year-old patient with long bone bowing and multiple fractures of femur

**Figure 2:** Right femur bowing after 2 fractures in the middle section and right tibia bowing after 3 fractures

**Figure 3:** Left femur bowing after 3 fractures in the mid region and left tibia bowing after 2 fractures in the midpoint

**Figure 4** An 8-year-old patient with bilateral femur and tibia bowing treated orthopedically

**Figure 5:** A 3-year-old patient with 4 fractures of the tibia and anterior bowing

**Figure 6:** A 9-year-old patient with numerous femur fractures and almost 90° angulation
Discussion
The physicians used Kirschner wire, Kuntscher nails, Rush nails, Ender nail and most recently elastic nailing, until development the method. All the techniques had good short-term results, with good bone alignment and prevention of the re-fracturing of the bone. But, the problem with these materials that patients outgrew very fast. Like secondary bowing and re-fracturing. It appeared when the growth plates went too distant from the rod and the bone outgrew the nail. Also, the low stability of the nails was a down factor, as nail loosening and slippage became a problem after the bone healed and grew in length and diameter. Regarding this fact, the first telescopic rod system was developed by Bailey and Dubow, soon after being improved to the Sheffield rod in the UK. The basic goal of telescopic nailing was achieved with these new systems, meaning that one can obtain a long-lasting osteosynthesis in a growing bone that can have good long-term results and can decrease the risk of secondary fractures and bowing once the nail starts to tear and cannot be outgrown by the bone. The problem with these systems became clear when the children had to start moving after the surgery and the extensive soft tissue damage to the joint surface and capsule became an evident impending movement. These proved to be a problem as joint stiffness and pain prolonged the postoperative recovery period with a decrease in life quality and ambulation. This telescopic rod achieves this by giving the operator the opportunity to insert the components in the distal epiphysis under Rx control so that the growth plate takes minimal damage and the joint surface is protected.

Conclusion
Telescopic nails methods reduced the rate of re-operation from 51% to 27%. This difference is less noticeable in the long term due to mechanical complications and secondary joint problems. The healing of the bone and rigid system gives the kid the chance to possess a traditional life without the danger of secondary fractures that would damage the nail and demand a second surgical intervention. The goal of the telescopic rod is to offer the right solution for the Osteogenesis imperfecta patient to possess a traditional life without the permanent fear of getting a fracture that would mean long periods of cast immobilization or surgical interventions with uncertain results.

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