Titanium Elastic Nails Successfully Splint Subsequent Femoral Fractures in a Child with Osteogenesis Imperfecta

Authors: **Lutomia MBL**, MBChB, FCS Orth(ECSA), **Lakati KC,** MBChB, FCS Orth(ECSA).
Affiliation: Egerton University Medical School, Nakuru orthopaedic Centre, ICRC Geneva.*E-mail:* mlumbasi77@yahoo.com

Summary

The current trend in the management of fractures in children with osteogenesis imperfecta (OI) is the use of expandable nails. These may not be widely available in resource- constrained settings. We report a case of a 9 year old female with a peri-prosthetic fracture treated with premeasured pre-contoured titanium elastic nails allowing rapid mobilization. The nails also provided adequate splintage to subsequent proximal fractures of the same femur a few months later, obviating need for further surgeries. Titanium elastic nails without fluoroscopy are an option for treating fractures in children with OI for the average orthopaedic surgeon in the developing world.

Introduction

Osteogenesis Imperfecta (OI) is a heritable disorder with variable phenotypic expression. The inherent disorder of type 1 collagen synthesis may be associated with blue sclera, dentinogenesis imperfecta, scoliosis, ligamentous laxity and easy bruisability (1-3). Left alone, the children suffer multiple fractures due to severe osteoporosis and the cycle of immobilization and fracture can render their life difficult and restrict ambulation.

Various operative options for the fractures are available. The current trend has been use of expandable nails and rods (3-6). The ideal implant should elongate as the child grows, protect the whole bone and prevent subsequent fractures or at least prevent multiple re- operations from displacement, prevent implant failure, migrations and angulation. Ability to be inserted without fluoroscopy may be advantageous in the developing world.

The use of the non-expandable but widely available Titanium Elastic Nails in the treatment of fractures in OI is not widely reported in this continent. We report its use without fluoroscopy as an option for orthopaedic surgeon in the developing world.

Case report

A 9 year old girl with Osteogenesis Imperfecta type 1, presented with a distal third right femur fracture after a minor fall at school. She had had multiple bilateral femur fractures from as early as the age of 1 year which were managed by various non-operative methods including hip spica, traction and plaster casts.

At the age of 6 years, an intramedullary rush rod was inserted in her right femur [Figure.1] after a subtrochanteric fracture, and at the age of 7 the same procedure was done on her left femur after a similar fracture.

She recovered and had a fracture free period of 2 years, during which she was ambulating without walking aids. She had not been on any bisphosphonates. She presented to our practice, at the age of 9 years. Her radiographs revealed severe osteopenia with two intramedullary rush rods in situ in both femurs, with detectable cortical notching by the tips of these implants. She was started on alendronate (MenofosR Giant Pharmaceuticals Limited) 30mg once weekly (1mg/kg for 30kg child) and calcium/vitamin D supplements. Her life in school was normal with only slight limitation of contact activity.

Three months later, she suffered a distal femur fracture at the tip of the rush rod on the right side at the notched cortex (Figure 2). A decision was made to extract the rush rod and replace it with 2.5mm Synthes Titanium Elastic Nails via two portals, one proximal at the greater trochanter, and the other distal, just proximal to the growth plate. The nails were measured off the radiographs. No imaging was available and so the fracture site was accessed through a 2cm incision to afford reduction. The nails were then inserted, one antegrade, crossing the centre of the distal growth plate, and the other retrograde, advanced into the neck. The nails were then cut and bent over the cortex to use the cortex to pull them apart as

the child grows. The decision not to violate the distal femur growth plate by the retrograde nail was because this was a peripheral entry site, but the antegrade nail was freely advanced into the central part of the growth plate as Bailey and others have already shown that this hardly interferes with growth (7). Violation of a growth plate at a peripheral site usually causes an epiphysiodesis at that site resulting in angular deformation.

The child was mobilised on a walking frame within one week, with partial weight bearing and was back to school in three weeks on crutches. She was ambulating without walking aids 5 weeks after surgery. After 2 months of unaided ambulation, she felt sudden pain in her right upper thigh as she was walking in school, and was brought in for a check. Radiographs revealed an undisplaced mid-shaft fracture of the right femur (Figure 3). She was advised to rest for a week and mobilised on a walking frame and then crutches. The child soon ambulated without walking aids with good progression of fracture healing (Figure 4). After four months of uneventful life, the child reportedly fell while playing and presented with a painful limp on an otherwise full weight bearing limb. Radiographs revealed an adequately splinted proximal shaft fracture of the same femur, and the child was advised to use a single crutch till pain subsided (Figure 5). Within two weeks she was bearing full weight (Figure 6).

Discussion

The generalised osteoporosis in OI renders these children vulnerable to frequent fractures of long bones and progressive deformities. In contrast to normal children whose fractures are often managed non-operatively, surgery is often necessary in the long-term management of Osteogenesis Imperfecta patients (3-6).

Intra-medullary nailing has generally been embraced as the most optimal method of surgical management that reduces fracture incidences and enables early ambulation. Sofield and Millar described multi-level osteotomies and realignment using intramedullary devices (4). The challenges with these surgeries were blood loss, infection and inability of the rods to accommodate growth (4).

Rigid expandable rods were then introduced by Bailey and Dubow to solve the problem, which allowed for longitudinal growth (7). These implants also had associated challenges including disassembly, failure to elongate, bending, migrations and osteotomy-related complications (8-10). Despite these, the outcomes (ambulation and fracture rates) have greatly improved.

The availability of these expandable rods locked or not, has been a major challenge in our part of the world. This has necessitated innovative alternative fixation methods. Titanium Elastic Nails provide a good option in this respect with guaranteed early ambulation, and biomechanical stability in the event of a fracture. Panzica et al have reported on use of TEN nails with additional osteotomy to manage a femoral shaft fracture in a patient with Osteogenesis Imperfecta (11). Imaging was utilised in their case. They allowed full weight bearing quickly and the recovery was uneventful. Their follow up which was up to 7 months, did not report any new fracture and it was not clear from the paper if their patient was also on bisphosphonates. They avoided the epiphyseal line distally and both nails were inserted retrograde.

In our case there was no imaging utilised intra operatively, and the nails were inserted antegrade from the greater trochanter through the distal epiphyseal line and the other retrograde into the neck of femur. The nails were then hooked over the cortex at their respective insertion sites, so that they will pull out differentially with continued growth, just like the hooked rush rods were pulling out distally (they were hooked at the trochanter) as the child grew (Figure 3 and Figure 4).

The postoperative period was uneventful with quick return to full weight bearing. Though she subsequently suffered another fracture proximally, the rods splinted it adequately and she did not require another procedure to deal with the new fracture (Figure 3). We feel that this was due to the strong biomechanical protection afforded by the rods, from metaphysis to metaphysis (12). Understandably, without the aid of fluoroscopy, the shaping of the nails was not as perfect as to offer the widest separation at the fracture site, but they still held the reduction well enough to ambulate fairly quickly. Her re-fracture was probably due to overexerting an already osteoporotic bone, because by this time the bisphosphonate may not have had tremendous effect. She was advised to rest for a week and was ambulant on crutches by the end of the second week. This was remarkably shorter than one would have expected had we needed to re-operate.

In conclusion, use of intramedullary Titanium Elastic Nails to manage femur fractures in osteogenesis imperfecta children gives good biomechanical stability and al-

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Figure 1-Radiograph showing Rush rods in situ and healed subtrochanteric fracture

Figure 2-Fracture at tip of Rush rod

Figure3-TEN in situ

Figure 4-Healed lower third fracture,with undisplaced midshaft fracture

Figure 5-Another fracture well splinted in Proximal femur

Figure 6-child bearing full weight

lows early mobilisation of the patient. These nails also may subsequently offer adequate splintage of any new fractures that may occur, thereby reducing the number of subsequent operations.

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