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Case Report

Humerus nail for tibial reconstruction in adolescent with osteogenesis imperfecta – A rare surgical case report

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ABSTRACT

Osteogenesis Imperfecta (OI) is a rare connective tissue disorder known for excessive bone fragility caused by collagen mutations. The common orthopaedic problems related to bone fragility include frequent multiple bone fractures, progressive deformity of long bones leading to impaired ambulation. Surgical correction of long bone deformities in OI is conventionally done using distraction osteogenesis (illizarov fixator), elastic intramedullary nailing, rigid extramedullary fixation using plates, after osteotomy. Intramedullary fixation appears to be an ideal choice for correction of recurrent deformity in the long bone and the devices used previously include telescoping rods, single or dual non-elongating nails (rush nail, TENS). Recently in a case report of 3 individual patients, a humerus nail has been used to reconstruct femur in adolescents with OI. We report a case of humerus nailing for reconstruction of tibia in an adolescent OI male with excellent outcomes which is first of its kind and not reported previously by any other authors.

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1. Introduction

Osteogenesis imperfecta (OI) is a rare congenital hereditary connective tissue disorder caused by mutations in collagen with the common clinical presentation of excessive bone fragility.¹ Clinical manifestations of OI are wide spectrum and could vary from severe forms in the perinatal period to the subtle forms, which could be hardly identified and comparable to a normal person. However, the common orthopaedic manifestations seen in the OI patients, due to excessive fragility are frequent fractures, progressive deformity of long bones and impaired ambulation.^{2,3} The treatment of OI requires a multidisciplinary approach includes both medical and surgical treatment. Medical management includes use of bisphosphonates therapy to increase bone mineral density and reduce long bone fractures rate and surgical management for fracture fixation

or deformity correction.⁴ Goals of surgical management in OI are to minimize the incidence of fracture, restore the bone axis and avoid any progressive deformity. The recommended surgical implant in OI is a load-sharing intramedullary (IM) device, with the largest diameter which is possible, due to its better biomechanical property in fragile bone over plate construct and avoid plate related complications, such as bony resorption from stress shielding, implant failure, progressive deformity and subsequent fractures at the plate ends. However, the surgical management in OI, especially OI patients in adolescent age group with tibial or femoral fracture or bony deformity, are still problematic due to its particular abnormal bony anatomy (such as short limb, non-anatomical alignment secondary to previous injury/surgery, and narrow and non-linear medullary canal with super physiologic bowing) resulting in difficulty of implant selection which is suitable for medullary canal size and bone length.³ Although there is an advanced surgical system, like telescopic rod, which is

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specifically designed for patients with OI and are available in many different sizes that allow fixation in all age groups but due to unavailability of this implant everywhere, including our country makes surgical management difficult. Moreover, pediatric IM standard traditional devices such as Rush pin/TENS might not be appropriate in these adolescent OI patients due to its small size and inability in providing rotational stability.⁵ Recently, there has been studies which reported that the small IM interlocking nail, such as humerus nail, could be used in fixation of femoral fractures in normal adolescent patients due to some of its advantages. The humerus nail being an IM locking device with small diameter and shorter length than conventional tibial nail which is appropriate for small-sized adolescent tibial anatomy, can be used for tibial fixation in adolescent OI patients.³ We report the first case of humerus nail being used for tibial reconstruction in these patients.

2. Case Report



Fig. 1:

2.1. Surgical technique

The patient was placed supine position under spinal anesthesia and pneumatic tourniquet. Plate was removed through the same old incision scar. Multi-level corrective osteotomy performed with saw and osteotome. Medullary canal was recanalized with the help of drill bits of increasing diameter followed by recanalization with hand reamers. Tibia was reduced with acceptable alignment. The entry point was made at the proximal tibia similar to the regular tibial nailing of adult bone followed by passing of guide wire in the Centro-medullary direction and the medullary canal was gently prepared by hand reaming using 6, 6.5 and 7-mm T-reamers. Then a 6.5-mm diameter Interlocking humeral nail (Sharma) with 240-mm length was inserted followed by proximal and distal locking screw insertion under fluoroscopic guidance. The bone gaps created due to the cortical splits during reaming were filled with bone grafts obtained from the anterior closing wedge osteotomy site. No additional wiring or implantation was performed. The operative time was 2 hours with minimal blood loss, and the total length of hospital stay was 4 days.

2.2. Postoperative Care and Rehabilitation

Sutures removed on the 15th day post op. Patient was allowed for knee and ankle ROM exercises and wheel chair mobilization for the first 6 weeks followed by weight bearing as tolerated. The proximal and distal osteotomy sites both showed clinical and radiological union in progress at 6 weeks follow up.

3. Discussion

OI is heritable disorder of collagen synthesis that commonly presents as bone fragility with multiple fractures and deformities. These bony problems usually require surgical management like fracture fixation or reconstruction by a load-sharing IM device.⁶ However, the surgical fixation in OI patients, especially in the older children with tibial fracture or deformity, is very difficult due to the abnormal anatomy of tibia resulting in problems with implant selection such as a mismatch with conventional tibial IM nail, the poor fixation stability of standard pediatric IM devices, and the high cost and unavailability of advanced telescopic rod. This study aimed to present the usefulness of humeral nail fixation as a surgical tool for reconstruction of tibia in adolescent OI patients. The humeral nail in tibial fixation for adolescent OI patients has many advantages. Firstly, humeral nail compared to conventional tibial IM nail, is available in smaller diameter and shorter length is more suitable with these patients tibial anatomy with narrow medullary canal and short limb. Secondly, humeral nail has narrow width and has a lateral bend at its proximal end which allows its insertion with minimal physeal violation, and 90-100 degrees of the cephalomedullary angle for

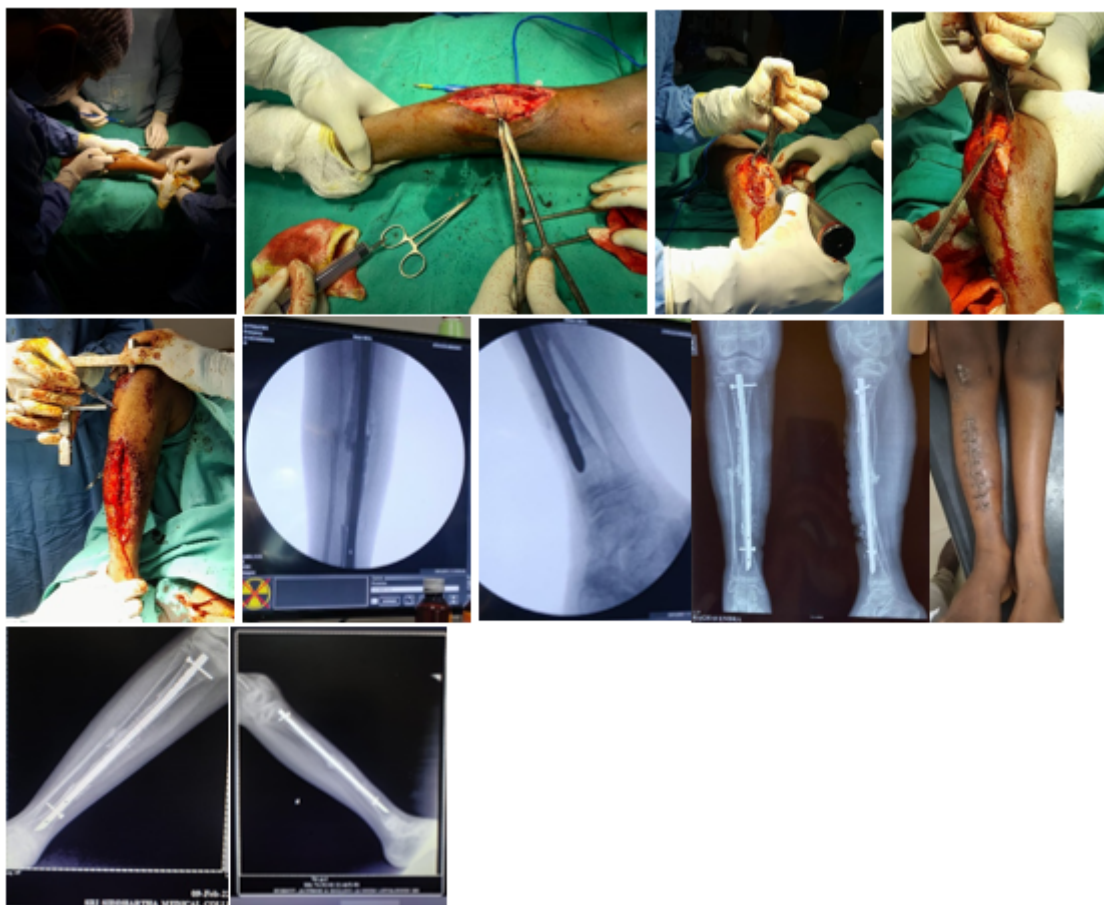


Fig. 2: 6 weeks follow up x-rays showing fracture union in progress

proximal locking blade or screw. Thirdly, because of the interlocking property of nail, the humeral nail would offer superior biomechanical benefits than gold standard Rush pin fixation as it provides rotational stability and leg length control, especially for those patients with multilevel corrective osteotomy. However, there were also limitations for using humeral nail for tibial reconstruction in these patients but are minimal, like proximal tibial physeal damage cannot be avoided which predisposes for future growth disturbances of proximal tibia. Absence of Herzog's bend limits the nail entry to a more proximal and posterior area of tibial articular surface. The results of this study showed that the use of humeral nail in tibial fixation was possible and could be used in proximal, middle or distal fractures. Moreover, this feature can be used for surgical fixation of fracture and non-union, or in corrective osteotomy. This study also demonstrated the favourable outcome with 100% fracture healing and without the implant-related complications such as infection, non-union, or AVN. Therefore, we concluded that tibial fixation with humeral nail in adolescent OI patients is one of the possible options with satisfactory outcomes.

4. Source of Funding

None.

5. Conflict of Interest

None.

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