

Severe Femur Angular Deformity in Osteogenesis Imperfecta – a case report

Mohd Zaharial, MZ

Orthopaedic Department, Hospital Universiti Sains Malaysia

INTRODUCTION:

Osteogenesis imperfecta (OI) is a hereditary disease of connective tissue characterized by the loss of bone density and mass that increases the fragility of the bones. Patient usually manifests as multiple fractures and bony deformities. One of the challenge in managing fractures in OI is the complexity of brittle bone and angular deformity, with the aim of getting better alignment, improving the mobility while providing internal support for the bone.

REPORT:

A case of 8 years old boy, presented with severe deformity of the left thigh. He had history of fall in 2021 and diagnosed with closed fracture femur. Patient subsequently underwent open reduction and plating of the left femur which was complicated with peri implant fracture. Removal of implant was done, and then treated non operatively. Consequent visit showed severe deformity of the femur while the fracture united. Patient was also given 2 course of iv pamidronate during clinic follow up. On examination, clinically there is severe varus deformity of the thigh with significant limb leg discrepancy. This is evident by 90° angulation deformity of the proximal 1/3rd of the femur on xray. He then underwent operation of multiple osteotomy and intramedullary insertion and k-wiring of left femur. Post operatively he was put on derotational bar which planned to be kept until fracture heal to prevent rotational instability.



Figure 1: Preop femur xray



Figure 2: Post op femur xray

CONCLUSION :

In dealing with osteogenesis imperfecta patient, there will always be a challenge to correct the deformity while dealing with the bone quality. An adequate pre operative evaluation and thorough surgical planning are important to get the best alignment without compromising the nerve and vessels, besides deciding in the implant to provide stability to the bone.

REFERENCES :

1. Morales et al, Cureus, January 2023.
2. Cho et al, Clinics in Orthopedic Surgery Vol 12, No 4, 2020