International Journal of Science and Research (IJSR)
ISSN: 2319-7064
Impact Factor 2024: 7.101

Anaesthetic Management in a Patient with Osteogenesis Imperfecta for Left Femur Nailing

Dr Uthkala B Hegde, Dr Janaki Vijayan

[1]Corresponding Author

Abstract: ***Osteogenesis imperfecta (OI) is an inherited connective tissue disorder primarily affecting the bones, sclera, and inner ear, resulting from a mutation in the type I collagen gene. According to the Sillence classification, OI is categorized into four types: Type I: Inherited in an autosomal dominant (AD) pattern, typically characterized by mild bone fragility. Type II: Lethal either in utero or during the perinatal period. Types III & IV: Both are inherited in an autosomal dominant manner, with more severe manifestations than Type I. The skeletal abnormalities in OI arise from defective ossification of endochondral bone, leading to bone fragility, hypermobile joints, and kyphoscoliosis. The disorder may also involve multisystem complications, including cardiac valvular lesions, cor-pulmonale, neurologic abnormalities, hyperhidrosis, cleft palate, metabolic derangements, susceptibility to malignant and non-malignant hyperthermia, obstructive uropathy secondary to renal and ureteric calculi, and platelet dysfunction.***

Keywords: Bone fragility, collagen mutation, autosomal dominant, osteogenesis imperfecta, skeletal abnormalities, multisystem complications.

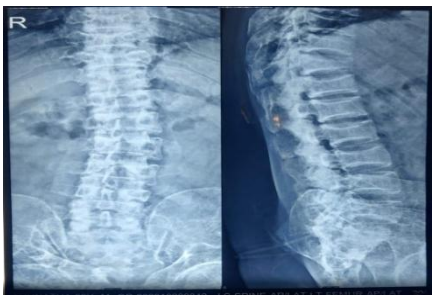
1. Case Report

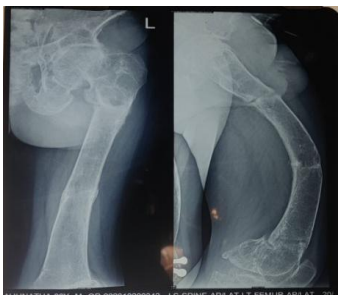
A 38-year-old male presented with complaints of pain over the shaft of the left femur for the past three months. The patient is a known case of osteogenesis imperfecta, characterized by short stature, brittle bones, hypermobile joints, and kyphoscoliosis. He has a history of recurrent fractures of long bones, for which he underwent two uneventful surgical interventions under subarachnoid block.

On general examination, the patient was short-statured (130 cm) and weighed 40 kg. He was afebrile with normally colored sclera. Vital signs, including pulse and blood pressure, were within normal limits. Respiratory examination revealed a barrel-shaped chest with bilateral equal air entry. Airway assessment showed acceptable flexion and extension at the neck, adequate mouth opening, and absent dentition, classified as Mallampati Class 2. Kyphoscoliosis was noted on spine examination. Other systemic examinations were unremarkable.

Routine blood investigations, including coagulation profile, were normal. Electrocardiogram (ECG) findings were normal. Chest X-ray revealed thin, gracile ribs with a normal cardiac silhouette. X-ray of the spine showed dorsolumbar kyphoscoliosis. The patient was assessed as ASA Grade 2 and scheduled for left femur nailing.

In the operating room, the patient was connected to all standard ASA monitors. Lumbar puncture was performed using a paramedian approach, and 2.6 ml of 0.5% Bupivacaine with 30 mcg of Buprenorphine was administered. Sensory block was achieved up to the T8 level. The patient remained hemodynamically stable throughout the surgery.



2. Discussion

The best anaesthetic technique in this patient is regional block. As it avoids the need for laryngoscopy, tracheal intubation associated risk of odontoaxial dislocation, fracture mandible, cervical vertebral and teeth injury.1 Conduction block makes the development of hyperthermia less likely as compared to GA. In our case the anatomical deformity was much severe than physiologic abnormality. Presence of dorso – lumbar kyphoscoliosis made the regional anaesthesia difficult. Kyphoscoliosis can predispose these patients to inadvertent dural puncture and coupled with short stature, made it difficult to predict the level of block produced.

3. Conclusion

Regional anaesthesia is the technique of choice in such cases, but when general anaesthesia is considered in view of proposed surgical procedure or due to relative contra-indication of regional block, meticulous attention is required especially with the use of neuromuscular blocking agents, inhalational agents, airway management, patient positioning and acute pain management.

References

[1] Bhandari G, Shahi KS, Bhadoria P, Bhalotra AR, Sandhya OD, Arya M. Osteogenesis imperfect: No place for imperfect anaesthesiologist. Indian J Anaesth.
[2] Bhardwaj M, Kaur K, Johar S, Ruchi S, Hooda S. Anaesthetic management in a patient with osteogenesis imperfecta and a fractured femur. South Afr J Anaesth Analg.