

# Anesthetic Management of Femur Fracture in a Pediatric Patient with Osteogenesis Imperfecta

Ayushi Patel<sup>1</sup>, Aniket R Aghara<sup>2</sup>, Kamla Mehta<sup>1</sup>, Saurin Panchal<sup>1</sup>, Vismit Gami<sup>1</sup> and Dev Desai<sup>1\*</sup>

<sup>1</sup>Department of Anesthesiology, SMT. SCL General Hospital, Smt. NHL municipal Medical College, India

<sup>2</sup>Department of Anesthesiology, GMERS Morbi, India

## \*Corresponding Author

Dev Desai, Department of Anesthesiology, SMT. SCL General Hospital, Smt. NHL Municipal Medical College, India.

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## Abstract

*Osteogenesis Imperfecta (OI) presents challenges in anesthesia due to fragile bones, risk of cervical spine instability, and potential for difficult airway management. We present the case of a 7-year-old male with OI Type I undergoing open reduction internal fixation of a right subtrochanteric femur fracture. Despite anticipated airway difficulties and musculoskeletal abnormalities, meticulous planning and vigilant anesthesia management ensured a successful outcome.*

**Keywords:** Osteogenesis Imperfecta, Pediatric Anesthesia, Femur Fracture, Anesthetic Management, Perioperative Care

## 1. Introduction

Osteogenesis Imperfecta (OI), commonly known as brittle bone disease, is a rare genetic disorder characterized by fragile bones, often resulting in frequent fractures with minimal trauma. This condition, caused by mutations in the collagen type I genes, presents a spectrum of severity ranging from mild to severe forms, each with its unique clinical manifestations [1]. OI has a birth prevalence of approximately 6–7/100,000 [Steiner et al., 1993] [2]. The discovery of the first gene associated with recessive osteogenesis imperfecta occurred in 2006 [3].

It is the leading genetic cause of bone fragility, occurring in approximately 1 out of every 15,000 to 20,000 births [4].

The higher bone fragility makes the vertebrae and extremities more susceptible to fractures, whereas fractures of the facial bones are rare [5].

In osteogenesis imperfecta (OI) patients and their parents, platelet function was frequently abnormal, with impaired PF3 release and elevated serum pyrophosphate (PPi) levels observed in many subjects. Despite these abnormalities, thrombin-induced platelet aggregation remained normal [6].

The SILENCE classification system categorizes Osteogenesis Imperfecta (OI) into four distinct types, each with its unique clinical features and inheritance patterns [7].

1. Type I: This is the most common form of OI and is inherited in an autosomal dominant fashion. It is characterized by blue sclera (the white part of the eye), fragile bones prone to fractures, hyperextensible joints, progressive hearing loss, and a dental condition known as dentinogenetic imperfecta.

2. Type II: OI Type II is the most severe form and is usually lethal either in utero (before birth) or during the perinatal period (around the time of birth). Infants with Type II OI typically have extremely fragile bones and may experience multiple fractures even before birth.

3. Type III: Also inherited in an autosomal dominant manner, Type III OI is characterized by severe bone fragility, skeletal deformities, and a tendency to develop fractures. Patients with Type III OI often experience a shortened lifespan, typically dying in childhood or adolescence due to complications such as cardiopulmonary issues.

4. Type IV: Similar to Type III, Type IV OI is inherited in an autosomal dominant fashion. However, it is distinguished by skeletal deformities without accompanying ocular (related to the eyes), audiological (related to hearing), or dental abnormalities. Despite the skeletal issues, individuals with Type IV OI generally

have a normal lifespan.

Patients with OI are particularly susceptible to fractures during routine medical procedures, including shifting or positioning for anesthesia administration. Additionally, anatomical features such as a short neck and restricted mouth opening pose difficulties in airway management, necessitating careful consideration during intubation.

Moreover, individuals with OI are vulnerable to complications such as malignant hyperthermia, attributed to muscle atrophy, which necessitates avoiding triggers like succinylcholine. The presence of kyphoscoliosis and disc prolapse further complicates the administration of regional anesthesia, often making general anesthesia the preferred choice.

## 2. Case Presentation

A 7-year-old male, known case of OI Type I, presented with a fracture of the right subtrochanteric femur. His medical history included recurrent long bone fractures, lumbar disc prolapses, and musculoskeletal deformities. Pre-anesthetic evaluation revealed short stature, fragile bones, kyphoscoliosis, and Mallampati Class III airway. Considering the risk of cervical spine instability, manual in-line stabilization was ensured during airway management. Difficult intubation cart was prepared, and after preoxygenation, induction was performed with fentanyl and propofol. Endotracheal intubation was challenging due to limited mouth opening and musculoskeletal abnormalities, but successful with a size 5 cuffed tube. Anticipating vulnerability to malignant hyperthermia, succinylcholine was avoided.



## 3. Anesthesia Management

This included preparation of a difficult intubation cart with

necessary equipment. Essential monitors for NIBP, SPO<sub>2</sub>, and ECG were attached and continuously monitored. Induction of general anesthesia was achieved with fentanyl and propofol, followed by mask ventilation and administration of atracurium. Despite difficulties in inserting the laryngoscope blade due to cervical spine stabilization, endotracheal intubation was successfully performed orally using a cuffed tube. The airway was confirmed, and the tube was secured.

## 4. Discussion

Osteogenesis Imperfecta (OI) presents significant challenges in the perioperative management of patients, especially in pediatric cases requiring surgical intervention. This discussion highlights the complexities and considerations essential for anesthetic management in a 7-year-old male with OI Type I undergoing open reduction internal fixation for a right subtrochanteric femur fracture.

### 4.1 Airway Management

Airway management in patients with OI is particularly challenging due to musculoskeletal abnormalities such as short neck, limited mouth opening, and potential cervical spine instability. In this case, the pre-anesthetic evaluation revealed a Mallampati Class III airway, indicating a potential for difficult intubation. Manual in-line stabilization of the cervical spine was essential to prevent atlantoaxial dislocation, a common risk in OI patients. The preparation of a difficult intubation cart, including video-laryngoscopy, provided alternative options to ensure successful airway management. Despite the challenging anatomy, endotracheal intubation was accomplished using a size 5 cuffed tube, demonstrating the importance of preparedness and skill in managing difficult airways.

### 4.2 Anesthesia Induction and Maintenance

The induction of anesthesia in OI patients requires careful selection of agents to mitigate risks associated with the condition. In this case, fentanyl and propofol were chosen for induction, avoiding succinylcholine due to the risk of malignant hyperthermia, a potential complication in OI attributed to muscle atrophy and other underlying abnormalities. Atracurium was used for muscle relaxation, providing a safer alternative. Continuous monitoring with non-invasive blood pressure (NIBP), pulse oximetry (SPO<sub>2</sub>), and electrocardiography (ECG) ensured real-time assessment of the patient's physiological status throughout the procedure.

### 4.3 Surgical Considerations

The surgical intervention for a subtrochanteric femur fracture in an OI patient is inherently complex due to the fragility of the bones and the high risk of intraoperative fractures. This necessitates meticulous handling and precise surgical techniques. In this case, the orthopedic team was prepared for potential complications, and the collaboration with the anesthetic team ensured that any intraoperative challenges could be managed promptly and effectively.

#### 4.4 Multidisciplinary Approach

The successful management of this case underscores the critical role of a multidisciplinary team approach. Anesthetists, surgeons, and other healthcare professionals must collaborate closely, each bringing their expertise to address the unique challenges presented by OI. The anesthetists' familiarity with the nuances of OI, including the risks of cervical spine instability and malignant hyperthermia, was crucial in formulating an effective anesthetic plan. The orthopedic surgeons' expertise in handling fragile bones minimized the risk of additional fractures and ensured the best possible surgical outcome.

#### 5. Postoperative Care

Postoperative care for OI patients involves monitoring for complications such as respiratory distress, which can result from kyphoscoliosis and other musculoskeletal deformities. Pain management is also a critical aspect, requiring a balance between effective analgesia and minimizing the risk of respiratory depression. In this case, the anesthetic team provided appropriate pain control while ensuring continuous monitoring to detect any early signs of postoperative complications.

#### 6. Conclusion

The anesthetic management of pediatric patients with Osteogenesis Imperfecta (OI), particularly during orthopedic surgeries, demands a highly specialized approach due to the unique challenges posed by the condition. In this case of a 7-year-old male with OI Type I undergoing open reduction internal fixation of a right subtrochanteric femur fracture, meticulous preoperative evaluation and planning were paramount. Key considerations included the risk of cervical spine instability, potential for difficult airway management, and the need to avoid triggers for malignant hyperthermia. The use of manual in-line stabilization during intubation, preparation of a difficult airway cart, and avoidance

of succinylcholine contributed to the successful management of this complex case. The collaboration of a multidisciplinary team, including anesthetists familiar with the nuances of OI, ensured a favorable outcome. This case underscores the importance of individualized anesthetic strategies to mitigate the inherent risks and enhance the safety of pediatric patients with OI undergoing surgery.

#### References

1. Gupta, D., & Purohit, A. (2016). Anesthetic management in a patient with osteogenesis imperfecta for rush nail removal in femur. *Anesthesia Essays and Researches*, 10(3), 677-679.
2. van Dijk, F. S., Cobben, J. M., Kariminejad, A., Maugeri, A., Nikkels, P. G., van Rijn, R. R., & Pals, G. (2011). Osteogenesis imperfecta: a review with clinical examples. *Molecular syndromology*, 2(1), 1-20.
3. Etc MCS. Myocardial extraction from neonatal rats. HHS Public Access. *Physiol Behav*. 2019;176(3):139-48.
4. Masroor, T., Chappell, A. G., & Shahzad, F. (2022). Anesthetic and Surgical Considerations in Osteogenesis Imperfecta: A Case Report of Mandible Fracture Management. *Eplasty*, 22.
5. Al-Osaimi, A., Samman, M., Al-Shakhs, M., Al-Suhaim, F., & Ramalingam, S. (2014). An unusual case of atrophic mandible fracture in a patient with osteogenesis imperfecta and on oral bisphosphonate therapy: Case report. *The Saudi dental journal*, 26(2), 68-73.
6. Hathaway, W. E., Solomons, C. C., & Ott, J. E. (1972). Platelet function and pyrophosphates in osteogenesis imperfecta. *Blood*, 39(4), 500-509.
7. Górnik-Właszczuk, E., Majewski, J., Szczygieł, R., & Dusiel, A. (2013). Anaesthesia in children with osteogenesis imperfecta—report of 14 general anaesthetics in three children. *Anaesthesiology Intensive Therapy*, 45(2), 85-88.

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