

Total Intravenous Anesthesia in a Child with Osteogenesis Imperfecta for Femoral Fracture Fixation: A Case Report

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ABSTRACT:

Osteogenesis imperfecta (OI) is a genetic collagen disorder characterized by extreme skeletal fragility, complicating perioperative care for pediatric patients with any fractures. Systemic risks include restrictive lung disease, coagulation abnormalities, and predisposition to hypermetabolism and hyperthermia, along with airway challenges from cervical instability. A seven-year-old girl with a femoral shaft fracture was scheduled for titanium elastic nailing. She had multiple prior fractures from trivial trauma, a family history of OI, blue sclerae, and dental caries. After careful premedication, padding, and warming, total intravenous anesthesia was given with propofol-enabled gentle intubation and positioning. Surgery proceeded uneventfully, with stable hemodynamics and no hyperthermia. Multimodal analgesia facilitated smooth recovery. Multidisciplinary planning, meticulous handling, and avoidance of volatile agents ensured safe outcomes in this fragile patient.

Keywords: Hyperthermia; Hypermetabolism; Osteogenesis Imperfecta; Total Intravenous Anesthesia.

INTRODUCTION

Osteogenesis imperfecta (OI) is a genetic collagen disorder characterized by extreme skeletal fragility, which complicates the perioperative care of pediatric patients presenting with traumatic multiple fractures.¹ The condition necessitates a multidisciplinary approach due to systemic risks, including restrictive lung disease, potential coagulation

disturbances, and a heightened predisposition for hypermetabolism and hyperthermia.² Furthermore, the structural anomalies inherent to this connective tissue disorder, such as cervical spine instability and narrow airway architecture, frequently necessitate specialized protocols for tracheal intubation to avoid iatrogenic trauma.

CASE REPORT

A seven-year-old girl arrived at our hospital with a history of a fracture in the right femoral shaft and was scheduled for Titanium Elastic Nailing (TENS). She underwent a comprehensive evaluation at the pre-anesthetic clinic two days ahead of the surgery. Upon taking a detailed medical history, it was discovered that she had suffered five previous long bone fractures, which were treated accordingly, with the recent fracture resulting from trivial trauma while playing at home. Her family history was positive for features of Osteogenesis Imperfecta (OI) in her father and one sibling. Clinical observations showed blue sclera (Fig. 1) and numerous dental caries, which are indicative of having multiple fractures from minimal trauma. All her laboratory tests, including Complete Blood

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Fig 1. Bilateral Blue sclera present in child

Count, coagulation profile, serum calcium levels, and renal function assessments, were normal. Serum Alkaline Phosphatase levels were slightly elevated. An echocardiogram was performed, revealing findings of levocardia with situs solitus.

During the airway assessment, the patient was classified as Mallampati class I (**Fig. 2**), exhibiting normal neck mobility, with no anticipated difficulties in intubation. However, contingency plans, including the availability of a video laryngoscope and smaller-sized endotracheal tubes, were prepared due to the potential cervical spine fragility associated with OI. Upon her arrival in the preoperative area, the patient displayed signs of anxiety and was administered intravenous midazolam at a dosage of 0.5 mg/kg, 45 minutes before the transfer to the operating room, to mitigate anxiety and facilitate a smooth induction process.

The operating room was meticulously prepared, ensuring that the operative table was well-padded with gel pads and cotton rolls at all pressure points. Warming devices, special forced-air warmers, and a room temperature of 24 degrees Celsius were positioned to maintain normothermia, as patients with OI may experience impaired thermoregulation. All personnel involved were informed regarding the necessity of gentle handling to prevent iatrogenic fractures. The patient's physical status was classified



Fig 2. Mallampati grading with protruding tongue

as ASA II, and informed written consent was secured from her parents, outlining the risks of fractures during positioning, airway management, and the potential for increased bleeding.

Upon arrival in the operating room, standard monitoring equipment was applied, including a five-lead ECG, non-invasive blood pressure monitor, pulse oximeter, capnography, and a temperature probe. Intravenous access was established cautiously in the left upper limb with a 22 G cannula, minimizing tourniquet pressure. Pre-oxygenation was performed using 100% oxygen for three minutes. Induction was conducted with intravenous propofol (2.5 mg/kg, administered through slow titration) and fentanyl (two µg/kg). Muscle relaxation was achieved with atracurium (0.5 mg/kg).

Tracheal intubation was performed using a 4.5 mm cuffed endotracheal tube via direct laryngoscopy, employing gentle jaw thrust and minimal neck extension techniques while avoiding cricoid pressure. Total intravenous anesthesia (TIVA) was maintained using a propofol infusion at a rate of 100–150 µg/kg/min, with an oxygen-air mixture delivering a fractional inspired oxygen concentration (FiO₂) of 40–50%. Intermittent boluses of atracurium (0.1–0.15 mg/kg) and fentanyl (0.5–1 µg/kg) were provided as required for analgesia and relaxation.

Ventilation was managed using a volume-controlled strategy with tidal volumes set at 6–8 mL/kg and the respiratory rate adjusted to maintain end-tidal CO₂ within the range of 35–40 mmHg. Special attention was devoted to positioning the patient. Prophylactic intravenous antibiotics (cefuroxime at a dosage of 30 mg/kg) were administered 30 minutes before incision. The surgical procedure comprised closed reduction under image intensifier guidance, followed by the retrograde insertion of two titanium elastic nails (TENS) through the distal femoral metaphysis. Intraoperative fluoroscopy exposure time was kept to a minimum. The total duration of the surgical procedure was 75 minutes, with negligible blood loss recorded at less than 50 mL, while hemodynamic parameters remained stable throughout (heart rate ranging from 90 to 110 bpm, blood pressure between 90–110/50–60 mmHg, and oxygen saturation ranging from 99 to 100%). Normothermia was successfully maintained, with core temperatures between 36.5°C and 37.2°C. There were no occurrences of hypertension, tachycardia, hyperthermia, or hypermetabolism.

Post-surgery, the child was reversed with

neostigmine (0.05 mg/kg) and glycopyrrolate (0.01 mg/kg). Extubation was performed when the patient was fully awake, obeying commands, exhibiting adequate muscle strength, and demonstrating stable vital signs. Postoperative pain management was implemented using a multimodal approach tailored to the child's age and bone fragility associated with OI. The patient was monitored in the Post-Anesthesia Care Unit (PACU) for a duration of two hours, with pain assessed using the FLACC (Face, Legs, Activity, Cry, Consolability) scale. Intravenous paracetamol (15 mg/kg) was administered every six hours, with the first dose provided intraoperatively

DISCUSSION

Meckel's diverticulum is the most common congenital OI posing significant anesthetic challenges, including skeletal fragility, difficult or traumatic airway management, cervical spine instability, respiratory compromise, bleeding tendency, and increased blood loss.³ TIVA with propofol and opioid infusion was used for maintenance to avoid volatile anesthetic agents.⁴ To minimize fracture risk, the patient was transferred and positioned gently with adequate padding. An LMA was used to avoid forceful laryngoscopy

and excessive neck manipulation.⁵ Respiratory status was closely monitored with pulse oximetry and capnography. Blood loss was anticipated, monitored, and appropriate blood products were kept available. Multimodal analgesia, including regional analgesia, was used to reduce opioid requirements and postoperative respiratory depression.⁶ Careful preparation for potential significant blood loss and coordinated multidisciplinary management are essential for safe perioperative care.⁷

CONCLUSION

Effective perioperative management of pediatric OI involves thorough preoperative evaluation, cautious handling to avoid iatrogenic fractures, precise airway management, and total intravenous anesthesia to reduce hypermetabolism and hyperthermia. Utilizing multimodal analgesia and a well-coordinated multidisciplinary strategy guarantees hemodynamic stability, effective pain relief, and a safe recovery, as illustrated in this case of uncomplicated TENS.

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