Anesthetic Management of a Parturient with Osteogenesis Imperfecta Type 1 for Cesarean Delivery

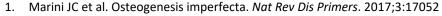


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Background

- Osteogenesis Imperfecta Type 1 (OI-1) is a rare autosomal dominant connective tissue disease characterized by normal but insufficient collagen production, resulting in fracture-prone bones, scoliosis, and mild short stature.¹
- Parturients with OI present several anesthetic concerns:²
 - Skeletal anomalies (difficult neuraxial block)
 - Potential difficult airways & risk of dental damage
 - Potential coagulopathy/PPH

- Fractures w/ extremity manipulation (positioning, tourniquets)
- Decreased pulmonary reserve (restrictive pattern)
- Hearing loss
- Ideal mode of delivery remains elusive, though C-sections are common due to the risk of maternal pelvic fractures during vaginal delivery.



2. Yan M et al. The Anesthetic Management of a Parturient With Osteogenesis Imperfecta Type I Undergoing Cesarean Delivery. *Cureus*. 2021;13(3):e13849



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Case Description

- A 24-year-old lady, 155cm and 61kg, G1P0 at 36w3d gestation presented with an intractable headache and mildly elevated BP.
- Medical History: OI-1 with pathologic vertebral fractures at T2-3, L1, L5, dextroscoliosis, migraines, and easy bruising.
- Denied any hearing impairment, neck immobility, dental involvement, or cardiopulmonary symptoms.

Plan: Elective 1º C-section for pre-eclampsia with severe features.
 Received an L3-L4 spinal block with 1.5mL 0.75% bupivacaine, 0.15mg morphine, and 15mcg fentanyl → Achieved a T4 sensory level.

Considerations:

- Careful positioning using adequate padding.
- NIBP cuff cycle set to every 5 mins. [A-line prepared in case of HD instability]
- Surgery was complicated by PPH with an EBL of 1.3L, managed supportively.
- Received a 24-hour magnesium infusion for seizure-prophylaxis, and reported resolution of her headache after delivery.
- POD 3: Complained of unilateral rib pain CXR negative for fractures.
- POD 4: Discharged in stable condition.



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Learning Points

- Comprehensive approach when managing OI-1 pregnancies.
- Neuraxial Anesthesia:
 - Avoids airway & cervical spine manipulation.
 - Reduces risk of maternal aspiration.
 - Minimizes neonatal depression due to anesthetics.
- In case spine deformities preclude neuraxial anesthesia and **General Anesthesia** is required, multiple points must be kept in mind:
 - 1- Hyperextension of the cervical spine can lead to atlantoaxial dislocation or fractures.
 - 2- Depolarizing neuromuscular blockers should be avoided for risk of contraction-induced fractures.
 - 3- Dentinogenesis imperfecta may be present in OI-1, with increased risk of tooth damage or loss during intubation.

