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.

1. Marini JC et al. Osteogenesis imperfecta. *Nat Rev Dis Primers*. 2017;3:17052
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.