

Schwartz N

Shriners' Hospital for Children, Philadelphia , Philadelphia , Pennsylvan, USA

CASE PRESENTATION

8-year-old, 21 kg male presents with history of grade III OI status post femoral fractures following a fall. He is scheduled for intramedullary rodding of bilateral femoral fractures and spica casting.

PMH: OI grade III, history of 61 prior fractures requiring 20 surgeries. Meds: Morphine 1 mg Q2prn, Oxycodone 2mg Q4prn, Zofran 4mg IV Q6prn, Physical examination and airway assessment were otherwise adequate. NPO for 8 hours.

Anesthetic management

Inhalational induction. Blood pressure monitored by sphygmomanometer.

Endotracheal intubation accomplished easily. Right radial arterial line inserted for intra-arterial pressure monitoring.

Lumbar epidural catheter inserted under fluoroscopic guidance with catheter tip confirmed with fluoroscopy and contrast to L1 midline level.

– Inhalational anesthesia maintained with sevoflurane. Epidural Infusion with ropivacaine. Dexmedetomidine was started one hour prior to the completion of surgery.

Uneventful emergence and early post-anesthetic care. Patient extubated without difficulty and transferred to PICU. He remained pain free for 3 days with epidural analgesia supplemented by morphine PCA.

Osteogenesis imperfecta (Lobstein disease, brittle-bone disease, blue-sclera syndrome) is a genetic bone disorder characterized by a deficiency of Type-I collagen arising from an aminoacid substitution of glycine in the collagen triple helix structure. OI affects an estimated 6 to 7 per 100,000 people . Types I and IV are the most common forms, affecting 4 to 5 per 100,000 people.

OI is classified into types I – VIII. OI Type I is the mildest and most common form of the disorder accounting for 50 percent of the total OI population. Type II is the most severe form. Most are stillborn or die shortly after birth, usually from respiratory failure. Type III is the most severe type among children who survive the neonatal period.

At birth, infants generally have mildly shortened and bowed limbs, small chests, and a soft calvarium and multiple long-bone fractures at birth, including many rib fractures. The presentation is for frequent fractures of the long bones and bowing and progressive malformation. Children have a markedly short stature. The sclerae may be tinted blue and they may have dentinogenesis imperfecta.

ANESTHETIC CONSIDERATIONS

- Specific issues related to pre-existing fractures or deformities
- Increased risk of intraoperative fractures
- BP Monitoring- A-line vs BP cuff
- Positioning - Pad all pressure points
- Difficult intubation-
- Direct laryngoscopy may cause mandibular or cervical fracture.
- Visualization of vocal cords may be difficult.
- Consider fiberoptic aid
- Dentition may be defective or prone to trauma during laryngoscopy
- Use of Succinylcholine with care
- Fasciculations may cause fractures
- Regional anesthesia may be beneficial
- Respiratory
- Kyphoscoliosis and pectus excavatum
- Decreased Vital Capacity and chest wall compliance
- Can result in hypoxemia and V/Q mismatch
- Regional Anesthesia

- Coagulation studies recommended
 - Prolonged bleeding times even with normal platelet count
 - Consider Desmopressin / DDAVP
 - Platelet dysfunction
 - Hyperthermia
 - Hyperhidrosis
 - Increased serum thyroxine levels
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