Anesthetic Considerations for Pediatric Patients with Osteogenesis Imperfecta

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Learning Objectives:

• Describe preoperative considerations in managing pediatric patients with Osteogenesis Imperfecta (OI)
• Formulate an appropriate anesthetic plan for patients with OI
• Avoid any further fractures in OI patients peri-operatively
Pathophysiology

• “Brittle bone disease”
• Estimated at 1 in 13,500
• Genetic disorder of connective tissue
• Most OI has mutation in genes that code for collagen type I alpha chains, \textit{COL1A1} and \textit{COL1A2}
• Collagen is the main component of organic bone matrix and a key ingredient in bone tissue integrity
Pathophysiology

• Abnormal collagen synthesis leads to bone fragility and other skeletal deformities

• Multiple subtypes
  - Type I-most common, mildest
  - Type II-most severe/lethal
  - Type III-severe, but survivable
  - Type IV-severity between type I and III
  - Many other subtypes with similar clinical presentation as above
Clinical Features

- Bone fragility
- Blue or grey discoloration of the sclera
- Dental discoloration, tooth fracture, and attrition
- Dysplasia of the mandible and maxilla
- Decreased muscle mass
- Rarely associated with posterior fossa compression, Chiari malformation, spinal cord syrinx formation and hydrocephalus
Diagnosis

• Molecular tests analyze structure and quantity of type I collagen synthesis

• Definite diagnosis is by clinical traits
Common Surgeries Performed

- Orthopedic surgeries for fracture repair
- Intramedullary rod implant to improve function and ambulation
- Deformity corrections
- Telescopic rods for vertical height gain
Preoperative Evaluation

• Airway evaluation: assess OI severity
• Excessive neck extension may lead to atlantoaxial subluxation and dislocation
• Reasons for potentially difficult airway
  • Brittle bones
  • C-spine instability
  • Short neck
  • Edentulous
Preoperative Evaluation

- Airway techniques
  - Direct laryngoscopy: avoid in patients with C-spine instability and consider videolaryngoscope or fiberoptic bronchoscope for intubation
  - Mask: avoid excessive force
  - LMA: least invasive way to ventilate, and a great rescue airway device

- Avoid further fractures!
Preoperative Evaluation

- Pulmonary Evaluation
  - Restrictive lung disease
  - Kyphoscoliosis
  - Pulmonary hypertension
  - Cor pulmonale

- Cardiac Evaluation
  - Premature atherosclerosis
  - Valvular disease
Preoperative Evaluation

Hematologic Evaluation

• Poor collagen function on platelet-endothelial cell may lead to bleeding diathesis

• Platelet dysfunction from capillary fragility (qualitative, not quantitative)

• Coagulation test to rule out platelet dysfunction for surgery and regional anesthesia
Intraoperative Implications

Anything can cause iatrogenic fractures!

- PIV placement
- Tourniquets placement
- Blood pressure cuff
- Surgical position
- Transport
- Careful padding needed
Intraoperative Implications

Succinylcholine

• Hyperkalemic arrhythmia in immobile patients
• Succinylcholine induced fasciculations can cause fractures!
• Twitch monitor can cause fractures!
Intraoperative Implications

Hypermetabolic state

• Can be triggered by atropine/glycopyrrolate
• Symptoms: excessive diaphoresis, hyperthermia, tachycardia, and tachypnea
• Avoid routine use of warming devices without monitoring temperature
• This hyperpyrexia reaction not associated with MH (malignant hyperthermia)
Brittle Bone Myths

Recent study in the Journal of Pediatric Orthopedics

- 37 OI patients underwent 96 orthopedic procedures
- NIBP cuffs used in 81 surgeries
- Tourniquets applied to lower extremity at a pneumatic pressure of 250 mm Hg in all patients
- No iatrogenic fractures with NIBP cuff use
- No fractures associated with tourniquet use
- One patient had a humerus fracture from preoperative patient positioning


Incidence of Fractures From Perioperative Blood Pressure Cuff Use, Tourniquet Use, and Patient Positioning in Osteogenesis Imperfecta.
Regional Anesthesia

- Preferred method in OI patients with severe cardiac/pulmonary diseases
- Peripheral regional anesthesia
  - Nerve stimulator at high voltage at cause fractures!
  - Ultrasound guidance is preferred
- Neuroaxial anesthesia
  - Platelet dysfunction in OI patients can result in epidural hematoma
Postoperative Disposition

• Postoperative sedation to avoid further fractures
• Hemorrhagic diathesis from platelet dysfunction
• If the patient has restrictive lung disease may need PICU admission postoperatively
Conclusions:

• Identify severity of OI
• Preoperative evaluation
  - Airway
  - Cardiopulmonary
  - Hemotologic
• Fracture, fracture, fracture!
• Avoid succinylcholine
• Consider regional anesthetic options
• Postoperative disposition
References:


