Birth/neonatal history: Osteogenesis imperfecta (OI) and worsening hypercapnic respiratory failure

Autosomal Dominant: Collagen-3- hydroxylation defect
- Type I collagen production
- Lethal
- Severe
- Moderate
- Severe to lethal
- Rhizomelia, neonatal fractures, “popcorn” metaphyses, short stature

Autosomal Recessive: Collagen-3- hydroxylation defect
- Type I collagen production
- Mild
- Non-deforming
- Prepubertal fractures, deafness, aortic regurgitation

Case description, cont.

Airway management: Maintain spontaneous ventilation, avoid facial fractures from facemask ventilation, avoid spinal cord injury from DL and neck extension
- NIPPV continued in the OR with anesthesia machine
- Patient sedated from opioids given in NICU – Attempted FOI but unsuccessful due to patient movement

Figures (a) skeletal survey, (b) R lower extremity, and (c) R upper extremity radiographs show extensive BL rib fractures, radial, ulnar, femoral, tibial and fibular fractures.

- Sevoflurane given via NIPPV to provide quick onset and offset sedation while preserving SV
- FOI through AirQ LMA attempted, but patient desaturated during scope placement and ventilation was inadequate via LMA. Saturation recovered after LMA removal and resumption of NIPPV.
- FOI was then successfully performed while maintaining ventilation, oxygenation, and sevoflurane delivery via NIPPV.

Discussion
Anesthetic implications of OI:
- Airway management
  - Risk of cervical spine or mandibular injury
  - GlideScope and LMA use reported in case reports
- Positioning injuries
  - BP measurement: NIBP-induced fractures, may need arterial line
- Vascular access: fractures from tourniquet placement
- Comorbidities: aortic root dilation, mitral valve prolapse, kyphoscoliosis, pulmonary HTN
- Hyperthermia
  - Many case reports describe TIVA for OI
  - Evidence for association with MH is weak
  - Can develop intraoperative hyperthermia
  - Not associated with hypermetabolism
  - Responsive to standard cooling measures

Neonatal noninvasive airway management:
- CPAP: Provides lung recruitment by providing a continuous level of pressure in the airways
- NIPPV:
  - Facilitates ventilation and oxygenation by augmenting airway pressure with inspiration and improving upper airway patency
  - Can be synchronized and delivered by conventional ventilator
  - Benefit may be from increased mean airway pressure or pressure changes
  - Inconsistent results for improved TV and gas exchange compared to NCPAP
  - More studies need to assess role in avoiding intubation to reduce incidence of bronchopulmonary dysplasia

References