Anesthetic Managment of a Patient with Diastrophic Dysplasia Requiring Cesarean Delivery

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Background

- Diastrophic dysplasia (DTD)
  - Autosomal recessive, rare form of skeletal dysplasia
  - Characteristic limb shortening, severe scoliosis, cleft palate, club foot and joint contractures

- Anesthetic implications

Short Limbs → Difficult IV and NIBP

Spine instrumentation → decreased neck ROM, difficult neuraxial

Small oral opening, decreased TM distance → difficult airway

Case Presentation

41-year-old G4P0212 with a history of diastrophic dysplasia, scoliosis status post correction with Harrington rods, and two prior cesarean deliveries pre-term for maternal respiratory decompensation with known difficult airway.

Scheduled for third cesarean delivery with general anesthesia at 36 weeks 1 day.

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Case Presentation (cont.)

Airway History:

- GETA (2015) - grade I view with Glidescope, 6.0 mm ETT passed easily
- First c-section – grade III view (secretions) with pediatric Glidescope using low-profile D blade, 2 attempts by attending anesthesiologist
- Second c-section – attempted awake fiberoptic intubation, failed. Airway secured via awake Glidescope using low-profile pediatric D blade. Grade IIb view.

Planning and Execution of Cesarean Delivery Under General Anesthesia

- Access: 2 US PIVs, 20G and 18G
- Blood ordered and immediately available
- Airway on DOS: TM < 3 FB, MO <3 FB, good cervical ROM, adequate inter-incisor distance
- NPO > 8 hours
  - Pre-medicated: sodium citrate
- RSI
- First pass airway plan: asleep video laryngoscopy with McGrath 3 blade, 5.5 mm ETT
  - First attempt by CA3 with grade IIb view, 5.5 mm ETT passed easily
- Backup airway plan: Glidescope with pediatric low-profile blades, fiberoptic scope, and pediatric supraglottic airways (sizes 2-3) available
- Cesarean delivery uneventful and patient extubated in OR, no post-delivery anesthetic complications

Learning Points

- Neuraxial anesthesia vs general anesthesia for cesarean delivery in DTD parturients?
  - Very little literature on anesthetic management of these patients
  - Some case reports exist detailing successful epidural anesthesia for cesarean deliveries, but the balance of adequate and unpredictable block in a patient with baseline tenuous respiratory status and airway is difficult
- Obtaining appropriate vascular access is critical
  - If unable to use NIBP, will require either non-invasive continuous hemodynamic monitoring or invasive arterial line
  - If unable to obtain IV access with ultrasound guidance, consider CVC
- Make a defined airway plan with multiple modalities readily available
  - Physical exam and thorough review of any prior anesthetics and airway documentation is vital
  - Awake fiberoptic intubations for planned cesarean deliveries under GA could be necessary
  - In the case of emergent cesarean delivery requiring GA, mobilize difficult airway response teams/surgical colleagues to be readily available
  - If available, can consider contacting ECMO team to be on standby

Lagoy JS, Kofford ND, Gosselin BI, Russell MA, Morley SD. Management of a Parturient with Diastrophic Dysplasia. A A Case Rep. 2015 Jul 1;5(1):6-8. doi: 10.1213/XAA.0000000000000162. PMID: 26125691.

Porter H, Mendonca C. Anaesthesia for Caesarean section in a patient with diastrophic dwarfism. Int J Obstet Anesth. 2007 Apr;16(2):145-8. doi: 10.1016/j.ijoa.2006.08.012. Epub 2007 Jan 30. PMID: 17270421.