

Anesthetic management of a parturient with Ehler-Danlos syndrome complicated by known difficult airway and local anesthesia resistance undergoing Cesarean section

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Background

- Ehler-Danlos syndrome (EDS) is a group of connective tissue disorders causing increased extensibility and fragility of collagenous tissues, posing unique anesthetic considerations
 - higher risk of difficult intubation
 - local anesthesia resistance due to leakage through highly permeable connective tissue
- In this population, neuraxial anesthesia may pose a higher risk of postdural puncture headache, spontaneous dural rupture, increased spinal deformities, and increased bleeding
- There are no clear recommendations for either general or neuraxial anesthesia in obstetric patients with EDS

Case

- 31 y/o G1P0 patient at 36w6d with a history of EDS (hypermobile type), suspected local anesthesia resistance, and known difficult airway requiring fiberoptic intubation presents for elective primary Cesarean section
 - s/p cervical fusion, limited neck extension, failed intubations with video laryngoscopy leading to canceled case
- She reports inadequate analgesia from local anesthesia during prior abdominal surgeries, dental anesthesia, and facet blocks for chronic back pain but has never had neuraxial anesthesia
- After extensive counseling, the anesthetic plan was to trial spinal anesthesia via CSE, with GETA via awake fiberoptic intubation as a backup
- A CSE was performed with 1.9 mL of 0.75% bupivacaine in 8.25% dextrose, 15 mcg fentanyl, 150 mcg morphine, and 5 mcg dexmedetomidine administered intrathecally
 - CSE placement placed without complication
 - FHR remained in the 120-130s
 - Bilateral T4 levels to cold and a negative Allis clamp test were achieved prior to incision
 - Uneventful intraoperative course

Teaching points

- Neuraxial anesthesia can be successfully used as the primary anesthetic in Ehler-Danlos patients undergoing C-section
- EDS has several subtypes that may influence choice of anesthesia
- Given poor experiences with local anesthesia, patients present with considerable anxiety, so extensive counseling of the risks and benefits of each anesthetic technique is essential
 - Priority of the mother's life over the baby's, even in the event of fetal decelerations, was made clear