Anesthetic Management for Cesarean Delivery of a Parturient with Ehlers-Danlos Type III Syndrome associated with Postural Orthostatic Tachycardia Syndrome: A Case Report and Literature Review

Abstract Type: Case Report/Case Series
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Purpose: Ehlers-Danlos syndrome (EDS) consists of a group of inherited connective tissue disorders. Type III mainly involves joint hypermobility and is commonly associated with postural orthostatic tachycardia syndrome (POTS), an autonomic disorder that manifests as orthostatic intolerance and tachycardia. We report the anesthetic management of a parturient with EDS Type III associated POTS having repeat cesarean delivery.

Clinical Features: A 37 year old woman, G2P1, presented at 37 weeks gestational age for repeat cesarean section. Her medical history was significant for EDS Type III associated with POTS, mitral valve prolapse, and paroxysmal atrial fibrillation. She previously had an urgent cesarean delivery for oligohydramnios and breech presentation, where airway management was complicated by failed intubation by two anesthesiologists and maintained by laryngeal mask airway.

Pulse oximetry, non-invasive blood pressure, ECG, and fetal heart rate monitoring were established. A preload of 1L Ringer’s Lactate solution was administered, and a radial arterial line was placed. An epidural catheter was placed at the L2-3 interspace, and used to administer Fentanyl 100mcg and 20mL of 0.5% Bupivacaine (without Epinephrine) in small increments and slowly over 15min to achieve a T4 level block bilaterally. The patient remained hemodynamically stable, and a healthy baby was delivered uneventfully with APGARs 9 and 9. An infusion of Oxytocin 0.5U/min was started after delivery, and Epidural Morphine 3mg was administered. She was monitored in the PACU for 12 hours, and her recovery period was uneventful with good pain management.

Discussion: Only two cases with details about anesthetic management of cesarean delivery for parturients with POTS have been described in literature [1, 2]. Corbett described a case using an epidural technique that required conversion to a general anesthetic [1]. Jones described an attempt to use a titrated epidural that was complicated by poor sensory block, requiring conversion to a spinal anesthetic. Their patient required a total of 8mg of Phenylephrine to maintain hemodynamic stability, and experienced episodes of orthostatic intolerance during her recovery period [2].

Our case report highlights the successful use of a carefully titrated epidural technique for a parturient with EDS Type III associated with POTS and a history of difficult intubation. We were able to use epidural anesthesia as our sole anesthetic technique without the requirement to convert to general or spinal anesthesia, and thus avoided hemodynamic instability in our patient.

References