Abstract # 216

Anesthetic Management of a Parturient with Klippel-Feil Syndrome Presenting for Cesarean Delivery

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Introduction: Klippel-Feil Syndrome (KFS) is a rare congenital syndrome with an incidence of 1 in 40,000 births that is characterized by abnormalities in the formation of the cervical spine.1 KFS patients present with limited cervical mobility and are at increased risk of neurologic injury and/or devastation with airway manipulation.1 The increased risk of cervical injury and an anatomically difficult airway should never be underestimated, especially when caring for a gravid obstetric patient with KFS.

Case Report: A 22-year-old woman with KFS presented for scheduled delivery of her second child. Her past medical history included morbid obesity with a BMI of 40, situs inversus, atrial septal defect repair, moderate aortic insufficiency, hypertension, and multiple fusion abnormalities of her cervical spine including fusion of C2-3, C4-5, and C7-T1. She had a short neck with severely limited lateral rotation and moderate limitation upon neck extension. She also reported new onset radiculopathy in her upper extremities. One year prior, the patient had delivered an infant via cesarean section under spinal anesthesia with no complications. Spinal anesthesia was performed, and a difficult airway cart with a fiberoptic bronchoscope was readily available. The cesarean section was completed without incident.

Discussion: The etiology of KFS is largely unknown, and the presentation is varied with the classical triad of a short neck, low hairline, and decreased range of motion only presenting in 40-50% of patients.2 KFS patients often present with abnormalities including scoliosis, heart disease, renal abnormalities, and auditory involvement.2 Renal abnormalities can be serious and should be screened with a basic renal ultrasound. Cardiovascular abnormalities, mainly septal defects, have been reported.1 Patients often present with limited overall cervical mobility as well as hypermobility of the unfused cervical segments leading to increased risk of neurologic injury with airway manipulation or minor trauma, especially in the second and third decades of life.1 Because of the dynamic nature of KFS, a previous uneventful anesthetic record in a KFS patient is a poor predictor of neurologic outcome in a subsequent case. Regional anesthesia has been successfully reported in patients with KFS but is often technically challenging.1 If required, awake fiberoptic intubation is the preferred method of airway management.1 A parturient who presents with a cervical spine disorder presents unique and complex challenges. Proper preparation, planning, and communication are the key to ensuring the greatest margin of safety when providing anesthesia for this subset of patients.

References