A Case of Difficult Intubation Due to Anatomic Airway Changes in a Patient with Ehlers-Danlos Syndrome

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Introduction:
Ehlers-Danlos Syndrome (EDS) is a mixed connective tissue disease which leads to changes in the synthesis and/or structure of collagen. There are six major and several minor classes of EDS, each with its own related symptoms and complications.

Symptoms include:
• Joint Hypermobility
• Easy Bruising
• Arterial Fragility
• Tendon Rupture
• Kyphoscoliosis
• Atlanto-axial Instability

Anesthetic complications include:
• Tracheal Rupture
• Difficult Intubation from TMJ Dysfunction/
  Occipitatlantoaxial Instability
• Local Anesthetic Resistance
• Barotrauma

Case Report:
A 15 year old girl presented for elective transoral odontoidectomy for cervical instability and dysphagia. The patient has a history of EDS and Chiari malformation complicated by ambulation difficulties, neurogenic bladder, and dysphagia. Her neurologic symptoms resolved after several procedures, including posterior fossa decompression and craniocervical spinal fusion. The patient’s neurologic symptoms returned 8 months prior to presentation following neck manipulation. At that time the patient also had a difficult intubation and ventilation, leading to cardiopulmonary arrest, resuscitation and ultimate intubation by video laryngoscopy. Due to the anticipated difficulty of mask ventilation and intubation, the anesthetic plan was to induce the patient with a ketamine titration to maintain spontaneous ventilation.

We were able to mask ventilate the patient after beginning the ketamine titration and subsequently administered succinylcholine for paralysis. Intubation was attempted with a Glidescope 3, however only the most inferior aspect of the airway was visualized. Oral and nasal fiberoptic intubation was then attempted, though the airway appeared anterior and retroflexed, causing difficulty in passing the endotracheal tube. An attempt was then made to open the airway with a laryngeal mask airway (LMA), followed by an intubating LMA, but neither LMA positioned correctly in the airway.

Case Report (cont’d):
A combination of the Glidescope and nasal fiberoptic bronchoscope was then tried. Once again, the airway was visualized, but was anterior and the endotracheal tube could not be positioned and passed. Intubating oral airways were then inserted and did not provide benefit. A pediatric Glidescope, which has a more anterior curvature than the Glidescope 3, allowed for visualization of the lower half of the airway. The pediatric Glidescope was used in combination with a nasal fiberoptic bronchoscope, which allowed visualization of the airway and successful passing of the endotracheal tube. After successful intubation, the patient’s prior imaging was reevaluated by the neurosurgeon, who noted the patient’s airway had previously shifted anterior and cephalad.

Case Report (cont’d):
Before caring for a patient with EDS, the anesthesiologist should review the type of EDS a patient has, if known, and should consider all possible complications that may arise.

EDS may lead to anatomic airway changes that can lead to difficulty in securing an airway.

A thorough review of an EDS patient’s imaging can help plan an intubation strategy.

Discussion:

Resources: