

[NM-140] Muscle relaxation worsens ventilation in a patient with Osteogenesis Imperfecta with a difficult airway.

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Introduction:

A 23 year old male with osteogenesis imperfecta and a broken right femoral rod presents for removal of the rod and a right femur osteotomy.

Management:

On pre-operative examination, the patient had signs of a potentially difficult airway. He had a Mallampati grade 3 view with a short neck, decreased temporomandibular distance, large barrel chest, and decreased cervical range of motion from multiple cervical compression fractures. His last surgery was in 2008, and there was no history of difficult airway management previously.

Our plan was to proceed with an oral endotracheal intubation after successful bag mask ventilation. A Glidescope was chosen as the primary laryngoscope with a fiberscope as backup.

The patient was placed in the 'sniff' position using a moderate ramp. Mask ventilation with an oral airway was established after induction with propofol 2.5 mg/kg and fentanyl 1 mcg/kg. Rocuronium 0.5 mg/kg was given. Mask ventilation progressively deteriorated as muscle relaxation ensued. The decision was made to proceed with laryngoscopy using the Glidescope #3. A view of the epiglottis and posterior arytenoids was attained, but intubation was unsuccessful due to the anterior position of the larynx despite use of the Glidescope stylet. A laryngeal mask airway was then placed, but ventilation continued to be difficult, with oxygen saturations between 30% and 50%. Fiberoptic approach was attempted through the left nare and then through the LMA, but no view could be established due to redundant tissue.

ENT was called after the initial failed intubation for possible surgical airway or rigid bronchoscopy. However, as the muscle relaxation waned, ventilation slowly improved through the LMA. At return of 1 twitch by train of four, neuromuscular blockade was reversed and the procedure cancelled.

The patient returned for surgery 2 weeks later, where he underwent a successful awake nasal intubation using a fiberscope and his surgery proceeded uneventfully.

Discussion:

Osteogenesis imperfect (OI) is a connective tissue disorder arising from defects in collagen formation. Characteristics of patients with OI include brittle bones, hypermobile joints, skeletal deformities, and blue sclera. Anesthetic implications in patients with OI include positioning injuries, respiratory compromise from skeletal abnormalities, difficult airways, and hyperpyrexia. Prior experiences with easy intubation can be deceiving because disease progression alters bony structure in the face and neck. Airway management can be particularly challenging as manipulation of the cervical spine can cause fractures. Also, they often have short necks, protruding mandibles, the presence of a pigeon chest, and large tongues (1). Given these characteristics, anesthetic management of patients with OI should be approached cautiously.

Interestingly, it is commonly believed that muscle paralysis facilitates mask ventilation. Our case demonstrates this does not reliably occur in a patient with a difficult airway.

References:

1. Oakley I, Reece LP. Anesthetic Implications for the Patient with Osteogenesis Imperfecta. AANA 2010; 78(1):47-53.
2. Warters et al. The Effect of Neuromuscular Blockade on Mask Ventilation. Anaesthesia, 2011; 66: 163-167.

