Successful use of an Air-Q LMA for an unanticipated difficult intubation in an infant with Desbuquois syndrome

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ABSTRACT
We discuss a case of successful use of an Air-Q LMA as a conduit for endotracheal intubation for unanticipated difficult intubation in an infant with Desbuquois syndrome. A 4-month old infant was scheduled for laparoscopic Nissen fundoplication and gastrostomy tube placement for severe reflux and recurrent aspiration. The patient proved to be a difficult intubation and was eventually intubated through an Air-Q LMA after multiple unsuccessful attempts with other airway devices. Desbuquois syndrome is an extremely rare genetic disorder. Owing to the rarity of the syndrome, there is a paucity of literature regarding anesthetic management of this disorder. This case report would offer insight regarding difficult airway management and anesthetic implications of Desbuquois syndrome.

Introduction

Desbuquois syndrome (DBQD) is an extremely rare genetic disorder

• Features include short stature and extremities, joint laxity and dislocations, narrow chest, kyphoscoliosis and dysmorphic facies including a flat face, micrognathia and a short neck

• Patients present with respiratory compromise at birth or during infancy secondary to chest deformity and recurrent aspiration pneumonias

Case Description

A 4-month old infant was scheduled for a laparoscopic Nissen fundoplication and gastrostomy tube placement. He was a FTNB born at 40-weeks via SVD. Hospital course was complicated by respiratory distress and aspiration pneumonia. Pt also had dysmorphic facies, cleft palate, laryngomalacia, narrow thoracic cage and limb anomalies. A diagnosis of Desbuquois Syndrome was made by Genetics. In the OR, the patient was placed on standard ASA monitors and pre-oxygenated with 100% FiO2. Intravenous induction was performed using propofol (3mg/kg) and fentanyl (1 mcg/kg) through a pre-existing peripheral IV. Once the ability to mask ventilate was established, rocuronium (1mg/kg) was administered. Easy mask ventilation was followed by direct laryngoscopy. Two attempts at DL revealed a Cormack/Lehane view 3. A third intubation attempt was made with a C-Mac video laryngoscope and was unsuccessful. A size 1 Air-Q LMA was then placed easily. A fiberoptic scope was introduced through the LMA and a 3.0 microcuffed ETT was placed successfully. Anesthetic course was otherwise uneventful. Dexamethasone was administered for airway edema from repeated airway instrumentation and decision was made to continue post-operative mechanical ventilation.

Patient was extubated on postoperative day 4 and discharged home after a week with no sequelae.

Discussion

Pediatric patients with DBQD can potentially present for a host of surgical procedures. Anesthetic concerns include:

- Difficult airway
- Dysmorphic facies, micrognathia
- Cervical spine
- C-spine instability, cervical spine kyphosis
- Aspiration risk
- Reflux, dysphagia
- Ventilatory compromise
- Restrictive chest deformity, recurrent respiratory infections
- Regional anesthesia challenging
- Kyphoscoliosis, vertebral anomalies
- Difficult vascular access
- Limb anomalies

- Dysmorphic facial features and a short, fused neck can lead to a difficult airway
- Particularly challenging in neonates and infants
- Supraglottic airways devices are a crucial part of the difficult airway algorithm
- Familiarity with existing devices available for pediatric use can significantly aid in airway management
- Limited data on the use of Air-Q LMA in the pediatric literature
- Air-Q LMA has been used successfully for airway rescue and as a conduit for endotracheal intubation, both blindly and with the aid of a fiberoptic scope.

Conclusion

Airway management in patients with DBQD can be particularly difficult. The Air-Q LMA may be used to facilitate endotracheal intubation in these patients

References

1. Desbuquois syndrome. NIH Genetic and Rare Disease Information Center. https://rarediseases.info.nih.gov/diseases/1818/desbuquois