Prone Intubation through an Air-Q in a Difficult to Ventilate and Intubate Neonate with Desbuquois Dysplasia

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Introduction

Desbuquois Dysplasia:

- Less than 50 cases of Desbuquois Dysplasia have been described in literature.
- It is an osteochondrodysplasia characterized by micromelic dwarfism, microretrognathia, facial dysmorphism, thoracic hypoplasia, kyphoscoliosis, and joint laxity with dislocations.
- Two forms have been distinguished on the basis of the presence (type 1) or the absence (type 2) of characteristic hand anomalies.
- Type 1 has a high mortality rate (>33%) from respiratory failure.
- Diagnosis is made by physical and radiologic findings and confirmed by the genetic screening of *CANT1* in type 1 and *XYLT1* in type 2.[3]
- Previous published literatures described successful ventilation in a patient with Desbuquois Dysplasia using a Cobra perilaryngeal airway. However, supine endotracheal intubation through this device was not possible.[2]

Case Report

Pre-operative History :

- A 3-day old, 2.1 kg ex-full term baby girl with syndromic physical findings was transferred to our hospital for management of respiratory distress.
- Her multiple anomalies included cleft palate without cleft lip, midface hypoplasia, micro/retrognathia, lowset thumbs, hypoplastic femurs, bilateral hip and knee dislocation.
- On admission, the patient only tolerated lying in a prone position with nasal cannula, and would otherwise desaturate. She had a near respiratory arrest event during supine TTE and had to be returned to the prone position immediately.
- Anesthesia was called for an urgent intubation given the worsening respiratory status.

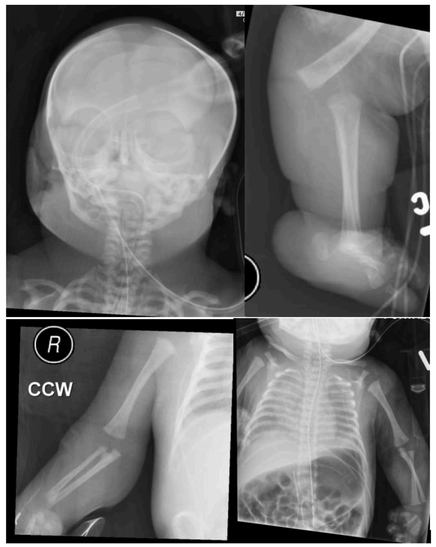

Figure 1: Radiologic findings from our patient include bilateral total knee and ankle joint dislocations, wrist and elbow subluxations, and hand contractures. In addition to facial characteristics, they are consistent with Desbuquois Dysplasia.

Intra-operative care:

- Patient was transported to the OR with standard ASA monitors in the prone position on nasal cannula.
- Glycopyrolate (0.1 mg), Dexmedetomidine (0.5 mcg/kg), and Ketamine (2 mg/kg) were slowly titrated with maintenance of spontaneous ventilation.
- Bag-mask ventilation of the patient in the supine position with an oral airway and two-handed technique was unsuccessful.
- Oxygen saturation improved with mask ventilation in the prone position with patient's head turned lateral. A 0.5 Air-Q LMA was inserted in the prone position with subsequent easy ventilation.
- To secure the airway, a 2.8 mm fiberoptic bronchoscope was passed through a Double Swivel Elbow connector attached to the Air-Q with a 3.0 uncuffed endotracheal tube (ETT) loaded. Ventilation was maintained via the Air-Q throughout the intubation process.
- After confirmation of ETT placement with the fiberoptic scope and capnography, the patient was paralyzed with Rocuronium (1 mg/kg).

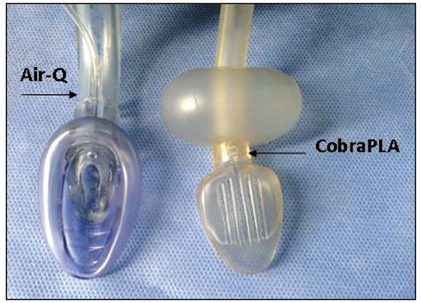

Figure 2: Air-Q and Cobra Perilaryngeal Airway[1]

Post-Operative:

- The patient was transported intubated to the NICU in a stable condition.
- The patient returned to the operating room two weeks later for a Laryngoscopy and Bronchoscopy by ENT with the intubation being described as "extremely difficult"
- A tracheostomy was placed at a later date, and the patient was placed on hospice care two months later.

Discussion

- Our case described the first successful emergent prone intubation in a 2.1 kg neonate with Desbuquois Dysplasia.
- The unique aspect of this intubating scenario was the inability of the patient to tolerate the supine position. Therefore, common airway techniques by ENT surgeons such as rigid bronchoscopy and tracheostomy would not be feasible.
- We believe that we demonstrated the first successful example of a prone intubation through an Air-Q LMA using a fiberoptic scope in this difficult patient population.
- Our technique may be applied to other syndromic patients that only tolerate prone positioning.

References

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