Case report

Use of CobraPLA™ for airway management in a neonate with Desbuquois syndrome. Case report and anesthetic implications

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Summary

We present the anesthetic management of an infant with Desbuquois syndrome (a rare form of micromelic dwarfism) with a possible difficult airway. The anesthetic implications of this syndrome are presented. The airway was managed with a new supraglottic device – the CobraPLA. Although intubation through this device was not possible in this instance, CobraPLA provided a satisfactory supraglottic airway. It was easy to insert and provided satisfactory conditions for positive pressure ventilation. The CobraPLA provides another option for airway management.

Keywords: CobraPLA; airway; Desbuquois syndrome

Introduction

Managing the difficult airway in neonates is one of the most challenging aspects facing anesthesiologists because of lack of tools of airway assessment compared with adults. In addition a number of syndromes and malformations are associated with difficult airway. Use of a supraglottic device to create a patent airway is recommended by the difficult airway algorithm in case of a failed intubation/ventilation scenario. Many types of supraglottic airway devices have been manufactured since the introduction of the laryngeal mask airway (LMA) into clinical practice by Brain et al. (1). We used the CobraPLA (a new supraglottic device) for management of an infant with Desbuquois syndrome with a potentially difficult airway.

Case report

A 3-week-old Hispanic male, weighing 2.3 kg was scheduled to undergo G-button placement for feeding access. The patient was born at 36 weeks gestation, weighing 2.04 kg, to a 24-year-old mother with limited prenatal care. The pregnancy was complicated by preterm labor and breech position. The fetus was found to have multiple congenital anomalies on a prenatal ultrasound, including renal pelviectasis, short limbs, and spinal anomalies. The infant was born by cesarian section under epidural anesthesia with APGAR scores of 5, 6, and 7 at 1, 5, and 10 min, respectively. In the neonatal unit the baby was alert and vigorous. Physical examination revealed a flat face, hypoplastic midfacies, pectus...
carinatum, sternal retractions, tachypnea, and inspiratory stridor. Evident also were multiple skeletal deformities: congenital dislocation of knees and elbows, subluxation of C5–6 and S2–3, overlapping digits and hypoplastic thumbs and great toes. Chest X-ray and CT showed normal lungs and a deformed thorax because of a short and deformed sternum and ribs. Cardiac ECHO revealed dextrocardia (secondary to rib and sternal anomalies), with normal cardiac chambers and moderate to severe tricuspid regurgitation with near systemic pulmonary pressure. Abdominal ultrasound revealed pelviectasis. A modified barium swallow showed discoordinated swallowing and sucking without gastroesophageal reflux and normal gastric emptying. Because of the skeletal abnormalities and the pulmonary hypertension a diagnosis of Desbuquois syndrome was considered.

All attempts to feed the baby failed because of cyanosis during feeding and a G-tube placement was recommended.

At preoperative evaluation the patient presented with a large face, microstomia (small mouth), micrognathia, very short neck, and very small deformed chest and sternum (Figure 1). In the OR, standard monitors were applied (ECG, noninvasive blood pressure, pulse oximetry) while the baby was placed on an infant warming blanket. The anesthesia was induced with sevoflurane in 100% O2 through a facemask and the anesthesia was supplemented with small doses of propofol 1 mg·kg⁻¹. In order to avoid extension of the cervical spine associated with laryngoscopy and tracheal intubation, a 0.5 size CobraPLA was inserted without any difficulty; the cuff was inflated so that a leak was heard at 20 cm H2O. A 2.2-mm neonatal fiberscope (LF-P, N25; Olympus America Inc., Melville, NY) armed with a 3.5-mm tracheal tube was inserted through the CobraPLA while the patient was ventilated with 2% sevoflurane. The vocal cords were easily (score 3 according to Brimacombe et al. (2) classification) visualized and the scope was advanced into the trachea. Further attempts to advance the tube along the fiberscope failed because of an acute angle of access into the larynx and the flexibility of the neonatal fiberscope. Considering the difficulty encountered and the fact that the surgery was quite short we decided to proceed with the surgery without intubation, under ventilation with the CobraPLA. The surgery lasted 15 min, the sevoflurane was discontinued and the CobraPLA removed 5 min later without any complications. The postoperative course was uneventful.

**Discussion**

Desbuquois syndrome is a rare skeletal dysplasia with presumed autosomal recessive inheritance. It is characterized by micromelic dwarfism (severe proportionate shortness of stature), narrow chest, vertebral and metaphyseal abnormalities, and advanced carpotarsal ossification. The findings in the hands are particularly distinctive with supernumerary ossification centers that cause deviation of the fingers. Dysmorphic facial features include a round flat face, prominent eyes, micrognathia, and long upper lip with flat philtrum. Severe, potentially lethal respiratory distress secondary to the small thorax is not uncommon. Survivors have developmental delay and generalized joint laxity, with dislocatable knees which is progressive (3). Cleft palate and mental retardation have been described (4). The differential diagnosis of Desbuquois syndrome include Kniest dysplasia, which is an autosomal dominant disorder (5), Catel-Manzke syndrome (normal birth length, slight joint laxity), and Larsen syndrome (short stature, pelvic changes similar to Desbuquois syndrome) (6).

Interestingly, we could not find any reports of anesthetic management in patients with Desbuquois
syndrome. The anesthetic implications include the following.

1. Possible difficult airway because of micrognathia, microstomia, hypoplastic midfacies (as in our case), and cleft palate. Consequently, all precautions for a difficult airway should be taken including the availability of a fiberoptic scope as well as supraglottic devices in case of difficult mask ventilation.

2. Joint hyperlaxity, including the cervical spine (as in our case), which requires attention to positioning and avoidance of cervical spine extension and its potential consequence: tetraplegia.

3. Possible difficulties with ventilation from restrictive pulmonary disease secondary to the deformed thorax.

4. Cardiac anomalies, which in our case revealed dextrocardia and tricuspid regurgitation with pulmonary hypertension.

5. Discoordination of swallowing and suck which can increase the risk of aspiration. In our case there was no gastroesophageal reflux and the gastric emptying was normal.

6. Urinary tract malformations with possible increased rate of urinary infections.

The laryngeal mask airway (LMA™) has been used in difficult airway cases in adult (7,8) as well as in pediatric patients (9). In children, the LMA has been used extensively in different difficult airway clinical situations (10–13). Considering the success of the LMA in the management of difficult airways, other supraglottic devices should also be useful in such situations.

The perilaryngeal airway (CobraPLA®, Engineered Medical Systems, Indianapolis, IN, USA) is a supraglottic device recently introduced into clinical practice. It is designed to be positioned in the hypopharynx where it abuts the structures of the laryngeal inlet. The CobraPLA is a supraglottic airway in the same class as the LMA and the cuffed oropharyngeal airway (COPA). The CobraPLA comprises a breathing tube with a wide distal end; a cuff is attached just proximal to the wide part. The cuff, when inflated, serves to seal off the distal end from the upper airway, thereby allowing positive pressure ventilation to be administered. The wide end is designed so that it holds both soft tissues and the epiglottis away from the distal CobraPLA, thereby allowing ventilation through the end of the device.

Extending distally from the wide part of the CobraPLA is a softened ‘tongue’ which facilitates passage into the hypopharynx by bending in the direction of the glottis as the CobraPLA is inserted (Figure 2). Once in place, it abuts directly against the glottis (i.e., against the aryepiglottic folds) with the anterior wall holding the epiglottis out of the way. The distal end has a number of slots or bars that allow ventilation through the end. The CobraPLA is made of biocompatible plastic materials and contains no latex.

Contraindications for the use of the CobraPLA are similar to those for the LMA and include: emergency surgical procedures, patients at risk for gastric regurgitation and vomiting, long-term ventilatory management, and patients with known pharyngeal abscesses or tumors. The CobraPLA is a single use device and is currently manufactured in eight sizes (0.5, 1, 1.5, 2, 3, 4, 5, 6) with four sizes suitable for pediatric patients (0.5, 1, 1.5, 2).

In a recent study in adults, Akca et al. (14) found the CobraPLA to have insertion and recovery characteristics similar to the LMA, but better airway sealing capabilities, which improve the ability to provide mechanical ventilation.

Agro et al. (15) successfully inserted the CobraPLA within 10 s and 57% of the patients had immediate effective ventilation, the rest requiring additional maneuvering. Gaitini et al. (16) compared three supraglottic devices (LMA, CobraPLA, and PAXpress) and found that the three devices were...
equivalent in providing effective and safe airway management in elective general anesthesia with mechanical ventilation, with the PLA offering some advantages such as a more effective seal and a better fiberoptic score. This means that a higher inspiratory pressure can be achieved with the CobraPLA during positive pressure ventilation and with better fiberoptic scores, intubation through the device might be achieved more easily.

Having used the CobraPLA in over 200 pediatric and neonatal patients with normal airways with excellent results, we felt comfortable using it in this patient with an anticipated difficult airway. The insertion and ability to ventilate were excellent. Although, we were able to pass a neonatal fibroscope through the device we were not successful in advancing the tracheal tube because of the fact that the neonate scope was not stiff enough to guide the tube especially given the acute angle of access in this patient.

In conclusion, this is the first report of anesthetic management in an infant with Desbuquois syndrome and the first report of use of a CobraPLA in an infant with possible difficult intubation. Although intubation through this device was not possible in this instance, CobraPLA provided a satisfactory supraglottic airway. It was easy to insert and provided satisfactory conditions for positive pressure ventilation. The CobraPLA provides another option for airway management.

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