Airway management of an infant with Pierre Robin syndrome for mandibular distraction osteogenesis

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ABSTRACT

Airway management of children with congenital craniofacial anomalies, especially those with Pierre Robin syndrome (PRS), which occurs in 1/8500–1/14,000 live births, is a challenge for the anesthesiologists. We report the case of a 60-day-old female PRS child who presented with severe upper airway obstruction and desaturation. The child underwent thin-sliced computed tomography followed by craniofacial reconstruction with mandibular distraction osteogenesis under general anesthesia. It is concluded that in children with PRS, securing airway even with fiberoptic bronchoscope can be difficult and may require additional airway assistance in the form of jaw thrust and tongue displacement during the procedure. Post-operative ventilatory assistance with nasal airway and continuous positive airway pressure may be helpful until effective mandibular distraction is achieved.

Key words: *Airway, Fiberoptic bronchoscope, Intubation, Pierre Robin syndrome*

Airway management of children with congenital craniofacial anomalies, especially those with Pierre Robin syndrome (PRS), which occurs in 1/8500–1/14,000 live births [1], is a challenge for the anesthesiologists. These patients usually present with failure to thrive and can have airway obstruction [2,3]. Although many airway instruments are available for difficult airway management of adults, the devices that can be used for pediatric patients are limited [4,5]. We report the airway management of an infant with PRS who developed severe dyspnea due to morphological abnormality.

CASE REPORT

A 60-day-old Pierre Robin syndromic female child weighing 2600 g with severe micrognathia, large midline cleft and glossoptosis presented with severe upper airway obstruction and desaturation. The infant was kept in a prone position in the pediatric intensive care unit (PICU) and was saturating between 90% and 94% with supplemental oxygen through nasal prongs.

On examination, the child had severe micrognathia, cleft palate, and desaturation even in the prone position and was in obvious distress. There was severe indrawing of the chest and the accessory muscles of respiration were acting. Cardiac examination, echocardiography, and blood investigations were essentially normal. Consent for tracheostomy, if required, was obtained from the parents. The child was posted for thin sliced computed tomography (CT), to be followed by craniofacial reconstruction with mandibular distraction osteogenesis (MDO).

Appropriate sized oral airways, endotracheal tubes (ETTs), Macintosh and Miller blades, and supraglottic airway devices (laryngeal mask and laryngeal tubes), C Mac video laryngoscope, and pediatric fiberoptic bronchoscope (FOB) were kept ready. Two senior anesthesiologists participated in airway management. Furthermore, an otorhinolaryngologist was included for the possibility of an urgent tracheostomy in view of the development of a "non-intubated, non-ventilated" condition. The child was brought to the operation theatre in a prone position with a saturation of 94%, then electrocardiogram and non-invasive blood pressure monitors were attached to the patient.

In the operation theater table, the child was put in a supine position and was preoxygenated with 100% oxygen using a Jackson Rees Circuit with a flow rate of 4 L/min with a size one anatomical face mask. Even with jaw thrust and chin lift, the child was struggling to breathe with minimal bag movement and end-tidal carbon dioxide tracing. Without jaw thrust, even before premedication, there was complete airway obstruction and use of oral airway also did not help in relieving the obstruction. While maintaining mask ventilation, the child was premedicated with 30 µg of glycopyrrolate with 94% saturation and heart rate of 136 beats/min.

Anesthesia induction was carried out using 8% sevoflurane in oxygen. The mask ventilation was possible, though difficult, and intubation was planned to be performed without using muscle relaxants. The initial plan was to intubate the child in a supine position using a C-Mac video laryngoscope with a pediatric D blade, but it was abandoned as even epiglottis was not visualized with a video scope. Although the child desaturated to 60% during the procedure, saturation soon picked up to 90% with the resumption of mask ventilation.

As the child came out of anesthesia, a nasopharyngeal airway was introduced, and the Jackson Rees Circuit was attached to it and the depth of anesthesia was maintained with sevoflurane to keep the child quiet. FOB (2.9 mm outer diameter) assisted intubation was then attempted in a supine position using a 3 mm inner diameter (ID) ETT introduced through the opposite nostril. Visualization of the larynx was difficult due to a lack of space posteriorly. Following jaw thrust and pulling the tongue out with a Magill's forceps, the larynx was visualized and the trachea was intubated. The child maintained saturation above 90% during the procedure.

After securing the airway, the FOB was inserted through the other nostril and distance from the nasopharynx to the nares were measured, which was 5.5 cm. The child was then shifted for CT and anesthesia was maintained with sevoflurane through the nasal ETT connected to the Jackson Rees Circuit. The child was breathing spontaneously throughout the procedure maintaining a SpO_2 >97%. After the CT, the patient was shifted to pediatric ICU and extubation was planned once she was fully awake and active.

During extubation, the nasal tube was pulled out up to the 5.5 cm mark and was maintained in that position, then the tube length was shortened by 7 cm and the ETT connector was attached. Thus, the same ETT was used as a nasopharyngeal airway and nasal continuous positive airway pressure (CPAP) was initiated through it. The child tolerated the nasal CPAP well and maintained a SpO_2 >94% with fraction of inspired oxygen (FiO_2) of 0.5. She was posted for elective mandibular distraction 2 days later after discussion with the multidisciplinary team.

On the day of surgery, the infant was brought to the operation theater with nasal CPAP and had a saturation of 96%. Though chest indrawing was minimal, the accessory muscles of respiration were acting. Before anesthesia induction, standard monitors were attached to the patient. She was premedicated with 30 µg of glycopyrrolate after documenting the starting SpO_2 96% and heart rate 144 beats/min. Just before induction, nasal CPAP was discontinued and inhalation induction was carried out through the previously placed nasal airway using 8% sevoflurane in oxygen maintaining spontaneous ventilation. A Magill's forcep was used to gently pull the tongue out to aid visualization and jaw thrust was also applied. FOB-assisted intubation was attempted nasally in a supine position using pediatric FOB, with a 3.5 mm ID ETT maintaining the depth of anesthesia with sevoflurane.

Intubation with FOB was difficult this time due to thick tenacious secretions adherent around the laryngeal inlet. Due to desaturation <60%, the procedure was discontinued twice to allow mask ventilation, and intubation was successful on the third attempt. There was no bradycardia during the procedure. The nasopharyngeal airway was removed after intubation, and then ETT was secured (Fig. 1). A thorough oral and pharyngeal suction were performed with the help of C-Mac after securing the ETT. The anesthesia was maintained with sevoflurane, fentanyl, and atracurium. After the procedure, the child was shifted back to the PICU for elective ventilation through nasal ETT (Fig. 2).

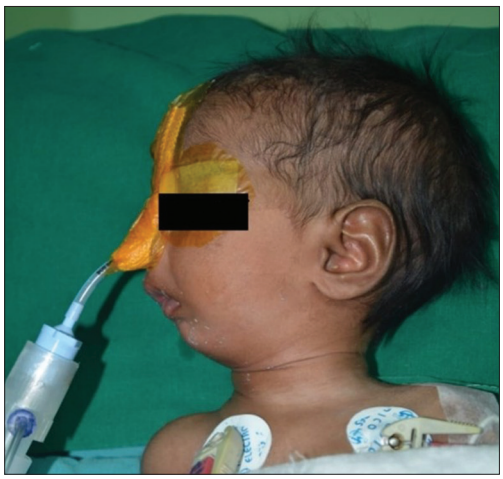

Figure 1: Patient after intubation

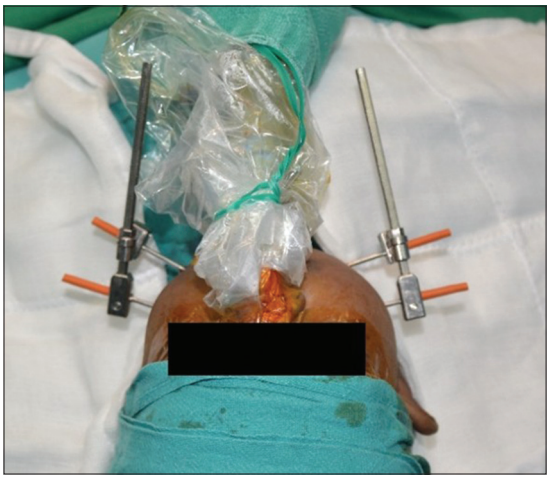

Figure 2: Patient after applying distractors

The child was ventilated for 1 more day and then extubated, but CPAP with nasopharyngeal airway was continued. Adequate mandibular distraction was achieved after 1 week. Following which, the nasal airway was removed and the child was put on high flow humidified oxygen and was able to maintain a supine position without respiratory distress. Supplemental oxygen was discontinued after 4 days, later the child was shifted to the ward and had an unremarkable recovery.

DISCUSSION

Pierre Robin sequence has been described as a triad of micrognathia, glossoptosis, and upper airway obstruction and is often accompanied by the cleft palate [5]. PRS often results in the upper airway obstruction or feeding difficulty secondary to micrognathia, glossoptosis, or a displaced tongue that comes in contact with the pharyngeal wall. Usually, progressive airway obstruction may become more noticeable in the 2nd month of life [6]. The immediate management includes positioning and placement of nasal pharyngeal airway [7]. These children may require surgical treatments including tongue lip adhesion, MDO, subperiosteal release of the floor of the mouth, and tracheostomy [8] if their airway obstruction

deteriorates or they fail to thrive. To have these procedures done, their airway needs to be secured first.

Currently, mandibular distraction or the lengthening of the mandibular bone [9,10] is the most common surgical procedure performed to correct a retracted tongue and the airway obstruction, produced in patients with mandibular hypoplasia. Mandibular development is the key for clearance at the base of the tongue to prevent subsequent upper airway obstruction. Distraction osteogenesis is a *de novo* biological bone formation process where traction pressure is applied [9,10]. MDO corrects micrognathia, gradually lengthens the mandible, and can be performed safely in infants. This procedure successfully prevents obstructive sleep apnea in most cases, thus avoiding the need for tracheostomy.

CONCLUSION

In children with PRS, securing airway even with FOB can be difficult and may require additional airway assistance in the form of jaw thrust and tongue displacement during the procedure. Post-operative ventilatory assistance with nasal airway and CPAP may be helpful until effective mandibular distraction is achieved.

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