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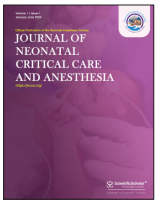

Case Report

Airway Management in a Neonate with Pierre–Robin Syndrome: Challenges for the Anesthesiologist

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ABSTRACT

Pierre–Robin sequence (PRS) is a developmental abnormality starting from 9 to 10 weeks of gestation. Hypoplasia of the mandible progresses eventually to a receding mandible, large tongue, and airway obstruction which may be associated with cleft palate and recurrent respiratory infections. The craniofacial abnormalities make the airway difficult posing a threat to the anesthesiologist. We present a case of difficult airway in a 14-day-old neonate having PRS associated with Hirschsprung's disease posted for laparotomy.

Keywords: Pierre–Robin sequence, Neonate, Difficult airway, Anesthesia management

INTRODUCTION

Pierre–Robin sequence (PRS) is characterized by micrognathia, glossoptosis, and airway obstruction.[1] Since birth, these neonates may exhibit a multitude of challenges such as respiratory distress, feeding difficulty, gastroesophageal reflux, bronchial micro-aspiration, pulmonary infections, obstructive sleep apnea, and failure to thrive.[2] The PRS affects approximately up to 1 in 14,000 newborns a year.[3] It may be isolated or present in association with other congenital deformities.

Craniofacial dysmorphology makes ventilation, oxygenation, and intubation challenging in these patients. Flexible fiberoptic bronchoscopy is considered gold-standard technique for securing the airway. However, certain adjuvants such as laryngeal mask airway (LMA) and I-gel can prove to be a savior in emergency conditions.

Here, we report the successful establishment of an airway in a neonate having PRS with Hirschsprung's disease, wherein an I-gel was used as a guide for endotracheal (ET) intubation.

CASE REPORT

A 14-day-old male neonate weighing 2.2 kg presented to the pediatric surgery department with complaints of abdominal distension and inability to pass stool for three days. The baby was born full-term by normal vaginal delivery without any instrumentation. He had micrognathia, macroglossia, and a small chin. On examination, his general condition was poor, the baby was lethargic, his heart rate was 160/min, his blood pressure was 90/54 mmHg, and his color was pink with active reflexes [Figure 1]. His per abdominal examination revealed a tense, distended abdomen with tympanic notes in all quadrants. Blood investigations showed that hemoglobin was 15.3 g/dL, total leukocyte count was 7100/μL, platelet was 1,60000/μL, blood urea was 21.49 mg/dL, serum creatinine was 0.37 mg/dL, serum sodium+ was 137 mmol/L, and serum potassium+ was 3.90 mmol/L. Viral markers were negative. X-ray abdomen showed multiple air-fluid levels suggesting dilated bowel loops and an empty rectum. Similar findings were seen in ultrasonography-abdomen, i.e., a dilated sigmoid colon. In view of clinical features such as micrognathia, macroglossia, and small chin along with radiological and sonographic findings, he was diagnosed as a case of PRS with Hirschsprung's disease and was planned for elective laparotomy and leveling biopsy.

There were many anticipated anesthetic challenges in this case, such as neonatal age, poor hydration, poor general condition, difficult airway, full stomach, high risk of aspiration, hypoxia, increased opioid sensitivity, obstructive sleep apnea, and high chances of hypothermia and micro-aspiration. Keeping all aspects in mind, the temperature of the operation theater was optimized to 32°C, and all the emergency equipment and drugs were kept ready along with neonatal difficult intubation cart including ET tubes

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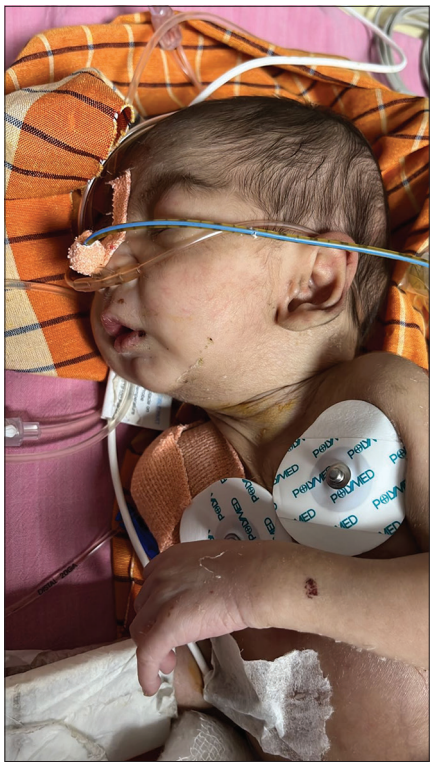

Figure 1: Neonate with Pierre–Robin sequence.

(ETTs), airway, LMA, I-gel, bougie, and cricothyrotomy set. Our plan-A for airway management was direct laryngoscopy and intubation, plan-B was insertion of a supraglottic airway device and intubation through it, i.e., passing a 2.5 mm uncuffed tube through I-gel size no. 1, and plan-C was securing a surgical airway. Hence, an ENT surgeon was also called at the time of induction.

Pre-oxygenation was done with 100% oxygen (O_2) for 5 min after putting a roll under the shoulder. Then, premedication was done with injection glycopyrrolate 0.02 mg iv and midazolam 0.2 mg iv and the patient was induced with injection ketamine 5 mg iv and sevoflurane 5% with O_2:air (50:50). Direct laryngoscopy was performed by a trained anesthesiologist but the glottic opening was not visible (CL – grade IV) even after external manipulation of the larynx and ET intubation was not possible. Hence, as per our plan-B, I-gel size 1 was inserted and an uncuffed ETT of 2.5 mm ID size without a connector was passed into the trachea through it. After satisfactory confirmation of tube position by auscultation and capnography, I-gel was removed carefully while maintaining the ETT *in situ*. Then, the position of ETT was reconfirmed, and the tube was secured. Spontaneous ventilation was maintained until the successful insertion of the ETT. Thereafter, anesthesia was maintained with O_2:air (50:50), sevoflurane, and IV fentanyl 3 µg. As the patient maintained adequate ventilation with a 2.5 mm ETT, no attempts were made to change the tube size to avoid additional trauma and mucosal edema. However, an oral pack was inserted to minimize air leak and prevent chances of aspiration. Thereafter, assist-control ventilation was done using a JR circuit and no palpable leak was present. Hence, surgery was allowed to start, a transverse loop colostomy was done, and biopsy samples were taken. The surgery was completed within 60 min. Keeping in mind the possible post-operative respiratory complications, the ETT was kept *in situ* for 24 h. The patient maintained stable vital parameters on spontaneous ventilation and was extubated uneventfully the next day.

DISCUSSION

Neonates and infants are known to be challenging to ventilate and intubate. As this was a case of intestinal obstruction with an anticipated risk of difficult intubation and micro-aspiration, securing a definitive airway was essential. Flexible fiberoptic-aided intubation is considered as gold standard for difficult intubation,[3] but unfortunately, it was not available in our set-up. Furthermore, its availability is limited due to cost and high maintenance. Various other techniques have been demonstrated for successful intubation in these patients. These include LMA, fiberoptic scope, retrograde wire, GlideScope, Shikani scope, Airtraq, and Air-Q scope.[4-8] Supraglottic airway devices have been proven to be a boon in the management of difficult airway. An Air-Q LMA would have been an ideal choice to guide the ETT in place; however, unfortunately, it was also not available in our setup.[9]

Induction was achieved with ketamine and sevoflurane so as to maintain spontaneous ventilation. Bag and mask ventilation was avoided to minimize the chances of aspiration. Literature has revealed the use of an LMA as a conduit for the insertion of an ETT in a case of PRS.[10] It is easy to perform, less traumatic and less time consuming than making multiple attempts at laryngoscopy or blind tracheal intubation. I-gel is known to have various advantages such as ease of insertion, effective seal pressure adequate to prevent aspiration, and lack of inflatable cuff, hence, less chances of mucosal damage.[11] Here, we used an I-gel to guide ET intubation due to the unavailability of neonatal fiberoptic and intubating LMA in our set-up and considering its advantages. When a fiberoptic bronchoscope is not available, supraglottic devices can also be used to aid ET intubation in selected cases requiring a definitive airway apart from being used as a rescue device for ventilation.

CONCLUSION

A supraglottic device such as I-gel can successfully be used as a conduit for the insertion of an ETT in cases of difficult intubation, especially in resource-limited settings even in neonatal age group.

Ethical approval

Institutional Review Board approval is not required.

Declaration of patient consent

The authors certify that they have obtained all appropriate patient consent.

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Conflicts of interest

There are no conflicts of interest.

Use of artificial intelligence (AI)-assisted technology for manuscript preparation

The authors confirm that there was no use of artificial intelligence (AI)-assisted technology for assisting in the writing or editing of the manuscript and no images were manipulated using AI.

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