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Successful Intubation of a Patient with Pierre Robin Sequence Outside an Operating Theatre after Repeated Failed Trials: A Case Report

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Pierre Rubin sequence · Difficult intubation · Failed intubation · Fiber-optic intubation · Micrognathia · Glossoptosis · Airway obstruction · Distraction osteogenesis

Abstract
Pierre Robin sequence (PRS) is a rare congenital birth defect characterized by an underdeveloped jaw, backward displacement of the tongue, and upper airway obstruction. Babies born with PRS commonly experience trouble breathing and feeding, early in life, resulting from the tongue's position, smaller jaw size, and the cleft palate formation. In this report, we will discuss the approach of emergency intubation outside an operating theatre in a 2-week-old preterm female neonate (33 weeks) weighing 2.2 kg diagnosed with PRS and an associated complete cleft palate. She needed emergency intubation immediately after admission due to desaturation and respiratory distress.

Introduction

Micrognathia, glossoptosis, and airway obstruction are the three characteristics of Pierre Robin syndrome, also known as Pierre Robin sequence (PRS). In around 50% of cases, the condition is associated with a cleft palate [1].

Since neonates and infants with the PRS are known to be challenging to intubate, numerous airway management approaches have been described mainly in case reports and short series. Flexible fiber-optic-aided intubation is thought to be the gold standard for difficult intubation [2].

We will present a case of PRS, who had recurrent episodes of airway obstruction, accompanied by desaturation. Several intubation trials by neonatologists were done, but unfortunately, all failed. In this case report, we will describe our management plan for a successful intubation.

Case Report

A 2-week-old preterm female neonate (33 weeks gestational age) weighing 2.2 kg was referred to neonatal ICU in view of recurrent episodes of airway obstruction. She was diagnosed with PRS and an associated complete cleft palate.

After admission, the patient had several episodes of desaturation and respiratory distress due to airway obstruction. So, she was kept nil per oral and on intravenous fluids.

Neonatologists tried several maneuvers to maintain the airway, including prone and lateral position as well as nasopharyngeal airway but all these maneuvers failed. Written consent from the parent was obtained for tracheal intubation, and neonatologists tried to intubate her but their trials failed and edema of the airway started to worsen the condition.

The on-call anesthetist was called for help due to severe respiratory distress. He attended the call and the first step he did was to

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check the bag mask ventilation. Ventilation was possible with the two-hand technique using bag and mask ventilation.

Equipment for difficult intubation was prepared, including laryngeal mask airway (LMA), video laryngoscope with different blade types and sizes 0–1, as well as different sizes of endotracheal tubes for this age. After preparation of all equipment in NICU, sedatives and emergency medications were prescribed.

Trail of intubation started, and vital signs were continuously monitored. The heart rate at the start of the trial was 160/min, and oxygen saturation was well-maintained 97% with bag and mask ventilation.

Midazolam 0.5 mg was administered intravenously, ventilation was maintained after sedation. Macintosh blade size zero video laryngoscope was used and showed moderate to severe airway edema including the tongue and oropharynx. There was a lot of saliva and little blood from the previous attempts, so suction of airway secretions was performed.

After suction, there was no view for the larynx, so the blade was changed to Miller size 0. After changing the blade, the posterior commissure of the larynx was barely visible.

The first trial of intubation by anesthetist failed, so bag and mask ventilation was resumed, and oxygen saturation was maintained at 91–96%. Another attempt was started with a different technique, LMA was inserted and we checked the ventilation, ventilation was satisfactory.

Uncuffed tube size 3 mm was mounted on a flexible intubating fiberscope size 2.8 mm. The on-call anesthetist advanced the distal tip of the scope through the LMA and he barely visualized the larynx. While slowly advancing the scope to enter the larynx, additional 0.5-mg midazolam was administered intravenously. He could pass the scope through the larynx, and then he advanced the mounted tube slowly until it passed the laryngeal inlet. The tube position was confirmed by the scope. He removed the scope and started ventilation through the tube; chest auscultation confirmed bilateral equal air entry.

The connector of the tube was removed to facilitate removal of the LMA. A soft bougie was inserted in the tube to keep it in place, LMA was removed slowly, and then the bougie was removed. The connector was connected again, and finally, chest auscultation was performed to check the tube position.

Endotracheal tube was successfully inserted and well secured with adhesive tube holder at depth of 9 cm at the lip level, and the neonatologist connected it to the mechanical ventilator. Sedation was started and chest X-ray post intubation showed a satisfactory level of the tube and clear lungs bilaterally.

Discussion

The criteria defining PRS were identified by Dr. Robin, a French stomatologist, in 1923. Dr. Robin described the tongue obstructing the oral cavity with a small mandible and termed it Glossoptosis [1, 2].

The clinical criteria defining PRS are small mandible (Micrognathia), backward and downward displacement of the tongue base (Glossoptosis), and airway obstruction. In some literatures, PRS is described as consisting of micrognathia, glossoptosis, and cleft palate [3, 4]. The cleft palate is commonly associated with PRS, but it is not always present. However, airway obstruction is present in all PRS patients, and it confirms the clinical diagnosis [5].

Symptoms of PRS vary from mild to severe and mild cases might be unrecognized. This might explain the variable incidence of 1:5,000–1:85,000. Symptoms of PRS include patients presenting with respiratory and feeding problems. Respiratory problems include respiratory distress and airway obstruction so the neonate/infant might present with stridor, retractions with or without cyanosis. Feeding difficulty, reflux, and failure to thrive might also occur secondary to airway obstruction [5].

The management approach for a PRS patient necessitates multidisciplinary team care including specialties like plastic surgery, ENT, neonatology, radiology, and anesthesia. Patients with PRS need anesthesia for a number of procedures like tongue-lip adhesion, mandibular osteogenesis, Nissen fundoplication, gastrostomy, or tracheostomy in refractory airway obstruction or failure to thrive [4].

Airway management, of a PRS patient, can be particularly challenging to the anesthetist. Airway obstruction and difficult intubation might lead to perioperative complications including respiratory distress, hypoxia, and respiratory failure. PRS patients are more liable to adverse effects of opioids as the chronic airway obstruction and hypoxia make them more opioid sensitive [5, 6].

The airway management for a PRS patient can be approached using a number of techniques and airway equipment. The intubation can be performed under general anesthesia, sedation, or awake. In managing an expected pediatric difficult airway under anesthesia or sedation, it is crucial to try and maintain spontaneous ventilation. Airway devices and equipment that can aid the intubation and/or ventilation in a PRS patient include LMA, video laryngoscopes, fibre-optic scope, and retrograde wire [2, 7, 8].

In the setting of severe respiratory distress and impending failure, it is possible to place an LMA without anesthesia or sedation. This was described by Markakis et al. [9] in 1992, and then, in 2008, Asai et al. [10] reported a series of 5 PRS neonates in whom the LMA was placed without sedation or local anesthesia. The authors reported that all 5 neonates (2.8–3.5 kg) became calm after LMA placement and airway patency restored. Another report by Stricker et al. [11] described placing an awake LMA in PRS patients before anesthesia induction followed by fiber-optic scope introduction to facilitate endotracheal tube placement.

If general anesthesia is induced before securing the airway, it is essential to maintain spontaneous ventilation. Maintaining the ventilation whether spontaneous or assisted can be very difficult or even impossible because of airway obstruction. Accordingly, airway adjuncts must be kept ready and an algorithm for difficult airway management must be followed. Steps in the management of difficult intubation and/or difficult ventilation include call for help, 2-handed jaw thrust, using airways, to aid ventilation, whether oral pharyngeal or nasopharyngeal or LMA. Trials of endotracheal intubation of the patient should be performed, in case of refractory airway obstruction despite these maneuvers.

Emergency rigid bronchoscopy by Otolaryngologist in case of failed intubation. The final step in the "cannot intubate or cannot ventilate" scenario is emergency tracheostomy. Another rescue option is extracorporeal membrane oxygenation but is available only in specialized centers, might need time, and is suitable only for patient who weigh more than 2 kg and are more than 34 weeks of age.

Proper laryngoscopy skills for pediatric patients are a prerequisite for anesthesia management of PRS cases. In 1997, Henderson [12] described a paraglossal approach for intubation of PRS patients.

In 2008, Semjen et al. [13] used this technique in trying to intubate 6 patients with PRS. They successfully intubated 5 of the 6 PRS patients. In this technique, the blade of the laryngoscope is introduced in the right corner of the mouth and advanced using leftward and anterior pressure displacing the tongue to the left. The distance to the opening of the glottis is reduced using this technique to facilitate intubation.

A fiber-optic scope can be introduced through an LMA or nasally. A nasopharyngeal airway can be used in such cases, in the opposite nares, to deliver oxygen with or without anesthetic gases. If the patient is too small for the endotracheal tube to be mounted on the fibre-optic scope, the endotracheal tube can be placed orally while the scope is introduced nasally to visualize the glottic opening and the tube is advanced under vision [14].

Conclusion

A PRS patient with a complete cleft palate had respiratory distress and impending respiratory failure secondary to airway obstruction. She was successfully intubated with a hybrid airway technique using classic LMA and flexible intubating fiberscope.

Statement of Ethics

Written informed consent was obtained from the patient mother for publication of this case report and any accompanying images. Ethical approval is not required according to Dubai Health Authority Committee policies.

Conflict of Interest Statement

The authors have no conflicts of interest to declare.

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Author Contributions

Ahmed Hashey contributed to the manuscript, discussion, and conclusion. Nadine Nour contributed to the abstract and conclusion

Data Availability Statement

All data generated or analyzed during this case report are included in this article. Further inquiries can be directed to the corresponding author.

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