Case Report

Interim management of Pierre Robin sequence using a custom-made face mask

ABSTRACT

Pierre Robin sequence poses a great challenge for anesthesiologists during laryngoscopy and intubation, making oxygenation and ventilation difficult. The role of early surgical intervention is recommended for the improvement of the airway and overall survival of the neonate. The situation becomes even more challenging, when the neonate may not be fit for such surgical interventions. The present case posed such a challenge to the team. To the authors' knowledge, the decision to use a face mask as an interim life-saving measure was considered for the first time. This provided a greater window of opportunity for further course of action, only to be later managed by distraction osteogenesis of the mandible. The unconventional use of orthopedic appliances for the management of threatened airways may provide the clinician with time, where further management may be carried out. The present article will explain such a procedure that was carried out as a life-saving measure.

Keywords: Face mask, Interim management, Pierre Robin sequence

INTRODUCTION

Pierre Robin sequence (PRS) is classically an anomaly affecting the neonatal maxillofacial complex, presenting as a deformity associated with micrognathia, glossoptosis, and airway obstruction.[1] A cleft palate is also found in roughly 90% of patients with PRS.[2] While the sequence is relatively rare with a reported prevalence ranging from 1:2,000 to 1:30,000 of live births, the mortality rate in the condition has been reported to be as high as 30%.[3] The most concerning findings in PRS patients are micrognathia and the retroposition tongue; this lethal combination is bound to compromise the airway, leading to respiratory distress and obstruction of the airway and also a failure of neonates to thrive. Nevertheless, severe dyspnea immediately after birth is unusual. The immediate management of such anomalies poses a challenge to surgeons, neonatologists, and anesthesiologists. It may sometimes not be possible due to smaller body size, lesser blood volume, and difficulty securing the airway owing to a smaller size. The issues may be compounded if the neonate is premature. We faced one such difficulty in the management of a case of PRS, where interim measures like the placement of face mask were performed to allow more time and increase the window of opportunity prior to managing with a definite treatment plan of performing distraction osteogenesis.

CASE REPORT

A 1-day-old 2.2 kg term neonate born after normal vaginal delivery presented with severe dyspnea immediately following birth in a tertiary-level teaching hospital. The newborn presented with classical symptoms of PRS, namely, micrognathia, cleft palate, and upper airway obstruction. There were signs of airway obstruction which did not improve

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with a change in position. Manual bag-mask ventilation with high oxygen concentration and the use of airway adjunct devices was ineffective, neonate received an injection of midazolam. One attempt at intubation by an experienced physician in emergency glottis could not be visualized, with rapid desaturation at repeated attempts, a total of three attempts at intubation were made. Given losing control over ventilation, an emergency call was made to pediatric anesthesia and the otolaryngology team for definitive airway control. Immediate management was to relieve the upper airway obstruction, which was carried out using a supraglottic airway device (SAD) in this case I—gel was used. The device could not be placed *in situ* for a longer time. To avoid a permanent scar, tracheostomy was not considered the primary choice for management. The oral and maxillofacial team was consulted and a plan to carry out bilateral mandibular body osteotomy and placement of intraoral distractors was planned. However, due to the neonate's general debilitating condition, considering the surgery under general anesthesia was considered a high-risk procedure. The plan was to consider an interim option to allow for systemic improvement and also to relieve upper airway obstruction. Hence, it was planned to place a customized face mask that would allow for forward positioning of the mandible and aid in upper airway improvement. The device was fixed to the forehead and the chin and secured using conventional IMF screws placed under local anesthesia and continuous traction using an e-chain elastic [Figure 1]. There was a scant improvement in the neonate's saturation, but it increases the window of time for the pediatric team to work on improving the condition and make the neonate worthy of a definitive surgical procedure. On the 7th day, the child was considered for surgery; under general anesthesia, a preoperative computed tomogram was obtained for vector planning [Figure 2]. The neonate was operated on bilateral body osteotomy and placement of intraoral distractors. The child was placed in a latency period of 72 hrs, and later, the distraction period was carried out for 8 days with a frequency of 0.25 mm four times daily. The consolidation period was for 3 months; later, distractor removal was done under general anesthesia. The postoperative period was uneventful. Postoperative computed tomogram was suggestive of elongation in the length of the mandible with the formation of mixed density in the body of the mandible suggesting the formation of neo-calcification [Figure 3]. The child showed drastic improvement in facial appearance and overall oxygen saturation.

One of the dreaded airway-related syndromes in children is the Pierre Robin sequence characterized by the classic triad of micrognathia, glossoptosis, and cleft palate.[4] It

Figure 1: Pre-op placement of face mask

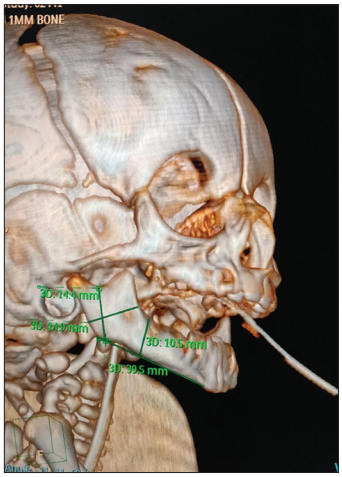

Figure 2: Pre-op CT scan

Figure 3: Post-op CT scan

poses challenges for anesthesiologists during laryngoscopy and intubation, making oxygenation and ventilation often difficult. An encounter with PRS in a neonate for airway management is even more challenging, where the reported incidence of difficult laryngoscopy (in neonates) is around 3.2%. In these children, progressive airway obstruction is generally prevalent in the second month of life.[5] However, there are no specific guidelines in the literature for anticipated and/or unanticipated difficult airway management

in neonates. Engelhardt T *et al*.[6] in 2019 proposed a step-wise algorithm and universal approach to airway management in pediatrics based on adult difficult airway society guidelines. In the event of upper airway obstruction, the mainstay options for immediate relief of obstruction are 2-handed jaw thrust and the use of airway adjuncts like oropharyngeal or nasopharyngeal airway and SAD. The role of SADs is well emphasized in the management guidelines for difficult airways in children and adults, as well as during neonatal resuscitation at the time of birth.[7] The use of such devices is still not definitive and may be associated with dislodgement and failure. The use of tracheostomy has been emphasized by Demke *et al*.,[4] in which 15 out of 31 neonates needed tracheostomy and decannulation after a mean period of 28 months. Another well-accepted technique is mandibular distraction osteogenesis, which can be a safe and effective way to avoid tracheostomy placement in selected neonates with the Pierre Robin sequence, with or without other associated syndromes. The cost to patients and families appropriately treated by mandibular distraction osteogenesis is substantially less than tracheostomy. Mandibular distraction does not involve ongoing maintenance, medical care, and a high risk of mortality associated with a tracheostomy. A support team of pediatric anesthesia, pulmonary, and otolaryngology specialists is required for the safe and successful selection and treatment of this group of patients with the Pierre Robin sequence using mandibular distraction. However, in the present case owing to the systemic condition the neonate could not be immediately taken under GA for placement of distractors. It was decided by the oral and maxillofacial team that a myofunctional appliance like a Petit face mask could be used to improve the window opportunity for further improvement of the general condition of the neonate. This innovative idea helped the team to buy time and improvise the treatment plan to make the neonate fit to receive general anesthesia. This has not been discussed in the literature as per the knowledge of the authors. Once the clearance was obtained and made fit as per ASA 3 for difficult intubation, the placement of the distractor was done which has already been discussed. The use of myofunctional appliances can be considered an alternative method of management. If the improvisation of such alternatives is achieved, then maybe the management of PRS can become more conservative in the future.

Declaration of patient consent

The authors certify that they have obtained all appropriate patient consent forms. In the form, the legal guardian has given his consent for images and other clinical information to be reported in the journal. The guardian understands that names and initials will not be published and due efforts will be made to conceal identity, but anonymity cannot be guaranteed.

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Conflicts of interest

There are no conflicts of interest.

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