Respiratory Failure in a Neonate with Pierre Robin Syndrome -A Challenging Proposition

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ABSTRACT

Pierre Robin Syndrome is a well-known difficult airway condition. We present a case of successful management of a neonate in severe respiratory distress with diagnosed Pierre Robin syndrome, pneumonitis and in impending respiratory failure. We wish to highlight a sequence of measures instituted for the child including use of a supraglottic device Igel for immediate airway rescue when intubation was nearly impossible and mask ventilation very difficult. We sutured patient's tongue with his lip near the chin to keep the upper airway patent as an intermediate intervention and we could subsequently successfully treat the concomitant lung infection. Child could be discharged from hospital alive and healthy.

KEY WORDS

Airway, igel, neonate

INTRODUCTION

Pierre-Robin syndrome (PRS) presents many a times as a life threatening emergency. The challenges posed by anatomical changes, a difficult intubation in small babies with pneumonitis and acute respiratory distress have taken its toll on a number of children in the past.[1] After taking written informed consent from parents, we present here, successful management of such an emergency in a neonate with PRS.

CASE REPORT

A twenty five day old male small for gestational age baby was brought to the neonatal intensive care unit (NICU) in an emergency because of difficulty in breathing, bluish discoloration of sole and poor feeding. Taking patient history in the neonatal intensive care unit from the parents, baby was confirmed to be full term, normal vaginal delivery who cried immediately after birth with a birth weight 2.35 kg. Baby was pink till 2nd day of life after which he developed bluish discoloration of sole, difficulty in breathing and refusal to feed. On examination, the baby was undernourished, dyspnoeic with high respiratory rate (70/min) and severe subcostal and intercostal chest retractions. (fig.1)

He was being kept prone as he could not maintain saturation in the supine position. Pierre Robin syndrome with cleft palate, micrognathia, retrognathia and glossoptosis were confirmed. Feeding was being done through nasogastric tube. No other associated congenital abnormality was documented. Breathing was labored and noisy with crepitations all over the chest and resting SpO_2 75-80% on room air. Oxygen supplementation was started immediately using an oxygen hood. His blood counts suggested acute infection with a high total count and high neutrophil count. Airway examination under anaesthesia was planned to find out other causes of respiratoy distress and noisy breathing. After taking appropriate consents and risk explanation to the parents, child was shifted to the operating room

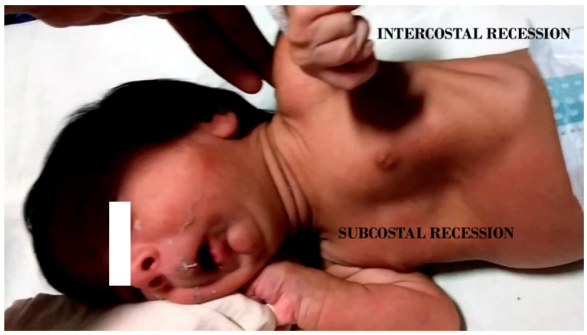

Figure 1. PRS neonate in severe respiratory distress requiring prone position to maintain saturation. Note the intercostal and subcostal retractions.

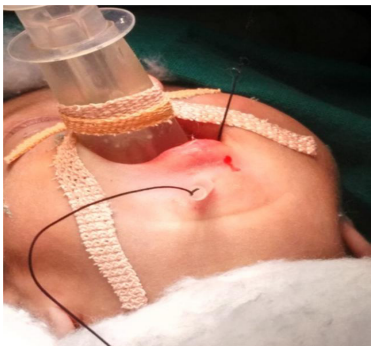

Figure 2. A non-absorbable suture bite taken from the tongue to be secured to the chin.

approximately an hour after this initial evaluation. Standard monitoring was attached and intravenous fluid infusion was started. Plan of airway management included sedating the child with a bronchodilator inhalational agent in prone position maintaining spontaneous respiration. Sevoflurane in 100% oxygen was administered through face mask, starting with 2% and gradually increasing till the jaw was perceived as relaxed. The child continued to be ventilated by face mask, with IPPV although considerable effort was required to keep the mouth open by pushing the mandible downward and forward. Two operator manual ventilation was required with oropharyngeal airway in place and despite all efforts, multiple episodes of desaturation to SpO_2 <70% occurred. When adequate depth of anaesthesia was achieved and SpO_2 stabilised at >95%, baby was turned to supine position and intubation attempted by experienced anaesthesiologist. Cormack Lehane was Grade IV with considerable use of force. Soon enough the child desaturated and he was again placed in the prone position. Three such attempts were made thereafter a decision to use supraglottic device was taken. A couple of attempts using Proseal laryngeal mask airway proved futile and after each attempt, the child had to be placed in the prone position to relieve obstruction of airways. At this time, use of IGel size No. 1 was contemplated. Use of Igel has not previously been described in literature for emergency airway management in a neonate with PRS. Igel, in this case could be inserted with ease and the fit was also found appropriate. The child could now be ventilated manually.

A fibreoptic laryngoscope could now be introduced through I-gel and airway assessment of associated airway abnormalities upto the carina was done. Arytenoids and vocal cords were seen to be slightly edematous. A decision was now made to pull the tongue out and fix to the chin. This would lessen the respiratory distress in the postoperative period. A more definitive treatment such as mandibular retraction was not possible as consent could not be obtained for the same. Two bites were taken almost midway in the tongue about an inch apart with non-absorbable sutures. One end of these sutures was then taken out between the lower lip and the chin. (fig. 2)

Both were then threaded over a segment of size 3.0 endotracheal tube and after taking them out through the opposite side, were tied over the tube. (fig. 3). This was done to prevent the cut through of the sutures. Throughout the procedure baby was on assisted ventilation and no muscle relaxant was given. Intravenous fentanyl 05 microgram and paracetamol 10 mg was given for pain relief. After the procedure, sevoflurane was gradually decreased; baby was brought out of anaesthesia. When baby regained adequate tone and his tidal volume was satisfactory, I gel was removed and baby was given oxygen by face mask. The child was put prone again after the procedure in an oxygen hood with oxygen flow at 10 lt/min and was observed for 45 minutes. Post operatively with a respiratory rate of 56/min, SpO_2>95% on oxygen hood, the baby was shifted to NICU in the prone position. Feeding was started after 6 hrs through nasogastric tube. The baby was monitored daily for signs of respiratory distress and gain in weight. The baby began to tolerate supine positioning in a progressively better manner in the following days. The pneumonitis was simultaneously managed. The tongue suture gave way on 10^{th} postoperative day. He remained in the NICU till 70^{th} day of life after which he was successfully discharged. At the end of his hospital stay he had gained weight to 2.73 kg, was comfortable in supine position with RR=40/min, SpO_2 of 96-98% on room air and was accepting spoon feeding.

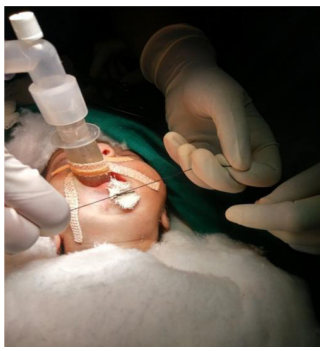

Figure 3. The tongue pulled away from the pharyngeal wall and secured by means of non-absorbable sutures near the chin. Note the arrangement of a small diameter endotracheal tube over the gauze piece on the chin.

DISCUSSION

Children with Pierre Robin syndrome often present with respiratory distress and feeding difficulties soon after birth. Due to the anatomical airway abnormalities micro-retrognathia and glossoptosis, the large tongue falls back in the supine position and causes upper airway obstruction preventing normal respiration and leading to hypoxemia.3 If the obstruction is unrelieved for a long time, chronic hypoxia may lead to cor pulmonale and right heart failure.[4] These children present with several anaesthetic challenges. Airway management during anaesthesia is often difficult because the airway obstruction increases when the child is turned supine. Even mask ventilation may be difficult because the receded mandible does not provide an adequate mask seal. In addition, these patients are very difficult to intubate. Sedation and muscle relaxation should not be used before securing the airway, as airway obstruction is more likely to increase when soft tissues are relaxed.[5] Due to feeding difficulties the children present with poor weight gain and signs of aspiration pneumonia. Other concerns in a neonate include maintainance of body temperature, increased sensitivity to muscle relaxants, inhalational agents and harmful effects of using 100% oxygen for long periods of time. Premature infants may have a transitional circulation and immature liver function.[6] In our case the neonate presented to the emergency with low birth weight, respiratory distress and peripheral cyanosis.

Awake fibreoptic intubation is not performed as an emergency procedure and it was impossible in this case due to the worsening respiratory distress in supine position. For the same reason, emergency tracheostomy could not be performed though we had an emergency surgical airway and standby ear nose throat (ENT) specialists ready for any eventualities. Tracheostomy in a neonate has its own inherent problems.[7]

Case reports have been described previously using classic LMA as means of securing airway in Pierre Robin syndrome.[8,9] However use of Igel has not been described. I gel has certain advantages like ease of insertion even in the prone position (keeping the head to the side). It also has a lower cost, may be used as an intubation conduit specially in such difficult airway cases. In our case we were not able to intubate the child in two laryngoscopy attempts. The baby could not be kept supine for a long period as he was rapidly desaturating. We were successful in using Igel for securing the airway in an emergency. By using a swivel connector it was possible to insert a fibreoptic laryngoscope through the Igel for airway examination and at same time ventilate the baby. With use of airway gadgets and tongue sutures we were successfully able to manage the airway subsequently treat the lung infection and save the life of this child.[10]

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