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Case Report

Anaesthetic Challenges in Children with Pierre-Robin Sequence: A Case Series

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ABSTRACT

Pierre Robin sequence is associated with a triad of micrognathia, glossoptosis and airway obstruction. Securing the airway is of utmost importance in these patients. Use of vagolytic premedication, nebulisation with lignocaine to obtund airway reflexes, use of other airway devices (SGA) and equipment to assist in different airway scenarios helps in anaesthetising the patients.

Pierre Robin sequence (PRS) is defined as the clinical triad of micrognathia, glossoptosis, and airway obstruction. Securing the airway is of utmost importance as it is complicated by cleft palate and limited mouth opening, resulting in difficult mask ventilation and intubation [1-2]. Pathogenesis is attributed to hypoplasia of the mandibular area before 9 weeks in utero causing a posterior position of the tongue that prevents palatal shelves from closing on the midline [2]. It was first documented by a French stomatologist, Pierre Robin in 1923 with an incidence of 1:8500 live births [3].

Case Series

Case report 1: A female term neonate aged 15 days weighing 2.5 kg with bilateral septic arthritis of hip and knee associated with Pierre Robin sequence and congenital heart disease (ASD - 2mm) was posted for arthrotomy and debridement. Clinical examination revealed micrognathia, retrognathia, cleft palate and glossoptosis. A difficult airway cart was prepared and GA with a supraglottic airway device was planned. Induction was done using the Jackson Rees circuit (J R Circuit) and with IV Thiopentone 5 mg.kg-1, Fentanyl 2 µg.kg-1 and Sevoflurane. Under deeper planes of anaesthesia, LMA™ Ambu No.1 with a gastric suction port was inserted. Oxygen, air and Sevoflurane mixture were used for maintenance. Standard ASA monitoring was done. Recovery was smooth and post-op uneventful.

Case report 2: A female infant aged 3 months weighing 3.5 kg, diagnosed with Hydrocephalus with Pierre Robin sequence was posted for VP shunt. The child also had an ASD with hypothyroidism. On examination, the child had retrognathia and a cleft palate. GA was planned with a difficult airway cart ready. Induction was done with J R circuit and IV Thiopentone and Fentanyl, after confirming the ability to mask ventilate, IV propofol was given in graded dose. With the child in spontaneous ventilation, laryngoscopy revealed CL grade-3, there were 2 failed intubation attempts and on 3rd attempt using shoulder roll, Micro cuff ™ tracheal tube No.3 with stylet was inserted aided by external laryngeal manoeuvre under deeper levels of sedation. After confirmation of adequate ventilation, a neuromuscular blocking agent was given. Maintenance was done by air, oxygen, Sevoflurane and Atracurium. Standard ASA monitoring was done. Recovery was smooth, the child was extubated and post-op was uneventful.

Case report 3: A male term neonate aged 12 days weighing 2.8 kg was diagnosed to have bilateral inguinal

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hernia with Pierre Robin sequence posted for right herniotomy. On examination, the neonate had micrognathia, cleft palate, short neck, spinal agenesis, left femoral agenesis and left club foot. GA with Caudal anaesthesia was planned. A difficult airway cart was prepared. Inhalation induction was done with Sevoflurane, after securing the IV-line ketamine and propofol were given. Adequate mask ventilation was confirmed and under deeper planes of anaesthesia, LMA Ambu™ size 1 was inserted. Caudal anaesthesia was given with 3ml of 0.1% plain Levobupivacaine. Standard ASA monitoring was done. Maintained with air, oxygen and Sevoflurane. Recovery was slightly delayed at the end of surgery and post-op was uneventful.

Case report 4: A female preterm neonate aged 45 days (post-conceptional age 40weeks) born at 34 weeks weighing 2.48 kg was diagnosed to have hydrocephalus secondary to meningitis, Pierre Robin sequence and ASD (L->R) was posted for VP shunt. On examination, the neonate had retrognathia and cleft palate. GA was planned. A difficult airway cart was prepared. Induction was done with J R circuit and IV Thiopentone, Fentanyl after confirmation of adequate mask ventilation, IV Propofol was given in graded dose. On spontaneous ventilation, laryngoscopy was done by an experienced anaesthesiologist with a miller blade that showed CL grade 3. After three failed attempts to intubate, the trachea was intubated with No. 3.0 Micro cuff™ tracheal tube aided with stylet, Macintosh blade with external laryngeal manoeuvre and shoulder roll. A neuromuscular blocking agent was given only after confirmation of adequate ventilation. Standard ASA monitoring was done. Maintenance was done with a Sevoflurane mixture. Extubated after adequate efforts, Apnoeic episodes occurred after extubation. Due to increased opioid sensitivity in preterm neonates and because of delayed recovery probably due to opioids, reversal was done by IV Naloxone 4mcg, after which recovery was smooth and uneventful post-op.

Discussion

Airway management in neonates and infants with craniofacial abnormalities is an anaesthesiologist nightmare. Pierre-Robin Sequence (PRS) especially during the neonatal period is very challenging due to airway obstruction by receding mandible, large tongue and cleft palate associated with feeding difficulty and recurrent respiratory infection [4]. Also, there may be more opioid sensitivity from chronic airway obstruction and hypoxia [5]. Hence, a preoperative evaluation is of utmost importance in patients with PRS. A thorough history of apnoea, respiratory complications, hospital stays, protracted intubation, tracheostomy, feeding, growth, and development history should be noted. There may also be an association with cardiovascular and neuromuscular dysfunctions [4]. Interventions like premedication with a vagolytic drug to abolish vagal hyperactivity, reducing the airway secretions [2], Nebulisation with 4% Lignocaine to provide surface anaesthesia obtunding airway reflexes for laryngospasm is useful only in bigger children [6].

Bag and mask ventilation is often challenging in PRS patients requiring Jaw thrust, nasopharyngeal or oropharyngeal airway, sometimes even a Supraglottic airway (SGA) may be used to relieve upper airway obstruction. We were able to mask ventilate our patients without using any aids.

Availability of SGA devices with a gastric port for neonates has minimal options presently, even the most popular I-gel™ has no gastric port for size 1. With PRS neonates having high-risk aspiration and difficult airway, a choice of more beneficial SGA becomes the utmost important decision in the difficult airway cart. We selected LMA Ambu™ size 1 which has a suction port for neonates and is used routinely.

Proper laryngoscopy should never be ignored when intubating PRS neonates. Henderson described a technique in PRS patients whereby using a paraglossal approach may be more effective than standard laryngoscopy [7]. Though a difficult intubation cart may be assisted with a Video laryngoscope, fibreoptic bronchoscope and other new airway devices, in our experience the LMA has proved to be the most beneficial for airway when endotracheal intubation is not required [8]. Studies by Templeton et al reported LMA™ and the Air-Q™ to be effective for airway management in patients with PRS [9]. Sevoflurane or Isoflurane can be used for maintenance. Studies also show Ketamine, Dexmedetomidine, Remifentanil as beneficial with minimal postoperative respiratory depression [4]. PRS neonates have airway concerns post-operatively too, airway obstruction can occur due to airway oedema secondary to surgical manipulation and muscular hypotonia following anaesthesia. To overcome this obstruction certain manoeuvres are helpful like prone positioning, insertion of small size tracheal tubes or nasal airway if available in suitable size through both the nostrils and/or tying the tongue to the chin or rarely tracheostomy could be performed [10]. Hence post-op monitoring is of utmost importance and should be done meticulously.

Conclusion

Safe and successful airway management in Pierre Robin sequence starts with a good preoperative airway assessment, preparation, anticipation and planned anaesthesia management. Inhalational induction with proper bag and mask ventilation, use of supraglottic airway devices wherever feasible and good post-op vigilance are essential.

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