



CASE STUDY

ANESTHETIC IMPLICATION OF 10 DAY OLD INFANT WITH PIERRE ROBIN SYNDROME POSTED FOR TONGUE LIP ADHESION

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ABSTRACT

Pierre Robin Sequence (PRS) consisting of Micrognathia, Retrognathia, Glossoptosis and cleft palate present with airway obstruction and feeding difficulties with or without other congenital anomalies. These patients come into category of difficult ventilation and difficult intubation. We present a case report of a 10 days old female child, weighing 2.5kg, suffering from PRS with cleft palate, Micrognathia, Retrognathia, Glossoptosis, airway obstruction, feeding and breathing difficulty posted for tongue lip adhesion. Thorough preoperative airway assessment was done. Use of transparent round mask and two hand jaw thrust were beneficial for ventilation of the child with receding chin. Inhalational induction was performed with Sevoflurane in oxygen. The child was ventilated with facemask. PRS patients are at risk of postoperative airway obstruction and chronic hypoxia. Use of long acting opioids was avoided and patient was kept under observation in NICU.

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INTRODUCTION

Pierre Robin syndrome (abbreviated to PRS, and also known as Pierre Robin sequence, Pierre Robin malformation, Pierre Robin anomaly) is a congenital condition of facial abnormalities in humans. PRS is a sequence, i.e. a chain of certain developmental malformations, one entailing the next. The main features are cleft palate (U shape), retrognathia (abnormal positioning of the jaw or mandible), micrognathia (small mandible) and glossoptosis (airway obstruction caused by backwards displacement of the tongue base). Above features make intubation difficult. The syndrome is generally diagnosed clinically shortly after birth. The infant usually has respiratory difficulty especially when supine. In prone or lateral position obstruction caused by tongue fall is relieved. Infants usually have difficulty in feeding and breathing. Because of chronic hypoxia and feeding difficulty there can be growth retardation. Airway obstruction is characterized by snorty breathing, apnea, difficulty taking a breath, or drop in oxygen saturation. A tongue lip adhesion is performed to bring the tongue forward, effectively opening up the airway. Mandibular distraction can be effective by moving the jaw forward to overcome the upper airway obstruction caused by the posterior positioning of the tongue.

Case report

A 10 days old female child weighing 2.5kg known case of PRS, posted for tongue tie adhesion. Patient was born with caesarian section because of Non progress of labour. Patient was referred to civil hospital for congenital anomaly where she was diagnosed with PRS. Patient developed upper airway obstruction with chest wall retraction many times. On examination she was conscious, crying, moving all four limbs. Respiratory examination reveals chest wall indrawing in supine position. Cardiovascular examination reveals no abnormality. Airway examination reveals micrognathia, retrognathia and cleft palate suggestive of difficult airway. Mouth opening was adequate with midline defect in soft palate. Her hematological, biochemical and radiological investigations were within normal limit. Bed side echocardiograph was normal. Difficult airway trolley was prepared which included RBS mask, transparent masks of appropriate sizes, small size oropharyngeal and nasopharyngeal airways, laryngeal mask airway of size 1, uncuffed endotracheal tubes of sizes 2 to 4, small size stellates, cricothyroidotomy set. In the operating room monitors including pulse oximeter, NIBP, ECG and capnography were employed. She was premedicated with injGlycopyrrolate 4ug/kg, inj Ondansetron 0.08mg/kg, paracetamol 5 mg/kg. Long acting opioid was avoided. Preoxygenation was done using round mask and JacksonRee's circuit. Inhalational induction was performed with Sevoflurane in oxygen. The child was ventilated with facemask although effort was required to keep her mouth open by pushing her mandible downward and forward.

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Figure 1. Per operative airway



Figure 2. Intra-operative management



Figure 3. After tongue lip adhesim

After achievement of proper depth of anesthesia, surgeon were asked to start surgery. In between surgery child had to be ventilated with oxygen and sevoflurane once. As surgery was of short duration, it was done with sevoflurane only. Surgeon took care of minimal blood loss, proper suctioning and they kept gauze piece between teeth to prevent any blood going in mouth cavity. Intraoperative steroid was given. Post operatively she was vitally stable, maintaining saturation. She was conscious, crying and moving all four limbs. She was sent to NICU postoperatively for post-operative care and monitoring.

DISCUSSION

Patients with PRS require anaesthesia for a variety of procedures including Direct Laryngoscopy and Bronchoscopy, Tongue Lip Adhesion, Mandibular Distraction Osteogenesis, tracheostomy, radiologic procedures, gastrostomy tubes, and Nissen fundoplication. Micrognathia, retrognathia, cleft palate, glossoptosis can present significant challenges for the anaesthesia provider including airway obstruction and difficult intubation. Preoperative evaluation is of utmost importance in patients with PRS. Anaesthetist should obtain full history of apnea (central and/or obstructive), respiratory complications, hospital stays, protracted intubation, tracheostomy, feeding, growth, and development. PRS may be associated with cardiovascular (corpulmonale, vagal hyperactivity) and neuromuscular (brainstem dysfunction, central apnea) dysfunctions. Facial abnormalities give an idea about obstructive apnea and respiratory distress. The clinical findings may be supported by various studies including sleep monitoring for episodes of spontaneous oxygen desaturation, desaturations during feeding, during sleep and during phonation. Nasoendoscopy and bronchoscopy are invaluable adjuncts to determining the site of airway obstruction, as there

may be more sources of airway compromise than the tongue base itself, such as in laryngomalacia, tracheomalacia, or other subglottic obstructions. Part of the assessment should also include evaluation of patients in different positions and how well positioning resolves upper airway obstruction. Plain X-ray, CT scan or MRI may be useful to diagnose of the cause or site of obstruction or bony or soft tissue. Premedication must include a vagolytic drug as these patients are prone to vagal hyperactivity and to reduce the airway secretion. Chronic hypoxia induced pulmonary hypertension and airway obstruction can lead to many complications. So, she was posted for tongue lip adhesion. Based on airway examination intubation was suspected difficult. The procedure was short. Surgeon assured minimal blood loss, proper suctioning and prevention of blood going to oral cavity by keeping gauze piece between teeth. That's why surgery was done under sevoflurane only without intubation. But it is always advisable to do surgery after securing airway.

Conclusion

A thorough airway assessment, preoperative preparation, inhalational induction postoperative vigilance are important in management of patients with Pierre Robin Sequence.

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