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Dr. Niranjni S
Final Year Postgraduate, Department of Anaesthesiology, Saveetha Medical College and Hospital, Kuthambakkam, Tamil Nadu, India

Dr. Harish Kumar J
Final Year Postgraduate, Department of Anaesthesiology, Saveetha Medical College and Hospital, Kuthambakkam, Tamil Nadu, India

Dr. Lakshmi
Head of the Department of Anaesthesiology, Saveetha Medical College and Hospital, Kuthambakkam, Tamil Nadu, India

Corresponding Author:
Dr. Niranjni S
Final Year Postgraduate, Department of Anaesthesiology, Saveetha Medical College and Hospital, Kuthambakkam, Tamil Nadu, India

Anaesthetic management of a infant with Pierre robin sequence

Dr. Niranjni S, Dr. Harish Kumar J and Dr. Lakshmi

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Abstract
Pierre Robin sequence (PRS) is characterized by the clinical triad of micrognathia (small mandible), glossoptosis (backward, downward displacement of the tongue), U or V shaped cleft palate and airway obstruction defines the Pierre Robin sequence (PRS). Airway obstruction and respiratory distress are clinical hallmarks. It occurs in 1:8500 live births with an equal male-to-female ratio. The mechanical theory which states that initiating event is mandibular hypoplasia which keeps the tongue high within the oral cavity; this in turn causes a cleft palate by preventing the closure of the palatal shelves, this syndrome is now called as Pierre robin sequence. These cases pose a challenge to the anesthetist as they present at a very young age, may have associated congenital anomalies, varying degrees of difficult airway, need repeated corrective surgeries, and lastly the airway has to be shared with the surgeons.

Keywords: Pierre robin sequence (PRS), difficult airway

Introduction
Cleft lip and palate (CLP) is one of the most common congenital malformations, associated with difficulty in feeding, speech development and facial disfigurement. Depending on the extent of the defect, different surgical manipulations may be required in various stages such as early orthopedic manipulation of the dental arch followed by cleft lip repair, palate repair surgery for velopharyngeal incompetence and maxillary advancement at a later stage. These cases pose a challenge to the anesthetist as they present at a very young age, may have associated congenital anomalies, varying degrees of difficult airway, need repeated corrective surgeries, and lastly the airway has to be shared with the surgeons.

Case details
A 11 months old girl of 7 kg presented with complaints of inability to feed, difficulty in phonation, deformity in face, with complaints of snoring. She was born my FTND and there was no history of cyanotic spells since birth. No history of repeated URTI, regurgitation. No history of frequent ear infections. No history of pulmonary complications. Immunized up to date. History Consanguineous marriage of her parents. Normal developmental milestones. She was posted for cleft palate repair and cheiloplasty.

General examination: Alert, active infant complete unilateral cleft lip and palate. No plallor, not icnteric, no cyanosis, no signs of heart failure. Neck movements normal. Mallampatti classification could not be elicited.

Vitals: PR – 98 bpm, RR – 22/ min, SPO2 – 99%, afebrile.

Systemic examination: RS – bilateral air entry present. No added sounds. CVS – S1 S2 present, no murmurs.
P/A – soft, non-tender. CNS – slert, active, moving all four limbs.
All blood investigations were normal. 2D ECHO – normal.
COVID 19 – negative. CXR – normal

Fig 1: Child with micrognathia

Anesthetic management

- Patient was kept nil per oral for four hours.
- On the day of surgery an intravenous line secured by 24G cannula in left hand.
- Premedicated with Inj. Glycopyrrolate – 0.07 mg, inj. Midazolam – 0.3 mg, inj. Emeset – 0.7 mg, inj. Fentanyl – 14 mics.
- ECG, SPO2, NIBP, end tidal capnometry, precordial stethoscope, temperature monitored by rectal probe were attached after shifting the patient inside the operation theatre
- The patient was preoxygenated with 100% 02 for 5 mins using facemask. Inj. Propofol 14 mg IV given.
- Bag and mask ventilation was tired and found to be adequate.
- Inj. Succinylcholine 2mg/kg IV given. IPPV given for 30 seconds
- Laryngoscopy with miller blade no 1 was used.
- Larynx was not visualised, hence a backward, upward, rightward pressure was given which makes larynx visible. Finally, trachea was intubated with RAE endotracheal tube no 3.5.
- Bilateral air entry checked, ETCO2 confirmed and tube was fixed in midline at 9 cm
- Throat packing was done. Inj. Dexa 0.7 mg IV given. Maintenance of anesthesia is best done with Sevoflurane
- Inj. Atracurium 3.5 mg IV given and top up of 0.7 mg IV given. Intraop vitals were stable. Blood loss of 30 ml. Total IV fluids of 140ml given.
- Throat pack was removed. Paracetamol suppository – 80 mg for post op analgesia.
- Left infraorbital nerve block was given with 1 ml of 0.25% bupivacaine.
- Patient was reversed with 0.35 mg of neostigmine and 0.08 mg of glycopyrrolate. Extubation was uneventful.
- Post extubation was normal and patient was shifted to PICU for further monitoring.

Fig 3: Image of intubated child

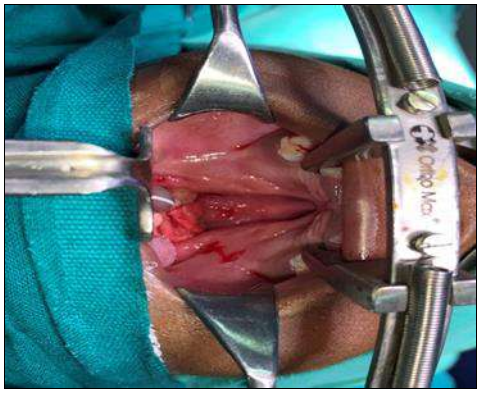

Fig 4: Image showing cleft palate

Discussion

Pierre Robin sequence (PRS) was originally described as consisting of micrognathia (which he termed "mandibular hypotrophy") and glossoptosis (an abnormal posterior placement of the tongue), which result in airway obstruction and feeding difficulties. Family members of PRS infants have a higher incidence of cleft lip and palate Cleft palate is associated with deletions on 2q and 4p, and duplications on 3p, 3q, 7q, 78q, 10 p, 14q, 16p, and 22q. Micrognathia is associated with deletions in 4p, 4q, 6q, and 11q, and duplications on 10q and 18q. Pierre Robin sequence is related to several other craniofacial anomalies and may appear in conjunction with the findings characteristic of several different syndromes. Infants with PRS should be evaluated in a team setting to assess the anatomic findings, delineate the source of airway obstruction, and address feeding issues to maximize growth and minimize obstruction. Anesthetist 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 (cor pulmonale, vagal hyperactivity) and neuromuscular (brainstem dysfunction, central apnea) dysfunctions. Facial abnormalities give an idea about obstructive apnea and respiratory distress. A multidisciplinary approach is ideally suited for this task, consisting of specialists from plastic and reconstructive surgery, pediatric otolaryngology, pediatric pulmonology, speech pathology, nursing, pediatric anesthesia, and neonatology. Preoperative evaluation is of utmost importance in patients with PRS.

Management of airway in infants and children with craniofacial abnormalities present difficult airway access for an anaesthesiologist. Airway obstruction is the hallmark of PRS due to its cardinal features of receding mandible, large tongue and cleft palate. Apart from obstruction in airway, difficulty in feeding and recurrent respiratory infections are of primary concerns in patients with PRS. Pre-operative evaluation is of utmost importance, as PRS may be associated with cardiovascular (PDA, right heart failure, increased vagal activity), neuromuscular (central apnoea), skeletal and soft tissue abnormalities.

In 60% of patients, PRS is associated with Stickler, Velocardiofacial, Treacher-Collins syndromes. Pre-operative echocardiography should be performed if any of these syndromes are suspected. Further assessment includes examination of patient in various positions and which position resolves upper airway obstruction. Prone position relieves airway obstruction in 70% of patients. Radiological

tests may be performed to evaluate bony or soft tissue abnormalities. “Normal maxillo-facial angle is less than 90°, if the angle is more than 100° then visualization of glottis becomes difficult with direct laryngoscopy”.

Premedication with glycopyrrolate or atropine can decrease airway secretions and vagal hyperactivity. Surface anaesthesia of airway with 4% lignocaine nebulisation prevents holding of breath and laryngospasms during intubation. Ventilation may also be difficult in PRS patients. Shirley D’Souza *et al*. suggested the use of dexmedetomidine for intubation due to its analgesic and sedative-hypnotic effects with minimal respiratory depression.11 Jaw thrust, nasopharyngeal or oropharyngeal airway or LMA may be used to relieve upper airway obstruction. Difficult intubation can be assisted with fiberoptic bronchoscope, Glidescope, Air-Q, Airtraq, retrograde wire, LMA.

Tariq Hayat *et al*. reported a 2 year old child with PRS where intubation was done using Air-Q intubating LMA. Parul Mallick *et al*. reported a case or twenty one month old boy with PRS scheduled for cleft palate repair where endotracheal intubation was done through a LMA using a modification of adult intubating stylet. Mukhopadhyay conducted a study on six paediatric patients with PRS and Treacher-Collins syndrome in which successful intubation was done by pulling tongue forward following induction. Maintainence of anaesthesia is done with sevoflurane as well as isoflurane. It may be added with ketamine, dexmedetomidine, remifentanil, which causes minimal postoperative respiratory depression.

Post-operatively, airway edema leading to airway obstruction secondary to surgical manipulation and muscular hypotonia following anaesthesia or closure of palatal cleft is of prime concern. As this can lead to “hypoxia, negative pressure pulmonary odema and death”. This can be prevented by maneuvers like prone position, insertion of nasopharyngeal airway, tying the tongue to the chin or tracheostomy.

Conclusion

Syndromes with craniofacial abnormalities can be a real challenge in terms of airway management. The key to success is effective preparation, presence of personnel with expertise in difficult pediatric airway management, regular training and familiarity with the difficult intubation equipment, teamwork, and following simple algo - rhythms for difficult airway management. A thorough airway assessment, preoperative preparation, inhalational induction and Para glossal technique of intubation and postoperative vigilance are the common denominators in successful and safe airway management patients with Pierre Robin Sequence. The optimal management for these patients would comprise utilization of familiar equipment and modifying techniques to enable successful airway control while avoiding airway complications.

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