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Airway Management for Pierre Robin Sequence: An Anesthetic Challenge.

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ABSTRACT

Pierre Robin Sequence (PRS) consisting of micrognathia, 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 3years old female child, weighing 9.5kg, suffering from PRS with right lacrimal sac abscess and history of recurrent failed intubation, feeding difficulty along with failure of development of speech was posted for endoscopic DCR. Thorough preoperative airway assessment was done using COPUR scale and NCCT neck for detail airway dimension. Use of silicone round mask and two hand jaw thrust were beneficial for ventilation of the child with receding chin. The spontaneous ventilation was maintained till the airway was secured by performing inhalational induction with Sevoflurane followed by intubation using paraglossal approach of direct laryngoscopy with Miller's blade 1 and uncuffed ETT no 4.PRS patients are at risk of postoperative airway obstruction and respiratory depression. Anticipating this catastrophe steroid was administered intra-operatively, use of long acting opioids was avoided and patient was kept under observation in PACU for 24 hours after extubation.

Keywords: Airway obstruction, Difficult ventilation, Difficult intubation, Paraglossal Technique, Pierre Robin Sequence.

INTRODUCTION

One of the dreaded airway disorders is Pierre Robin Sequence (PRS) also called as Pierre Robin Syndrome. It is defined as a combination of micrognathia, glossoptosis and cleft palate.[1] Hypoplasia of the mandibular area prior to 9 weeks in utero causes a posterior position of the tongue that prevents palatal shelves from closing on the midline.[2] Pierre Robin Sequence was first described in 1923 by Pierre Robin, a French somatologist and is estimated to affect 1 in 8500 live births.[3] PRS patients have varied presentation and in order to determine the appropriate treatment patients are categorized based on the severity of airway obstruction.[1]

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CASE REPORT

A three year old female child weighing 9.5kg suffering from PRS, with right lacrimal sac abscess was posted for endoscopic dacroycystorhinostomy (DCR). She was a diagnosed case of PRS and had feeding difficulty along with mild upper airway obstruction. She preferred sleeping in prone position and repeatedly inserted her right hand fingers in her mouth probably to pull her tongue forward to relieve the airway obstruction. She was fed with nasogastric tube since birth. She was posted for cleft palate repair surgery twice but the surgery got postponed in view of failed intubation. She had swelling on medial side of both the eyes and complaint of watering from both eyes since birth.

On examination, her chin appeared receding. Mouth opening was adequate with midline defect in hard palate. COPUR scale score was 12. Infant feeding tube no 8 was in situ. No other congenital anomaly was detected.

Her haematological, biochemical and radiological investigations were WNL. Room air SpO_2 was 100 %. Incidental finding in MRI of orbit and sinuses showed narrowing of supraglottic airway with diameter of 4mm. Hence NCCT neck was done for detail airway dimensions and supraglottic, glottic and subglottic diameters were 6mm, 2mm and 8mm respectively.

Difficult airway trolley was prepared which included RBS mask, silicone masks, conell masks of appropriate sizes, small size oropharyngeal and nasopharyngeal airways, laryngeal mask airway of size 1.5 and 2, uncuffed endotracheal tubes of sizes 2

to 4, small size stellates, pediatric fibreoptic bronchoscope, rigid bronchoscope, cricothyroidotomy set and tracheostomy kit.

In the operating room monitors including pulse oximeter, ECG, NIBP and capnography were employed. Patient was nebulised with 1.5 ml 4% Lignocaine 15min prior to induction. She was premedicated with inj Glycopyrrolate 4ug/kg, inj Ondansetron 0.08mg/kg, inj Midazolam 0.03mg/kg and inj Fentanyl 1 ug/kg. Preoxygenation was done using round silicone mask and Jackson Ree'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. When adequate depth of anaesthesia was achieved, direct laryngoscopy was done with Miller's blade no 1 and paraglossal approach was used to visualize the glottis. When glottis was visualized inj Propofol 1 mg/kg i.v. was administered. Patient was intubated with uncuffed ETT no 4 and throat packing was done. The child was paralyzed with 0.1mg/kg Vecuronium and anaesthesia was maintained with O_2, N_2O (50:50), Sevoflurane 2-3% and inj Vecuronium 0.02mg/kg.

Intraoperative vitals remained stable. Inj Paracetamol 10 mg/kg i.v, Paracetamol suppository, Inj Hydrocortisone 2mg/kg and Inj Dexamethasone 0.25 mg/kg were administered towards the end of surgery. On resumption of spontaneous breathing patient was reversed and extubated smoothly. She was observed in PACU for any airway obstruction postoperatively for 24 hours.

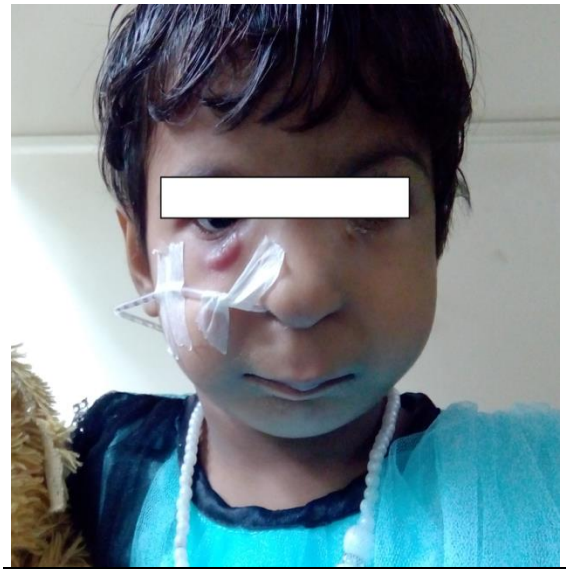

Figure 1: Preoperative photograph showing receding chin

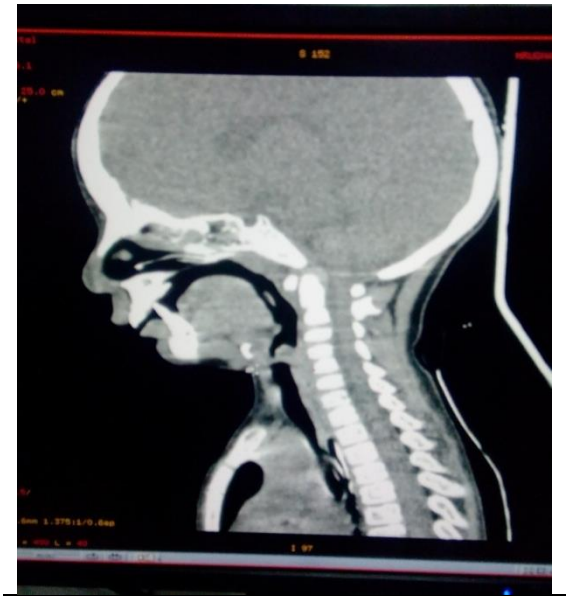

Figure 2: NCCT for neck reported supraglottic, glottis and subglottic diameters to be 6mm, 2mm and 4 mm respectively.

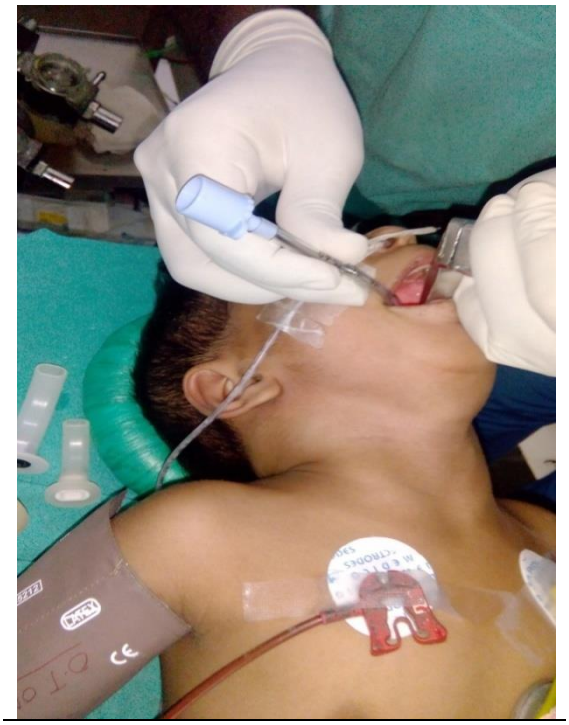

Figure 3: Paraglossal technique for intubation with Miller's blade no 1.

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. The clinical triad of PRS can present significant challenges for the anaesthesia provider

including airway obstruction and difficult intubation. [1]

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 (cor pulmonale, vagal hyperactivity) and neuromuscular (brainstem dysfunction, central apnea) dysfunctions. Facial abnormalities give an idea about obstructive apnea and respiratory distress. [2]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.[4] Plain X-ray, CT scan or MRI may be useful to diagnose of the cause or site of obstruction or bony or soft tissue abnormalities.[5] Maxillo-facial angle is measured on lateral x-ray is normally less than 90^{o}, if the angle is greater than 100^{o} then it is difficult to visualize the glottis during direct laryngoscopy.[6] In our case detailed airway examination was done with COPUR scale (Chin, Opening of mouth, Previous H/O intubation/OSA, Uvula, Range of neck movement) and NCCT neck was done for detail airway dimensions.

Premedication must include a vagolytic drug as these patients are prone to vagal hyperactivity and to reduce the airway secretion.[2] Nebulisation with 4% Lignocaine provides surface anesthesia to airway, prevents breath holding and laryngospasm in response to intubation and also provides preoxygenation.[5] Preoperative sedative administration is a matter of choice of the anaesthetist.[1]

The ventilation may be also difficult in the patients with PRS in view of retrognathia. To relieve the obstruction two handed jaw thrust, nasopharyngeal airway, oropharyngeal airway, LMA may be used. If patient cannot be ventilated then go for laryngoscopy and intubation, if not successful emergent bronchoscopy with rigid bronchoscope is done by otolaryngologist to ventilate the patient. Emergency tracheostomy and Extracorporeal membrane oxygenation are the last resorts in pediatric cannot ventilate cannot intubate situation.[1] In the above case silicone round mask and two hand jaw thrust were found to be beneficial for ventilation in view of retrognathia.

Intubation can be performed using various techniques without sedation, with sedation or under general anesthesia. The hallmark of managing the pediatric difficult airway is maintaining the spontaneous ventilation.[1] Shirley DSouza et al used Dexmeditomedine and Ketamine for intubation and emphasized on the use of Dexmedetomidine which is a highly selective alpha 2 agonist, taking advantage of its sedative-hypnotic and analgesic effects with minimal respiratory depression.[7]

Several devices can be used to assist the difficult airway in PRS including fibreoptic bronchoscope, retrograde wire, Glidescope, Sikhani scope, Air-Q scope, Airtraq, LMA[1]. In a study by Rasch DK et al (1986) suggested that children with obstructive symptoms should have laryngoscopy prior to anaesthetic induction. If the glottic opening is visualized, inhalational induction can proceed. If the glottic structures cannot be visualized, then the anaesthetist must choose between awake oral or nasal intubation, elective tracheostomy, or fiberoptic intubation.[8]

Proper laryngoscopy skills are essential when intubating patients with micrognathia and paraglossal technique helps in visualization of glottis by reducing the distance to glottis opening.[1]P. Mukhopadhyay et al (1992) did study on 6 pediatric patients with PRS or Treacher Collins Syndrome and they performed intubation with a assistant pulling the tongue forward after induction followed by direct laryngoscopy and intubation.[9]

If a fiberoptic scope is used, it can be placed orally through an LMA or nasally to visualize the glottic opening.[1]Marston AP et al (2012) conducted a study on 23 neonates with PRS demonstrated that endotracheal intubation could be done safely using conventional laryngoscopy in 37 % patients and in rest 63 % flexible fibreoptic bronchoscopy assisted intubation was used successfully.[10]Tariq Hayat Khan et al (2013) reported the intubation of a two year old child with PRS by using Air-Q intubating laryngeal mask airway.[11]Parul Mullick et al (2005) reported a case in which LMA insertion along with the use of a modified adult intubating stylet facilitated blind endotracheal intubation in a twenty one month old boy with Pierre Robin syndrome posted for cleft palate repair.[12]

Maintenance of anesthesia is best done with Sevoflurane though Isoflurane can also be used. Inhalational anaesthetic may be supplemented with an ultra short acting opioid like Remifentanil or a alpha two agonist as the post operative sedation can be avoided so that there is no post operative respiratory depression as these patients are opioid sensitive.[1] Rasch DK et al recommended use of only a potent inhalational for maintenance of anaesthesia in such patients.[8]

The postoperative concerns occur secondary to airway obstruction due to reduction of the airway by the operation and the muscular hypotonia following anesthesia and surgery or closure of palatal gap may also cause airway obstruction. Some maneuvers

described to overcome this obstruction include prone positioning, insertion of small size endotracheal tubes or nasal airway if available in suitable size through both the nostrils and/or tying the tongue to the chin or doing tracheostomy.[1] P. Mukhopadhyay advocate postponement of closure of cleft palate in patients with PRS for longer duration than ordinary cases of cleft palate alone in view of serious postoperative airway obstruction in these patients.[9] In the study conducted by Tariq Hayat Khan et al postoperatively the surgeons had to tie the tongue at two points to the alveolar ridges in view of respiratory obstruction occurring post operatively.[11] We have used steroids to minimize the possibility of postoperative airway edema and have avoided use of long acting opioids intraoperatively and postoperatively. We would also stress upon postoperative monitoring in these patients for signs of airway obstruction.

CONCLUSION

A thorough airway assessment, preoperative preparation, inhalational induction and paraglossal technique of intubation and postoperative vigilance are the common denominators in successful and safe airway management patients with Pierre Robin Sequence.

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REFERENCES

1. Cladis, Franklyn, Kumar Anand, Grunwaldt Lorelie, Otteson Todd, Ford Mathew, Losee Joseph E. Pierre Robin Sequence: A Perioperative Review. Anesthesia and Analgesia. August 2014; 119(02): p 400-412.
2. Bissonnette B,Bruno Bissonnette, Igor Luginbuehl, Bruno Marciniak, Bernard J. Dalens. Rapid recognition and Perioperative Implication; Pierre Robin Syndrome;McGraw-Hill Education, 2006. http://www.accessanesthesiology.com
3. Heather Naumann, Andrew Pittaway: Anesthesia for an Infant with Pierre Robin SequenceandPyloric Stenosis;PBLD-Table#25. ;http://www2.pedsanesthesia.org/meetings/2010winter/syllabus/pdfs/pblds/Table%20-%2025.pdf
4. Noopur Gangopadhyay et al. Pierre Robin Sequence. Semin Plast Surg. 2012 May; 26(2); 76-80.
5. Yosha Prasad: ATOTW 250 – The Difficult Paediatric Airway; www.totw.anaesthesiologist.org, 13/02/2012: Page 1 of 13.
6. D.J. Hatch: Airway management in cleft lip and cleft palate surgery; BJA. 1996; 76: 755-756.
7. Shirley Dsouza et al. Intubation using dexmedetomedine and ketamine in a child with Pierre Robin sequence. J Anaesthesiol Clin Pharmacol. 2012; 28(4):545-6.
8. Rasch DK et al. Anesthesia for Treacher Collins and Pierre Robin syndromes: a report of three cases. Can Anesthesia Soc J. 1986 May; 33(3 pt 1); 364-70.
9. P Mukhopadhyay, P Mukherjee, M. Adhikari. Problems in Anesthetic Management of Pierre Robin and Treacher Collin Syndromes. Indian Pediatrics Journal. September 1992; 29: 1170-1172.
10. Marston AP,Lander TA, Tibesar RJ, Sidman JD. Airway management for intubation in newborns with Pierre Robin Sequence. Laryngoscope. 2012; 122(6); 1401-4. [Pub med]
11. Tariq Hayat Khan, Amna Ghayas , Ayesha, Samreen Khush bakht, Adeel Ahmed Khan, Naeem Khan. CASE REPORT-Successful intubation with air-Q in Pierre Robin Syndrome. Anesthesia Pain and Intensive care. 2013; 17(1):71-74.
12. Parul Mullick, Vandana Talwar, Anup Gogia. The Laryngeal mask- An aid for difficult intubation in a child with Pierre Robin Syndrome-A Case Report. Indian Journal of Anesthesia; Feb 2005; 49(1): 51-53.

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