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Case Report

Anaesthetic management in Pierre Robin syndrome with bicuspid aortic valve: A case report

Shobha V[1*], Saurabh Mittal[2], K.V. Rawat[3]

[1]Assistant Professor, [2]Junior Resident, [3]Professor, [1,2]Dept. of Anesthesiology and Critical Care, [3]Dept. of Plastic Surgery, [1-3]Himalayan Institute of Medical Sciences, Dehradun, Uttarakhand, India

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Abstract

Pierre Robin Sequence (PRS) is an uncommon complex congenital syndrome with a clinical triad of micrognathia, glossoptosis and cleft palate posing a risk of upper airway obstruction and difficulty in feeding with or without multiple secondary abnormalities. We report a case of 3.5yr male child suffering from PRS along with left renal agenesis, bilateral CTEV, bilateral inguinal hernia, pectus carinatum, bicuspid aortic valve where successful ventilation and intubation was done and taking a tongue stitch along with nebulization and prone position helped to maintain airway during recovery.

Introduction

Pierre Robin Sequence (PRS) was first documented by a French somatologist, Pierre Robin in 1923 with an incidence of 1:8500 live births.[1] It comprises of "micrognathia, glossoptosis and U or V shaped cleft palate" either found in isolation or in association with other anomalies.[2] Hypoplasia of mandible before 10th weeks of gestation prevents the palatal shelves from fusing in the midline resulting in small receding mandible and large tongue, hence causing airway obstruction.[3] Airway management is further complicated by cleft palate and limited mouth opening, resulting in difficult mask ventilation and intubation.[3,4]

Case Report

A 3.5year old baby boy weighing 10kgs was posted for cleft palate repair under general anaesthesia. He was delivered prematurely at 33 weeks of gestation weighing 2kgs but had bluish discolouration of face and upper limbs with respiratory distress and multiple congenital anomalies. Hence was admitted in NICU for 2 months. At birth, on evaluation he had short receeding mandible, large tongue, cleft palate, left renal agenesis, bilateral CTEV and inguinal hernia, pectus carinatum, echocardiography showed Patent ductus arterioses (PDA) of 2mm with left to right shunt and 26mmHg gradient, Bicuspid aortic valve. During his ICU stay he was fed with nasogastric tube and had recurrent chest infections. He was diagnosed case of Pierre Robin Syndrome along with difficulty in breathing and mild airway obstruction, breath holding spells, cyanosis which gradually subsided with increasing age. On examination, he revealed resting Spo_2 of 92% on room air without any signs of respiratory infection, pulse rate 110/mim, NIBP 92/58 mmHg, systolic murmur in aortic area on auscultation. Routine investigations were within normal limits, USG abdomen showed absent left kidney, chest X-ray showed cardiomegaly, Echocardiography revealed Bicuspid aortic valve (gradient 20/4) and absent PDA with LVEF 70%. (Fig. 1, 2).

In the operating room, difficult airway trolley was prepared. After attaching standard monitors, Inj, Glycopyrrolate 0.05 mg IV was given. Inhalational induction done with Sevoflurane, gradually increasing from 3% to 8% along with 100 % oxygen. Direct laryngoscopy with Mcintosh blade No.1 was performed after achieving adequate depth of anaesthesia. After visualising glottis, Inj. Propofol 20 mg IV was administered and intubated with uncuffed ETT No. 4. Bilateral air entry confirmed along with waveform capnography followed by administration of Inj. Atracurium 5mg and Inj. Fentanyl 20mcg IV and mechanical ventilation started. Throat packing was done. (Fig. 3)

Anaesthesia was maintained with FiO_2 50% by adding air, Sevoflaurane 1.5-2%, Inj. Atracurium 1mg. The surgery lasted for 90 mins, after which anaesthetic agent was turned off and Inj. Paracetamol100mg IV and Inj. Dexamethasone 2.5mg IV were administered. Patient was reversed with Neostigmine and glycopyrrolate after resumption of spontaneous respiration. The child was then extubated after achieving adequate tidal volume. Following which child developed upper airway obstruction and SpO_2 decreased to 60%. Immediately nasopharyngeal airway was inserted and face mask was applied with 100% O_2 along with jaw thrust. Airway was made patent by a tongue stitch and prone

*Corresponding Author: Shobha V, Assistant Professor, Dept. of Anaesthesiology, Himalayan Institute of Medical Sciences, Dehradun, Uttarakhand, India
Email: drshobha.v.v@gmail.com
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position. SpO_2 improved to 95%. Adrenaline nebulization done and he was observed in PACU postoperatively for 24 hrs. The tongue stitch was removed on next day and discharged home on 4th post operative day.

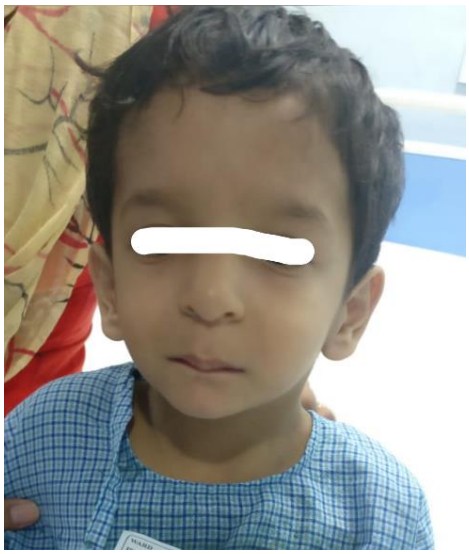

Fig. 1: Child showing micrognathia

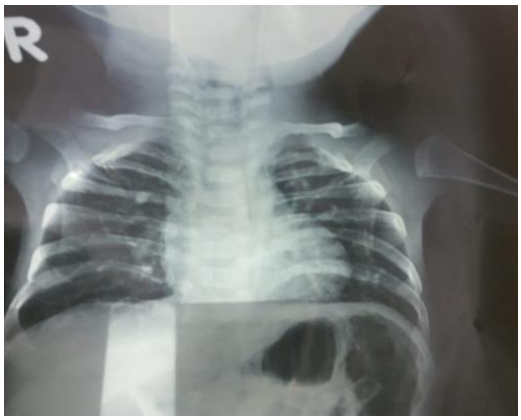

Fig. 2: Chest X-ray showing cardiomegaly and chest deformity

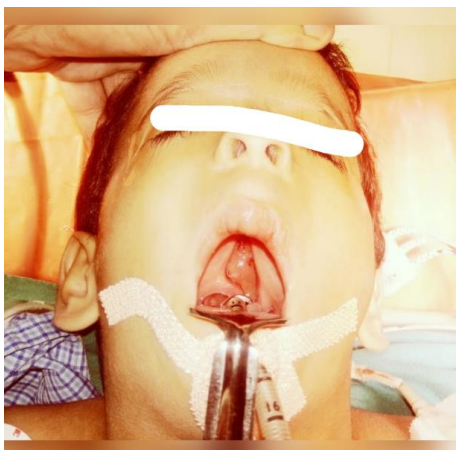

Fig. 3: Cleft palate after successful intubation of child

Discussion

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.[2] 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 cardiovasular (PDA, right heart failure, increased vagal activity), neuromuscular (central apnoea), skeletal and soft tissue abnormalities.[3] 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.[2,5] 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.[6-8] Radiological tests may be performed to evaluate bony or soft tissue abnormalities.[9] "Normal maxillo-facial angle is less than 90°, if the angle is more than 100° then visualization of glottis becomes difficult with direct laryngocopy".[10]

Premedication with glycopyrrolate or atropine can decrease airway secretions and vagal hyperactivity.[3] Surface anaesthesia of airway with 4% lignocaine nebulisation prevents holding of breath and laryngospasms during intubation.[9] 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.[2] Tariq Hayat et al. reported a 2 year old child with PRS where intubation was done using Air-Q intubating LMA.[12] 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.[13] 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.[14]

Maintainence of anaesthesia is done with sevoflurane as well as isoflurane. It may be added with ketamine, dexmedetomidine, remifentanil, which causes minimal post-operative respiratory depression.[2]

Post-operatively, airway edema leading to airway obstruction secondary to surgical manupilation 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.[2,7] In a study conducted by Tariq Hayat

et al reported post-operative respiratory obstruction, which was relieved by tongue tie at two points to the alveolar ridges.[12] Post-operative use of opioids or sedatives in these patients can precipitate grave respiratory obstruction.

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References

1. Naumann H, Pittaway A: Anesthesia for an Infant with Pierre Robin Sequence and Pyloric Stenosis; PBLDTable#25.
2. http://www2.pedsanesthesia.org/meetings/2010winter/syllabus/pdfs/pblds/Table%2, %2025.pdf
3. Cladis, Franklyn, Kumar A, Lorelie G, Todd O, Mathew F et al. Pierre Robin Sequence: A Perioperative Review. *Anesth Analgesia* 2014;119(02):400-12.
4. Bissonnette B, Bissonnette B, Luginbuehl I, Marciniak B, Bernard J. Dalens. Rapid recognition and Perioperative Implication; Pierre Robin Syndrome; McGraw- Hill Education, 2006. http://www.accessanesthesiology.com
5. Breugem C, Courtemanche D. Robin Sequence: Clearing Nosologic Confusion. *Cleft Palate Craniofac J* 2010;47:197-200.
6. Kaufman MG, Cassady CI, Hyman CH, Lee W, Watcha M, Prenatal Identification of Pierre Robin Sequence: A Review of the Literature and Look towards the Future. *Fetal Diagn Ther* 2019;39: 81-9.
7. Von Ungern-Sternberg BS, Saudan S, Petak F, Hantos Z, Habre W. Desflurane but not sevoflurane impairs airway and respiratory tissue mechanics in children with susceptible airways. Anesthesiology 2008;108:216-24.
8. Mackay DR, Controversies in the diagnosis and management of the Robin sequence. *J Craniofac Surg* 2011;22:415-20.
9. Gangopadhyay N. Pierre Robin Sequence. *Semin Plast Surg* 2012;26(2);76-80.
10. Yosha Prasad: ATOTW 250 – The Difficult Paediatric Airway; www.totw.anaesthesiologist.org, 13/02/2012: Page 1of 13.
11. D.J. Hatch: Airway management in cleft lip and cleft palate surgery; BJA. 1996; 76: 755-756
12. Shirley Dsouza. Intubation using dexmedetomedine and ketamine in a child with Pierre Robin sequence. *J Anaesthesiol Clin Phar* 2012;28(4):545-6.
13. Khan TH, Ghayas A, Ayesha, Bakht SK, Khan AA, Khan N. CASE REPORT Successful intubation with air-Q in Pierre Robin Syndrome. *Anesth Pain Intensive Care* 2013;17(1):71-4.
14. Mullick P, Talwar V, Gogia A. The Laryngeal mask- An aid for difficult intubation in a child with Pierre Robin Syndrome-A Case Report. *Indian J Anesth* 2005;49(1):51-53.
15. P Mukhopadhyay, P Mukherjee, M. Adhikari. Problems in Anesthetic Management of Pierre Robin and Treacher Collin Syndromes. *Indian Pediatr J* 1992;29:1170-2.

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