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ANAESTHETIC IMPLICATIONS IN PATIENT WITH PIERRE ROBIN SEQUENCE

Anaesthesiology

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ABSTRACT

Infants with Pierre Robin Sequence (PRS) presents a challenge to the Anaesthesiologist as infants mostly present with a hypoplastic mandible and breathing difficulty. Tongue gets displaced due to the smaller mandible. PRS is not a syndrome in itself but rather a sequence with one malformation leading to another. Exact cause is not known but hereditary cause has been postulated. We present a case of 4 month old male baby who was to be operated for bilateral ureter implantation.

KEYWORDS

INTRODUCTION:

Pierre Robin Sequence is characterised by a classical triad of micrognathia, glossoptosis and airway obstruction.[1] This heterogeneous birth defect has a prevalence of 1 per 8500 live births.[2] Ratio of male to female is 1:1. Hereditary cause is postulated and autosomal recessive inheritance is possible. The mechanical theory for PRS is most accepted which states that the initial event, mandibular hypoplasia occurs between 7th and 11th week of gestation which keeps the tongue high up in the oral cavity causing a U shaped cleft palate. Micrognathia occurs in 91.7% of the cases. The combination of micrognathia and glossoptosis can cause respiratory and feeding difficulty.[3] Other systemic aberrations may be seen. Otitis media is the most common ottic anomaly present in 80% of the patients. Nasal deformities are uncommon.[4] Laryngomalacia occurs in approximately 10-15% of patients with PRS. Speech defects are seen frequently. Cardiovascular findings may present as benign murmurs, patent ductus arteriosus, atrial septal defect, pulmonary stenosis, or pulmonary hypertension. Prevalence of cardiac defect may vary from 5-58%. musculoskeletal system are the most frequent systemic anomalies. Infants with pronounced micrognathia may present with severe respiratory distress or failure to thrive.[5,6]

Case Report:

We present a case of 4 month old baby who was to be operated for bilateral ureteric implantation. Patient was born with full term normal vaginal delivery, immediately after birth patient had severe respiratory distress and cyanosis for which patient was intubated. Patient was diagnosed with severe laryngomalacia and underwent supraglottoplasty. Patient was extubated 4 days after the surgery. Patient had history of hospitalization twice after that for the complaint of respiratory distress. Patient was then evaluated for Pierre Robin syndrome. On examination patient had micrognathia, low set ears, incomplete cleft palate, left choanal stenosis. Patient had stridor, subcostal retractions, murmur was heard on auscultation, HR-134/min. Patient had difficult IV access. Twenty-two gauge IV cannula was present on scalp. Weight of patient was 3.2 kg. On investigation complete haemogram (14gm%), coagulation profile, urine complete examination was within normal limits. Echo revealed atrial septal defect of 1.2 mm for which cardiology opinion was done and no active intervention was required from cardiology side. In USG abdomen hydronephrosis was present in bilateral kidneys and vesicoureteric junction obstruction was seen. Informed written high risk consent was taken in view of difficult airway and cardiac defect. Post-operative ventilation consent was taken. Ventilator was arranged in paediatric ICU. On the day of surgery difficult airway cart was kept ready. Routine monitors were attached (ECG, SPO2, temperature probe). IV line was secured in left side external jugular vein as mostly veins were thrombosed due to multiple hospitalisations. Injection fentanyl 1 microgram/kg, injection Thiopentone 5 mg/kg was given. Bag and mask ventilation was done with 100% oxygen and Sevoflurane. We were able to mask ventilate the patient. Check laryngoscopy was done and glottic strutures were visible. Injection Attracurium 0.5mg/kg was given. Bag and mask ventilation was done for next 3 min. Airway was secured with 3.5 mm ID endotracheal tube. Patient was maintained on 50% oxygen, 50% nitrous oxide and 1 MAC of sevoflurane was achieved. Thirty five ml of fluid (1/2 NS+10% Dextrose) was given in the first hour. Injection 1 microgram/kg fentanyl was repeated. Injection Paracetamol 24.5 gm was given. Surgical duration was 90 minutes. Incision site was infilterated with 2 ml Injection bupivacaine plain 0.25%. All the anaesthetic gases were stopped after respiratory efforts were seen. Injection Atropine 100 microgram + Injection Neostigmine 0.175mg was given for reversing muscle relaxation. Endotracheal tube was removed after oral suctioning after the patient was awake taking adequate tidal volume and respiratory rate. Patient was observed for 20 minutes in the OR. His vitals were HR-145/min, RR-40/ min, spo2 100%. Child was awake and crying. Patient was then shifted to recovery room with oxygen supplementation via nasal prongs at 4 l/ min.

DISCUSSION:

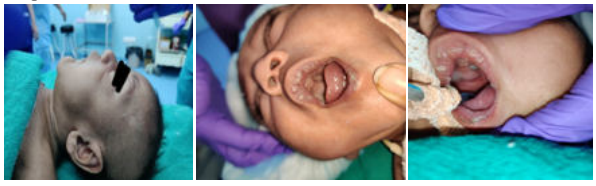
Pierre Robin Sequence is a challenging airway to the Anaesthetist. Patients with PRS may present to the Anaesthesiologist for variety of interventions for e.g, cleft palate repair, bronchoscopy etc. Hence difficult airway cart should be kept ready with LMA, Fibroscope as patient might have difficult airway and airway obstruction. Patient should be assessed meticulously during pre-anaesthetic visit. Mouth opening and palate should be assessed as cleft palate may be present. Unrecognized obstructive or central sleep apnea should be confirmed via various sleep studies. Patient's airway obstruction should be checked in all the positions to see in which position patient's airway obstruction is relieved most. Radiographic assessment should be done to rule out the cause of obstruction, for which X-ray STN or CT scan of neck can be done. MRI neck can be done to rule out any soft tissue abnormality. Maxilla-facial angle should be calculated if it is greater than 100^0 than it is a marker suggesting glottis structures will be visualised with difficulty. Echocardiography should be done to rule out any cardiac malformation. Proper systemic examination should be done as multiple malformations may be associated. We did detailed airway examination with COPUR scale (chin, opening of mouth, previous history of intubation/OSA, uvula, range of neck movement), score was 8. These patients might be difficult to ventilate hence, ability to bag and mask ventilation should be confirmed.

Intubation can be done using various techniques, without sedation, with sedation or under complete relaxation using intravenous agents and muscle relaxant. We were able mask ventilate our patient easily hence we intubated our patient under complete anaesthesia. Maintenance of anaesthesia is best done with sevoflurane or isoflurane. Major concern in the post-operative period is airway obstruction. Endo tracheal tube should be removed when the patient is awake. We can

also put the child in prone position, a nasal airway can be inserted or some surgeons may prefer to tie the tongue to the chin. Shirley DSouza et al in 2012 used Dexmedetomedine and Ketamine for intubation. They emphasized on the use of Dexmedetomedine. It is a highly selective alpha 2 agonist, taking advantage of its sedative-hypnotic and analgesic effects with minimal respiratory depression.[7] In 1986 Rasch DK et al suggested that children having 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] Similarly we also did laryngoscopy prior to induction.

CONCLUSION

Meticulous pre-operative evaluation, planned and gentle induction of anaesthesia and postoperative vigilance during postoperative period can lead to successful management of the patients with Pierre Robin Sequence.



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