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Case Report

A case report on anaesthetic challenges in Patau syndrome: Navigating craniofacial and cardiac defects

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ABSTRACT

The anaesthetic management of paediatric patients with Patau syndrome presents unique challenges, particularly when combined with craniofacial and cardiac defects and limited airway resources. This case report highlights our experience in managing a syndromic child with a difficult airway in a resource-constrained environment. The inability to secure the airway adequately can pose significant perioperative risks. In this case, we successfully utilized an oesophageal bougie as an alternative technique to secure the airway. This report emphasizes the importance of adapting to limited resources and employing innovative approaches to ensure optimal patient care in challenging situations.

1. Introduction

Patau syndrome is the third most common autosomal trisomy disorder, after Trisomy 21 and Trisomy 18, with a prevalence rate of 8 to 15 per 100,000 live births.[1] Patau syndrome phenotype is characterized by multiple craniofacial and congenital organ malformations leading to difficult ventilation and intubation.[2] Cleft palate is reported in 75% of cases and is associated with increased risk of pulmonary aspiration and infection.[2] This risk further increases with age due to development of thoracic kyphoscoliosis resulting in reduced lung volumes and ineffective cough.[3,4] As the paediatric patients have poor functional residual capacity, unsuccessful intubation can lead to hypoxemia, bradycardia and even cardiac arrest.[5] The airway devices for difficult airway in paediatric patients are often restricted due to unavailability and smaller sizes, hence a well formulated strategies are often required.[5,6]

2. Case Report

A 2 years 6 months old male child weighing 8.5 kg, a known case of Patau syndrome (Trisomy 13) was planned to undergo laparoscopic orchiopexy surgery for bilateral non-palpable undescended testis. The physical examination revealed craniofacial anomalies like short-webbed neck, micrognathia, bulbous nose, prominent occiput, sharp canines, incisors tooth decay and limb defects like swollen palms and soles with clenched fingers and toes (Figures 1 and 2). The birth history suggested preterm assisted vaginal delivery at 36 weeks of gestational age, with 1.2 kg birth weight. Although baby cried immediately after birth but, was dyspnoeic and shifted to ventilator in Neonatal Intensive Care Unit for 2 weeks due to Respiratory distress syndrome (RDS). The boy had a history of weak cry with poor rooting, suckling reflexes, feeding difficulty, regurgitation of milk through the mouth and nose till the age of 2 years, without any obvious oropharyngeal deformity. The general examination revealed delayed developmental milestones, poor nutritional status, small scrotal sac, normal intelligence quotient (IQ) for age with

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congenital acyanotic heart disease (moderate size= 8mm ostium secundum atrial septal defect with mild tricuspid regurgitation, no pulmonary arterial hypertension with left to right shunt) detected through the 2D echocardiography. All lab investigations were within normal limits.

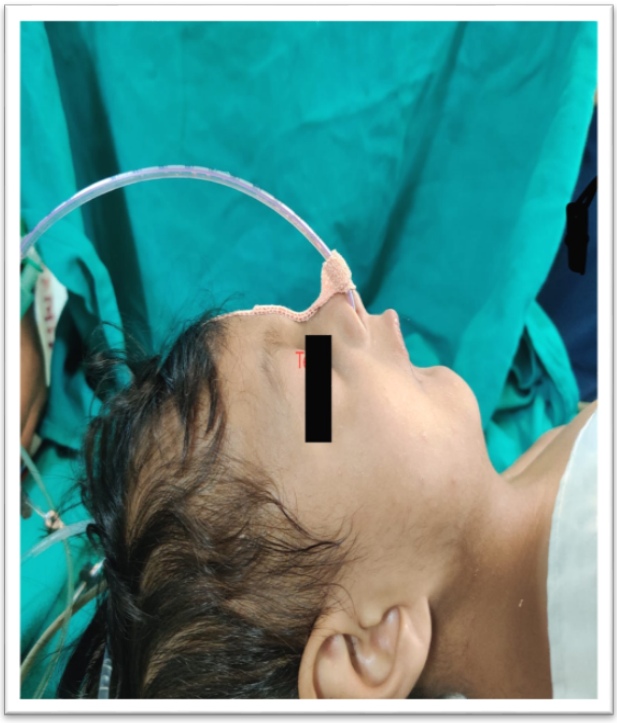

Fig. 1: Depicts micrognathia

His parents were explained about the anticipated high risk and an informed consent taken. The boy was premedicated and ASA standard monitors were attached. An otorhinolaryngologist was also informed to be prepared for an urgent tracheostomy due to the development of situation "cannot intubate, cannot ventilate". The patient was administered general anaesthesia with midazolam, fentanyl and ketamine while maintaining spontaneous ventilation with 100% oxygen. The supraglottic airway device (I-Gel) of size 1.5 was inserted without giving any muscle relaxant. The patient was ventilated manually but the bilateral equal chest rise was not achieved. Few manipulations were done but the airway was not effective, hence the device was taken as a failure and planned for endotracheal intubation immediately. The patient was ventilated with gentle bag mask ventilation in between the attempts of securing airway. During direct laryngoscopy, switch from mac-intosh blade to miller's straight blade was done to lift the epiglottis for better view. As larynx was situated very anteriorly, an attempt to intubate the child failed even after application of a paediatric bougie, burp or intubating LMA. Due to unavailability of paediatric stylet and paediatric video laryngoscope, third attempt

Fig. 2: Showing craniofacial abnormalities and clenched fingers

of intubation was done using an esophageal bougie as shown in (Figure 3) with mild Burp and a 4.0 mm ETT was rolled over the oesophageal bougie of size 8F. The patient was then shifted to controlled ventilation after administration of muscle relaxant. The perioperative fluid requirements and analgesia were maintained. At the end of the surgical procedure, the patient was reversed for the residual neuromuscular blockade & extubated.

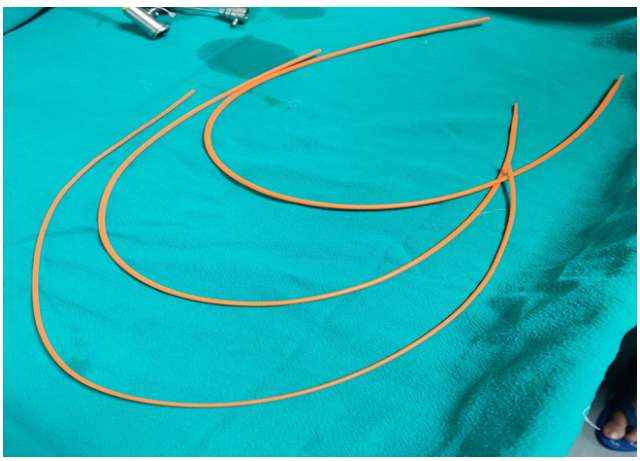

Fig. 3: Showing an oesophageal bougie

3. Discussion

Infants and children with Patau Syndrome are associated with various facial and dental abnormalities leading to difficult ventilation and intubation.[2] The anaesthetic management of such patient always demands a strategic and stepwise conventional approach in a resource constraint condition to overcome anticipated airways difficulties and prevent cardiac complications. The peri-operative goals were set in order to maintain systemic vascular resistance, stroke volume, adequate analgesia, avoidance of hypoxemia, hypercarbia, hypothermia, and prevention of increased pulmonary vascular resistance. In cardiac cases, both induction and intubation should be as smooth to avoid vasopressor responses and haemodynamic fluctuations.

Conventionally, ketamine had been considered the induction agent of choice in such patients owing to its effect of an increase in systemic vascular resistance and a decrease in the right to left intra cardiac shunt due to its sympathomimetic effects.[7] When administered in a dose of 1-2 mg/kg intravenous, it does not increase peripheral vascular resistance in children with congenital heart disease, including those with pulmonary vascular disease provided there is no hypoventilation. The anaesthesia in patients with congenital heart disease is always challenging and there are no evidence-based recommendations for its peri-operative management.[8] Intravenous or inhalation induction could also be carried out, also etomidate or moreover a combination of ketamine and etomidate would have been an alternative as an induction agent in this patient.[7] Good pre-medication was important to reduce anxiety and make parental separation easy, smooth induction as it would reduce catecholamine release and avoid hyper cyanotic spell due to the reversal of shunts. The opioids administration is usually associated with excellent haemodynamic stability and provide deeper plane of anesthesia for intubation without muscle relaxants in difficult airway both for adults and children with heart disease.[9]

The options for handling cases of anticipated difficult airway included awake intubation using fiberoptic bronchoscope, insertion of supraglottic airway device, elective invasive airway intervention and/or emergency lifesaving airway procedure. Before intubation attempt in the anticipated difficult airway, we should also determine the benefits of a non-invasive versus invasive approach to airway management especially in neonatal and paediatric patients. Whenever, the selected non-invasive approach for securing airway gets failed or is not feasible, then we should prepare for an elective or emergency invasive intervention. The noninvasive airway devices are such as rigid laryngoscopes with blades of different size and design, lighted or optical stylets, supraglottic airway devices, introducers, tube exchangers, adult & paediatric sized bougies, malleable stylets, video laryngoscopes, flexible intubation scopes and fiberoptic bronchoscope. The literature is insufficient to evaluate which devices are most effective when attempted first after failed intubation, nor is the literature sufficient to evaluate the most effective order of devices to be used for attempted intubation of an anticipated difficult airway.[10] Thus, the child was planned for placement of I-Gel or endotracheal intubation under deeper plane of anesthesia without muscle paralysis following successful mask ventilation so that the surgery could be cancel and child could be awake if failed to secure airway.

This might not be regarded as a primary approach in such cases but it's good to continue airway management post induction of general anaesthesia after calculation of the risk benefits ratio. We had also restricted the frequency of attempts at supraglottic airway placement or tracheal intubation to avoid potential trauma and airway complications like desaturation. Several case reports stated that applying burp on the larynx had led to successful intubation in a patients with difficult airway after failed direct intubation.[11] As the larynx was situated very high, the commonly used aids to assist tracheal intubation like burp and paediatric bougie failed. Therefore, we used an oesophageal bougie mimicking like an intubating bougie to roll endotracheal tube over it and facilitate tracheal intubation. An oesophageal bougie is a device made up of polyvinyl chloride (PVC) with a blunt round tip, commonly used to relieve esophageal strictures. This 8F sized device helped us to reach vocal cords due to its semi curved stature as shown in Figure 3 and secure airway successfully when conventional intubating aids failed to reach anterior larynx and advanced airway devices were unavailable. An inhalation agent with oxygen and air as a carrier gas were the preferred method for maintenance of anaesthesia since ages. N2O may also cause increases in peripheral vascular resistance in non-cyanotic children, hence it was avoided in this case. The extubation should be smooth and was attempted only when the patient's haemodynamics, core body temperature and the reflexes were within normal limits.

4. Conclusion

In conclusion, securing a difficult airway and perioperative management in a syndromic child with craniofacial and cardiac anomalies in a limited resources scenario is a challenging task to perform, but with proper planning and guarded strategies, successful tracheal intubation can be achieved without compromising oxygenation and hemodynamic stability using an oesophageal bougie.

5. Source of Funding

None.

6. Conflict of Interest

None.

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