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Apert Syndrome: Anaesthetic Challenges In Child Posted For VP Shunting.

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ABSTRACT

Apert Syndrome or "Acrocephalosyndactyly" is a rare syndrome comprising of multiple congenital anomalies, like turribrachycephaly, midface hypoplasia, syndactyly, orbital hypertelorism, small mandible etc., most of which complicate the airway management. Raised intracranial pressure and difficult venous access are other major challenges in the anesthetic management. We hereby describe our experience of anesthetic management of a 5 year old child with this syndrome posted for Ventriculoperitoneal shunting.

Keywords: Apert Syndrome, Anesthetic Challenges, Ventriculoperoitoneal Shunt.

INTRODUCTION

Apert syndrome, also referred to as acrocephalosyndactyly, occurs at frequency of 1 in 160,000 live births. The characteristic features of apert syndrome include turribrachycephaly (high steep flat forehead and occiput), midface hypoplasia, orbital hypertelorism, low set ears and small mandible. Cleft palate occurs in approximately 30 % of these patients. Congenital cardiac disease (ASD, VSD) is one of the more common visceral anomalies(10 % cases). Genitourinary anomalies like cryptorchidism, hydronephrosis also occur in 10% cases. Syndactyl of hands and feet, fusion of 2nd to 4th digit, fusion of cervical vertebrae can occur. Mental retardation is common, raised ICP (intracranial pressure) can also occur.[1] Systemic nature of this condition often poses a challenge to anesthetists during such procedures. Though difficulty related to airway management is major concern, one should be aware of other complications and difficulties caution with premedication due to raised ICP and difficulty in intravenous access due to syndactyl. We report our experience in the anesthetic management of case of Apert syndrome posted for VP (ventriculoperitoneal) shunting.

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CASE REPORT

A 5-year-old child with a history of enlarging head since birth presented to our facility. He was born by term normal vaginal delivery and no perinatal complications. He also complained of fused digits in both upper and lower limbs. He had delayed milestones. There was no history of seizures, weakness of any limbs or bowel and bladder incontinence. There was no history of cyanosis, jaundice or known cardiac disease. No history of previous surgery. There was no history of upper respiratory tract infection. Patient did give a history of snoring, but there was no history suggestive of obstructed sleep apnea. However, history of lethargy, vomiting was present.On examination, brachycephaly, depressed nasal bridge, mid face hypoplasia, hypertelorism [Figure 1], and syndactyly [Figure 2 & 3] of all the four limbs were present. Also downward gaze was seen (sun setting sign).On systemicexamination, chest was clear bilaterally and heart sounds werenormal. Airway evaluation revealed Mallampati grade II with high arched palate [Figure 4] and adequate neck movement. Teeth were malaligned. Relevant investigations including echocardiography werenormal. General anaesthesia was planned for the procedure.Difficult airway cart was kept ready. No premedication was given.In the operating roomstandard monitors were attached. Intravenous access was obtained with 22 G cannula on right lower limb after applying EMLA cream. A

shoulder roll was kept to align axis in view of large head. Modified rapid sequence intubation was done in view of risk of aspiration. The patient was induced with glycopyrrolate 0.1 mg, fentanyl 30 µg, thiopentone 75 mg and 18 mg of rocuronium was given. Cricoid pressure was applied as soon child was sedated until inflation of cuff. The patient was intubated with 4.5 sized uncuffed endotracheal tubes, which was fixed at 14 cm. The Cormack-Lehane view was Grade 1. Laryngoscopy was easy although there was problem with intubation due to angulation of glottis. The anesthesia was maintained with oxygen, nitrous oxide, isoflurane, fentanyl. A total volume of 75 ml of 20% mannitol was also administered. The patient maintained an intraoperative temperature of 36-36.2°C. The patient suffered a blood loss of hardly 100 ml which was replaced with crystalloids. The patient was hemodynamically stable maintaining a systolic blood pressure of 90-110 mmHg. After the completion of surgery, the patient was assessed and on ensuring that the patient was fully awake trachea was extubated.

Figure 1: Showing hypertelorism, midface hypoplasia, flattened nasal bridge

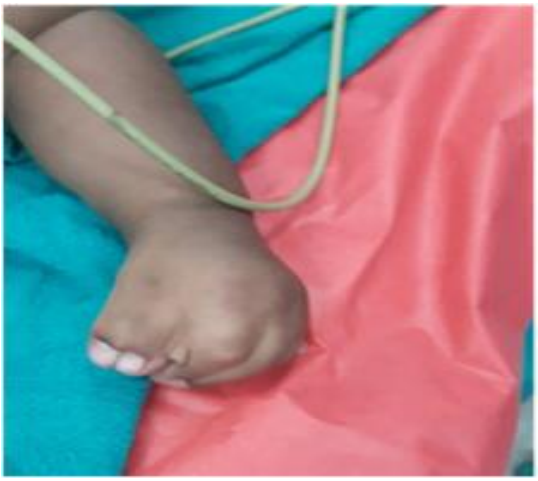

Figure 2: Syndactyl of hands

Figure 3: Syndactyl of feet

Figure 4: High arched palate

DISCUSSION & CONCLUSION

Apert syndrome is an autosomal dominant disorder caused due to the mutation of fibroblast growth factor recptor-2 (FGFR-2) on chromosome 10q.The majority of cases are sporadic, resulting from new mutations with a paternal age effect.[2] Unique fibroblast growth factor receptor 2 mutations lead to to an increase in the number of precursor cells that enter osteogenic pathway.[3] Ultimately, this leads to increased subperiosteal bone matrix formation and premature calvaria ossification during fetal development . Once a suture becomes fused , growth perpendicular to that suture becomes restricted and the fused bones act as a single bony structure. Compensatory growth occurs at the remaining open sutures to allow brain growth. However, complex, multiple sutural synostosis frequently extends to premature fusion of the sutures at the base of the skull causing brachycephaly, midfacial hypoplasia, shallow orbits , hypertelorism, foreshortened nasal

dorsum, maxillary hypoplasia and mandibular prognathism as seen in our case.[4]

As there are chances of difficult intubation, the following classical features should be evaluated preoperatively:

- High Mallampati score
- Reduced thyromental distance
- Reduced sternomental distance
- Restricted atlanto-occipital joint movement
- Restricted temporo-mandibular joint function
- Limited mouth opening

Intubation aids, such as Combitube, laryngeal masks and emergency tracheotomy sets must be available.[5] The major airway abnormality is midfacialhypoplasia which make them more prone for obstructive sleep apnoea and difficult mask ventilation. Use of oral airway, supraglottic airway devices, judicious use of narcotics and sedatives is advisable to avoid airway obstruction during induction and emergence of anaesthesia. Different sizes and types of masks should be available. Due to the increased incidence of supraglottic obstruction, a nasopharyngeal airway may be useful in the immediate postoperative period. If the child is CPAP/BiPAP dependent, then the child must be sent to an appropriate ward setting to accommodate this.Cervical spine anomalies occur in 71% of these patients and mainly consist of complex fusion of the spine at C5-C6 level.[6] This however, is of little consequence in intubation of these patients as positioning for intubation involves the movement at upper cervical spine levels.

Cartilaginous abnormality of trachea, tracheal stenosis, choanal atresia and inability to clear airway secretions leads to significant rise in airwaymorbidity in these patients.[7] These children tend to haveprofuse airway secretions making them more prone for bronchospasm. In an emergency situation it may require reintubation.[8]

These patients are vulnerable to develop hypothermia as significant heat loss occurs through the exposed head and blood loss. Moreover, the patients with Apert syndrome are known to have excessive sweating.

Another challenge is a difficult intravenous access as these children come frequently for various procedure. Syndactyly also limit cannulation and pulse oximetry monitoring. For short procedure intravenous access can be avoided while in emergency situation intraosseous or intramuscular route can be used. Surgeries involving extensive blood loss may require central venous catheter insertion.[9] Abnormal skull development may lead to shallow orbits causing protrusion of the eyes and inability of the lids to close, known as exorbitism. Due to inadequate lid closure eyes are particularly susceptible to damage and hence eyes should be lubricated, taped and padded. Consideration should be given to avoid pressure point.[10]

Tosun and Sener's study showed that Apert syndrome was in parallel with G6PD deficiency. G6PD deficiency is an enzymatic hereditary disorder leading to hemolytic anemia as a result of red blood cell destruction. The main problem in G6PD deficiency is that hemolysis can be precipitated by a number of factors, such as oxidant drugs, eating fava beans, or intercurrent infection. Drugs that may induce hemolysis include sulphonamides, chloramphenicol, aspirin, acetaminophen, penicillin, and streptomycin. Therefore, one must avoid drugs that may potentially induce hemolysis as result of G6PD deficiency. Analgesia should be maintained with i.v opioids in titrated dosing and therefore avoiding acetaminophen and NSAIDS.[11]

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