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Original Research Paper

Anesthesiology

ANAESTHETIC CONCERNS AND CHALLENGES IN A CASE OF APERTS SYNDROME AND ITS PERIOPERATIVE MANAGEMENT

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ABSTRACT

Apert syndrome is a congenital autosomal dominant disease characterized by craniosynostosis, brachycephaly, midface hypoplasia , hypertelorism , choanal stenosis , multidigit hand and foot syndactyly.

Anaesthetic implications include difficult airway, hyperactive airway disorder, difficult intravenous access, temperature dysregulation, repeat surgical procedures.

Thorough pre - operative assessment of airway should be performed. Peri -operative management should include difficult airway cart preparation, temperature monitoring and watch for post-operative signs for airway obstruction.

Regional anaesthesia may be difficult in these patients due to anatomic variations and therefore ultrasound guidance will be helpful.

We report a case of 3-year-old girl of Aperts syndrome who presented to our institute for syndactyly release

KEYWORDS :

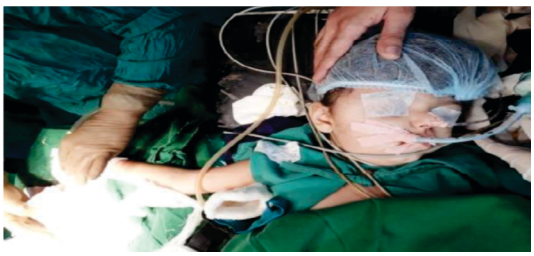
INTRODUCTION:

Aperts syndrome is a congenital autosomal dominant disease characterized by various facial abnormalities like brachycephaly, midface hypoplasia, hypertelorism other congenital abnormalities like hydrocephalus, cardiac and genitourinary defects , gastrointestinal defects. It may present a number of challenges to anesthesiologists like difficult airway,difficult intravenous access, temperature dysregul ation. Here we report a case of 3 year old girl with aperts syndrome who presented to our institue for syndactyly release.

CASE REPORT:

- A 3 year old female child of 15kgs with k/c/o Aperts syndrome was presented to our instituite. She was born by term normal vaginal delivery with birth weight of 2.5kg with no perinatal complications . Immunised and h/o develomental delay present. No history of any seizures,cardiac disease, jaundice or cyanosis. Child was operated for cleft palate at the age of 1 ½ years. Patient had recurrent episodes of pneumonia which was treated by I.V antibiotics.
- On examination she was found to have brachycephaly ,with flattened occiput with midface hypoplasia , proptosis, down-slanting of palpebral fissures , hypertelorism, and depressed nasal bridge. Dental findings followed crowding , impaction , missing teeth with adequate mouth opening, large tongue, high arched palate and with Mallampati classification 2 . Child was found to have profuse secretions and sweating.
- On systemic examination harsh breath sounds were noted for which nebulisation was advised one day prior to surgery.
- Her blood investigations were within normal limits with hb of 10.4g/dl. Her 2D echo showed LVEF 60% and structurally normal heart. Was accepted under ASA 2.
- Child was kept nil by mouth 4 hours before surgery. All routine monitors like ECG, NIBP, SPo2 and temperature were attached and baseline parameters were noted. 24 G IV cannula was taken on right foot (difficult cannulation).

- Child was premedicated with Inj.glycopyrrolate 0.06mg , midazolam 0.75mg , inj fentanyl with 40 mcg . Adequate bag mask ventilation was confirmed with airway. Difficult airway cart was prepared with LMA and fiberoptic .
- Child was intubated with 4.0 uncuffed ET tube at the second attempt and was maintained with sevoflurane.
- Proper eye care was taken by applying ointment and padding. Surgery lasted for 2 hours and was uneventful. USG guided left axillary block was given for post operative analgesia with 4ml of 0.25% bupivacaine . Child was reversed with neostigmine and glycopyrrolate .Spontaneous breaths taken. Child maintained saturation with 6ltrs of O2 via Hudson mask. Patient was shifted to PICU for observation.



DISCUSSION:

- Genetically the defect is in chromosome 10 affecting FGF receptor 2 gene. Airway dysmorphism carries a risk of difficult mask ventilation and intubation due to midface hypoplasia. About 71% have complex fusion of C5-C6spine. Cartilageneous abnormality of trachea, tracheal stenosis, choanal atresia may rise to significant airway morbidity. Therefore difficult airway cart should always be prepared before the induction.
- Use of regional anaesthesia could itself be difficult due to anatomic variations in shoulder joint and related structures. Profuse secretions make them prone for bronchospam which needs post op monitoring.
- Another challenge includes difficult intravenous access as these children come more frequently for various procedures. For short procedures introsseous or intramuscular route can be used.
- Due to profuse sweating, they are more prone for

hypothermia,hence all precautions to maintain normothermia and temperature monitoring is essential. As proptosis and inadeqaute eye closure could result in eye injury, adequate lubrication and eye padding should be done.

CONCLUSION:

Dealing with a case of Aperts syndrome is very challenging for anaesthetist. So, thorough preoperative assessment, a proper anesthesia plan as well as a back up for difficult airway management are very important in these patients.

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