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ANESTHETIC MANAGEMENT OF A RARE CASE OF APERT SYNDROME WITH CLEFT PALATE FOR LONG BONE HUMERUS FRACTURE.

Anaesthesiology

Dr. Siddhant R Umbarkar	3rd Year Junior Resident, Department of Anesthesiology, DVVPF'S Medical College and Hospital, Ahilyanagar.
Dr. Nikhil Karkhanis	Associate Professor, Department of Anesthesiology, DVVPF'S Medical College and Hospital, Ahilyanagar.
Dr. Khushboo Damani	Professor, Department of Anesthesiology, DVVPF'S Medical College and Hospital, Ahilyanagar.
Dr. Shashank Mohole	Associate Professor, Department of Anesthesiology, DVVPF'S Medical College and Hospital, Ahilyanagar.
Dr. Lohit Sureshchandra Vaishnao *	3rd Year Junior Resident, Department of Anesthesiology, DVVPF'S Medical College and Hospital, Ahilyanagar. *Corresponding Author

ABSTRACT

Introduction: Apert syndrome is a rare disorder which generally occurs sporadically, although an autosomal dominant inheritance has been described[1]. Its features have clinical relevance to anaesthetists as some degree of upper airway obstruction due to reduced nasopharyngeal volume and choanal patency is certain along with lower airway compromise. **Case Summary & Management:** A 7 year old female weighing 25 kg with left proximal humerus one third shaft fracture posted for internal fixation with TENS nailing. Patient is a known case of APERT Syndrome

- Syndactyly (Connected Fingers)
- Blue Sclera, Exophthalmos
- Broad forehead, Cleft palate
- Abnormal facies with delayed developmental milestones
- Dextrocardia
- Irregular, crowded and buck teeth with MPC IV with American Society of Anesthesiology grade IV.

This paediatric patient with difficult airway was managed with general anesthesia with ETT no. 5.5 and supraclavicular block using Injection Bupivacaine 0.25% (p) 20 ml given in supraclavicular region. **Conclusion:** In Children with APERT syndrome anesthetists face multiple problems while dealing with them, a risk of difficult mask ventilation and intubation due to mid face hypoplasia and craniofacial anomalies. The proper planning and difficult intubation trolly should be kept ready while dealing with a patient with APERT Syndrome.

KEYWORDS

INTRODUCTION

Apert syndrome is a rare disorder which generally occurs sporadically, although an autosomal dominant inheritance has been described[1].

These features have clinical relevance to anaesthetists as some degree of upper airway obstruction due to reduced nasopharyngeal volume and choanal patency is certain. Obstructive sleep apnoea is present in almost 50%[2] and cor pulmonale may result[3]. There have also been descriptions of complete or partial cartilage sleeve abnormalities of the trachea, which has been postulated to cause lower airway compromise[4].

Children with this syndrome require general anaesthetics for a number of different operations and procedures. It has been suggested that they have a much higher incidence of peri-operative respiratory complications, particularly bronchospasm, than other children[5].

Case Summary

A 7 year old female weighing 25 kg with left proximal humerus one third shaft fracture posted for closed reduction and internal fixation with TENS nailing.

Patient had history of fall from from bed.

Patient is a known case of APERT Syndrome

- Syndactyly (Connected fingers)
- Blue Sclera, Exophthalmos
- Broad forehead, Cleft palate
- Abnormal facies with delayed developmental milestones
- Dextrocardia
- Irregular, crowded and buck teeth with MPC IV with American Society of Anesthesiology grade IV.

Patient had no known drug allergies. Her surgical history was insignificant. Preoperative laboratory studies were within normal limits. Premedication consisted of midazolam, 10 mg intranasally.

The patient was brought to the operating room and monitored with standard electrocardiograph, blood pressure, pulse oximetry, and temperature equipment. An uneventful insertion of a 24-gauge peripheral intravenous catheter. Injection Ondensetron 2 mg IV given.

Preoxygenation done with Mask no 2 with 10 litres of O2 for 3 min. Primarily general anesthesia with supraclavicular block was given. For premedication Inj Midazolam 0.25 mg, Injection Nalbuphin 3 mg, Injection Glycopyrollate 0.08 mg IV given.

For induction Injection Propofol 60 mg IV, Inj Fentanyl 60 mg IV and Inj Atracurium 15 mg IV given. Abolition of Eyelash reflex and jaw relaxation was confirmed. Intubation with ETT No. 5.5 oral enforced endotracheal tube was atraumatic on first attempt with triples maneuver. After Bilateral breath sounds were auscultated and end tidal CO2 was noted on the capnography tube was taped and secured. Anaesthesia was maintained with O2, Air and Sevoflurane.

Supraclavicular block using Injection Bupivacaine 0.25% (p) 20 ml given in supraclavicular region.

The patient was maintaining Saturation of 100% on Jackson Rees circuit with assisted Mode of ventilation VT-240-300 Freq-12.

The patient was vitally stable throughout the procedure.

After procedure vitals of the patient were P-124 bpm, BP-100/64mmHg and RR-20/min.

Inj Neostigmine 1.25mg + Inj Glycopyrollate 0.1 mg given IV as a reversal.

The patient was shifted to SICU on O2 support.

DISCUSSION

Apert Syndrome was first described by the French pediatrician Eugene Apert in 1906.[6] It is a rare autosomal dominant disease with an incidence of around 1 per 160 000 live births.[7] Genetically, the defect is in chromosome 10 affecting fibroblast growth factor receptor 2 gene. This mainly affects digits and cranium causing fusion of multiple digits of both upper and lower limbs and premature fusion of cranial sutures.[8] These children also have other associated anomalies like cardiac defects, polycystic kidneys, and pyloric stenosis.

These children require a number of different operative procedures and Anesthetists face multiple problems while dealing with them, first of it being the airway management. Airway dysmorphism carries a risk of difficult mask ventilation and intubation due to mid face hypoplasia.[9,10] Children with Apert syndrome have profuse secretions with inability to clear these that may cause increase in airway irritability, higher incidence of bronchospasm, and repeated respiratory tract infection.[11] Craniofacial anomalies are often associated with airway obstruction, especially during sleep, and can cause obstructive sleep apnea,[12,13] and therefore are prone to becoming obstructed on induction and emergence. Preoperative evaluation and optimization is important in these patients. Airway adjutants like appropriate size airway, LMA, endotracheal tubes, and emergency tracheostomy kit has to be kept ready.[14] Use of regional anesthesia techniques as an adjuvant to general anesthesia help to decrease the requirement of opioids during intra- and post-operative period. This reduces the risk of airway obstruction. Regional anesthesia itself could be difficult in these patients due to anatomical variations in shoulder joint and related structures. Ultrasonography (USG)-guided regional anesthesia techniques may help us to overcome this problem.[15]

Through screening for associated anomalies like cardiac defects, polycystic kidneys and pyloric stenosis should be done preoperatively.[16]

Another challenge for the anesthetist is intravenous access. Limb deformity and multiple operative procedures make intravenous access more difficult in these patients. As a result, some anesthetists feel that for short procedures, e.g., change of dressings or computed tomography (CT) scans, intravenous access is not mandatory. In an emergency, an intraosseous or intramuscular route can be used.[10]

CONCLUSION

In Children with APERT syndrome anesthetists face multiple problems while dealing with them, a risk of difficult mask ventilation and intubation due to mid face hypoplasia and craniofacial anomalies.

The proper planning and difficult intubation trolly should be kept ready while dealing with a patient with APERT Syndrome.

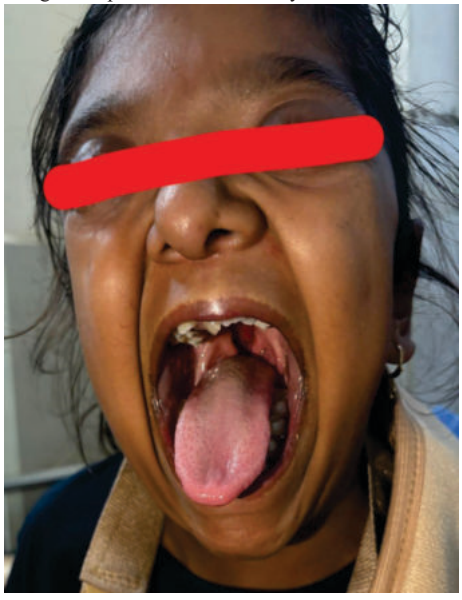

Figure no. 1: Airway of Patient

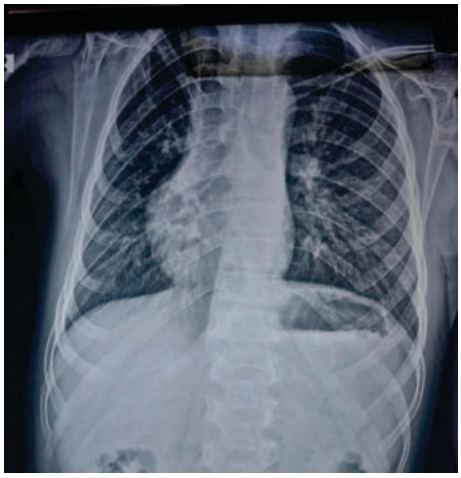

Figure no. 2: Chest Xray Showing Dextrocardia.

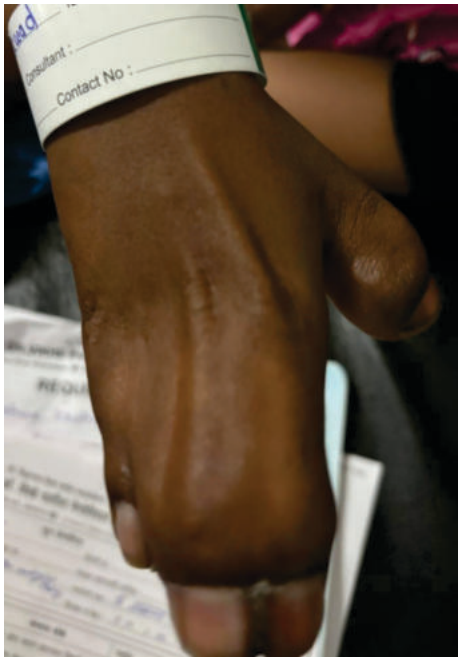

Figure no. 3: Picture of Hand Showing Syndactyly.

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