Journal of Institute of Medicine Nepal
Institute of Medicine, Kathmandu, Nepal

OPEN ACCESS

Case Report

JIOM Nepal. 2021 Apr;43(1):50-53.

Anesthetic Management of Apert Syndrome: A Case Report

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Submitted
Jan 10, 2021

Accepted
Mar 16, 2021

ABSTRACT

Apert syndrome is a rare autosomal dominant disease associated with abnormalities of skull, face and limbs. These patients present for different types of surgeries. To the anesthesiologists,they pose special challenge during airway management, manifesting as difficulty in bag-mask ventilation and increased incidence of airway obstruction. Here, we report a case of a child with Apert syndrome undergoing syndactyly release under general anesthesia, in whom severe bronchospasm and a failed bag-mask ventilation occurred during the emergence of anesthesia requiring re-insertion of laryngeal mask airway.

Keywords
Anesthetic management, Apert syndrome

INTRODUCTION

Apert ayndrome is a rare congenital autosomal dominant disease occurring due to fibroblast growth factor receptor 2 genetic defect in chromosome 10 resulting in craniofacial and limbs malformation.[1] It was first described by a French pediatrician, Eugene Apert and occurs in 1:160000 live births.[2] Due to prevention of apoptosis of cells, these malformations present as a premature fusion of cranial sutures resulting in craniosynostosis and fusion (cutaneous and/or bony) of both hands and feet. The characteristic features include brachycephaly, mid face hypoplasia, hypertelorism, exorbitism and syndactyly of hands and feet. These children mainly present to the operation theatre for the release of syndactyly, craniosynostosis correction, dental and orthopedic surgeries.The major challenge for the anesthesiologist is during airway management.

CASE PRESENTATION

A 9-year-old female child weighing 20 kg,with the characteristic features of Apert syndrome (Figure 1-4) presented for the release of syndactyly of left hand. She had brachycephaly, mid face hypoplasia, depressed nasal bridge, slightly hypoplastic chin, hypertelorism and exorbitism.

Fig 1. Picture showing brachycephaly, mid face hypoplasia, hypertelorism and depressed nasal bridge

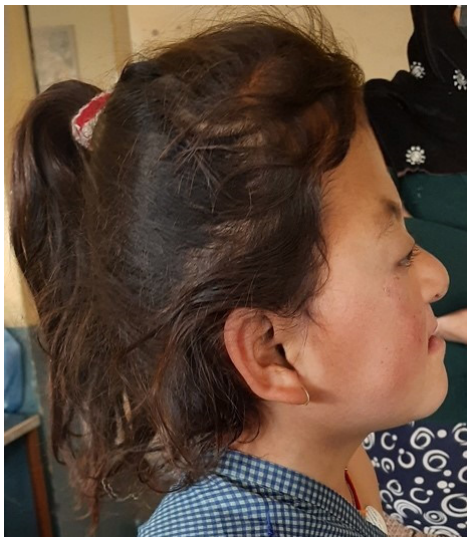

Fig 2. Picture showing hypoplastic chin

She had a normal birth history and had normal developmental milestones. She had no features of raised intracranial pressure. She had the history of prior two surgeries for the syndactyly of right hand and the documents related to anesthetic management were not available. There was no history of snoring or any features suggestive of obstructive sleep apnea. Her airway examination revealed a high arched palate, slightly hypoplastic chin and misalignment of multiple teeth. Her all laboratory investigation reports were within normal limits.

An intravenous access was secured in right hand and patient was premedicated with glycopyrrolate 0.1mg, 30 mins before the surgery. Patient was preoxygenated, followed by administration ofinj Fentanyl 30 mcg. Patient was induced with propofol 60 mg followed by an insertion of size 2.5 laryngeal

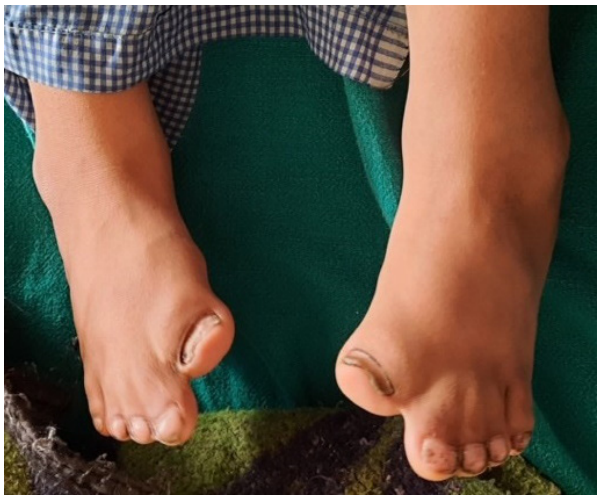

Fig 3. Picture showing syndactyly of feet

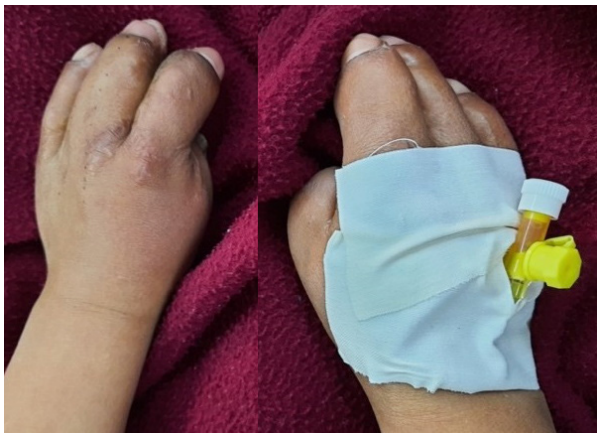

Fig 4. Picture showing syndactyly of hands (intravenous cannula in situ in the right hand)

mask airway (LMA). Anesthesia was maintained with Isoflurane, and with spontaneous breathing. Intraoperative temperature monitoring was done using nasopharyngeal temperature probe. Patient received intravenous paracetamol 300 mg, and a wrist block with 0.25% bupivacaine (6 ml) at the end of surgery. After completion of surgery, LMA was removed with patient in deeper plane of anesthesia. Spontaneous ventilation was inadequate, so bag-mask ventilation with Guedel's airway was attempted in conjunction with triple airway maneuver. Airway obstruction was not relieved even with continuous airway pressure, leading to drop in oxygen saturation. Patient was re-induced with propofol 20 mg and LMA was re-inserted. Upon auscultation there was severe bronchospasm for which we administered salbutamol puffs via LMA and injectable hydrocortisone 50 mg. The bronchospasm relieved slowly, followed by normalization of oxygen saturation. After assuring adequate ventilation and with patient fully conscious, LMA was removed. The subsequent emergence from anesthesia was uneventful.

DISCUSSION

Though a rare entity, patients with Apert syndrome may be encountered multiple times in the operation theatre, being posted for corrective surgeries. The venous access may be difficult due to the operating field being the limb itself, or due to multiple previous surgeries or deformed limb. In our case the syndactyly release was planned in left hand so we had an access in right hand.

These patients may have associated cardiac anomalies, so if a history is suggestive, echocardiography should be done preoperatively. Since our patient had a good functional status, we assumed her to have normal cardiac function.

The most commonly encountered complications is related to airway during induction and emergence of anesthesia.[3-4] History of obstructive sleep apnoea (OSA) is suggestive of difficult airway. Due to mid facial hypoplasia, airway obstruction occurs frequently. In our case after removal of LMA, there was an airway obstruction which could not be managed with airway maneuvers or oropharyngeal airways. Ventilation could only be possible after the insertion of a supraglottic airway device. Mid facial hypoplasia and the associated anatomical alternations might have caused failed ventilation during the deep plane of anesthesia, compounded by associated bronchospasm. Bronchospasm has been reported to occur in such patients due to excessive secretion and inability to clear the secretions. In our patient, we had premedicated with antisialogogue. Excessive secretion leading to blockage of tube have been reported previously.[5] In our experience, when oropharyngeal airways and airway maneuver techniques fail to maintaina patent airway, a supraglottic airway device can be helpful. Always keeping an LMA of appropriate size standby, would be prudent. Choanal stenosis may be present in such patients precluding insertion

Table 1. Anesthetic concerns in patients with Apert Syndrome.

Anesthetic concern	Cause	Implications
Difficult bag mask ventilation	Mid face hypoplasia	Use of airway adjuncts, LMA
Airway obstruction	Mid face hypoplasia, high incidence of OSA	Use of airway adjuncts, multimodal analgesia, lesser dose of opioid, use of regional anesthesia whenever possible
Difficulty in inserting nasopharyngeal airway	Choanal atresia, reduced nasopharyngeal volume.	Avoidance or use of a smaller airway
Difficult intubation	Fusion of C5-C6 spine	Difficult intubation cart
	Bamboo trachea	Smaller size ETT
Bronchospasm	Increased secretion, increased airway susceptibility	Premedication with antisialogogues, reduced airway instrumentation
Difficult venous access	Surgery in the limbs, multiple surgeries	Staged operation, intraosseous needle
		Dressing without intravenous access
Difficult regional anesthesia	Abnormal anatomy of shoulder, limited abduction of upper arm	Use of ultrasound guided block
Increased intracranial pressure	Ongoing brain growth despite fusion of suture	Proper history and examination, avoidance of drugs maneuvers that increase ICP
Eye injury	Exorbitism (inability of total eyelid closure)	Ensure well lubrication, taping and padding of eye
Cardiac risk	Cardiac anomalies	Proper history, echocardiography if required
Temperature regulation	Excessive sweating increased chance of hyperpyrexia	Temperature monitoring

of nasopharyngeal airway. Forceful insertion may traumatize and should be avoided. The cervical vertebre may be fused at varying levels leading to limited neck mobility and cause difficulty in intubation.[6] Tracheal anomalies have also been reported.[7] Trachea is defined to be like a bamboo requiring a smaller than anticipated ETT during intubation.

Opioids may worsen airway obstruction, thus multimodal analgesic technique should be used to decrease the perioperative opioid requirement. We used paracetamol and wrist block for the purpose of decreasing the dose of fentanyl. Regional anesthesia may be challenging because of abnormal anatomy of shoulder joint and ultrasound may be a useful aid.[3] These patients may have protruded eyes with incomplete lid closure. In order to prevent eye injury, proper eye taping and use of eye lubricant is recommended. Also, these patients tend develop intraoperative hyperthermia if active warming is done. So, temperature monitoring should always be done.

Features of raised intracranial pressure should always be looked for as they have premature suture closure.[8] Sometimes it is difficult to insert a pulse oximeter due to total fusion of all the fingers.

The anesthetic concerns and its implications are tabulated in Table 1.

CONCLUSION

In conclusion, a proper anesthetic plan to deal with airway complications is a must whenever we are anesthetizing a case of Apert syndrome for whatsoever surgery.

CONSENT

Informed consent was was obtained from the mother of the patient for publication of this case report and accompanying images.

CONFLICT OF INTEREST

None declared.

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