Anaesthesia Concerns in Apert Syndrome - A Case Report

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Received Date: 07-16-2019; Accepted Date: 07-24-2019; Published Date: 07-28-2019

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Abstract

Apert syndrome (AS) a type of acrocephalosyndactyly is a rare congenital disorder with autosomal dominant mode of transmission that consists of craniofacial synostosis, midfacial hypoplasia and bilateral limb syndactyly. Patients present in early childhood for multiple surgeries which make it imperative to know about various anaesthetic implications like difficult airway ventilation, airway hyper reactivity, associated congenital anomalies, increased airway secretions and deranged temperature thermoregulation associated with this syndrome. The patient should be thoroughly evaluated preoperatively and managed accordingly. We discuss the successful management of a three and a half years old male child with Apert syndrome.

Keywords

Apert syndrome; Acrocephalosyndactyly; Difficult airway

Introduction

Apert syndrome is a rare autosomal dominant disorder characterised by craniosynostosis, craniofacial anomalies & severe symmetrical syndactyly (cutaneous & bony fusion) of the hands & feet. Apert syndrome is named after the French Physician Eugene Apert who described the syndrome acrocephalosyndactyly in 1906 [1]. It poses an anaesthetic challenge because of the difficulties related to airway management, intravenous access, associated systemic anomalies (cardiac defects, polycystic kidneys & pyloric stenosis) & increased incidence of bronchospasm & wheezing. We report our experience in the anaesthetic management of a case of Apert syndrome, referred to us for syndactyly release.
Case Report

A three and a half years old male child, already diagnosed as a case of Apert syndrome, was admitted to our hospital for scheduled bilateral hand syndactyly release. The child was already operated for bicornoral synostosis at the age of nine months & for syndactyly release at the age of 1 year. This child had midfacial hypoplasia involving maxillary & zygomatic bones with orbital proptosis, frontal bossing & fusion of multiple digits in all the limbs. Any coexisting malformations of the heart, digestive system or urogenital systems were excluded by medical history & previous medical records. All blood investigations showed normal results. Preoperative evaluation of the child revealed harsh vesicular breath sounds for which preoperative nebulisation was done.

After an informed parental consent, the child was scheduled for an elective surgery under general anaesthesia. The patient was taken to the operation theatre. The first problem which was encountered in this patient in operation theatre was difficult peripheral venous access. An intravenous 22G cannula was secured with difficulty in left lower limb in operation theatre. Routine monitoring (HR, RR, SpO2 with peripheral plethysmograph) done besides temperature monitoring. Another difficulty encountered was to apply the routine saturation probe because of all the 4 limbs syndactyly & therefore an ear lobe probe was used in this patient. Premedication was given with 10µgm/kg atropine as the patient had increased airway secretions. General anaesthesia was induced with 5mg/kg Thiopentone & 2mg/kg Succinylcholine. There was some difficulty in achieving a close fit of the face mask (because of abnormal facial contour) & a slight airway obstruction during mask ventilation which was relieved by using oral airway. Tracheal intubation was done with uncuffed endotracheal tube size 5, under direct laryngoscopy & confirmed by auscultation & ETCO2 (capnography). Anaesthesia was maintained with Sevoflurane (1MAC) with N2O & oxygen & intermittent muscle relaxation with atracurium. During anaesthesia secretions were frequently aspirated through the ETT. Eyes were properly lubricated with eye ointment, taped & padded to avoid injury to eyes which could be aggravated due to presence of proptosis. No active warming was used as the patient’s body temperature increased intraoperatively from 36.5°C to 37.8°C & patient was sweating profusely. Paracetamol suppository 20mg/kg & intravenous fentanyl 2µgm/kg were given for intraoperative analgesia. The surgery lasted for 4 hours. Reversal was uneventful without any signs of respiratory depression.

Discussion

Apert syndrome is a rare genetic disorder characterized by acrocephalosyndactyly. It is inherited as autosomal dominant trait or is sporadic with mutation in fibroblast growth factor receptor 2 gene on chromosome 10q26 [2]. The retarded skull growth affects the brain development, which may result in mental retardation but may have normal intelligence also as was in our case. These children require a number of different operative procedures & anaesthetists face multiple problems while dealing with these patients. Thorough screening
for associated anomalies like cardiac defects, polycystic kidneys & pyloric stenosis should be done preoperatively [3].

As these patients have characteristic proptosis, special attention must be paid to the eyes to avoid corneal or compression ophthalmic injury. Intravenous access may be extremely difficult in these patients depending on severity of syndactyly. Varied types of pulse oximeter probes (finger, earlobes) preferably with adhesive sensors must be readily available as normal pulse oximeter finger probes will be difficult to apply because of syndactyly & deformed fingers & toes as was in our case. These patients tend to sweat a lot and unlike other patients undergoing syndactyly surgery do not appear to need warming when undergoing peripheral surgery to their limbs. If they are actively warmed, there is a risk of pyrexia, so temperature should always be monitored [4]. Most patients with Apert syndrome experience some degree of airway obstruction as a result of small nasopharyngeal & oropharyngeal dimensions, particularly if choanal atresia & or tracheal stenosis are present. Also cartilagenous abnormalities of the trachea, fusion of the cervical vertebra, angular deviation of the trachea & midfacial hypoplasia may contribute to difficult intubation in these patients [5]. Various airway adjuncts & difficult airway cart must be readily available. Obstructive sleep apnea is common & must be addressed early to avoid development of corpulmonale [6]. Use of regional anaesthesia techniques as an adjuvant to general anaesthesia or as a sole technique helps to avoid the complications of general anaesthesia but regional anaesthesia itself can be difficult in these patients due to various anatomical variations. Ultrasonography guided regional anaesthesia techniques may help to overcome the problem [7].

**Conclusion**

Anaesthetic management of a case of Apert syndrome is a challenging task. Prior knowledge about the difficulties in management of these patients will help in the proper planning in perioperative care & uneventful recovery of these patients.

**References**