DOI: 10.7860/JCDR/2022/50287.16236

Case Series

Anaesthesia Section

Anaesthetic Management and Literature Review of Syndromic Craniosynostosis in Infants-A Case Series

SMRITI SINHA[1], REENA R KADNI[2], JOEL CHAKRAVARTHY[3], VARGHESE K ZACHARIAH[4]

ABSTRACT

Craniosynostosis is a rare congenital defect. This group of patients is heterogeneous, presenting as a single suture defect in otherwise well child to Syndromic Craniosynostosis (SC) affecting multiple sutures as cranial defect associated with extracranial defects. Syndromic Craniosynostosis is found to be associated with Apert, Crouzon, Pfeiffer, Muenke and Saethre-Chotzen syndrome. These cases are generally associated with raised Intracranial Pressure (ICP), difficult airway and congenital multiple organ involvement. The case series describes the successful anaesthetic management of three and half-month-old male infant with Crouzon syndrome presented with raised ICP and difficult airway, planned for third ventriculostomy and seven-month-old male infant with Apert syndrome with syndactyly for correction surgery. A literature review about anaesthetic concerns related to SC is also described. The literature search was performed in the PubMed database with terms SC and anaesthetic management.

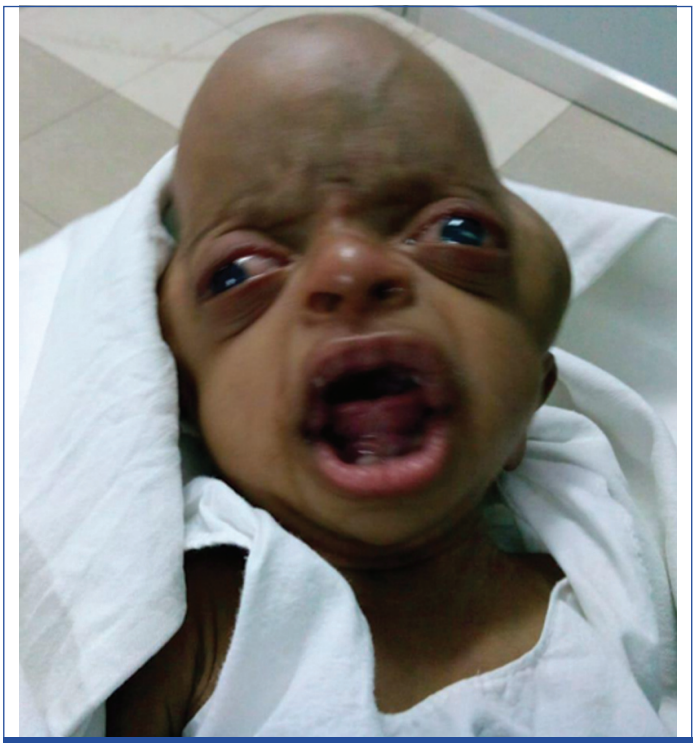
Keywords: Apert syndrome, Crouzon syndrome, Hydrocephalus, Raised intracranial pressure

INTRODUCTION

Craniosynostosis occurs in one in 2500 live births. The syndromic association occurs in 20% of craniosynostosis patients [1]. The SC occurs mostly due to mutation in genes coding for Fibroblast Growth Factor Receptors (FGFR); which is autosomal dominant in inheritance. Out of all SC, Muenke syndrome is the most common syndrome, followed by Crouzon and Pfeiffers syndrome. Apert syndrome has the lowest prevalence [1]. The premature fusion of one or more cranial sutures leads to abnormal head shape and makes them prone to airway obstruction and raised ICP [2-4]. Such patients need surgery for control of raised ICP, delay the deformity, correction of cranial vault and for associated condition correction like syndactyly repair. The current manuscript discusses anaesthetic management of two infants with Apert syndrome and Crouzon syndrome and the literature review of SC.

Case 1

A three and half-month-old male infant, presented with hydrocephalus since birth. The birth history was not available as the patient was brought from an orphanage. The current weight was 2.8 kg (<5 percentile) and head circumference 38 cm (15 percentile). The infant presented with a few episodes of vomiting, feeding problem, bilateral coronal synostosis with raised ICP and was diagnosed as Crouzon Syndrome. The patient was planned for endoscopic third ventriculostomy [Table/Fig-1].



[Table/Fig-1]: Facial feature of case 1, Crouzon Syndrome.

After correction of dehydration and maintaining fasting guidelines, the patient was taken up for the surgery keeping special anaesthetic considerations in mind [Table/Fig-2]. In the operating room, standard American Society of Anaesthesiologists (ASA) monitors were attached, 24G intravenous cannula was in situ and the patient was induced with intravenous fentanyl 2 µg/kg and a

Variables	Patient 1- Crouzon syndrome	Patient 2- Apert syndrome
1. Clinical investigations	Blood counts, renal and coagulation profile were normal with haemoglobin of 10.2 gm/dL. On fundoscopy, papilledema was present and 2D Echocardiography revealed the atrial septal defect. Magnetic Resonance Imaging brain and whole spine revealed brachycephaly, dilated third and lateral ventricle, prominent dural sac and tethering of cord.	All the blood investigations were within normal limits. 2D Echocardiography showed mild tricuspid regurgitation with normal pulmonary vasculature.
2. Features of anaesthetic considerations	Difficult mask ventilation- Depressed nasal bridge, hydrocephalus, midface hypoplasia, beak and narrow nose, large flat forehead. Difficult laryngoscopy- High arched palate, macroglossia Raised Intracranial Pressure (ICP) Cardiac involvement- Atrial Septal Defect eye care for proptosis	Difficult airway- large head compared to body size, modified Mallampati Grade 3, flat facial profile, large forehead, depressed nasal bridge, high arched palate, macroglossia, micrognathia, bifid uvula, epidermoid cyst over gingiva, short neck, maxillary hypoplasia. Cardiac involvement- Mild tricuspid regurgitation eye care for proptosis

[Table/Fig-2]: Clinical investigations and features of anaesthetic considerations in Syndromic Craniosynostosis (SC).

graded dose of propofol 2 mg/kg. Bag and mask ventilation was managed by two hand techniques, head tilt, chin lift, continuous positive pressure and oropharyngeal airway. Laryngoscopy was anticipated to be difficult, intravenous xylocard 1.5 mg/kg and atracurium 0.5 mg/kg were given to blunt intubation response. Gentle direct laryngoscopy showed Cormack Lehane view grade 2, endotracheal intubation was done with size 3.5 endotracheal tube and fixed at 10 cm. The anaesthesia was maintained with oxygen and air (50:50) with isoflurane, intermittent doses of fentanyl, dexamethasone 0.1 mg/kg and paracetamol 30 mg/kg suppository. Appropriate eye care was given as the patient had proptosis and exposure keratitis. To avoid any rise in ICP; intubation response was blunted, neutral position of the head, avoidance of hypoxia and hypercarbia were practised. To minimise left to right shunt through Arterial Septal Defect (ASD), all the measures were taken to prevent a rise in systemic and pulmonary vascular resistance. The haemodynamic parameters, airway pressure and end-tidal CO_2 were normal with adequate depth of anaesthesia throughout the surgery. The surgery continued for two hours 20 minutes. Blood loss was minimal and the patient remained euthermic. After completion of surgery neuromuscular block was reversed and on the assurance of complete recovery, the trachea was extubated. The infant had an uneventful course and was discharged on the fifth postoperative day.

Case 2

A seven-month-old male infant, presented with a history of absent fingers since birth for syndactyly repair. The infant was a pre-term baby, born by caesarean section because of premature rupture of membrane and abnormal shape of the head, which was diagnosed in term ultrasonography. The conception was normal, from non consanguineous marriage. The baby cried immediately after birth. The birth weight was 2.75 kg (<5 percentile), head circumference 38 cm (15 percentile) and body length 57 cm (15 percentile). The patient had an abnormal shape of head, brachycephaly, craniosynostosis and was diagnosed with Apert syndrome. The repair of the cranial vault was done at two months of age. The patient's father was under treatment for schizophrenia for the last 10 years.

The milestones of the infant were normal. He was immunised to date. He had proptosis, hypertelorism, large low set ears, large philtrum, no history of seizures, any upper respiratory tract infection, weakness of any limbs, cyanosis, jaundice, bowel and bladder incontinence [Table/Fig-3].

[Table/Fig-3]: Facial features of case 2, Apert syndrome.

Fasting guidelines were followed preoperatively. Mask ventilation and laryngoscopy was anticipated to be challenging and a difficult airway cart was kept ready. General anaesthesia was the plan with Laryngeal Mask Airway (LMA) placement with spontaneous ventilation [Table/Fig-2]. The standard ASA monitors were attached to the patient and were induced with inhalational induction using oxygen and sevoflurane, mask ventilation was difficult so the two-hand technique and oropharyngeal airway were used, after which IV cannula was secured in the left hand. The infant weighed 7 kg (<15 percentile). Intraoperatively fentanyl 2 µg/kg intravenously, paracetamol suppository 30 mg/kg and bilateral wrist block with 4 mL of 0.25% bupivacaine was used for analgesia. The airway was secured by Laryngeal Mask Airway (LMA) Proseal size 1.5, adequate ventilation was assured by end-tidal carbon dioxide monitoring. The anaesthesia was maintained using oxygen and air (50:50) with sevoflurane to maintain a Monitored Anaesthesia Care (MAC) of one. The patient remained haemodynamically stable and euthermic throughout the surgery. After completion of the surgery, LMA Proseal was removed when the patient was fully awake.

DISCUSSION

The SC affects multiple cranial sutures and leads to midface hypoplasia with other associated defects of the skull base and limb abnormality, while non syndromic/simple SC involves a single suture [2]. The major challenges faced during the surgery were raised ICP, airway obstruction, difficult bag and mask ventilation, high arched palate, macroglossia, retrognathia, exorbitism, abnormal feeding pattern making them prone for electrolyte disturbances. There can also be disturbances in intellectual and neurologic development along with associated congenital heart diseases.

These children can come for various surgeries like increased ICP, severe exophthalmos, obstructive sleep apnea (OSA), craniofacial deformity, prevention of neurologic sequelae and psychosocial concerns. The preoperative evaluation should be tailored to each patient as the severity of the disease and presentation may vary between patients. The difficult airway is a common feature in SC that can make mask ventilation and laryngoscopy challenging [1,2,5,6]. Any history of snoring and OSA mandates the multidisciplinary team involvement to manage the anticipated difficult airway. OSA can be treated by non-invasive ventilation including Bilevel Positive Airway Pressure (BiPAP) and Continuous Positive Airway Pressure (CPAP) [1]. The preanaesthetic evaluation and optimisation should focus on optimising the respiratory condition of patients as preoperative wheeze is common in Apert syndrome leading to the postponement of surgeries [7,8]. Premedication should be titrated as over sedation can cause airway obstruction in these children.

The infant with Crouzon syndrome was planned for endoscopic third ventriculostomy, in view of raised ICP. The less invasive nature of surgery was associated with less blood loss. The invasive surgeries required for correction of craniosynostosis are of long duration and carry the risk of extensive blood loss, hypothermia and venous air embolism [2,5,9]. Recent advanced surgeries are done by the minimally invasive endoscopic release of fused sutures with new fixation devices like helmets, springs and distractors [10]. This has helped in decreasing the duration of surgery, complications, blood loss and re-fusion of the suture. This development has led to staged surgery and needs a multidisciplinary approach [11]. Preoperative optimisation with erythropoietin and iron supplement, intraoperative controlled hypotension and antifibrinolytics are found to be effective in minimising blood loss in literature [2,6,12].

A study done by Lionel KR et al., found that the incidence of the difficult airway was predicted to be 76% in patients of SC, which required awake fiberoptic bronchoscopy and video laryngoscopy for intubation, while in another study, the incidence was found to be almost 58% [6,12]. In the current series, both the infants were labelled as difficult airways in terms of the bag and mask ventilation, which needed two hand techniques, head tilt and chin lift. For patient with Crouzon syndrome, intubation was performed

using Millers blade and for patient with Apert syndrome LMA proseal was used to secure the airway. To manage difficult mask ventilation, two-handed technique, head tilt, chin lift, jaw thrust, oropharyngeal airway, lateral position and continuous positive pressure are recommended. Classical and Proseal LMA are found to be useful to manage the difficult airway in the paediatric age group [13]. There are newer devices that can be used for endotracheal intubation like paediatric sizes of Glidescopevideo laryngoscope, Miller blade, Airtraq optical laryngoscope and fiberoptic bronchoscope [14].

The association of raised ICP is found to be maximum with Crouzon and Pfeiffer syndrome (50-70%), followed by Apert syndrome (40-50%) [1]. The induction of anaesthesia was planned to avoid any rise in ICP by careful selection of drugs, blunting the hemodynamic responses of intubation by intravenous Xylocard administration and adequate depth of anaesthesia. An appropriate position, based on the need of surgeons was provided keeping in mind to facilitate venous drainage to escape rise in ICP. Hypercarbia and hypoxia were avoided. Ocular proptosis may require special attention and protection with padding along with joints and peripheral nerves.

For invasive surgeries, additional monitors like arterial catheters and central venous access should be considered along with standard ASA monitoring requirements. Cardiac involvement is common among Crouzon and Apert syndrome, which should be well investigated [5]. A coagulation profile is a must. Hearing impairment and compromised cognitive functions in SC patients can make communication difficult. The postoperative period may get complicated due to large intraoperative blood loss and fluid shift, which mandates intensive care and monitoring. In current series, at the end of the surgery both the patients were extubated in view of non invasive surgery, and followed-up for five days.

CONCLUSION(S)

The treatment of craniosynostosis usually involves surgery to unlock the bones and reshape the skull,hence anaesthetic management plays a major role in the outcome of the procedure. The anaesthetic considerations depend on the age of patients, associated system aberrations and type of planned surgery. It mandates a well-planned, multidisciplinary approach.

REFERENCES

[1] Mathijssen INJ. Gideline for care of patients with the diagnoses of craniosynostosis. Working group on craniosynostosis. Journal of Craniofacial Surgery Sept 2015;26(6):1735-807.
[2] Thomas K, Hughes C, Johnson D, Das S. Anesthesia for surgery related to craniosynostosis: A review. Part 1. Paediatric Anesthesia. (2012);22:1033-41.
[3] Sharma A, Patel N, Arora S, Ramachandran R. Child with Saethre-Chotzen syndrome: Anesthetic management and literature review. Acta Anaesthesiol Belg. 2014;65(4):179-82.
[4] Vaughan C. Anesthetic management of children with craniofacial anomalies. CRNA. 1997;8(4):123-34.
[5] Pearson A, Matava CT. Anaesthetic management for craniosynostosis repair in children. BJA Education. 2016;16(12):410-16.
[6] Lionel KR, Moorthy RK, Singh G, Mariappan R. Anaesthetic management of craniosynostosis repair– A retrospective study. Indian J Anaesth. 2020;64(5):422-25.
[7] Elwood T, Sarathy PV, Geiduschek JM, Ulma GA. Respiratory complications during anaesthesia in Apert syndrome. Paediatric Anesthesia. 2001;11(6):701-03.
[8] Nargozian C. Apert syndrome. Anesthetic management. Clin Plast Surg. 1991;18(2):227-30.
[9] Hughes C, Thomas K, Johnson D, Das S. Anaesthesia for surgery related to craniosynostosis: A review. Part 2. Paediatric Anesthesia. 2013;23:22-27.
[10] Practor MR, Meara JG. A review of the management of single-suture craniosynostosis, past, present, and future. J Neurosurg Paediatr. 2019;24:622-31.
[11] Lee JM, Gee E, Liu CA. Anesthesia for innovative paediatric surgical procedures. Anesthesiol Clin. 2020;38(3):493-508.
[12] González Cárdenas VH, Vanegas Martínez MV, Rojas Rueda ME, Guevara NS, Prada JR, Baquero P. Anaesthesia in craniosynostosis. Colomb J Anesthesiol. 2014;42:199–204.
[13] White MC, Cook TM, Stoddart PA. A critique of elective paediatric supraglottic airway devices. Paediatr Anaesth. 2009;19(Suppl 1):55-65.
[14] Doherty JS, Froom SR, Gildersleve CD. Paediatric laryngoscopes and intubation aids old and new. Paediatr Anaesth. 2009;19(Suppl 1):30-37.

PARTICULARS OF CONTRIBUTORS:
1. Assistant Professor, Department of Anaesthesia and Critical Care, Manipal TATA Medical College, Manipal Academy of Higher Education, Jamshedpur, Jharkhand, India.
2. Senior Consultant, Department of Anaesthesiology, Bangalore Baptist Hospital, Bangalore, Karnataka, India.
3. Consultant, Department of Anaesthesiology, Bangalore Baptist Hospital, Bangalore, Karnataka, India.
4. Senior Consultant, Department of Anaesthesiology, Bangalore Baptist Hospital, Bangalore, Karnataka, India.

NAME, ADDRESS, E-MAIL ID OF THE CORRESPONDING AUTHOR:
Smriti Sinha,
Dlx 81, Carnation, Ashana Garden, Sonari, Jamshedpur, Jharkhand, India.
E-mail: smritii576@gmail.com

PLAGIARISM CHECKING METHODS: [Jain H et al.]
- Plagiarism X-checker: May 13, 2021
- Manual Googling: Oct 30, 2021
- iThenticate Software: Jan 20, 2022 (1%)

ETYMOLOGY: Author Origin

AUTHOR DECLARATION:
- Financial or Other Competing Interests: None
- Was informed consent obtained from the subjects involved in the study? Yes
- For any images presented appropriate consent has been obtained from the subjects. Yes

Date of Submission: **May 12, 2021**
Date of Peer Review: **Sep 18, 2021**
Date of Acceptance: **Dec 22, 2021**
Date of Publishing: **Apr 01, 2022**