Anesthesia Management in Craniosynostosis Surgery: A Retrospective, Single-center Experience

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ABSTRACT

Aim and objective: We report our experience with anesthesia management of craniosynostosis surgeries at a tertiary care children’s hospital.

Materials and methods: We conducted a retrospective analysis of craniosynostosis surgeries performed at our institute over the last 3 years.

Review results: Twelve children underwent reconstructive surgery over a period of 3 years. Eight patients underwent IV induction with propofol followed by atracurium. In four patients where difficult airway was anticipated, an inhalational induction with sevoflurane was performed. Anesthesia was maintained using air, oxygen, and sevoflurane delivered through a closed circuit, and dexametomidine @ 0.5 µg/kg/hour. Fentanyl and paracetamol were used for analgesia. Monitoring included standard ASA monitors and additionally, arterial blood pressure and urine output monitoring. Tranexamic acid was used to reduce bleeding. There was no incidence of major intraoperative complications viz. venous air embolism, massive hemorrhage, or hemodynamic instability. No patient needed vasoactive infusion support.

Conclusion: Surgery for craniosynostosis poses several challenges for anesthesia. Having a protocolized approach to anesthesia management and transfusion can result in good outcomes.

Clinical significance: Raised intracranial pressure, obstructive sleep apnea, syndromic associations are common in craniosynostosis. Surgery in infancy poses additional challenges of massive blood loss. The anesthetist has to be vigilant and step up monitoring to detect and manage perioperative complications.

Keywords: Anesthesia management, Craniosynostosis, Retrospective review.

Research and Innovation in Anesthesia (2021): 10.5005/jp-journals-10049-0107

INTRODUCTION

Craniosynostosis is a disorder of skull development that occurs due to premature fusion of one or several cranial sutures. The incidence is approximately 1 in 2,500 births. It often occurs as an isolated condition but may be part of a syndrome in up to 40% of cases. The more common associations include Pfeiffer, Apert, Crouzon, Muenke, Saethre-Chotzen, and Carpenter syndromes.¹

Early fusion of the cranial sutures results in restricted growth of the skull perpendicular to the affected suture, with compensatory bone growth parallel to the affected suture to accommodate the growing brain. The resultant deformity of the skull and face leads to the obvious esthetic abnormality, but may also be associated with increased intracranial pressure (ICP), hindbrain herniation, and obstructive sleep apnea (OSA).

Surgery in infancy is advocated for better neurodevelopmental outcomes but poses several challenges for anesthesia. Syndromic craniosynostosis has its added anesthesia implications peculiar to the syndrome complex. We report our experience with anesthesia management of craniosynostosis surgeries at a tertiary care children’s hospital.

MATERIALS AND METHODS

We performed a retrospective analysis of all craniosynostosis surgeries performed at our hospital for 3 years from 2018 to 2021. Institutional Review Board approval for the study and publication of results was obtained. Data were retrieved from hospital records and perioperative anesthesia charts. Results are presented as simple mean and percentages and depicted in tabular form.

RESULTS

Twelve children underwent reconstructive surgery over a period of 3 years. The demographic details and the type of sutural involvement are listed in Table 1. Three patients were syndromic; two with Crouzon’s syndrome, and one with Apert’s syndrome. Three patients had symptoms suggestive of OSA. In four patients, a difficult airway was anticipated. Six patients had symptoms suggestive of raised ICP viz. irritability, visual defects, and proptosis. One patient had a history of seizures. A 3-month-old infant was operated using an endoscopic approach, the rest had open surgery. The average duration of anesthesia and surgery was 4 hours 50 minutes.

Preoperative investigations included hemogram and coagulation profile. All patients underwent echocardiography, no patient had a structural cardiac anomaly. One patient had severe pulmonary hypertension. Standard fasting guidelines were
followed. A high-risk consent, consent for transfusion of blood and blood products, and postoperative ICU admission and ventilation were obtained in all patients. The difficult airway trolley was readied in all patients. All patients had an intravenous line inserted before anesthesia induction and were posted as 1st on the operation theater list.

Eight patients, who were deemed to have an “easy airway” on preoperative assessment, received sedative premedication. Inj. midazolam 0.05 mg/kg IV in the preoperative holding area. All patients received glycopyrrolate 4 μg/kg and fentanyl 2 μg/kg before induction. Eight patients underwent IV induction with propofol followed by atracurium as a muscle relaxant. In four patients where difficult airway was anticipated, an inhalational induction with sevoflurane was performed. After confirming the ability to mask ventilate, atracurium was administered to three of these patients. In one patient, following sevoflurane induction, a check laryngoscopy was performed, and atracurium was administered only after confirming the glottic view. Ten patients were intubated on the 1st attempt using a direct laryngoscope with a Macintosh blade. Two patients with syndromic craniosynostosis needed frowa® intubating introducer for intubation, and one patient in this subset was intubated in the 3rd attempt.

Anesthesia was maintained using air, oxygen, and sevoflurane delivered through a closed circuit, and dexmedetomidine @ 0.5 μg/kg/hour. Intermittent fentanyl boluses (1 μg/kg/hour) and inj. paracetamol (15 mg/kg) were used for perioperative analgesia.

Monitoring included standard ASA monitors and additionally, arterial blood pressure and urine output monitoring. In all patients radial arterial access was secured for sampling and continuous blood pressure monitoring. Two peripheral venous cannulas (1 preexisting, and another large bore–22G secured after induction) were cited for volume replacement. For temperature management, convective warmer (Bair Hugger®) and fluid warmer were used.

Inj. tranexamic acid was administered in a loading dose of 10 mg/kg followed by infusion of 1 mg/kg/hour till the end of surgery. Preoperative hemoglobin averaged 12.42 g/dL (9.3–14 g/dL). Average intraoperative blood loss was 18.2 mL/kg (4.5–34 mL/kg). Ringer’s lactate was administered @ 6 mL/kg/hour. All patients received packed cell transfusion. No patient received fresh frozen plasma or platelet transfusion. Hemodynamics were maintained within acceptable limits, and no patient needed vasopressors or inotropes. Dexamethasone 0.2 mg/kg was administered after induction. All patients received ondansetron 0.1 mg/kg before extubation. Arterial blood gas estimations showed a tendency of patients to develop a base deficit averaging –7 mEq/L by the end of surgery. Eleven patients were extubated on the table after they achieved extubation criteria. One patient with OSA and severe PH was electively ventilated and extubated on postoperative day (POD) 1. All patients were shifted to the intensive care unit following surgery. Significant facial edema developed in all patients following surgery and settled in 3 days. Enteral feeding was started on POD 1. Postoperative analgesia was managed using IV paracetamol QDS and IV tramadol TDS. The average ICU stay was 2.5 days and the average hospital stay was 6.3 (5–9) days. There was no incidence of major intraoperative complications viz. venous air embolism, massive hemorrhage, or hemodynamic instability. No patient needed vasoactive infusion support. Hemoglobin dropped by 1 g/dL on POD 1 as compared to the immediate post-surgery value. However, no patient needed postoperative packed cell transfusion. There were no postoperative complications viz. CSF leak, or flap infection.

**Discussion**

We report a single-center experience of anesthesia management of craniosynostosis surgery. Although there are several publications detailing surgical and anesthesia aspects in international journals, we found only a few case reviews and some case reports from India after an extensive literature search. While this could well be due to a tendency to not publish, we believe that surgery for craniosynostosis is not very commonly performed in many centers in India. We share our experience of managing these surgeries and discuss the myriad implications of such procedures.

The surgeries in our hospital were performed independently by two neurosurgeons, assisted by two plastic surgeons. Although anesthesia was administered by different consultants from our department, we have a set protocol, which maintains uniformity in management.

Children with craniosynostosis have a high incidence of sleep-disordered breathing, ranging from snoring to OSA. Although multifactorial, a predominant contributing factor is midface hypoplasia. Children with syndromic craniofacial malformations are more likely to have more severe OSA with an incidence as high as 87%. Central apnea may occur as a result of pressure on the respiratory centers due to an underlying Chiari malformation or due to narrowing of the cranio cervical junction.

Several factors can cause increased ICP in children with craniosynostosis, including craniocelebral disproportion, abnormal intracranial venous drainage, and hydrocephalus. Obstructive sleep apnea can exacerbate these factors or even cause raised ICP by itself, due to its effects on CO2, a potent vasodilator, and its

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**Table 1: Patient demographics**

<table>
<thead>
<tr>
<th>Parameters</th>
<th>Mean (range)</th>
</tr>
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<tbody>
<tr>
<td>Age (months)</td>
<td>38.6 (3–72)</td>
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<tr>
<td>Weight (kg)</td>
<td>13.08 (4.5–20)</td>
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<tr>
<td>Preoperative Hb (g/dL)</td>
<td>12.42 (9.3–14)</td>
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<tr>
<td>Intraoperative blood loss (mL/kg)</td>
<td>18.2 (4.5–34)</td>
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<tr>
<td>Duration of hospital stay (days)</td>
<td>6.3 (5–9)</td>
</tr>
</tbody>
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<table>
<thead>
<tr>
<th>Gender</th>
<th>Number of patients</th>
<th>Percentage</th>
</tr>
</thead>
<tbody>
<tr>
<td>Male</td>
<td>7</td>
<td>58.3</td>
</tr>
<tr>
<td>Female</td>
<td>5</td>
<td>41.7</td>
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<table>
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<tr>
<th>Features of raised ICP</th>
<th>Number of patients</th>
<th>Percentage</th>
</tr>
</thead>
<tbody>
<tr>
<td>Difficult airway</td>
<td>4</td>
<td>33.3</td>
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<tr>
<td>Syndromic association</td>
<td>3</td>
<td>25</td>
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<table>
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<tr>
<th>Shape of head</th>
<th>Number of patients</th>
<th>Percentage</th>
</tr>
</thead>
<tbody>
<tr>
<td>Dolicocephaly</td>
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<td>16.7</td>
</tr>
<tr>
<td>Brachycephaly</td>
<td>7</td>
<td>58.3</td>
</tr>
<tr>
<td>Frontal plagiocephaly</td>
<td>2</td>
<td>16.7</td>
</tr>
<tr>
<td>Trigonocephaly</td>
<td>1</td>
<td>8.3</td>
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<table>
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<tr>
<th>Type of surgery</th>
<th>Number of patients</th>
<th>Percentage</th>
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</thead>
<tbody>
<tr>
<td>Invasive/open</td>
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<td>91.7</td>
</tr>
<tr>
<td>Endoscopic</td>
<td>1</td>
<td>8.3</td>
</tr>
</tbody>
</table>

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cardiovascular effects on arterial blood pressure and thus cerebral perfusion pressure.\textsuperscript{12} Raised ICP may be present in infants with isolated single suture craniosynostosis but is more common in syndromic craniosynostosis with multiple suture involvement.

Surgery for craniosynostosis correction includes total calvarial reconstruction, fronto-orbital advancement, or posterior cranial vault reconstruction. These procedures are typically performed in late infancy. Fronto-orbital advancement and reconstruction procedures are performed with the patient in the supine position and involve a wide bicoronal incision followed by a frontal craniotomy and separation of the orbital rim. The excised bones are then cut, refashioned, and replaced using wires or reabsorbable plates and screws.

For surgery done in infancy, the physiologic nadir of hemoglobin may coincide with the timing of surgery, necessitating an increased need for transfusion. Performing surgery after 6 months, after the nadir of hemoglobin, but before the bone deformity gets too extensive due to skull growth and ossification of the dura may reduce transfusion requirements.\textsuperscript{13}

Minimally invasive craniosynostosis surgery techniques, e.g., endoscopic strip craniectomy, spring-assisted craniosynostosis surgery are associated with less blood loss and reduced hospital stay.\textsuperscript{14} Data from the Pediatric Craniofacial Surgery Perioperative Registry and other groups demonstrates additional benefits of shorter anesthesia and surgery durations, lower rates of postoperative intubation, and lower ICU utilization.\textsuperscript{15} Several large volume centers report shifting nonsyndromic patients directly to the postsurgical ward with no complications following endoscopic surgery.\textsuperscript{16,17}

Preoperative evaluation should include a good history and evaluation to identify comorbidities. Our laboratory testing includes a hemogram, coagulation profile, and type and cross match. Three units of packed cells are reserved for every case. Average transfusion in craniosynostosis surgery can range from 50 to 100 mL/kg.\textsuperscript{4,5} Greatest bleeding occurs during scalp dissection and osteotomy.

Several strategies to reduce blood use intraoperatively have been described including the use of local vasoconstrictors, autologous blood salvage, preoperative use of iron supplements, and recombinant erythropoietin, intraoperative hemodilution, and accepting lower transfusion triggers. Establishing a unit policy and transfusion protocol helps reduce transfusion requirements.\textsuperscript{18,19}

The antifibrinolytic drug tranexamic acid has been shown to reduce blood loss in craniosynostosis surgery. Different dose regimens of tranexamic acid have been used to reduce the blood loss in various surgeries ranging from 10 to 100 mg/kg, followed by 1–10 mg/kg/hour infusion.\textsuperscript{20,21} Low-dose tranexamic acid (5 mg/kg) is just as effective as high doses (50 mg/kg).\textsuperscript{21} A combined intra- and postoperative infusion has been effective in reducing overall transfusion rates in CS surgery.\textsuperscript{22}

Our surgeons use a cocktail of lignocaine with epinephrine, hyaluronidase, and saline to infiltrate the scalp. We use tranexamic acid bolus at 10 mg/kg followed by 1 mg/kg/hour till the end of surgery. Some centers start vasopressor infusions routinely to combat anesthesia-induced vasodilatation and to maintain hemodynamics in the face of ongoing continuous blood loss.\textsuperscript{23} It is our practice to start blood early, and none of our patients needed vasopressor support.

VAE is a possible complication during neurosurgical procedures due to the presence of noncollapsible veins and the surgical site being above the level of the heart. The incidence of this complication may be lower in minimally invasive procedures.\textsuperscript{16,24}

Hypothermia is a problem since the large surface area of the head is exposed to the atmosphere. Even mild hypothermia can impair coagulation enzyme function and is associated with increased bleeding and transfusion requirements. We monitored core and skin temperatures during surgery and used convective warming systems to prevent heat loss. All infusions were administered through a fluid warmer.

Surgery can be performed with patients in several positions—supine, prone, or the modified sphinx position, depending on the type of CS. Our patients were all operated on in the supine position, with head elevation to reduce bleeding. Careful attention must be given to the prevention of pressure injury.

During cranial vault reconstruction procedures in the supine position, surgeons prefer that the eyes not be taped closed because the eyes are within the surgical field. Syndromic children often have propotosis. Our surgeons perform tarsorrhaphy to protect the eyes. Ophthalmic ointment should be applied and careful attention paid to prevent eye injuries. When a surgeon is operating near the orbits, the oculocardiac reflex may be triggered and bradycardia observed. It is usually self-limiting and reverts as the surgeon releases the pressure.

It is important to secure the endotracheal tube well, due to the relative inaccessibility of the airway. Reconfirming the correct tube position after patient positioning is important. We use loban\textsuperscript{a} to secure the tracheal tube which anchors it well and prevents loosening of the tube sticking from saline or blood.

Invasive arterial pressure monitoring is recommended because it allows for the immediate detection of hypotension (due to VAE, or bleeding) and frequent blood sampling.\textsuperscript{1} It is difficult to assess blood loss, as there is considerable seepage into and underneath surgical drapes and the irrigation fluid further makes accurate assessment challenging. In addition to steady blood loss that occurs during the procedure, precipitous hemorrhage can also occur with inadvertent dural venous sinus tears or disruption of large emissary veins. We found that postoperative ooze into the wound dressing resulted in a dip in the hemoglobin on POD 1. For these reasons, we find it convenient and useful to insert the arterial line and remove it only on POD 2 or 3 when the patient stabilizes.

CVP monitoring has proven to be not very useful to guide fluid therapy, hence we avoided central venous cannulation in our patients. Implementation of central venous pressure monitoring in infants undergoing complex craniofacial reconstruction has not resulted in a reduction in the incidence or duration of hypotension.\textsuperscript{2} Adequate venous access is vital; 22-G or larger catheters are preferred. Blood and products are better transfused through a peripheral line. The only indication for inserting a central venous access line is the absence of good peripheral lines, or the need to start vasopressors.

**Conclusion**

We report our experience with perioperative management of surgery for craniosynostosis. Obstructive sleep apnea, raised ICP, and difficult airway are common associations in craniosynostosis. Intraoperative bleeding is expected, and having a protocolized approach to anesthesia management and transfusion can result in good outcomes.
REFERENCES


