Perioperative management of patients undergoing craniosynostosis repair

Rajni Mathur[1], Tuhin Mistry[2*], Gaurav Sharma[3], Vandana Mangal[4]

Senior Professor[1,4], Department of Anaesthesiology, S.M.S. Medical College, Jaipur, Rajasthan, India. Post-Doctoral Fellow in Regional Anaesthesia[2], Department of Anaesthesiology, Ganga Medical Centre and Hospitals Pvt Ltd, Coimbatore, Tamil Nadu, India. Associate Professor[3], RUHS College of Medical Science, Jaipur, Rajasthan, India.*

Craniosynostosis is a type of craniofacial malformation because of premature fusion of cranial suture. The primary treatment is surgical correction which is usually done in stages over years to get best possible outcome. We did a retrospective analysis of five years from 1st August 2011 to 30th July 2016 in which we included twelve patients of craniosynostosis requiring corrective surgeries under general anaesthesia in our institution.

Keywords: Craniosynostosis; general anaesthesia; reconstructive surgery; blood loss; syndromic child

Introduction

Craniosynostosis is the premature fusion of one or more cranial sutures, resulting in an abnormal shape of head either from a primary defect of ossification (primary craniosynostosis) or as part of other syndromes (secondary craniosynostosis). It may be simple or single suture synostosis (only one suture) and complex or compound or multiple synostosis or craniosynostosis (more than one suture). Though craniosynostosis is a fairly prevalent disorder (1 per 2000 live births), syndromic craniosynostosis (SC, with other body deformities or systemic involvements) are relatively rare. The commonly associated syndromes include Crouzon, Apert and Pfeiffer, Saethre-Chotzen and Muenke syndromes with incidence of 1 in 25,000;1 in 100,000; 1 in 100,000; 1 in 25,000 to 50,000 and 1 in 10,000 live births respectively.[1]

The management of children with craniosynostosis is a multidisciplinary team approach. Providing anaesthesia for these patients presents unique challenges in terms of difficult airway, blood loss, prolonged anaesthesia in paediatrics, syndrome-specific issues, metabolic disturbance and post-operative complications.

*Correspondence: Tuhin Mistry
E mail: dr.tuhin2014@gmail.com
https://orcid.org/0000-0003-1904-4831
Received: 08/09/2018
Accepted:24/12/2018
DOI: http:/doi.org/10.4038/slja.v27i1.8378

Case histories

We did a retrospective review of all the craniosynostosis cases done within five years (from 1st August 2011 to 30th July 2016) at a tertiary care center (Department of Anaesthesiology, S.M.S. Medical College, Jaipur, Rajasthan, India). We included twelve patients of craniosynostosis requiring corrective surgeries under general anaesthesia. Case specific presentation, examination findings and investigations in our patients are described in Table 1.

Discussion

Corrective surgeries involve craniofacial reconstruction to separate the fused bones which is mostly started in early childhood and performed in stages over years. Pre-anesthesia checkup (PAC) is undoubtedly an extremely important part of the anesthetic care for cranial vault surgery in patients with SC. Babies with craniosynostosis, particularly in case of fusion of multiple sutures or SC may develop increased intracranial pressure as their skulls don't expand enough to make room for growing brains which may lead to various manifestations including visual difficulties even blindness, nausea and vomiting, somnolence, headaches, seizures, brain damage and death in rare instances.[2]

Table 1: Case-based presentation and management of patients

Age/Sex	History	Examination and Investigations	Associated syndrome	Surgery	Postoperative period
14yrs male	reflux of food into nasal cavity, unable to close the eyes completely, abnormal shape of head and face since birth	B/L proptosis, Hypertelorism and fused fingers of both hands (Type III syndactyly). systemic examination, blood investigations, Chest X-ray, ECG and 2D echo findings were within normal limits	Apert Syndrome	cranial vault remodelling with fronto-orbital advancement	satisfactory recovery discharged after 7 days
19yrs male	swelling at root of nose since birth which was gradually increasing in size and increase distance between the eyes. The mass increases in size during coughing	no other physical or mental abnormalities on clinical examination. CT scan findings revealed cystic lesion on right side of nose with a skull defect at floor of anterior cranial fossa at cribriform plate of the ethmoid on right side lateral to the nasal septum	-	Craniotomy with excision of sac and box osteotomy for hypertelorism correction	Uneventful, discharged after 8 days
1yr male	deformity of head and face, both upper and lower limbs (fused fingers and toes) since birth with delayed developmental milestones, h/o upper and lower respiratory tract infection	craniosynostosis of coronal suture, open anterior and posterior fontanelle, gaping of sagittal suture, frontal bossing/L parietal eminence exaggerated. Increased nasofrontal angle, hypertelorism, B/L proptosis, hypoplasia of mid face, high arched palate with cleft palate, B/L syndactyly of hands (fused 2^{nd},3^{rd},4^{th}& 5^{th} digits), B/L fused toes. USG brain & 2D echocardiography were within normal limits	Apert Syndrome	craniotomy with fronto-orbital advancement	Uneventful, discharged after 7 days.
13yrs female	swelling over left forehead and bulging of left eyeball which started and gradually increased over 1 year with no history of headache, vomiting and seizure. She underwent percutaneous balloon pulmonary valvuloplasty for valvular stenosis 11 years back	swelling was bony hard, smooth contoured, round with abnormal protrusion of left eyeball. fibrous dysplasia of left frontal bone and roof of orbit.	-	Deroofing of left orbit and frontal bone contouring were done under GA	Satisfactory, discharged after 5 days.
19yrs female	Headache, swelling of right forehead and protrusion of right eye for 5 months. Hypothyroidism for 2 years (was on l-thyroxin) and seizure disorder for 4 years (was on levetiracetam, carbamazepine and clobazam)	swelling was bony hard, diffuse, proptosis of right eyeball. fibrous dysplasia of right frontal bone and skull base	-	Cranioplasty with right optic nerve decompression	Blurring of vision 6 hours after surgery. Examination revealed congested right eye, no finger counting or hand movement, but perception of light and projection of rays were present with pale optic disc. she was given pulse methyl prednisolone

					therapy and moxifloxacin. stayed 4 days in hospital and discharged.
2yrs male	swelling over parietal and occipital region since birth	a soft cystic globular swelling present over occipital region, overlying skin is thin, no prominent vascular markings, positive transillumination test. He was diagnosed to have occipital encephalocele with craniosynostosis. NCCT and MRI of brain were suggestive of occipital encephalocele with herniation of cerebellar hemisphere.	-	intraop cardiac arrest (ventricular tachycardia followed by bradycardia) which was revived, and patient was shifted to ICU after surgery	was on inotropic support and mechanical ventilation because of shock and respiratory failure. patient had another episode of cardiac arrest 4 hours after surgery during ICU stay but could not be revived inspite of all efforts.
22yrs male	increased distance between two eyes and depressed nasal bridge since birth. She was operated for squint and nasal dorsal augmentation with cranial bone graft one year back.	systemic examination, blood investigations, Chest X-ray, ECG and 2D echo findings were within normal limits	-	Box osteotomy under GA	satisfactory recovery. discharged after 7 days
11yrs female	Operated previously with Le Forte II advancement	Mid face hypoplasia, class IV malocclusion & mid-face hypoplasia	Crouzon's syndrome	Le Forte III advancement osteotomy	Satisfactory, Discharged after 6 days
1yr female	Deformity of head and face since birth	Asymmetry of the head		B/L Fronto-orbital advancement with cranial vault remodelling	Uneventful discharged after 7 days
2yrs female	Increasing head size more than normal since birth, past history: right parietal burr hole with ventricular tap done 2 months back but as the CSF pressure was very low, VP Shunt was not done	Increase circumference of head with prominent forehead, sunset sign in both eyes(hydrocephalus)	Craniosynostosis with aqueductal stenosis with type I Arnold Chiari malformation	Fronto-temporo-parietal remodelling	After the surgery, patient was shifted to ICU under antibiotics, steroid and anticonvulsant. On the second post-operative day she had one episode of seizure following which she went into asystole. CPCR started

					and revived but inspite of all efforts she passed away on 3rd post-operative day.
8yrs male	Asymmetric growth of skull and abnormal appearance of both eyes which gradually became apparent after birth	B/L proptosis, NCCT & CECT brain revealed type I Arnold Chiari malformation and luckenschaden skull	Crouzon's syndrome	B/L fronto orbital advancement with Le Forte III	Uneventful discharged after 9 days
18yrs female	Increased distance between eyes since birth, difficulty in learning and decreased performance in school	Distance between the medial canthi was 35 mm	-	box osteotomy, medialisation of orbit and dorsal nasal augmentation with bone graft	Satisfactory, discharged after 8 days

Airway should be assessed thoroughly during PAC as most of the patients are in paediatric age. Cartilaginous abnormalities of the trachea, fusion of the cervical vertebrae, tracheal stenosis and angular deviation of the trachea contribute to respiratory morbidity or difficult intubation in SC.[3] In addition, presence of wheeze and obstructive sleep apnoea complicates the scenario. We should be prepared with difficult airway cart to manage loss of airway patency, risk of difficult intubation and respiratory complications during general anaesthesia. In our 3rd case, patient had respiratory infection due to the underlying condition and increased secretions.

SC, including Apert's syndrome, can have associated atrial septal defect, ventricular septal defect or patent ductus arteriosus.[4] Our 4th case of craniosynostosis was associated with pulmonary stenosis for which she underwent percutaneous balloon pulmonary valvuloplasty 11 years back.

Laboratory evaluation usually includes a complete blood count, coagulation studies, liver and renal function tests, blood sugar, electrolytes, chest x-ray, ECG, type and cross match for adequate volumes of packed red blood cells. A written and informed consent as well as high risk consent should be taken from parents after full explanation about the intraoperative complications, blood transfusion, possibility of post-operative stay in intensive care unit (ICU) and mechanical ventilation.

The anaesthesiologist must be ready for airway problems, intubation difficulties, blood loss and massive blood transfusion, perioperative haemodynamic changes, disseminated intravascular coagulation, venous air embolism, positional injury, hypothermia and even the problems which may arise from different systemic involvements in SC. Monitoring should include pulse oximeter, non-invasive blood pressure, electrocardiograph, end tidal CO_2, airway gases and pressures as per American Society of Anesthesiologists guidelines as well as advanced monitoring like arterial blood pressure, central venous pressure (CVP), temperature, urine output etc. Point-of-care testing for haematocrit, electrolytes and acid-base balance is highly desirable. Adequate venous access preferably with two large bore cannula is mandatory. For paediatric age group 24 to 20-gauge cannulae were appropriate in our patients. Central access and CVP monitoring is not routinely practiced by us but is used if adequate peripheral access cannot be obtained and in case of huge blood loss or to manage venous air embolism (VAE). VAE as a complication is reported with incidence ranging from 2.6% to 82.6% as detected by precordial Doppler monitoring or echocardiography.[5]

We used intravenous atropine or glycopyrrolate and fentanyl as premedication. Induction usually carried out by either intravenous induction agent like thiopentone, etomidate or inhalation agent sevoflurane. We use succinylcholine or rocuronium for laryngoscopy and intubation and for maintenance we preferred atracurium or vecuronium and sevoflurane. In presence of increased intracranial pressure, high dose sedative premedication, ketamine, and succinylcholine was avoided.

We used 0.9% saline and lactated ringer's solution for maintenance, to replace the volume loss and to

replace a portion of the blood loss. Colloids and inotropes were used as per requirements. An accurate determination of ongoing blood loss during surgery and a precise restoration of that represent one of the major concerns. Blood saving begins with evaluation of the patient's bleeding risk, type, technique and length of surgery, number of cranial sutures involved and patients age, weight and physical status. Blood should be in the operating room before surgery begins as estimated blood volume (EBV) is limited to 80 ml/kg. Common sources of blood loss are the subgaleal tissues, fresh ends of the cut bones and dural venous sinuses. After osteotomy, blood loss is usually slow and continuous. It may be impossible to accurately measure blood loss, as much of the loss is absorbed by and/or under the drapes, surgical gowns, on sponges and in suction tubing and reservoirs mixed with saline irrigation. It is best advised to start transfusion at the start of skin incision and to keep up with the ongoing losses. We also transfused fresh frozen plasma and platelets for correction of microvascular bleeding in the setting of massive transfusion when coagulation tests could not be obtained in a timely manner.

During craniosynostosis surgery a large surface area is exposed to the atmosphere, which may result in excessive heat loss. We monitored core as well as peripheral temperature intraoperatively and normothermia was maintained by covering the body with cotton roll, using warm intravenous fluids and convective forced-air warming devices.

Post-operative care should be provided in postoperative anaesthesia care unit or ICU because of lengthy surgery, unstable haemodynamics, elective ventilation and good nursing care. Out of the 12 cases described, 10 patients were discharged after successful recovery, but one patient (case 6, Table1) had arrhythmia and one patient (case 10, Table 1) developed seizures in the post-operative period. Appropriate antibiotics, anticonvulsant, good analgesics and maintenance of haematocrit level are the mainstays of postoperative care.

Conclusion

Management of craniosynostosis is challenging for an anaesthesiologist in view of paediatric patients with difficult airway, prolonged duration of surgery, high likelihood of significant blood loss, massive blood transfusions and post-operative complications. With proper planning, intraoperative vigilance, and postoperative care, patient outcome is satisfactory.

References

1. Derderian C, Seaward J. Syndromic Craniosynostosis. Seminars in Plastic Surgery. 2012;**26**(2):64-75. https://doi.org/10.1055/s-0032-1320064 PMid:23633933 PMCid:PMC3424695
2. Faberowski LW, Black S, Mickle JP. Craniosynostosis: an overview. Am J Anesthesiol 2000;**27**(2):76–82.
3. Morris GP, Cooper MG. Difficult tracheal intubation following midface distraction surgery. PediatrAnaesth 2000; **10**: 99-102. https://doi.org/10.1046/j.1460-9592.2000.00466.x
4. Breugem CC, Fitzpatrick DF, Verchere C. Monozygotic twins with Apert syndrome. Cleft palate craniofac J 2008;**45**:101-104. https://doi.org/10.1597/06-149.1 PMid:18215098
5. Faberowksi LW, Black S, Mickle JP. Incidence of venous air embolism during craniectomy for craniosynostosis repair. Anesthesiology 2000;**92**:20-3. https://doi.org/10.1097/00000542-200001000-00009