An Infant with Apert Syndrome and Tetralogy of Fallot for Craniosynostosis Correction: Anesthetic Challenges

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Abstract

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► Apert’s syndrome
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► paradoxical air embolism
► tet spell

Apert syndrome (AS), popularly known as acrocephalosyndactyla, a rare autosomal dominant disorder characterized by craniosynostosis, craniofacial anomalies, and severe symmetrical syndactyly of hands and feet. Ten percent of children with AS can have associated congenital cardiac anomalies. Association of complex cyanotic heart disease with craniosynostosis is very rare. So far, only one case has been reported in the literature. The craniosynostosis corrective surgery is associated with the risk of massive bleeding or venous air embolism, which can cause paradoxical air embolism and precipitate cyanotic spell, which makes the anesthesia more challenging. In this report, we present the anesthetic challenges of an 8-month-old infant with AS and tetralogy of Fallot for craniosynostosis correction.

Case Report

An 8-month-old infant (weight: 8 kg) presented with the typical features of AS, such as flattening of bilateral frontal prominences with fused cranial bones, ocular hypertelorism, down-slanting palpebral fissures, and proptotic eyes, depressed nasal bridge, wide nose with a bulbous tip, fused middle and third fingers on the left hand, and fused second and third toes in both feet (►Fig. 1A-C). The child had central cyanosis, with oxygen saturation of 82%. Airway examination revealed short webbed neck with restriction of neck movements. Cardiac examination revealed features of TOF. Echocardiography showed a subaortic ventricular septal defect (5-mm defect) with bidirectional shunting, 20% aortic overriding with severe infundibular stenosis with a peak gradient of 107 mm Hg. Computerized tomography of skull revealed fused coronal suture with open sagittal, metopic, and lambdoid sutures. She was posted for bifrontal craniotomy with right frontal orbitotomy. The child was on propranolol 5 mg bid for preventing infundibular spasm.

After consultation with a multidisciplinary team, it was decided to proceed with craniosynostosis correction before the surgical correction for TOF. All standard monitors were connected and induction was carried out using 4% sevoflurane with 100% oxygen. Once the child was asleep, an intravenous access was established and the anesthetic plane was deepened with fentanyl, ketamine, and a small dose of propofol. Phenylephrine (5 µg) bolus was given along with induction drugs to maintain the systemic vascular resistance (SVR). After confirming the adequate bag and mask ventilation, the child was paralyzed with atracurium, was intubated using a video laryngoscope (C-MAC size 1, video laryngoscope, KARL STORZ, Germany) with 4.5-uncuffed endotracheal tube (ETT) which was reinforced with 5.5-mm ETT to prevent kinking (from the angle of mouth to connector level) as described by...
Singh et al. A 22-G peripheral venous access, a 24-G right radial arterial line, and a 5.5-F right subclavian triple lumen central venous catheter were inserted and strict measures were taken to avoid air entainment. Tranexamic acid (20 mg/kg) bolus followed by an infusion at 1 mg/kg/h was given to reduce the bleeding. Bilateral scalp block was given using 8 mL of 0.2% ropivacaine before the skin incision. Fentanyl (up to 8 µg/kg) and morphine (0.75 mg) and 20 mg/kg of paracetamol were given for analgesia. Intraoperatively, measures were taken to avoid "Tet Spell" by maintaining the SVR using low dose of noradrenaline (0.02–0.05 µg/kg/min) and by avoiding factors that can increase the pulmonary vascular resistance (PVR), such as hypercarbia, hypoxia, acidosis, hypothermia, and high positive end expiratory pressure. Intraoperatively, the saturation was maintained between 85% and 93% with an inspired oxygen concentration of 60%. The procedure lasted for 4 hours with 170 mL of blood loss (30% of blood volume) which was replaced with 100 mL packed red cell concentrates and 50 mL of fresh frozen plasma and 25 mL of cryoprecipitate. Since we were expecting another 50 to 60 mL of blood loss in the postoperative wound drain and we did not want to dilute the blood with crystalloids, and to avoid postoperative hypotension and coagulopathy, we had given the blood products. The child was hemodynamically stable, warm, and awake at the end of surgery. The trachea was exubated and shifted to high dependency unit with fentanyl (0.4–0.5 µg/kg/h) and ketamine (0.02–0.03 mg/kg/h) infusion for postoperative analgesia. Child had good recovery and got discharged on the 7th postoperative day.

Discussion

AS accounts for 4.5% of all cases of craniosynostosis and often associated with cardiac and urological anomalies with a prevalence of 1 in 65,000 to 75,000 live births.1 There are several anesthetic concerns while anaesthetizing these children. Firstly, they can have upper airway obstruction due to the reduction in the nasopharyngeal size and choanal patency, can have lower airway compromise due to the associated anomalies of the tracheal cartilage.6 In some cases, they can even present with severe obstructive sleep apnea and the cor pulmonale. In our case, though the history and the preoperative examination did not reveal airway obstruction, the child required an oral airway to maintain the airway patency soon after induction and an hour after extubation. Secondly, they can have congenital cervical spinal fusion, especially at C5 and C6 (seen in 68% of cases), can lead to difficult intubation. This infant also had a short and webbed neck with restriction in neck movements which made us select C-MAC video laryngoscope for intubation (Fig. 1A, B). Thirdly, premature closure of cranial suture can result in elevated intracranial pressure. So, it is imperative to prevent further increases in the intracranial pressure by assuring a smooth induction, intubation and emergence. Lastly, AS can have associated cardiac anomalies as seen in this case that had TOF. The mortality of children with congenital heart disease undergoing major surgery is 16%.2 Since this child had TOF along with AS, measures were taken to maintain adequate preload to stent open the stiff, noncompliant RVOT; the afterload was maintained with noradrenaline infusion to minimize the right to left shunt, thereby promoting pulmonary blood flow; and measures were taken to avoid increase in the PVR which will decrease the pulmonary blood flow, thus worsening the cyanosis.

Specific risks related to craniosynostosis corrective surgery include major blood loss and venous air embolism (VAE).6,7 In our patient, tailoring the surgical procedure to involve minimal dissection of the bone over the venous sinuses aided in reducing blood loss. The incidence of VAE has been reported as high as 83%, mostly occurring without hemodynamic compromise and only about 1 to 2% being clinically significant.7 The occurrence of VAE in presence of TOF can cause paradoxical air embolism, which can increase morbidity or mortality significantly.

Conclusion

A syndromic infant coming for craniosynostosis surgery with an underlying congenital cyanotic heart disease and a difficult airway poses a lot of challenges to the neuroanesthesiologist. Maintaining both the cardiovascular and cerebrovascular homeostasis is of paramount importance for a successful outcome. In our case, smooth induction and extubation, adequate preparedness to face the massive blood loss, appropriate blood, and fluid transfusion to prevent hypovolemia, VAE, administration of low dose of vasopressor to maintain the SVR, and adequate postoperative pain relief all helped in the rapid recovery.

Note

This case report abstract is submitted as conference paper and was presented in the ISNACC-2019 (Feb 15–17th) at Gurugram, Haryana, India.

Conflict of Interest

None declared.

Authors’ Contributions

All authors made material contributions to the handling of this case and to the intellectual content of this article.

Note

This manuscript has not been submitted elsewhere for publication. Only abstract was submitted in ISNACC 2019 conference.
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