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Anesthetic Management of a Child with Frontonasal Encephalocele and Hemifacial Microstomia Syndrome for Repair of Encephalocele and Microstomia Correction

Selvendiran Panneerselvam[1] Georgene Singh[1] Ananth P. Abraham[2] Keta Thakkar[1]

[1] Department of Neuroanaesthesia, Christian Medical College, Vellore, Tamil Nadu, India
[2] Department of Neurological Sciences, Christian Medical College, Vellore, Tamil Nadu, India

Address for correspondence Keta Thakkar, MD, DNB, PDF, DM, Department of Neuroanaesthesia, Christian Medical College, Vellore, Tamil Nadu, 632004, India (e-mail: keta0819@gmail.com).

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Abstract

Anesthetic management of a child with hemifacial microstomia and frontonasal encephalocele can be challenging due to factors such as a difficult airway, associated systemic conditions, young age, and prolonged operating time. Hereby, we outline the intraoperative management of a 3-year-old child undergoing repair of multiple defects requiring multidisciplinary involvement of neurosurgery, plastic surgery, and anesthesia team. In our case, a size 0 mask and hyper-angulated D-blade video laryngoscopy were used to prevent sac compression and facilitate intubation. Blood conservation strategies included tranexamic acid, precise fluid management, and maintenance of normothermia. Extubation was carefully planned, considering blood loss and airway edema, and performed when the child was fully awake, ensuring a stable postoperative period and successful recovery. This report emphasizes the importance of meticulous preoperative assessment, readiness for a challenging airway, efficient management of intraoperative blood loss, and the value of teamwork.

Keywords
- encephalocele
- microstomia
- perioperative

Introduction

Frontonasal encephaloceles are relatively uncommon congenital malformations that manifest as a clinically visible facial mass along the nose, the location and size of which vary depending on the variety.[1] We present a case of anesthetic management of a child with frontonasal encephalocele and facial hemiatrophy planned for bifrontal craniotomy with repair of the encephalocele and correction of microstomia. This case report provides novel insights into managing rare craniofacial anomalies, highlighting advanced techniques and interdisciplinary teamwork.

Case Report

A 3-year-old girl child, weighing 9.7 kg, presented with complaints of swelling on the forehead and seizure disorder along with hypertelorism, elongation of face, receding chin, and deviation of the mouth to the right side (►**Fig. 1**). The child had global developmental delay and bilateral profound hearing loss. A computed tomogram revealed a bony defect above the nasion through which there was protrusion of the meninges with multiple facial anomalies (►**Fig. 2A**), which was confirmed by magnetic resonance imaging (►**Fig. 2B**). The child was planned for excision and repair of the

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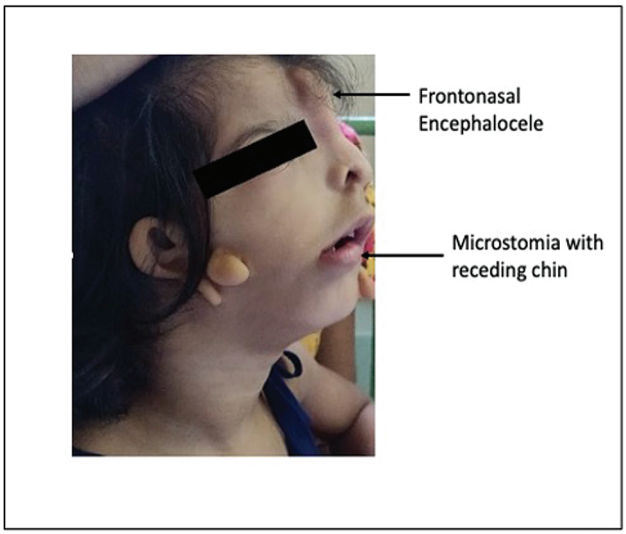

Fig. 1 Child having the frontonasal encephalocele with hypertelorism, elongation of face, receding chin, and a deviation of the mouth to the right side.

encephalocele, followed by microstomia correction. On preanesthetic evaluation, a difficult airway was anticipated due to the presence of a lobulated encephalocele sac on the forehead and right side of the root of the nose, severe retrognathia, and absent right mandibular ramus and condyle.

After confirming adequate fasting status and obtaining informed consent from the parents, the child was shifted to the operation theater. Standard American Society of Anesthesiologists' recommended monitoring practices were initiated in the operating room. A difficult airway cart with appropriate-sized airways, stylets, and laryngeal mask airway was kept ready. As the child did not have intravenous (IV) access in situ, steal induction with 8% sevoflurane in 6 L/min of 100% oxygen, avoiding nitrous oxide, was done, and a 24 G peripheral IV access was secured. IV propofol (20 mg) and fentanyl (20 mcg) were administered to facilitate mask ventilation with a size 0 mask and a size 1 (70 mm) airway, carefully avoiding compression on the encephalocele. After confirmation of the adequacy of mask ventilation, an additional dose of propofol was administered. A quick and gentle check-laryngoscopy done with hyperangulated D-blade video laryngoscopy revealed a Cormack–Lehane grade of 1. The child was then paralyzed with atracurium (5 mg). The trachea was intubated with a 4 mm cuffed South Pole RAE (Ring–Adair–Elwyn) tube, which was railroaded on an appropriately shaped stylet, and a throat pack was inserted. Additional IV access was secured post-induction, and the radial artery was cannulated. A fluid warmer for administering IV fluids, prewarming with a forced air-warming blanket, and lukewarm fluids for cleaning and irrigation were utilized to maintain the core temperature. Mechanical ventilation was initiated, and the lungs were ventilated with oxygen and air with FiO_2 of 0.5, sevoflurane, and additional IV doses of atracurium. Tranexamic acid 15 mg/kg over 20 minutes was used as a loading dose before skin incision, followed by 2 mg/kg/h as a maintenance dose. Multimodal pain management was provided with incision-site local anesthesia infiltration with continuous fentanyl infusion of 1 mcg/kg/h and IV paracetamol 100 mg. After the repair by the neurosurgeons, the plastic surgeons performed microstomia correction surgery. The maximum allowable blood loss was 260 mL, whereas the estimated total blood loss was 200 mL over 8 hours, which was replaced with 150 mL of irradiated packed red cells.

Additionally, goal-directed fluid therapy using pulse pressure variation (PPV) guidance was administered with 600 mL

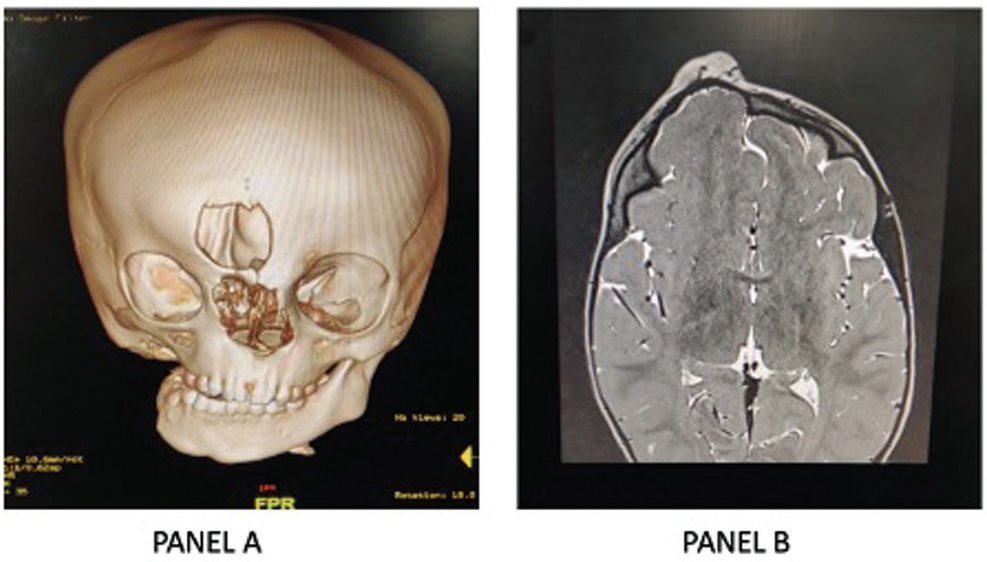

Fig. 2 PANEL A: CT (reconstruction) revealed a defect in the anterior right frontal lobe through the defect with multiple facial anomalies like dysplasia of the right hemimandible, angle, condyles, and coronoid process with a resultant deviation of the chin to the right side with retrognathia. PANEL B: MRI (T2) shows a defect in the anterior right frontal lobe with meningeal protrusion with dysplasia of the right frontal lobe. CT, computed tomography; MRI, magnetic resonance imaging.

crystalloids and 100 mL colloids. The total intraoperative urine output was 250 mL. Arterial blood gas analysis at the end of the surgery was done, and a packed cell volume (PCV) of 31.7, lactate of 1.3, and serum calcium of 1.20 were found. After completion of the procedure, adequate oral suctioning was done, the throat pack was removed, and a cuff leak test was performed, which was positive. Awake extubation of the trachea was done following the reversal of neuromuscular blockade with neostigmine and glycopyrrolate. The child was then shifted to the intensive care unit for observation and monitored in the postoperative period, which was uneventful. The consent for writing the case report was obtained from the child's guardian.

Discussion

It is crucial to adequately prepare for airway management when dealing with hemifacial microstomia patients with anterior encephalocele.[1,2] Managing the airway in children with frontonasal encephalocele is expected to be challenging due to the presence of a protruding sac, which can make mask ventilation difficult.[1] Additionally, determining the element of a blocked nose is essential, which can be assessed from history and whether the child breathes with an open mouth. In addition, parental presence during the induction of anesthesia can be helpful against the backdrop of a difficult airway, the inability to premedicate, and the need to avoid crying or straining during induction.[1] We used a size 0 mask with an appropriate size airway to avoid compression on the sac with the mask, which can rupture.[1] Inhalation induction performed with the readiness of a difficult airway cart enabled us to assess the airway while the child was spontaneously breathing. The encephalocele repair procedure was to be followed by microstomia correction, which usually requires nasal intubation, which was impossible in this case. Hence, the trachea was intubated with a South Pole RAE tube. The first-pass success rate of video laryngoscopy is 100% in children with hemifacial microstomia. Therefore, we decided to use the hyper-angulated D-blade, facilitating an adequate glottic visualization.[3,4]

Blood loss is another concern while managing a pediatric case with craniofacial deformity.[3] Our blood conservation and fluid management strategies included tranexamic acid, preoperative calculation of maximum allowable blood loss, continuous hemodynamic monitoring, following trends of dynamic parameters of fluid responsiveness like PPV, reviewing hemoglobin values, and ensuring adequate urine output. More recently, using the RD Rainbow sensors in the pediatric age group in our institute has enabled us to monitor the vital signs, hemoglobin, and oxygenation status, ensuring a stable intraoperative period. Extubation readiness should also be evaluated while managing such a case.[3] In our case, long-duration procedures, blood loss, difficult airway, possible airway edema, and temperature were all considered and optimized before extubation.

A thorough preoperative evaluation, meticulous planning during the perioperative period, and multi-disciplinary teamwork achieved a successful outcome.

Conflict of interest
None declared.

References

1 Mahajan C, Rath GP. Anaesthetic management in a child with frontonasal encephalocele. J Anaesthesiol Clin Pharmacol 2010; 26(04):570–571
2 Gallagher DM, Hyler RL, Epker BN. Hemifacial microsomia: an anesthetic airway problem. Oral Surg Oral Med Oral Pathol 1980; 49(01):2–4
3 Meier N. Anesthetic considerations for pediatric craniofacial surgery. Anesthesiol Clin 2021;39(01):53–70
4 Thakkar KD, Hrishi AP, Sethuraman M, Vimala S. Management of a difficult airway scenario in a case of Hurler's syndrome with a D-blade video laryngoscope. J Neuroanaesth Crit Care 2021;08(03): 214–216

