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# ENCEPHALOCELE REPAIR IN INFANT WITH CONGENITAL HEART DISEASE – AN ANAESTHETIC CHALLENGE

**Anaesthesiology**

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## ABSTRACT

Encephalocele is a neural tube defect leading to herniation of cerebrospinal fluid, brain tissue and meninges through a defect in skull bone. These defects can be repaired surgically during infancy. The anaesthetic management of occipital encephalocele is challenging mainly due to difficulty in securing a definitive airway, preventing the rise of intracranial pressure and maintaining stable hemodynamics with prone position of the patient. Encephalocele has multiple syndromic associations like Von Voss Cherstvoy syndrome, Knobloch syndrome, Meckel Gruber syndrome, Jouberg syndrome, Walker Warburg syndrome, Klippel Feil syndrome and many more. These syndromes further increase the risk of perioperative complications.

This is a case report describes the anaesthetic challenge in a case of occipital encephalocele repair in a 4 day old infant with congenital heart disease.

## KEYWORDS

Occipital encephalocele, congenital heart disease, atrial septal defect, ventricular septal defect, general anaesthesia, prone position.

## INTRODUCTION:

Encephelocele is a neural tube defect characterized by sac – like protrusions of the brain and the membranes that cover it through openings in the skull. It accounts for 10 – 20% of all craniospinal dysraphisms[5]. The incidence of encephalocele is 1 in 10,500 births[6]. A repair surgery is generally performed during infancy. It is most commonly associated with hydrocephalus, which would require surgical shunting. The infant may also have other congenital abnormalities like cyanotic or acyanotic heart diseases, spine deformities, limb deformities, craniofacial malformations, etc. The post-operative neurological outcome depends on the size of defect and associated congenital abnormalities. Special attention needs to be given to positioning of patient during intubation as well as the surgery to avoid rise in intracrainial pressure without hampering the surgical site vision.

This case report describes regarding the anaesthetic precautions and practices followed while managing a case of occipital encephalocele repair.

## CASE REPORT:

A 25 year old female, gravida one parity one, by date 37.2 weeks pregnant came to casualty of MGM hospital in latent labour and the patient was shifted to labour room for further management. A female child weighing 2.15 kilograms was delivered. Delivery was uneventful.

The neonate was born with a swelling in the occipital region of the head, which was approximately 10cm X 8cm.

A neurosurgical review was done for the patient and a surgical excision and repair was planned. On examination the child was drowsy, responding to painful stimulus with perioribital swelling and hypertelorism was present, with restricted passive neck movements due to occipital swelling. Local examination of the swelling suggested a 6cm X 8cm pedunculated oval shaped swelling in the occipital region with no visible bleeding or local rise of temperature. CT scan Brain plain along with the swelling reported a 2cm X 2.1 cm cranial defect is noted along the midline involving bilateral parietal bones at the level of posterior fontanelle. This is causing herniation of bilateral superior and inferior parietal lobules and parts of occipital lobe including posterior horn of lateral ventricle. Herniated brain shows areas of T2 FLAIR s/o gliosis. some areas of herniated brain shows restricted diffusion s/o ? infarct. No evidence of hydrocephalus. MRI Brain was suggestive of a 2 x 2.1 cm sized cranial defect along the midline involving bilateral parietal bones at the level of posterior fontanels; gliosis and compressive ischemic changes were also noted in the herniated brain. 2D Echogram suggested a large 5.3 mm ostium secondum (Atrial septal defect with left to right shunt) with small muscular 2.2 mm ventricular septal defect with left to right shunt. Cardiologist review was done regarding the same advised to continue with surgery with moderate risk of perioperative cardiac events.

The patient was kept nil per oral 4 hours prior to surgery and was started on maintenance fluid at the rate of 12ml/hr and the parents were explained the high risk of intraoperative cardiac events as well as need of ventilatory support post operatively. Preoperatively, the patency of 24 gauge intravenous cannula in left dorsum of hand was confirmed and care was taken to keep the burrette iv drip set clear of any air bubbles. Premedication was done with Inj. Glycopyrrolate 0.004 mg/kg, Inj. Midazolam 0.05 mg/kg and Inj. Fentanyl 2 mcg/kg.

### *Intraoperative Management:*

Then patient was brought in the operation theatre, started on 4litre/min oxygen via nasal prongs and all standard monitors were attached.

Intubation was planned in supine position. The large encephalocele swelling would make the intubation difficult. Hence, the child was positioned supine on was protected using a doughnut shaped head support. But as the sac was too big, it did not rest in the doughnut support. Hence, the baby was lifted and her head was placed beyond the edge of the table with an assistant supporting the encephalocele and taking adequate care so as to prevent rupture while another assistant stabilized the baby's body.

After pre oxygenation, patient was induced with Inj. Propofol 2 mg/kg. Sevoflurane was used as the inhalational agent.

Bag mask Ventilation was done in left lateral position of head as shown in Fig 1. An oropharyngeal airway of size 000 was inserted to maintain adequate chest rise during ventilation.

After confirming adequate mask ventilation, a non depolarizing muscle relaxant Inj. Atracurium 1mg/kg was given. Three minutes later, neonate was intubated in supine position after taking the patient at the edge of the bed, using millers blade 0 with uncuffed endotracheal tube number 3. Air entry was confirmed with adequate chest rise and capnography. The endotracheal tube was fixed at 10 cm mark and the airway was kept in situ (Fig 2). Patient was haemodynamically stable throughout induction. Anaesthesia was maintained with O2, air and sevoflurane.

Urinary catheter was inserted. Prone position was given to the neonate with adequate padding of pressure areas. Head was supported on a horseshoe padding. The ECG leads were switched to the back. A precordial stethoscope was secured in place. (Fig 5)

Multimodal analgesia was maintained by a per-rectal paracetamol suppository of 40 miligrams and two micrograms of Inj. Fentanyl was given every hour.

An approximate 80 millilitres of blood loss occurred intraoperatively, patient was started on PRBC via infusion pump at the rate of 10 ml/hr. A total of 33 ml PRBC was given intraoperatively.

A loading dose Inj. Fosphenytoin 30 mg/kg body weight that is 66 miligrams was given intraoperatively.

A deep plane of anaesthesia was maintained using Inj. Atracurium, oxygen, air and sevoflurane at a minimum alveolar concentration of 2% w/v. The neonate was hand ventilated using a Jackson rees circuit with a 0.5 L bag.

Total intraoperative fluid given was 70 ml of ringer lactate and urine output was approximately 10 ml. The duration of the surgery was approximately 2 hours.

Post operatively, patient was switched to supine position. A reversal of Inj. Neostigmine 0.1mg and Inj. Glycopyrrolate 0.016mg was given after spontaneous breathing was established. The neonate was extubated after noticing regular breathing efforts and adequate regain of power in all four limbs.

Post extubation, she was monitored in the postoperative area for any signs of desaturation and O2 was given at 4L via neonatal Hudson's mask.

***Intraoperative Finding:***

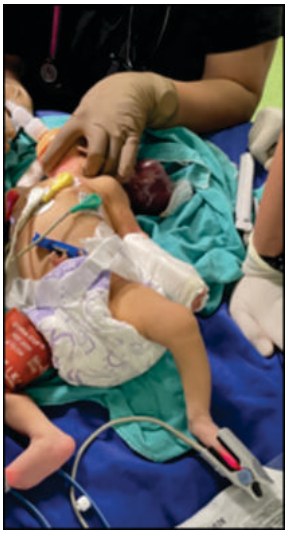

***Fig1:*** *Mask Ventilation In Lateral Position Of Head. Occipital Encephalocele Swelling Seen*

***Fig2 :*** *Inubated photo of neonate with airway 00 in situ. Hypertelorism seen*

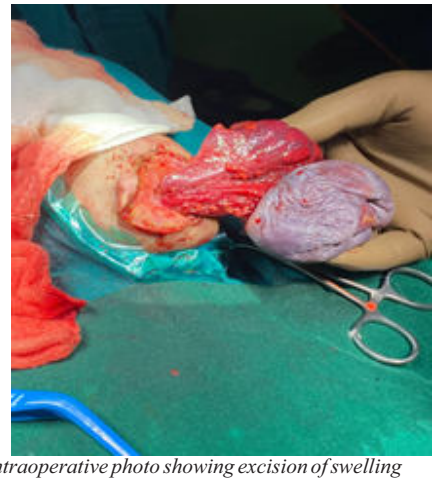

***Fig 3 :*** *Intraoperative photo showing excision of swelling*

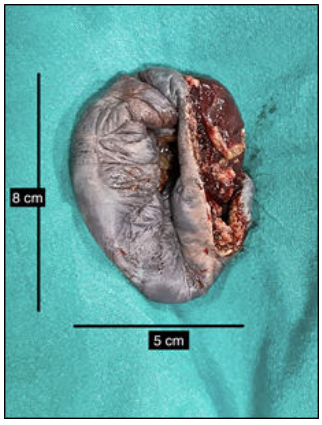


***Fig 4 :*** *Post Excision Specimen*

***Post-operative NICU care***

After half an hour of monitoring in the post operative area the patient was shifted to the neonatal intensive care unit for further monitoring and management, where the patient was maintained on oxygen by hood and weaned slowly from oxygen support. Breast-feeding was restarted the next day until then maintenance fluid was given 8 ml/hr. The baby was transferred to step down ward after 5 days of NICU stay and was discharged from hospital on post-operative day 20.

**DISCUSSION:**

Encephalocele is the protrusion of neural element from a defect in the cranium. As per literature, 75% of encephalocele are occipital[3]. These children are more likely to have other associated conditions like club-foot, hydrocephalus, exostrophy of bladder, Klieppel – fiel syndrome or congenital cardiac defects[3]. Infants with encephalocele have shown increased incidence of latex allergy leading to intraoperative cardiac arrest or bronchospasm. Hence, latex free gloves were used for all procedures.

Major challenge in this case was airway management due to large size of encephalocele. Pediatric patients physiologically have small amount of functional residual capacity, hence reducing their apnea time. Due to this reason, an error during intubation can cause more harm in a neonate as compared to adults.

Induction of anesthesia should be done keeping in mind the neonatal cerebral physiology and all effort should be towards maintaining adequate cerebral blood flow and intracrainial pressure (ICP) which may require hyperventilation. The goal of induction is to avoid increase in ICP owing to associated with hypoxia, hypercapnia and volatile anaesthetic-induced increase in cerebral blood flow[2].

We used Inj. Propofol and sevoflurane as the inducing agent, owing to its smooth induction with rapid recovery. The intubation was done in supine position with encephalocele swelling at the edge of the OT table and an assistant supporting the swelling. It is of utmost importance to support the swelling as stretching or rupture of the encephalocele membrane might cause a rise in the intracranial pressure and might

lead to further herniation of brain tissue. However, there are other methods of intubation in case of a large occipital encephalocele like: 1) in lateral position of the head. 2) in supine position while placing the swelling over a doughnut shaped cushion 3) needle decompression of encephalocele sac under sterile conditions. However, the resultant rapid decompression of ventricular system may lead to fatal complications such as cardiac arrest due to traction on cerebral neuronal pathways[4].

Prone position should be given while ensuring adequate bolsters under chest and pelvis while the abdomen remains free. As, any rise in intraabdominal pressure would impede ventilation, compress the vena cava and increase epidural venous pressure and bleeding[2]. The horseshoe head rest may cause congestion of face and tongue and sore on malar prominences[2]. Extreme flexion of neck may cause accidental intrabronchial intubation or kinking of the tube inside the mouth.

Congenital heart diseases, which may otherwise remain asymptomatic, may become symptomatic under the influence of anaesthetic agents or mechanical ventilation or excess blood loss, which occurs in a neurosurgical procedure. The aim is to maintain an average mean blood pressure of 40-50 mm Hg and just adequate positive pressure ventilation, so as to prevent reversal of shunt from acyanotic to cyanotic.

Inspite of all the difficulties, the anaesthetic as well as surgical management was carried out successfully.

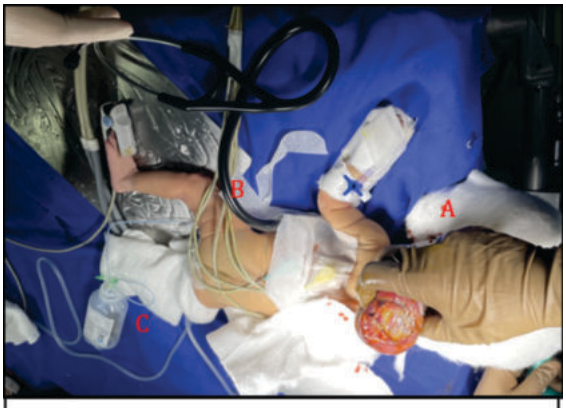


Fig 5: Neonate in prone position
A – Horseshoe head rest
B – Precordial stethoscope
C – Urinary catheter attached to 100 ml empty bottle acting like a urinary bag

## CONCLUSION:

Encephalocele repair poses challenges for both the anaesthetist as well as the neurosurgeon. Paediatric neuroanaesthesia poses an added risk due to immature nervous system development. In this case, apart from the large encephalocele, there was additional risk of perioperative cardiac events mainly due to reversal of shunt due to positive pressure ventilation. However, all these perioperative complications can be managed successfully with meticulous preoperative planning and intraoperative vigilance.

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