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## ANAESTHETIC CHALLENGES IN THE MANAGEMENT OF PEDIATRIC ENCEPHALOCOELE REPAIR: RETROSPECTIVE CASE SERIES

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**ABSTRACT: INTRODUCTION:** Encephalocele is the protrusion of the cranial contents beyond the normal confines of the skull through a defect in the calvarium and is far less common than spinal dysraphism.<sup>1</sup> Anaesthetic challenges in management of occipital meningoencephalocele include securing the airway with intubation in lateral position, intraoperative prone position and its associated complications, careful securing of the endotracheal tube and accurate assessment of blood loss. These babies also have associated congenital anomalies, gastrointestinal malrotation, renal anomalies, cardiac malformations and tracheoesophageal fistula, making anaesthetic management even more difficult. Meticulous anaesthetic management is crucial for early repair of encephalocele to prevent any sequel.<sup>2</sup> **METHODS:** To identify the anaesthetic challenges, perioperative and postoperative complications during encephalocele repair, 20 cases were studied retrospectively from 2012 to 2014 at Department of Anaesthesia, Department of Neurosurgery, MR Medical College, Gulbarga. **RESULTS:** 20 cases of encephalocele repair were undertaken during the study period. Out of these 12 (60%) were male and 8(40%) female. Age range was 1 day to 6 years. Most common type of encephalocele was occipital 12(60%), which posed a difficulty during positioning & intubation, followed by occipito- cervical 4(20%), Parietal 2(10%), Fronto- nasal 1(5%) & Fronto- naso- ethmoidal 1(5%). Most of the patients were extubated successfully on table, only one patient required post-operative ventilator support for a day. Peri-operative complications included bronchospasm (15%), followed by hypotension, tachycardia, laryngospasm, hypoxia, accidental extubation (10% each) & bradycardia, endobronchial intubation (5%). **CONCLUSION:** Children with Encephalocele are prone to have peri-operative complications which can be managed by meticulous anaesthetic management.<sup>3</sup> Early surgical management of encephalocele is not only for cosmetic reasons but also to prevent tethering, rupture and future neurological deficits.

**KEYWORDS:** Occipital encephalocele, bronchospasm, anaesthesia.

**INTRODUCTION:** Encephalocele is a congenital anomaly where herniation of the cranial contents occurs through a congenital defect in cranium. It account for 10-20% of all craniospinal dysraphisms.<sup>1</sup> Occipital encephalocele generally occur through a bony defect in the occipital bone. It may extend into the foramen magnum and can involve the posterior arch of Atlas. Occipital encephalocele represent approximately 85% of lesions. The underlying basis for the extrusion of brain tissue protruding from the meninges and CSF in encephalocele is a primary abnormal mesodermal defect. Giant occipital encephalocele associated with microcephaly and micrognathia is extremely rare. In these patients, there can be associated lesions including agenesis of corpus callosum, hypoplasia of cerebellum, meningocele, and hydrocephalus.

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Most infants present with hydrocephalus are usually associated with Arnold- Chiari type II malformation (downward displacement of the cerebellum into the brain stem and cervical canal with medullary kinking). This may cause cervical cord compression, during extension for intubation leading to brain stem compression.

Airway management may be difficult in patients with significant hydrocephalus. Most patients show a diminished response to hypoxia, and may be more susceptible to post-operative apnoeic episodes. The effect of positioning in neonates for induction of general anaesthesia must also be considered. No direct pressure should be applied to the exposed neural placode.<sup>4,5</sup>

**METHODS:** All the available medical records of children who underwent excision and repair of encephalocele over a period of two years (2012 to 2014) were analysed. For each child, data were collected by detailed review of records related to pre-anaesthetic evaluation, intraoperative course and postoperative complications.

Pre-operative evaluation included age, sex, weight, height, Hb%, electrolytes, chest x-ray, site of lesion, CSF leak from the sac, neurological presentation, associated systemic abnormalities, venous and airway assessment. All the patient had their MRI and CT Scan of relevant part to see the extent of disease.

Intraoperative data included anaesthetic technique, intraoperative monitoring (heart rate, saturation, ventilation, BP, Urine output, temperature), fluids infused, blood loss and transfusion. Intraoperative parameter such as heart rate and blood pressure were recorded. Below and above 20% alteration from baseline values of heart rate and BP were regarded as abnormal. Respiratory complications (hypoxemia, hypercarbia, bronchospasm, laryngospasm), endobronchial intubation, accidental extubations were noted. Miscellaneous complications like hypothermia, dislodgement of intravenous catheter were reviewed.

Post-operative complications like nausea, vomiting, mechanical ventilation, surgical complications were reviewed.

**ANAESTHETIC TECHNIQUE:** A standard anaesthetic technique was followed for all children. Intravenous line was secured with or without sedation. Those who needed sedation were given midazolam 0.05mg/kg body weight. Intravenous induction was done with thiopental sodium (3-5mg/kg). Vecuronium (0.08mg/kg) was used to facilitate tracheal intubation.<sup>6</sup> The intubation was performed mostly in supine position (16 cases) putting placode within the padding made in the shape of a doughnut. In patients associated with large hydrocephalus pillow behind shoulders were kept during intubation. Intubations in lateral position (4 cases) were done in patients with large EMC. Repair of EMC was done in prone position whereas in patients with large hydrocephalus VP Shunt was inserted in supine position followed by EMC repair in prone position. Anaesthesia was maintained with oxygen and Isoflurane with controlled ventilation. Fentanyl was used for analgesia. Ringer Lactate was the fluid of choice in most patients, but isolyte -p was also used especially in neonates. At the end of surgery, residual neuromuscular block was reversed with calculated dose of Neostigmine and atropine and the trachea was extubated when the patient fully awake.

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**RESULTS:** Out of a total 20 children operated, The youngest were 1 day old infants while the oldest child presented at 6 years of age (median=1 month, mode=1 month). More male children (n=12) were operated than female child (n=8) with male: female ratio of 3:2.

Age	Male	Female	Total no	Percentage
0-60 days	3	2	5	25
>60 days-1 year	6	5	11	55
>1 year	3	1	4	20
<b>Total</b>	<b>12</b>	<b>8</b>	<b>20</b>	

Table 1: Demographic details

Site	No (%)
Occipital encephalocoeles	12 (60%)
occipito- cervical	4 (20%)
Parietal	2 (10%)
Fronto- nasal	1(5%)
Fronto- naso- ethmoidal	1(5%)

Table 2: Site of encephalocoele

Most common type of encephalocele was occipital and occipito cervical.

Associated anomalies	No
Double encephalocoele with split pons	1
Chiari 3 malformation	2
Meningomyelocoeles	4
Syrinx	4
Co-existing medical conditions	4
• Upper respiratory tract infections	
• Electrolyte imbalance	4

Table 3: Associated anomalies

One patient had a double encephalocoele (one atretic and other was occipital) with dermal sinus tract and limited dermal myelocschsis and a split pons.

Electrolyte imbalance was present in 4(20%) as hypokalaemia, hyperkalaemia, hyponatraemia. Two of the children were also anaemic and malnutrition was present concurrently.

Complications	No (%)
<b>Cardiovascular</b>	
• Bradycardia	1 (5%)
• Tachycardia	2 (10%)
• Hypotension	2 (10%)

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<b>Respiratory</b>	
• Hypoxia	2 (10%)
• Bronchospasm	3 (15%)
• Endobronchial intubation	1 (5%)
• Accidental extubation	1 (5%)
• Laryngospasm	2(10%)
<b>Total</b>	<b>14</b>
<b>Table 4: Intra operatively</b>	

The duration of anesthesia ranged from 2hrs-3hrs, an average of 2hrs, in few cases especially with hydrocephalus there were problems during intubation. Among the 14 intraoperative complications respiratory was most common as seen in 8 patients than cardiac complication (Table 3).

Among respiratory complications Bronchospasm, hypoxemia, laryngospasm, accidental extubation were more common. Endobronchial intubation was found in 1 case. Cardiac complications in the form of bradycardia, tachycardia and hypotension created problems during the operative procedure. Intraoperative blood loss was 20 ml- 130 ml, an average of 75 ml. Other uncommon complications were hypothermia 2(10%), facial edema 2(10%).<sup>8,9,10.</sup>

Post operatively 3 patients presented with rupture one of whom succumbed before surgery to shock. Of the other 2 patients, in whom repair of sac was performed, the patient with meningocele improved while the patient with encephalocele succumbed to meningitis and shock.

The other 17 encephaloceles treated surgically did well with no immediate surgical mortality. One child with Chiari III malformation succumbed 6 months postsurgery to unrelated causes. Shunt was performed in 4 cases due to the development of hydrocephalus.

Wound site infection CSF leak was seen in 4 cases which was conservatively managed with 1 patients requiring re-exploration and repair. The case with fronto nasal encephalocele developed ocular CSF leak which was conservatively managed.

<b>Complications</b>	<b>No</b>
Hydrocephalus	4
Wound infection	4
CSF leak	4
Re exploration & repair	1
<b>Table 5: post op complications</b>	

At the end of surgery 16 cases were extubated on the operating table. 4 cases had delayed recovery and one of them needed postoperative ventilation. The ICU stay was 2 to 7 days whereas the hospital stay was 4-24 days.

**DISCUSSION:** Encephalocele is hernial protusion of neural elements in sac. Meningocele is protrusion of only meninges. Early excision is recommended to prevent infection and rupture. Anaesthesiologist encounters many challenges during excision of meningocele or encephalocele in

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occipital region including difficult airway management, prone positioning, protection of neural placode, assessment of volume status and prevention of hypothermia.

Common congenital anomalies associated may be club foot, hydrocephalus with Chiari's malformation, extrophy of bladder, prolapsed uterus, Klippel feil syndrome and cardiac defects. Children with meningoencephalocele are likely to have varying degrees of sensory and motor deficits.<sup>11,12</sup>

Anaesthesia may induced with intravenous or inhalational method. Intubation can be done awake in lateral position or supine position by placing the neural placode in doughnut shaped support. Other method is placing the head at the edge of the table, supported by one person and elevating the body off the table while supporting the pelvis by other. Another method was described by Mowaffi is placing the baby supine on platform made by silicon supports kept one above other till the height matches with encephalocele sac and head was supported in hollow cushion protecting sac.<sup>13</sup>

The perioperative complications pose challenges to the anaesthesiologist and more common are respiratory complications followed by cardiovascular complications. Respiratory complications are hypoventilation, sleep apnoea, bronchospasm, laryngospasm, prolonged breath holding as a result of structural derangement of post medullary respiratory control centre or in its afferent and efferent pathways.

Cardiovascular complications included bradycardia, hypotension and tachycardia. Brainstem compression and coning causes most of the cardiac complications including cardiac arrest when Chiari malformation is associated with EMC.<sup>14</sup>

The timing of surgery, usually in the first 48 hours after birth, is important because an increased infection rate is associated with delayed surgery.

Paediatric endotracheal tubes should be uncuffed until at least six years of age. A correctly sized tube allows adequate ventilation with a small audible leak of air present when positive pressure is applied at 20cm H<sub>2</sub>. The main problems encountered were particularly difficult intubation owing to huge hydrocephalus, huge EMC in occipital region and dorsal, anatomical and physiological variations in the paediatric airway.

Hypothermia occurred commonly owing to the age group of the patient and the use of general anaesthetic agent as they depress the thermoregulatory response in children.

Heat is lost from the core to the cooler peripheral tissues, particularly in non-shivering thermogenetic neonates. Prolonged hypothermia can lead to a profound acidosis and impaired tissue perfusion, which have been associated with delayed awakening from anaesthesia, cardiac irritability, respiratory depression, increased pulmonary vascular resistance and altered drug responses.<sup>1</sup>

Preoperative evaluation especially of cardiac, gastrointestinal, genitourinary system is very important because embryologically they are formed concurrently with malformed neurologic system. Literature review suggests that 37% of MMC are associated with congenital heart lesions however present study did not show any of those. The short trachea is found in 36% chest radiograph of MMC patients, few of them get endobronchial intubation. In our study we could not diagnose short trachea preoperatively but endobronchial intubation occurred in 2 patients probably owing to short trachea.<sup>1</sup>

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Children with meningoencephalocele have an increased incidence of latex allergy<sup>7</sup>, which can manifest as intraoperative cardiovascular collapse and bronchospasm.

The neurological prognosis in such children depends on the amount of neural tissue that has herniated through the sac. The neural tissue is often dysplastic and gliotic but the presence of microcephaly with a large posterior encephalocele containing significant brain tissue is a predictor of poor neurological outcome. The decision regarding surgery is dependent on various factors including the amount of neural tissue in the sac, other congenital anomalies, etc. The decision must involve the family and other medical personnel.

Process of recovery is crucial as well. The criteria for extubation include intact cough and gag reflex, a negative inspiratory force of 15-30 cm H<sub>2</sub>O, a force vital capacity breath in excess of 10 ml/kg, an air leak around the tube with inflation pressures less than 25 cm H<sub>2</sub>O as well as the maintenance of acceptable oxygenation and ventilation on minimal ventilator support. Adequate reversal of neuromuscular blockade is demonstrated by either a sustained arm lift or leg lift. Extubation should be performed only when the child is awake and breathing well. In our study 32 cases were extubated on the operating table but 5 patients had delayed recovery, one of them needed post-operative ventilation. The possible causes of delayed recovery are hypothermia, inadequate reversal, prolonged effect of the anaesthetic agent, electrolyte imbalances and malnutrition.

Despite aggressive surgical and medical management 15-30% of neonates with EMC die within the first 5 year of life, majority of deaths are attributable to severe CM II. Developmental abnormalities of brain such as corpus callosum agenesis, microgyria, porencephalic cyst, and arachnoid cyst are commonly associated apart from Chiari malformations.<sup>15,16</sup>

**LIMITATIONS OF THE STUDY:** It is a retrospective study and carried out in small number of children without any neonatal or paediatric backup, therefore, the study may not reflect the true morbidity and mortality.

**CONCLUSION:** The children with EMC have several associated CNS abnormalities which are very prone to have perioperative complications. Most of the complications are manageable through early and precise diagnosis, meticulous preoperative preparations, vigilant intraoperative monitoring and preparation for anticipated complication.

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