



Case Report

Anaesthesia Approach for Cerebral Hemispherotomy in a Child with Hemimegaencephaly and Cortical Dysplasia with Intractable Epilepsy

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Abstract

Cerebral Hemispherectomy (CH), is an advanced and rare surgical treatment to reduce or eliminate intractable seizures in children with Hemimegaencephaly (HME) and Cortical Dysplasia (CD). Aim of early surgical intervention is to have a seizure free patient.

Presenting a case of a 12 month, 12 kg child, with seizures from day 13 of birth in the form of small jerks lasting for 3 to 4 minutes multiple times a day and was prescribed a multidrug regimen of anticonvulsants since then. Brain MRI showed right megalencephaly with dysplastic cortex of white matter. The patient was induced with standard intravenous induction, endotracheally intubated, and maintained on normocapneic volume controlled ventilation.

The duration of the surgery was approximately 9 hours. A blood loss of almost the patient's entire blood volume was observed (80 ml/kg), with subsequent hypotension, fall in hemoglobin and metabolic disturbances accompanying massive blood loss. There were no intraoperative seizures. The patient was ventilated electively for 3 days postoperatively, with no postoperative complications and uneventful Pediatric Intensive Care Unit (PICU) stay.

Keywords: *pediatric neurosurgery, intractable seizures, hemimegalencephaly, cortical dysplasia, cerebral hemispherotomy.*

Background

Intractable seizures due to hemimegalencephaly (HME) with cortical dysplasia requires a highly skilled, multispecialty approach in a multidisciplinary centre, involving a team of neurosurgeons, neuroanaesthetists, paediatricians, neurologists, pediatric intensivists.

Epilepsy is a common neurological disorder presenting with seizures. Prevalence in India is 6 per 1000.¹

Intractable epilepsy is defined as when 2 or more seizures occur per month for a period of 2 years or more, and a failure to control the seizures by a combination of suitable first line and second line anticonvulsant medications.

Patients suffering from intractable seizures can be evaluated for surgical control of seizures, following an extensive evaluation and drawing up an appropriate management plan involving a team of physicians, surgeons and intensivists.²

There exist various approaches to surgical management of intractable seizures, customised for the needs of the patient by the neurosurgeons and neurologists. The aim of surgical management is, ideally, to have a seizure-free patient, with minimal complications, for an improved quality of life.

Anesthetic approach to cerebral hemispherotomy is thus specific to the type of surgery planned.^{3,4}

Hemimegalencephaly (HME) is a rare congenital neurological condition, with prevalence ranging from 1 to 3 cases per 100 epileptic children. It occurs due to a spontaneous mutation in the third week of gestation, in which one half of the brain is abnormally larger than the other half, accompanied by malformed neurons, leading to seizures that are often intractable and difficult to control with a regime of multiple anticonvulsant drugs. Hemimegalencephaly is frequently also associated with hemiparesis, psychomotor growth retardation, feeding difficulties in addition to seizures. Early surgical intervention gives the patient a possibility of seizure-free and improved quality of life. Thus, a cerebral hemispherotomy is definitely a promising therapeutic option for controlling seizures.⁵

Cortical dysplasia is a condition with localised malformation of the cerebral cortex involving either the temporal lobes or the frontal lobes, or both. It's prevalence ranges from 1% to 14% in patients with cortical abnormalities. ⁵ Brain MRI shows focal cortical thickening or thinning, areas of focal cerebral atrophy and blurring of the grey matter-white matter junction. The extent of cortical dysplasia on brain MRI would help with assessment of the extent of surgical dissection required and anticipated complications, both intraoperatively and postoperatively.

Hemispherotomy was first performed by Dandy in 1923 as the treatment for diffuse hemispheric glioma.⁶

Cerebral hemispherotomy is a difficult procedure requiring considerable skill as the vasculature is malformed with the loss of normal anatomy, leading to blood loss intraoperatively. In addition,

the patients who commonly undergo their procedure are children, who have a higher blood volume to body weight ratio.

Cerebral hemispherotomy could be either anatomical or functional

Anatomical cerebral hemispherotomy: It is a precise method involving isolation of the affected hemisphere, that is, the seizure focus or foci, from the normal, healthy region. This approach is difficult to achieve in practice, with considerable morbidity and mortality, massive blood loss, complications like delayed hydrocephalus.

Functional cerebral hemispherotomy: There are 4 goals to this approach, namely, disconnection of the corticothalamic tract (internal disconnection of the internal capsule and corona radiata), resection of the medial temporal structures, total corpus callosotomy, and disconnection of the orbitofrontohypothalamic tract (disruption of the frontal horizontal fibers). This approach is widely accepted and safer than anatomical resection.⁷

Case Report

A 12 month old male child weighing 12 kilograms presented with seizures in the form of jerks lasting 3 to 4 minutes multiple times a day with abnormal posturing and without loss of consciousness, starting from day 13 of birth. The patient was placed on a multipurpose anticonvulsant regimen.

The patient was a full term, vaginally delivered child, APGAR scoring was not available, and developed neonatal jaundice on day 2 of life, requiring phototherapy.

The patient was shown to have a history of feeding difficulties such as frequent choking on feeds, frequently suffering from upper respiratory tract infections, lower respiratory tract infection probably due to aspiration requiring bronchodilator and antibiotic treatment, starting 10 months of age.

The patient was on a combination of Vigabatrin, Topamac and Levitiracetam to control his seizures.

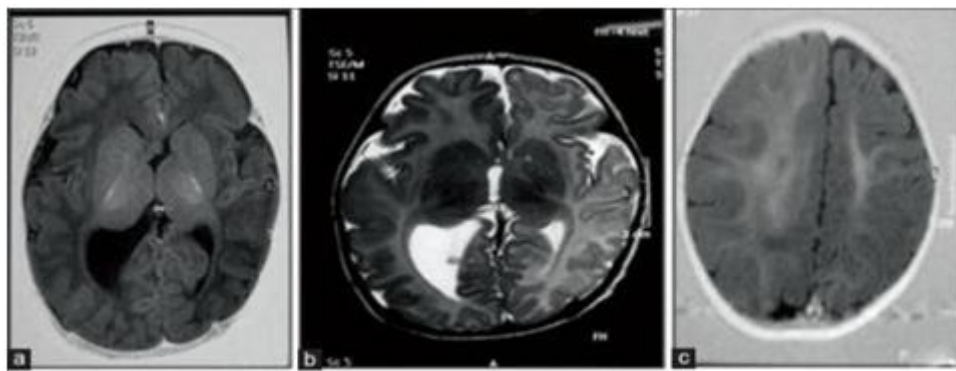
On examination, the patient was not very active, hypotonic, had delayed milestones, absent

spontaneous leg movements. Examination of the respiratory system, gastrointestinal system, genitourinary system and cardiovascular system showed no abnormality.

Coagulation profile, liver function tests, serum creatinine, serum electrolytes, arterial blood gas analysis were within normal limits.

Chest X ray showed right upper lobe consolidation

Brain MRI showed right megalencephaly with dysplastic cortex. (Figure 01)



MRI showing widespread cortical dysplasia in right hemisphere with right sided hemimegalencephaly

Figure 01

Surgical Approach

Navigation guided right frontotemporal craniotomy for a functional right hemispherotomy was planned.

Anaesthesia Management

- The patient was given his morning dose of anticonvulsants before shifting him to the operation theater.
- Sedation in the ward and antihistamine agents was avoided to avoid respiratory depression and triggering seizures, respectively.
- Preoperative hydration was done with 2 ml/kg normal saline.
- The patient was preoxygenated. Adjuvant medications like Fenatnyl at the dose of 5 microgram/kg and atropine at dose of 0.01 milligram/kg were given.
- Thiopentone sodium at the dose of 3 milligram/kg was chosen as the induction agent. Muscle relaxation as Vecuronium at the dose of 1 milligram/kg was given.
- Airway was secured with a 4.5 number non-cuffed endotracheal tube. The patient was ventilated using volume control ventilation at

a tidal volume of 10millilitre/kg, respiratory rate of 16 to 18 breaths per minute.

- Intravenous access was secured with one 20 gauge peripheral line. One 22 gauge peripheral line was used for muscle relaxant infusion for maintenance of anaesthesia. A triple lumen right internal jugular venous access for rapid fluid, blood and blood product infusion was also secured.
- Invasive radial arterial blood pressure monitoring was done throughout the surgery and continued postoperatively.
- Urinary catheterisation for monitoring urine output and establishing adequacy of fluid administration was done.
- A rectal probe for temperature monitoring was secured.
- The patient was positioned supine with a 10 degree neck extension and the head was rested on a well-cushioned horseshoe rest.
- Anaesthesia was maintained using a combination of oxygen, air and Desflurane along with an infusion of Atracurium at 7 microgram/kg/min.

- End tidal carbon dioxide (EtCO₂) was kept between 32 to 35 mm Hg, mean arterial pressure between 50 to 60 mm Hg.
- A surface warmer was used to maintain normothermia.
- Appropriate antibiotics, antacid, steroids were given.
- Blood and blood products were kept ready in the operating room.

Monitoring

- Electrocardiogram (ECG)
- Oxygen saturation (SpO₂)
- End tidal carbon dioxide (EtCO₂)
- Noninvasive blood pressure (NIBP)
- Central venous pressure (CVP)
- Invasive arterial blood pressure (IBP)
- Urine output
- Rectal temperature
- Bispectral index (BIS)
- Blood sugar
- Arterial blood gas sampling
- Serum electrolytes

Intraoperative Observations

- Total operative time was about 9 hours.
- Blood pressure fell by about 30 mm Hg, and was corrected by using boluses of crystalloids, blood and blood products and knot ropes, as needed.
- Tachycardia resulting from hypotension.
- Blood loss was calculated to be approximately 700 milliliters. In a child weighing 12 kilograms, total blood volume equals approximately 960 to 1000 milliliters, assuming a blood volume of 80 milliliters/kg. Thus, it can be calculated that 70% to 73% of the patient's total blood volume was lost.
- Urine output was maintained at 1 milliliter/kg/hour.
- Acidosis on arterial blood gas analysis was corrected using sodium bicarbonate.
- Mild hypothermia up to 34 degree Celsius was observed and surface warmer

temperature adjusted to raise core temperature appropriately.

- No hypoglycaemia was observed.
- No seizures occurred intraoperatively and postoperatively.
- No cerebral congestion or brain bulging occurred.
- No venous air embolism occurred.
- Total fluid, blood and blood products infused equalled a volume of 1300 milliliters.
- Hemoglobin dropped to 6 gram percent in the postoperative period.
- No pulmonary oedema or coagulopathy occurred as a consequence of massive blood transfusion.

The patient was electively ventilated and shifted to the pediatric intensive care unit (PICU).

Intensive Care Management

- The patient had 2 seizures in the postoperative period.
- Anticonvulsant medications were adjusted in dosage and frequency as per the neurologist's recommendations.
- The patient was weaned off the ventilator and the endotracheal tube removed on day 3 postoperatively.
- The patient showed no improvement in physical activity and hypotonia.

The patient was discharged after 20 days of hospital stay, on a 2 drug anticonvulsant regimen.

Anticipated Anaesthesia Problems with Suggested Solutions

- Anaesthesia management of CH and Cortical dysplasia is extensive, highly challenging, and requires continuous vigilance.⁸
- Basic principles of neuroanaesthesia need to be followed. The optimal partial pressure of carbon dioxide (PCO₂) range is between 32–34 mmHg in order to achieve maximum brain relaxation and a loading dose of steroids (dexamethasone) is also useful.⁹
- Cerebral hemispherotomy generally involves infants and small children, hence, it

is technically difficult for accessing intravenous, intra arterial and central jugular or subclavian lines for monitoring.¹⁰

- Cerebral hemispherotomy requires a large craniotomy and severe to massive blood loss is anticipated.¹¹ Furthermore, cortical dysplasia has abnormal anatomy and vasculature that can contribute to intraoperative bleeding and subsequent events like anaemia, electrolyte and metabolic disturbances and coagulopathy. Blood investigations like hemoglobin and haematologist monitoring, arterial blood gas analysis, and a coagulation profile needs to be done perioperative for quick ongoing corrections.¹² As haemodynamic instability or even hypovolemic shock is expected due to massive blood loss, blood, fresh frozen plasma, platelets and colloids should be available.
- It is mandatory to have a central venous access for central venous pressure monitoring as urine output is not a very good indicator of fluid status. CVP may also not be a very good predictor of response to fluid management with such massive blood loss. In such cases, pulse pressure variability and systolic pressure variability have been shown to be better predictors of response to fluid management than CVP and pulmonary artery occlusion pressure. If available, these monitoring modalities may be utilised¹³
- There is an inherent risk of venous air embolism during craniotomy and dissection, detected by sudden fall in end tidal carbon dioxide, needs to be informed to the surgeon immediately and controlled¹⁴
- Sudden cerebral edema, brain bulging, congestion, sudden rise in BIS, end tidal carbon dioxide, may be indicative of intraoperative seizures⁴, due to anaesthetic factors like proconvulsant anaesthetic agents, light anaesthesia hypo-/hypercapnia, hypoxemia, metabolic factors like hypoglycemia, hypo-/hypernatremia,

hypocalcemia, hypomagnesemia or uremia.⁹ Instant cold saline irrigation and drugs like benzodiazepines or barbiturates are useful in controlling seizures. However, care should be taken during cold saline irrigation as this may cause severe bradycardia and sinus arrest during irrigation¹⁵

- Hypothermia is prevented by suitable warming devices.
- Neurogenic pulmonary edema is a common complication after cerebral hemispherotomy and extensive subcortical resection. It is indicated by severe decrease in cardiac index, bradycardia, increase in systemic vascular resistance, and increase in alveolar to arterial gradient¹⁶
- Adequate depth of anaesthesia is to be maintained, as a light plane can induce intraoperative seizures. Hence, a bispectral index of 50 to 70 (BIS) is helpful¹⁷ The postauricular placement of the bispectral electrodes is a suggested alternative to conventional frontal lobe placement.
- Enzyme induction caused by anticonvulsant medications can increase the dose and frequency of most anaesthesia drugs, therefore plasma levels of these drugs have to be optimized before going in for surgery.
- The action of anaesthetic agents on electroencephalogram (EEG) is complex and requires a balanced titration and a well thought out plan for the surgery¹⁸
- The proconvulsant and anticonvulsant effects of anaesthetic agents have to be considered while choosing drugs for general anaesthesia
- Severe bradycardia/sinus arrest may occur during Amygdala hippocampectomy, that is not seen during routine anterior temporal lobe resection. This is due to surgical limbic system stimulation resulting in enhanced neural vagal activity¹⁹
- Inotropes to maintain MAP
- It is not prudent to extubate because of long duration of surgery, slow emergence,

haemodynamic instability/hypovolemic shock, massive blood loss requiring massive blood transfusion, laryngeal edema and postoperative seizures leading to airway compromise if extubated.

- Postoperative period for cerebral hemispherotomy is equally stormy requiring continuous monitoring. Early postoperative seizures are expected with the potential for it progressing to status epilepticus. Neurogenic pulmonary oedema can be expected. Metabolic changes, electrolyte disturbances and anaemia to be assessed and treated aggressively.
- DVT prophylaxis is another major consideration. Appropriate measures for its prevention need to be applied.
- Postoperative surgical complications like cerebral hematoma, cerebral edema, late hydrocephalus can occur requiring further surgical intervention.
- Anticonvulsant medications need to be continued and then tapered as per the neurologist's/paediatrician's suggestions.

Conclusion

In a child of hemimegalencephaly and cortical dysplasia with intractable epilepsy, the aim of functional hemispherotomy is the reduction in frequency of seizures and improvement in quality of life.

The challenge for anaesthesiologist starts right from the preoperative evaluation, including an understanding of the antiepileptic drugs and their interaction with anaesthesia agents, the prolonged operative time and technical difficulties in resection and separation of the affected hemisphere, the intensity of bleeding involved during resection due to cortical dysplasia requires anaesthesiologist to be on their toes. The requisition of invasive monitoring adds to the challenge of induction.

These patients are susceptible to hypovolemic shock, hypothermia and electrolyte and metabolic

disturbances, managing these is a continuous process.

Prevention and detection of perioperative and early postoperative seizures causing brain bulging and congestion is a challenge by itself.

Postoperative haemodynamic and ventilator respiratory care is to be continued as a team with the paediatrician, till the child is off ventilator and able to manage his airway to prevent aspiration.

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