

## Case report

**ANAESTHETIC MANAGEMENT OF MOYAMOYA DISEASE IN A SIX-YEAR-OLD CHILD (ANAESTHETIC MANAGEMENT OF MOYAMOYA DISEASE)**Shubhi Singhal<sup>1</sup>, Neerja Gaur Banerjee<sup>1</sup>, Devang Bharti<sup>1</sup>, Mohandeep Kaur<sup>1</sup><sup>1</sup>Atal Bihari Vajpayee Institute of Medical Sciences (ABVIMS) & Dr. Ram Manohar Lohia (RML) Hospital, New Delhi, India

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**Abstract**

**Introduction:** Moyamoya disease is a rare form of the chronic cerebrovascular disorder. Patients develop progressive stenosis or occlusion of the terminal portion of the internal carotid arteries and a proximal portion of the anterior and middle cerebral arteries. The compensatory collateral circulation that develops is weak and small, prone to hemorrhage, aneurysm, and thrombosis. **Case Report:** We report a case of a six-year-old boy with Moyamoya disease, presenting with complaints of seizures and weakness of the left side of the body. A bilateral encephalo-myo-synangiosis (EMS) was successfully performed on the child under general anesthesia without any perioperative complications. **Conclusion:** Revascularization surgery in a patient with Moyamoya disease is a high-risk procedure with an increased incidence of perioperative neurological sequelae. However, patients can be successfully perioperatively managed by ensuring adequate cerebral blood flow and minimizing cerebral metabolic oxygen consumption rates.

**Key words:** Moyamoya disease; Encephalomyosynangiosis; Paediatric anesthesia

**Introduction**

Moyamoya disease is a rare form of the chronic cerebrovascular disorder that leads to progressive stenosis of the terminal portion of the internal carotid arteries and the proximal portions of anterior and middle cerebral arteries. Collaterals compensate for this blockage, and on cerebral angiography, they give the characteristic appearance of a „puff of smoke” („Moya Moya” in Japanese)<sup>1</sup>. This compensatory collateral circulation is weak and small, prone to hemorrhage, aneurysm formation, and thrombosis. Various surgical anastomotic techniques are used to ensure extracranial to intracranial revascularization, augmenting the blood flow to affected hemispheres. Encephalo-myo-synangiosis is one such technique, which involves the placement of the superior temporal artery over the cerebral hemisphere.

**Case Report**

A six-year-old boy, weighing 15 kg, presented at our institution with complaints of generalized

tonic-clonic seizures (1–2 episodes per day) and left-sided weakness for 15 days. Contrast-enhanced magnetic resonance (MR) imaging of the brain with magnetic resonance angiography (MRA) revealed bilateral distal internal carotid artery occlusion, right middle cerebral artery and bilateral anterior cerebral artery attenuation, and presence of multiple collaterals. This led to the diagnosis of Moyamoya disease, and we planned a surgical procedure bilateral encephalo-myo-synangiosis for this child.

At the preoperative assessment, the child responded well to the treatment of Phenytoin 60 mg intravenously (IV) twice, Sodium Valproate 250 mg IV once, and oral Aspirin 75 mg (PO) once daily. He was seizure-free for the last 48 hours prior to the operation. Premedication included 3 mg midazolam intranasally, and the mother accompanied the patient to the operation room. Two IV cannulas (20G) were secured under local anesthesia, and fentanyl 30 mcg, along with a fluid bolus of 200 ml over 20 minutes, was administered. Monitoring included pulse oximetry (SpO<sub>2</sub>), five lead electrocardiography (ECG), invasive blood pressure,

central venous pressure (CVP), end tidal-carbon dioxide (EtCO<sub>2</sub>), temperature, and bispectral index (BIS). We induced anesthesia using propofol 45 mg IV (in titrated dosage) and vecuronium 1.5 mg IV for muscle relaxation. After administering lidocaine 22.5 mg IV, the trachea was intubated with an uncuffed, 5.0 mm ET tube, fixed at 15 cm. We maintained anesthesia with isoflurane 1% in 60% oxygen/air mixture and vecuronium infusion (1 mg/hr). Throughout the procedure, a mean arterial pressure of 50–80 mmHg and EtCO<sub>2</sub> of 30–35 mmHg was maintained. Forced air warmer and warm fluids were used to ensure optimal body temperature. Analgesia consisted of fentanyl infusion (15 mcg/hr) and paracetamol infusion 200 mg IV. Total blood loss during the five-hour-long procedure was 110 mL, which was replaced completely using packed red blood cells. After completing the procedure, Ondansetron 1.5 mg was given and residual neuromuscular blockade was reversed using slow IV administration of glycopyrrolate 150 mcg and neostigmine 750 mcg. When the patient started generating adequate tidal volume and could perform a sustained head lift, trachea was extubated after gentle oral suctioning. No adverse events were noted intraoperatively. Postoperative stay of the child in the hospital was uneventful.

## Discussion

The main goal of both anesthetic and surgical management in Moyamoya disease patients is to maintain cerebral perfusion. These patients have reduced cerebral blood flow, which is sufficient to match the basal cerebral metabolic oxygen consumption rate. In children, even a transient decrease in cerebral blood flow due to any external factor may lead to new neurological deficits. Thus, all the factors which either decrease cerebral blood flow or increase cerebral metabolic oxygen consumption rate need to be avoided.

Crying causes hyperventilation and a consequent decrease in the cerebral blood flow. Allaying the patient anxiety by allowing the parent to accompany the child, the distraction with toys and music and minimizing pain with intranasal or oral preparations, local anesthesia for painful procedures, excellent perioperative analgesia reduce the risk of crying. Cerebral autoregulation is impaired

in Moyamoya disease patients, predisposing the patient to episodes of both hypotension and hypertension. The weak collateral circulation is especially sensitive to blood pressure changes and might rupture during hypertension episodes. Hypotension at the time of induction may be prevented by sufficient preloading and slow induction of agents in a titrated manner. Maintaining an adequate depth of anesthesia at all times avoids hypertension. Pharmacological agents, like lidocaine, may also be used to blunt the hemodynamic stress response.

To maintain adequate cerebral perfusion, EtCO<sub>2</sub> should be kept in the high normal range. Normothermia should also be ensured as hypothermia may lead to vasospasm and reduced cerebral blood flow, while hyperthermia may trigger ischemic events. There is no recommendation regarding the use of any particular inhalational anesthetic for Moyamoya disease patients. Some authors even suggest complete avoidance of such agents for fear of cerebral steal phenomenon<sup>2</sup>. We decided to use isoflurane because it increases cerebral blood flow and causes a dose-related decrease in cerebral metabolic oxygen consumption rate<sup>3</sup>. Also, isoflurane's cerebral protective effects have been well documented during transient cerebral ischemia in adults undergoing carotid endarterectomy<sup>4</sup>.

The blood's oxygen carrying capacity is of prime importance in these patients, and anemia should be corrected in the preoperative period itself. Similarly, the threshold for intraoperative blood transfusion should be decreased. Real-time monitors to detect cerebral ischemia are especially useful in the intraoperative period. In our case, electroencephalogram electrodes could not be placed, as they were interfering in the surgical field, and near-infrared spectroscopy was not available. So, we employed a bispectral index for this purpose. Although the manufacturer of the device clearly states that the bispectral index is not intended to monitor cerebral ischemia, many cases have been reported where the bispectral index has been successfully used to detect cerebral ischemic events.<sup>5-8</sup>

Unless contraindicated, extubation should be performed after completion of the procedure, in the operation theatre itself or early in the postoperative period. This facilitates early postoperative neurological examination. Care should be taken to

prevent pain and postoperative nausea and vomiting after the surgery.

### **Conclusion**

Revascularization surgeries in patients with Moyamoya disease are high-risk procedures with an increased incidence of perioperative neurological sequelae. However, such cases can be successfully managed perioperatively by ensuring adequate cerebral blood flow and minimizing cerebral metabolic oxygen consumption rates.

### **References**

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