VOLUME - 15, ISSUE - 05, MAY - 2026 • PRINT ISSN No. 2277 - 8160 • DOI : 10.36106/gjra

Original Research Paper

Anaesthesiology

ANAESTHETIC MANAGEMENT OF MOYA-MOYA DISEASE IN PEDIATRIC PATIENT UNDERGOING DIRECT REVASCULARISATION SURGERY: A CASE REPORT

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ABSTRACT

Background: Moya-Moya disease is a chronic progressive cerebrovascular disorder characterised by stenosis or occlusion of the intracranial internal carotid arteries and development of fragile collateral vessels. These patients are at high risk of cerebral ischemia or haemorrhage during the perioperative period. Anaesthetic management focuses on maintaining adequate cerebral perfusion, avoiding hypotension, hypocapnia, and hypovolemia, and ensuring hemodynamic stability. We report the anaesthetic management of a 3-year-old Female diagnosed with Moya-Moya disease, posted for Direct Revascularisation surgery. A carefully planned anaesthetic strategy emphasising maintenance of cerebral perfusion, normocapnia, normovolemia, and avoidance of sympathetic fluctuations was implemented. The intraoperative and postoperative course remained uneventful with no neurological deterioration.

KEYWORDS : Moya-Moya Disease, Cerebral Perfusion, Anaesthesia, Ischemia, Collateral Circulation, Neuroprotection.

INTRODUCTION

Moya-Moya disease is a rare cerebrovascular disorder characterised by progressive stenosis of the terminal internal carotid arteries and their proximal branches, leading to the formation of abnormal collateral vessels at the base of the brain. The angiographic appearance resembles a "puff of smoke," which gives the disease its name. Patients commonly present with ischemic strokes, transient ischemic attacks, seizures, or intracranial haemorrhage. Anaesthetic management is particularly challenging due to impaired cerebral auto-regulation and dependence on fragile collateral circulation. This case highlights the anaesthetic considerations and management strategies in a paediatric patient undergoing direct revascularisation surgery.

CASE REPORT

A 3-year-old female child weighing 12 kg, diagnosed with Moya-Moya disease, was scheduled for direct revascularisation surgery. The patient had a history of generalised tonic-clonic seizures for two years, progressive upper limb weakness, and recent onset focal seizures with left-sided hemiplegia for one month. She was on levetiracetam and aspirin therapy. On preoperative evaluation, the child was awake, conscious, and haemodynamically stable, with neurological examination revealing left-sided hemiplegia. Airway assessment was unremarkable. Laboratory investigations showed haemoglobin of 9.6 g/dL, normal renal and liver function tests, and an INR of 1.1. MRI brain demonstrated a non-hemorrhagic infarct in the right ACA/MCA watershed region, while MR angiography revealed the characteristic "puff of smoke" appearance consistent with Moya-Moya disease.

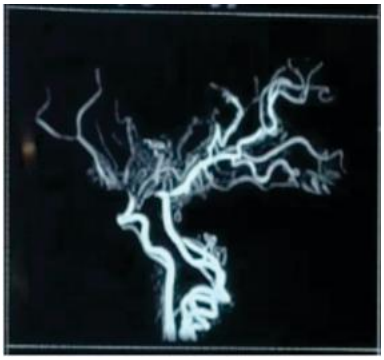

Figure 1: Puffed Smoke Appearance on MR Angiography

Adequate preoperative hydration was ensured and anticonvulsant and anti-platelet therapy were continued. Peripeheral intravenous line 20G was secured preoperatively. Care was taken to avoid prolonged fasting, anxiety, and crying to prevent hypocapnia-induced cerebral vaso-constriction. Premedication with intravenous midazolam (0.03mg/kg) was administered cautiously. Intraoperative monitoring included standard ASA monitors (ECG, Spo2, NIBP) ,end-tidal CO_2 monitoring, temperature probe and urine output measurement. Induction was performed with pre-oxygenation followed by administration of propofol (2mg/kg),remifentanil (4ng/ml) using target-controlled infusion, and atracurium (0.75mg/kg). The airway was secured using direct laryngoscopy with a 4.5 mm internal diameter microcuffed endotracheal tube, and correct placement was confirmed by capnography and auscultation. Anaesthesia was maintained using sevoflurane in an oxygen-air mixture along with remifentanil infusion and intermittent atracurium. Ventilation was adjusted to maintain normocapnia with end-tidal CO_2 between 35–40 mmHg. The surgery lasted five hours with a blood loss of 180 ml. The patient received crystalloids and packed cell transfusion (200ml) and urine output was 120ml. At the end of the procedure, smooth extubation was performed after reversal of neuromuscular blockade with neostigmine (0.05mg/kg) and glycopyrrolate (8mcg/kg) ensuring minimal sympathetic stimulation.The patient was shifted to the paediatric intensive care unit for close monitorinmg. Neurological status was assessed regularly, and adequate analgesia was provided with paracetamol suppositories (20mg/kg) with opioids (Inj.Tramadol) administered if required. The postoperative period remained uneventful, with no evidence of neurological deterioration or complications

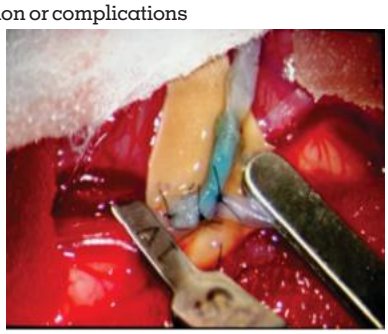

Figure 2 : Critical Anastomosis Step in Direct Cerebral Revascularisation using STA-MCA Bypass

DISCUSSION

Moya-Moya disease has a incidence of 0.35 to 2.3 per 100,000 worldwide. In india the prevelance range is between 3.2 to 10.5 per 100,000. Moya Moya disease is more common in females peaking in children between 3 to 8 years. Moya-Moya disease presents as seizures, hemiparesis, TIA, Cognitive delays. Moya-Moya disease presents significant anaesthetic challenges due to impaired cerebral blood flow and reliance on fragile collateral circulation. Cerebral perfusion in these patients is pressure-dependent, making them highly susceptible to ischemia during episodes of hypotension. Carbon dioxide levels play a crucial role, as hypocapnia can lead to cerebral vasoconstriction and ischemia, while hypercapnia may cause a steal phenomenon, worsening cerebral perfusion. Maintaining hemodynamic stability during induction, laryngoscopy, and extubation is essential to prevent fluctuations in cerebral blood flow. Adequate fluid management is necessary to maintain euvolemia and support cerebral perfusion, while normothermia must be ensured to avoid increased cerebral metabolic demand. A well-planned anaesthetic strategy focusing on these principles can significantly reduce perioperative complications and improve outcomes

CONCLUSION

This case highlights the importance of meticulous perioperative anaesthetic management in patients with Moya-Moya disease. Maintenance of cerebral perfusion, avoidance of hemodynamic fluctuations, and strict control of ventilation are critical in preventing perioperative neurological complications. With careful planning, vigilant monitoring, and adherence to neuroprotective strategies, safe anaesthetic outcomes can be achieved in such high-risk patients

REFERENCES

1. Scott RM, Smith ER. Moyamoya disease and moyamoya syndrome. N Engl J Med. 2009;360:1226–1237.
2. Gupta DK, et al. Anaesthetic management of Moyamoya disease. J Neurosurg Anesthesiol. 2011;23;100-109
3. Kato H, et al. Perioperative management in Moyamoya disease. Stroke. 2015;46;293-298
4. Smith ER. Surgical and anaesthetic considerations in Moyamoya disease. Neurosurg Clin N Am. 2010;21;3;517-524