

**ANAESTHETIC APPROACH FOR ELECTIVE CAESAREAN SECTION IN A  
PREGNANT PATIENT WITH MOYAMOYA DISEASE – CASE REPORT**

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**ABSTRACT**

Moyamoya disease (MMD) is defined by vaso-occlusive changes at the internal carotid and cerebral arteries, leading to proliferation of an abnormal vascular network at the base of the brain. The disease is manifested by ischemic, hemorrhagic strokes or transient ischemic attacks. Pregnant and puerperal MMD patients may be at a higher risk of developing new strokes as well as during the peripartum period. Since hyperventilation-induced cerebral ischemia and hypertension are provoked by active labor, elective cesarean section has been recommended to avoid neurologic complications for pregnant women with moyamoya disease. We report the successful use of epidural anesthesia in the management of a term parturient with moyamoya disease undergoing cesarean section.

**KEYWORDS:** MMD(Moya moya disease), ICA(Internal carotid artery), MCA (Middle carotid artery), PCOM (Posterior communicating artery), COW(Circle of willis), DVT( deep vein thrombosis).

**INTRODUCTION**

Moyamoya disease(MMD) is a rare disease named after the Japanese word describing the characteristic “puff of smoke” appearance seen on cerebral angiography. It is defined by vaso-occlusive changes at the internal carotid and cerebral arteries, leading to proliferation of an abnormal vascular network at the base of the brain manifested by ischemic, hemorrhagic strokes or TIA. The disease predominates in females at 2:1 ratio. The prevalence of MMD is higher in Japan, more common in children and females; there may be a familial predisposition. Genes located on chromosomes 17q25 recently 8q23 was discovered. MMD has a bimodal age distribution, with peaks in the first and fourth decade of life.

**CASE REPORT**

26 year old G2P1L1 was admitted with diagnosis of MMD. She had a history of stroke 2 years back; which was after her first child birth. Neurosurgical consultation was obtained and MMD diagnosis was confirmed through MRI Angiograph brain, showed moderate to severe stenosis of right ICA and right MCA and mild to moderate stenosis of left ICA (C5, C6 and C7 segments) and MCA (M2 segment), azygous A2 segment, moderate stenosis of right PCOM associated with multiple fragile tiny collaterals along the basal aspect of COW- likely represents moyamoya disease.(fig 1) She was treated with Aspirin. Her left sided weakness had improved over a period of 2 years. Patient’s father had a similar episode of CVA 10 years back, he was not on any treatment. His weakness improved gradually.



(Fig 1): Without Any Residual Neurological Deficits.

Patient remained without any further symptoms throughout pregnancy. Cardiology consultation was obtained, Echocardiogram showed Ejection fraction of 60% with no structural heart abnormalities and advised to continue antiplatelet throughout the pregnancy. Elective Cesarean section was indicated due to maternal disease.

Pre-anesthetic checkup was done before the procedure Antiplatelet was stopped 5 days before the surgery according to ASRA (American society of regional anaesthesia) guidelines to minimize the bleeding risk. Patient was accepted under ASA III. A controlled neuroaxial anaesthesia (Epidural) was chosen.

After the consent for anaesthesia was taken patient was shifted to OT. Standard ASA monitors were applied and

baseline parameters were recorded. After Intravenous preload with lactate Ringer's solution, under aseptic precautions an epidural catheter was placed in L2-L3 interspace. Incremental injections of 0.5% bupivacaine and fentanyl 50mcg was injected through epidural catheter every 4-5 min interval. Total volume of 14 ml (5+5+4) was injected. Level of sensory analgesia to pinprick was attained upto T10 and surgery was started. Healthy male infant was delivered. Oxytocin 15units/hr was given as slow iv infusion. Patient was haemodynamically stable throughout the procedure. Blood loss was 1300 ml. Post-operative analgesia was provided with inj. Bupivacaine 0.125% of 8ml/hr infusion through epidural catheter. Catheter was removed 24 hours after the procedure and antiplatelet medication was restarted.

### DISCUSSION

Moyamoya disease is a rare disease named after the Japanese word describing the characteristic "puff of smoke" appearance that is seen on cerebral angiography. It has most often been reported in the Japanese, is more common in children and females and there may be a familial predisposition. Hypercoagulability, venous stasis and endothelial lesion, common during pregnancy, may contribute to the occurrence of stroke. Therefore pregnant MMD patients have more vulnerable cerebral vessels and may be at a higher risk than pregnant women without the disease. Mismatch of cerebral blood flow, cerebral metabolic rate of oxygen (CMRO<sub>2</sub>), due to hypotension can aggravate ischemia, for example, dehydration secondary to vomiting in the last trimester of pregnancy and blood loss during delivery. Hypertension secondary to pain-induced increased sympathetic drive could lead to intracranial bleeding; hence, it is crucial to maintain hemodynamic stability and adequate pain control by early initiation of labor epidural with adequate hydration.

Elective Caesarean section has been recommended for the pregnant patient with moyamoya disease to avoid possible deleterious effects of pain, hyperventilation and bearing-down during labour. General anaesthesia has been used and has the potential advantages of decreasing the cerebral metabolic rate for oxygen which may confer some protection against ischaemia. General anesthesia in patients with severe anxiety may be beneficial for the prevention of hypocapnia. In contrast, general anesthesia may produce low neonatal APGAR score at birth due to fetal circulation of drugs administered to the mother. Here is also a risk of bronchoaspiration and difficult tracheal intubation during general anesthesia. The hypertensive response to intubation may precipitate intracerebral haemorrhage and although this response may be obtunded by the use of adjuvant drugs such as vasodilators, sympathomimetic antagonists and rapidly acting opioids, care must be taken to avoid hypotension and consequent cerebral ischaemia. Hypocapnia may cause neurological deficit, so use of capnography during controlled ventilation is mandatory. Anesthesia with

propofol has been successfully used for cesarean deliveries in MMD patients. Continuous propofol infusion reduces hypertensive response to laryngoscopy and intubation in the prevention of hemorrhagic stroke.

Sevoflurane in the cerebral vasculature creates two different effects. It reduces the need for metabolic reactions by vasoconstriction at low doses, and it creates vasodilation at high doses. It is known that clinical doses of sevoflurane preserve brain physiology better than isoflurane. In the study by Bora DINC *et al.*, chose to use anesthesia maintained with sevoflurane.

Regional anaesthesia avoids these risks but the associated sympathetic block may lower cerebral perfusion pressure causing ischaemia distal to the stenotic vessels. Spinal anaesthesia may reduce endogenous catecholamine levels with labor pain relief, resulting in decreased systemic vascular resistance. Furthermore, pain relief can prevent hypocapnia caused by hyperventilation. Spinal anaesthesia was associated with convulsions and hemiparesis in a child. Sharma and colleagues used extradural anaesthesia for Caesarean section in a parturient at 31 weeks' gestation and reported advantages that included continuous monitoring of conscious level and cerebral function in an awake patient. They emphasized the need to avoid hypotension but relied solely on crystalloid preload and uterine displacement to negate the effects of sympathetic block and did not provide details of haemodynamic changes. Because the parturient at term is particularly susceptible to hypotension during extradural anaesthesia, more active management may be necessary to maintain cerebral perfusion. However, large upward swings in arterial pressure should also be avoided because of the potential for intracerebral haemorrhage. In the study done by W. D. Ngan Kee and C. D. Gomersal found that using an infusion of ephedrine in addition to fluids and lateral tilt, arterial pressure was well maintained without marked fluctuations.

Hypercoagulability, venous stasis and endothelial lesion, common during pregnancy, may contribute to the occurrence of stroke. Therefore, pregnant MMD patients have more vulnerable cerebral vessels and may be at a higher risk of developing stroke, hence elective caesarean section has been recommended.

We had selected regional Epidural anaesthesia over GA to avoid the risk of haemodynamic response of intubation, which may precipitate intracerebral haemorrhage. Regional anaesthesia avoided the risk of GA but associated with sympathetic blocked and hypotension may lower cerebral perfusion and increased the risk of ischemic episode. Patients coagulation status should be optimized at the time of epidural catheter placement and level of anticoagulation must be carefully monitored during the period of catheterization to prevent the risk of spinal hematoma. Indwelling catheter should

not be removed in the presence of therapeutic anticoagulation.

Cases with hemorrhagic stroke as the initial manifestation are considered more severe than those with ischemic strokes. Multiple strokes or transient ischemic attacks (TIAs) can provoke mental deterioration and may be fatal due to hemorrhagic stroke, particularly at term or during labor.

The optimal treatment for ischemic MMD has not been determined. Hemorrhagic cases may require drainage and removal of the hematoma, depending on size and location. In ischemic cases, treatment is aimed at increasing blood flow to hypoperfused areas by creating collateral circulation. Surgery is performed in an attempt to reduce the formation of abnormal blood vessels and decrease the risk of hemorrhagic stroke produced by hemodynamic fluctuations in vessels associated by MMD. Early neurosurgical intervention is made to revascularize directly, indirectly or both. Direct procedures are more commonly used in adults, while indirect procedures are used in children. Platelet antiaggregants, vasodilators, calcium channel blockers and corticosteroids have been used with variable results. Antiplatelet agents are more commonly used than anticoagulants due to the known risk of hemorrhagic stroke in these patients. Diverse surgical procedures have been used such as superficial temporal artery-middle cerebral artery (STA-MCA) anastomosis, encephalo duro arterio synangiosis (EDAS) and encephalo duro arterio myo-synangiosis (EDAMS). Prognosis is more favorable with the STA-MCA surgery with prompt increase in the blood supply.

## CONCLUSION

It is essential to have a multidisciplinary team approach as MMD is rare. Bottom line is to optimize the patient's of moyamoya disease and accordingly plan for intraoperative management to maintain an optimal end organ perfusion and oxygen delivery to the mother and fetus. We concluded that Epidural anesthesia provides an opportunity for neurological assessment and hemodynamic stability of the patient intraoperatively and better post-operative analgesia can be provided through epidural catheter, hence may be preferred over general and spinal anaesthesia.

## Consent

Informed consent for the publication of the case report was obtained from the patient.

## Conflicts of Interest

The authors declare that they have no conflicts of interest.

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