Mooyamoya disease is a non-inflammatory, non-atherosclerotic and non-amyloid cerebral vasculopathy more often affecting Japanese and their descendents. It is an uncommon condition characterized by bilateral stenosis or occlusion of the terminal portion of Internal Carotid and or proximal portion of Anterior Cerebral or Median Cerebral Arteries. Diagnosis is confirmed by Brain Angiography, where pathological vessels are presented similar to ‘cigarette smoke cloud’ hence the origin of the Japanese word 'Moyamoya'. It affects primarily children and young adults, and is more prevalent among females. With the evolution of the disease, IQ may be decreased by up to 60% in a period of 5 to 9 years. In general, the disease is associated to other morbidities, such as asthma, Down syndrome and hypothyroidism.

Anaesthetic management of such patients must take into consideration the importance of preserving an adequate balance between brain blood flow and oxygen consumption. Patients are at risk of cerebral ischemia and hemorrhage, Neurological deterioration may occur during pregnancy.

CASE REPORT
A 28 years old female, para-1, 61 kg, was admitted for observation with a plan for elective caesarean section at 37 weeks of gestation. She had been diagnosed angiographically to have moyamoya disease 4 years ago after sudden onset of aphasia for which she was thoroughly investigated. Neurological assessment was Broca's Aphasia secondary to Lt. MCA territory infarct. MRI brain revealed Lt. Inf. Frontal lobe infarct. Cerebral DSA showed evidence of large and medium vessel involvement bilaterally. ESR -90/1st hr, CRP-6.34 ng/L, Low Complement, Weakly +ve ANA was reported. She was treated with Azathioprin and antiplatelet since diagnosis and made a good recovery with residual difficulty in framing sentences-slow and delayed articulation of sentences, but no other neurological deficit. Arterial pressure was in the range of 110-130/70-90mmHg. She had associated bronchial asthma and hypothyroidism. Her TSH was 4.70 microIU/ml, Free T4-1.20 ng/100ml, Hb-12.3gm%, TLC-8900/cumm, N:\L:M:B:3, FBS-90mg%; PPBS-98mg%. She was receiving Thyronorm 75 µg for hypothyroidism and salmeterol inhaler for bronchial asthma. She was premedicated with inj. ranitidine 50mg, metoclopramide 10mg iv and 0.3 molar soln. of sodium citrate 30ml orally.

Left radial artery and central venous cannulation were performed under local anaesthesia before induction of GA to monitor IBP, CVP and ABG. Self retaining urinary catheter was introduced to measure urinary output.

General anaesthesia was induced with thiopentone (2.5%) followed by succinylchline-50mg after adequate preoxygenation. Endotracheal intubation was accomplished under cricoid pressure using 7.5mm ETT. A tilt of 15 degrees to left was given. Anaesthesia was maintained with 50% \textsubscript{N}2O in \textsubscript{O}2 and Sevoflurane (2%) in Closed circuit. A healthy 2.25 Kg female baby was delivered with Apgar scores of 8 (1 min) and 10 (5min).

After clamping of umbilical cord, fentanyl 100mcg, oxytocin 10units, atracurium 30mg was given iv. Infusion of nitroglycerine was kept ready to be used but not required as the BP was around 116/84-130/90 mmHg. Blood loss was approximately 500ml. A total of 1200 ml of Lactated Ringer's soln was given in the perioperative period. To maintain normothermia, all IV fluids were warmed; the temperature of OT was raised to \textdegree\textsubscript{C}. Nasopharyngeal probe was used to record the temperature. Mechanical ventilation was adjusted to keep ETCO\textsubscript{2} around 32-36 mmHg. The procedure lasted for 50 minutes and extubation was achieved without complication. Patient was transferred to PACU. Her vitals were monitored including ABG at regular intervals for next 24 hrs, which did not reveal any abnormality. No post-op deterioration of the neurologic findings noted.

DISCUSSION
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. In our case General anaesthesia was administered because of the potential advantages of decreasing the CMRO\textsubscript{2} which might confer some protection against ischemia. The objective of choosing general anesthesia was to maintain stable brain and systemic hemodynamics. Hypertensive response associated with...
Laryngoscopy and intubation should be adequately controlled with pharmacological means because of the potential for intracerebral hemorrhage. Hypovolemia and Hypotension should similarly be avoided for consequent cerebral ischaemia. Hence utmost care must be taken in using regional anaesthesia in these patients.\textsuperscript{4,5} Normocapnia should be maintained as there is evidence that both hypocapnia and hypercapnia decrease brain blood flow in these patients.\textsuperscript{6} Sevoflurane being a dose dependent cerebral vasodilator was used for anaesthetic maintenance. Initially in low doses it induces indirect vasoconstriction by decreasing metabolic needs and, in higher doses, causes vasodilatation due to its intrinsic effects. Sevoflurane preserves brain physiology better than isoflurane, even in clinically high concentrations. Nitrous oxide may also cause brain vasodilatation.\textsuperscript{7}

Hematocrit values should not be above normal because they may trigger cerebral ischemia, and anemia should be corrected in the preoperative period to maximize oxygen transportation. It has been suggested that hematocrit values should remain between 30\% and 42\% with mild hemodilution which may be beneficial for decreasing blood viscosity.

Normothermia is also recommended because it has been observed that these patients may present with vasospasm in the presence of hypothermia, while hyperthermia may also trigger ischemic events.

Infrared spectroscopy is a noninvasive method for continuous transcranial monitoring of cerebral oxygenation. It should be used in case where risk of cerebral ischemia is suspected. In these situations, bilateral monitoring is recommended.\textsuperscript{1} So transcranial oximetry monitoring may be very useful in patients with MoyaMoya disease. But unfortunately it was not available at our institution.

Although there are some established anaesthetic approaches for moyamoya disease, all co-morbidities should be thoroughly evaluated by the anaesthesiologist and specific perioperative approaches should be determined on a case-by-case basis. In our opinion General Anaesthesia can be a better option in these patients.

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