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Revascularisation Surgeries in Pediatric Patients With Moyamoya Disease: An Anesthesia Point of View (Implications)

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Abstract

Background: Moyamoya disease is a progressive occlusive cereberovascular disorder that usually presents as recurrent strokes in pediatric population. There is paucity of literature on anesthetic management of pediatric patients in the Indian subcontinent. The main objective of our study was to evaluate the perioperative course and outcome of children undergoing revascularization surgery of Moyamoya disease. *Methodology*: A series of 11 patients aged between 5 months-10 years age group were analyzed overa period of one year. *Conclusion* – Anaesthetic management involves maintainence of cerebral blood flow and cerebral perfusion pressure and normothermia to avoid perioperative ischemic complications.

Keywords: Moyamoya disease (MMD), Revascularization, Anaesthetic management, Scalp block, Magnetic Resonance Imaging (MRI).

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Introduction

Moyamoya disease (MMD) is a chronic cerebrovasculopathy of unknown etiology first described in 1957 by Takeuchi and Shimizu, as characterized by progressive stenosis and occlusion at the terminal portion of the internal carotid artery and an abnormal vascular network at the base of the brain in the circle of Willis. To compensate for the blood flow around the occlusive region, a fine vascular network develops that resembles "puffs of smoke", thus, the Japanese term "moyamoya" (Japanese for misty).[1-3] The unique appearance of moyamoya vessels elucidated by Suzuki and Takaku in 1969 triggered international recognition of MMD.[3] Kudo named it officially as the Spontaneous occlusion of the circle of Willis.[4]

The risk factors for peri-operative complications, predominantly the cerebral ischemic events in patients with MMD are: history of transient ischemic attacks, severity of disease, type of revascularization procedure, significant reduction in hematocrit, intraoperative hypotension, intra-operative hypercapnia and reduction in circulating blood volume.[5-7] The prime objective of treatment for MMD is to improve cerebral blood flow; however, medical treatments appear to be

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ineffective in preventing ischemic and hemorrhagic events.[8,9] Surgical revascularization represents an optimal therapeutic option.[10]

Peripheral nerve blockade minimizes the perioperative complications thereby providing an admirable monitoring of neurological status and also maintains haemodynamic stability. In our present study we performed scalp block as an anesthesia management strategy in pediatric moyamoya patients of age above 3 years. We observed improved pain management with decreased postoperative pain, headaches and crying in children using scalp block as an anesthesia implication.

Epidemology

MMD was originally noticed exclusively in East Asia, Nishio (1964) and Nishimoto et al. (1965, 1966) regarded it as a vascular malformation. They considered it an entity peculiar to Japan.[15,16] It is now progressively diagnosed around the world and represents an important source of childhood stroke.[17,18] The diagnosis of MMD is mainly based on angiographic findings and a majority of these cases are reported in Asia.[19] Even in Japan, the overall incidence of MMD remains below 1 per 100,000. Its incidence ranges between 0.086 in USA to 0.54 per 100,000 patients globally.[20,21] 2.2 for blacks and 0.5 for Hispanics as compared with whites.[21] The incidence peaks in two age groups: children who are approximately 5 years of age and adults in their mid-40s.[22] Females are affected nearly twice as often as males.[23] Familial occurrence accounts for about 15% of patients.[24] It accounts for one-fifth of the identifiable cerebral arteriopathies in childhood stroke up to most common cerebrovascular disease in children in East Asia.[25,26] In children, unilateral involvement occurs about 18%[24] and progress to bilateral involvement within 2 years.[27]

Materials and Methods

A retrospective study was undertaken on revascularization surgeries for MMD over a period of one year. Institutional ethical committee approval was obtained for the study. Data collection included clinical presentation, the demographic profile of the patient, diagnosis, surgical procedure, intra-operative course, postoperative outcome at discharge.

Diagnosis

Cerebral angiography remains the gold standard to confirm the diagnosis of MMD Clinical picture includes transient ischemic attacks, slow cognitive decline, headaches, dizziness, seizures, visual impairment, involuntary movements, hemiparesis, monoparesis, sensory impairment or cerebral infarction.[28,29]

Surgical treatment of moyamoya typically uses the external carotid artery (ECA) as a source of new blood flow to the ischemic hemisphere. Two general methods of revascularization are used:

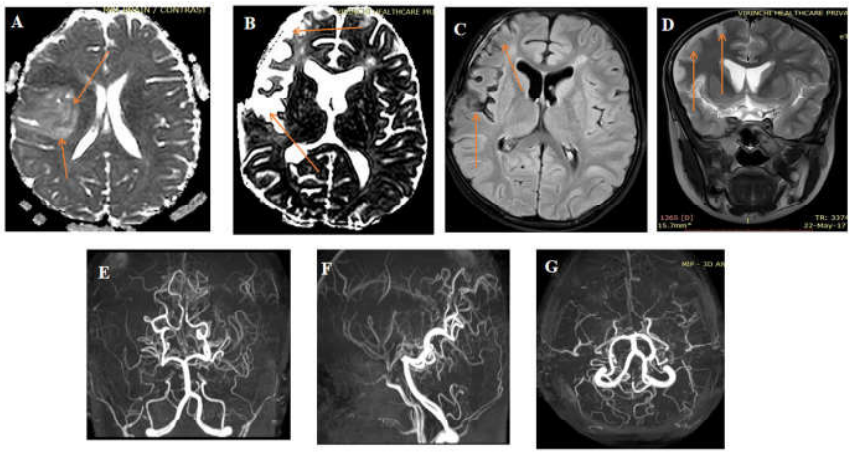

Fig. 1: Radiological assessment for MMD in pediatric patients

(**A)-** Subcaute infarct in right temporal region; (**B) –** Diffusion weighted sequence- showing acute infarcts in left frontal region; **(C)** – FLAIR- LT SIDE- acute infarct in temporal lobe, Right side – subacute infarct in right temporal region; **(D)** – coronal T2W1- infarcts in temporal lobe; **(E)** – MRA-stenosis of Internal carotid artery, Middle cerebral artery and anterior communicating artery. (ICA, MCA, ACA); **(F)** – MRA- puff of smoke appearance; **(G)** – MRA stenosis of MCA, ICA and ACA

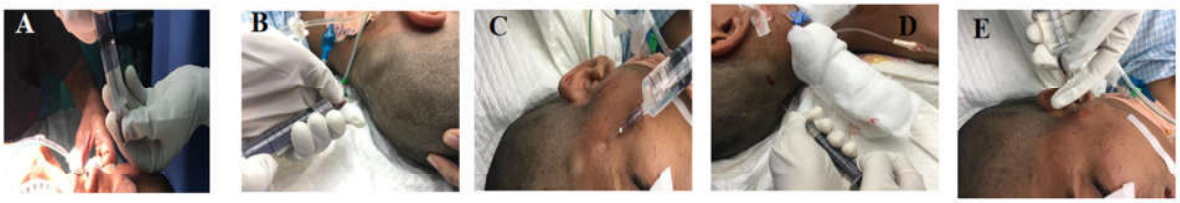

Fig. 2: Scalp block procedures done in pediatric patients for pain management

(A) Supraorbital and supratrochlear nerve block; (**B)** Greater occipital nerve block; (**C)** Zygomaticotemporal nerve block; (**D)** Lesser occipital nerve block; E) Auriculotemporal nerve block.

direct and indirect. In direct revascularization, a branch of the ECA (usually the superficial temporal artery) is directly anastomosed to a cortical artery. Indirect techniques involve the placement of vascularized tissue supplied by the ECA such as dura, temporalis muscle, or the superficial temporal artery (STA) itself in direct contact with the brain, leading to the growth of new blood vessels to the underlying cortex. The direct bypass techniques that have been proposed include STA to the middle cerebral artery (MCA), occipital artery (OA) to MCA, and middle meningeal artery to MCA anastomoses.[30] The indirect techniques include encephalomyosynangiosis(EMS),encephalo duro arteriosynangiosis (EDAS),[31-33] encephalo duro arterio myo synangiosis (EDAMS), encephalo myo arterio synangiosis, multiple cranial bur holes,[33] and omental transposition (Fig. 1).

Use of scalp block in children decreases the adverse effects of opioids (nausea, vomiting, respiratory depression, itching) and postoperative analgesia along with hemodynamic stability, Mean arterial pressure (MAP), and HR responses to skull pin placement and scalp incision in patients undergoing craniotomy. We performed scalp block in patients more than 3 years old with 0.2% ropivacaine after induction with general anesthesia. Patients were more comfortable with stable hemodynamics both intra operative and postoperative. The safe dose of ropivacaine is 2.5-3.0 mg/kg body weight (Fig. 2).

Results

Statistical analysis was done using SPSS version 17. Data are presented as number (%) or mean ± SD or median. We analyzed the records of 11 patients with MMD who underwent re-vascularisation procedures at our center. Out of 11 patients two children underwent indirect revascularization procedures and nine children had direct revascularization procedures (two patients EDAMS, one STA+MCA, eight STA+MCA + EDAMS) during the study period. Four patients had postoperative neurological complications. The demographic details are presented in Table 1 which includes the mean age (4.8 ± 2.88 years), weight (16.8 ± 8.86 kg) and gender (Male: 05, Female: 06) of children who underwent revascularization procedure for MMD respectively. Clinical features include Fever, LOC, headache, seizures, neurologic deficit, ischemic stroke and haemmorhage these are presented in Table 2. Surgical treatment details of patients are presented in Table 3. Post surgical data presented in Table 4 comprises of baseline parameters like haemoglobin, hematocrit , mean intra operative and postoperative factors associated with post-procedure hospital stay, blood transfusions and post-operative hemoglobin and post-operative complications. In our study we have given blood transfusion to 2 children out of 11 children. One of the child was re-operated.

Table 1: Demographic profile

Variables	N = 11 (No. of patients)	MEAN ± S.D, %
Age (years)	Min – 5 months Max – 10 years	4.86 ± 2.88
Weight (kg)	Min -5.8 Max – 34	16.8 ± 8.86
Gender (M/F)	05:06	M- 45.5% F- 54.6%
UL/BL	03:08	UL- 27.3% BL- 72.7%

Table 2: Clinical Features

Surgical Treatment	N= No. of pateints	Percentage (%)
Direct Revascularisation		
STA-MCA	1	9.1
STA-MCA+EDAMS	8	72.7
STA-MCA+EDAS	0	0
Indirect Revascularisation		
EDAMS	2	18.2

Table 3: Surgical Treatments

Surgical Treatment	N= No. of pateints	Percentage (%)
Direct Revascularisation		
STA-MCA	1	9.1
STA-MCA+EDAMS	8	72.7
STA-MCA+EDAS	0	0
Indirect Revascularisation		
EDAMS	2	18.2

Table 4: Post Surgical Data

Baseline parameters, intra operative data, ICU stay, hospital stay	Mean ± SD & %
Hemoglobin (g%)	11.42 ± 0.82
Haematocrit	34.3 ± 2.86
Duration of surgery (hrs)	2.5 ± 0.7
Duration of anaesthesia (hrs)	4.08 ± 0.76
Intraoperative fluids (ml)	576.36 ± 279.25
Urine output (ml)	204.55 ± 156.29
Blood loss (ml)	153.18 ± 127.36
ICU stay (days)	1.09 ± 0.53
Hospital stay (days)	5.36 ± 1.50
Blood transfusion	(n= 4) 36.4%
Post op Hemoglobin (g%)	9.35 ± 1.27
Post op complication	(N= 2)18.2%

Discussion

The prime goal of anesthetic management during revascularization is to maintain a balance between O_2 supply and demand.[35,36] Identification of risk factors for peri-operative complications and outcomes related to the use of anesthesia agents, adequate pain control, increased use of regional anesthesia and better monitoring techniques in providing high quality and safe patient care to patients with MMD is also of significance. CBF is maintained by avoiding hypotension & maintaining normocarbia, appropriate depth of anesthesia and analgesia for minimizing and prevention of increase in $CMRO_2$ associated with pin application, laryngoscopy, intubation and surgical stimulus.

Although no specific anesthesia technique has been precisely shown to decrease perioperative complications in moyamoya patients, several methods for optimizing intraoperative cerebral hemodynamics are commonly used to help minimize this risk. Like, use of Intravenous anesthetics as opposed to inhalation agents are associated with reduced regional CBF in moyamoya patients,[37] which could cause an increased risk for cerebral steal syndrome and perioperative ischemic complications. Maintainence of mean arterial blood pressure about 10% above preoperative baseline throughout the surgical procedure has shown to decrease perioperative complications. In most of the cranial procedures hypocarbia is used to achieve brain relaxation through global cerebral vasoconstriction, this technique is largely opposed as meticulous maintenance of normocarbia throughout the procedure is essential to minimise the risk for ischemic complications. Contrary to most cranial procedures, mannitol is avoided to maintain adequate intravascular volume throughout the procedure.

Most of the children presented with ischemic stroke whereas hemorrhage was rare in our study. Standard monitoring in the form of HR, ECG, NIBP, SpO_2, $ETCO_2$ & temperature was carried out for all patients. Intra-arterial catheters were placed in either a radial artery or dorsalis pedis artery for continuous BP recording. It is extremely important to prevent peri-operative crying. This requires proper pre medication, smooth inhalational or IV induction and good post op pain management.

All children had received IV Fentanyl (2µg/ kg), Paracetamol for postoperative analgesia on a fixed schedule. Older children had received scalp block after induction. In our study, we have used the scalp block as the anesthesia consideration to reduce the perioperative complications post revascularization. Induction was achieved in our study with Sevoflurane in small children. In older children, who had an IV cannula, IV induction with graded doses of Propofol was carried out. Atracurium 0.5mg/kg was given to facilitate tracheal intubation. Anaesthesia was maintained with Sevoflurane and opioids with air and oxygen mixture (50:50). We avoided N_2O as it can cause cerebral vasodilation leading to intra-cerebral steal. Maintenance of normotension is recommended in MMD to prevent ischemic insults. In our centre, balanced anaesthesia technique was used and the main goals was the maintenance of normovolemia by adequate fluid therapy and urinary output. It is imperative to maintain normothermia during re-vascularisation surgery. We used heating blankets to maintain body temperature.

A decreased hematocrit due to anaemia or perioperative blood loss places the MMD patients at risk of cerebral ischemia. Hematocrit 30-42% has been proposed as adequate. In our study, 4 patients had low postoperative Hemoglobin and 2 patients developed new stroke and TIA leading to prolonged duration of hospital stay. Postoperative

ischemic complications were fewer in patients who have combined STA-MCA bypass with EDAMS procedure as compared with indirect bypass. In our study, none of the children developed headaches except one case who showed headache with transient weakness of the eyelids. Some of the children displayed seizures preoperatively. We also observed that the risk of neurological deterioration (weakness of upper and lower limbs, left and right hemeperesis) was higher in patients who underwent indirect vascularisation compared to those of direct anastomatic procedures.

There have been a few studies in which researchers have used a scalp block for a craniotomy. The authors Nguyen A etal., Pinosky ML etal of 2 studies also used the same NB technique as ours but in adults, and they also reported that the block yielded effective analgesia.[38,39]

Conclusion

Moyamoya disease with unknown etiology results in a challenge in determining medical treatment. Re-vascularization surgery remains the only viable option to decrease further ischaemic episodes and neurologic deterioration. Proper pre-operative evaluation is the most effective method to achieve good results. Anaesthetic management of MMD should focus on the maintenance of adequate cerebral blood flow and cerebral perfusion pressure ensuring adequate cerebral oxygenation and to avoid ischemic complications. Scalp block was found to be very effective in reducing postoperative pain and crying in children and it also reduces the risk of ischemic neurological complications there by decrease the hospital stay of the patient.

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