Original Research

ANAESTHETIC MANAGEMENT OF PEDIATRIC PATIENTS WITH MOYAMOYA DISEASE UNDERGOING REVASCULARIZATION SURGERY: A CASE SERIES

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ABSTRACT

Background: Moyamoya disease is a rare, chronic, progressive cerebrovascular disorder characterized by stenosis of the intracranial internal carotid arteries and its branches with development of compensatory fragile collateral vessels.

Aim: To highlight important anaesthetic considerations and perioperative management strategies in patients undergoing Encephaloduroarteriosynangiosis (EDAS PROCEDURE) for Moyamoya disease, with focus on minimizing complications and optimizing patient outcomes.

Methods: In this case series ten paediatric patients aged 3-15 years diagnosed with moyamoya disease who underwent EDAS under general anaesthesia were included. In addition to standard ASA monitoring, invasive arterial pressure monitoring was performed. Ventilatory parameters were adjusted to maintain normocapnia. Data regarding preoperative and intraoperative hemodynamic parameters, duration of surgery and perioperative complications were recorded.

Results: All patients presented with ischemic symptoms. Intraoperative hemodynamic stability was maintained in all patients with episodes of hypertension and tachycardia treated accordingly. Three patients developed transient hypotension after induction, which responded to fluid bolus and vasopressor therapy. The mean duration of surgery was 222.7 ± 26.02 minutes. One patient required postoperative ventilatory support. No patient developed postoperative neurological deficits. The mean ICU stay was 1.6 ± 0.69 days.

Conclusion: Careful maintenance of normotension, normocapnia, normovolemia, and normothermia is essential for safe anaesthetic management in paediatric Moyamoya revascularization surgery.

Keywords: Moyamoya Disease, Anaesthesia, Revascularization Surgery, EDAS Procedure.

INTRODUCTION

Moyamoya is a rare chronic cerebrovascular condition characterized by progressive stenosis of intracranial internal carotid artery (ICA) and its proximal branches, the Anterior and Middle cerebral arteries (ACA and MCA) resulting in increased risk of stroke.

The stenosis of these key vessels leads to development of dilated perforated branches of ICA that provides collateral perfusion to the ischemic brain distal to the occlusion.

The angiographic appearance of these dilated vessels resembles “moyamoya” which means “puff of smoke” in Japanese.

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Moyamoya disease has incidence of 0.35 cases/1,00,000 people globally, with Asian predominance particularly in Japan, Korea, and China. In Japan incidence is 0.54 and prevalence is 6.03 cases/1,00,000 people. This much higher incidence in East Asia is related to genetic factors (notably RNF213 gene mutation). The disease has female predominance with female to male ratio 1.8:1 Age of presentation is typically bimodal in Moyamoya, one is pediatric peak at 5-15 years and other middle age peak at 35-45 years.

Moyamoya syndrome occurs when it is associated with other conditions like sickle cell disease, neurofibromatosis type I, history of cranial irradiation, trisomy 21 and cardiac or renal disease.

Symptoms of the disease are either Ischemic or Hemorrhagic in nature. Clinically ischemic moyamoya presents as Stroke or TIA that reverse within 24 hours. There may be development of dystonia or severe headache, classically look headache behind the eye. Chronic cases also present

with intellectual or motor disability. Children usually present with ischemic symptoms.

Adults more commonly have hemorrhagic presentation due to rupture of fragile collateral vessels or rupture aneurysm, like headache, seizures, monoparesis or hemiparesis, visual impairment or any other.

The diagnosis of Moyamoya disease is established by neuroimaging such as MRI and MRA with six vessels Digital Subtraction Angiography (DSA) as gold standard for diagnosis. DSA also allows staging (Suzuki grading I – IV) of the disease.

There is no proven benefit from medical treatment in case of Moyamoya disease, although antiplatelet therapy is routinely used for preventing stroke in these patients.

The definitive treatment for Moyamoya is revascularization surgery that can be either direct or indirect. In direct revascularization surgery STA - MCA bypass is performed by anastomosing superficial temporal artery to branch of middle cerebral artery. This procedure immediately increases cerebral blood flow and is effective in adults with advanced ischemia.

Though direct revascularization is superior it is technically demanding in children as vessels are too small. Various indirect bypass techniques are described such as Encephalo-duro-arterio-synangiosis (EDAS), Encephalo-mayo-arterio-synangiosis (EMAS), and Encephalo-duro-mayo-arterio-synangiois (EDMAS). In these techniques vascularized tissues (artery, dura and muscle) are placed in contact with the brain surface to stimulate angiogenesis and enhance cerebral perfusion. Most of the data related to moyamoya disease is from East Asian countries like Japan and South Korea, while limited studies have done for Indian scenario. We did a case series of pediatric patients having moyamoya disease undergoing indirect revascularization surgery.

MATERIALS AND METHODS

Ten children of either sex, aged 3 years to 15 years, weighing 10 – 55 kg, ASA physical grade II-IV were operated for surgical revascularization procedure for Moyamoya disease. All patients were admitted a day prior to surgery. IV access and blood drawn for grouping and cross-matching on previous day. Patients were mostly on medications like anticonvulsant, steroid and antiplatelets. A thorough pre-anesthetic checkup was done and investigations including complete blood count, renal and liver function tests, X-ray chest, 2D echo, CT angiography and MRI brain were done for all patients.

On the day of surgery patient counselling was done and written informed consent was obtained from parents. No sedation or premedication was given in the preoperative ward. In the operating room after applying monitors inj. glycopyrrolate (0.004 mg/kg), inj. ondansetron (0.15 mg/kg), inj. fentanyl (1 mcg/kg) were given. Preoxygenation was performed for five minutes. Induction agent propofol (2.5 mg/kg) was used and tracheal intubation was facilitated by IV inj. Succinyl choline (1.5-2 mg/kg) and Intubation was done with proper size cuffed ET tube and confirmed with bilateral air entry. Anesthesia was maintained with oxygen, air (1:1), and sevoflurane (MAC 0.8 to 1), with loading inj. Vecuronium (0.08 mg/kg) and intermittent top-ups of vecuronium. Tidal volume was maintained at 8-10 ml/kg and respiratory rate 12-16 breaths/min to keep end-tidal CO2 at 35-40 mmHg. Foleys catheterization was done to monitor urine output.

Monitoring included standard parameters (ECG, pulse oximetry, non-invasive blood pressure), invasive blood pressure, central venous access in certain cases, urine output and temperature. Fluid management consisted of crystalloids at 4-6 ml/kg/hr, with attention to maintaining urine output at least 2 ml/kg/hr. Reversal was performed with measures to prevent sudden tachycardia or hypertension during extubation. Pain was managed with IV paracetamol and diclofenac suppository.

Duration of surgery, intraoperative blood loss, and perioperative complications like brady/ tachycardia, hypo/hypertension, hypo/hypercapnia, hypo/hyperthermia were recorded.

In this study continuous variables were expressed as mean ± standard deviation, and categorical variables as frequency and percentage. Statistical analysis was performed using Microsoft Excel 2019.

OBSERVATIONS AND RESULTS

Table 1: Demographic data

1	Number of patients		10
2	Age group (3-15 years)	3-5 years	3
		6-10 years	4
		11-15 years	3
3	Weight (kg)	3-5 years	13.33 ± 1.88 kg
		6-10 years	21.75 ± 3.35 kg
		11-15 years	50 ± 4.08 kg
4	Gender	Male	4
		Female	6
5	Procedure	Rt EDAS	5
		Lt EDAS	4

		Rt EDAS + Rt EMAS	1

Ten children underwent indirect revascularization procedures during study period. The demographic details are presented in Table 1. Preoperatively the systolic BP under 150 mm hg and diastolic blood pressure under 90 mm hg was maintained. Intraoperative BP was closely monitored through invasive arterial monitoring and SBP and MAP was maintained as baseline ± 20 mm hg. Three patients developed transient hypotension after induction of which two responded with bolus IV fluid and one patient required IV me phentermine single dose.

All patients were fasted for at least 8 hrs before surgery. Hydration through NBM hours was assured with average rate of fluid administration 4-6 ml/kg/hr to maintaining urine output of at least 2 ml/kg/hr. Average blood loss was around 100 ml, only one patient required intra op blood transfusion.

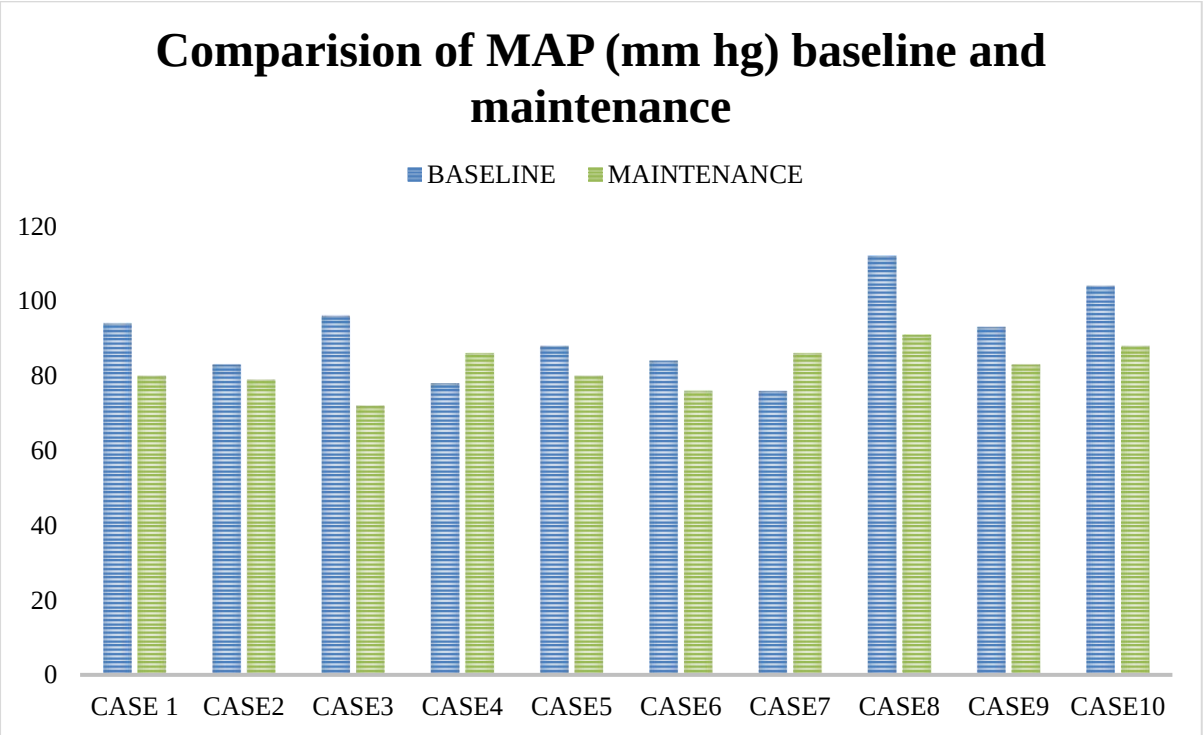
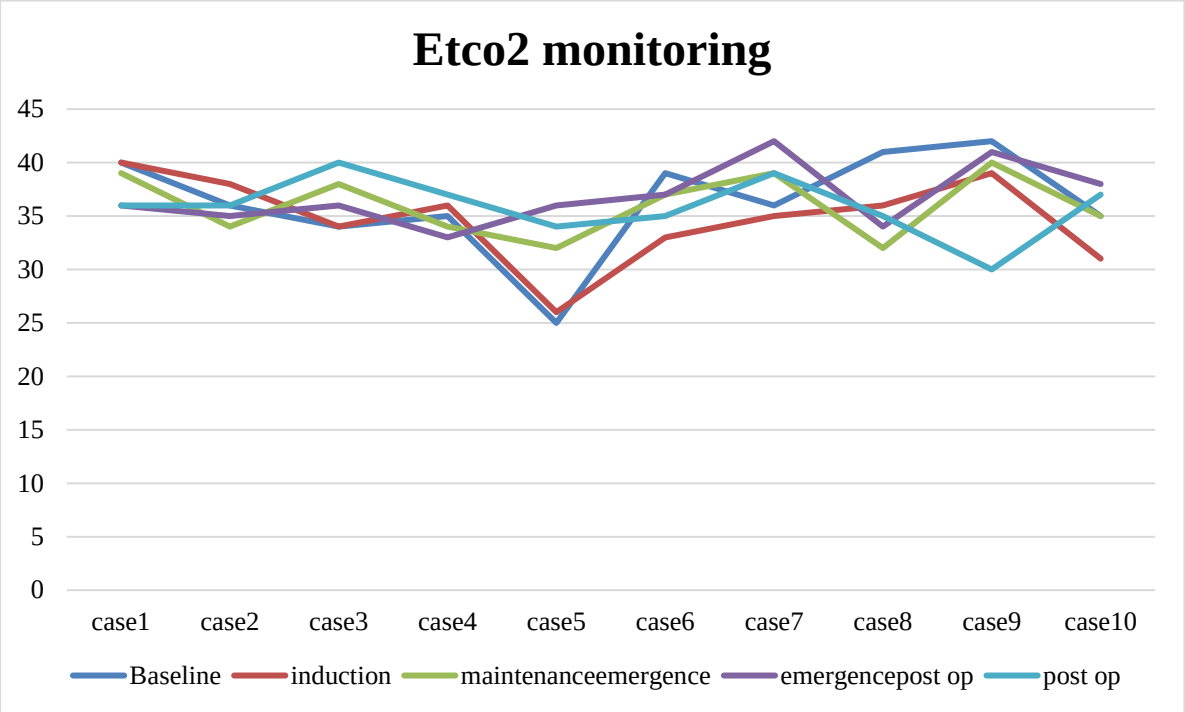


Table 2: Clinical Data Analysis

No.	Variable	Result
1	Preoperative hemoglobin (g/dL)	11.84 ± 1.06
2	Duration of surgery (mins)	222.7 ± 26.02
3	Intravenous fluids (ml)	701 ± 411
4	Urine output (ml)	416 ± 202
5	Blood loss (ml)	109.5 ± 52
6	Patient requiring blood transfusion	1
7	Post operatively ventilated	1
8	Post-operative ICU stay in days	1.6 ± 0.69
9	Discharge from hospital (days)	4.1 ± 0.99

Tidal volume and rate of respiration were set to according to age and weight of patients with target EtCO2 around 35-40 mm Hg, avoiding fluctuations.



Average duration of surgery was 4-5 hours with duration of ICU stay 1-2 days. One Patient was ventilated postoperatively for 1 day; others were extubated on table. Discharge from hospital was within 4-5 days postoperatively.

DISCUSSION

The goal of this case series was to evaluate effectiveness of balanced anesthesia in minimizing perioperative hemodynamic instability in surgery for Moyamoya disease having compromised cerebral perfusion. Moyamoya patients have reduced cerebral perfusion and rely heavily on pressure dependent flow. Previous studies have demonstrated that patients with Moyamoya disease have compromised cerebrovascular reserve and impaired cerebral autoregulation. Iwama et al. reported that perioperative hypotension is a major risk factor for ischemic complications in pediatric Moyamoya patients. Therefore, maintaining stable hemodynamics is a key component of anesthetic management. (6)

In pediatric patients' cerebral metabolic rate of oxygen consumption (CMRO2) is higher than adults. So Moyamoya children have significant mismatching of CBF and CMRO2. Anesthesiologists must pay attention to maintaining normovolemia, normotension, normoxia, normocapnia and normothermia, as abnormalities of these physiological parameters have shown to be associated with development of complications. (7, 9)

In our study all ten patients received IV anesthetic induction and maintenance with sevoflurane MAC< 1. Choice of anesthetic technique remains controversial in Moyamoya surgery. Sato et al. demonstrated that inhalational anesthesia may reduce cortical blood flow in patients with Moyamoya disease, suggesting a potential advantage of total intravenous anesthesia (TIVA) (5). Similarly, Kikuta et al. reported that propofol anesthesia was associated with lower intracranial pressure and improved cortical blood flow compared with sevoflurane anesthesia. (4)

We gave balanced anesthesia using opioid, propofol and low dose sevoflurane, which helped to maintain stable hemodynamics.

Carbon dioxide levels play a crucial role in regulating cerebral blood flow. Both hypocapnia and hypercapnia can adversely affect cerebral perfusion in Moyamoya patients. Hypocapnia causes cerebral vasoconstriction, which may reduce perfusion to already compromised brain regions. Conversely, hypercapnia can lead to vasodilation of normal cerebral vessels, producing a "steal phenomenon" in which blood is diverted away from ischemic territories.

Hydration during NBM hours and intraoperative period was maintained by giving IV crystalloids guided through urine output (> 2ml/kg/hr). Gardner Yelton SE *et al.* suggested to maintain hyper hydration with 1.5× maintenance IV fluids for minimum 12 h before anesthesia. (9)

Pain and crying cause hyperventilation and hypercarbia that causes constriction of cerebral blood vessels, causing relative blood flow reduction in moyamoya vessels, occasionally precipitating TIAs in children. (3)

We gave multimodal analgesia (short acting opioid+ paracetamol+ NSAID) to achieve pain relief without causing extra sedation.

Cerebral autoregulatory response to hypotension was substantially diminished in children, so hypotension is to be avoided. (6)

Hypertension can cause significant mismatch in CBF and CMRO2, therefore a deep plane of anesthesia should be provided that will decrease the relatively high CMRO2 in children while maintaining adequate CBF.

Hypercarbia due to excessive sedation causes cerebral vasodilation, in which blood preferentially flows through low-resistance normal vessels instead of moyamoya vessels, which worsens mismatch. This *cerebral steal* phenomenon divert blood away from ischemic territories and precipitate complications. (2)

Significant decreases in perfusion have been observed both during hypocapnia and hypercapnia in patients with moyamoya disease using laser doppler flowmetry. (10)

Thus careful perioperative anesthetic management with control of physiological parameters is necessary for favorable outcomes in pediatric patients with Moyamoya disease.

Summary of management

- Normotension
  - Maintain MAP within 20% baseline; treat drops promptly with fluids/ vasopressors
  - Hypotension ->inadequate CPP -> ischemic stroke.
  - Hypertension->risk of hemorrhage
- Normocapnia
  - Maintain Etco2 around 35-40 mm Hg
  - Hypocapnia -> cerebral vasoconstriction -> ischemia
  - Hypercapnia ->cerebral vasodilation in normal vessels -> “steal phenomenon” -> ischemia in diseased territories
- Normothermia
  - Maintain core temperature 36-37°C
  - Hyperthermia -> increases cerebral metabolic rate of oxygen ($CMRO_2$) ->demand-supply mismatch
  - Hypothermia -> increases blood viscosity -> worsens ischemia
- Normovolemia
  - Maintain euvolemia avoiding both dehydration and fluid overload
  - Hypovolemia reduces cerebral perfusion pressure (CPP)
  - Hypervolemia may increase intracranial pressure (ICP)

Limitations

First, we had small sample size as Moyamoya disease is a rare condition and only ten paediatric patients undergoing indirect revascularization surgery were included. Second, this was a single-center observational case series without a control group, which limits the ability to draw definitive conclusions regarding the superiority of a particular anaesthetic technique. Third, long-term neurological outcomes were not evaluated.

CONCLUSION

Indirect revascularization relies on long term collateral development in Moyamoya patients. Disciplined perioperative anaesthetic management minimizes ischemic and neurological complications.

Conflict of interest

None.

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