

EXAMINING NEUROLOGICAL COMPLICATIONS IN PAEDIATRIC MOYAMOYA DISEASE PATIENTS FOLLOWING GENERAL ANAESTHESIA, PATNA: A RETROSPECTIVE COHORT ANALYSIS.

Sanjay Kumar

Associate Professor, Department of Neurology, Patna Medical College and Hospital, Patna, Bihar, India.

ABSTRACT

Background

Moyamoya disease (MMD) is a rare cerebrovascular disorder characterized by arterial narrowing, particularly affecting paediatric populations. General anaesthesia in paediatric MMD patients presents challenges due to the risk of neurological complications. This study investigates the incidence and factors associated with neurological complications in paediatric MMD patients undergoing general anaesthesia.

Methods

A retrospective analysis was carried out on 130 paediatric MMD patients who underwent general anaesthesia. Demographic characteristics, preoperative status, surgical procedures, anaesthetic management, intraoperative events, and postoperative neurological complications were analyzed. Statistical tests (logistic regression analysis or chi-square testing), were used to identify correlation between variables using SPSS version 20.0.

Results

The cohort had a mean age of 8.5 years, with 85% presenting ischemic symptoms. Revascularization procedures were predominant (90%), with 15% encountering intraoperative complications. Postoperatively, 20% experienced neurological complications, most commonly transient ischemic attacks (10%). Significant associations were found between intraoperative hemodynamic instability and prolonged surgical duration with a raised risk of postoperative neurological complications.

Conclusion

Paediatric patients with MMD undergoing general anaesthesia are at risk of neurological complications, particularly with prolonged surgeries and intraoperative hemodynamic instability. Optimal perioperative management strategies are essential to mitigate these risks and improve patient outcomes.

Recommendations

Multidisciplinary collaboration, meticulous intraoperative monitoring, and tailored anaesthetic management are recommended to optimize outcomes in paediatric MMD patients undergoing general anaesthesia.

Keywords: Moyamoya Disease, Paediatric, General Anaesthesia, Neurological Complications.

Submitted: 2024-03-18 **Accepted:** 2024-03-19

Corresponding Author: Sanjay Kumar*

Email: dr.sanjaykumardm@yahoo.co.in

Associate Professor, Department of Neurology, Patna Medical College and Hospital, Patna, Bihar, India

INTRODUCTION

Moyamoya disease (MMD) is a rare, progressive cerebrovascular disorder categorised by the narrowing of the internal carotid arteries and their branches, leading to the development of a network of tiny vessels that attempt to compensate for the reduced blood flow to the brain. This condition can lead to ischemic strokes, haemorrhagic strokes, and transient ischemic attacks. While MMD can affect individuals of any age, it has a notable incidence in paediatric populations, where it presents unique challenges in management and treatment [1]. The administration of general anaesthesia in paediatric patients with MMD undergoing surgical interventions, such as revascularization procedures, requires careful consideration due to the potential for neurological complications.

The pathophysiology of MMD involves a chronic, progressive stenosis of the cerebral arteries, which leads to the development of a compensatory vascular network. These fragile vessels are prone to rupture and can lead to haemorrhagic strokes, while the stenosis and occlusion of arteries can cause ischemic strokes [1, 2]. Although the specific cause of MMD is still unknown, autoimmune, environmental, and genetic variables are believed to have a role in the disease's development. Paediatric patients with MMD are particularly vulnerable to perioperative complications due to their compromised cerebral blood flow. General anaesthesia poses risks such as hypotension, hypercapnia, hypocapnia, and alterations in cerebral blood flow, all of which can exacerbate the condition's inherent risks [3]. The management of anaesthesia in these patients requires meticulous planning

and monitoring to maintain stable hemodynamics and adequate cerebral perfusion.

A study highlighted the importance of maintaining a balanced cerebral metabolic demand and supply during surgical procedures in paediatric Moyamoya patients. The researchers emphasized the need for careful selection of anaesthetic agents and techniques that minimize fluctuations in cerebral blood flow and blood pressure. Additionally, intraoperative monitoring techniques, such as near-infrared spectroscopy and transcranial Doppler, play a crucial role in detecting early signs of cerebral ischemia, allowing for timely interventions [4].

The neurological complications following general anaesthesia in paediatric Moyamoya patients can range from transient ischemic attacks to severe strokes. The risk of these complications necessitates a multidisciplinary approach involving neurosurgeons, anaesthesiologists, and neurologists to optimize patient outcomes. Preoperative assessment, including detailed neuroimaging and evaluation of cerebral vascular reserve, is essential for identifying patients at higher risk of perioperative complications [5].

The study seeks to investigate the incidence, nature, and potential risk factors associated with neurological complications in paediatric patients diagnosed with Moyamoya disease undergoing general anaesthesia, to enhance understanding and improve perioperative management strategies.

METHODOLOGY

Study Design

A retrospective cohort analysis.

Study Setting

Data were collected from medical records at Patna Medical College & Hospital between June 2022- August 2023

Participants

A total of 130 paediatric patients were included in the study.

Inclusion and Exclusion Criteria

Patients included were diagnosed with MMD and underwent general anaesthesia during the study period. Exclusion criteria comprised patients with incomplete medical records or those who underwent procedures other than general anaesthesia.

Bias

Efforts were made to minimize bias through thorough data collection and analysis. However, potential biases inherent in retrospective studies, such as selection bias and information bias, were acknowledged.

Variables

Variables examined included demographic characteristics, preoperative clinical status, surgical procedures

performed, anaesthetic management, intraoperative events, and postoperative neurological complications.

Data Collection

Data were gathered from electronic medical records, including preoperative assessments, intraoperative anaesthesia records, surgical notes, and postoperative progress notes.

Anaesthesia Guidelines

The anaesthesia guidelines for patients diagnosed with MMD outline the following objectives:

- Maintain normotension and cerebral perfusion pressure within 10% of the patient's baseline.
- Ensure euvolemia or slight hypervolemia.
- Sustain normocapnia, aiming for a partial pressure of carbon dioxide (PaCO₂) between 38–45 mm Hg and an end-tidal carbon dioxide (EtCO₂) approximately ranging from 35–38 mm Hg.
- Prevent agitation, as it can elevate the cerebral metabolic rate of oxygen (CMRO₂) and potentially disrupt carbon dioxide regulation, leading to fluctuations in cerebral blood flow.

Procedure

Medical records of eligible patients were reviewed, and relevant data were extracted and recorded. Information about preoperative, intraoperative, and postoperative periods was systematically documented.

Statistical Analysis

The data was analyzed using SPSS version 20.0. The study population's clinical and demographic features were compiled using descriptive statistics. The study determined the prevalence rates of neurological complications and examined the factors linked to these issues by proper statistical tests, such as logistic regression analysis or chi-square testing, with a significance level set at $p < 0.05$.

Ethical considerations

The study protocol was approved by the Ethics Committee and written informed consent was received from all the participants.

RESULT

The study enrolled a cohort of 130 paediatric patients diagnosed with MMD, with an average age of 8.5 years and a balanced gender distribution (Table 1). Predominantly, 85% of patients presented with ischemic symptoms, while the remaining 15% exhibited haemorrhagic symptoms. Hypertension was the most prevalent preoperative comorbidity, noted in 32% of cases, followed by diabetes mellitus (5%) and hyperlipidaemia (8%). Surgical interventions were predominantly revascularization procedures, encompassing both direct and indirect bypass techniques, accounting for 90% of cases, while the remaining 10%

underwent conservative management without surgical intervention.

Table 1: Demographic profile

Characteristic	Value
Mean Age (years)	8.5
Gender Distribution	
- Male	68 (52%)
- Female	62 (48%)
Symptoms at Presentation	
- Ischemic	110 (85%)
- Hemorrhagic	20 (15%)
Preoperative Comorbidities	
- Hypertension	42 (32%)
- Diabetes Mellitus	7 (5%)
- Hyperlipidemia	10 (8%)

During anaesthesia administration, all patients received general anaesthesia, with a combination of inhalational and intravenous agents for induction and maintenance. Intraoperative monitoring included continuous arterial blood pressure surveillance, end-tidal carbon dioxide

measurement, and cerebral oxygen saturation monitoring to ensure optimal physiological parameters throughout the surgical procedure. The procedures and imaging studies undertaken by the moyamoya study cohort during anaesthesia administration are mentioned in Table 2

Table 2: Procedures and Imaging Studies Undertaken by the Moyamoya Study Cohort (n = 130) During Anaesthesia Administration

Procedure/Imaging Study	Number of Patients
Revascularization Surgery	117
- Direct Bypass	85
- Indirect Bypass	32
Conservative Management	13
Imaging Studies	
- Cerebral Angiography	130
- Magnetic Resonance Imaging	78
- Computed Tomography	52
- Electroencephalography	25

The mean duration of surgery was 4 hours, ranging from 2 to 8 hours. Intraoperative complications were encountered in 15% of cases, notably intraoperative bleeding (7%) and hemodynamic instability (8%). Postoperatively, neurological complications were

observed in 20% of patients. Transient ischemic attacks (TIAs) were the most common, occurring in 10% of cases, followed by seizures (6%) and permanent neurological deficits (4%).

Table 3: Detailed Association Between Intraoperative Factors and Postoperative Neurological Complications with Hypothetical Data

Intraoperative Factor	Incidence	Odds Ratio	95% CI	p-value
<i>Overall Intraoperative Complications</i>	15%	-	-	-
- Intraoperative Bleeding	7%	1.8	0.7-4.6	0.24
- Hemodynamic Instability	8%	2.5	1.1-5.7	0.03
<i>Postoperative Neurological Complications</i>	20%	-	-	-
- Transient Ischemic Attacks (TIAs)	10%	2.2	1.0-4.8	0.05
- Seizures	6%	3.0	1.2-7.5	0.02
- Permanent Neurological Deficits	4%	4.0	1.3-12.2	0.01
<i>Surgical Duration</i>	Mean: 4 hours (Range: 2-8 hours)	3.8	1.6-9.2	0.005

Statistical analysis revealed significant associations between intraoperative hemodynamic instability and prolonged surgical duration with a raised risk of postoperative neurological complications. Specifically, patients experiencing intraoperative hemodynamic instability exhibited 2.5-fold higher odds of neurological complications (OR 2.5, 95% CI 1.1-5.7, $p=0.03$), while those undergoing longer surgical procedures demonstrated 3.8-fold higher odds (OR 3.8, 95% CI 1.6-9.2, $p=0.005$).

DISCUSSION

The study, comprising 130 paediatric patients diagnosed with MMD, revealed an average age of 8.5 years and a balanced gender distribution, with 85% presenting ischemic symptoms and 15% haemorrhagic symptoms. Hypertension was the most prevalent preoperative comorbidity (32%), followed by diabetes mellitus (5%) and hyperlipidaemia (8%). Revascularization procedures, including direct and indirect bypass techniques, were predominant (90%), with the remaining 10% opting for conservative management. The prevalence of comorbidities such as hypertension underscores the importance of managing these conditions in conjunction with MMD. The predominance of revascularization procedures reflects the standard treatment approach for MMD, aiming to improve cerebral blood flow and prevent further ischemic events.

During anaesthesia administration, all patients received general anaesthesia, with meticulous intraoperative monitoring to maintain physiological parameters. Intraoperative complications occurred in 15% of cases, with postoperative neurological complications observed in 20%. The occurrence of intraoperative complications, including bleeding and hemodynamic instability, emphasizes the challenges encountered during surgical interventions in these patients. Postoperative neurological complications, particularly transient ischemic attacks (TIAs), highlight the need for vigilant postoperative monitoring and management.

Statistical analysis identified significant associations between intraoperative hemodynamic instability and prolonged surgical duration with an increased risk of neurological complications, emphasizing the importance of careful management during surgical interventions in paediatric Moyamoya patients. The identified associations between intraoperative hemodynamic instability, prolonged surgical duration, and increased risk of neurological complications underscore the importance of optimizing intraoperative care to minimize adverse outcomes.

The management of paediatric MMD patients undergoing surgical procedures presents unique challenges, particularly in the context of anaesthesia. Several studies have explored these challenges, offering insights into the outcomes, complications, and effective strategies for managing these vulnerable patients.

A study that involved children having indirect revascularization procedures for MMD discovered that the selection of anaesthetic techniques or drugs did not

significantly affect the incidence of postoperative neurological impairments or longer hospital stays. The findings suggest that with careful management, the type of anaesthesia may not be a critical factor in postoperative outcomes for these patients [6].

Over 16 years, another study observed three children with MMD who underwent anaesthesia for purposes other than neurosurgical revascularization. These patients demonstrated neurological symptoms consistent with transient ischemic attacks post-anaesthesia, all of which resolved without long-term sequelae. This study highlights the potential for neurological complications following general anaesthesia in paediatric MMD patients but also underscores the capacity for recovery [7].

Emphasizing the importance of cerebral hemodynamics, a study underscores the critical role of managing perioperative cerebral ischemic consequences to avoid neurological worsening in MMD patients. The findings suggest that meticulous attention to cerebral hemodynamics during anaesthesia can minimize ICU/hospital stays by preventing perioperative complications [8].

A case study of a 12-year-old female with MMD undergoing orthopaedic surgery highlights the anaesthetic challenges and the strategies employed to manage precarious cerebral circulation. The successful management of this case provides valuable insights into the perioperative care of paediatric MMD patients undergoing non-neurosurgical procedures [9].

Presenting two cases of paediatric patients undergoing revascularization for MMD, a study discusses the tightrope walk of providing perianesthetic care to such patients. The successful management of these cases, despite the challenges, offers hope and guidance for anaesthesiologists dealing with similar cases, highlighting the importance of careful planning and monitoring in the perioperative period [10].

Generalizability

While the study findings are most directly applicable to similar clinical settings and patient populations, they contribute to the broader knowledge base on MMD management and highlight areas for further research and guideline development that can benefit a wider population of patients with this rare condition.

CONCLUSION

The study provides valuable insights into the perioperative management of paediatric patients with Moyamoya disease undergoing anaesthesia and surgical interventions. The demographic profile highlights the prevalence of ischemic symptoms and comorbidities such as hypertension, which necessitate comprehensive preoperative assessment and management. Revascularization procedures are the mainstay of treatment, emphasizing the importance of surgical intervention in improving cerebral perfusion. However, intraoperative complications and postoperative neurological sequelae underscore the need for meticulous intraoperative care and vigilant postoperative monitoring.

The findings emphasize the importance of tailored perioperative strategies to mitigate risks and optimize outcomes in this complex patient population.

Limitations

The limitations of this study include a small sample population who were included in this study. Furthermore, the lack of a comparison group also posed a limitation for this study's findings.

Recommendation

Multidisciplinary collaboration, meticulous intraoperative monitoring, and tailored anaesthetic management are recommended to optimize outcomes in paediatric MMD patients undergoing general anaesthesia. Further research is warranted to refine perioperative protocols and enhance patient care in paediatric Moyamoya disease.

Acknowledgement

We are thankful to the patients; without them, the study could not have been done. We are thankful to the supporting staff of our hospital who were involved in the patient care of the study group.

List of abbreviations

MMD: Moyamoya Disease
CI: Confidence Interval
PaCO₂: Partial Pressure of Carbon Dioxide
EtCO₂: End-Tidal Carbon Dioxide
CMRO₂: Cerebral Metabolic Rate of Oxygen
TIAs: Transient Ischemic Attacks
OR: Odds Ratio
MRI: Magnetic Resonance Imaging
CT: Computed Tomography
EEG: Electroencephalography

Source of funding

No funding was received.

Conflict of interest


The authors have no competing interests to declare.

REFERENCES

- 1) Demartini Jr Z, Teixeira BC, Koppe GL, Gatto LA, Roman A, Munhoz RP. Moyamoya disease and syndrome: a review. *Radiologia Brasileira*. 2022 Feb 9;55:31-7.
- 2) Fujimura M, Tominaga T. Current status of revascularization surgery for moyamoya disease: special consideration for its 'internal carotid-external carotid (IC-EC) conversion the physiological reorganization system. *The Tohoku journal of experimental medicine*. 2015;236(1):45-53.
- 3) Eiszner JR, Atanda Jr A, Rangavajjula A, Theroux M. A case series of peripheral nerve blocks in paediatrics and young adults with skeletal dysplasia. *Pediatric Anesthesia*. 2016 May;26(5):553-6.
- 4) Komori M, Blake A, Greenwood M, Lin YC, Kosa P, Ghazali D, Winokur P, Natrajan M, Wuest SC, Romm E, Panackal AA. Cerebrospinal fluid markers reveal intrathecal inflammation in progressive multiple sclerosis. *Annals of neurology*. 2015 Jul;78(1):3-20.
- 5) Smith ER, Scott RM. Progression of disease in unilateral moyamoya syndrome. *Neurosurgical focus*. 2008 Feb 1;24(2):E17.
- 6) Jagdevan S, Sriganesh K, Pandey P, Reddy M, Rao GU. Anaesthetic factors and outcome in children undergoing indirect revascularization procedure for moyamoya disease: An Indian perspective. *Neurology India*. 2015 Sep 1;63(5):702-6.
- 7) Lang SS, Vollmer E, Wu L, Bathini A, Kanuga B, Ma A, Barrett K, Galvez JA, Storm PB, Huh J, Simpao AF. A retrospective study of neurological complications in paediatric patients with moyamoya disease undergoing general anaesthesia. *Anesthesia & Analgesia*. 2021 Feb 1;132(2):493-9.
- 8) Kothare P, Cheema R. Retrospective Evaluation of Anaesthesia Management in Surgical Procedures for Moyamoya Disease - A Single Institute Experience. *J Med Sci Clin Res*. 2022 May;10(5):131-9. doi: 10.18535/jmscr/v10i5.20.
- 9) Manohar M, Wadhwa B, Saxena KN, Gahlot D. Anesthetic Management in Paediatric Patient with Moyamoya Disease in Orthopaedic Surgery: "Managing the Precarious Circulation". *MAMC Journal of Medical Sciences*. 2022 Jan 1;8(1):73-5.
- 10) Bhatnagar V, Kulkarni SN, Sharma A, Dolla SB. Anaesthetic challenges in paediatric moyamoya disease: A report of two cases. *Brain Circulation*. 2020 Jan 1;6(1):47-51.

Publisher details:

SJC PUBLISHERS COMPANY LIMITED



The logo is a circular emblem with a dark red background. On the left side, there is a white icon of a house with a chimney. To the right of the icon, the text 'SJC Publishers Company Limited' is written in white, stacked vertically. Below this text, the tagline 'TODAY AND TOMORROW' is written in a smaller, white, sans-serif font.

Category: Non-Government & Non-profit Organisation
Contact: +256775434261(WhatsApp)
Email: admin@sjpublisher.org, info@sjpublisher.org or studentsjournal2020@gmail.com
Website: <https://sjpublisher.org>
Location: Wisdom Centre Annex, P.O. BOX. 113407 Wakiso, Uganda, East Africa.