

Review Article

Indirect revascularization for moyamoya disease: A pediatric neuroanesthesiology perspective

Karla E. K. Wyatt, Sandi K. Lam¹, Nisha Gadgil¹Department of Anesthesiology, Baylor College of Medicine/Texas Children's Hospital, ¹Department of Neurosurgery, Division of Pediatric Neurosurgery, Baylor College of Medicine/Texas Children's Hospital, Houston, TX 77030, USAE-mail: *Karla E. K. Wyatt - karla.wyatt@bcm.edu; Sandi K. Lam - sklam@texaschildrens.org; Nisha Gadgil - ngadgil@bcm.edu

*Corresponding author

Received: 21 May 18 Accepted: 03 August 18 Published: 02 November 18

Key Words: Anesthetic management, anesthetic monitoring, cerebral ischemia, cerebral perfusion pressure, moyamoya, neuroprotective

ILLUSTRATIVE CASE

The patient is a 4-year-old male with no significant prior medical history who presented with acute onset of right arm and leg weakness. Neurological examination was significant for 4/5 strength in the right arm and 2/5 strength in the right leg. MRI of the brain was significant for acute cortical infarct of the left frontal lobe and FLAIR signal change of the deep frontal and periventricular white matter consistent remote ischemic insult [Figure 1a and b]. Cerebral angiogram demonstrated complete occlusion of the left distal internal carotid artery, dilated basal collateral "moyamoya" vessels, and no significant external carotid transdural collaterals (Suzuki grade 3) [Figure 2]. In preparation for revascularization surgery, the patient was pre-medicated with intravenous dexmedetomidine 0.5 µg/kg, for anxiolysis prior to parental separation. Subsequent to operating room transfer, standard monitors were applied. Intravenous induction with fentanyl 2 µg/kg, lidocaine 2 mg/kg, propofol 2 mg/kg, and rocuronium 0.1 mg/kg was administered to quickly induce unconsciousness, maintain hemodynamic stability, and blunt the sympathetic response to endotracheal intubation. The patient's vital signs remained near his baseline blood pressure of 90/52 mm Hg, 80 beats per minute (bpm), and pulse oximetry of 99%. Following intubation, the patient was placed on mechanical ventilation, pressure control mode, to achieve an end-tidal CO₂ of 35–40 mm Hg, with tidal volumes of 5–7 ml/kg. Anesthesia was maintained with sevoflurane at 0.9 MAC and a dexmedetomidine infusion at 0.2 µg/kg/h. A right radial arterial line was placed to maintain

a blood pressure range within 10% of his baseline, and to monitor acid base status and hematocrit. Intravascular volume was supported with an electrolyte-balanced crystalloid throughout the case. The patient underwent left sided cerebral revascularization employing encephaloduroarteriosynangiosis and dural inversion without complications. During the revascularization, the patient experienced a brief episode of hypotension that resolved with a small dose of phenylephrine 0.5 µg/kg and a crystalloid fluid bolus of 10 ml/kg. Following the reversal of neuromuscular blockade, the patient had an uneventful emergence from anesthesia. The dexmedetomidine infusion was continued during

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Quick Response Code:	Website: www.surgicalneurologyint.com
	DOI: 10.4103/sni.sni_151_18

How to cite this article: Wyatt KE, Lam SK, Gadgil N. Indirect revascularization for moyamoya disease: A pediatric neuroanesthesiology perspective. *Surg Neurol Int* 2018;9:224.

<http://surgicalneurologyint.com/Indirect-revascularization-for-moyamoya-disease-A-pediatric-neuroanesthesiology-perspective/>

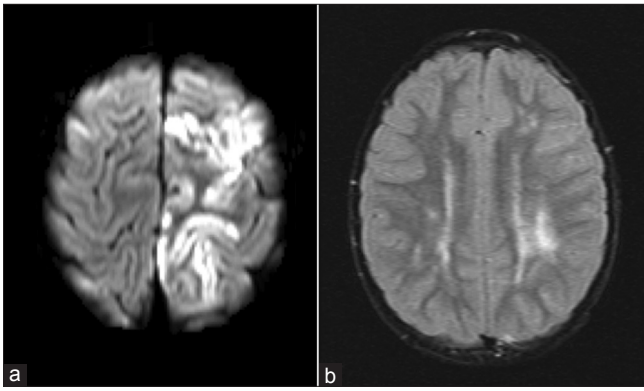


Figure 1: (a) Diffusion weighted magnetic resonance imaging demonstrating diffusion restriction of the left posterior frontal lobe suggesting acute infarct. (b) T2-weighted FLAIR magnetic resonance image demonstrating signal change in the left periventricular subcortical white matter suggestive of remote ischemia

emergence to blunt the sympathetic and hyper-excitatory state associated with the stages of anesthesia, extubation, and post-operative pain. The patient's post-operative course was uneventful, and he was discharged home with outpatient rehabilitation therapy. Repeat cerebral angiogram performed 6 month post-operatively demonstrated extensive new collaterals (arrows) from the left middle meningeal and superficial temporal arteries providing excellent revascularization of the left middle cerebral artery territory (Matsushima grade A) [Figure 3]. The patient has been followed for 6 year post-operatively and has had no recurrent ischemic events; his right sided hemiparesis has resolved, and his neurological exam at last follow-up was unremarkable.

BACKGROUND

Moyamoya disease, a name coined by Suzuki and Takeuchi,^[10,12] which in Japanese means “something hazy, like a puff of cigarette smoke drifting in the air,” is a progressive cerebrovascular disorder often mimicking this appearance on cerebral angiogram.^[11] The pathogenesis remains unknown; however, genetic, infectious, and inflammatory processes have been implicated in the etiology. Four major types of moyamoya disease have been classified in the literature: ischemic, hemorrhagic, epileptic, and “other.”^[12] Escalating narrowing and proliferation within the distal internal carotid, proximal anterior cerebral, and/or proximal posterior cerebral arteries along with tributaries leads to a network of collateral vessels. This collateral vasculature maximally dilates to accommodate cerebral blood flow, albeit patients with moyamoya have an overall reduction in cerebral blood flow when compared to healthy individuals.^[9] Fluctuations in arterial gas tension and mean arterial pressure can lead to decreases in cerebral oxygen delivery. Symptomatology includes



Figure 2: Preoperative left internal carotid angiogram, anteroposterior projection demonstrates severe stenosis of the distal left internal carotid artery and extensive basilar collateralization

transient ischemic attacks (TIAs), recurrent ischemic and/or hemorrhagic strokes, headache, and/or seizures. Moyamoya ultimately progresses to partial or complete unilateral or bilateral occlusion of the internal carotid artery. Histopathology of vasculature in patients with moyamoya disease demonstrates an eccentric fibrous intimal thickening, hyperplasia of the smooth muscle cells, and lipid proliferation, without evidence of inflammation or atheromatous changes. This disease is rare in the pediatric population. In children, moyamoya disease typically manifests with an ischemic stroke or TIA, unlike adults who commonly present with an intracranial hemorrhage.^[5,6,9,11] When moyamoya disease is associated with sickle cell disease, neurofibromatosis, Down's syndrome, and other underlying comorbidities, it is assigned the secondary classification of moyamoya syndrome.^[8]

EPIDEMIOLOGY

Moyamoya disease has a known ethnic and genetic predisposition. The prevalence of moyamoya is highest among Asian countries as compared with the rest of the world. Japan has an estimated incidence of 0.35 to 0.94 per 100,000 people, with a female-to-male ratio of 2:1.^[5] In the United States, the overall incidence is 0.086 per 100,000 people, with the highest ethnicity-distinct incidence seen among Asian Americans, followed by African Americans and Hispanics.

CLINICAL PRESENTATION, WORKUP, AND DIAGNOSIS

Symptomatology suggestive of moyamoya begins with head computed tomography scan and magnetic resonance imaging (MRI)/magnetic resonance angiography (MRA)

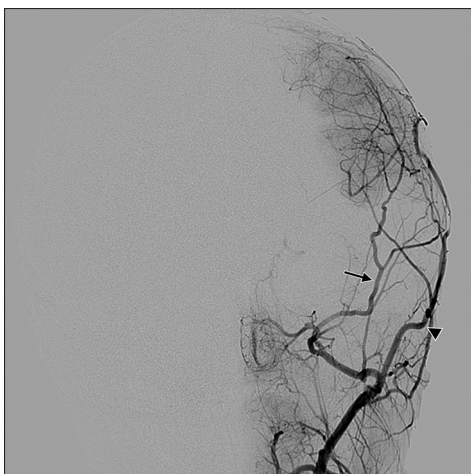


Figure 3: Postoperative left external carotid angiogram, anteroposterior projection obtained 6 months postoperatively demonstrates dilated middle meningeal branch (arrow) and superficial temporal artery (arrowhead) with extensive intracranial collateralization

to delineate acute versus chronic ischemic infarct. The gold standard for diagnosis of moyamoya disease remains the cerebral angiogram; however, MRA provides a less invasive alternative with concise imaging results.^[2,3,7] Moyamoya disease is classified into 6 stages depending on disease progression [Table 1].

Medical treatment is aimed at slowing the progression of vascular changes, reducing the risk of thrombosis, optimizing perfusion in stenotic areas, and seizure control. This is often achieved with a combination of antiplatelet agents, vasodilators, and anticonvulsants. When conservative management strategies are ineffective and/or patients demonstrate evidence of reduced cerebral perfusion, revascularization procedures are performed. Surgical revascularization has been shown to reduce the risk of stroke recurrence.^[3] Direct revascularization in pediatric patients is less common: recipient and donor vessels are smaller, and operative times are longer than indirect methods, and temporary clamping required for direct anastomosis risks interruption of transcortical collaterals. Therefore, it is the practice at our institution to perform indirect revascularization for pediatric moyamoya disease. Extent of revascularization as determined from post-operative cerebral angiogram has been classified into 3 categories according to the Matushima grading system [Table 2].

ANESTHETIC MANAGEMENT

A multitude of anesthetic techniques and medications have been used for the induction and maintenance of anesthesia in children undergoing interventional and surgical revascularization for moyamoya disease. Pre-eminent to the anesthetic approach is a thorough understanding of the patients' pathology, symptoms, and

Table 1: Suzuki grading system of moyamoya disease

Grade	Angiographic findings
I	Narrowing of the terminal internal carotid artery
II	Initiation of deep moyamoya collaterals
III	Intensification of moyamoya collaterals
IV	Development of transdural collaterals from the external carotid artery
V	Intensification of external carotid artery collaterals and reduction of moyamoya collaterals
VI	Complete internal carotid artery occlusion and disappearance of deep moyamoya collaterals

Table 2: Summary of Matsushima grades from post-revascularization cerebral angiogram

Grade	Description
A	Area perfused by revascularization >2/3 MCA territory
B	Area perfused by revascularization between 1/3 and 2/3 MCA territory
C	Area perfused by revascularization <1/3 MCA territory

MCA: Middle cerebral artery

any underlying comorbid conditions that may further confound anesthetic options.

PREMEDICATION

Anti-anxiolytics, when there are no contraindications, are safe and favorable in helping to blunt the sympathetic and hyperventilatory responses associated with crying, agitation, and parental separation. It is prudent to consider which type of premedication to use so as to avoid sympathetic surge, hypertension, and hypotension.

MONITORING

In addition to the standard American Society of Anesthesiologists^[1] monitors with evaluation of oxygenation, circulation, and temperature, the neuroanesthesiologist should consider an arterial line, central venous monitoring, electro-encephalography (EEG), and urine output monitoring. The arterial line allows for close hemodynamic monitoring with the ability to also trend arterial oxygen and carbon dioxide tensions and blood loss. EEG enables the monitoring of cerebral blood flow and possible hypoperfusion during the surgical procedure.^[7,9] Urine output helps to guide intravascular volume status.

INDUCTION AND MAINTENANCE OF ANESTHESIA

Both intravenous and inhalational induction of anesthesia is safe and effective in the child with moyamoya. More critical than the technique chosen is the awareness of the

potential dangers associated with either. The overarching goals are to maintain cerebral perfusion pressure and thereby cerebral blood flow, avoid hypotension and hypertension, maintain normocarbia, and blunt the sympathetic responses that can increase cerebral oxygen consumption.^[7,9]

Children, unlike adults, have a high cerebral metabolic oxygen consumption rate (CMRO₂) coupled with a high cerebral blood flow (CBF). As such, pediatric patients with moyamoya are at an increased risk for cerebral ischemia once CBF is compromised. Healthy cerebral vasculature will autoregulate changes in mean arterial pressure to maintain CBF; however, in moyamoya this response is diminished for both hypotensive and hypertensive episodes.^[7,9] The most advantageous anesthetic will balance CMRO₂ and CBF, ideally lowering the CMRO₂ requirements while maintaining optimal CBF. Pertinent factors that increase CMRO₂ include sympathetic stimuli (i.e., pain, inadequate depth of anesthesia, and tracheal intubation), which can be controlled with a host of intravenous agents and volatile anesthetics. Mild hypothermia has been associated with a reduction in CMRO₂, and possible neuroprotection against cerebral ischemia.^[7] Nonetheless, the degree of hypothermia substantiating this effect has not been validated in studies and, as such, normothermia is advocated. The decoupling of cerebral autoregulation, and thereby CBF, is seen with dose-dependent increases of volatile anesthetics. Core protection of CBF focuses on maintenance of intravascular volume with non-glucose containing crystalloids, careful titration of volatile agent, and the avoidance of hypo/hypertension. CBF monitoring is not routinely employed for moyamoya surgery however, mean arterial pressure (MAP) has long served as a surrogate of autoregulation. The blood pressure limits for autoregulation in pediatric patients with moyamoya is unknown. Ideally, MAP should be maintained at or above the child's baseline (with respect to the high normal limit for the age of the child) with the utilization of vasopressors, crystalloid, and blood products, where necessary. Aggressive systolic and diastolic parameter management becomes prudent when underlying cardiopulmonary or renal disease is present. Ongoing research is investigating the role of near-infrared spectroscopy and total tissue hemoglobin extrapolated data as a measure of intraoperative functional autoregulation to determine the optimal patient specific mean arterial pressure perioperatively.^[4]

VENTILATION

Arterial carbon dioxide acts to vasodilate the cerebral vasculature (increasing CBF) at high tensions and as a vasoconstrictor (decreasing CBF) at lower tensions.

As such, hyperventilating patients with moyamoya can potentiate cerebral ischemia, and this has been shown to cause marked cerebral hypoperfusion through EEG and xenon-based inhalational methods when PaCO₂ decreases below 29 mmHg.^[4,7,9] Moreover, cerebral dysfunction secondary to hyperventilation can persist beyond these periods due to a “steal” phenomena, whereby the dilated moyamoya collaterals shunt blood flow to the new ischemic areas following termination of hyperventilation—further compromising the diseased areas. Similar, studies have demonstrated that hypercapnia in patients with moyamoya does not produce the characteristic vasodilatory response seen in healthy individuals.^[8] For these reasons, the maintenance of normocarbia is optimal.

EMERGENCE AND POSTOPERATIVE EVALUATION

The goals of emergence coincide with those of the induction of anesthesia. An immediate postoperative neurological exam is judicious. Extubation of the trachea should occur in a smooth manner so as to avoid the increase in CMRO₂ associated with coughing, bucking, agitation, and pain. This can be accomplished with the careful titration of short acting opioids, alpha-2 adrenergic agonists, lidocaine, and/or supplemental short-acting beta-blockers. The postoperative destination will be institution-and surgeon-specific; it is important that the admitting units have the capability to continuously monitor blood pressure, neurological exam, oxygen saturation, hematocrit, and urine output.

CONCLUSION

Moyamoya is a rare but austere disease in the pediatric population. Its presentation and management in children can be complicated by underlying congenital conditions and syndromes. Early diagnosis and treatment are cardinal in preventing neurologic sequelae. Surgical management is effective at reducing symptoms and ischemic recurrence. These children may present to the neuroanesthesiologist for a myriad of procedures. The perioperative anesthetic goals of care are to maintain cerebral perfusion pressure, and thereby cerebral blood flow, avoid hypotension and hypertension, balance normocarbia and normothermia, and blunt the sympathetic responses that can increase cerebral oxygen consumption. Fostered and safe anesthetic management can collaboratively optimize surgical and patient outcomes.

Financial support and sponsorship

Nil.

Conflicts of interest

There are no conflicts of interest.

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