

Introduction

Moyamoya disease (MMD) is a steno occlusive disease at the terminal portions of the intra cranial internal carotid arteries and compensatory development of a hazy network of small collateral vessels at the base of the brain. Although this disease is old, we bring forth some new insights. Due to changing technology and more advanced anesthesia medications, techniques and newer procedures and increasing complexity of the patient's diseases, we review five cases and anesthesia related to their management.

Pathophysiology

These abnormal vessels look like a puff of cigarette smoke in angiography, which is described as "moyamoya" in Japanese. The disease causes ischemic stroke, intracranial hemorrhage, headache, seizures, and transient ischemia attack in children and in adults. Medical treatment including antiplatelet, anticoagulant and cerebral vasodilator drugs can still be challenging even with better noninvasive diagnostic techniques for diagnosis of the MMD. Surgery remains the only viable option to decrease further ischemic insult and neurologic deterioration. Revascularization surgery procedures have shown to provide symptomatic benefit in a majority of these patients by augmenting collateral cerebral blood flow (CBF). The revascularization is achievable either by directly anastomosing an external carotid artery branch to a cortical artery (EC-IC bypass)or indirectly by placing a vascularized tissue pedicle in direct contact with the brain(EDAM, EDAS), leading to an ingrowth of new blood vessels to the cortex. Both procedures remain controversial in patients with atherosclerotic disease⁵ but both are commonly used for stroke prevention in moyamoya patients.

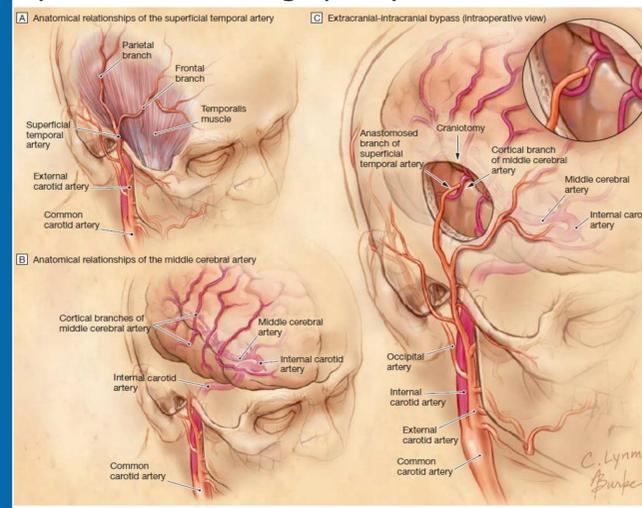
	Patient 1	Patient 2	Patient 3	Patient 4	Patient 5
Age/Sex	30/F	30/F	55/F	46/F	51/F
Presentation	TIA, Headache Post-partum vision change	TIA	TIA PreveC-IC , EDAM	Multiple Rt sided stroke	TIA
Current Surgical procedure	EDAM	EC-IC bypass	Percutaneous Balloon angioplasty	EDAM	EC –IC Bypass
MRA/MRI findings	B/L supra Clinoid ICA occlusion , Lt>Rt	B/L supra Clinoid ICA occlusion	Rt Craotid art stenosis	Rt MCA superior div infarct , Lt sided weakness	B/L supra Clinoid ICA occlusion CT- chronic left frontal and parietal infarcts
Medications for MMD	Lovenox	Lovenox	Plavix	Plavix	nil
Anesthetic technique	Art line , ET tube Sevo+Remifentanil	Art line , ET tube Sevo+Remifentanyl	Art line , ET , Sevo	Art line , ET , Remi	Art line , ET Sevo+Remifentanyl
IV fluid	crystalloids	Crystalloid	Crystalloid	Crystalloid	Crystalloid
I/O Steroid	nil	nil	nil	dexamethasone	dexamethasone
Anti epileptic	Keppra	Keppra	nil	Keppra	Keppra
Blood loss	50 cc	50cc	5	150 cc	75
Mean Temp					
Vasopressors	PhE + Eph	PhE(I)	PhE + Eph	PhE(I)	PhE (I)
Urine Output	440 cc	1400	600	940 cc	1430
Duration of surgery	174 min	250 min	172 min	258 min	250 min
Post op events	Headache	none	none	Stroke	none

Goal of anesthetic management

The anesthetic management of these patients has evolved over the years with an increased understanding of the disease. These have specifically resulted from the identification of risk factors for perioperative complications and outcomes related to the use of anesthetic agents, importance of pain control, the increased use of regional anesthesia, and better monitoring techniques in providing high quality and safe patient care to patients with MMD. The goal for anesthesia is to maintain the balance between the oxygen supply and demand which is even more important in MMD to avoid neurologic morbidity because of the underlying pathophysiology. CBF should be maintained by avoiding hypotension and maintaining normocarbia while an increase in cerebral oxygen consumption (CMRO2) associated with laryngoscopy, tracheal intubation, and surgical events can be minimized by the use of appropriate depth of anesthesia.

Thus, safe and effective anesthetic care of these patients involves understanding the pathophysiology of the disease and its anesthetic management while minimizing the risk of perioperative stroke.

We report 5 cases of adult patients with MMD who underwent different surgical procedures I,e Ec-IC bypass, EDAM and precut balloon angioplasty



Peri operative management and complication

The effects of anesthetics, surgery, and perioperative stress response on severely altered cerebral hemodynamics are complex. Evidence suggests that sparing vital collateral vessels and minimum brain retraction during surgery are essential to avoid perioperative complications. Postoperative ischemic complications are fewer in patients who have combined procedures (STA-MCA bypass with EDAMS) compared with indirect bypass. Moreover, patients who underwent indirect revascularization had a greater risk of neurologic deterioration than those who had direct anastomotic procedures

Moyamoya remains a disease with unknown etiology that results in a challenge in determining medical treatment.

Early diagnosis and effective management are

necessary before any ischemic event and neurologic impairment occurs.

Surgery remains the only viable option to decrease further ischemic insult and neurologic deterioration.⁸ Surgical revascularization procedures have been shown to provide symptomatic benefit in 87% of patients.⁵⁰ As a result, MMD patients are likely to require anesthesia for multiple diagnostic and surgical procedures.

Thus, safe and effective anesthetic care of these patients involves understanding the pathophysiology of the disease and its anesthetic management

References

- 1 Soriano SG, Sethna NF, Scott RM. Anesthetic management of children with moyamoya syndrome. *Anesth Analg.* 1993;77:1066–1070.
- 2 Moyamoya Disease: A Review of the Disease and Anesthetic Management; Tariq Parray, MD, Timothy W. Martin, MD, MBA, and Saif Siddiqui, MD (*J Neurosurg Anesthesiol* 2011;23:100–109)