Anesthetic Management of Child with Moyamoya Disease for Pial Synangiosis

Craig D. McClain, MD, MPH
Boston Children’s Hospital and Harvard Medical School
Case Presentation

• 14 year old male with bilateral moyamoya disease is referred from an outside hospital and presents for bilateral pial synangiosis
• H/o multiple TIAs
• Old, completed stroke in right hemisphere
• Mild left sided weakness, but otherwise – neurologically intact
• Pt. has history of craniopharyngioma treated with surgery and radiation
• Patient also has pan-hypopituitarism secondary to resection of craniopharyngioma
• Other medical history is non-contributory
What blood vessels are involved with moyamoya disease?

A. Internal carotid arteries
B. Vertebral arteries
C. Remnants of the Vein of Galen
D. External carotid arteries
E. All of the above
Sorry – I disagree

Try again
CORRECT !!
PATHOPHYSIOLOGY OF MOYAMOYA DISEASE
Moyamoya Disease

• Bilateral narrowing of the terminal internal carotid arteries in the supraclinoid area
• Results in long term progressive cerebral ischemia
• Secondary development of abnormal, weak, thin collateral arteries in Basal Ganglia/Circle of Willis area
• These collaterals are what gives rise to the name moyamoya, meaning “like a puff of smoke” in Japanese
• The following slide shows the circle of Willis and the internal carotid arteries

Moyamoya Disease

- Moyamoya disease is a neurovascular disorder that has a bimodal peak of presentation.
- In the first decade of life, the disease tends to present with strokes or TIAs.
- In the 5th decade of life, presentation tends to be with hemorrhage and the consequences of intracranial bleeding.

Natural History

• The natural history of moyamoya disease is quite variable.
• Progression can be slow with infrequent events, or progress can be fulminant resulting in a rapid decline with permanent neurologic impairment.
Anesthetic Goals

• Maintenance of adequate cerebral blood flow is the primary goal of anesthesia when managing these patients.

• The anesthetic and surgical management can sound fairly straightforward on paper. However, putting these plans into practice can be fraught with pitfalls.
Autoregulation

- It is unclear what degree of autoregulation these patients possess. They do appear to have some degree of autoregulation, but it is unclear what the limits are. It would appear that the limits of autoregulation in moyamoya patients are somewhat individualized and narrower than the general population.
CMRO$_2$/CBF Physiology

• Healthy children typically have a higher Cerebral Metabolic Rate of Oxygen consumption (CMRO$_2$)
• Moyamoya vessels and normal cerebral collaterals are maximally dilated at baseline
• Blood flow typically just meets oxygen demand in the awake moyamoya patient
• As mentioned above, cerebrovascular autoregulation is impaired
This graph shows the normal cerebral autoregulatory limits for healthy adult patients.
In a normal, healthy brain, there is a relative luxury of blood flow for cerebral oxygen demand.
In a brain with moyamoya, the CBF is just meeting the oxygen demand. Thus, these patients are extremely sensitive to any decrease in CBF. Even minor decreases in CBF may lead to ischemia and stroke.
RADIOGRAPHIC FEATURES
• What is the Suzuki grade of this angiogram?
  
  A. Grade I
  B. Grade II-III
  C. Grade III-IV
Sorry – I disagree

Try again
CORRECT !!
Suzuki Grade

• Disease severity classified into stages based on description of Suzuki, et al in 1969.
• The presence of the classic “puff of smoke” appearance of collaterals on angiogram corresponds to the intermediate stages of the Suzuki grading system.
Suzuki Grade

• Grade I – narrowing of ICA apex
• Grade II – initiation of moyamoya collaterals
• Grade III – progressive ICA stenosis with intensification of moyamoya collaterals
• Grade IV – development of ECA collaterals
• Grade V – intensification of ECA collaterals and reduction of moyamoya collaterals
• Grade VI – total occlusion of ICA and disappearance of moyamoya collaterals

• The following images are simply examples and not meant to be this patient’s images
Clockwise –
Suzuki I, II-III and III-IV
Axial flair MR with ivy sign (red arrow, bright signal in sulci for slow flow)
Normal flow (green arrow, black sulci).
Our Patient

• Pt has undergone MRI/MRA that showed bilateral moyamoya disease with Ivy sign
• Pt then had a cerebral angiogram that showed bilateral moyamoya disease with Suzuki grade IV on right and Suzuki grade II on left
ASSOCIATED CONDITIONS
• Please select the single best answer that describes pathologies associated with moyamoya disease.

A. Down’s syndrome
B. Neurofibromatosis
C. History of intracranial radiation
D. Hemoglobinopathies
E. All of the above
Sorry – I disagree

Try again
• A number of other pathologies are associated with moyamoya disease including Down’s syndrome, various hemoglobinopathies (namely sickle cell disease), neurofibromatosis, and patients who have had intracranial radiation (specifically for sellar masses).
Associated Conditions

• Common – 50-75%
  – Moyamoya disease without other disease
  – Asian ethnicity

• Less common (moyamoya syndrome) – 10-20%
  – Sickle cell disease
  – Neurofibromatosis type I
  – History of intracranial radiation
  – Down’s syndrome

• Rare (moyamoya syndrome) - <10%
  – Congenital cardiac disease
  – Renal artery stenosis
  – Hyperthyroidism
  – Giant cervicofacial hemangiomas

Associated Conditions

• It is crucial to understand that these associations occur frequently and to plan care with consideration of the comorbidities (e.g. congenital cardiac disease with Down’s and endocrinopathies with a history of sellar tumors)
OUTCOMES WITH TYPE OF MANAGEMENT
Outcomes with surgical management are superior to medical management alone.

- True
- False
Sorry – I disagree

Try again
CORRECT !!
Medical Management

- Medical management is limited to drugs that either decrease the headaches associated with moyamoya (calcium channel blockers, may also decrease TIAs) and antiplatelet agents such as aspirin.
- Agents such as calcium channel blockers must be used cautiously as they may cause hypotension.
- Other aspects of medical management include encouraging aggressive hydration and avoidance of activities that may cause excessive perspiration or hyperventilation.

[NEJM 2009; 360: 1226-37.]
Surgical vs. Medical Management

- Although more work needs to be done to answer this question definitively, a meta-analysis performed in 2005 indicated that the estimated rate of symptomatic progression in patients who have had surgery for moyamoya is 2.6% (1156 patients).

- That same year, a paper reported that the rate of disease progression with medical management alone is high.
Surgical vs. Medical Management

- Up to 2/3 of patients with moyamoya will have symptomatic disease progression over a 5 year period.
- In the majority of patients, even asymptomatic ones, moyamoya will progress without intervention.
- The current evidence indicates that while there is real risk to surgery, patients who have undergone surgical procedures to correct cerebral blood flow issues for moyamoya will have a much lower rate of symptomatic disease progression.
- Further, anecdotal evidence would indicate that this improvement is long lasting (at least for decades).

SURGICAL APPROACHES
What is involved with pial synangiosis? Please select the single best answer.

A. Ligation of the superficial temporal artery and anastomosis to the ICA
B. A direct surgical approach that involves EC-IC bypass
C. An indirect surgical approach approximating a target EC branch to the surface of the brain
D. Suturing a target vessel on to the dura.
E. A, B and C
F. A and C
G. All of the above
Sorry – I disagree

Try again
CORRECT !!
Surgical Approaches

• Broadly speaking, surgical approaches to treatment of moyamoya disease can be divided into direct and indirect approaches. There is no clear cut superior approach and a variety of centers have their own biases for why they prefer one over the other.

• Surgical treatment generally utilizes the external carotid to provide a new source of blood flow to the affected hemisphere.

• A complete discussion of the relative merits of direct and indirect approaches is beyond the scope of this presentation. There is a volume of literature addressing this controversy that interested readers should pursue. Please see the SNACC bibliography for moyamoya disease.

Direct Surgical Approaches

• With direct approaches, a branch of the external carotid (often the superficial temporal artery) is directly anastomosed to a cortical artery. This is also called an EC-IC bypass.

• Historically, the direct approach has been more common in adults where an immediate increase in cerebral blood flow is crucial.

• Direct techniques can be difficult in children because of the small size of the vessels involved.

Indirect Surgical Approaches

• Indirect approaches involve the placement of vascularized tissue (such as the temporalis muscle or the superficial temporal artery, supplied by the EC) in direct contact with the brain which leads to ingrowth of new vessels that ultimately will provide increased CBF to the affected hemisphere.

Indirect Surgical Approach

• The indirect approach takes advantage of the fact that chronically ischemic cortical tissue expresses a variety of angiogenic factors including metalloproteinases, transforming growth factor β-1 and vascular endothelial growth factor.

• These factors will encourage angiogenesis when there is a source of blood flow in close proximity

Indirect Surgical Approach

• Indirect approaches include encephaloduroarteriosynangiosis, encephalomyoarteriosynangiosis, drilling of burr holes, pial synangiosis

• A report of 143 patients treated with pial synangiosis showed significant reduction of stroke after the surgical procedure

• 67% of patients had strokes preoperatively, 7.7% had strokes in the perioperative periods and only 3.2% had strokes at at least one year follow-up

• The long term risk of stroke appears to be about 4% as measured in a group of patients with at least 5 year follow-up

Pial Synangiosis

• The pial synangiosis procedure consists of
  – Identification and mapping of the target vessel (often the STA)
  – Dissection and skeletonization of the target vessel
  – Opening a craniotomy under the vessel
  – Opening the dura in a stellate fashion
  – Opening the arachnoid
  – Suturing the target vessel to the pia mater
  – Closing

STA Dissection

This photograph shows the target vessel dissected out with the dura open underneath. Note the stellate fashion of dural opening which is designed to provide more area of dural edge to help further encourage vessel ingrowth.
STA-Pia Mater Attachment

In this photograph, the target vessel is being sutured to the pia mater.
INTRAOPERATIVE ANESTHETIC MANAGEMENT
Which of the following would be useful monitors for assessing adequacy of cerebral blood flow? Please select the single best answer.

A. Invasive blood pressure
B. EEG
C. ET CO2
D. All of the above
Sorry – I disagree

Try again
CORRECT !!
Monitoring During Revascularization

- The most important aspect of a safe anesthetic in these patients is maintenance of adequate cerebral blood flow.
- A variety of methods can help estimate the adequacy of CBF including ETCO$_2$, invasive blood pressure monitoring and EEG.
Intraoperative Electroencephalography
Intraoperative EEG Monitoring

• It is difficult to obtain a practical direct measurement of cerebral blood flow intraoperatively.
• MAP and ETCO$_2$ are surrogates for CBF and cerebral perfusion in the setting of intact autoregulation.
• Moyamoya patients may have some degree of autoregulation, but the limits would appear to be much narrower than patients with normal brains.
Intraoperative EEG Monitoring

• Much experience with intraoperative EEG for monitoring for cerebral ischemia during carotid surgery (CEA)

• Slowing on EEG has been associated with acute changes in CBF (i.e. hyperventilation) in pts with moyamoya

EEG

- Monitored by EEG tech from neurology
- Concerned about slowing
  - Can indicate decrease in CBF and parenchyma at risk of ischemia
- Interventions
  - Will depend on milieu of situation
    - May involve adjusting ventilation or interventions to attempt to normalize systemic BP to improve CBF
    - Propofol decreases CMRO$_2$

EEG Monitoring

• A 2011 retrospective study looked at one hospital’s experience with EEG monitoring during pial synangiosis, total of 220 patients.
• Slowing occurred in 100 cases
  – Correlated with specific operative manipulations
    • Suturing vessel to pia
    • Closure of craniotomy
• Generally occurred bilaterally, independent of side of intervention
Anesthetic Approach

• This procedure will invariably be performed under general anesthesia.

• The patients should be admitted to the hospital the night before surgery for preoperative hydration with 1.5x maintenance fluids until they go into the OR. This minimizes the hypotensive effects of the vasodilatory agents we utilize.

• The morning of surgery, the patients have a modified array EEG placed in the preoperative area if they are old enough to tolerate it when awake. In very young patients, EEG leads are placed after induction of anesthesia.
Anesthetic Approach

• Patients may be premedicated with IV midazolam and brought to the OR.
• A baseline EEG obtained and monitored prior to IV induction.
• General anesthesia may be maintained with a combination of an opioid infusion (fentanyl, sufentanil, or remifentanil) and inhaled agent.
  – The cocktail of drugs is really a matter of semantics. Practitioners must simply be aware of the pitfalls of maneuvers that decrease CBF and plan accordingly.
• The patients are placed in head pins or a gel head ring (at the surgeon’s discretion).
Anesthetic Approach

• It is appropriate to be aggressive with fluids, which is somewhat the opposite of classic fluid management approaches taught for intracranial surgery.

• The most severely affected side is generally operated upon first.

• If everything has been stable and there are no concerns, it is fine to proceed to the second side in order to accomplish synangiosis bilaterally in a single anesthetic.
Anesthetic Approach

• If slowing occurs, it is imperative to immediately ensure that cerebral blood flow is optimized.
  – Ensure the patient’s ventilation is appropriate and inadvertant hyperventilation (even very mild) has not occurred.
  – Optimize blood pressure. This may require fluid bolus or use of vasoactive agents.
  – If these maneuvers fail, small (0.2-0.3 mg/kg) doses of propofol may help alleviate the slowing without decreasing blood pressure.
Anesthetic Approach

• Care must be taken on awakening to ensure a lucid patient that can follow commands while still having adequate pain control.
• Early and frequent neurologic exams are crucial in this patient population.
POSTOPERATIVE CARE
Postoperative Period

• Is the patient cured at the end of the procedure if things have gone well?
  – Yes
  – No
Sorry – I disagree

Try again
CORRECT!!
Postoperative Care

• After an indirect surgical approach, the patient is fundamentally the same as they were preoperatively.
• However, because of the surgery, they are now set up for success.
• Vessel ingrowth can take months to accomplish so the same concerns of aggressively maintaining adequate cerebral blood flow apply in the immediate postoperative period.
Postoperative Care

• These patients should go to the ICU postoperatively for close neuromonitoring.
• Pain control and aggressive treatment of postoperative vomiting is crucial.
• There are reports of children stroking and dying in the immediate postoperative period from excessive crying due to poor pain control.
• The following three slides illustrate this concept
This is the preoperative relationship of vessels, bone and cortex.
The only change in the immediate postoperative period is the vessel has been sutured to the pia.
The brain does not have any new source of cortical blood flow, yet.
This picture illustrates the ingrowth of new vessels as a result of the angiogenic factors in the CSF encouraging angiogenesis due to proximity of the target vessel.