PIAL SYNANGIOSIS FOR MOYAMOYA DISEASE

Surgical Specialty:	Neurosurgery
Authors:	Eugene Carragee, BS, MD, and Andrew Infosino, MD, University of California- San Francisco, San Francisco, CA

Background:

- General considerations
  - Moyamoya disease is a rare progressive vasculopathy that primarily affects the cerebral vasculature resulting in stenoses of the distal internal carotid artery and its branches.[1]
  - Stenosis of the cerebral vasculature can result in cerebral ischemia, which commonly presents in pediatric patients as ischemic strokes, transient ischemic attacks, cognitive decline, aphasia, headaches, or epilepsy.[1, 2]
  - While medical management is possible, definitive treatment of Moyamoya disease is surgical. The goal of surgical intervention is to improve cerebral perfusion by either bypassing stenotic regions or to promote the growth of new cerebral vasculature. Surgical techniques typically involve a combination of direct revascularization and indirect revascularization.[3]
    - Direct revascularization involves the anastomosis of an unaffected extracranial artery directly to an affected intracranial artery.
    - Indirect revascularization involves the induction of vascular growth factors and subsequent growth of new collateral blood vessels within the brain following the placement of unaffected blood vessels on the surface of the brain.
  - Pial synangiosis is an example of indirect revascularization and typically involves suturing the superficial temporal artery onto the pial surface of the brain.[4]
  - The stenotic vasculature associated with Moyamoya disease disrupts the ability to maintain cerebral blood flow necessary to match cerebral oxygen demand. Therefore, the major intraoperative anesthetic goal is to maintain cerebral perfusion.
- Patient Considerations
  - Moyamoya disease-specific considerations
    - Preoperative and postoperative ischemic attacks may be triggered by episodes of hyperventilation, such as crying, which can cause hypocarbia and associated cerebral vasoconstriction.[1]
    - Intraoperative anesthetic goals:
      - Maintain euvolemia.
      - Maintain cerebral perfusion pressure (CPP) by preventing decreases from baseline mean arterial pressure (MAP).
      - Prevent hypocarbia which may cause cerebral vasoconstriction.

- Moyamoya Associated comorbidities/syndromes
  - Moyamoya syndrome is defined as the diagnosis of Moyamoya disease with a co-existing inherited disorder. The most commonly associated inherited syndromes in conjunction with Moyamoya Disease are: Sickle cell disease (SCD), neurofibromatosis type 1 (NF1), and Trisomy 21.[5]
    - Comorbid SCD: Increased risk of stroke and ischemic sequelae
    - Comorbid NF1: Increased risk of neurofibromas and gliomas
    - Comorbid Trisomy 21: Increased risk of additional vascular anomalies with increased stroke risk

Anesthetic Planning:

- Pre-Anesthetic Evaluation
  - Preoperative workup for Moyamoya disease includes cerebral and cerebrovascular imaging (commonly, MRI, MRA, CTA, or cerebral angiography). These imaging studies may require sedation or general anesthesia, especially in younger patients.
  - Patients with Moyamoya disease often present with seizures and are prescribed antiseizure medications, which should be continued throughout the perioperative period, including the day of surgery. [6]
  - Patients diagnosed with Moyamoya disease are frequently prescribed antiplatelet agents. The decision to continue or hold these medications in the perioperative period should be made in conjunction with neurosurgery.
  - Baseline blood pressures documented during preoperative visits prior to surgery should be used to direct intraoperative hemodynamic goals.
  - Pre-admission for intravenous (IV) hydration at 1.5 times maintenance fluids is institution-dependent and can help maintain hemodynamic stability and prevent hypotension during induction.[6]
- Specific or unique room set-up requirements
  - Drugs/infusions
    - Premedication: anxiolysis with dexmedetomidine or midazolam can be used to mitigate crying and hyperventilation which can cause cerebral ischemia.[7]
    - Phenylephrine infusion to augment/maintain blood pressure perioperatively.
  - Monitors
    - Standard American Society of Anesthesiologists (ASA) monitors.
    - Arterial cannulation for continuous blood pressure monitoring to ensure adequate cerebral perfusion should ideally be placed in the right upper extremity rather than the lower extremities. Arterial lines are often placed after the induction of anesthesia in pediatric patients.
    - Electroencephalogram monitoring has not been shown to be predictive of intraoperative ischemic events.[8]
    - Near-infrared spectroscopy (NIRS) to measure cerebral oximetry: some institutions utilize NIRS, but one study did not identify clinically relevant intraoperative ischemia.[9]
  - Blood availability
    - Patients should be typed and cross-matched; the potential for blood loss should be discussed with neurosurgery preoperatively.
  - Pediatric Intensive Care Unit (PICU) Bed Availability
    - Patients are recovered in the PICU for close hemodynamic monitoring and post-operative pain management.

Intraoperative Considerations:

- General
  - Avoid changes in intracranial pressure or cerebral perfusion pressure to maintain adequate cerebral blood flow to meet cerebral metabolic oxygen demand.[6]
- Induction
  - Either a mask or IV induction is acceptable as long as hemodynamic stability is maintained throughout and IV access is achieved quickly after induction.
  - Relaxation with non-depolarizing neuromuscular blockade is typically utilized for intubation.
  - Propofol in combination with a short-acting opioid with or without lidocaine can be used to mitigate the hemodynamic response to laryngoscopy.[10]
  - Phenylephrine infusion to maintain adequate blood pressures can be started prior to the induction of anesthesia. If an inhalational induction is utilized, phenylephrine infusion should be immediately available as soon as IV access is obtained.
- Airway
  - Secured with an appropriately sized endotracheal tube.
- Access
  - Peripheral vascular access is likely adequate, and central venous access is rarely required for the administration of vasoactive medications or central venous pressure monitoring.[6]
- Positioning
  - The operating room bed is usually turned 90 or 180 degrees; circuit extensions may be necessary.
- Maintenance
  - Utilization of IV propofol as a maintenance anesthetic may improve cerebral perfusion by decreasing ICP secondary to decreased cerebral blood flow.[7]
  - Typical maintenance regimens include a combination of propofol infusion, a short-acting opioid infusion such as remifentanil or fentanyl, dexmedetomidine infusion, and an inhaled anesthetic. No specific maintenance anesthetic has demonstrated superior outcomes. The focus should be on ensuring adequate cerebral perfusion by maintaining blood pressure goals.[6]
- Hemodynamic/Physiologic goals
  - Goal: Normotensive to Hypertensive
    - Patients with Moyamoya disease are unable to maintain cerebral perfusion at lower MAPs in comparison to healthy patients due to abnormal vasculature. MAPs should be maintained above 85% of baseline blood pressure as deviations below 85% of baseline pressure are associated with negative neurologic outcomes postoperatively.[11]
    - As hypertension is better tolerated in patients with Moyamoya disease than hypotension, some centers target a MAP of 5-10 mmHg above baseline by utilizing a phenylephrine infusion.[6]
- Fluid management
  - Goal: Euvolemia to hypervolemia
    - Fluid management goals for this procedure are euvolemia to hypervolemia (which is atypical for neurosurgical procedures) to maintain CPP.[7]
    - Some centers aim for a urine output of > 4 mL/kg during pial synangiosis as a marker of adequate hydration.[7]

- Ventilation
    - Goal: Maintain normocarbia (end-tidal CO_2 of 38-42 mmHg)
        - Hypocarbia secondary to hyperventilation results in cerebral vasoconstriction and decreased overall cerebral perfusion.[11]
        - Hypercarbia can result in the shunting of blood to healthy cerebral vasculature worsening perfusion of areas affected by Moyamoya disease via a "steal" phenomenon.[11]
- Temperature management
    - Goal: Maintain normothermia
        - Hyperthermia can result in increased cerebral metabolic oxygen demand that is not adequately matched by the affected cerebral vasculature secondary to Moyamoya disease.[12]
        - Hypothermia was historically used to decrease cerebral metabolic oxygen demand but is no longer recommended as it has not been shown to improve outcomes.
- Emergence/disposition
    - Goal: Early extubation to facilitate neurologic examination with adequate pain control.
        - A smooth extubation with minimal coughing may be facilitated by continuing either remifentanil or dexmedetomidine infusions until the patient is extubated.[6]
        - If a remifentanil infusion has been utilized, consider small bolus doses of long-acting opioids at the conclusion of the procedure to facilitate postoperative pain management and avoid hyperalgesia associated with the discontinuation of remifentanil infusions.[6]
        - Disposition to PICU to ensure adequate postoperative pain control, maintenance of hemodynamic goals, and frequent serial neurologic exams.
- Post-op Care
    - Postoperative pain management
        - Postoperative pain, hyperventilation, and crying have been associated with cerebral infarcts in patients treated for Moyamoya disease.[9]
        - Plan for multimodal pain management postoperatively: scheduled acetaminophen, short-acting opioids, and eventual transition to oral opioids.[6]
        - A discussion of the addition of non-steroidal anti-inflammatory drugs should be made in conjunction with the surgical and intensive care teams.
    - Hemodynamic management
        - Intravenous vasopressors are commonly employed to maintain MAP goals postoperatively. The patient can then be gradually transitioned to oral alpha agonists such as midodrine.[6]
    - Antiplatelet and antiseizure medications are typically continued in the postoperative period.[6]

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Reviewed by:

Reviewer #1: Debnath Chatterjee, MD, University of Colorado

Reviewer #2: Alina Lazar, M.D., Lurie Children's Hospital

Reviewer #3: Bridget K. Pearce, M.D., University of Michigan

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