orphanaesthesia

Anaesthesia recommendations for patients suffering from

Moyamoya disease

Disease name: Moyamoya disease

ICD 10: I67.5

Synonyms: Moyamoya means “something hazy, like a puff of cigarette smoke” in Japanese. It is also referred to as progressive intracranial occlusive arteriopathy.

The angiographic changes associated with moyamoya are caused by numerous heterogeneous and pathologic processes. The disease process is characterized by progressive occlusion of one or both internal carotid arteries and their proximal branches. As a result of reduced blood flow in the anterior circulation, collateral circulation composed of small vessels develops near the apex of the carotid artery, meningeal vessels, and dural/basilar branches of the external carotid artery. The angiographic abnormalities are usually bilateral, with differences in severity between sides. The affected vessels and collateral vessels are generally maximally dilated; consequently, patients are vulnerable to ischemic injury with small changes in cerebral blood flow. Patients with associated conditions, such as neurofibromatosis type I, sickle cell disease, trisomy 21, or history of radiotherapy for intracranial tumor (i.e., optic glioma, craniopharyngioma, or pituitary tumor) are referred to as having moyamoya syndrome. Patients without associated risk factors are said to have moyamoya disease. Patients with unilateral disease—with or without associated risk factors—are identified as having moyamoya syndrome. In up to 40% of patients with unilateral moyamoya, contralateral disease may develop, particularly in those of younger age.

Medicine in progress

Perhaps new knowledge

Every patient is unique

Perhaps the diagnostic is wrong

Find more information on the disease, its centres of reference and patient organisations on Orphanet: www.orpha.net

Typical surgery

- Direct revascularization (STA-MCA bypass): The superficial temporal artery (STA) is sutured to a distal branch of the middle cerebral artery (MCA), which is temporarily clamped during surgery. This procedure is also known as extracranial to intracranial (EC-IC) bypass.

- Indirect revascularization (EDAS, pial synangiosis, EMS): The encephaloduroarteriosynangiosis (EDAS) procedure involves dissection of the STA. Through a craniotomy, the STA is sutured to the dura mater, and the cranium is replaced. Pial synangiosis is a modification of the EDAS procedure in which the arachnoid is opened and the STA is sutured directly to the pia mater. In the encephalomyosynangiosis (EMS) procedure, the temporalis muscle is dissected and placed onto the surface of the brain through a craniotomy.

It is important to note that the target vessel is *not* clamped and anastamosed in patients who undergo indirect revascularization procedures. Instead, angiogenesis with increased proximal blood flow is expected to occur in areas at risk of ischemia. Thus, these patients should be considered neurologically the same after surgery as they were before surgery because ingrowth of the new vessels takes several months. Consequently, maintaining adequate cerebral blood flow remains a primary concern in the postoperative period.

Type of anaesthesia

General anesthesia is recommended for any of the revascularization surgeries described above. Physicians should conduct a thorough preoperative neurologic assessment to determine whether any neurologic deficits are present, as these patients are predisposed to transient ischemic attacks (TIAs) and strokes. Beginning in the preoperative area, and continuing postoperatively in the intensive care unit, anesthesiologists must maintain conditions that optimize cerebral blood flow and minimize the risk of ischemic or hyperemic neurologic injury.

Hyperventilation induces cerebral vasoconstriction, which increases the risk of ischemic injury. In the preoperative period, efforts should be made to prevent the child from crying (with associated hyperventilation). The patient can be kept calm with involvement of the family and/or administration of premedications. Hypercarbia can result in regional cerebral vasodilation and cerebral blood flow “steal.” Therefore, normocarbia should be maintained during the intraoperative and postoperative periods. Postoperative analgesia must be provided to prevent crying or agitation, and care must be taken to avoid postoperative respiratory depression.

Intraoperative and postoperative blood pressure should be maintained close to the patient’s baseline. To be considered a baseline blood pressure, the measurement must be obtained in a calm child with an appropriately sized blood pressure cuff. Review of recent clinical records may be necessary. Patients with moyamoya are at high risk of developing cerebral ischemia with decreases in blood pressure. Preoperative hydration with intravenous fluids the night before surgery and special attention to replacing fluid deficits in the operating room can minimize hypotension during anesthesia. Some centers administer intravenous fluids at 1.5-times the maintenance rate the night before surgery. Care should also be taken to avoid sudden hypertension.

It is paramount to avoid conditions that may increase intracranial pressure (ICP). Hypoxia, hypercarbia, and pain may induce cerebral vasodilation with an increase in ICP. Therefore, maintaining oxygenation, normocarbia, and an adequate depth of anesthesia with a balanced anesthetic is critical. If hypertonic medications must be given to treat elevated ICP (i.e., mannitol or hypertonic saline), the anesthesiologist must be vigilant to avoid hypotension after diuresis.

Postoperative extubation to enable early and serial neurologic examinations is ideal. Postoperative monitoring of arterial blood pressure should also be considered. Continuation of intravenous fluids will decrease the risk of postoperative hypotension. As previously mentioned, adequate analgesia must be provided to prevent agitation or crying (with associated hyperventilation).

Necessary additional diagnostic procedures (preoperative)

Patients with moyamoya may require frequent diagnostic tests, including brain MRIs and angiograms. The considerations described above for revascularization surgery still apply during anesthesia for diagnostic procedures.

Particular preparation for airway management

The risk of aspiration depends on associated conditions. Patients with moyamoya are not necessarily at high risk of aspiration. During induction, the goals are to maintain normotension and normocarbia in order to maintain steady cerebral blood flow. Patients with Down's syndrome may need preoperative cervical spine films and cervical in-line stabilization. The latter is important both for airway management and for positioning; excessive cervical flexion, extension, or rotation must be avoided.

Particular preparation for transfusion or administration of blood products

Cross-matched blood products should be available. Patients with sickle cell disease may have an extensive transfusion history, so they may have antibodies that prolong the time to acquire cross-matched blood products. In patients with sickle cell disease, preoperative transfusion (within 7 days) may be considered to maximize the amount of hemoglobin A. Patients with chronic transfusion maintenance regimens should be transfused preoperatively in consultation with their hematologist.

Particular preparation for anticoagulation

Patients are usually on aspirin preoperatively, and this regimen may be continued perioperatively. For direct approaches, including EC-IC bypass, heparin may be required before temporary vessel clamping.

Particular precautions for positioning, transport or mobilisation

None.

Probable interaction between anaesthetic agents and patient's long term medication

Patients with seizure disorders who take anticonvulsant medications may have upregulated cytochrome P450 metabolic pathways. Upregulation of these pathways can result in prolonged action of neuromuscular blockers and other medications that depend on these metabolic pathways.

Anaesthesiologic procedure

For both direct and indirect surgical approaches, anesthetic goals include maintaining oxygenation, normocarbia, and normotension to minimize the risk of perioperative ischemia as described above. The anesthetic regimen must take into account neuromonitoring when used. Burst suppression may be required for direct revascularization procedures. In addition to using routine anesthesia monitors (including temperature), the anesthesiologist should place an arterial line for close monitoring of the blood pressure and to obtain serial labs if needed. Large-bore intravenous access and a Foley catheter to help assess volume status are also indicated. Central venous access may be considered in patients at particularly high risk for cerebral ischemia who may require vasopressor support.

Particular or additional monitoring

Continuous electroencephalography (EEG) with a modified montage and somatosensory evoked potentials (SSEPs) are often monitored. The various benefits of such advanced neuromonitoring techniques are well described in the literature and are specific to institutional practices.

Possible complications

Perioperatively, the primary concern is cerebral ischemia and stroke. Ischemia may manifest as EEG slowing or changes in the SSEPs. Intraoperative hemorrhage and seizures may also occur. Postoperative complications include TIAs, strokes, and seizures.

Postoperative care

These patients are at considerable risk for TIAs or strokes postoperatively. Early extubation is preferred to allow for serial neurologic exams. Patients should be admitted to a setting where continuous arterial blood pressure monitoring and frequent neurologic exams are possible, such as the intensive care unit. As previously discussed, providing postoperative

analgesia to prevent hyperventilation from crying or agitation is essential to minimize the risk of cerebral vasoconstriction and ischemia.

Information about emergency-like situations / Differential diagnostics

caused by the illness to give a tool to distinguish between a side effect of the anaesthetic procedure and a manifestation of the disease:

Delayed awakening: consider residual anesthetics, neuromuscular blockade, hypoglycemia, hypothermia, hypercarbia, hypoxia, and cerebral ischemia. May need head CT.

Ambulatory anaesthesia

N/a.

Obstetrical anaesthesia

Patients with moyamoya syndrome should be treated with tight blood pressure control. Avoid both hypotension and hypertension. Peripartum pain control is crucial to preventing pain-related hyperventilation and resultant cerebral vasoconstriction. Because cerebral aneurysms may develop with disease progression, these patients are at higher risk of intracranial hemorrhage than the general population. The development of cerebral aneurysms may be secondary to chronic hypertension or regional vessel constriction in moyamoya.

Literature and internet links

1. Scott RM, Smith ER. Moyamoya disease and moyamoya syndrome. N Engl J Med 2009;360: 1226-1237.doi:10.1056/NEJMra0804622
2. Soriano SG, Sethna NF, Scott RM. Anesthetic management of children with moyamoya syndrome. Anesth Analg 1993;77(5):1066-1070
3. Hannon KE. Pial synangiosis for treatment of Moyamoya syndrome in children. AORN J 1996; 64(4):540-554
4. Vendrame M, Kaleyias J, Loddenkemper T, Smith E, McClain C, Rockoff M, Manganaro S, McKenzie B, Gao L, Scott M, Bourgeois B, Kothare SV. Electroencephalogram monitoring during intracranial surgery for moyamoya disease.Pediatr Neurol 2011;44(6):427-432.doi: 10.1016/j.pediatrneurol.2011.01.004
5. Smith ER, McClain CD, Heeney M, Scott RM. Pial synangiosis in patients with moyamoya syndrome and sickle cell anemia: perioperative management and surgical outcome. Neurosurg Focus 2009;26(4):E10.doi:10.3171/2009.01.FOCUS08307
6. Chong CT, Manninen PH. Anesthesia for cerebral revascularizтion for adult moyamoya syndrome associated with sickle cell disease. J Clin Neurosci 2011;18(12):1709-1712.doi:10. 1016/j.jocn.2011.03.026
7. Prakash A, Manninen P, Venkatraghavan L. Anesthetic considerations for extracranial to intracranial bypass (EC-IC) surgery: control of blood pressure. Can J Anaesth 2008;55(1): 4735651-4735652
8. Adams HP, Jr, Kassell NF, Wisoff HS, Drake CG. Intracranial saccular aneurysm and moyamoya disease. Stroke 1979;10:174-179
9. Lee JK, Williams M, Jennings JM, Jamrogowicz JL, Larson AC, Jordan LC, Heitmiller ES, Hogue CW, Ahn ES. Cerebrovascular autoregulation in pediatric moyamoya disease. Paediatr Anaesth. 2013;23(6):547-556.doi:10.1111/pan.12140.

Last date of modification: May 2014

This guideline has been prepared by:

Author
Monica Williams, Department of Paediatric Anaesthesia and Critical Care Medicine, Johns Hopkins University School of Medicine, Baltimore, MD, USA
mwill151@jhmi.edu

Co-Author
Jennifer K. Lee, Department of Anaesthesiology and Critical Care Medicine, Division of Paediatric Anesthesia, The Johns Hopkins University School of Medicine, Baltimore, MD, USA

Co-Author
Edward S. Ahn, Department of Neurosurgery, The Johns Hopkins University School of Medicine, Baltimore, MD, USA

Peer revision 1
Craig McClain, Department of Anaesthesiology, Perioperative and Pain Medicine, Children's Hospital Boston / Harvard Medical School, Boston, USA
Craig.McClain@childrens.harvard.edu

Peer revision 2
Edward Smith, Department of Neurosurgery, Children's Hospital Boston / Harvard Medical School, Boston, USA
Edward.Smith@childrens.harvard.edu