Anesthetic Management of Child With Moyamoya Disease With Severe Blood Loss

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GOALS
1. Understand the Moyamoya disease (MMD), clinical presentation, diagnosis
2. Discuss the implications of severe blood loss in a patient with MMD
3. Know anesthetic monitoring techniques to be used in patient with severe blood loss
4. Discuss anesthetic and pharmacological interventions to reduce intraoperative blood loss.
5. Understand the anesthetic management of patients with MMD.
6. Develop strategies to avoid perioperative cerebral ischemia in MMD patients.
7. Describe how to balance the conflicting goals for anesthetic management in these patients

Stem Case
6 year old, 15 kg female with severe thoracic lordosis and scoliosis was scheduled for posterior spinal reconstruction. Her past medical history was significant for MMD, severe restrictive lung disease and neurofibromatosis type 1. Her past surgical history included posterior mediastinal tumor excision and cerebral revascularization procedure without any anesthetic complications.

PE: HR 96, BP 110/69, R 26, T 36.5. Physical examination was unremarkable except for scoliosis.

Patient was well sedated with midazolam to avoid anxiety or hyperventilation before coming to the operating room. Standard ASA monitors were placed, followed by smooth inhalation induction with sevoflurane, placement of two peripheral intravenous lines and easy endotracheal intubation with 5.0 mm cuffed oral ETT. Central venous line was placed in the right internal jugular vein and arterial line was placed in left radial artery. Regional intracerebral hemoglobin oxygen saturation was measured using near-infrared spectroscopy.

Somatosensory evoked potentials (SEPs) were used to monitor lower and upper limb function during surgery and motor evoked potentials (MEP) were used to monitor motor tract function.

With the patient in prone position anesthesia was maintained using 0.4% isoflurane with oxygen and air supplemented with remifentanil and propofol infusions. Dopamine infusion was used to keep to keep systolic blood pressure above 90mmhg or mean arterial blood pressure above 50mmhg. During the surgical period of 5 hours patient lost approximately 925ml of blood and urine output was 185ml.

Patient received 350ml of cell saver blood, 350ml of PRBC, 40ml of platelets, 1800ml of crystalloids and 500ml of 5% albumin.

SEPs, MEPs remained consistent with baseline. Intraoperative SpO2 was maintained at 99-100% and rSO2 did not change from the baseline.

The patient at the end of surgery moved her extremities and was extubated. She was admitted in intensive care unit with emphasis on good pain control. She was discharged on sixth postoperative day without any neurodeficit.

QUESTIONS.
1. What is MMD? How does MMD present in children? Diagnosis?
2. What are your concerns in this patient?
3. How would you schedule this patient?
4. Would you stop the patient’s medications?
5. Risk factors for cerebral ischemic events in MMD? How would you monitor for cerebral ischemia?
6. What monitors would you use if you are expecting major blood loss?
7. What anesthetic techniques would you adopt?
8. Would you advise autologus predonation or use isovolumic hemodilution?
9. Any concerns about positioning the patient if she is going to be in prone position?
10. What anesthetic techniques would you employ to decrease blood loss?
11. What pharmacological measures can be adopted to minimize blood loss?
12. What would be effect of hypothermia in this patient? What are your goals for ventilation?

DISCUSSION OUTLINE

Moyamoya Disease is a rare cerebrovascular disease seen both in children and adults with variable progression and presentation. It is characterized by angiographic evidence of progressive stenosis or occlusion of terminal portions of the internal carotid arteries and the proximal portion of the anterior and middle cerebral arteries. The posterior cerebral arteries may also be involved. It is probably a genetically inherited disease, an autosomal dominant disease with low penetrance. (1)

MMD is usually symptomatic, although asymptomatic cases have been reported The disease causes frequent transient ischemia attacks in children and intracranial hemorrhage in adults. The most common clinical presentation in children is repeated transient ischemic attacks (TIA) occurring during crying, exercise, coughing or straining. The children may also present with slow cognitive decline, headaches, dizziness, seizures, visual impairment, involuntary movements, hemiparesis, monoparesis, sensory impairment or cerebral infarction. (2)(3)

If MMD is suspected, the workup starts with a head computed tomography (CT) scan looking for evidence of intracranial hemorrhage. Patients may be evaluated with an MRI/magnetic resonance angiography (MRA) which can show acute infarcts, chronic infarcts and a cortical ischemic pattern. (4)(5) Cerebral angiography remains the gold standard to confirm the diagnosis of MMD.

The medications used in medical management are: antiplatelet agents (usually aspirin or ticlopidine), vasodilators such as calcium channel blockers (nimodipine, nicardipine) and pentoxifylline. (3)(6)(7) Anticonvulsants are used in those having seizures. Children less than 6 years of age are given aspirin at 81mg per day and older children are prescribed a larger dose. Aspirin is continued lifelong at some centers but is usually stopped 7-10 days prior to surgery and replaced by low-dose low molecular weight heparin (LMWH).(8) LMWH 0.5mg per kg twice a day subcutaneously is administered in some centers for neurologically unstable patients who need reversible anticoagulation prior to surgery and angiography. Some neurosurgeons continue aspirin until the time of surgery; this may increase intraoperative bleeding for which platelet transfusion may be needed. Aspirin is restarted after the surgery on the first postoperative day. (8)
The risks depend on a variety of factors including the severity of the disease, type of surgical or diagnostic procedure, the amount of blood loss, and hemodynamic instability. Risk factors for perioperative complications predominantly cerebral ischemic events in patients with MMD are:

- History of transient ischemic attacks, severity of disease, type of revascularization procedure,

Patient undergoing posterior spinal fusion can have massive blood loss. Massive blood loss is defined as a loss of one blood volume within 24 hours or loss of 50% blood volume in 3 hours.

The priorities in this surgical case include maintaining oxygenation and tissue perfusion by transfusion of blood products and maintaining hemodynamic stability to ensure adequate CPP. These patients need wide bore peripheral intravenous cannulae in addition to the central venous line for restoration of circulating volume.

These patients should be scheduled as the first cases of the day in the operating room to avoid dehydration which may result in decreased cerebral blood flow. If the patient's surgery is delayed for some reason the patient may be given clear fluids as per NPO guidelines. If the patient is admitted on the day of surgery IV fluids may be started in the preoperative area.

The monitoring for the surgical procedure involving hemodynamic instability in addition to the standard ASA monitors require invasive arterial blood pressure monitoring, central venous pressure monitoring, urine output, and near infra-red spectroscopy (NIRS) monitoring. Cerebral ischemia is commonly monitored by EEG, although near infra-red spectroscopy (NIRS) and transcranial doppler monitoring have been used. EEG changes may indicate that areas of the brain are experiencing ischemia. Cerebral ischemia appears as slowing of frequencies with reduction or loss of amplitude, depending on the severity of ischemia. Anesthetic effects on the EEG are global, which helps to distinguish general from regional ischemia.

SSEP are used to monitor functional integrity of sensory tracts from peripheral nerves to the cerebral cortex including the dorsal spinal tract. MEP are used to monitor functional integrity from the cerebral cortex to muscle including anterior spinal tract. If the blood supply of cerebral cortex is compromised both SSEP and MEP will be affected. Ischemia will cause an increase in latency and decrease in amplitude. SSEP and MEP are also influenced by anesthetic agents, hypotension, hypothermia, hemodilution, hypercarbia, hypocarbia and hypoxia.

Patients with MMD have compromised cerebral circulation, so surgical procedures involving blood loss and hemodynamic instability increase the risk hypotension causing decreases cerebral blood flow and risk of ischemic damage. This may be compounded by a decrease in oxygen delivery because of blood loss. The goal for anesthesia is to maintain the balance between oxygen supply and demand. This can be achieved by maintaining adequate cerebral perfusion pressure (CPP), normocarbia, normothermia and an appropriate depth of anesthesia to avoid increase in cerebral metabolic rate for oxygen (CMRO₂) by anesthetic or surgical events.

Induction and maintenance
Inhalation induction should be carefully titrated to avoid hypotension and impairment of cerebral autoregulation. The choice of non-depolarizing muscle relaxant should be less likely to cause
histamine release and any hemodynamic changes. Implications of the use of succinylcholine for failed or difficult airway should be considered in patients with neurological deficits. For children with an intravenous line already in place, or adults, the use of an intravenous induction is a logical choice. Thiopental, propofol, etomidate or ketamine may be used. (12)

The choice of anesthetic technique will be dictated by the type of surgical or diagnostic procedure and required monitoring. Anesthesia can be maintained with volatile agent, nitrous oxide, and opioids. To avoid interference with neurologic monitoring the patient can be maintained on TIVA (total intravenous anesthesia) using propofol and opioids with or without a low dose of volatile agent. All volatile agents have been used in patients with MMD. (10)(14)

Some studies have shown improved outcome with TIVA using propofol, while other studies have shown no significant difference as compared to isoflurane. (15)

**BLOOD LOSS**

Decreased hematocrit due to preexisting anemia places patient with MMD at higher risk of cerebral ischemia. If severe, anemia should be corrected preoperatively with blood transfusion to improve oxygen delivery.

Acute surgical blood loss perioperatively decreases the oxygen carrying capacity in the body placing the MMD patient at risk for cerebral ischemia. Clinical signs of acute blood loss will depend on amount and severity of blood loss. Blood loss if not corrected will be manifested by tachycardia, hypotension, decrease in oxygen saturation decreased urine output, ECG changes, hypovolemic shock and death. It becomes critical to regularly check the hematocrit intraoperatively and transfuse blood as necessary. A high hematocrit with associated increased viscosity is regarded as a risk factor for cerebral infarction in patients with MMD. (11)

Certain measures can be taken to reduce blood loss and transfusion requirement but in a patient with MMD they may not be always practical. The patient’s pelvis and shoulders should be supported to avoid pressure on abdomen and inferior vena cava. This will reduce blood flow through collateral venous plexus and decrease blood loss. Pt blood lost in surgical field is salvaged intraoperatively and processed RBC’s are transfused back. This reduces the transfusion requirements.

Preoperative autologus blood donation and acute normovolemic hemodilution are useful techniques to decrease the need for blood transfusion. They may not always be an option in children who have compromised cerebral circulation Hemodilution if excessive can itself cause cerebral ischemia by reducing oxygen carrying capacity of the blood since hemodilution will decrease the oxygen carrying capacity of blood with risk of cerebral ischemia. (11).What constitutes an ideal hematocrit in a patient with MMD is debatable, but a hematocrit of 30-42% has been proposed as adequate. (16). Hypotensive anesthetic techniques should be avoided because potential risks of cerebral ischemia are greater. Many of the anesthetic agents and medications used have the potential to lower the blood pressure, and therefore must be carefully titrated.

Pharmacological agents that can reduce bleeding in surgical patients include antifibirinolytic agent’s epsilon-aminocaproic acid and tranexamic acid. (17)

**Blood Pressure and Intravascular Volume**

It is important to maintain the blood pressure at or above baseline preoperatively as well as perioperatively. Hyperosmotic drugs should be avoided as they cause dehydration and
hypotension. Surgical procedures involving extensive surgical blood loss and hemodynamic instability can result in decreased CPP. In such patients central venous pressure, arterial blood pressure and urine output must be carefully monitored which together should provide a good indication of volume status. (3) Vasopressors have been used to maintain the CPP at the baseline. (12)

Temperature
Hypothermia can occur during long surgical procedures; especially the large body area is exposed. Hypothermia increases perioperative bleeding by enzyme inhibition of both clotting factors and sequestration of platelets. Blood loss increases because coagulation is temperature dependent process. It is therefore generally agreed that normal body temperature should be maintained perioperatively. Blood warmers, forced warm air blankets and ambient room temperature should be employed.

Ventilation
Changes in ventilation will not have any major effect on blood loss, but affects the CBF. Changes in ventilation will affect the CBF and can be a major factor in determining neurological complications. In Moyamoya patient’s hypercapnia dilates the normal cerebral vessels, while the diseased vessels show minimal response resulting in less blood flow to areas supplied by diseased vessels. (9) Hypocapnia induced by hyperventilation causes cerebral vasoconstriction and the areas of brain supplied by diseased vessels are more at risk of cerebral ischemia. (9) Hyperventilation induced by crying has been seen to trigger TIA in children with MMD. (12)

REFERENCES.


