Case Skeleton for a Patient with Sickle Cell Disease and Moyamoya for Pial Synangiosis

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Objectives:

- Discuss and understand the anesthetic considerations for a patient with complicated sickle cell disease who is undergoing general anesthesia.

- Discuss and understand the pathophysiology of moyamoya disease.

- Discuss and understand the preoperative, intraoperative, and postoperative management of patients with moyamoya disease.

- Discuss and understand the implications of general anesthesia in patients with sickle cell disease complicated by a history of TIA and/or CVA.

- Discuss and understand the effects of general anesthesia on cerebral blood flow and cerebral physiology.

Case History:

A 10 yo African American male with a history of sickle cell disease is diagnosed by angiogram with moyamoya disease after a TIA. He is scheduled for pial synangiosis.

Questions:

What is sickle cell disease? In particular, what are the advanced complications of sickle cell disease? What are the possible neurologic complications of sickle disease? For those who present with TIA or stroke, how are these patients initially treated? What are the risks of this treatment regimen? What are the options if this treatment regimen does not work?

What is moyamoya disease? Is this different than moyamoya syndrome? How can sickle cell disease lead to moyamoya syndrome? What is the typical presentation of moyamoya syndrome? What happens to cerebral blood flow in patients with moyamoya and how does this couple with cerebral metabolic consumption rate of oxygen? How does hypoventilation or hyperventilation affect these patients?
Preoperative evaluation:

This 10 yo child presents with no current focal neurological deficits. He had a history of several transient ischemic attacks (TIA) in the past that resolved with blood transfusions. Per the parents, he is currently on a transfusion protocol. Physical exam is normal.

Questions:

What additional preoperative evaluation would you need to proceed? Are there any services that would need to be consulted prior to surgery? Is there any evidence that surgery is beneficial? What is your target hematocrit prior to surgery? Would you continue existing anticoagulation medications? Would you have this patient be admitted the same day to surgery or would you pre-admit this patient for any reason? What are your anesthetic concerns for patients with sickle cell disease?

Anesthetic Plan:

Questions:

What are the surgical treatments of moyamoya disease? What is your anesthetic plan for the case? How do the considerations of dealing with sickle cell disease and moyamoya disease under general anesthesia mesh or conflict?

Intraoperative Care:

Questions:

What are your hemodynamic goals during the induction of anesthesia? During the maintenance? Why is this important? What are your goals for the temperature, CO2, and pH during the case? Why are these goals important for this particular case? What monitors would you use for this patient? Would you use EEG monitoring intraoperatively? Would you want blood products available? Is using an inhalational agent better than an intravenous agent? Would you use opioids? What is your plan for emergence?

Postoperative Care:

Questions:

Does this patient need the ICU post-operatively? How will you control their pain post-operatively and why is that important? What are your major concerns postoperatively? Any studies postoperatively?
Surgical Outcome:

Questions:

What are the long-term outcomes in patients who undergo these procedures? How do these outcomes compare to patients undergoing medical management only?

Discussion:

Sickle cell disease is a congenital hemoglobinopathy characterized by deformed red blood cells. This leads to acute episodic attacks of pain and pulmonary compromise, widespread organ damage, and early death. States such as hypoxia, acidosis, hypotension, venous stasis, vasoconstriction, and hypothermia should be avoided in the perioperative period as these can all cause erythrocytes to sickle that leads to vaso-occlusion and organ dysfunction.

Approximately 100,000 people have sickle cell anemia in the United States and about 10% of children with sickle cell anemia have a stroke before the age of 20. Initially, medical management is attempted. A patient is started on a transfusion regimen if he/she has had a previous stroke or if he/she had an elevated transcranial doppler study > 200 cm/sec. Transfusion therapy decreases the stroke risk about 10-fold. These transfusions must be continued indefinitely. However, up to 43% of patients with sickle cell anemia and strokes will develop moyamoya-like collaterals on imaging studies. These patients have a 5-fold increase for recurrent stroke compared to those without evidence of collateral vessels.

Moyamoya syndrome is a chronic occlusive cerebrovascular disorder of unknown origin. It is characterized by progressive stenosis of the bilateral supraclinoid internal carotid arteries with resultant formation of tortuous arterial collaterals at the base of the brain, which reconstitute the distal branches of the cerebral circulation. At the molecular level, there seems to be an intimal thickening and narrowing of the lumen. Over time, this can progress to a partial or complete occlusion of the cerebral arteries. Other normal collateral vessels in the ischemic hemisphere are maximally dilated. Moyamoya disease is the idiopathic presentation of angiographic moyamoya changes while moyamoya syndrome is the presentation of angiographic moyamoya changes found in association with another known pathological state.

Patients typically present with transient ischemic events and strokes. Behavior such as blowing balloons, whistling, and crying (behaviors that result in hyperventilation and consequent hypocapnia) can precipitate these symptoms. Cerebral blood flow is decreased in patients with moyamoya syndrome compared to healthy individuals. Also, the degree of the cerebral autoregulatory response to hypotension is unclear in children with moyamoya disease. Hence, because of mismatching of CBF and CMRO2, children are more prone to developing neurologic deficits during hypotensive episodes. Arterial CO2 tensions are also important as hypocarbia has been shown to lead to cerebral ischemia in these patients via
pH mediated decreases in cerebral blood flow. Alternatively, hypercapnea does not seem to improve cerebral blood flow.

Patients with sickle cell anemia may have repeated end organ injury from sickling in a variety of vascular beds, abnormal regulation of vasoconstriction, and intimal hyperplasia. There is an association between sickle cell disease (as well as other hemoglobinopathies) and moyamoya disease. The presence of moyamoya is associated with an increased incidence of stroke.

For this presentation, consultation with hematology services would be ideal. Often, the patients are referred from the hematologist for angiographic and surgical evaluation. The ideal hematocrit is unclear. However, the important consideration is optimizing the timing of the surgical procedure soon after the patient's routine transfusion (as part of the transfusion protocol) to minimize the presence of sickle prone erythrocytes. Aspirin should be continued perioperatively. Our practice is to admit the patient to the hospital the night before the surgery for intravenous hydration in order to minimize hemodynamic perturbations during induction of anesthesia.

Intraoperatively, the patient should be kept normotensive, normothermic, normocarbic, well hydrated, and maintain a normal pH. These maneuvers will lessen the likelihood of sickling as well as maintain adequate cerebral blood flow. Good peripheral access is necessary. In addition to standard ASA monitors, an arterial line should be placed as well as a precordial doppler. Our institution also uses intraoperative EEG monitoring to look for slowing as a surrogate marker of inadequate cerebral blood flow. This necessitates having a dedicated individual who is experienced in assessing real-time EEG tracings. The hemodynamic goals are to maintain normotension throughout. Alterations in cerebral blood flow can severely compromise these patients. Therefore, the maintenance of adequate cerebral blood flow is the main priority to minimize the risk of perioperative strokes. An optimal balance between cerebral metabolic consumption rate of oxygen and cerebral blood flow is the desired result. In order to decrease CMRO2, an adequate depth of anesthesia in necessary during painful stimuli: laryngoscopy, tracheal intubation, placement of head pins, and surgical incision. Maintenance of adequate intravascular volume is crucial. The approach to these patients is significantly different from the classic anesthetic approach to craniotomies where a relatively “dry” patient is the goal. In this setting, keeping the patient relatively “dry” may lead to severe postoperative complications such as stroke. There is no single “best” technique in terms of agent selection. The importance is matching CBF to CMRO2. Maintain normocarbia as potential problems exist with both hypo and hypercarbia.

Postoperatively, these patients should be cared for in an intensive care unit. As with all intracranial neurosurgical procedures, frequent neurologic checks are the most sensitive means of following the patient’s postoperative course. Avoiding hypotension and hypocapnia is crucial in the postoperative period. It is vital to understand that at the end of the procedure, the patient is still at risk for significant neurologic events. The surgery sets the patient up for success but the development of collaterals from the synangiosis will take months to occur. Thus, the same concerns that existed both pre- and intraoperatively remain in the postoperative period. Maintenance of adequate IV hydration is essential until
the patient can recover fully and drink well (which typically occurs 2-3 days post-op). Aspirin therapy should be continued throughout the perioperative period.

It should be noted that many of these patients with sickle cell disease have developed opioid tolerance from repeated exposure to opioids during pain crises, so higher than average doses of opioids may be required postoperatively for pain control. Pain control is very important as crying can lead to hyperventilation which may lead to cerebral vasoconstriction and subsequent hypoperfusion. There should be a low threshold to give supplemental oxygen to prevent hypoxia, and to promote incentive spirometry.

Surgical outcome for this patient population is quite favorable. The perioperative stroke rate in these patients is 5.3% per hemisphere while the stroke rate is 4% for moyamoya patients without sickle cell anemia. The rate of stroke years later in these patients appears to be minimal. The limited data on long-term outcomes appears to be quite favorable with surgical approaches offering decreased rates of neurologic events in pediatric patients as compared to only medical management.

References:


10) Ganesan V. Moyamoya: to cut or not to cut is not the only question. A paediatric neurologist’s perspective. Dev Med Child Neurol. 2010 Jan;52(1):10-3.