

## **PBLD – Perioperative Management of Stroke in an Adolescent with Moyamoya and Sickle Cell Disease Presenting for Pial Synangiosis**

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### **GOALS:**

- Discuss the perioperative management of sickle cell disease including preoperative evaluation, erythrocyte transfusion strategies, pain control, and possible complications such as stroke.
- Discuss the pathophysiology and surgical treatment of moyamoya.
- Identify the appropriate consultations and perioperative care for surgical treatment of moyamoya in a patient with sickle cell disease

### **CASE DESCRIPTION:**

The patient is a 15 year old female with sickle cell disease (SCD) hemoglobin SS scheduled for six-vessel cerebral arteriogram. One month ago she was diagnosed with a stroke after she was admitted to the hospital with symptoms of left-sided weakness, drooling, and aphasia. MRI revealed acute right tempoparietal ischemia and subacute left basal ganglia ischemia. She made a good recovery with only mild residual left hemiparesis and mild aphasia. The patient has not been compliant regarding SCD follow-up. Four months ago she had a therapeutic abortion for a pregnancy that was revealed during work-up for a pain crisis in the abdomen and lower back. She was recently seen by nephrology for hypertension with evidence of left ventricular hypertrophy on echocardiography.

### **Questions:**

1. What is the pathophysiology of SCD?
2. What are the preoperative considerations and anesthetic goals for a patient with SCD?
3. Would you consider transfusing this patient prior to the procedure? What are the current perioperative transfusion guidelines for SCD?
4. What measures should be taken for medical optimization of this patient?
5. What are the possible perioperative complications of SCD? What are the predictors of SCD related perioperative complications?
6. What are your thoughts regarding the patient’s hypertension? Should this be treated pre-operatively?

**Case History (Continued):**

Cerebral angiography reveals high-grade narrowing of the right internal carotid artery and severe stenosis of the left anterior cerebral artery. Neurosurgery recommends bilateral extracranial to intracranial bypass (right pial synangiosis and left burr holes) for moyamoya.

**Questions:**

1. What is moyamoya? Is moyamoya disease different from moyamoya syndrome?
2. What is the pathophysiology and natural history of moyamoya?
3. What is the management of a patient with moyamoya?
4. What pre-operative information do you want prior to scheduling this case?
5. The patient's medications include aspirin, Folic Acid, penicillin and Keppra. Do you recommend continuing these medications until surgery? Should any other medications be initiated?
6. What is your anesthetic plan? What monitors do you plan to use? Which neurophysiologic monitors would you use?
7. What is your approach for analgesia?
8. How does SCD complicate the intraoperative management of this neurosurgical procedure?

**Intraoperative Care:**

Anesthesia was induced with midazolam, propofol and fentanyl followed by rocuronium. Following successful oral intubation by direct laryngoscopy, the non-invasive blood pressure measured 216/125 with a MAP of 175 mmHg and HR of 130.

**Questions:**

1. What is your goal blood pressure for this particular case?
2. What vasoactive agents would you use to lower or increase this patient's blood pressure?
3. What agents will you use for maintenance of anesthesia in this patient?
4. What are your goal parameters for the patient's PaCO<sub>2</sub>, temperature, and fluid management?
5. What is your threshold for blood product transfusion during this case?
6. Should you give a diuretic and/or mannitol for better surgical exposure?
7. Do you plan to extubate the patient in the operating room?

**Postoperative:**

The patient was extubated in the OR and transported to the ICU.

1. What perioperative complications can occur after surgical treatment of moyamoya?

**DISCUSSION:**

Sickle cell disease results from a mutation in the beta-globin gene resulting in abnormal hemoglobin S. Sickle cell trait is a heterozygous state where erythrocytes contain hemoglobin A

and S and follows a benign clinical course. The severe form of sickle cell anemia occurs with inheritance of paired mutant beta-globin genes that produces hemoglobin SS. Hemoglobin SS is unstable in the deoxygenated state and can precipitate out of solution causing the characteristic sickling of erythrocytes. These impaired cells can break down and release free hemoglobin, triggering a cascade of impaired nitric oxide signaling, oxidative stress, inflammation and endothelial damage. The chronic expression of SCD results in vascular destruction and organ damage marked by intermittent acute vasoocclusive pain crises. Other major SCD complications include acute chest syndrome and stroke. Transcranial doppler (TCD) screening is an important method to identify stroke risk. Chronic transfusion therapy is recommended for those identified as high risk patients.

The perioperative management of SCD includes consideration of erythrocyte transfusion, maintenance of oxygenation and normothermia, standard fluid management to avoid hypovolemia, adequate analgesia, and avoidance of acidemia. There is significant debate on the appropriate guidelines for the use of perioperative prophylactic PRBC transfusion. It is generally recommended, however, that high risk surgical procedures on patients with greater disease severity warrant perioperative transfusion acknowledging the associated morbidity. Predictors of perioperative risk for SCD includes the type of surgery, age, severity of disease as assessed by frequency of SCD complications.

Moyamoya is characterized by a progressive occlusion of the terminal portion of the internal carotid arteries and proximal branches. A network of fragile, capillary-sized collateral vessels develops over time. The typical angiogram resembles a “puff of smoke,” which, in Japanese is moyamoya. Moyamoya disease is an idiopathic condition often involving bilateral internal carotid arteries, whereas moyamoya syndrome is associated with other conditions such as SCD that cause a similar vasculopathy. Clinically, children present with ischemic signs and symptoms, whereas hemorrhagic events are more common in adults. Diagnosis is made with cerebral angiography. The goal of treatment for moyamoya is to preserve or improve cerebral blood flow, to halt the development of additional collateral vessels, reduce symptoms and prevent strokes. Medical therapy includes antiplatelet agents and calcium-channel blockers. Progressive disease is usually treated surgically through a variety of approaches. The surgical approach has a good safety profile with a greater than 85% symptomatic benefit. The perioperative risks include additional ischemic events and intracranial hemorrhage. Perioperative management goals are to avoid hypocarbia, hypovolemia, and hypotension. Attempts should be made to avoid **agitation, anxiety, and pain**.

The combination of SCD and moyamoya poses a significant risk of perioperative morbidity. The anesthetic management of these patients requires a careful and thoughtful approach to avoid disastrous complications.

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