PBLD- Table

A 9-year-old patient with Williams Syndrome and mid-aortic syndrome requiring left heart bypass for a thoracoabdominal aortic bypass

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Objectives

1. Review the anesthetic concerns of patients with Williams syndrome
2. Describe the anesthetic considerations in a patient with mid-aortic syndrome
3. List types of monitoring that would be advantageous in an extra-anatomic thoracic to descending aortic bypass
4. Discuss options for single lung ventilation in children
5. Understand the various options for spinal cord protection during aortic surgery

A 9 year old, 21 kg, with Williams syndrome and hypertension was diagnosed with mid-aortic syndrome. Her aorta was stenotic between the celiac and superior mesenteric arteries and now she is scheduled for a thoracoabdominal aortic bypass.

1) What is Williams Syndrome?
2) What is the pathophysiology of William's Syndrome?
3) What are the cardiac abnormalities associated with William's syndrome? What are the non-cardiac symptoms and signs associated with William's syndrome?
4) What are the anesthetic considerations in patients with William's Syndrome? Should pediatric cardiac anesthesia be involved in this case?

Our patient had undergone previous grafting of her ascending aorta with reimplantation of a coronary artery, multiple pulmonary artery stents and balloon dilations, as well as previous stenting of the mid-aortic coarctation. However, she continued to have lower limb claudication and uncontrolled hypertension despite 3 anti-hypertensive agents. Therefore, she was scheduled for an extra-anatomic thoracic to abdominal bypass and revascularization of the kidneys using left heart bypass.

1) What concerns do you have for induction of anesthesia? How would you induce general anesthesia? Would you place invasive monitoring (arterial line) prior to induction?
2) What access do you want for this case? What type of invasive monitoring would you use? A-line? Central line? Transesophageal echocardiography (TEE)?

3) Would you do one-lung ventilation for this case? If so, how would you isolate the lung? Bronchial blocker? Double lumen ETT? Right mainstem intubation? What are the risks and benefits of each?
4) Would you use an anti-fibrinolytic (i.e. tranexamic acid, aminocaproic acid) for this case?

After induction of general anesthesia, a large thoracoabdominal incision was made to expose the left chest and entire aorta. During this period, the patient was hypotensive.

1) What antihypertensive medications would you have given this patient prior to her procedure? Which ones would you stop, and for how long?
2) What are your goals for blood pressure management in this patient? What are the risks associated with hypotension in this patient?
3) What vasopressors would you use to maintain a normal blood pressure? What are the advantages and disadvantages of different vasopressors or inotropes?

After the heart was safely exposed, heparin was administered and left heart bypass was initiated for the thoracic anastomosis portion of the surgery.

1) What is left heart bypass? How is this different from full cardiopulmonary bypass? Why was left heart bypass used in this operation?
2) What monitoring do you need for left heart bypass?
3) What are you concerned about during initiation and separation from left heart bypass?
4) What do you need to communicate with the perfusionist during left heart bypass? What does the perfusionist need to communicate to you?

After the thoracic anastomosis was completed, the patient was separated from left heart bypass. The remaining surgery required aortic clamping and unclamping as the SMA, IMA and renal arteries were re-anastomosed to the new aortic graft.

1) How would you manage this portion of the surgery? Would you choose crystalloid, albumin or blood for fluid resuscitation? What other resuscitation medications would you prepare?
2) What methods would you employ for spinal cord protection?
3) What other concerns do you have during this period of aortic clamping? What about specific to a patient with William's Syndrome?

Mild-moderate hypothermia was used for additional spinal cord protection. During aortic unclamping, there was a period of hypotension requiring intermittent re-clamping while additional volume was readministered. No significant coronary ischemia was noted.

The procedure was completed in 12 hours.

1) What is your plan for pain control postoperatively? Would a regional technique be appropriate? Why or why not?
2) Would you extubate this patient in the operating room?
3) What do you expect in the first 24 hours postoperatively?

The patient was successfully resuscitated and went to the intensive care unit. She required additional blood and vasopressor support overnight. She was successfully extubated the next day with nurse-controlled analgesia.

Case Summary:

Williams-Beuren syndrome (WS) was first described in 1961 and occurs in 1;20,000 live births. The classic description of children with this syndrome includes supravalvular aortic stenosis, mental retardation, behavioral traits ("cocktail personality"), elfin facies, and neonatal hypercalcemia. In addition, other vascular manifestations include peripheral pulmonary artery stenosis in up to 80% of patients, narrowing of the ascending and descending aorta, ostial or diffuse coronary artery stenoses, and stenoses of the renal and mesenteric arteries.

Williams syndrome is the result of a 7q11.23 deletion, which codes for the elastin gene – an important component of normal arterial smooth muscle. Elastin present in large arteries allow for the windkessel effect – the hydrodynamic energy stored during systole and released during diastole. Patients with WS produce 85% less elastin than normal patients, resulting in hypertrophy of smooth muscle cells. The resulting intimal thickening of large vessels causes the characteristic supravalvular aortic stenosis as well as stenosis other major systemic and pulmonary vessels.

The left-sided obstructive lesions may consist of a combination of a narrowed sino-tubular junction, abnormally thickened and restricted aortic valve leaflets, and/or narrowing of the ascending aorta. Due to the increased afterload, the left ventricle hypertrophies, resulting in diastolic dysfunction. The presence of peripheral pulmonary artery stenoses may lead to right ventricular hypertrophy. The hypertrophy of one or both ventricles results in a propensity for subendocardial ischemia. Coronary artery malperfusion is a particular problem in patients with WS. Patients may have abnormal coronary anatomy, but in addition, the abnormal flow characteristics in the aortic root caused by the left sided obstructions also make these patients at high risk for myocardial ischemia.

In Williams syndrome, the majority of sudden cardiac death has been attributed to myocardial ischemia with common features including rapid hemodynamic decline and a lack of response to aggressive resuscitation. In a series of 19 cases of sudden cardiac deaths, 11 events were associated with the administration of sedative or anesthetic drugs for catheterization or noncardiac surgical procedures. After review of available autopsy data, patients with either coronary artery abnormalities or biventricular outflow obstruction likely at the highest risk of sudden cardiac death.

In order to mitigate the risk of myocardial ischemia, several components of anesthetic management are key. These include avoiding tachycardia and arrhythmias, providing adequate volume resuscitation prior to induction to normalize preload, maintaining contractility of the heart, minimizing medications that decrease systemic vascular resistance, aggressively treating hypotension, and avoiding increases in pulmonary vascular resistance.

As mentioned above, the elastin deficiency in WS may lead to intimal thickening of large vessels, such as the aorta, mesenteric, and renal arteries. Therefore, a potential solution is to bypass the stenoses with an extra-anatomic thoracoabdominal graft. Risks during aortic clamping include myocardial ischemia, spinal cord ischemia and paralysis, and renal or bowel ischemia. As in adults, the aortic cross clamp may result in increased afterload on an already hypertrophied left ventricle with marginal coronary perfusion, placing the patient at increased risk for myocardial ischemia. Clamping the aorta at the thoracic level may decrease spinal cord perfusion pressure (Spinal cord perfusion pressure = Aortic pressure – cerebral spinal cord pressure) and increase the potential for spinal cord ischemia and paralysis.

Like a thoracoabdominal aortic aneurysm repair, an extra-anatomic thoracoabdominal bypass procedure may be done with a clamp-and-sew technique, left heart bypass, cardiopulmonary bypass, or circulatory arrest to aid in the aortic anastomoses during aortic clamping. Left heart bypass is unique in that the sole purpose is to provide flow to the lower body but does provide the ability to oxygenate, cool or warm, remove air, or add volume. Therefore, the anesthesiologist must be prepared to manage preload, treat acid/base abnormalities, provide adequate oxygenation and ventilation, and thermoregulate the patient. Unclamping (and weaning from left heart bypass) requires adequate volume resuscitation to maintain preload and careful attention to the lactic acid released into the circulation, which may precipitate more hypotension or arrhythmias.

Another important concern in descending aortic surgery is spinal cord protection. Several methods of spinal cord protection have been studied including lumbar spinal drains, mannitol, and steroids. However, the most effective methods are hypothermia and maintaining adequate spinal cord perfusion pressure. Even mild hypothermia (32^{o}C) doubles spinal cord ischemic tolerance. Spinal cord perfusion pressure can be maintained either by increasing systemic blood pressure or decreasing the cerebral spinal cord pressure, via a lumbar spinal drain. While there is data in adults suggesting that a lumbar spinal drain may be beneficial for high-risk thoracoabdominal aortic aneurysm repairs, there are no data regarding pediatric patients.

References

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