

Case Skeleton

PBLD – Table #12

AN ON-CALL NIGHTMARE: EPISTAXIS IN AN ANTICOAGULATED VAD PATIENT

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1. Goals:

- a. Discuss the cardiovascular complications and anesthetic considerations of Kawasaki disease.
- b. Review the preoperative evaluation of patients with a left ventricular assist device (LVAD)
- c. Evaluate the different types of LVADs available for the pediatric population
- d. Describe the concerns in the anesthetic management of patients with LVADs undergoing non-cardiac procedures
- e. Identify and prevent early peri-operative complications in pediatric patients on LVADs
- f. Discuss the safest way to recover this patient population

2. Case description

A 2-year-old, 15 kg child with a history of Kawasaki disease now awaits heart transplantation while being supported with the Berlin EXCOR LVAD. His course leading to the LVAD was tortuous, beginning with medical management and ending with both a CABG and mitral valvuloplasty to address Kawasaki-induced coronary artery aneurysm and severe mitral regurgitation. As these repairs failed to halt the progression of left ventricular dysfunction, an LVAD was ultimately placed.

Questions:

What is Kawasaki disease? What are its most common manifestations? How is it medical managed? What are the anesthetic implications of Kawasaki disease? What are common causes of heart failure in the pediatric population? What types of MCS are available in the pediatric population? What are the advantages/disadvantages of VAD vs ECMO? How do we determine when to place a VAD?

Case history and physical examination (continued):

While awaiting transplantation, the patient is anticoagulated with warfarin and aspirin. Nutrition is taken orally, but in an attempt to bolster the patient's weight, a nasojejunal feeding tube is placed. After an initial failed attempt at passing the tube through the right nostril, the tube is finally placed down the left nare. Minutes thereafter, blood is noted from the patient's nose and mouth, appearing in a continuous stream. External compression and packing do not stop the bleeding.

ENT is urgently consulted and the patient is scheduled for nasal exploration and hemostasis. You're called about the case, and on your arrival to evaluate the patient, you note the following vitals: temperature 36.1°, heart rate 88, RR 23, blood pressure 82/54, and SpO2 98% on room air. LVAD settings include: rate of 90; % systole: 45; stroke volume 30 ml and driving pressure: 235/-30 mmHg. No fibrin deposits are observed and the pump has an adequate filling/emptying cycle.

Questions:

Why is anticoagulation and/or antiplatelet therapy necessary in patients on LVADs? What are normal settings for a Berlin EXCOR VAD? How do you examine the LVAD? What information is needed to assure that the device is working well? What LVAD settings can the physician control?

Preoperative studies:

EKG: NSR with left axis deviation, deep Q-waves in lead V6 and aVL, and left ventricular hypertrophy.

Echocardiogram: systolic flow from the outlet cannula is observed in the ascending aorta. The inlet cannula is seen in the left ventricular apex with laminar flow into it by color and spectral Doppler. There is mild aortic regurgitation. The LV is moderately to severely hypertrophied and interventricular septum is dyskinetic. The RV size and systolic function is normal.

Labs: The only abnormal laboratory value is an INR of 3.7. The patient is T+C.

Questions:

Is the presence of deep Q-wave in lead V6 and ST depression in infero-lateral leads concerning? What is important in the echo evaluation of a LVAD patient? How does the LVAD support affect right ventricular function? Is the INR value concerning? Would you reverse the anticoagulation on this patient *before* surgery? What are the risks of doing so?

What are the anesthetic considerations in patients with an LVAD with poorly controlled epistaxis? What anesthetic options would you consider for this patient? How would you induce anesthesia? If the patient has no IV access would you do an inhalation induction? How will you manage the airway? What monitors would you need for the procedure?

Intraoperative care:

The patient has a PICC line for IV access. Anesthesia is induced with midazolam (0.1 mg/kg) and ketamine (1 mg/kg) and maintained with increments of midazolam (0.05 mg/kg) ketamine (0.5 mg/kg). The airway is managed with intermittent facemask with O₂ at 4 LPM.

Questions:

If the patient develops hypotension during the procedure, how would you treat it?
What are the potential causes of hypotension in a patient with a VAD? How would you determine the cause in this patient?

Surgical Procedure:

The ENT surgeon places SURGIFLO® Hemostatic Matrix constituted with thrombin and oxymetazoline in the right nostril. After pinching the septum for 10 min, the bleeding stops.

Questions:

What are the systemic effects of oxymetazoline? Where should the patient recover?

Postoperative care:

The patient recovers well in the PACU and no further bleeding is noted from the right nostril. As the LVAD continues to function appropriately and hemodynamics remains stable, the patient is transferred to the step down unit for further monitoring.

3. Discussion outline

Kawasaki disease (KD) is an inflammatory disease of unknown autoimmune origin probably triggered by an infectious insult. Has a male/female predominance of 1.5:1 and affects all ethnic groups. In the US is more common in winter months. It is a disease of young children peaking between 1-2 years of age and 80% of the cases are under 4 years old. It courses through an **acute phase (1st 10 days)** with high fever (> 39°) for more than five days and a least four of the following symptoms:

- Changes in extremities including erythema or edema.
- Rash within 5 days of onset (Polymorphous exanthema)
- Conjunctivitis

- Changes in the lips and oral cavity (e.g. erythema, lip cracking and/or mucosal injection)
- Cervical lymphadenopathy

Cardiovascular (CV) manifestations in the acute phase include pericarditis and/or myocarditis. The presentation includes tachycardia, gallop rhythm and systolic murmur if mitral regurgitation present. ECG changes are frequent including prolonged PR and nonspecific ST-T changes. At the end of the first week coronary involvement can be seen. Q waves are indicative of myocardial infarction as in our patient. Other non-CV manifestations include arthritis or arthralgia, abdominal pain and irritability. Laboratory findings are nonspecific but acute phase reactants and platelets are elevated. Elevated platelet count contributes to coronary thrombosis.

The sub acute phase (11-25 days) is characterized by improvement on the inflammatory reaction with rash, fever and lymphadenopathy disappearing but significant cardiovascular changes may be present. On echo about 20% of the patient will have coronary artery aneurysms defined by a coronary dimension greater than 3 standard deviation of any of the three proximal segments. Dilated cardiomyopathy in KD is secondary to ischemia secondary to coronary aneurysm thrombosis and/or myocarditis. **The convalescent phase** lasts until the elevated ESR and platelet count normalize.

The medical management of acute phase KD includes immunosuppression with immunoglobulin, steroids, aspirin and supportive therapy. During the sub-acute and convalescent phase in-patients with coronary aneurysm therapy is targeted to prevent coronary thrombosis with antiplatelet agents. One third of the aneurysms become obstructed and are associated with myocardial infarction, arrhythmias, or sudden death.

The **anesthetic considerations of KD** in acute phase include those related with bleeding from the oropharynx due to the mucosal injection. Beyond the initial phase

cardiovascular manifestation is the main concern. The pre-anesthetic evaluation should include 12-lead EKG and 2-D echo to rule out coronary arteries involvement. Early detection and treatment of ischemia is mandatory in these patients. This patient is the typical presentation of *isolated left ventricular failure* secondary to LAD coronary aneurysm thrombosis. For this reason LVAD support is adequate in this patient and ECMO is not needed.

ECMO is indicated in biventricular failure, pulmonary hypertension, or combined left ventricular and respiratory failure. Disadvantages of ECMO relative to a VAD include an increased systemic inflammatory response due to the presence of the oxygenator, less decompression of the heart (and thus relatively increased myocardial oxygen consumption), and the need for higher levels of sedation and even muscle relaxation. Moreover, the anti-coagulation needs of ECMO are higher than that for LVADs. There is good evidence that patients supported with LVADs while waiting for a heart transplant have a better outcome than those supported with ECMO.

Common causes of heart failure in the pediatric population include congenital heart disease, cardiomyopathy (of which idiopathic is the most common), pulmonary hypertension, and myocarditis. Metabolic and genetic processes can also contribute.

There are **two types of LVAD** are available in the pediatric population: pulsatile and continuous flow devices. The most commonly used pulsatile device is the **Berlin EXCOR LVAD** and is suitable for all pediatric patients including neonates. It is available with several pumping chamber sizes (10-80 ml) and provides pulsatile flow delivered through a pneumatically driven thin membrane pump. In systole the pump moves compressed air into the diaphragm causing the ejection. In diastole negative pressure is generated to aid in the filling of the chamber. The maximum systolic positive pressure generated is 350 mmHg and the maximum negative driving pressure is (-) 100 mmHg. The pump rate can be adjusted between 30 – 150 beats/min and the systolic time can be adjusted between 20 – 70% of the cycle.

Importantly, the Berlin EXCOR pump is transparent, thus allowing visualization of filling, emptying and thrombus formation. If there is any thrombus formation in the pump or cannulas, the pump must be exchanged to avoid systemic embolization.

Continuous flow devices include axial devices like the **Heart Mate II** and centrifugal devices like the **Heart Ware**. Both devices are simpler, intra-corporeal and less thrombogenic but are limited by the patient size. Heart Mate II has only one moving component, with no valves, vent or compliance chamber reducing the device complexity. It is only available for patients with a body surface area $\geq 1.4 \text{ m}^2$. The Heart Ware device consists of a centrifugal blood pump integrated inflow cannula, an outflow graft, and a percutaneous driveline. This device has been used in patients $> 17 \text{ kg}$ but is not available for toddlers or infants.

The incidence of stroke in patients with Berlin EXCOR is high ($\sim 30\%$). Most patients are thus anticoagulated and may also be on antiplatelet agents. Anticoagulation and platelet inhibition is monitored with thromboelastography and platelet aggregometry (target inhibition of 30%). Unless bleeding is severe, anticoagulation is not reversed due to the risk of clotting the pump and systemic embolization.

The anesthetic considerations for LVAD patients include maintaining systemic vascular resistance (SVR) because of the relatively fixed cardiac output. Hypotension and hypertension are not well tolerated. Hypotension can be caused by hypovolemia, which is easily diagnosed by the lack of filling of the LVAD chamber, or by vasodilation (LVAD fillings is normal). Fluids and alpha-agonists should be first-line response to hypotension. Hypertension will increase the afterload and impede the device's ability to eject. This will be seen as incomplete emptying at the pump and heard via the high-pressure alarm. Hypertension should be treated by deepening anesthesia and/or by the judicious administration of vasodilators. As a temporary measure the measure the driving pressure can be raised, but this carries the risk of worsening hypertension with its attendant risks.

The **echo exam** of an adequately functioning LVAD should show minimal opening of the aortic valve, a decompressed LV chamber, and a midline inter-ventricular septal position. The inflow cannula should be aligned with the mitral inflow creating laminar flow. Right ventricular function and pulmonary vascular resistance can also be assessed by septal position, ventricular size, and the severity of tricuspid regurgitation.

Treatment of epistaxis can range from conservative management with external pressure and oxymetazoline, to more invasive nasal packing, to ultimately surgical cautery. Packing can be performed with gauze or a hemostatic agent like SURGIFLO® Hemostatic Matrix or SURGICEL®. Both are more effective in the presence of thrombin. Oxymetazoline, though useful in the control of bleeding, should be used cautiously because of its potential to cause significant hypertension. Anesthesia for epistaxis can vary from deep sedation for short procedure up to general anesthesia with endotracheal intubation for airway protection in profuse bleeding. Inhalation induction can be risky in patients with large amounts of swallowed blood because the risk of aspiration. A combination of midazolam and ketamine provides sedation, analgesia and amnesia without causing major changes in the LVAD hemodynamics.

The **postoperative management** of LVAD patients for minor procedures can be done in regular PACU. Pediatric anesthesiologist should become familiar with these devices since its use is becoming widespread.

4. REFERENCES:

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