Inability to ventilate? Child with history of hypoplastic left heart syndrome for a laceration repair with undiagnosed plastic bronchitis.

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Objectives:

1. Describe the anatomy and physiology of single ventricle palliative repair.
2. Describe the preoperative assessment of a patient with Fontan physiology.
3. Identify the goals of perioperative management of a patient with Fontan physiology.
4. Discuss the complications of palliative repair in patients with single ventricle physiology.

Case History:

A 5-year-old, 16kg male, presented for repair of a forehead laceration after a fall from a bunk bed. The child’s past medical history was significant for Hypoplastic left heart syndrome (HLHS) s/p staged palliation and currently s/p non-fenestrated Fontan procedure.

Questions:

What is a Fontan? What are the different stages of palliation for HLHS? How does the anatomy and subsequently physiology differ from a biventricular heart?

Case history and physical examination (continued)

Patient’s parents stated that he had been doing well from a cardiac standpoint. His oxygen saturation (SpO2) was usually 98% at home. He was able to play with kids his age but did fatigue easier than others. His home medications included aspirin and lisinopril. Patient had no complications with anesthesia after his cardiac surgeries. In our pre-operative area, the child was anxious and complaining of a headache. He had a 3cm right frontal laceration without any signs of active bleeding. His vital signs were notable for SpO2 93% on RA. Upon auscultation, the patient had a normal heart rate and rhythm. He exhibited effortless breathing and normal breath sounds in bilateral lung fields. No other signs or symptoms of trauma noted.

Questions:

Is the oxygen saturation relevant? What is your differential for relative hypoxemia in this child? Is this patient optimized for surgery? Do you need any other information prior to proceeding to the operating room (OR)? What are the anesthetic implications of this patient’s abnormal physiology? Would you involve any members of other services to optimize this patient? What tests would you order prior to proceeding to the OR? Would you delay the case if the tests were not immediately available?
Pre-operative studies:

Patient’s lab work showed normal electrolytes and a normal creatinine level. His hemoglobin was 13 g/dL with a normal platelet count. A TTE revealed non-obstructed cavopulmonary flow and good ventricular contractile function. No atrioventricular valve insufficiency or outflow tract obstruction noted.

Questions:

What do you think of these studies? What are the important aspects to consider when reviewing an echo report? Is there any other information you need prior to proceeding to the OR? What is your plan for induction? Maintenance? In addition to standard ASA monitors, do you need any other monitors? How are you going to ventilate this child?

Intraoperative care:

Patient was taken to the OR and general anesthesia was induced with etomidate and rocuronium after placement of standard ASA monitors. The child was uneventfully intubated, however, after intubation significantly diminished breath sounds were noted on the right. Normal breath sounds were noted on the left side. Patient was saturating 90-92% on 100% inspired oxygen.

Questions:

What would you do next? What is your differential? Can we let the surgeon proceed since we are maintaining oxygen saturations? Are we still ventilating the patient adequately? How do we know? Do we need to involve any other services’ help? Who would you consult?

Intraoperative care:

The endotracheal tube (ET) position was reassessed by direct laryngoscopy and was confirmed to be in the good position. The ET was suctioned with a flexible suction catheter with no improvement. Subsequently, fiberoptic bronchoscopy was performed through the ET. White material was noted in the right main bronchus. Otolaryngology (ENT) team was consulted intraoperatively for evaluation and possible bronchoscopy. ENT surgeons performed a rigid bronchoscopy and a large bronchial cast was retrieved, which improved breath sounds. Patient’s ET was suctioned numerous times for increased thin clear secretions. Patient’s head laceration was repaired uneventfully.

Questions:

Would you extubate the child at the end of the procedure? Why or why not? Where should the child recover? What are the immediate concerns in the post-operative period?

Post-operative care:
The child was transported intubated to the PICU secondary to copious secretions from the endotracheal tube. The child was extubated on post-operative day two and a diagnosis of plastic bronchitis was established based on pathology results.

Discussion:

Outcomes and survival of patients with congenital heart disease continues to improve, however, children with congenital heart disease undergoing non-cardiac surgery have an increased risk of morbidity, perioperative cardiac arrest and 30-day mortality. Numerous congenital cardiac anomalies cannot be repaired anatomically and undergo palliative repair. Single ventricle physiology refers to several conditions that are not amenable for biventricular repair and result in a single ventricle ejecting blood to the systemic circulation with passive pulmonary blood flow. Conditions such as Hypoplastic Left Heart Syndrome (HLHS), tricuspid atresia, pulmonary atresia (with intact septum), double inlet left ventricle, double outlet right ventricle, unbalanced AV canal and heterotaxy syndrome will frequently undergo palliation resulting in single ventricle physiology.

The single ventricle palliation is a 3-step staged procedure. The first stage involves the single ventricle ejecting blood to systemic circulation and parallel pulmonary circulation through an aorto-pulmonary shunt. In the second stage repair (Bi-directional Glenn), a cavo-pulmonary shunt redirects venous return from SVC into the pulmonary circulation. Pulmonary blood flow at this stage is passive and the ventricle is pumping blood systemically. The third stage (Fontan procedure) involves directing the venous return from IVC into the pulmonary circulation. After completion of third stage, the pulmonary and systemic circulations are separated and arterial oxygenation is normalized.

Even successfully palliated lesions can have significant degree of cardiac and other end-organ dysfunction. Progressive contractile dysfunction, arrhythmias, thromboembolism, pressure and volume overload are the most common residual effects of palliative repair.

Preoperative assessment of patients with a Fontan physiology should be aimed at obtaining information on original anatomy, type of surgical repair, post-operative complications, current medications, and need for optimization. Transthoracic echo and cardiac catheterization reports can help ascertain current anatomy, any residual shunts and various intracardiac pressures.

In patients with Fontan physiology, the target systemic venous pressure is 10-15 mm Hg and the appropriate atrial pressure is 5-10 mm Hg. This results in a “driving” hydrostatic pressure or transpulmonary gradient 5-8 mm Hg. Therefore, normal blood flow depends on unobstructed venous return, patent anastomotic connection, low pulmonary vascular resistance, and low intrathoracic pressure. In spontaneously breathing patients most of the blood flow through the lungs occurs during inspiration. Cardiac function (systolic and diastolic), normal
sinus rhythm, competence of the atrio-ventricular valve and absence of outflow obstruction are important determinants for the Fontan circulation at the level of the univentricular heart.

Intraoperative management of a patient with Fontan physiology presents more challenges and clinical dilemmas. Cardiac output is decreased by acute increases in pulmonary vascular resistance, arrhythmias and decreased transpulmonary gradient. Increased pulmonary vascular resistance will decrease ventricular filling and have negative effect on cardiac output. Hypoventilation in spontaneously breathing anesthetized patients may result in hypoxemia, hypercarbia, acidosis and atelectasis with net effect of increased pulmonary vascular resistance. In addition, surgical stress may also cause pulmonary vasoconstriction. On the other hand, excessive positive pressure ventilation with increased intrathoracic pressure and diminished blood flow though the lungs will increase pulmonary artery pressure and depress cardiac output and oxygenation. Most of the literature suggests careful use of positive pressure ventilation to maintain lung volumes and gas exchange. Increased risk of arrhythmias and subsequent negative effect on cardiac output may require prophylactic placement of defibrillating pads to treat arrhythmias expeditiously. Sinus node dysfunction and atrial reentry tachycardia are common cause of morbidity in patients with a univentricular heart. Careful assessments of intravascular volume helps to maintain adequate preload and transpulmonary gradient. Patients with Fontan physiology are at higher risk for bleeding because of increased venous pressure, presence of collateral vessels and thromboembolic prophylaxis with anticoagulation therapy. Use of inhalational agents also has to be judicious secondary to risk of myocardial depression and vasodilation.

Patients with Fontan physiology may require higher level of care with possible ICU admission even after “simple” procedures. Balance of pain control and adequate ventilation may necessitate postoperative ventilation, with careful assessment of the patients prior to extubation. Involvement of a cardiology consultant familiar with the patient’s anatomy and physiology is important for decision-making process in immediate postoperative period. Plastic bronchitis is an extremely rare complication after Fontan operation. Pathogenesis of this condition is not clear, however, this complication is potentially fatal and may require heart-lung transplantation.
Fontan circulation.

Common atrium → Single Ventricle → Aorta → SVR

Pulm veins ← PVR ← Pulm arteries ← SVC, IVC

Transpulmonary gradient = CVP – Common atrial pressure

References:


