Hypoplastic Ventricle, Hypertrophied Tonsils, Severe Apnea Hypopnea Index, What Next?

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Objectives: Describe the physiology of total cavopulmonary connection (Fontan)

Discuss anesthetic management goals for a patient with Fontan physiology

Describe the physiologic effects of morbid obesity and obstructive sleep apnea on patients with Fontan physiology

Formulate an anesthetic plan for patients with Fontan physiology presenting for non-cardiac surgery

Case Description:

16 year old male with severe sleep apnea by sleep study presents for tonsillectomy and adenoidectomy. Patient was born with hypoplastic left heart syndrome (HLHS) and underwent a series of cardiac staging procedures culminating with a Fontan procedure in 1999. His weight is 108 kg (body mass index or BMI of 44).

Questions:

What is a Fontan procedure? What are some of consequences of Fontan physiology? How will you assess whether this patient is appropriate and/or optimized for surgery? What information do you need from your history and physical to make this assessment? What events during a sleep study suggest severe sleep apnea? Do you want any other preoperative diagnostic or laboratory studies?

Preoperative assessment and studies:

The patient has had no recent illnesses. He is sedentary and does not exercise. He reports no dyspnea at the level of activity he engages in. He has no history of arrthymias or syncopal episodes. His baseline oxygen saturation fluctuates between the high 80's and low 90's. The patient takes aspirin and lisinopril and has been on those medications for some time with no recent adjustments in dose. On exam he is in no apparent distress and his lungs are clear to auscultation.

The patient's sleep study shows an apnea hypopnea index (AHI) of 125 events per hour. His oxygen saturation frequently dipped into the 60's and 70's. His nadir was 9% during the study. EKG shows normal sinus rhythm. A recent echocardiogram shows normal right ventricular function with trivial neoaortic regurgitation and stenosis, but is otherwise unremarkable for a child with history of HLHS with Fontan procedure.

Questions:

What are the consequences of untreated severe sleep apnea? Is there any relationship between obesity and sleep apnea in children? What are the consequences of untreated severe sleep apnea in a patient with Fontan physiology? What are the common treatment options for sleep apnea in children? Are the options any different for a patient with Fontan physiology?

Preoperative Discussion:

With untreated severe sleep apnea the patient's pulmonologist and cardiologist are concerned about the long term risk of pulmonary hypertension and subsequent right ventricular dysfunction. Noninvasive positive pressure ventilation (CPAP or BiPAP) is commonly used in the treatment of OSA. This is not without hemodynamic consequences, however. Some specialists suggest that if CPAP/BiPAP is considered in a patient with Fontan physiology, that it should be trialed under controlled conditions (in a cardiac catheterization laboratory), so that pulmonary blood flow, oxygen saturation, and cardiac output can be closely monitored.

Questions:

Do you wish to make any adjustments to this patient's medications prior to surgery? What instructions for preoperative fasting should this patient receive? Can the patient receive sedation prior to induction? Does this patient require prophylaxis against bacterial endocarditis? What type of induction is most appropriate (inhalational or intravenous)? What agents do you wish to administer for induction? Do you want to use a neuromuscular blocker for intubation? Does the patient need to be intubated with an endotracheal tube or is a laryngeal mask airway (LMA) preferable? How do you wish to maintain anesthesia? Is there an optimal intraoperative plan for ventilation? What is your plan for intraoperative and postoperative analgesia? Plan for emergence? Where should the patient be monitored postoperatively? Is he a candidate for ambulatory surgery?

Discussion:

The Fontan procedure or total cavopulmonary anastomosis is the final surgery for many patients with single ventricle lesions. This procedure is usually undertaken between age 2 and 6 depending on the preferences of the surgeon and/or cardiologist. The goal of the Fontan procedure is to divert the flow of blood from the inferior vena cava to the pulmonary arteries so that the majority of systemic blood return is directed to the lungs. Cardiac output is essentially dependent on pulmonary blood flow.

Congenital heart defects requiring a Fontan procedure may be a manifestation of a genetic disorder or syndrome (e.g. Trisomy 21, DiGeorge Syndrome, etc.). Important anesthetic considerations may apply to other organ systems as well. These patients have survived multiple cardiac surgeries and potentially prolonged complicated hospitalizations. Review of old records is critical in creating an appropriate anesthetic plan.

While there are general principles which should be applied to the anesthetic management of Fontan patients, there are no specific anesthetic agents which are recommended. Children with compensated Fontan physiology can safely undergo inhalational induction of anesthesia. Patients with ventricular

dysfunction, AV valve insufficiency, and decreased cardiac output should be induced with agents that have minimal myocardial depression.

Vascular access can be challenging in this patient population due to history previous interventions. Central venous access is tempting but presents a risk of thrombosis. Monitoring of the patient's blood pressure may be altered depending on whether one of the subclavian arteries was sacrificed or utilized for creation of Blalock-Taussig shunt during earlier procedures.

While spontaneous ventilation in patients with Fontan physiology promotes improved venous return and pulmonary blood flow, spontaneous ventilation under general anesthesia may be inadequate to prevent hypercarbia, hypoxemia, and atelectasis. Short inspiratory times, limited peak inspiratory pressure, low respiratory rates, and avoidance of excessive positive end expiratory pressure will help maintain adequate pulmonary blood flow as long as intravascular volume is maintained.

Maintenance of intravascular volume is critical to the Fontan patient, as they are preload dependent. If preload is decreased, pulmonary blood flow and cardiac output will follow. Minimizing fasting times is essential and preoperative intravenous hydration may be required if prolonged fasting is unavoidable. Fontan patients may require fluid volumes in excess of common requirements, especially if positive pressure ventilation is used. Due to elevated venous pressure, coagulation defects, venous collaterals and antithrombotic therapy perioperative bleeding is often increased. In Fontan patients, the risk of thrombosis must be carefully weighed against the risk of perioperative bleeding.

Arrhythmias are common after Fontan procedures. Maintenance of sinus rhythm is crucial, as non-sinus rhythms are usually very poorly tolerated. Adequate cardiac output in the single ventricle patient is dependent on the atrial contribution to ventricular filling. Bradycardia is poorly tolerated as the ability to increase stroke volume is significantly diminished.

With the advances in the diagnosis and treatment of pediatric congenital heart defects, a significant number of these patients are surviving beyond infancy and early childhood. Subsequently, many of these patients present for non-cardiac surgery as they mature and will require careful anesthetic planning.

References:

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