Esophagostroduodenoscopy (EGD)/Colonoscopy in a Patient with Failing Fontan Circulation

Can’t You Just Do a MAC?

Moderators:
Michelle Schlunt MD, Associate Professor of Anesthesiology, UCLA
Charles Lee MD, Pediatric Anesthesiologist, Sunset Kaiser Permanente

Objectives:
1. Review univentricular physiology and surgical palliation for the single ventricle patient
2. Understand medical management and surgical treatment for a patient with failing Fontan circulation
3. Review complications with pediatric esophagostroduodenoscopy
4. Discuss anesthetic management (MAC vs. GA) in a patient with failing Fontan circulation undergoing EGD/colonoscopy

Case History:
LM is a 14-yr-old patient scheduled for an add-on EGD/colonoscopy. She was admitted for a 7-day history of abdominal pain and diarrhea. Over the past 24 hours she has had three episodes of hematemesis. She is 34 kg and 148 cm tall. Her room air oxygen saturations are 73-85%. The pediatric gastroenterologist tells you she is adding a percutaneous liver biopsy to the EGD/colonoscopy since the patient is being worked up for possible heart transplantation. She isn't sure what the original heart problem was. It will be very quick.

Upon review of the patient’s medical records, you find she has a history of dextrocardia/tricuspid atresia/pulmonary atresia/hypoplastic right ventricle. She underwent extracardiac Fontan at five years of age and subsequent revision with fenestration at age 13 along with a MAZE procedure for atrial flutter. She most recently underwent interventional right pulmonary artery stent placement in the cath lab.
**Questions:**

What is Fontan physiology? What is an extracardiac Fontan? Are there other types of Fontan completion? Does it matter? What is the life expectancy with a single ventricle? Shouldn’t her oxygen saturations be higher? Do you need any further preoperative testing or labs?

Physical examination of the patient reveals a petite, cachectic girl with marked abdominal distention and breathing rapidly. A recent right/left heart catheterization shows a mean pulmonary artery pressure of 16 mm Hg and left ventricular end-diastolic pressure (LVEDP) of 9 mm Hg. A recent echo demonstrated good single ventricle function and a patent Fontan. However yesterday’s chest x-ray shows bilateral pleural effusions.

Medications: Lasix, ASA, Eplerenone, Sildenafil, Bosentan, Coumadin, Cholecalciferol

**Questions:**

Are these numbers good? What is the transpulmonary gradient? Is it important? What is causing the pleural effusions? Is her abdominal distention of concern or is just related to her diarrhea? Why is she being worked up for a heart transplant if she has good ventricular function on her echocardiogram? Why is it necessary to do a liver biopsy? What are the signs and symptoms of a failing Fontan circulation?

Her laboratory values from today are:

- **Na 139**  K 3.2  Cl 108  CO2 27  Bun 12  Cr 0.7
- **WBC 4.1**  HGB 12.4  HCT 39.6  PLTS 310
- **PT/INR 16.8/1.7**  PTT 29.4
- **ALP 54 (31-103)**  AST 26 (7-36)  ALT 9 (4-45)
- **Total Protein 3.5 (6.2-8.3)**  Alb 2.1 (3.7-5.1)

Her parents arrive and request no intubation due to her prior protracted time on the ventilator after her last surgery. The patient has had multiple prior surgeries and interventions. She and her parents are both extremely anxious.

**Questions:**

Do you agree with the parents? Is it necessary to intubate? Will you give premedication? What is your anesthetic plan if you proceed with just sedation per the parents’ request? Propofol? Ketamine? Dexmedetomidine? Any special monitoring needed? Does positioning affect the
Fontan circulation? Does this procedure need to be done in the operating room or is the GI suite fine? What are the possible procedural complications with EGD/colonoscopy? Do you need to give endocarditis prophylaxis? What is protein-losing enteropathy? How is it diagnosed and what causes it? How is it treated? Does it affect your anesthetic plan?

You proceed with propofol sedation and oxygen supplementation via nasal cannula. The gastroenterologist passes the endoscope, but the patient keeps coughing. The gastroenterologist complains she can't do the procedure under these conditions. The nurse alerts you the oxygen saturation is 67% now.

Questions:
Do you continue with sedation or proceed to GA and intubate? Can you just give narcotic to stop the coughing? Isn't spontaneous ventilation preferred for Fontan patients? What is plastic bronchitis?

You continue to proceed with sedation and avoid intubation. On EGD, gastric varices are noted but no apparent bleeding. Colonoscopy is then performed followed by the liver biopsy. Upon completion, oxygen saturation is 79% on supplemental oxygen via nasal cannula.

Questions:
Can she be recovered as usual in the GI suite? Can she go back to her room on the pediatric basic floor? Does she still need the supplemental oxygen? Is she at risk for bleeding from the percutaneous liver biopsy? What are the therapeutic options for a failing Fontan circulation? What are the outcomes following heart transplantation for failing Fontan circulation?

Discussion:
Fontan and Baudet first described their surgical treatment for tricuspid atresia in 1971. The ultimate goal was to create a pathway by which systemic venous blood would enter the pulmonary circulation bypassing the need for a right ventricle. The surgical procedure originally consisted of ligation of the main pulmonary artery, anastomosis of the superior vena cava (SVC) to the distal right pulmonary artery (PA), anastomosis of the right atrial appendage to the proximal right PA via an aortic valved homograft, closure of the atrial septal defect, and placement of a pulmonary valved homograft into the inferior vena cava (IVC). The “ventricularized” atrium would serve to support the pulmonary circulation. The systemic and pulmonary circulations were now in series driven by the single functional ventricle.

Since that time multiple modifications have taken place. Atriopulmonary connections are no longer desired, but rather total cavopulmonary connections, after demonstration of the
detrimental effects related to right atrial dilatation over time. These detrimental effects included turbulent flow with energy loss, thrombus formation, and arrhythmia development. The first stage of palliation at birth involves a systemic-to-pulmonary artery shunt, which inevitably results in volume overload for the univentricular heart. Volume overload results in a dilated, hypertrophied ventricle that is preload dependent. The second stage of palliation, the bi-directional Glenn shunt performed at four to six months of age, which involves an end-to-side anastomosis of the SVC to right PA, serves to partially volume unload the single ventricle. Staging allows for adaptation of the single ventricle to preload reduction and adjustment of the upper body systemic venous and lymphatic systems to the increase in venous pressure before Fontan completion. The Fontan, usually performed at 18 -24 months of age, remains the final surgery in the staged palliation approach for many forms of congenital heart disease with a functional single ventricle. Fontan completion results in complete separation of the systemic venous return to the pulmonary circulation. IVC flow is directed to the pulmonary circulation via an extracardiac conduit. The prior modification, the lateral tunnel which involved a baffle constructed in the lateral portion of the right atrium to direct IVC return to the pulmonary circulation has largely been abandoned. The advantages of the extracardiac conduit include improved laminar flow, minimal atrial suture lines, no intracardiac prosthetic material, and the ability to be performed without the use of cardiopulmonary bypass.

“Working” Fontan circulation depends on adequate systemic venous return, low pulmonary vascular resistance (PVR), a competent atroventricular valve, and a well-functioning systemic ventricle. Fontan physiology creates systemic venous hypertension with pulmonary arterial hypotension. Pulmonary blood flow is passive as there is not a subpulmonary ventricle present. The driving force or transpulmonary gradient is equal to the difference in central venous pressure minus the common atrial pressure. The ideal transpulmonary gradient is 7-8 mm Hg. The single ventricle alone must overcome the resistance of the systemic arterial, systemic venous and pulmonary circulation in series. Fenestration of the Fontan allows for preload delivery to the systemic ventricle should the PVR become prohibitively high, thereby limiting pulmonary blood flow and pulmonary venous return to the common atrium, however fenestration is no longer routinely performed.

Survival at 25 years post Fontan is 70%. Fontan physiology is not normal. There appears to be a slow late attrition rate following Fontan completion resulting in what many feel to be the inevitable, a failing Fontan circulation. This is most likely related to chronic systemic venous hypertension and decreased preload with low cardiac output. Symptoms develop at an advanced stage of declining Fontan circulation. Symptoms related to the failing Fontan circulation include fatigue, decreased exercise tolerance, volume retention, syncopal episodes, dyspnea and decreasing oxygen saturation. Normal expected oxygen saturation is 90-95% in a well-functioning non-fenestrated Fontan. Important causes of decreasing oxygen saturation in Fontan patients include: presence of a fenestration with its R→L shunt, venovenous or venoatrial collaterals, pulmonary arteriovenous malformations, pulmonary vein obstruction and pleural effusions.

Approximately 50% of patients have diastolic dysfunction 10 years post-Fontan, and 50% of patients over the age of 30 have signs and symptoms of heart failure. Risk factors for myocardial dysfunction are a morphologic systemic right ventricle and longer duration since
Fontan completion. Impaired diastolic relaxation results in an increase in LVEDP with a subsequent increase in left atrial pressure (LAP) that is eventually transmitted to the pulmonary vascular bed. An LVEDP >12 mm Hg indicates significant ventricular dysfunction. Therapy follows that of chronic biventricular heart failure: ACE inhibitors, calcium channel blockers, pulmonary vasodilators, beta-blockers and diuretics.

Failing Fontan circulation affects multiple organ systems with protein-losing enteropathy (PLE) being one of the most worrisome occurring in 3-15% of patients. This carries a 50% mortality rate at five years from the time of diagnosis. The mean time to onset of PLE following Fontan completion is seven years. The etiology is unclear, but is probably related to the chronic systemic venous hypertension and low cardiac output state. Chronic systemic venous hypertension is transmitted to the SVC, IVC and portal vein system. Increased abdominal venous pressures lead to intestinal congestion, obstruction of the lymphatic system, and enteric protein loss. Loss of albumin leads to a decrease in vascular oncotic pressure promoting peripheral edema, pleural and pericardial effusions, and ascites. Chronic diarrhea is related to edema of the bowel wall causing malabsorption. This ongoing protein loss also includes anticoagulant proteins promoting a hypercoaguable state, and a loss of immunoglobulins creating an immunodeficiency state. A low cardiac output state via compensatory mechanisms shunts blood away from the mesenteric circulation decreasing oxygen delivery that affects intestinal mucosal cell function. The low cardiac output state may also trigger inflammatory mediators such as α-tumor necrosis factor. Diagnosis is made by a low serum albumin without the presence of associated liver or renal disease and elevated α-1 antitrypsin fecal levels. Therapy is directed at increasing cardiac output: inotropic support, pulmonary vasodilators such as Sildenafil and Bosentan, possible fenestration of the Fontan conduit, multi-site pacing if not in sinus rhythm, and afterload reduction. Symptomatic therapies include corticosteroids to address the inflammatory component and possibly stabilize intestinal capillary and lymphatic membranes and unfractionated heparin as chronic intestinal vascular congestion interferes with the production of heparin sulfate (these glycosaminoglycans have been shown to regulate albumin losses). Additional treatment modalities include diuretics, exogenous albumin administration, calcium replacement, IV immunoglobulin replacement, and a high-protein diet. Successful treatment for protein-losing enteropathy occurs in only 25% of patients.

Hepatic venous pressure may be 3-4 times above normal following Fontan completion. Low cardiac output contributes to hepatocyte hypoxia leading to hepatic congestion with subsequent hepatic fibrosis and cirrhosis. There is a correlation with the degree of abnormal hepatic pathology and time from Fontan completion. The duration of hepatic fibrosis and cirrhosis may increase the risk of developing hepatic adenomas and hepatocellular carcinoma. Monitoring for hepatic changes includes liver transaminases, bilirubin and prothrombin time (PT)/international normalized ratio (INR) along with suggested serial imaging with contrast computed tomography or abdominal ultrasound. Liver biopsy still remains the gold standard for diagnosis.

Due to the lack of a subpulmonary ventricle, there is absence of pulsatile flow and low mean pulmonary artery pressures that underfill the pulmonary vascular bed increasing PVR over time. Pulsatile flow is important for the sheer-stress-mediated release of nitric oxide from the endothelium and recruitment of pulmonary capillaries to maintain their patency. Plastic
bronchitis is a rare complication occurring in <2% of Fontan patients. There is no clear etiology, but is possibly related to obstruction of lymphatic flow. Patients present with respiratory symptoms of persistent cough, wheezing and expectoration of bronchial casts formed by leakage of proteinaceous material into the airways. These bronchial casts may cause significant airway obstruction resulting in severe hypoxia. Therapy consists of pulmonary vasodilators, mucus-thinning agents such as acetylcysteine, bronchodilators, aerosolized tissue plasminogen activator to dissolve bronchial casts, and bronchoscopic lavage/removal of casts.

Maintenance of sinus rhythm is important to maintaining preload to the systemic ventricle. Ten years following Fontan completion over 50% of patients will develop arrhythmias. The most common arrhythmia is intra-atrial reentrant tachycardia. Factors promoting arrhythmogenesis are systolic/diastolic dysfunction of the single ventricle, atrioventricular valve regurgitation, atrial dilatation, intra-atrial tunnels, and prior multiple cardiac surgeries with resultant atrial scarring. Therapy involves anti-arrhythmic agents, radiofrequency ablation, resynchronization therapy, and AICD placement for associated malignant arrhythmias. Surgical options include right atrial reduction plasty, MAZE procedures and conversion to an extracardiac conduit.

Patients with Fontan physiology have abnormal coagulation profiles. Increased platelet reactivity and decreased levels of proteins C, S and anti-thrombin III lead to a hypercoaguable state. With failing Fontan circulation, distended and sluggish blood flow via the Fontan pathway serve to increase the thrombotic risk. Formation of silent pulmonary microemboli may detrimentally increase PVR. On the opposing side, anticoagulant abnormalities related to decreased levels of Factors V and VII showing a rise in the INR may also be present with hepatic dysfunction. The use of chronic anticoagulation for the predominant hypercoaguable state remains controversial, which may leave these patients at higher risk for subsequent stroke.

Surgical options for the failing Fontan physiology first attempt to address any correctable issues such as conduit stenosis, branch pulmonary artery stenosis, baffle leak or pulmonary vein obstruction. If these have been ruled out as a complicating cause for the failing Fontan, the next alternatives include take-down of the Fontan or conversion if a prior atrio pulmonary connection was performed. The final surgical option is orthotopic heart transplantation.

Approximately 2% of heart transplantations are performed for congenital heart disease. It is difficult to predict the proper time for pursuing heart transplantation for the failing Fontan. The presence of sequelae such as protein-losing enteropathy and plastic bronchitis despite preserved ventricular function has been associated with a decrease in survival following transplantation. Hence, earlier listing is encouraged. However, the shortage of donor hearts as well as prior allosensitization from multiple prior transfusions can make finding a suitable donor in a timely fashion very difficult.

Patients with a failing Fontan circulation present the anesthesiologist with a unique set of management issues. For preoperative evaluation, it is extremely helpful if the patient’s care has occurred primarily in your facility. Therefore records such as a recent catheterization report, cardiac magnetic resonance imaging or echocardiogram are readily available, as well as the patient’s cardiologist for further consultation. It is very important to have a comprehensive understanding of the patient’s cardiac defect, prior interventions, and their current functional
status. Unfortunately, the majority of the time this isn’t the case, and most of the medical records are not obtainable. Many of the patients and their families are not even sure of their actual diagnosis and the nature of the multiple interventions that may have occurred in the past. Sadly, this only becomes worse as the patients become older.

The main goals for anesthetic management involve: decreasing PVR, maintaining adequate preload, maintaining sinus rhythm, avoiding myocardial depression, and decreasing afterload. There is no specific anesthetic technique or agent recommended. The plan needs to be tailored to the individual patient taking into account the complexity of the proposed procedure. A preoperative physical exam and review of recent laboratory work should be done looking for abnormalities suggestive of Fontan dysfunction such as peripheral edema, clotting derangements and polycythemia associated with chronic cyanosis. The choice of sedation versus general anesthesia for EGD/colonoscopy should take into account the risk of aspiration, especially with insufflation and the presence of ascites in this particular patient. There is a 2.3% complication rate with pediatric EGD with cardiopulmonary events constituting 80% of the complications and gastrointestinal (GI) complications, such as GI bleeding, the second largest group. Two-thirds of the cardiopulmonary complications were related to hypoxia. Patients undergoing IV sedation for the procedure had a five times higher risk of cardiopulmonary complications compared to those receiving general anesthesia. Other identified risk factors for cardiopulmonary complications included: younger age, higher American Society of Anesthesiology classification, and female gender.

An intravenous (IV) induction for a patient with failing Fontan circulation is preferred as most of the induction agents cause a drop in preload and SVR and even myocardial depression that may require immediate resuscitation. However, intravenous access may be difficult as these patients have undergone multiple procedures in the past, and causing high anxiety in these patients will only serve to increase their PVR. Maintenance of spontaneous ventilation enhances systemic venous return to the pulmonary circulation, however spontaneous ventilation must be adequate to avoid hypoxia and hypercarbia that will increase PVR. If a patient is receiving positive pressure ventilation, then one should provide for short inspiratory times, lower peak airway pressures, 10-15 milliliters/kilogram tidal volumes and avoidance of excessive PEEP. End-tidal carbon dioxide (CO₂) monitoring underestimates arterial CO₂ in the presence of a R→L shunt.

The choice for invasive monitors depends to some extent on the anticipated complexity and length of the procedure. A diagnostic EGD/colonoscopy is very different from a lengthy endoscopic retrograde cholangiopancreatogram in the prone position that probably warrants an arterial line. The use of a central venous line is somewhat controversial due to the theoretical risk of thrombosis in the Fontan pathway. However, if there is a high likelihood that inotropic support will be needed for established ventricular impairment either during the procedure or postprocedure then it should be placed if access is available. The use of ultrasound may help to determine patency of central vessels that may have thrombosed in the past. The use of transesophageal echocardiography (TEE) intraoperatively is of substantial benefit as it gives a real-time evaluation of ventricular loading and function, however is not applicable in this particular case.
Hypovolemia is poorly tolerated in these patients. These patients appear to have a higher venous capacity and may have higher volume requirements than estimated from the usual paradigms. However, excessive volume loading is not well tolerated either and may further worsen ventricular dysfunction. It is beneficial to schedule these patients first in the morning or early placement of an IV to minimize dehydration issues.

Endocarditis prophylaxis is somewhat controversial since the new guidelines in 2007. EGD/colonoscopy is considered a low-risk procedure for bacteremia, yet in a survey of gastroenterologists, 50% administer endocarditis prophylaxis to patients with high-risk or moderate-risk for a low-risk procedure despite the opposing recommendations.

Regardless of the choice of sedation versus general anesthesia, these particular patients are at high risk for postprocedural complications. A patient such as this would not be eligible for outpatient status or be able to return back to the basic pediatric floor afterwards. Patients such as this need to be monitored for 24 hours in either a step-down intensive care unit (ICU) or actual ICU if step-down is not available. Continuous pulse oximetry monitoring provides early warning of ventricular dysfunction or respiratory depression related to postprocedural pain medications.

A patient with failing Fontan circulation presents many challenging issues to the anesthesiologist. It is imperative to have a thorough understanding of the interaction between our anesthetic techniques and single ventricle physiology. It is these “simple” and “quick” procedures where this knowledge is truly tested.

References: