Chapter 29

Extracardiac Fontan

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Case Scenario

A 15-year-old teenager presents for excision of a lipoma on her back. She was born with complex single-ventricle anatomy consisting of dextro-transposition of the great arteries, pulmonary atresia, and a hypoplastic left ventricle. As a neonate, she underwent placement of a modified Blalock–Taussig shunt. Subsequently, she had a bidirectional Glenn procedure at 6 months of age, followed by an extracardiac Fontan procedure at 2 years of age. She has been doing well at home and her cardiologist has been seeing her every 6 months. She reports some difficulty keeping up with her peers in gym class but no significant change over the past several years. Her current medications are aspirin and enalapril. Vital signs are heart rate 84 beats/minute, respiratory rate 18 breaths/minute, and SpO_2 95% on room air. Hemoglobin is 15 g/dL and all other laboratory values are within normal limits.

Echocardiography reveals:

- *Mildly depressed right ventricular function*
- *Mild tricuspid regurgitation*

Key Objectives

- Describe this patient's complex single-ventricle anatomy.
- Review the palliative surgeries that culminate in Fontan physiology.
- Describe the preoperative assessment of a patient with Fontan physiology.
- Review the intraoperative monitoring and management goals for a patient with Fontan physiology.
- Discuss postoperative disposition in patients with Fontan physiology having noncardiac surgery.

Pathophysiology

How can this patient's cardiac anatomy at birth be described?

The patient's cardiac anatomy and underlying physiology must be considered based on the two primary cardiac lesions: dextro (d)-transposition of the great arteries (d-TGA) and the hypoplastic left ventricle (LV).

- In d-TGA, a normal relationship exists between the heart's atrial and ventricular connections (*atrioventricular concordance*) but the ventricles and great arteries are incorrectly connected (*ventriculoarterial discordance*). (See Chapter 21.) Blood from the superior and inferior vena cavae (IVC) enters the right atrium (RA), passes through the tricuspid valve into the right ventricle (RV), and then into a right-sided and anterior aorta. On the other side of the heart, the left atrium (LA) connects to the LV via the mitral valve. From there, blood passes into the left-sided, posterior pulmonary artery.
  - With d-TGA blood flow in the pulmonary and systemic circulations exists in parallel (deoxygenated blood incorrectly flows to the systemic circulation and oxygenated blood returns to the pulmonary circulation, which is incompatible with life) rather than in series as normally occurs.
  - For the infant to survive after birth, a connection between the two circulations, such as a patent foramen ovale, atrial septal defect (ASD) or ventricular septal defect, must be present to allow for mixing and delivery of oxygenated blood to the rest of the body.
  - If an inadequate atrial connection is present, an urgent balloon atrial septostomy must be performed to create an ASD. This is typically performed either in the intensive care unit (ICU) with ultrasound guidance or in the cardiac catheterization lab with fluoroscopy. The femoral vein is accessed and a balloon-tipped catheter is passed into the RA, across the foramen ovale, and into the LA. With the balloon inflated, the catheter is then forcefully withdrawn back into the RA, thereby increasing the size of the atrial septal connection and allowing for improved mixing of atrial blood.
- This patient was also born with pulmonary atresia and a hypoplastic LV, which means that the main pulmonary artery and the LV were underdeveloped. Pulmonary atresia anatomy varies and can range

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from a fused trileaflet valve to a long segment of atresia, with all variations causing complete pulmonary outflow tract obstruction requiring an alternate source of pulmonary blood flow (PBF), which in this case was the patent ductus arteriosus (PDA). The PDA supplied blood to the pulmonary arteries from the aorta. Generally a prostaglandin (PGE_1) infusion is required to maintain patency of the PDA until a surgical source of blood flow (such as a modified Blalock–Taussig [mBT] shunt) can be created. Hypoplasia of the LV means that the LV cannot support cardiac output (CO) and the patient therefore will require single-ventricle palliation. Because of the transposition, the aorta originated from the RV, the only functional ventricle for this infant.

What was the first step in cardiac surgical palliation for this patient?

During the neonatal period, this patient required an mBT shunt, an artificial tube graft sewn between the subclavian or innominate artery and the pulmonary artery in order to supply blood flow to the pulmonary arteries. The patient's PDA was then ligated. This procedure may or may not require cardiopulmonary bypass (CPB) depending on individual patient factors. Because the aorta was of normal size, this patient did not require a Norwood procedure (which would have included aortic reconstruction) for her first surgery; she required only an mBT shunt to provide PBF.

Clinical Pearl

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What are the benefits of an mBT shunt and how does it affect pulmonary blood flow?

Anatomically, the mBT shunt is similar to the PDA in that it provides blood flow to the pulmonary arteries. Unlike a PDA, it provides a stable source of PBF that is not dependent on a prostaglandin infusion. The mBT shunt size is usually 3.5–4.0 mm, depending on patient size and the size of the shunt relative to the size of the aorta and pulmonary arteries. The length of the mBT shunt and the location of the arterial anastomosis (subclavian vs. innominate) also determines the amount of PBF provided. As pulmonary vascular resistance (PVR) is lower than systemic vascular resistance (SVR), this promotes PBF through the mBT shunt while limiting pulmonary overcirculation. Ultimately, the goal is to have a balanced Q_p:Q_s (clinically indicated by an oxygen saturation of 75%–85%) and to have flow to both pulmonary arteries allowing for symmetrical growth. (See Chapter 1 for Q_p:Q_s discussion.)

The patient's arterial oxygen saturation is determined by the oxygen saturation of systemic venous blood returning to the RA (mixed venous saturation), the oxygen saturation of pulmonary venous blood entering the RA across the ASD, and the amount of flow returning to the heart from both of these circulations. If an adequate ASD is not present, it will be necessary to perform either a balloon atrial septostomy or an atrial septectomy to ensure adequate mixing of oxygenated and deoxygenated blood. Unlike a balloon atrial septostomy, an atrial septectomy requires the use of CPB.

Clinical Pearl

Factors determining the patient's arterial oxygen saturation include the saturation of systemic venous blood returning to the right atrium (mixed venous oxygenation), the saturation of pulmonary venous blood entering the right atrium across the ASD, and the amount of flow returning to the heart from both of these circulations.

What monitoring concerns exist in patients who have had mBT shunts?

After the mBT shunt is taken down there can subsequently be distortion, stenosis, or aneurysmal dilation at the site of the proximal shunt insertion on the subclavian artery which could limit blood flow through the vessel. More rarely, the subclavian artery is sacrificed when the mBT shunt is taken down. Thrombosis of the artery is also possible. In each case, blood pressure measurements may not be accurate in the ipsilateral arm. Usually it is best to avoid the affected arm, but if it is necessary to use the ipsilateral arm for blood pressure measurements, comparison with measurements on other extremities is advised.

Clinical Pearl

In a patient with a history of an mBT shunt, blood pressure measurements may not be accurate on the affected side due to distortion, stenosis, or thrombosis of the subclavian vessel. This applies to both noninvasive and direct arterial blood pressure measurements.

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What is a bidirectional Glenn procedure and what criteria must be met before proceeding with it?

The bidirectional Glenn is also known as a ***superior cavopulmonary anastomosis*** (SCPA) wherein the superior vena cava (SVC) is anastomosed to the pulmonary artery (PA); this is usually the second stage of single-ventricle palliation. (See Chapter 27.) This procedure is performed using CPB. To allow time for PVR to decrease from the higher levels seen in neonates, a BDG is usually not performed before 3 months of age. Low PVR is essential for the success of the BDG, as PBF will now be passively supplied via the SVC–PA anastomosis.

Once on CPB, the mBT shunt is taken down. The SVC is disconnected from the RA and reanastomosed to the right pulmonary artery so that all venous flow from the upper body, including cerebral and upper extremity drainage, bypasses the heart and passively drains directly into the pulmonary circulation. Because the pulmonary arteries are in continuity, both lungs are supplied with blood as it flows from the SVC into the right and left pulmonary artery, hence the term "bidirectional."

What are the expected oxygen saturations and hematocrit for patients following a BDG?

When the BDG is performed the previous mBT shunt is replaced with a new shunt: the SVC-to-PA anastomosis. Because deoxygenated blood from the IVC returns to the common atrium and mixes with oxygenated pulmonary venous return, the patient's oxygen saturations do not change significantly after the BDG and remain 75%–85%. This persistent cyanosis triggers a compensatory increase in red blood cell production and oxygen carrying capacity leading to expected hematocrits of 40%–45%.

What is the Fontan procedure?

The Fontan procedure, also known as a ***total cavopulmonary anastomosis***, is the final palliative stage for patients with single-ventricle physiology. In this procedure IVC blood flow is baffled to the pulmonary arteries and therefore, given the previous BDG, all systemic venous blood return now bypasses the RA and flows directly into the pulmonary circulation.

In this era the Fontan procedure is most commonly performed using an extracardiac conduit that consists of a tubular GORE-TEX® graft directing IVC flow to the pulmonary arteries; thus all systemic venous return is now passively directed to the pulmonary circulation. Pulmonary blood flow then returns from the lungs to the heart via the pulmonary veins, passes through the common atrium into the single RV, and (owing to the d-TGA) is pumped out of the aorta. The pulmonary and systemic circulations are now separated and a series circulation exists. Pulmonary blood flow depends on the pressure gradient between the central venous pressure (CVP) and the pressure in the common atrium, which is called the transpulmonary pressure or ***transpulmonary gradient (TPG)***. This pressure difference allows blood flow to move through the lungs without the assistance of the pumping action of the heart. (See further details of single-ventricle palliative procedures in Chapters 26, 28, and 30.)

Clinical Pearl

After a Fontan procedure PBF depends on the pressure gradient between the central venous pressure and the pressure in the atrium, which is called the transpulmonary gradient. This pressure difference allows blood flow to move through the lungs without the assistance of the pumping action of the heart.

What surgical techniques can be used for the Fontan procedure?

Two main types of Fontan procedures are currently performed by surgeons, either the lateral tunnel or the extracardiac conduit, both of which result in similar physiology.

- The ***lateral tunnel Fontan*** is usually preceded by a hemi-Fontan procedure (HF) which is a modification of the BDG. The HF incorporates part of the RA into the connection between the SVC and the PA but is otherwise physiologically similar to a BDG. An advantage to the HF for the lateral tunnel Fontan sequence is that after the atrial anastomosis during the HF, creating the lateral tunnel baffle in the RA is often a more straightforward and shorter procedure than creating an extracardiac conduit. The lateral tunnel Fontan allows for direct placement of a fenestration allowing flow between the Fontan pathway and the common atrium. (See Chapter 28.)
- In an ***extracardiac Fontan***, the RA is bypassed completely and all systemic venous flow (SVC and IVC) is directed to the pulmonary arteries. An advantage to the extracardiac Fontan is that since the atrium is not part of the connection and thus does not see significantly elevated central pressures, the incidence of atrial arrhythmias may be lower than observed with the lateral tunnel Fontan. Additionally, there is minimal suturing of atrial tissue in the

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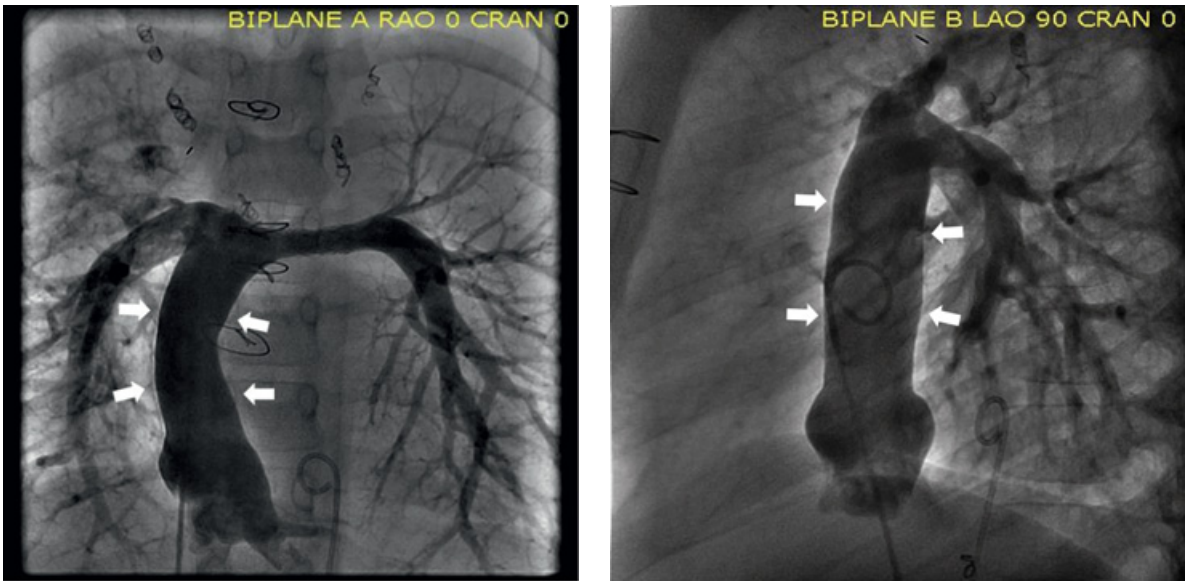

Figure 29.1 (A, B) Extracardiac Fontan. An angiogram is performed in the AP and lateral projection in the lower aspect of the extracardiac Fontan connection (white arrows). The Fontan connection is tube-like and smooth walled. Courtesy of Russel Hirsch, MD.

extracardiac conduit, which may further decrease the risk of arrhythmias. (See Figure 29.1A and B.)

Clinical Pearl

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What is a fenestration and what potential advantage does it offer?

In a patient with a newly created Fontan pathway, it is essential that PVR remain low. If the pulmonary impedance increases, this can result in a reduction of blood flow through the pulmonary vascular bed. A fenestration is a surgically created 4 mm hole in the lateral tunnel baffle, created to allow systemic venous blood to "pop-off" into the atrium when pulmonary impedance increases, resulting in a right-to-left shunt. This "pop-off" helps to maintain CO in the presence of higher resistance in the pulmonary bed, but it can also cause arterial desaturation due to the blood moving from the right (deoxygenated) side to the left (oxygenated). Once the pulmonary vascular bed has adapted to the new physiology, the fenestration can be closed in the cardiac catheterization lab, with the timing dependent on the patient's clinical condition.

While the fenestration is straightforward to place when creating a lateral tunnel Fontan, it is slightly less so in the extracardiac conduit and hence the intra/extracardiac conduit Fontan procedure was developed [1]. An extracardiac conduit is used so the atrium is not subjected to the higher pressures that can be seen with the lateral tunnel Fontan. However, in some patients, a small portion of the atrium is connected to the conduit and called a fenestration. Importantly, this fenestration is not placed near the sinus node, which decreases the risk of atrial arrhythmias.

Clinical Pearl

A fenestration in a lateral tunnel Fontan can behave as a "pop-off" by allowing right-to-left shunting from the IVC to the atrium. This helps to maintain CO in the presence of higher resistance in the pulmonary bed but can cause arterial desaturation due to the blood moving from the right (deoxygenated) side to the left (oxygenated).

What should the patient's expected oxygen saturations be if a fenestration is open? If it is closed?

With Fontan physiology and an open fenestration, arterial oxygen saturations can be expected to be in the range of 80%–90% depending on the degree of shunting occurring.

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Without a fenestration (as in this patient) or once the fenestration is closed, saturations should be above 90%.

Clinical Pearl

Patients with a fenestrated Fontan can have oxygen saturations in the range of 80%–90%. Once closed, oxygen saturations should be near normal, in the upper 90s.

What are the considerations regarding PVR in a patient with Fontan physiology undergoing noncardiac surgery?

In a patient with a Fontan palliation, CO must traverse the pulmonary vascular bed without the advantage of a pumping ventricle, which makes maintenance of low PVR critical. Elevations in PVR can compromise both oxygen saturation and CO. Intraoperatively, it is important to avoid hypoxia, hypercarbia, hypothermia, acidosis, and inadequate anesthesia, in order to keep PVR low and maintain flow through the Fontan pathway. Priority should be given to correcting respiratory or metabolic acidosis, as pH is a potent effector of pulmonary vascular tone. In patients with a fenestrated Fontan, oxygen saturation is a simple method to evaluate the degree of right-to-left shunt, as more flow across the fenestration (which could indicate increased PVR) will lead to more systemic oxygen desaturation. Elevations in PVR are not well tolerated in patients with Fontan physiology and should be minimized.

Clinical Pearl

Elevations in PVR can compromise both oxygen saturation and CO as CO must traverse the pulmonary vascular bed without the advantage of a pumping ventricle.

What potential issues can develop in patients with long-standing Fontan physiology?

Long-term issues can develop in patients with Fontan physiology and the likelihood of complications increases with age. Potential issues include Fontan failure, end-organ failure, arrhythmias, and thrombosis. ***Atrial arrhythmias*** are common in Fontan physiology, and the incidence increases with age. This may be due to stretching of the atrium secondary to both high pressures and the proximity of the Fontan connection to the sinus node. Older Fontan patients may require ablation procedures or require a pacemaker or implantable cardioverter-defibrillator.

Significant risk exists for the development of ***vascular thrombosis*** secondary to potentially poor cardiac function, dilated atria, nonpulsatile PBF, arrhythmias, and coagulation abnormalities. Many patients are on antithrombotic medications, such as aspirin, which may also contribute to perioperative coagulation abnormalities.

About 70% of patients with Fontan circulation develop ***myocardial dysfunction*** and failure at the 10-year mark as the systemic ventricle becomes dilated, hypertrophic, and hypocontractile, with deterioration in both systolic and diastolic function. A patient with Fontan failure may have signs of chronic systemic venous congestion affecting multiple organ systems, including hepatomegaly, splenomegaly, ascites, and exercise intolerance. Fontan failure may present as ***protein-losing enteropathy (PLE)***, in which patients lose proteins and coagulation factors through their intestinal tract resulting in severe diarrhea, electrolyte abnormalities, hypercoagulability, hypocalcemia, hypomagnesemia, and ascites. About 13% of patients have PLE at 10-year follow-up, and the prognosis is poor, with an approximately 60% five-year survival after diagnosis [2]. ***Plastic bronchitis*** *is* another manifestation of Fontan failure, in which mucofibrinous bronchial casts form in the airways and can cause potentially life-threatening airway obstruction. Additionally, end-organ damage to the liver, kidneys, or lungs can develop separately from overt heart failure. This end-organ damage is thought to be multifactorial and may be secondary to high central venous pressures, poor tissue oxygenation, and/or lack of pulsatility of PBF. (See Chapter 30.)

Clinical Pearl

Older patients with Fontan physiology are at increased risk for long-term complications including decreased CO, hepatic dysfunction, atrial arrhythmias, and thrombosis. Preoperative evaluation should evaluate for these sequelae. This includes review of a recent echocardiogram and electrocardiogram, as well as laboratory testing including coagulation studies.

Anesthetic Implications

What fasting instructions should be given to the patient and family?

The American Society of Anesthesiologists (ASA) guidelines for preoperative fasting should be followed in all patients, including those with Fontan physiology. These guidelines mandate that the fasting times for children be 6 hours for formula and solids, 4 hours for breast milk, and

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2 hours for clear liquids. In patients with Fontan circulation, intravascular volume is a major determinant of central venous pressure and flow through the pulmonary vasculature, thus hypovolemia from prolonged fasting is poorly tolerated. Patients, with help from their families, should be strongly encouraged to continue clear liquids until 2 hours before the start of the anesthetic. This often necessitates that the procedure be scheduled as first case of the day to minimize schedule delays. If fasting is prolonged or case start time is unpredictable, consideration should be given to preoperative intravenous (IV) fluid administration.

Clinical Pearl

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Should this patient's enalapril and aspirin be withheld preoperatively?

Many patients with Fontan palliation take angiotensin converting enzyme inhibitors (ACEi) for afterload reduction of the systemic ventricle. These medications have been associated with hemodynamic lability, refractory hypotension, and vasoplegia under anesthesia, especially in the adult population [3, 4]. The half-life of enalapril is <12 hours; it is generally advised to hold the morning dose on the day of surgery. The decision whether to discontinue this medication before a procedure should be carefully considered and discussion with the patient's cardiologist may be useful. If the procedure is emergent or the patient did not follow instructions to hold the morning dose, the case would usually not be delayed or cancelled. However, vasopressin (0.04 units/kg intravenously or 0.5 milliunits/kg/minute infusion) should be readily available to counteract refractory hypotension.

Patients with Fontan circulation are often on antiplatelet agents or anticoagulants due to their increased risk for thromboembolism. Depending on the anticipated procedure, antiplatelet therapy may be temporarily withheld perioperatively. Bridging therapy with a shorter acting anticoagulant may be needed for patients on warfarin until treatment can be continued, with the timing depending on the procedure. The precise timeline for discontinuation and reinitiation of therapy should include a thoughtful discussion and agreement between the surgeon, cardiologist, and anesthesiologist.

What preoperative testing is required for this patient?

Due to the multisystem organ effects of Fontan physiology, a comprehensive metabolic panel within a year of the scheduled procedure is advisable in Fontan patients. In addition, a complete blood count should also be reviewed, recognizing that high hemoglobin and hematocrit are consistent with chronic cyanosis. At a minimum a 12-lead electrocardiogram (ECG) should be performed to assess rhythm disturbances, and a recent echocardiogram should be available to assess pathways, and valvular and cardiac function within 12 months of the surgical date. Repeat studies would be warranted prior to surgery if the patient has developed new symptomatology or a change in exercise tolerance in the interim.

Clinical Pearl

Repeat studies would be warranted prior to surgery if the patient has developed new symptomatology or a change in exercise tolerance in the interim since the last study.

Should a cardiology visit be scheduled preoperatively?

Most Fontan patients are closely followed by their cardiologist. Depending on the patient's functional status and the nature and invasiveness of the planned surgery, no additional testing may be required preoperatively. However, if the patient has been lost to follow-up or has not visited their cardiologist recently a careful history and physical to determine functional status is crucial. If evidence of decreased exercise tolerance or other concerning cardiac signs or symptoms are found in a patient with minimal or limited cardiology follow-up, the surgery should be rescheduled until a patient has seen a cardiologist. Ideally all patients with Fontan physiology having an anesthetic should have been seen by a cardiologist within the past 12 months. In patients who have routine cardiology visits, the cardiologist should be informed of the planned procedure so they have the opportunity to convey any concerns not made immediately evident by imaging and laboratory studies.

Clinical Pearl

If evidence of decreased exercise tolerance or other concerning cardiac signs or symptoms are found in a patient with minimal or limited cardiology follow-up, the surgery should be rescheduled until a patient has seen a cardiologist. Ideally all patients with Fontan physiology undergoing an anesthetic should have been seen by a cardiologist within the past 12 months.

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What aspects of the echocardiogram are most important to review?

The anesthesiologist should review the most recent echocardiogram or magnetic resonance imaging study, ideally dated within 6–12 months of the planned procedure. During review of the echocardiogram, one should note systemic ventricular systolic and diastolic performance, atrioventricular and aortic valve function, atrioventricular or aortic valve obstruction, ventricular outflow obstruction including coarctation, patency of the Fontan pathway, pulmonary artery patency, and atrial septal communication. Particular attention should be focused on the function of the single ventricle.

Can or should this patient have this procedure done at a freestanding outpatient center?

An elective procedure under anesthesia in a patient with Fontan physiology should ideally be performed at a hospital with appropriate personnel, equipment, and resources in the event of an adverse event. This would include clinicians who understand the physiology and implications of a Fontan circulation. In most circumstances, this would not be at a freestanding center.

Is prophylaxis for infective endocarditis indicated for this procedure?

Infective endocarditis (IE) prophylaxis may be required in a patient with Fontan circulation presenting for noncardiac surgery if they have a residual lesion. If this patient does not have such lesions, she would not require IE prophylaxis.

What monitoring should be recommended?

Standard monitoring as recommended by the ASA guidelines, including pulse oximetry, 5-lead ECG, noninvasive blood pressure, temperature, and end-tidal carbon dioxide (CO_2) monitoring are highly recommended for all cases in patients with Fontan circulation. This patient should only require standard monitors for this particular surgery.

Clinical Pearl

Standard ASA monitors with a 5-lead ECG should provide an adequate level of monitoring for patients undergoing low or moderate risk surgery. Patients with a poorly functioning Fontan circulation who are undergoing high-risk surgery should have an arterial line and central venous catheter placed for continuous blood pressure monitoring, assessment of volume status, and potential delivery of inotropic agents.

When might more invasive monitoring might be recommended?

When a Fontan patient has decreased ventricular function or is undergoing major surgery, particularly where significant volume shifts are expected, invasive arterial and CVP monitoring are strongly recommended. An arterial pressure line allows for continuous blood pressure monitoring and blood gas sampling. Monitoring the CVP trend can aid in assessment of volume status. It is important to remember that a central venous catheter with the tip in the SVC will not measure ventricular filling pressure, but instead will measure mean PA pressure since the SVC is connected to the PA in a patient with Fontan physiology. Obtaining a ventricular filling pressure during a noncardiac procedure in this patient will be impossible since there is no percutaneous method of accessing the atrium in a Fontan patient. Central venous pressure trends are followed as a reasonable alternative, recognizing that CVP in a Fontan patient is higher than a typical CVP in a patient with a normal heart.

Clinical Pearl

A central venous catheter with the tip in the SVC will not measure ventricular filling pressure but instead will measure mean pulmonary artery pressure, since the SVC is connected to the pulmonary artery in a patient with Fontan physiology.

What is the TPG and what is its relationship to the CVP in a Fontan patient?

In patients with a Fontan circulation, the TPG is the primary driving force promoting PBF and subsequent CO. The TPG is defined as the difference between CVP and pulmonary venous atrial pressure. In a well-compensated Fontan, CVP is usually 10–15 mm Hg and pulmonary venous atrial pressure is usually 5–10 mm Hg, giving an ideal TPG of about 5–8 mm Hg. In addition to CVP trend, transesophageal echocardiography can be used for intraoperative assessment of ventricular preload and function as well as to monitor for the presence of emboli.

Clinical Pearl

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What are the anesthetic goals for this surgery in this patient?

The main goals for anesthetic management in a patient with well-functioning Fontan physiology are the following:

- Maintain adequate preload: replace fasting volume appropriately and keep abreast of third space loss and blood loss.
- Avoid agents that significantly depress myocardial contractility.
- Maintain sinus rhythm.
- Avoid increases in PVR.

What are the options for induction of anesthesia in this patient?

Given the patient's age and physiology a peripheral IV line should be placed for induction of anesthesia. Induction agents with minimal effect on myocardial contractility, CO and PBF should be selected. Etomidate and ketamine are both good choices for induction as they preserve myocardial contractility and vascular tone with little to no effect on PVR. Propofol should be administered judiciously, as it may cause both profound decreases in venous return and myocardial depression. If a muscle relaxant is necessary, avoid those with histamine-releasing properties, as they can result in hypotension and tachycardia. Rocuronium, vecuronium, cisatracurium, and succinylcholine are all good selections because of their stable hemodynamic properties. One must also be cognizant during induction that airway obstruction and/or difficult ventilation or intubation can lead to acute elevations in partial pressure of CO_2 which can increase PVR and negatively impact CO.

What are the effects of spontaneous ventilation on Fontan physiology? What are the considerations for this patient for this procedure?

Since PBF is dependent on passive venous drainage from the SVC and IVC into the pulmonary circulation, any ventilation strategy impeding intrathoracic venous return will have a negative effect on PBF and subsequently CO. During spontaneous ventilation, negative intrathoracic pressure increases antegrade flow from the SVC, IVC, and hepatic venous circulation into the pulmonary arterial tree, allowing more blood to be available for gas exchange. The benefits of spontaneous ventilation must be compared to the risks of hypoventilation and hypercarbia under anesthesia with sedatives. For this surgery, the patient will need to be prone; therefore spontaneous ventilation either with or without a definitive airway device may be contraindicated. A discussion with the surgeon is warranted to determine the length of the procedure and if, in this case, the position can be altered from prone to lateral. A short procedure or a change in position from prone may make spontaneous ventilation a better option for this patient.

Clinical Pearl

Since PBF is dependent on passive venous drainage from the SVC and IVC into the pulmonary circulation, any ventilation strategy impeding intrathoracic venous return will have a negative effect on PBF and subsequently CO.

What are the effects of positive pressure ventilation on this patient physiology?

Positive pressure ventilation (PPV) via an endotracheal tube can increase intrathoracic pressure, diminish venous return, PBF, and CO in patients with Fontan physiology. High peak inspiratory pressures and high positive end-expiratory pressures (PEEP) can decrease venous return and inhibit PBF. If PPV is indicated because of patient or procedural factors, the ventilatory goals should be as follows: low tidal volumes (6–8 mL/kg), low respiratory rates, short inspiratory times, and low (physiologic) PEEP. This ventilation strategy will optimize Fontan flow and CO.

What considerations are important during the maintenance phase of anesthesia?

Preservation of CO is a key goal during the maintenance of anesthesia. This is accomplished by ensuring adequate preload for good ventricular filling, minimizing agents which decrease contractility, and avoiding increases in PVR which can compromise ventricular filling and CO. Hypoxia, hypercarbia, acidosis, hypothermia, inadequate analgesia or anesthesia, excessive mean airway pressure, and PEEP can all increase PVR and should be minimized or avoided. A balanced anesthetic is ideal for maintenance. A low concentration of inhalational agent in conjunction with opioids, benzodiazepines, and/or dexmedetomidine should provide hemodynamic stability.

Is regional anesthesia feasible or useful in patients with Fontan physiology?

The risk-benefit profile of regional anesthesia should be examined critically and executed with caution. Close attention

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should be paid to any existing coagulopathy as well as the use of antiplatelet agents or anticoagulants. Epidural anesthesia has been successfully employed in patients with Fontan circulation; however, a spinal anesthetic is not recommended due to the hypotension and bradycardia that may accompany the sympathectomy occurring with spinal local anesthetics. Adequate monitoring and appropriate fluid administration are recommended if a neuraxial procedure is performed [5].

What are the options for treatment if pulmonary artery pressures acutely increase intraoperatively?

Treatment for an acute increase in PA pressure in a patient with Fontan physiology under anesthesia includes administration of 100% oxygen, fluid, and pulmonary vasodilators such as inhaled nitric oxide and inhaled prostacyclin analogues.

Are patients with Fontan circulation candidates for laparoscopic surgery?

Patients with a Fontan circulation have successfully undergone laparoscopic abdominal surgery. Key goals include keeping intraabdominal pressure during insufflation at or <10 mm Hg, ensuring adequate ventilation, and maintaining intravascular volume during the procedure. Potential complications due to the induced pneumoperitoneum include hypercarbia, gas embolism, hypotension, pneumothorax, and mediastinal emphysema. Additionally, if there is a fenestration in the Fontan circuit, there is a risk of paradoxical CO_2 embolism. If unrecognized, these complications may be potentially catastrophic. With careful management, most patients with a well-functioning Fontan circulation can tolerate laparoscopic abdominal surgery, which allows them to benefit from the reduced postoperative pain and recovery time compared with open surgery.

What is the appropriate postoperative disposition in this patient after this surgery?

This patient, with a well-functioning Fontan and minimal comorbidities, should be able to have outpatient surgery. She can be discharged home after an uneventful anesthetic once she is tolerating fluids by mouth, pain issues are addressed, and when she meets post-anesthesia care unit discharge criteria. If any instability or adverse events occur during the anesthetic, consideration must be given to admitting her for further observation, depending on the nature of the event.

What is the long-term prognosis for this patient?

With improvements in surgical techniques and overall management of patients following the Fontan procedure, long-term survival continues to improve as well. However, overall survival for patients with Fontan physiology is significantly lower than that of the general population due to the development of complications as described above. Recent reports indicate that approximately 74% of Fontan patients will survive without transplantation and the 30-year transplant-free survival rate is only 43% [2].

References

1. R. A. Jonas. The intra/extracardiac conduit fenestrated Fontan. *Semin Thorac Cardiovasc Surg Pediatr Card Surg Annual* 2011; **14**: 11–18.
2. A. Kay, T. Moe, B. Suter, et al. Long term consequences of the Fontan procedure and how to manage them. *Prog Cardiovasc Dis* 2018; **61**: 365–76.
3. C. C. Ajuba-Iwuji, S. Puttreddy, B. G. Maxwell, et al. Effect of preoperative angiotensin-converting enzyme inhibitor and angiotensin II receptor blocker use on hemodynamic variables in pediatric patients undergoing cardiopulmonary bypass. *World J Pediatr Congenit Heart Surg* 2014; **5**: 515–21.
4. C. Hollmann, N. L. Fernandes, and B. M. Biccard. A systematic review of outcomes associated with withholding or continuing angiotensin-converting enzyme inhibitors and angiotensin receptor blockers before noncardiac surgery. *Anesth Analg* 2018; **127**: 678–87.
5. M. Tiouririne, D. G. De Souza, K. T. Beers, et al. Anesthetic management of parturients with a Fontan circulation: a review of published case reports. *Semin Cardiothorac Vasc Anesth* 2015; **19**: 203–9.

Suggested Reading

Kiran U., Aggarwal S., Choudhary A., et al. The Blalock and Taussig shunt revisited. *Ann Card Anaesth* 2017; **20**: 323–30.

McClain C. D., McGowan F. X., and Kovatsis P. G. Laparoscopic surgery in a patient with Fontan physiology. *Anesth Analg* 2006; **103**: 856–8.

Nayak S. and Booker P. The Fontan circulation. *Cont Educ Anaesth Crit Care Pain* 2008; **8**: 26–30.

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