Chapter

69 The Fontan Patient

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A 16-year-old male status post-Fontan completion presents to the emergency room with a two-day history of abdominal pain and emesis. Radiography reveals a small bowel obstruction. Surgery is consulted with the plan being a laparoscopic enterostomy. Following initial resuscitation, his vitals are T: 38.4, HR: 110, BP: 90/58, RR 22, and SpO_2: 88% RA.

During preoperative evaluation, the patient is 170 cm and 70 kg, and appears comfortable. He has 1–2+ pitting edema in the bilateral lower extremities, clear respirations, and a strong pulse. His abdomen is tender in the right lower quadrant and his airway exam is unremarkable. Hepatosplenomegaly is appreciated.

On assessment of the patient's exercise tolerance prior to illness, he tolerates day-to-day activities, but doesn't exercise.

Labs are significant for a leukocytosis of 22,000/UL with an elevated neutrophil count. His C-reactive protein (CRP) is 4.

His primary cardiac disease was tricuspid atresia with associated pulmonary atresia and a ventricular septal defect. He initially underwent a modified Blalock–Taussig shunt in the first week of life, followed by a bidirectional Glenn at age three months. He underwent lateral-tunnel Fontan with fenestration at three years. His post-Fontan course has been complicated by episodes of supraventricular tachycardia for which he is on sotalol.

An echocardiogram performed in the emergency room reveals normal ventricular function with mild-to-moderate atrioventricular valve regurgitation and right-to-left shunting visualized across the fenestration.

What Are the Different Types of Fontan Procedures, and What Are Their Associated Issues?

Different surgical techniques have been used over time to restore a series circulation in patients with single ventricle physiology (Figure 69.1). Initially, atriopulmonary Fontan repairs were most common in which the right atrium would be anastomosed to the pulmonary artery. The rationale for this procedure was that the right atrium would become "ventricularized" and serve as a subpulmonic pumping chamber. With time, it became evident that such remodeling did not occur, and that complications such as atrial arrhythmias and thrombus formation were prevalent.

To minimize the risk of atrial dilation, de Leval et al. introduced the total cavopulmonary anastomosis. Initially conceived as the intracardiac lateral tunnel, this repair brings the superior vena cava to the pulmonary artery, and baffles the inferior vena cava blood to the inferior aspect of the pulmonary artery using an intraatrial conduit. While a small amount of native atrium is retained in this repair to provide growth potential, the risk of atrial arrhythmia and thrombosis is theoretically minimized.

The most recent modification to the Fontan repair has been the introduction of the extracardiac conduit. Here, an interposition graft is placed outside of the heart connecting the transected inferior vena cava and the underside of the pulmonary artery. The impetus for this repair was to avoid pulmonary or systemic venous obstruction in patients with small atria or anomalous venous return. Due to its ease of placement, it has become widely adopted as the technique for Fontan completion. Drawbacks to the extracardiac conduit include a lack of growth potential and a theoretic risk of thrombosis due to the prosthetic graft. Outcomes between the lateral tunnel and extracardiac Fontan have essentially been equivalent.

What Is the Time Frame for Fontan Completion Surgery?

The usual time frame for Fontan completion is between the ages of two to five years. As the patient

https://doi.org/10.1017/9781108668736.070 Published online by Cambridge University Press

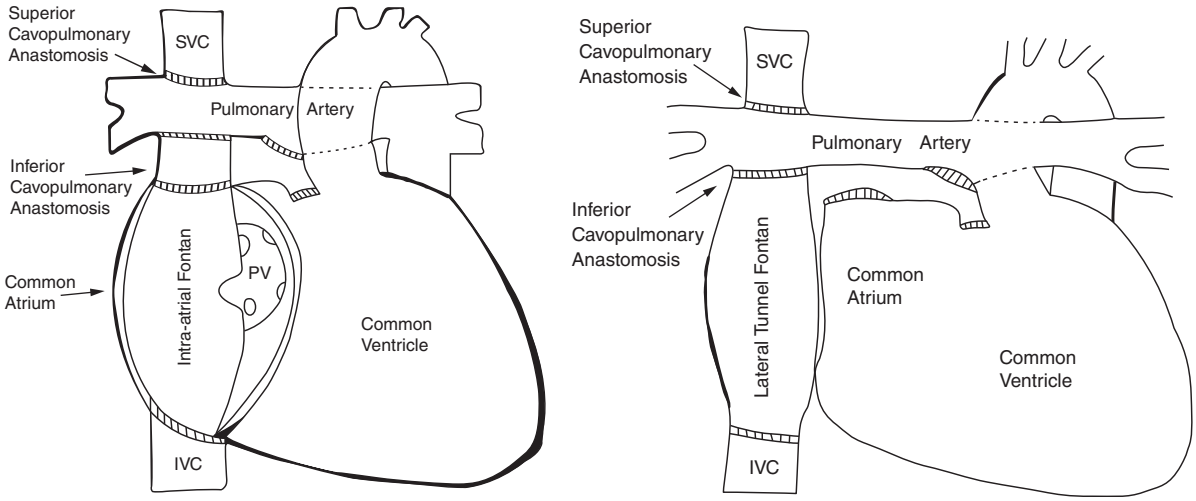

Figure 69.1 Illustration of intra-atrial Fontan (right) and lateral tunnel Fontan (left). SVC and IVC, superior and inferior vena cava, respectively. Illustration by Adam C. Adler, MD.

grows, the percentage of blood return from the lower extremities increases. The blood returning to the lungs from the upper extremities and head becomes a smaller percentage of total venous return and the patient becomes progressively more cyanotic. The ideal age for Fontan completion varies, but attempts to balance the years the patient is cyanotic and the need for revisions, as these conduits do not grow with the patient.

What Is the Purpose of Fenestrating a Fontan, and What Implications Does This Have on Saturation?

A fenestration is a hole created at the time of the Fontan completion between the conduit and the pulmonary venous atrium. This can be accomplished with both the lateral tunnel and extracardiac Fontan. The goal of the fenestration is to maintain cardiac output at the expense of a decrease in saturation in those patients who may be at risk of having significantly elevated Fontan (pulmonary arterial) pressures. This may be the case in patients with elevated pulmonary vascular resistance, ventricular dysfunction, or atrioventricular valve regurgitation.

Because filling of the systemic ventricle is dependent on passive blood flow through the pulmonary vasculature, any factor that adds resistance to this pathway can lead to a decrease in cardiac output and further elevations in both the Fontan and central venous pressures. The fenestration in this setting can be thought of as a "pop-off" valve in situations where these pressures increase. During a period of increased peak inspiration pressure, blood can return to the ventricle through the fenestration which serves as a right-to-left reversible shunt.

Saturations should be in the high 80s to low 90s in patients with fenestrations, with some variability due to the extent of right-to-left shunting across the fenestration.

What Are Other Unique Causes of Desaturation in a Fontan?

Beyond a fenestration, resting saturations <90% may indicate intrapulmonary arteriovenous shunting or the development of systemic venous-to-pulmonary venous collaterals. Both act as extracardiac right-to-left shunts.

Is There Such a Thing as a "Good" Fontan?

The ideal Fontan circulation would have normal sinus rhythm, low atrial pressures, and a low transpulmonary gradient (the difference between the pulmonary arterial pressure and the pulmonary wedge or left atrial pressure).

Implicit in this setting would be preserved systolic and diastolic ventricular function, and no residual

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inflow or outflow obstructions. Examples of such obstructions to flow include:

- pulmonary arterial or pulmonary venous stenosis
- elevations in pulmonary vascular resistance
- atrioventricular valve regurgitation or stenosis
- systolic or diastolic ventricular dysfunction
- ventricular outflow tract obstruction
- aortic insufficiency or stenosis
- recurrent coarctation of the aorta

While Fontan circulation provides a means of palliation for patients with single ventricle physiology, significant long-term sequelae remain. Over time, lack of a pumping mechanism providing active preload to the lungs leads to progressive issues. Each time the pulmonary or intrathoracic pressure is elevated (i.e., under stress, exercise, or Valsalva), stress in the form of elevated pressure is placed on the venous circulation. The physiologic consequence of the Fontan is cavopulmonary hypertension with a relative decrease in ventricular preload, stroke volume, and cardiac output. In addition, patients with single morphologic right ventricles develop complications related to a right ventricle ejecting against high pressures.

What Are the Signs of a Failing Fontan?

Fontan failure can present early after the Fontan procedure or late, but in either case is defined by an inadequate cardiac output and the sequelae of an elevated central venous pressure: pleural effusions, ascites, and peripheral edema.

Soon after surgery, failure may occur secondary to residual obstructions, myocardial injury, or underappreciated preoperative risk factors (elevated pulmonary vascular pressures). Unless addressed promptly, such situations can result in a vicious cycle in which cardiac output and pulmonary function rapidly worsen. Management options depend on the etiology of failure, and include repair of the residual obstruction, take-down of the Fontan, creation of a fenestration if not already present, or mechanical circulatory support.

Late failure following Fontan surgery may be in the form of progressive exercise intolerance and the insidious onset of multiorgan dysfunction. This may represent the cumulative effects of primary ventricular dysfunction and/or progressive increases in pulmonary vascular resistance over time. Elevated pulmonary pressure results in chronic hepatic venous congestion often progressing to resulting in portal hypertension and progression to hepatic failure or cirrhosis. Esophageal varices may be observed in 25% of patients although variceal bleeding is rare. Hepatorenal insufficiency may also develop. With the increasing number of Fontan patients surviving long term, focus on early recognition, diagnosis, and treatment and prevention of sequelae has become critical.

Describe the Sequelae of Protein Losing Enteropathy and Plastic Bronchitis in the Setting of the Fontan Circulation

Plastic bronchitis is a lymphatic flow disorder in which rubbery casts form within the pulmonary tree resulting in obstruction, coughing, and gas exchange disturbance.

Protein losing enteropathy (PLE) is a relatively common morbidity following the Fontan procedure. Symptoms include progressive dyspnea, large abdominal and/or pleural effusions, and peripheral edema including ascites. Patients often have intestinal malabsorption, hypoalbuminemia, hypogammaglobinemia, and lymphopenia. While the exact etiology is unknown, it is theorized that over time, low cardiac output results in intestinal vascular hypoperfusion and especially intestinal mucosal hypoperfusion.

The morbidity and mortality in patients with PLE remains high with treatment focusing on prevention and symptom amelioration especially in the form of inotropic support, diuretic therapy, and protein (albumin) replacement.

Does it Matter Whether the Systemic Ventricle Is a Morphologic Right or Left Ventricle in Predicting Fontan Failure?

While the right ventricle is capable of supporting the systemic circulation, clinical experience both with congenitally corrected transposition, and patients undergoing an atrial baffle repair for transposition suggests a high incidence of systemic right ventricular failure over time. In patients with single ventricle physiology, data has been mixed, but suggests that a systemic right ventricle is a risk factor not only for Fontan failure, but also death. The right ventricle is morphologically different from the left ventricle with its extensive trabeculations and thinner myocardium. Thus, the right ventricle is less apt to eject against high pressures for many years (Figure 69.2).

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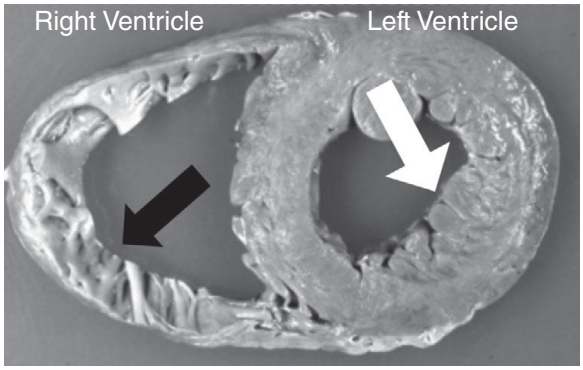

Figure 69.2 Gross cardiac specimen of the left and right ventricle demonstrating the difference in ventricular wall morphology

Is It Possible to Have Both Preserved Systolic Function and a Failing Fontan?

Yes. Two major clinical phenotypes of Fontan failure are observed: those with failure in the setting of systolic ventricular dysfunction, and those with failure in spite of preserved systolic function. This latter group accounts for up to two-thirds of adult Fontan patients who die or require transplantation. Mechanisms accounting for this paradox include elevations in pulmonary vascular resistance and the complex effects of this circulation on the liver, kidneys, vasculature, and neurohumoral system.

What Information Is Useful in the Preoperative Evaluation of a Fontan?

Thorough evaluation entails knowledge of the patient's anatomic substrate and past surgical interventions. Recent catheterization data including interventions, cardiac imaging, cardiac rhythm, and oxygen saturations should be reviewed.

Laboratory values should aim at identifying end-organ dysfunction especially cardiac, liver, and renal dysfunction. Labs of particular significance include liver function tests (including tests of coagulation), blood urea nitrogen and creatinine values to assess renal function, a complete blood count to assess platelet levels due to the risk of thrombocytopenia due to sequestration, and albumin levels if there is concern for a protein-losing enteropathy. Recent changes in functional status or excessive weight gain should raise concern for failure.

As Fontan physiology lends itself to arrhythmias, a history suggestive of rhythm abnormalities or documented arrhythmias may signal the potential need for cardioversion or pacing in the perioperative period.

Echocardiography can reveal areas of concern in the Fontan circuit, with specific attention to echocardiographic assessment of: ventricular function, the atrioventricular valve, ventricular outflow tract, and pulmonary arterial and venous flows.

Beyond the components of the normal physical exam, attention should be paid to the potential presence of pleural effusions, hepatosplenomegaly, signs of cirrhosis, ascites, or peripheral edema. Such findings suggest elevated venous pressure and potentially failing circulation. Baseline saturations should also be noted as any significant desaturation may indicate the presence of an intra- or extracardiac shunt. Lastly, as adequate hydration is critical to maintaining the hemodynamics of a Fontan, volume status should be assessed.

What Intraoperative Monitors Would You Consider Using in the Patient Described in this Chapter?

Concerns in this patient include not only his Fontan circulation, hypovolemia, and the potential for sepsis in the setting of bowel obstruction. When taken together with the need for positive-pressure ventilation, anesthesia, and laparoscopy, this patient is at significant risk for hemodynamic deterioration during this procedure. In addition to routine monitoring, one may also consider using cerebral near–infrared spectroscopy (NIRS) and arterial and central venous lines.

Case continued: Once in the operating room, the patient undergoes a rapid sequence induction with 1 mg of midazolam, 50 mcg of fentanyl, 70 mg of propofol, and 80 mg of succinylcholine. The initial blood pressure after intubation is 92/42, but trends down to 68/40 following arterial line placement with 0.6 v/v% isoflurane.

How Would You Manage Hypotension in This Setting?

Hypotension in this patient following induction and intubation is likely multifactorial. The onset of positive-pressure ventilation in conjunction with hypovolemia are likely contributing to diminished

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ventricular filling, and thus a decreased cardiac output. The induction of anesthesia has also likely reduced systemic vascular resistance and increased venous capacitance, leading to a decrease in both blood pressure and preload. First-line treatment would entail volume resuscitation and the initiation of vasopressors. When time permits, volume repletion should be attempted prior to induction of anesthesia. Ventilator settings can also be reassessed to achieve the minimum peak pressures, positive end-expiratory pressure (PEEP) and rate that provide adequate oxygenation and ventilation.

What Is the Potential Impact of Laparoscopic Surgery on the Patient's Physiology?

The abdominal insufflation accompanying laparoscopy reduces lung compliance resulting in increased peak inspiratory and plateau pressures. Insufflation acts to further decreases systemic venous return, thereby decreasing cardiac output. If the Trendelenburg position is required, these effects are exacerbated. Steep reverse Trendelenburg positioning may reduce venous return on the basis of lower extremity pooling. Additionally, CO_2 used for abdominal insufflation may result in hypercapnia with a subsequent increase in pulmonary vascular resistance.

These negative effects can be mitigated if the minimal amount of insufflation pressure is used. This strategy must be communicated with the surgeon in advance of the procedure. If hemodynamic instability remains, consideration should be given to performing an open procedure.

Should This Patient Be Extubated?

Provided adequate hemodynamics, oxygenation, and ventilation, extubation is appropriate. Spontaneous ventilation improves passive venous return and therefore cardiac output.

Pain control is paramount, and can be achieved with a combination of narcotics, acetaminophen (assuming no significant hepatic derangements), ketorolac (assuming preserved renal function), and/or transverse abdominis plane blocks. Inadequate pain control results in consistently elevated PVR.

Where Should This Patient Recover?

For the first postoperative night, it may be advisable to recover this patient on a floor where the staff is familiar with the patient's physiology. If such a floor is not present, the intensive care unit may be the most appropriate.

Suggested Reading

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