Chapter 30

Failing Fontan

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Case Scenario

A 20-year-old female with a past medical history of anxiety and hypoplastic left heart syndrome palliated to a fenestrated lateral tunnel Fontan presents for wisdom teeth extraction. She underwent Fontan completion at age 3 years, with her initial postoperative course complicated by chylous effusions requiring chest tube placement and postoperative sinoatrial node dysfunction requiring placement of a dual chamber pacemaker. Two years ago, she was admitted with severe cyanosis and found to have restrictive flow across the intraatrial Fontan baffle at the level of the anastomosis with the inferior vena cava. She underwent stenting of the obstruction without recurrence and since then has maintained appropriate follow-up with her cardiologist.

She states that she has been doing well and denies syncopal episodes, chest pain, or palpitations. Pulse oximetry shows a room air saturation of 91%. On examination, she is alert but anxious. Electrocardiogram shows a paced rhythm at 70 beats/minute. Recent cardiac catheterization confirmed the echocardiographic findings and noted a small patent baffle fenestration. Fontan pressure was measured as 20 mm Hg and several systemic-pulmonary venous collateral vessels were coiled.

A recent echocardiogram shows:

- *A morphologic right ventricle*
- *Mild tricuspid valve regurgitation*
- *Mildly impaired systolic function, ejection fraction 40%*
- *An unobstructed stented lateral tunnel Fontan*

Key Objectives

- Understand the Fontan procedure and physiology.
- Recognize symptoms, signs, and outcomes of failing Fontan physiology.
- Discuss preoperative assessment of Fontan patients.
- Develop a perioperative management plan for a patient with a failing Fontan.
- Discuss the postoperative disposition of Fontan patients.

Pathophysiology

What is Fontan physiology?

Initially proposed by Francis Fontan and Eugene Baudet as a surgical palliation for tricuspid atresia, the Fontan procedure has since been adapted as a palliation for a variety of congenital heart diseases resulting in single-ventricle (SV) physiology. Improvements in surgical technique and medical management have led to increased survival, making it more likely that these patients will present for a variety of noncardiac procedures. In general, patients with congenital heart disease (CHD) have an increased risk of 30-day mortality when compared to healthy cohorts [1]. To provide a safe perioperative course, it is essential to understand Fontan physiology.

In a normal circulation each ventricle pumps against one resistance, either pulmonary or systemic. The Fontan procedure directly routes systemic venous return to the pulmonary arteries, creating a series circulation whereby cardiac output (CO) is dependent on three elements: pulmonary blood flow (PBF), the transpulmonary gradient (TPG), and the single ventricle. In other words, the single ventricle must provide the energy for blood to flow across three resistance beds: the systemic vascular bed, the cavopulmonary connection, and the pulmonary vascular bed. A reduction in PBF therefore results in both decreased CO and systemic deoxygenation. (See Chapters 26–29 for further details of single-ventricle palliation and Fontan physiology.)

Clinical Pearl

In a normal circulation each ventricle pumps against one resistance, either pulmonary or systemic. Cardiac output in a patient with Fontan physiology is dependent on maximizing blood flow across three resistances (systemic vascular, cavopulmonary connection, and pulmonary vascular) with one ventricle.

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What makes a "good" Fontan candidate?

Requirements for an ideal Fontan circulation can be divided into cardiac and pulmonary components.

- ***Cardiac requirements***
  - Sinus rhythm
  - Atrioventricular valve competency
  - Good ventricular function
  - Unobstructed systemic outflow from the single ventricle
- ***Pulmonary requirements***
  - Nonrestrictive, unobstructed Fontan connection from the systemic veins to the pulmonary arteries
  - Adequately sized pulmonary arteries without anatomic distortion
  - Near-normal pulmonary vascular resistance (PVR)
  - Unobstructed pulmonary venous return

In other words, the pulmonary artery pressure (PAP) should be low (ideally <15 mm Hg). The transpulmonary gradient (the difference between mean PAP and LA pressure) should also be low, below 7 mm Hg. It may be elevated when there is increased pulmonary venous pressure, which will impair adequate flow across the Fontan circulation. Pulmonary vascular resistance should be <4 Wood units/m^2. These measurements are indicators of potential pulmonary vascular disease.

What are the different types of Fontan procedure?

Over time several different anatomic variations of the Fontan have been performed and are outlined below in chronological order. Although the atriopulmonary Fontan is no longer performed, older surviving patients may still display this anatomic connection.

1. ***Atriopulmonary Fontan.*** Performed after a "classic" Glenn procedure, this method has become obsolete as a surgical option. A "classic Glenn" procedure anastomoses the superior vena cava (SVC) to the distal end of the right pulmonary artery (RPA). An atriopulmonary connection is then established between the proximal end of the divided RPA and the anatomic right atrium (RA). The atrial septal defect is closed and IVC blood flow is now directed toward the left PA to perfuse the left lung. The main PA is ligated, allowing blood to bypass the nonfunctional ventricle. (See Figure 30.1A.)
2. ***Total cavopulmonary anastomosis (TCPA).*** This may be performed after EITHER a bidirectional Glenn (BDG) or hemi-Fontan (HF) Stage II procedure. The BDG and HF are both procedures performed by surgically connecting the SVC and the pulmonary arteries, thus providing a source of PBF. *Although they differ anatomically, the physiologic result is the same.*
   - The ***BDG*** is created by dividing the SVC at its junction with the RA, oversewing the atrial end, and creating an end-to-side anastomosis between the SVC and the RPA.
   - The ***HF***, in contrast, keeps the SVC and RA in continuity when the SVC is connected to the RPA. However, homograft tissue is sewn across the superior cavoatrial junction to prevent blood flow from the SVC to the RA.

   The TCPA or "completion Fontan" is then generally performed 1–3 years after either the BDG or HF procedure. This completion adds IVC flow directly into the pulmonary bed as well, and commonly takes one of two forms as listed below. An opening, or a ***fenestration***, is often created between the RA and the conduit or baffle. This allows right to left (R-to-L) shunting in the setting of any increases in PVR and hence a means of increasing CO, albeit at the cost of normal oxygen saturations, as oxygenated and deoxygenated blood are then allowed to mix.
   - ***Lateral tunnel Fontan.*** (See Figure 30.1B.) The RA is septated and an intraatrial baffle is placed in order to direct blood from the IVC to the RPA.
   - ***Extracardiac Fontan.*** (See Figure 30.1C.) The IVC is disconnected from the RA and anastomosed using a conduit to the RPA.

Clinical Pearl

An opening, or fenestration, is often created between the RA and the conduit or baffle during the Fontan procedure, allowing a R-to-L shunt to occur in the setting of any increases in pulmonary vascular resistance. This results in increased systemic blood flow, albeit at the cost of normal oxygen saturations, as oxygenated and deoxygenated blood are then allowed to mix.

What are the advantages of a TCPA versus atriopulmonary Fontan?

It was initially thought that the atriopulmonary Fontan improved PBF secondary to a pulsatile atrium. However, over time the thin-walled atrial portion becomes dilated from exposure to higher than normal systemic venous pressures, leading to impaired contractility and energy loss. The atrium also becomes a site for the development of atrial arrhythmia and thrombus formation due to stasis.

https://doi.org/10.1017/9781108657341.031 Published online by Cambridge University Press

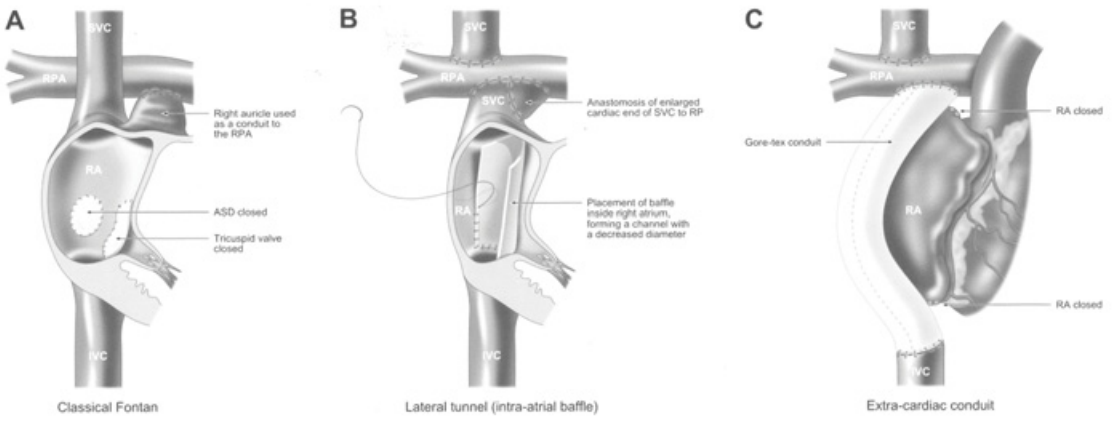

Figure 30.1 Fontan surgical techniques. (A) Classical atriopulmonary connection. (B) Lateral tunnel cavopulmonary connection. (C) Extracardiac cavopulmonary connection. From Y. d'Udekem et al. The Fontan procedure: contemporary techniques have improved long-term outcomes. *Circulation* 2007; **116**: I-157–64. With permission.

Patient's age (years)

	20	25	30	35	40	45	50	55	60
ASD	25	26	32	38	42	47	52	57	61
Valvar disease	29	31	36	40	45	49	54	59	63
VSD	28	30	36	40	44	49	53	59	63
Aortic coarctation	32	33	38	43	47	52	56	62	66
AVSD	33	34	39	44	48	52	57	62	66
Marfan syndrome	37	38	42	46	50	54	59	64	68
Tetralogy of Fallot	37	38	42	47	50	54	60	65	69
Ebstein anomaly	42	43	47	51	54	59	63	68	72
Systemic RV	46	48	51	55	59	63	67	72	76
Eisenmenager syndrome	57	58	62	65	69	73	77	81	84
Complex CHD	58	59	63	67	70	74	78	82	85
Fontan	64	65	68	72	75	78	82	86	91

Age difference:
- >40
- 30–40
- 20–30
- 10–20
- 5–10
- 2–5
- <2

Figure 30.2 The numbers represent the equivalent age having similar 5-year mortality rates. Colors reflect the difference between the relative age and the actual age of patients. ASD, atrial septal defect; AVSD, atrioventricular septal defect; CHD, congenital heart disease; RV, right ventricle; VSD, ventricular septal defect. From G. Diller et al. Survival prospects and circumstances of death in contemporary adult congenital heart disease patients under follow-up at a large tertiary centre. *Circulation* 2015; **132**: 2118–25. With permission.

Patients with TCPA have demonstrated improved outcomes and also have a decreased risk over time of atrial arrhythmias and thrombosis compared to patients with atriopulmonary Fontans. The lateral tunnel method includes the lateral wall of the RA, which allows for growth as the child grows. The extracardiac conduit decreases suture lines within the atrium and because it excludes atrial tissue, dilation is limited. This decreases arrhythmogenic foci. However, the graft used does not allow for growth potential, so this method is usually performed in patients large enough to accept a graft of adequate size for adult IVC flow.

Does the presence of CHD affect the onset of heart failure?

The progression of symptomatic heart failure in congenital heart patients, particularly patients with SV physiology, occurs decades before failure in patients with acquired heart disease. This is illustrated in Figure 30.2, where the equivalent age for adult patients with CHD is compared to that of patients without CHD [2]. For example, the 5-year mortality rate of a 35-year-old Fontan patient is equivalent to the 5-year mortality rate of a 72-year-old from the general population!

What is Fontan failure and what are the signs and symptoms?

Signs and symptoms of Fontan failure may include fatigue, dyspnea, growth failure, exercise intolerance, decreased activity level, syncopal/presyncopal episodes, palpitations, cyanosis, weight gain, edema, ascites, and cough with expectoration of mucoid cast-like material. These can be secondary to ventricular dysfunction, systemic atrioventricular valve insufficiency, atrial and/or ventricular arrhythmias,

https://doi.org/10.1017/9781108657341.031 Published online by Cambridge University Press

renal failure, hepatic insufficiency, protein losing enteropathy (PLE), and plastic bronchitis [3, 4].

Cardiac

- ***Ventricular function***. Systemic ventricular dysfunction is a common problem encountered over time and may be due to a history of ventriculotomy during staged palliative procedures or chronic hypoxemia. *Patients with a morphologic systemic RV experience a greater decline in ventricular function compared to those patients with systemic left ventricles*. Low CO can present as a decline in exercise tolerance and resting or exercise desaturation. Decreasing exercise tolerance may also result from a decreased response to β-stimulation secondary to limited preload reserve [5]. These patients show an abnormal response to increased heart rate, including a diminished contractile response and impaired diastolic filling [6]. Additional causes of low CO may be related to low flow across the Fontan pathway from physical obstruction across the pulmonary arteries, compression of pulmonary venous return, elevations in PVR, or atrioventricular (AV) valve dysfunction. Atrioventricular valve dysfunction results in elevated atrial pressures that may cause atrial dysrhythmias and decreased CO. Aortic stenosis will lead to ventricular hypertrophy and reduced ventricular compliance, while aortic regurgitation creates volume overload and systemic ventricular failure.
- ***Rhythm disturbances.*** Fontan patients may suffer from a multitude of rhythm disturbances including sinus node dysfunction, junctional rhythm, AV block, and supraventricular or ventricular arrhythmias. Risk factors include atrial suture lines, myocardial fibrosis, atrial dilation, AV valve regurgitation, and ventricular failure. Atrioventricular conduction block may occur due to intrinsic conduction abnormalities or as a consequence of surgery. If pacemaker implantation is necessary, epicardial pacemakers are used most commonly due to limited venous access to the atrium and the risk of endocardial lead thrombosis. It is imperative to maintain sinus rhythm in Fontan patients, as nonsinus rhythm can cause an acute elevation in atrial pressures with each ventricular contraction and is associated with decreased CO and long-term hepatic fibrosis [6, 7].
- ***Oxygen saturation***. Cyanosis may occur from persistent R-to-L shunting and pulmonary venous desaturation.
  - ***R-to-L shunting*** can occur secondary to excessive flow across a fenestration, a baffle leak, and/or veno-venous or veno-arterial decompressing collateral vessels. The formation of decompressing vessels (such as veno-venous collaterals) is not unusual in a Fontan patient where systemic venous pressure becomes elevated. This compensatory process alleviates pressure in the venous return pathway, while maintaining systemic ventricular preload. Patients who become desaturated may form aortopulmonary collaterals, which are vessels that carry oxygenated blood from the aorta to the pulmonary vascular bed in patients with poor PBF. These not only decrease systemic CO and but also cause volume overload on the single ventricle. If volume overload is significant, the patient may present for hemodynamic cardiac catheterization and possible coiling of decompressing vessels.
  - ***Pulmonary venous desaturation*** may occur due to intrinsic lung disease, pulmonary arteriovenous malformations, pleural effusions, or plastic bronchitis. Plastic bronchitis presents as tachypnea, cough, wheezing, and expectoration of bronchial casts. Leakage of proteinaceous material into the airways lead to cast formation and potential obstruction of the airways. It is a poor prognostic sign and an indication for transplant.
  - ***Pulmonary thromboembolic events*** can contribute to hypoxemia.

Hepatorenal

Chronic elevated systemic venous pressure, low CO, and end-organ hypoperfusion lead to hepatic and renal dysfunction. Patients may have mild elevations in bilirubin and abnormally elevated liver enzymes.

- ***Hypoalbuminemia*** and increased risk of hepatocellular carcinoma may occur.
- ***Protein losing enteropathy*** is characterized by lymphatic dysfunction with persistent hypoalbuminemia in the absence of renal or liver disease, with manifestations of ascites, peripheral edema, pleural effusions, and fat malabsorption. The etiology is not well known; theories include low CO, mesenteric vascular flow anomalies and elevated pressures, or autoimmune/inflammatory reactions. Initial management includes adjustments to existing anticongestive therapy, parenteral albumin, sodium restriction and diet modification, immunoglobulin replacement, or steroid therapy. Patients with obstructive lesions within the Fontan circuit are evaluated and obstructions may need to be relieved via transcatheter approach or surgical management. The presence of PLE is a poor prognostic sign and may be an indication for transplant.

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Hematologic

Hematologic disturbances may arise, including possible sequestration of platelets by the liver and spleen resulting in thrombocytopenia. ***Thromboembolism*** may occur due to atrial dilation, reduced CO, slow-moving venous flow, anatomic obstructions within the Fontan circuit, and arrhythmias. For this reason, patients are usually on anticoagulation medications [3, 4].

Table 30.1 summarizes the clinical manifestations of failing Fontan physiology along with possible etiologies.

What potential etiologies exist for the lower oxygen saturation in this patient?

Systemic oxygen saturation in a well-functioning Fontan patient should be between 90% and 95%. Potential causes of hypoxemia in the Fontan population include persistent R-to-L shunting and pulmonary venous desaturation.

What does an oxygen saturation of 100% indicate in a patient with a fenestrated Fontan?

A fenestration is often created between an extracardiac or lateral tunnel Fontan and the atrium to maintain ventricular preload while reducing elevated pressure in the Fontan circuit. This fenestration helps preserve systemic preload and CO when PVR is elevated and/or PBF is decreased. Decreased PBF may occur when there is an obstruction within the Fontan circulation or an increase in PVR. This occurs at the expense of systemic desaturation, as the systemic venous return will bypass the pulmonary circulation. An oxygen saturation of 100% raises the suspicion for obstruction or thrombus formation at the level of the fenestration.

Table 30.1 Laboratory Abnormalities in the Failing Fontan

Cardiac	Elevated brain natriuretic peptide (BNP)
Hepatic	Liver function tests Hyperbilirubinemia
Renal	Elevated creatinine
Hematologic	Anemia Polycythemia Lymphopenia Immunoglobulin deficiency Decreased or prolonged PT, PTT, INR
Electrolyte abnormalities	Hyponatremia Hyperglycemia
Protein-losing enteropathy	Hypoproteinemia Hypoalbuminemia Hypocalcemia + stool α_1-antitrypsin

INR, international normalized ratio; PT, prothrombin time; PTT, partial thromboplastin time.

Clinical Pearl

The systemic oxygen saturation in a patient with Fontan physiology is usually between 90% and 95%. Desaturation is abnormal and could indicate a failing Fontan, while saturation of 100% in a fenestrated Fontan could indicate thrombosis or obstruction at the level of the fenestration.

Anesthetic Implications

How can clinical status be assessed in a patient with Fontan physiology?

The preoperative evaluation of patients with Fontan physiology involves a thorough history and physical examination with attention to recent changes in health status, exercise capacity, hospital admissions, current medications, and additional comorbid conditions. Recent respiratory tract infections are of special concern, as they can lead to an increased incidence of bronchospasm or laryngospasm with increased airway resistance and PVR, resulting in lower PBF and CO. Physical evaluation should focus on the airway, heart, and lungs, as well as extremities. The assessment of the extremities allows for the evaluation of cyanosis, edema, and vascular access.

The patient with a well-functioning Fontan is warm, well perfused, and acyanotic. Auscultation of the heart should be without murmurs and oxygen saturation is typically between 90% and 95%. However, in a patient with a failing Fontan signs of systemic venous and hepatic congestion are often present. Vascular access may prove challenging due to previous surgeries and interventions. Preoperative hematologic studies including blood counts, electrolytes, and hepatic and renal function studies should be considered depending on the type of surgery. Table 30.2 describes possible laboratory findings in a patient with failing Fontan physiology [6, 8, 9].

Clinical Pearl

The functional status and comorbidities in patients with Fontan physiology can vary widely, from patients who are well-compensated to those with a failing circulation.

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Table 30.2 Failing Fontan

Complications	Etiology
Growth failure	Suboptimal cardiac output
Exercise intolerance	Chronotropic incompetence, abnormal pulmonary compliance with exertion, atrial distention
Depression	Limitations in functional status
Arrhythmias	Sinus node dysfunction, predominant junctional rhythm, AV block, supraventricular tachycardia/atrial tachycardia, ventricular tachycardia
Diminished cardiac output	Obstruction: Atriopulmonary, pulmonary arterial, pulmonary venous obstruction, AV valve inflow or ventricular outflow AV valve dysfunction Ventricular dysfunction: due to atrial distortion or dilation, AV valve dysfunction, chronic arrhythmias, impaired myocardial perfusion, abnormal ventricular morphology, prolonged cyanosis, or volume overload Thrombosis: systemic venous, atrial, or pulmonary
Cyanosis	Intracardiac right-to-left shunt, veno-venous collaterals, pulmonary AVMs, progressive increase in PVR
Pleural effusions	Elevated Fontan pressures
Plastic bronchitis	Unknown
Protein losing enteropathy	Unknown
Hepato-renal insufficiency	Low cardiac output, sepsis
Ascites	Portal hypertension from obstruction, hepatic failure, cirrhosis
Metabolic derangements	Decreased albumin, thrombocytopenia, hyperbilirubinemia, coagulopathy

AV, atrioventricular; AVM, arteriovenous malformation; PVR, pulmonary vascular resistance.

The patient's medications include albuterol, aspirin, enalapril, fluoxetine, and spironolactone. What preoperative considerations are involved?

Preoperative assessment includes knowledge of the current medication regimen and any recent changes consistent with worsening clinical status. Patients are frequently taking diuretics, digoxin, antihypertensives, antiarrhythmics, and anticoagulants.

Most cardiac medications should be continued up to the time of anesthetic induction. No consensus exists for preoperative diuretics. They are typically continued for low-risk surgery but withheld for higher-risk surgeries due to the risk of potential dehydration. Of note, patients on diuretic therapy may have electrolyte abnormalities that should be evaluated with preoperative laboratory studies. Angiotensin converting enzyme inhibitors (ACEi) are commonly withheld prior to surgical procedures requiring general anesthesia due to intraoperative hypotension that can be difficult to manage. If a patient is taking an ACEi prior to surgery, the recommendation at our institution is to stop the ACEi 24 hours prior to the procedure. If the patient took their ACEi on the day of the procedure, the decision to proceed with surgery is determined by the type and urgency of the surgical procedure: a procedure with the potential for large volume shifts and hemodynamic instability should be postponed if possible.

Aspirin is usually continued to prevent thrombosis, and patients on warfarin may need preoperative admission for monitoring and bridging to intravenous heparin. The patient's cardiologist and surgeon should be involved in discussions regarding the perioperative management of antithrombotic medications.

Clinical Pearl

The patient's cardiologist and surgeon should be involved in discussions regarding the perioperative management of antithrombotic medications.

What are the major preoperative anesthetic concerns for this patient?

Patients with failing Fontan physiology often have poor ventricular systolic function at baseline and are vasoconstricted in order to maximally augment their preload. These patients are also at risk for hemodynamically significant atrial arrhythmias. The patient in this scenario has RV dysfunction in addition to AV valve regurgitation. Her

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Table 30.3 Diagnostic Modalities and Their Utility in the Evaluation of the Failing Fontan

Diagnostic Modality	Evaluation
Electrocardiogram	• Rhythm • Pacing dependence
Echocardiography	• Ventricular function • AV valve function • Fontan pathway patency • Fenestration status/gradient
Cardiac catheterization	• Fontan conduit pressure/resistance • Quantification of ventricular function • Ventricular end-diastolic pressure • Quantification of PVR • Presence/embolization of collateral vessels or arteriovenous malformations
Chest radiography	• Heart size • Pleural effusions • Pulmonary edema

AV, atrioventricular; PVR, pulmonary vascular resistance.

cardiac catheterization demonstrated elevated Fontan pressures, which could indicate an obstruction to flow or significant myocardial dysfunction. Finally, in addition to the risk of arrhythmia secondary to the Fontan pathway, she is pacemaker-dependent and will thus require pacemaker management in the perioperative period.

Baseline hematologic and biochemical studies are important in the preoperative evaluation. In addition, several imaging and diagnostic modalities are required. These are summarized in Table 30.3.

With a history of complete heart block and a pacemaker, what considerations are important?

The management of any permanent pacemaker (PPM) or implantable cardioverter-defibrillator (ICD) device requires knowing the indication for placement, pacing mode, pacing rate, battery life, underlying rhythm, and any cardioversion or defibrillation events. For that reason, it is necessary to consult the electrophysiology team and to interrogate these devices in the preoperative setting.

The potential effects of electromagnetic interference (EMI) on device function are of concern in pacemaker-dependent patients. Possible sources of EMI include electrocautery (especially monopolar electrocautery), evoked potential monitors, nerve stimulators, external defibrillation, radiofrequency ablation, and extracorporeal shock wave lithotripsy. Electromagnetic interference may be misinterpreted as intrinsic cardiac signals, resulting in oversensing and inhibition of pacing in PPMs. In pacemaker-dependent patients, inappropriate inhibition of pacing due to EMI can cause bradycardia, sinus arrest, or ventricular standstill. In ICDs, EMI can cause noise, potentially resulting in inappropriate cardioversion/defibrillation. (See Chapters 23 and 49 for details of intraoperative pacemaker management.)

Clinical Pearl

The management of any PPM or ICD requires the knowledge of the indication for placement, pacing mode, pacing rate, battery life, underlying rhythm, and any cardioversion or defibrillation events. In pacemaker-dependent patients, inappropriate inhibition of pacing due to EMI can cause bradycardia, sinus arrest, or ventricular standstill. In ICDs, EMI can cause noise, which causes inappropriate cardioversion/defibrillation.

What anesthetic options exist for this patient?

General anesthesia, regional anesthesia, or sedation can be used in Fontan patients. In this case, the safest option is the use of local anesthesia and supplementation with intravenous anxiolytics such as midazolam. Spontaneous ventilation allows for optimal PBF without the need for positive pressure ventilation (PPV), and avoidance of general anesthesia prevents the potential risk of hemodynamic instability from systemic vasodilation. Intravenous fentanyl may be used to supplement sedation and as an adjunct for procedural discomfort. However, it is important to keep in mind that a diminished respiratory drive with subsequent increased carbon dioxide levels may lead to hemodynamically significant elevations in PVR.

Due to her extreme anxiety the patient desires general anesthesia. What is an appropriate plan?

Intravenous (IV) anesthetic agents may cause vasodilation, decreased preload, and myocardial depression. Due to her diminished ventricular function, a peripheral IV line should be placed prior to induction in this patient. For patients with severely diminished ventricular function it may be advisable to start inotropic support prior to induction. Although in patients with well-compensated physiology an inhalation induction may be performed safely if necessary, in a teenage or adult patient with decreased ventricular function an IV induction is preferable.

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This patient requires endotracheal intubation to protect the airway while providing general anesthesia. While the surgical team generally prefers nasotracheal intubation over orotracheal intubation for ease of access to the oral cavity, the need for nasotracheal intubation should be discussed with the proceduralist as these patients are at high risk for bleeding secondary to elevated venous pressures, hepatic dysfunction and preoperative use of anticoagulants, and the use of an oral endotracheal tube is often acceptable in light of these risks.

During anesthetic maintenance, the use of high concentration of volatile agents should be avoided due to the increased risk of arrhythmias and myocardial depression. Using low concentrations of inhalational agent in combination with the intermittent use of opioid or a short-acting opioid infusion can provide a hemodynamically stable anesthetic. Neuromuscular blocking agents are a useful adjunct for intubation and may be used as long as there is adequate reversal since residual weakness can lead to hypoventilation and hypercarbia, which are poorly tolerated. Finally, perioperative pain control in Fontan patients should be adequate, as uncontrolled pain increases catecholamine release and thus increases PVR. Useful adjuncts in this case can include the use of local anesthetic by the surgical team and intravenous acetaminophen if deemed appropriate.

Potential anesthetic problems can include intraoperative hypotension, thromboembolic events, unstable arrhythmias, significant bleeding, and coagulopathy.

Clinical Pearl

While the surgical team generally prefers nasotracheal intubation over orotracheal intubation for ease of access to the oral cavity, the need for nasotracheal intubation should be discussed preoperatively as Fontan patients are at high risk for bleeding secondary to elevated venous pressures, hepatic dysfunction, and preoperative use of anticoagulants. The use of an oral endotracheal tube is often acceptable in light of these risks.

What type of monitoring should be utilized?

Standard monitors should be placed prior to the induction of general anesthesia, including 5-lead ECG, noninvasive blood pressure monitoring, and pulse oximetry. Blood pressure measurements should be assessed in all four extremities, as discrepancies may exist due to prior surgical procedures or previous arterial cut-down attempts. For example, the presence of a previous classic Blalock–Taussig shunt may cause altered blood pressure readings in the upper extremity when compared to the contralateral side.

The use of invasive monitors depends on patient-specific conditions including pre-existing ventricular dysfunction, as well as procedure-specific factors, and should not be necessary for this case. The use of invasive arterial blood pressure monitoring and central line placement for central venous pressure (CVP) monitoring may be useful in managing large fluid shifts during major surgery. Monitoring CVP trends can help in assessing intravascular volume status but it is important to remember that this number will be a reflection of main PA pressure and not ventricular preload. Central venous access may also be beneficial to provide inotropic support. The routine use of central lines for noncardiac procedures is rare, as the risks of infection, thrombosis, and impaired venous return within the Fontan circulation may outweigh any potential benefits. Transesophageal echocardiography can be used for the assessment of ventricular preload and function when appropriate.

Due to the history of pacemaker-dependent sinus node dysfunction and the risk for arrhythmias, intraoperative placement of transcutaneous defibrillator pads is advised. The placement of invasive arterial blood pressure monitoring prior to the start of the case is not necessary since the upper extremities are readily accessible should the patient become unstable. Finally, a neuromuscular twitch monitor should be used to assure adequate reversal of neuromuscular blockade prior to endotracheal extubation.

Clinical Pearl

The routine use of central lines for minor noncardiac procedures is rare, as the risks of infection, thrombosis, and impaired venous return within the Fontan circulation may outweigh any potential benefits.

Should endocarditis prophylaxis be administered?

The need for bacterial endocarditis prophylaxis in Fontan patients is based on the type of surgical procedure. Current American Heart Association guidelines state that prophylaxis should be administered during dental procedures involving manipulation of gingival tissue, manipulation of the periapical region of teeth, or perforation of the oral mucosa in patients with the following [10]:

- Prosthetic cardiac valves, including transcatheter-implanted prostheses and homografts
- Prosthetic material used for cardiac valve repair, such as annuloplasty rings and chords

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- Previous endocarditis
- Congenital heart disease in the following categories:
  - Unrepaired cyanotic CHD, including palliative shunts and conduits
  - Completely repaired CHD with prosthetic material or device, whether placed by surgery or catheter intervention, during the first 6 months after the procedure
  - Repaired CHD with residual shunts or valvular regurgitation at the site or adjacent to the site of a prosthetic patch or prosthetic device
- Cardiac transplantation recipients with valvular regurgitation due to a structurally abnormal valve

In this case, the patient will require endocarditis prophylaxis because of her tricuspid regurgitation and the surgical palliation using prosthetic material within the lateral tunnel. Appropriate intramuscular (IM) or intravenous (IV) endocarditis prophylaxis consists of either ampicillin 2 g OR the choice of cefazolin or ceftriaxone 1 g. If the patient is allergic to penicillin, then clindamycin 600 mg should be used and can be administered either IM or IV.

What are the hemodynamic goals during anesthesia?

Intraoperative management goals include the following:

- ***Maintain sinus rhythm.***
- ***Maintain adequate preload*** to enhance PBF and CO. Dehydration can prove to be hemodynamically significant and detrimental for Fontan patients. Fasting intervals should be minimized to ensure adequate preoperative hydration and when possible Fontan patients should be scheduled as the first case of the day. Clear liquid intake should be allowed until 2 hours prior to surgery. In older patients, preoperative placement of IV access and IV fluid administration should be considered, particularly if the case is scheduled later in the day.
- ***Avoid or minimize increases in PVR,*** as increased PVR can compromise ventricular filling and CO. Hypoxia, hypercarbia, acidosis, hypothermia, inadequate analgesia or anesthesia, vasoactive drug use, excessive mean airway pressure, or compression of the lung by pleural effusion can all result in increased PVR.
- ***Support of ventricular function.*** Intraoperative management involves maintenance of adequate perfusion pressures with the use of inotropic support if necessary.
- ***Minimize afterload.***

Induction is uneventful, but as the case progresses the patient has persistent hypotension only transiently responsive to repeated fluid boluses. What is the differential diagnosis and what potential therapies should be considered?

The differential diagnosis for hypotension in this patient includes hypovolemia, vasodilation, cardiac depression, arrhythmia, and/or obstruction to flow across the Fontan circulation. This patient is susceptible to hypotension due to her preexisting depressed right ventricular function and elevated Fontan pressure. These factors are exacerbated by hypovolemia and any cause of decreased blood flow through the Fontan circulation. Fluid boluses may be required to counteract decreased preload. However, if the patient is no longer responding to fluid boluses it is appropriate to initiate inotropic support for persistent hypotension.

The initiation of PPV after intubation can also result in hypotension, as PPV can result in decreased PBF and CO due to decreased systemic venous return with increased intrathoracic pressure and an increase in PVR. Appropriate ventilatory parameters during PPV include moderately elevated tidal volumes, low respiratory rate, a long expiratory time, and minimal positive end-expiratory pressure to maintain low PVR and mean airway pressure. It would be beneficial to reassess ventilation strategies and consider starting inotropic support if necessary. A summary of common agents used for hemodynamic support and their effects on the Fontan circulation is presented in Table 30.4.

The patient in this scenario is also pacemaker-dependent and sudden loss of capture can lead to hypotension. Other potential arrhythmias include junctional rhythm and atrial arrhythmias such as flutter or fibrillation. Acute episodes of atrial tachycardia should be terminated early due to the risk of rapid deterioration. In the case of refractory hypotension, potential obstruction across the Fontan pathway due to thrombus formation should be considered. In this case, it is likely that the patient will also experience significant hypoxemia. If thrombus formation is suspected, then the patient should be urgently anticoagulated with heparin, taken for cardiac catheterization, and if necessary extracorporeal membrane oxygenation (ECMO) support may be considered.

Clinical Pearl

The first step in managing intraoperative hypotension should be intravenous fluid replacement. Should hypotension persist the initiation of inotropic therapies should be considered.

https://doi.org/10.1017/9781108657341.031 Published online by Cambridge University Press

Table 30.4 Agents for the Hemodynamic Management of the Failing Fontan

Agent	Effects
Oxygen	Lowers PVR
Dobutamine α-antagonist, β-agonist	Inodilator Lowers PVR, SVR Easily titratable
Dopamine α- and β-agonist	Inotropy and vasoconstriction Increased SVR and PVR (greater than epinephrine) Tachycardia/arrhythmogenic
Epinephrine α- and β-agonist	Inotropy and vasoconstriction Increased SVR and PVR Tachycardia/arrhythmogenic
Milrinone Phosphodiesterase type 3 inhibitor	Lusitropy Lowers PVR and SVR Not easily titratable
Inhaled nitric oxide	Decreases PVR
Nitroglycerin/nitroprusside	Decrease SVR Decreases preload

PVR, pulmonary vascular resistance; SVR, systemic vascular resistance.

The ECG rhythm changes abruptly to atrial flutter with a rapid ventricular response. What management options exist?

The acute management of atrial tachyarrhythmias is dependent on the hemodynamic status of the patient. In a stable patient, management includes rate control and cardioversion and, if persistent, anticoagulation to prevent thromboembolism. Options for rate control include ß-blockers, calcium channel blockers, or amiodarone. In unstable patients, atrial tachyarrhythmias can be terminated by electrical cardioversion, overdrive pacing, or drug therapy. Pharmacologic cardioversion can be used acutely. Amiodarone is the medication used most often; however, ibutilide or sotalol may also be used. Intraoperative atrial tachyarrhythmia management, particularly if the patient becomes unstable, can be supported by guidance from the cardiac electrophysiologist. Once a patient has experienced an atrial arrhythmia, recurrences are common, thus rhythm control is generally recommended.

Atrial tachyarrhythmias occur in 40%–50% of patients with a Fontan pathway, and these patients have an increased risk of sudden death, stroke, and heart failure [7]. The occurrence of atrial arrhythmias following Fontan surgery is determined by the type of Fontan procedure, with a lower incidence of atrial arrhythmias following the lateral tunnel and extracardiac operations compared to the classic atriopulmonary Fontan [11]. Risk factors for the development of tachyarrhythmias include atriopulmonary connection, right atrial dilation, history of AV valve surgery and/or AV valve regurgitation, and a history of immediate postoperative arrhythmias.

Patients with CHD, and Fontan physiology in particular, have anatomic and physiologic factors that can make conventional cardiopulmonary resuscitation (CPR) ineffective. In general, CPR may provide 10%–30% of normal blood flow to the heart and 30%–40% of normal blood flow to the brain. These numbers are lower in those patients with already compromised pulmonary and systemic blood flows and cerebral perfusion. Critical components of high-quality CPR include minimal interruptions in chest compressions, compressions of adequate rate and depth, avoidance of leaning between compressions, and avoidance of excessive ventilation. Finally, the early use of extracorporeal life support in patients who have failed CPR has been useful when the etiology of arrest is thought to potentially be from reversible causes [12].

Clinical Pearl

Patients with Fontan physiology have anatomic and physiologic factors that can make conventional CPR ineffective. The early use of extracorporeal life support has been useful in patients in whom the etiology of arrest is thought to be from potentially reversible causes.

What considerations exist with emergent surgery as opposed to a scheduled procedure?

Understanding the proposed surgery and its implications is important to provide a safe perioperative course. If a Fontan patient has to undergo emergency surgery without time for total optimization, it is important to do a preoperative evaluation of the patient's current status and a dedicated assessment of ventricular function. These patients will require both IV hydration and resuscitation before induction of anesthesia and intraoperative monitoring of volume and acid–base status. Transfusions may be necessary depending on the type of surgery.

Postoperative pain control may present a challenge in Fontan patients undergoing major surgery. Although epidural anesthesia has been successfully used, coagulation status should be evaluated prior to its implementation. Subarachnoid block is not recommended because of associated sympathectomy, hypotension, and bradycardia, which are poorly tolerated in Fontan patients. Some centers have implemented erector spinae plane blocks for thoracic pain coverage in patients who are either coagulopathic or who have received intraoperative

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anticoagulation. These blocks are believed to be safer options, as they are performed in a superficial plane, with lower risk of neurologic sequelae from hematoma formation [13].

How should postoperative care be managed?

Postoperative management of the Fontan patient is similar to management throughout the intraoperative period. It is imperative to maintain adequate intravascular volume and ventilation, normal acid–base status and CO during the immediate recovery period. Finally, adequate analgesia is necessary to avoid increases in catecholamines potentially leading to increases in PVR. However, analgesics should be carefully titrated to avoid hypoventilation. Patients with a pacemaker should have the pacemaker reevaluated by a member of the electrophysiology team prior to discharge.

Clinical Pearl

Hemodynamic goals for postoperative care include maintaining adequate CO and avoiding increases in PVR. Adequate postoperative analgesia can assist in achieving these goals.

What is the most appropriate discharge disposition for this patient?

Due to her new onset atrial arrhythmia, this patient should be admitted to an intensive care unit for monitoring postoperatively. In general, the recovery location should be discussed with the patient's cardiologist and surgeon, and arrangements should be made prior to the day of surgery. The postoperative disposition will depend on the patient's overall preoperative status, the type of surgery performed, and the perioperative course. A Fontan patient with well-compensated physiology undergoing wisdom teeth extraction can be considered for same day discharge if the perioperative course remains uncomplicated. If there is concern that the patient will be unable to maintain an adequate volume status and CO due to nausea, vomiting, or the inability to take fluids orally, then the patient should be admitted to an inpatient unit for IV hydration. On the other hand, patients with failing Fontan circulation undergoing a low-risk procedure such as this should still be considered for inpatient recovery despite an uncomplicated perioperative course. Escalation of care should depend on the patient's ventricular function, comorbid conditions, and intraoperative course.

Should this procedure be performed at an outpatient facility or ambulatory care center?

Although wisdom teeth extractions can be safely performed at an outpatient facility, in the case of the patient with failing Fontan, surgery should take place at a facility with an appropriate support system including cardiologists, nurses, and ancillary staff comfortable managing patients with CHD, in particular those with Fontan physiology.

When would a failing Fontan be considered for heart transplant?

Patients referred for transplant due to failing Fontan circulation have two different modes of presentation. These patients are categorized into those with systolic ventricular dysfunction and those with relatively preserved systolic ventricular function but suffering from the complications of the failed Fontan circulation. The primary indication for heart transplantation is systolic ventricular dysfunction. However, despite preserved ventricular function patients who demonstrate sequelae of a failed Fontan prior to transplant are at high risk of mortality, with a greater than threefold risk of death within one year after transplant compared to those with ventricular dysfunction [14].

Conclusion

Patients with Fontan circulation are experiencing improved long-term outcomes and longer life expectancy, and hence present more often for noncardiac surgery. A comprehensive understanding of the Fontan circulation is required to provide safe care to both well-functioning and failing Fontan patients in the perioperative period.

References

1. V. G. Nasr, S.J. Staffa, D. Zurakowski, et al. Pediatric risk stratification is improved by integrating both patient comorbidities and intrinsic surgical risk. *Anesthesiology* 2019; **130**: 971–80.
2. G. P. Diller, A. Kempny, R. Alonso-Gonzalez, et al. Survival prospects and circumstances of death in contemporary adult congenital heart disease patients under follow-up at a large tertiary centre. *Circulation* 2015; **132**: 2118–25.
3. B. J. Deal and M. L. Jacobs. Management of the failing Fontan circulation. *Heart* 2012; **98**: 1098–104.
4. D. J. Goldberg, R. E. Shaddy, C. Ravishankar, et al. The failing Fontan: etiology, diagnosis and management. *Expert Rev Cardiovasc Ther* 2011; **9**: 785–93.
5. H. Senzaki, S. Masutani, H. Ishido, et al. Cardiac rest and reserve function in patients with Fontan circulation. *J Am Coll Cardiol* 2006; **47**: 2528–35.

https://doi.org/10.1017/9781108657341.031 Published online by Cambridge University Press

6. S. S. Eagle and S. M. Daves. The adult with Fontan physiology: systematic approach to perioperative management for noncardiac surgery. *J Cardiothorac Vasc Anesth* 2011; **25**: 320–34.
7. A. Karbassi, K. Nair, L. Harris, et al. Atrial tachyarrhythmia in adult congenital heart disease. *World J Cardiol* 2017; **9**: 496–507.
8. A. D. Pitkin, M. C. Wesley, K.J. Guleserian, et al. Perioperative management of a patient with failed Fontan physiology. *Semin Cardiothorac Vasc Anesth* 2013; **17**: 61–5.
9. P. D. Bailey and D. R. Jobes. The Fontan patient. *Anesth Clin* 2009; **27**: 285–300.
10. W. Wilson, K. A. Taubert, M. Gewitz, et al. Prevention of infective endocarditis: guidelines from the American Heart Association: a guideline from the American Heart Association Rheumatic Fever, Endocarditis and Kawasaki Disease Committee, Council on Cardiovascular Disease in the Young, and the Council on Clinical Cardiology, Council on Cardiovascular Surgery and Anesthesia, and the Quality of Care and Outcomes Research Interdisciplinary Working Group. *J Am Dent Assoc* 2008; **139**: 3S–24S.
11. S. Ovroutski, I. Dahnert, V. Alexi-Meskishvili, et al. Preliminary analysis of arrhythmias after the Fontan operation with extracardiac conduit compared with intra-atrial lateral tunnel. *Thorac Cardiovasc Surg* 2001; **49**: 334–7.
12. B. S. Marino, S. Tabbutt, G. MacLaren, et al. Cardiopulmonary resuscitation in infants and children with cardiac disease: a scientific statement from the American Heart Association. *Circulation* 2018; **137**: e691–782.
13. C. Noss, K. J. Anderson, and A. J. Gregory. Erector spinae plane block for open-heart surgery: a potential tool for improved analgesia. *J Cardiothorac Vasc Anesth* 2019; **33**: 376–7.
14. E. R. Griffiths, A. K. Kaza, M. C. Wyler von Ballmoos, et al. Evaluating failing Fontans for heart transplantation: predictors of death. *Ann Thorac Surg* 2009; **88**: 558–64.

Suggested Reading

Jolley M., Colan S. D., Rhodes J., et al. Fontan physiology revisited. *Anesth Anal* 2015; **121**: 172–82.

Windsor J., Townsley M. M., Briston D., et al. Fontan palliation for single-ventricle physiology: perioperative management for noncardiac surgery and analysis of outcomes. *J Cardiothorac Vasc Anesth* 2017; **31**: 2296–303.

https://doi.org/10.1017/9781108657341.031 Published online by Cambridge University Press