Long-term Issues After the Fontan Procedure

Marion E. McRae, RN, MScN, CCRN-CSC-CMC, CCN(C), ACNP-BC, ACNPC

ABSTRACT

The Fontan procedure is used to treat various serious congenital heart defects. Although many people who have had the procedure live productively into adulthood, as they age, they face several health issues due to the physiology of the Fontan circulation. This article reviews the 4 types of Fontan procedures and the changes caused by the surgery, including single-ventricle physiology, nonpulsatile pulmonary perfusion, systemic venous hypertension, and intracardiac scarring, as well as their sequelae. Key nursing assessment items and possible treatment strategies are reviewed. Additional topics, including pregnancy in patients who have undergone the procedure, infective endocarditis prophylaxis, and health-related quality of life, are briefly discussed. Options for Fontan failure, including Fontan conversion or transplantation, are presented. Potential future solutions are outlined.

Keywords: arrhythmias, cardiac surgery, congenital heart disease, Fontan failure, single-ventricle repair

The Fontan procedure has been performed since 1971 to treat various serious congenital heart defects, including pulmonary atresia with intact ventricular septum, tricuspid atresia, hypoplastic left-heart syndrome, and other complex congenital heart diseases (CHDs) requiring a single-ventricle repair pathway. Many survivors of the procedure are now adults, are starting to experience many problems as a result of Fontan physiology (known as “Fontan failure”), and are seeking care in settings other than pediatric cardiology units. They may now be seen in adult cardiology, cardiac surgery, transplant, and emergency departments; obstetrical services; and other settings as they encounter noncardiac health problems. Understanding the unique Fontan physiology and how it affects assessment and treatment of these patients has become increasingly important for nurses working in areas other than pediatric cardiac settings. For hospitals that have continued to observe adults in pediatric cardiac settings, pediatric nurses increasingly will see patients with Fontan failure; therefore, understanding the long-term outcomes of the Fontan procedure is increasingly important.

Fontan Procedure

To understand Fontan physiology, clinicians must know what type of Fontan procedure an individual has undergone. There are 4 types of Fontan procedures (see Figure 1 for a description and illustration of each type of Fontan procedure). The oldest type is the atriopulmonary (AP) Fontan as originated by Fontan and Baudet and Kreutzer et al in the 1970s. The atrioventricular (AV) Fontan was developed in

Marion E. McRae is Nurse Practitioner-Congenital Heart Program, Advanced Health Sciences Pavilion, A3400-03 Cedars-Sinai Medical Center, 8700 Beverly Blvd, Los Angeles, CA 90048 (marion.mcrae@csahs.org or memcrae1@gmail.com). This paper was presented in part at the 22nd International Symposium on Adult Congenital Heart Disease, Toronto, Ontario, Canada, on May 31, 2012. The author declares no conflicts of interest.

DOI: 10.1097/NCI.0b013e31829744c7

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Figure 1: The 4 types of Fontan procedures. (A) The atriopulmonary Fontan. The right atrial appendage is anastomosed to the pulmonary artery (PA). (B) The atroventricular (Bjork) Fontan. The right atrial appendage is anastomosed posteriorly to the right ventricle with a graft anteriorly (may or may not contain a bioprosthetic valve) bypassing the absent tricuspid valve in tricuspid atresia. (C) The lateral tunnel Fontan. The superior vena cava (SVC) is anastomosed to the PA, and the inferior vena cava is anastomosed to the PA via an intracardiac tunnel using the lateral wall of the right atrium as part of the conduit. (D) The extracardiac Fontan. The SVC is anastomosed to the PA. The inferior vena cava is anastomosed to the PA via an extracardiac synthetic conduit.
the late 1970s by Bjork et al\textsuperscript{3} for individuals with tricuspid atresia and a right ventricle of least moderate size, and it is the only Fontan variant that maintains biventricular circulation. Therefore, individuals with an AV Fontan may not necessarily experience Fontan problems if the morphological right ventricle is of sufficient size and contributes significantly to cardiac output and pulsatile pulmonary perfusion. The next iteration of the procedure, the lateral tunnel (LT) Fontan, was developed by de Leval and colleagues\textsuperscript{4} in the late 1980s. The most recent Fontan procedure is the extracardiac (EC) Fontan developed in the 1990s by Marcelletti and colleagues.\textsuperscript{5} The latter 2 Fontan procedures are sometimes referred to as total cavopulmonary connections, because the superior and inferior venae cavae are connected directly to the pulmonary circulation.

Some institutions are currently performing and evaluating hybrid Fontan procedures in the population with hypoplastic left-heart syndrome. The hybrid Fontan procedure involves construction of an EC Fontan in the cardiac catheterization laboratory using a covered stent to direct blood flow from the inferior vena cava to the superior vena cava (SVC), after a surgical setup to complete the Fontan occurs intraoperatively during the Glenn shunt procedure (SVC to pulmonary artery [PA] anastomosis) and aortic arch reconstruction.\textsuperscript{6} Medium- and long-term outcomes of the hybrid approach are lacking and are not addressed in this article.

Survival free of death or heart transplantation after the Fontan procedure is about 75\% at 10 years, 68\% at 20 years, and 54\% at 25 years. When survival free of death or transplantation is examined in perioperative survivors (to account for the higher risk of surgery in earlier Fontan eras), the long-term survival is not significantly different between Fontan types.\textsuperscript{7}

Clinicians must know whether the Fontan procedure contained a fenestration and whether the fenestration is still patent. Fenestrations are sometimes performed on the LT and EC variants of the Fontan procedure (between the conduit and the common atrium) for the purpose of allowing right-to-left shunting if venous pressures get too high in the early postoperative period. The rationale for performing a fenestration is that it may result in fewer prolonged pleural effusions and improve cardiac output as blood flow bypasses the restrictive factor in the circuit, which is pulmonary vascular resistance.\textsuperscript{8}

Fenestrations come at the expense of lower oxygen saturations (in the mid-70s to low 90s, depending on how much shunting is occurring) and pose a risk for paradoxical embolization if any air or thrombus enters the venous system and crosses over into the arterial system through the fenestration. Paradoxical embolization may result in stroke, limb ischemia, and emboli to abdominal organs. Many institutions use hydrophobic air filters (with pore sizes less than 0.40 micron) on intravenous catheters to prevent air embolization. However, these devices cannot be used when infusing blood products, lipid infusions, or other drugs with a large molecular size, such as mannitol, as these products are larger than the pore size in the filters. With fenestrations and lower saturations, compensatory erythropoiesis occurs, resulting in higher than normal hematocrit levels. Clinicians need to keep in mind that the lower the saturations, the higher the hematocrit level must be kept to permit optimal oxygenation (generally at least 45\%). Fenestrations may close spontaneously over time if there is minimal flow through them, they may be closed in the cardiac catheterization laboratory with occlusion devices, or they may be closed by tightening sutures placed around the fenestration at the time of surgery.

**Fontan Physiology**

Unique physiological changes caused by Fontan circulation (with the exception of the AV Fontan) include (1) a single-ventricle circulation (sometimes with a systemic right ventricle), (2) passive (nonpulsatile) venous flow to the lungs instead of the normal pulsatile lung perfusion, (3) systemic venous hypertension, and (4) scarring in the heart (except for the EC Fontan).

**Sequelae of Fontan Physiology**

The sequelae of the 4 major physiological changes after the Fontan procedure can result in Fontan failure with cardiac and noncardiac manifestations. The sequelae of each of these changes are reviewed later. Key points in the nursing assessment of patients who have undergone the Fontan procedure and suggestions for treatment are summarized in Table 1.

**Single-Ventricle Circulation**

The single ventricle has increased work to perform, as it must pump blood to the body and also generate enough force to drive blood
### Table 1: Assessment and Long-term Management of Patients Who Have Undergone the Fontan Procedure

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<tr>
<td><strong>Single-ventricle physiology</strong></td>
<td>Brain natriuretic peptide has been validated as an indicator of heart failure in patients with a single ventricle. Assess for lung crackles, dyspnea, orthopnea, PND, decreased exercise tolerance, or chest pain.</td>
<td>The single ventricle must pump blood to both systemic and pulmonary circulation. Cardiac output is determined by the pulmonary vascular resistance the single ventricle must overcome.</td>
<td>Restrict fluid/salt intake. Treat with diuretics and ACE inhibitors (class IIa recommendation). Avoid drugs that increase fluid retention such as nonsteroidal anti-inflammatory drugs. Avoid bradycardia or tachycardia—see intracardiac scarring section. Consider Fontan conversion.</td>
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<tr>
<td>Heart failure</td>
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<td>Systemic atrioventricular valve regurgitation</td>
<td>Auscultate for systolic murmur at the fifth intercostal space left sternal border to apex (depending on cardiac position). Assess for dyspnea, orthopnea, PND, fatigue, lung crackles, and palpitations.</td>
<td>Systemic atrioventricular valve is a morphological tricuspid valve if the systemic ventricle is a morphological right ventricle and was not designed to pump at systemic pressures for years.</td>
<td>Consider heart transplantation for severe ventricular dysfunction. Systemic atrioventricular valve repair may be considered if moderate to severe regurgitation is causing symptoms or if surgery is being undertaken for other reasons. Treat with diuretics if clinically indicated. Implement heart rate control if arrhythmias such as atrial fibrillation are present as a result of atrial stretch.</td>
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<td>Nonpulsatile pulmonary perfusion</td>
<td>Determine whether the patient has an open fenestration. Monitor oxygen saturations for increasing hypoxemia (may be significantly worse with activity). Saturations in the nonfenestrated Fontan should be about 95%-96% because of coronary venous flow entering the systemic blood flow. Saturations in fenestrated Fontans depend on the size of the fenestration and whether right-to-left shunting is occurring (generally mid-70s to low 90s). Assess for decreased exercise tolerance or progressively worsening exercise tolerance in everyday activities.</td>
<td>Increased pulmonary vascular resistance results from the lack of pulsatile pulmonary perfusion and endothelial dysfunction. Surgically created fenestration may be present to permit right-to-left shunting if systemic venous pressures increase significantly. Endothelial dysfunction may increase systemic vascular resistance. Decreased exercise tolerance results from chronically elevated pulmonary vascular resistance. Hypercapnia and hypoxemia increase pulmonary vascular resistance.</td>
<td>Avoid hypercapnia. If persistent hypoxemia with an open fenestration, consider fenestration snaring or occlusion. If fenestrated, seek orders for acceptable oxygen saturations on room air before applying oxygen. Ensure adequate hematocrit levels in cyanotic patients (&gt;0.45%). Avoid hypercapnia and excessive hypoxemia. Avoid PEEP or use very low PEEP when patient is intubated. Extubate as soon as medically safe to avoid negative effects of positive pressure ventilation on cardiac output. Assess PA pressures on cardiac catheterization. Pulmonary vasodilators may be tried for pulmonary hypertension (some evidence of effectiveness of nitric oxide and sildenafil). Objectively assess exercise tolerance every few years or as needed with significant changes via a cardiopulmonary exercise test.</td>
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<td><strong>Systemic venous hypertension</strong></td>
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<td>Elevated neck vein pressure</td>
<td>Assess neck vein pressure—will be nonpulsatile and elevated.</td>
<td>Neck vein pressure will be nonpulsatile as a result of lack of pulsatile pulmonary flow. Neck vein pressure measures mean PA pressure, not central venous pressure. Expect mean PA pressure to be about 15 mm Hg in many patients with Fontan circulation (normal mean pulmonary artery pressure). Ensure that prescribers basing treatment decisions on neck vein pressure understand this point. Indwelling thermodilution PA catheters are not used in patients with Fontan circulation because of anatomy and thromboembolic risk.</td>
<td>Label transduced neck vein pressure as “mean PA pressure” or “Fontan pressure” not “CVP.” If concerned about fluid overload, look for other signs of edema or heart failure rather than elevated neck vein pressure and treat appropriately based on findings.</td>
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<td><strong>SVC syndrome</strong></td>
<td>Assess for edema of the face and head. Assess for prominent upper chest veins.</td>
<td>Venous drainage to the lungs is via gravity, and systemic venous pressure is elevated.</td>
<td>If SVC syndrome is present, avoid head of the bed &lt; 45° (use gravity to promote drainage of the head). Salt and fluid restriction as well as diuretics may be needed if no clot is present. Assess for clot in SVC via ultrasonography. Avoid any agents that can increase the likelihood of a clot in the SVC (eg, estrogens). Educate patient about not smoking or facilitate antismoking therapy. If an SVC clot is present, anticoagulate. Avoid venous neck catheters in SVC syndrome.</td>
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<tr>
<td>Venovenous collaterals or arteriovenous malformations</td>
<td>Assess for hypoxemia, clubbing, dyspnea, and reduced exercise capacity.</td>
<td>Venovenous collaterals or arteriovenous malformations may be present or reopen as a result of systemic venous hypertension and lack of hepatic factor in the pulmonary circulation in the AP Fontan.</td>
<td>If hypoxemia is present or is worsening, assess for venous collaterals or arteriovenous malformations that could be coil embolized, or consider Fontan conversion to include hepatic venous blood in the pulmonary circulation.</td>
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<td>Chronic venous insufficiency</td>
<td>Assess for telangiectasia, peripheral edema, lower limb pruritus, lower limb skin pigmentation changes (stasis dermatitis), varicose veins, and venous leg ulcers.</td>
<td>Elevated venous pressure often is more prominent in the lower limbs. Multiple femoral cardiac catheterizations and previous deep vein thrombosis also may contribute to venous insufficiency.</td>
<td>Venous compression hosiery may be useful. Elevate lower limbs when seated.</td>
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<tr>
<td>Portal hypertension and esophageal varices</td>
<td>Observe for upper gastrointestinal bleeding.</td>
<td>High systemic venous pressure is reflected in the gastrointestinal tract.</td>
<td>Use extreme caution inserting any invasive catheters or tubes down the esophagus, such as nasogastric tubes or transesophageal echocardiography probes. If such devices need to be inserted in patients with esophageal varices, ensure that a type and screen are available in the blood bank.</td>
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<td>Liver sinusoidal dilatation, liver fibrosis, cirrhosis, arterial liver nodules, and hepatocellular carcinoma</td>
<td>Observe for enlarged and firm liver with irregular edges (liver normally at the costal margin and soft). Review liver function tests for abnormalities (may be normal as a result of the lack of inflammation). Observe for ascites. Screen all patients with Fontan circulation for hepatitis B and C, as this is an additional risk factor for liver dysfunction.</td>
<td>High inferior vena cava pressures reflected to the liver can dilate the sinusoids, cause fibrosis, cirrhosis, and arterial nodules, which can lead to hepatocellular carcinoma. Patients transfused before 1992 (date of hepatitis screening of all blood) are at particular risk for hepatitis.</td>
<td>Liver ultrasound may be indicated on the basis of laboratory and physical assessment. Liver biopsy or transhepatic gradients on cardiac catheterization may be used to assess liver function. If liver failure is present, use hepatically metabolized or excreted drugs with caution. Educate patient to avoid ethanol and liver toxins. Refer patients with hepatic abnormalities to a hepatologist for ongoing follow-up.</td>
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<tr>
<td>Hypercoagulable state</td>
<td>Observe for signs of thrombi, such as deep vein thrombosis or pulmonary emboli.</td>
<td>Deficiencies in protein C; factors II, V, VII, and X; plasminogen; and antithrombin III are common. Higher factor VIII levels may be present.</td>
<td>Patients with fenestrated Fontans or those with known thromboemboli or atrial arrhythmias should be therapeutically anticoagulated. All patients with Fontans who are not on anticoagulants should be on low-dose aspirin (75–100 mg daily). Patients with Fontans who are not on anticoagulants should be on low-dose aspirin (75–100 mg daily).</td>
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<tr>
<td>Pleural effusions</td>
<td>Assess for decreased air entry to the lung bases, hypoxemia, and dyspnea. Observe for pleural effusions on chest x-ray, dullness to percussion over the lower thorax.</td>
<td>High systemic venous pressure causes fluid to move into the pleural space.</td>
<td>Restrict salt/fluid intake. Treatment with diuretics may be required. Pleurocentesis or chest tube insertion may be required if the pleural effusions are large or if the patient is dyspneic.</td>
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<tr>
<td>Pericardial effusion</td>
<td>Observe for dyspnea, orthopnea, hypotension, and poor peripheral perfusion.</td>
<td>High systemic venous pressure causes fluid to move into the pericardial space.</td>
<td>Diagnose and monitor with serial echocardiograms. If compression of the heart is starting to occur, drainage may be required.</td>
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<td>PLE</td>
<td>Assess for diarrhea, abdominal pain, hypoproteinemia (low serum protein and albumin levels), peripheral edema, ascites, hypocalcemia with muscle tetany, and immunodeficiency with frequent infections. If suspected PLE, serum protein and albumin levels should be evaluated as well as a fecal α-1 antitrypsin level (elevated in PLE).</td>
<td>Protein leaks into the gastrointestinal tract as a result of systemic venous hypertension, inflammation, low cardiac output, and increased mesenteric vascular resistance causing loss of serum protein with resultant peripheral edema, ascites, and loss of serum calcium. Immunodeficiency results from loss of immunoglobins.</td>
<td>Treat heart failure and low cardiac output. Other possible treatments include steroids, heparin (for anti-inflammatory effects), and high protein and low-fat diet. Fenestration can be considered in the nonfenestrated Fontan. Treat with calcium supplementation as needed. Use routine vaccinations to reduce infection risk. If medical treatment fails, Fontan conversion or heart/heart-lung transplantation should be considered.</td>
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<td>Plastic bronchitis</td>
<td>Monitor for plastic-like airway casts or plugs in patients with wheezing or hypoxemia.</td>
<td>Protein leaking into airways creates obstructive casts.</td>
<td>Treatment may require bronchoscopy for removal of casts.</td>
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<tr>
<td>Fontan circuit obstruction as a result of right pulmonary vein compression</td>
<td>Observe for dyspnea. Patients may be asymptomatic.</td>
<td>In the AP Fontan, the enlarged right atrium can compress the right pulmonary vein, resulting in obstruction.</td>
<td>MRI (or CT in the presence of MRI contraindications) is used to assess for pulmonary vein compression. Fontan conversion to an EC Fontan is indicated to relieve compression.</td>
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**Intracardiac scarring**

| IART                             | Watch for heart rates > 120/min with 1:1 or 1:2 conduction to the ventricle (flutter waves may be masked and mistaken for a sinus tachycardia). Assess hemodynamic effect of IART (eg, hypotension, heart failure symptoms, palpitations, presyncope). | IART in patients with a single ventricle can result in hemodynamic compromise in a short period of time due to decreased baseline cardiac output. Holter monitor or loop recorder may be useful in diagnosing arrhythmias in patients with palpitations and other symptoms where arrhythmias are not seen on 12-lead ECG or in-hospital monitoring. | Attempt to slow rhythm down with Valsalva maneuver or antitachycardia pacing if a pacemaker is present to assess for flutter waves. Atrial electrograms can be used in patients with temporary epicardial wires or permanent pacemaker leads to diagnose IART. Amiodarone, cautious β-blockade, or other antiarrhythmic agents can be used for rate and rhythm control. Cardioversion can be used for hemodynamically compromising or recurrent IART if no clot is present in atria. |

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<td>IART (continued)</td>
<td>Use anterior-posterior defibrillator pads to convert dilated right atrium in the AP Fontan. Higher energy may be needed for cardioversion. If persistent or recurrent IART, consider anticoagulation. With all arrhythmias, there should be a search for a hemodynamic cause, such as obstruction in the Fontan circuit or ventricular dysfunction and provision of appropriate treatment. Radiofrequency ablation can be considered if there is venous access to the right atrium. Consider the right atrial maze procedure for IART if reoperation is being undertaken.</td>
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<td>Atrial fibrillation</td>
<td>Observe for atrial fibrillation and assess hemodynamic response (eg, hypertension, presyncope, dyspnea). Rapid atrial fibrillation likely will be poorly tolerated in the patient with Fontan circulation due to low baseline cardiac output.</td>
<td>Atrial stretch from systemic venous hypertension and atrial incisions predisposes patients to atrial arrhythmias.</td>
<td>As with IART, bialtrial maze procedure should be considered if Fontan conversion is being undertaken. Anticoagulate with atrial arrhythmias to avoid thromboembolism.</td>
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<td>Sinus node dysfunction</td>
<td>Observe for bradycardia, junctional rhythm, atrial escape rhythm (unusual or varying P-wave morphology with a P-wave access other than 0°–60°). Assess hemodynamic effect of sinus node dysfunction (eg, hypertension, presyncope, dyspnea).</td>
<td>Sinus node may be injured during surgery or by venous hypertension, causing chronic atrial stretch. Left isomerism may occur (2 morphological left atria with no sinus node present).</td>
<td>Avoid negative chronotropic drugs such as β-blockers, nondihydropyridine calcium channel blockers (diltiazem, verapamil), or digoxin. A permanent pacemaker should be considered if sinus node dysfunction persists (epicardial leads needed).</td>
</tr>
<tr>
<td>Ventricular tachycardia</td>
<td>Assess for nonsustained or sustained ventricular tachycardia with or without symptoms (eg, presyncope, syncope, palpitations, hypotension).</td>
<td>AV Fontans may develop ventricular tachycardia from right ventricular outflow tract as a result of right ventriculotomy scarring.</td>
<td>Amiodarone or other antiarrhythmic agents may be considered. Fontan revision to an EC or LT Fontan can be considered for AV Fontans. Implantable cardioverter-defibrillator may be considered with epicardial patches.</td>
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<td>Pregnancy/contraception</td>
<td>Comprehensive evaluation by ACHD providers should be completed prior to conception. Preconception genetics consultation should be completed if not previously performed. Assess for use of warfarin and ACE inhibitors in individuals planning pregnancy. Fetal echo should be used if patient is pregnant to assess for fetal CHD. Avoid estrogen-containing contraceptives. For patients considering surgical sterilization, avoid laproscopic sterilization.</td>
<td>Pregnant patients with Fontan circulation are best managed in centers with expertise in the management of complex CHD and high-risk pregnancies. Prepregnancy evaluation permits assessment of capacity to safely tolerate pregnancy and discontinue teratogenic drugs before conception. ACE inhibitors are contraindicated in pregnancy because of potential fetal harm. Warfarin can cause fetal embryopathy in first trimester and bleeding at delivery. Note the risk of CHD transmission to fetus. Patients with Fontan circulation are at high risk for thromboembolism; estrogen increases thromboembolism risk. Laproscopic sterilization involves carbon dioxide insufflation, head down tilt position, and positive pressure ventilation, all of which can decrease cardiac output.</td>
<td>Use alternative heart failure drugs rather than ACE inhibitors. Weigh the risks of warfarin versus low-molecular-weight heparin in various stages of pregnancy and have patient make informed decision. Progesterone-only injections/implants can be used. Hysteroscopically inserted fallopian tube stents may be placed under local analgesic and sedative agents may be a better option.</td>
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<tr>
<td>IE</td>
<td>Assess patient knowledge of IE prophylaxis use.</td>
<td>Risk of IE is present with fenestrations, history of IE, valve replacement, valve repair or graft placement during cardiac surgery (first 6 months), with prosthetic device placement within first 6 months, or with patch leaks or shunts.</td>
<td>Follow the most recent IE guidelines. Patient education about IE use, avoidance of tattoos and piercings, and good dental hygiene is recommended.</td>
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<tr>
<td>Depression</td>
<td>Assess depression by self-report or by the use of depression scales.</td>
<td>Higher rates of depression are present in patients with Fontan circulation versus healthy controls.</td>
<td>Refer to psychiatry for treatment of depression. Avoid QT-prolonging antidepressants in the presence of arrhythmias.</td>
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Abbreviations: ACE, angiotensin-converting enzyme; ACHD, adult congenital heart disease; AP, atriopulmonary; AV, atrioventricular; CHD, congenital heart disease; CT, computed tomographic; CVP, central venous pressure; EC, extracardiac; ECG, electrocardiogram; IART, intra-atrial reentry tachycardia; IE, infective endocarditis; LT, lateral tunnel; MRI, magnetic resonance imaging; PA, pulmonary artery; PEEP, positive end-expiratory pressure; PLE, protein-losing enteropathy; PND, paroxysmal nocturnal dyspnea; SVC, superior vena cava.

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across the pulmonary bed as (with the exception of the AV Fontan) no ventricle exists to pump blood through the pulmonary circulation. Cardiac output is determined by the pulmonary vascular resistance that is present in a Fontan circulation. In addition, endothelial dysfunction occurs after the Fontan procedure, likely because of abnormalities in the nitric oxide pathway, and this dysfunction may cause increased systemic vascular resistance. The additional burden of the single ventricle having to overcome this increased systemic vascular resistance to eject blood into the aorta may interact with single-ventricle dysfunction to produce heart failure.

With a systemic right ventricle, the systemic AV valve is a tricuspid valve that is not structurally designed to function at systemic pressures for years, unlike a mitral valve. Systemic AV valve regurgitation may occur and further complicate heart failure. Moderate and severe AV valve regurgitation were present in 19% and less than 1%, respectively, of patients in the Contemporary Outcomes After the Fontan Study. Systemic AV valve regurgitation can be treated by valve repair or replacement if it is moderate to severe and causing symptoms.

The use of the neurohormonal biomarker brain natriuretic peptide has been validated in adult patients who have congenital heart disease (ACHD) of varying classes of heart failure and systemic ventricular dysfunction, including patients with a single-ventricle. The role of medications, other than diuretics, to treat heart failure in the failing single ventricle is less well documented than for biventricular physiology. The Pediatric Heart Network multicenter, randomized trial failed to demonstrate any advantage in ventricular function or heart failure severity when enalapril was administered to infants with a single ventricle. A large multicenter, randomized, double-blind trial of carvedilol versus placebo failed to show any improvement in clinical heart failure outcomes in pediatric patients with heart failure, some of whom had CHD. Notable were poorer outcomes in patients with a systemic right ventricle versus a systemic left ventricle. The American College of Cardiology/American Heart Association (ACC/AHA) 2008 ACHD guidelines make a class IIa recommendation (benefit greater than risk but additional studies with focused objectives needed) that treating single-ventricle dysfunction with angiotensin-converting enzyme inhibitors and diuretics is reasonable, but further study of medications treating heart failure in this patient population, particularly in adults, is needed.

In addition, little evidence is available for the use of cardiac resynchronization therapy or implantable cardioverter-defibrillators (ICDs) in patients who have undergone the Fontan procedure. As a result of lack of access from the systemic venous system into the heart with all types of Fontans except the AV Fontan unless a fenestration is present, the leads for cardiac resynchronization therapy and ICDs need to be placed on the epicardium. Although epicardial leads can be placed with a subxiphoid approach or minithoracotomy approach, extensive adhesions may preclude such placement, necessitating sternotomy or thoracotomy versus the usual transvenous approach used in individuals with a biventricular circulation. For this reason, the leads are typically implanted at Fontan revision surgery and tunneled into the abdomen for potential generator implantation, if needed, at a later date. One detractor of inserting ICDs in patients with ACHD is the higher incidence of inappropriate shocks in younger patients with ACHD. Referral for heart transplantation is undertaken for severe ventricular dysfunction, despite optimal medical therapy.

**Passive Venous Flow to the Lungs**

The lungs are not perfused by ventricular ejection, because no subpulmonary ventricle (other than in the AV Fontan) exists. The pressure difference between the right atrium and the left atrium drives flow through the pulmonary bed. Pulmonary vascular resistance is increased because of the lack of pulsatility (pulsatility induces shear stress-mediated release of endothelium-derived nitric oxide, a pulmonary vasodilator) and endothelial dysfunction from elevated levels of endothelin-1 (a pulmonary vasoconstrictor) found in patients who have undergone the Fontan procedure. If pulmonary vascular resistance increases above baseline values, for example, with hypoxemia or hypercapnia, pulmonary perfusion will decrease. Therefore, good oxygenation and avoidance of hypercapnia are useful in avoiding elevated pulmonary vascular resistance.
resistance. Ventilating with minimal or no positive end-expiratory pressure also facilitates venous drainage into the heart and can augment cardiac output. Early extubation, if the patient is otherwise stable, also negates the negative effects of positive pressure ventilation in patients who have undergone the Fontan procedure.

Patients with Fontan physiology generally have decreased exercise capacity as pulmonary vascular resistance needs to decrease to increase cardiac output during exercise. The chronically elevated pulmonary vascular resistance in patients with Fontan physiology is a limiting factor in exercise. Mean PA pressure also is known to increase during exercise with age in normal individuals with biventricular physiology. Therefore, patients with Fontan physiology increasingly may experience further reductions in exercise capacity as they age. Cardiopulmonary exercise testing is useful every few years to objectively and serially assess exercise tolerance.

Evidence from small trials of the effectiveness of pulmonary vasodilators such as nitric oxide and sildenafil has shown improved cardiac output and/or exercise capacity and lower PA pressure in patients who have undergone the Fontan procedure. Most of these trials involved short-term therapy only. A small pilot study with bosentan, a pulmonary vasodilator that targets endothelin, failed to show significant improvement in exercise capacity, oxygen saturation, or quality of life in patients with failing Fontan circulations. Larger studies of longer use of various pulmonary vasodilators in the Fontan physiology are needed.

**Systemic Venous Hypertension**

In the Fontan procedure (except the AV Fontan), blood entering the SVC flows either directly into the PA (LT or EC Fontan) or into the right atrium, which is directly connected to the PA (AP Fontan). Therefore, jugular venous pressure does not reflect intracardiac preload but rather PA pressure. Similarly, pressure transduced from a jugular or subclavian catheter does not reflect intracardiac preload but rather mean PA pressure. As pulmonary perfusion is nonpulsatile (except in the AV Fontan), the jugular veins are nonpulsatile. Indwelling PA catheters of the thermodilution type cannot be used in patients with Fontan physiology in intensive care units because of lack of anatomic access to the pulmonary circulation from the venous circulation, except in the AV Fontan.

In the AV Fontan, one would not want to leave an indwelling PA pressure catheter across the AV connection because of the risk of thrombus. Cardiac output calculated from an indwelling PA catheter of the thermodilution type would not provide correct calculations in a patient with Fontan circulation, as the assumptions underlying the calculation of cardiac output are based on normal intracardiac anatomy. The jugular venous pressure is frequently at the jaw or higher, and pressure transduced from a neck catheter often is greater than 15 mm Hg, reflecting mean PA pressure. Most bedside cardiac monitors do not have an option to label neck-catheter venous pressures as “Fontan pressure” or “mean PA” pressure; therefore, bedside caregivers must be knowledgeable about what this number means and communicate this information to all interdisciplinary team members, so that it is not misinterpreted and used inappropriately in treatment decisions. Treating a patient with a Fontan pressure of 15 mm Hg with diuretics may result in underfilling of the single ventricle and impair cardiac output. Individual assessment and recognition of the fact that the neck vein pressure reflects mean PA pressure must occur.

Early after the Fontan procedure, systemic venous hypertension can result in SVC syndrome. Later, catheters in the neck veins and clot or stenosis of the SVC can cause SVC syndrome. Several prothrombotic factors in patients with Fontan physiology (see later discussion) can increase the likelihood of SVC clot, as can smoking and the use of estrogen-containing medications, such as oral contraceptives. Superior vena cava syndrome can cause the head to be edematous and upper chest neck veins to be prominent. Elevation of the head of the bed to greater than 45° is used to decrease edema. Ultrasound surveillance for clots should be undertaken. Salt and fluid restrictions as well as diuretics may be indicated to decrease the edema if no clot is found.

Because of the systemic venous hypertension, venous channels closed off in embryological development may reopen when exposed to elevated pressures. New venous collaterals also can form, draining to an atrium, a pulmonary vein, the coronary sinus, or the inferior vena cava. Similarly, arteriovenous malformations (AVMs), which are abnormal connections between small arteries and veins, may occur as a result of systemic venous hypertension as well.
as exclusion of hepatic blood from the pulmonary circulation (in the AP Fontan) and higher levels of vascular endothelial growth factor in patients with Fontan physiology. Venous collaterals and AVMs may divert blood from the lungs and hence may result in hypoxemia. Venous collaterals and AVMs may be treated with coil embolization in the cardiac catheterization laboratory if they cause hypoxemia. Surgical revision to include hepatic venous blood into the pulmonary circulation when collaterals and AVMs are not amenable to transcatheter management is a class I recommendation (benefit greater than risk and procedure or treatment should be performed/administered) in the ACC/AHA 2008 ACHD guidelines. Systemic venous hypertension may cause chronic venous insufficiency, resulting in varicose veins and leg ulcers. Multiple femoral cardiac catheterizations and previous deep vein thrombosis are also predictors of chronic venous insufficiency in patients with Fontan physiology. Chronic venous insufficiency may require treatment with compression hosiery. High systemic venous pressures are transmitted to the liver via the inferior vena cava and hepatic venous circulation. High systemic venous pressures can lead to portal hypertension, esophageal varices, ascites, and liver changes, such as sinusoidal dilatation, liver fibrosis, centrilobar necrosis, cirrhosis, and hepatocellular carcinoma. Upper gastrointestinal bleeding can occur with varices. Bryant et al found arterialized liver nodules in patients with Fontan physiology, with higher mean right atrial pressures and higher systemic ventricular end-diastolic pressures versus those with lower pressures. The exact pathophysiology of the liver changes in the Fontan circulation is not completely understood, but reduced cardiac output and relative hypoxemia also may play a role.

Hepatic assessment is difficult, as liver enzymes, bilirubin, and international normalized ratio may be normal or only mildly elevated, because inflammation is not the primary pathophysiological mechanism as opposed to other liver pathologies such as hepatitis. The liver edge may feel irregular and firm. Jaundice is generally a late sign. Ultrasonography, elastography, and computed tomography/magnetic resonance imaging often are used for initial liver assessment. Liver biopsy and transhepatic gradients often are required to fully assess the liver. All patients with Fontan circulation should be assessed for liver dysfunction, and those with significant liver impairment should be referred to a hepatologist for ongoing management. Patient education about avoiding further liver injury with ethanol or other liver toxins should be provided. Individuals with significant liver dysfunction pose a higher risk for reoperation and for transplantation.

Individuals who had blood transfusions prior to 1992 when hepatitis screening of blood was instituted are also at risk for hepatitis B and C infections, which may add to hepatic injury. Therefore, all adults who have undergone the Fontan procedure who have not been screened for hepatitis B and C should undergo screening. Deficiencies in protein C; factors II, V, VII, and X; plasminogen; and antithrombin III as well as higher factor VIII levels may result from poor hepatic function and cause a hypercoagulable state. Low-dose aspirin (75-100 mg daily) is suggested, or if at higher risk for thrombus, such as with atrial arrhythmias or known atrial clot, therapeutic anticoagulation is needed. The ACC/AHA 2008 ACHD guidelines recommend warfarin as a class I intervention for patients with a documented atrial shunt, atrial thrombus, atrial arrhythmias, or a thromboembolic event. A recent multicenter, randomized trial comparing warfarin and aspirin for thromboprophylaxis after the Fontan procedure in patients without atrial arrhythmias showed an overall thrombosis rate of 19% two years postoperation and no difference in thrombosis rates between the 2 treatments. Newer anticoagulants such as dabigatran have not been studied in patients with Fontan physiology. High systemic venous pressure can cause fluid to move into the pleural or pericardial space, resulting in pleural or pericardial effusions. These effusions can generally be treated medically and often do not require drainage.

Protein can leak into the intestine and other interstitial spaces and cause protein-losing enteropathy (PLE). The pathophysiology of PLE is not completely understood but likely involves a combination of systemic venous hypertension, inflammation, low cardiac output, and increased mesenteric vascular resistance, leading to decreasing mesenteric perfusion. Protein-losing enteropathy is manifest as ascites, peripheral edema, diarrhea, abdominal pain, pleural and pericardial effusions, hypoproteinemia, and frequent infections. Chronic protein loss results in hypocalcemia with possible muscle tetany.
and immunodeficiency as a result of immunoglobulin loss. Protein-losing enteropathy occurs in about 4.2% of patients who have undergone the Fontan procedure, with a median time from operation to PLE diagnosis of 10 years; PLE has a mortality rate of 50% (5-year survival rate of 49%-59%). Protein-losing enteropathy is diagnosed by clinical symptoms, a low serum protein or albumin level, and a high fecal α-1 antitrypsin level. No highly effective treatments are available for PLE, but the general approach is to treat heart failure, administer steroids (including oral budesonide) and heparin (for its binding effect on cytokines involved in inflammation known to disrupt tight junctions and contribute to protein leakage), provide a high-protein and low-fat diet, and convert AP and AV Fontans to an EC Fontan. If these options fail or the patient is considered too high risk for Fontan conversion, heart transplantation is the only treatment.

Protein leaking into the airways can cause plastic bronchitis, a condition in which proteinaceous plastic-like casts obstruct the airways. Bronchoscopy may be required for cast removal.

Intracardiac Scarring
All types of the Fontan procedure except the EC Fontan leave behind scar tissue in the heart as the incisions heal. Scar tissue along with atrial distension from venous hypertension can lead to atrial arrhythmias and sinus node dysfunction. In particular, in patients with AP Fontans, a huge right atrium develops over time, which may explain the higher incidence of atrial arrhythmias seen in this type of Fontan connection. Moderate to severe AV valve regurgitation also may increase atrial stretch. The incidence of atrial arrhythmias increases up to 50% by 15 to 20 years postoperatively and is highest in those with an AP Fontan. The LT Fontan was designed to have less atrial scarring, with the hope that the rate of late atrial arrhythmias would be lower. A recent review of late arrhythmias after the Fontan procedure found that the incidence of supraventricular arrhythmias, such as intra-atrial reentrant tachycardia (IART), was 40% to 65% for the AP Fontan, and 13% to 60% for the LT Fontan at 15 years postoperation. Similar arrhythmias existed in 8% to 15% of patients with EC Fontans, but follow-up for this group was only from 5 to 12 years postoperation. Longer follow-up to determine which procedure is superior for arrhythmia prevention is needed. When the stasis of blood caused by atrial flutter and atrial fibrillation is added to the prothrombotic changes previously described, the potential for intracardiac clot and pulmonary emboli increases further. Atrial arrhythmias also decrease cardiac output as a result of the loss of atrial contraction contribution to cardiac output. Hence, the risk of heart failure increases further in those with atrial arrhythmias.

The most common atrial arrhythmia in patients who have undergone the Fontan procedure is IART, also known as incisional tachycardia, which is a reentrant atrial arrhythmia with a constant cycle length that usually has an atrial rate of 150 to 250/min (slower than typical atrial flutter that usually has a rate of about 300/min). Often, it is conducted 1:1 or 1:2 to the ventricle. Intra-atrial reentrant tachycardia with 1:2 conduction to the ventricle can easily be mistaken as sinus tachycardia. Identification of IART can be facilitated with a Valsalva maneuver or, in patients with temporary pacemaker wires or permanent pacemaker leads, by the use of atrial electrograms. See Figure 2 for an illustration of IART. Intra-atrial reentrant tachycardia can precipitate heart failure and hemodynamic compromise within a short period of time in patients with single-ventricle physiology, such as the Fontan, and should, therefore, be treated promptly. The ACC/AHA 2008 ACHD guidelines make a class I recommendation to be mindful of the high risk for symptomatic IART in adults who have undergone the Fontan procedure. Anticoagulation will need to be started with recurrent or persistent IART. Patients with an AP Fontan are at high risk for thrombus formation due to their enlarged right atria with sluggish flow; IART only serves to increase this risk. Embolization from the right atrium will result in pulmonary embolus. One study found a silent pulmonary emboli prevalence of 17% in patients who had undergone the Fontan procedure (60% AP Fontans).

Intra-atrial reentrant tachycardia can be treated with antitachycardia pacing and can be cardioverted if clinicians can determine that no clot is present in the atrium with a transesophageal echocardiogram. Cardioversion is best performed with defibrillator pads in the anterior-posterior position, especially in...
venous atrium to perform ablation is limited in LT and EC Fontans unless a fenestration exists. Yap and colleagues demonstrated an acute procedural ablation success of 53% in adult patients who had undergone a Fontan procedure, with freedom from IART recurrence after successful ablation of 89% at 6 months, 31% at 2 years, and 15% at 4 years. Older age at ablation was a risk factor for IART recurrence. Other options for IART treatment include antitachycardia pacing, but positioning epicardial pacing leads for good sensing may be difficult as a result of postoperative adhesions. If undertaking a Fontan conversion for Fontan failure, surgeons can use a right-atrial maze procedure for IART. If atrial fibrillation is present, a biatrial maze procedure is performed.

Sinus node dysfunction is common after the Fontan operation. Loss of sinus rhythm occurs as a result of atriotomies, injury to the sinus node or its arterial blood supply during surgery, venous hypertension with chronic atrial stretch, or left isomerism (2 morphological left atria with no true sinus node rather than a morphological right and left atrium). In the Contemporary Outcomes After Fontan Procedure multicenter study, 67% of patients were in sinus rhythm after Fontan, 9% had an atrial escape rhythm, 8% had a junctional escape rhythm, and 8% were paced. β-blockers and other negative chronotropic drugs such as nondihydropyridine calcium channel blockers should be used cautiously (low starting dose and slow upward titration) in patients with Fontan physiology with arrhythmias because of the propensity for sinus node disease and should be avoided in documented sinus node disease. Treatment of sinus node dysfunction is insertion of a permanent pacemaker. Epicardial pacemaker wires are needed because of lack of access to the ventricle from the SVC as a result of the Fontan procedure, except in the AV Fontan.

Ventricular tachycardia from the right ventricular outflow tract may develop in patients with an AV Fontan, to convert the massively dilated right atrium. Higher energy (often about 200 J) may be needed to cardiovert the IART. Radiofrequency ablation has shown mixed results for IART, with recurrence rates of 32% to 100% because of the difficulty of getting transmural lesions in the thick right atrium. Venous access to the systemic

Figure 2: (A) Appearance of intra-atrial reentrant tachycardia with 1:2 conduction on the electrocardiogram. (B) Valsalva maneuver slows down heart rate and exposes IART flutter waves (at arrow). Used with permission from Walsh EP, Cecchin F. Arrhythmias in adult patients with congenital heart disease. Circulation. 2007;115:537.
Other Issues

Pregnancy and Contraception

Low cardiac output may contribute to the increased incidence of spontaneous abortion and preterm labor as well as maternal heart failure noted in patients with Fontan physiology.53,54 The ACC/AHA 2008 ACHD guidelines have a class I recommendation that all women with a Fontan connection should have a comprehensive evaluation by a physician with ACHD expertise before planning a pregnancy. In addition, the guidelines make a class III recommendation (risk greater than benefit and procedure or treatment should not be performed/administered because it is not helpful and may be harmful) that pregnancy should not be planned in patients with Fontan circulation without consultation and evaluation at a comprehensive ACHD center with experience and expertise in maternal and prenatal management of complex CHD.14 For women who plan to become pregnant, genetic counseling may be indicated, if not already performed, to assess the likelihood of CHD in the offspring. During pregnancy, the fetus should be screened for CHD. For patients being treated with warfarin, consideration must be given to the risk of first-trimester fetal embryopathy as well as bleeding at delivery and what the best course of anticoagulation is during the pregnancy. Patients being treated with angiotensin-converting enzyme inhibitors need to have these withdrawn during pregnancy because of reports of fetal injury and death in the second and third trimesters. As a result of the increased risk of thromboembolism in patients with Fontan circulation, women seeking contraception should avoid using estrogen-containing contraceptives. Progesterone-only injections and implants are reasonable options. If a woman with a Fontan circulation elects surgical sterilization, laparoscopic sterilization can cause hazardous decreases in cardiac output as a result of carbon dioxide insufflation and head down tilting as well as positive pressure ventilation needed for the procedure. Hysteroscopically inserted fallopian stents placed under local analgesic and sedative agents may be an alternative. Sterilization procedures should be planned with the congenital cardiologist collaborating with a gynecologist familiar with Fontan circulation.52

Prevention of Infective Endocarditis

The 2008 AHA/ACC ACHD guidelines14 suggest the use of antibiotic drugs for infective endocarditis prophylaxis for patients with Fontan physiology with cyanosis (this would include fenestrations), with a previous history of infective endocarditis, with valve replacement, during the 6-month period after heart surgery (such as Fontan conversion), following an interventional procedure in which a prosthetic device was placed, or with residual patch leaks. Tattoos and piercings may increase the risk for bacteremia and should be discouraged.

Health-Related Quality of Life

Survivors of the Fontan procedure have a lower health-related quality of life (HRQOL) than age-matched controls55 and their siblings.56 Ongoing studies of HRQOL, as patients who have undergone the Fontan procedure age and develop Fontan failure, will be important to guide interventions. Patients with Fontan physiology also report higher levels of depression than healthy controls.57 Surveillance and treatment of clinical depression are vital as HRQOL declines. Self-report or clinical depression scales can be used. Referral to psychiatry may be needed for treatment. If arrhythmias are found, an antidepressant needs to be selected to avoid QT-interval prolongation and proarrhythmia effects.

Fontan Failure

The collective negative effects of the single-ventricle physiology, systemic venous hypertension, and intracardiac scarring lead to a pathophysiological state often referred to as “Fontan failure.” Although some authors consider only the cardiac manifestations of the Fontan physiology when diagnosing Fontan failure, researchers increasingly recognize that multisystem assessment needs to guide the diagnosis of Fontan failure. The clinical indicators of Fontan failure are listed in Table 2. No guideline is available as to how many indicators need to be present to diagnose the condition. Although many studies have attempted to address the predictors of Fontan failure to gain knowledge about when and how to intervene, the studies suffer from imprecise and variable definitions of Fontan failure, and few studies examine long-term outcomes after surgery.57–61 Fontan failure may result in decreased quality of life. Treatment is focused on the underlying cause of the Fontan failure where specific pathophysiology can be targeted, Fontan conversion to an LT or EC Fontan, or heart transplantation.

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venous cannula when venous flow goes directly to the lungs rather than first through the heart as in the LT and EC Fontans. In this situation, blood removed from the vena cava must be oxygenated extracorporeally, or an inflow cannula must be inserted into the PA confluence behind the conduit (which may be technically difficult). The cannulas may need length or size adjustment to accommodate the anatomy or afterload. Case reports are available on the use of the Berlin heart ventricular assist device as a bridge to recovery or transplantation in patients with Fontan circulation, albeit mostly pediatric patients. Generally, single inflow and outflow cannula are used instead of dual inflow and outflow cannula unless the Fontan circulation is taken down prior to implantation. First-line treatment of acute ventricular decompensation not responding to pharmacological treatment has been extracorporeal membrane oxygenation. However, this modality is suited only to short-term support, primarily as a bridge to urgent transplantation. Total artificial hearts may offer more options for patients with Fontan circulation, but experience with these devices is limited.

Heart Transplantation
The ACC/AHA 2008 ACHD guidelines recommend heart transplantation for severe single-ventricle dysfunction or PLE (class IIb, benefit greater than risk, procedure/treatment may be considered but additional studies with broad objectives needed/additional registry data would be helpful). Heart transplantation is difficult in the Fontan population as a result of allosensitization from homografts that may have been placed during surgery prior to the Fontan and from blood transfusions required perioperatively. Technical difficulties, such as the need for PA and SVC reconstruction at the time of transplantation, make the surgery long and often require donor hearts with extended lengths of arterial and venous connections. All of these factors result in longer wait times for a heart versus other non-ACHD adult patient waiting times. Increased bleeding from adhesions and liver dysfunction are common. Early outcomes for heart transplantation are poorer among patients with Fontan circulation than among patients without CHD in most but not all studies. Heart-lung transplantation may be needed as a result of high pulmonary vascular resistance.

Fontan Conversion
The ACC/AHA 2008 ACHD guidelines recommend conversion of an AP Fontan to an EC Fontan if the patient has recurrent atrial fibrillation or flutter without hemodynamically significant anatomic abnormalities (class IIa recommendation). A concomitant maze procedure is recommended at the time of Fontan conversion. Another indication for Fontan conversion would be pulmonary venous obstruction by the enlarged right atrium in the AP Fontan. Fontan conversion surgery to an EC Fontan with arrhythmia surgery has a reported operative survival rate of 86.7% to 99.2%.

Ventricular Assist Devices, Extracorporeal Membrane Oxygenation, and Total Artificial Heart
Although commonly used in end-stage heart failure for acquired heart disease and other types of biventricular failure of congenital origin, ventricular assist devices are rarely used for single-ventricle physiology because of the technical problems of insertion of the devices. Problems include placement of the venous cannula when venous flow goes directly to the lungs rather than first through the heart as in the LT and EC Fontans. In this situation, blood removed from the vena cava must be oxygenated extracorporeally, or an inflow cannula must be inserted into the PA confluence behind the conduit (which may be technically difficult). The cannulas may need length or size adjustment to accommodate the anatomy or afterload. Case reports are available on the use of the Berlin heart ventricular assist device as a bridge to recovery or transplantation in patients with Fontan circulation, albeit mostly pediatric patients. Generally, single inflow and outflow cannula are used instead of dual inflow and outflow cannula unless the Fontan circulation is taken down prior to implantation. First-line treatment of acute ventricular decompensation not responding to pharmacological treatment has been extracorporeal membrane oxygenation. However, this modality is suited only to short-term support, primarily as a bridge to urgent transplantation. Total artificial hearts may offer more options for patients with Fontan circulation, but experience with these devices is limited.

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**Future Prospects**

Laboratory studies are currently underway to develop a low-pressure, implantable pump for the Fontan circulation that would provide pulsatility to the pulmonary circulation and reduce systemic venous hypertension.\(^7\)–\(^8\) This type of pump would effectively convert the univentricular Fontan circulation into a biventricular circulation. This type of intervention might avoid some of the current problems with Fontan physiology. Initially, such pumps likely will be applied in situations of Fontan failure as a bridge to transplant or as a bridge to recovery, with the eventual goal of creating a biventricular Fontan circulation for all individuals undergoing this surgery.\(^7\)\(^9\)

**Summary**

The Fontan procedure has enabled many infants with severe CHD to survive to adulthood and lead productive lives. However, the single-ventricle physiology, nonpulsatile pulmonary perfusion, systemic venous hypertension, and intracardiac scarring created by the Fontan procedure lead to ongoing health issues that cause Fontan failure and increase morbidity and mortality rates over time. Nurses and other healthcare professionals will play a key role in knowledgeably assessing and treating these patients as adults to maximize HRQOL.

**Acknowledgment**

The author thanks Matthew Villagonzalo for preparation of the artwork in Figure 1.

**REFERENCES**


