Long-Term Outcomes After Repair of Congenital Heart Defects: Part 2

A review of four congenital defects, and the repairs and potential complications of each. Part 2 of a two-part article.

OVERVIEW: Many congenital heart defects can be repaired, but long-term monitoring is often required to forestall possible complications. This two-part article reviews 10 common congenital heart defects, their repairs, and their common long-term outcomes, along with the implications for nurses in cardiac and noncardiac settings alike. Here, in part 2, the author reviews four defects: tetralogy of Fallot, transposition of the great arteries, congenitally corrected transposition of the great arteries, and single-ventricle defects.

Keywords: cardiac surgery, congenital heart defect, congenital heart disease

Nurses in a variety of settings are likely to encounter patients with congenital heart disease (CHD), including adults whose CHD defect was repaired in childhood. Complications of CHD repair may show up years afterward, especially in patients who have not had regular examinations with a cardiologist. This two-part article reviews 10 common CHD defects and their repair, so that nurses may better understand how long-term complications can be avoided. Last month I reviewed six defects; this month, in Part 2, I review four: tetralogy of Fallot, transposition of the great arteries, congenitally corrected transposition of the great arteries, and single-ventricle defects. For reference, Figure 1 depicts the normal heart.

TETRALOGY OF FALLOT

Tetralogy of Fallot is the most common cyanotic CHD. (Cyanotic refers to the blue color of the mucous membranes and skin in fair-skinned people caused by significantly lowered oxygen saturation levels.) Tetralogy of Fallot, which accounts for about 5.4% of CHD in adults,[1] is a constellation of four defects: ventricular septal defect (VSD), pulmonic stenosis, an aorta that overrides the left and right ventricles and receives blood from both, and compensatory right ventricular hypertrophy (see Figure 2). About 25% of those with tetralogy of Fallot have an aortic arch on the right side rather than the left side of the body.[2]

Tetralogy of Fallot is repaired by closing the VSD and relieving the right ventricular outflow tract obstruction with a pulmonary valvotomy (see Figures 2A and 2B). Older adults with repaired tetralogy of Fallot may have had a right ventricular incision to perform the repair, possibly resulting in a scar that can interfere with electrical conductivity in the heart and cause ventricular arrhythmias. An adult patient

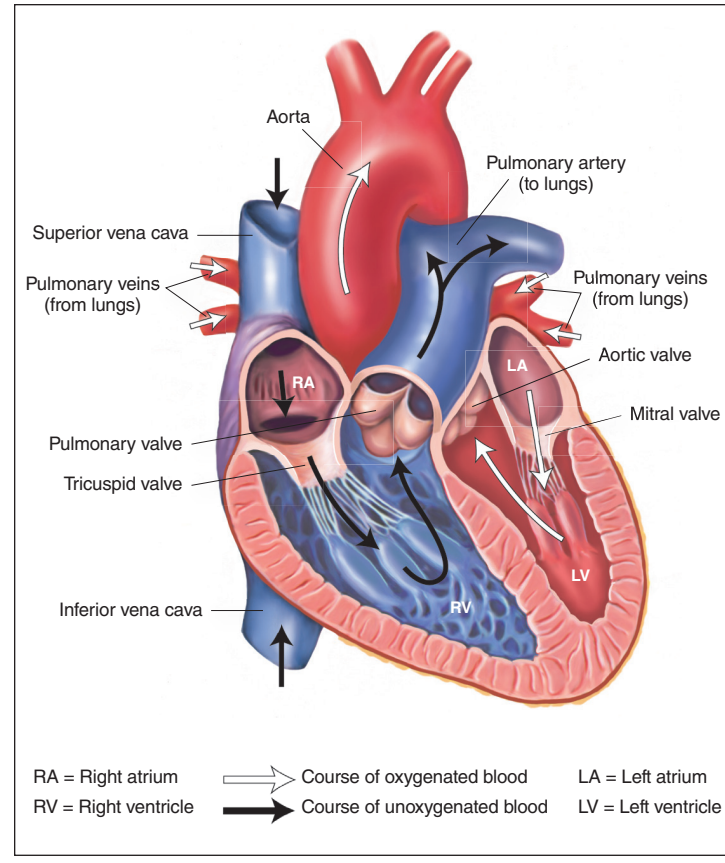
may have had a palliative shunt as an infant prior to the repair, such as a Blalock–Taussig shunt (a shunt between the subclavian and pulmonary arteries designed to increase pulmonary blood flow and relieve cyanosis). The prior use of a shunt may be evident if there is a scar on one or both sides of the chest from posterolateral thoracotomy. Nurses may not find a radial pulse on the side where the shunt was created, especially in older adults who had the subclavian artery disconnected from the aorta to make the shunt.[3] Such patients should not have blood pressure taken on that arm; it will not reflect central aortic pressure. If shunts have been constructed on both sides, the legs should be used for blood pressure measurement.

Long-term issues after tetralogy of Fallot repair include pulmonary valve regurgitation as a result of the pulmonary valvotomy.[4] Blood leaking backward through a regurgitant pulmonary valve into the right ventricle is well tolerated for many years, but eventually the ventricle will start to enlarge. When pulmonary regurgitation is present, a diastolic murmur is best heard at the left-upper sternal border. Other symptoms include dyspnea on exertion and decreasing exercise tolerance. Pulmonary regurgitation may necessitate replacement of the pulmonary valve, either surgically or with implantation of a transcatheter valve. Transcatheter valves can be implanted endovascularly (through the vascular system) or in a transventricular hybrid fashion (in which a 1-to-2-in. incision is made over the lower sternum and into the right ventricle to place the valve) if the valve can safely be anchored in the enlarged right ventricular outflow tract.

As a result of right ventricular dilatation, regurgitation of the tricuspid valve may develop. This can be suspected in cases of elevated jugular venous pressure, peripheral edema, or ascites. If there is significant tricuspid regurgitation, a systolic murmur may be heard at the left-lower sternal border; blood leaking backward enlarges the right atrium and may cause atrial arrhythmias that in turn may cause palpitations or presyncope. The 2014 guidelines for adult CHD management from the Pediatric and Congenital Electrophysiology Society (PACES) and the Heart Rhythm Society (HRS) recommend periodic ambulatory cardiac monitoring in patients with tetralogy of Fallot (Class IIA, Level B).[5] Treatment for atrial arrhythmias includes antiarrhythmia medications, catheter ablation, or a maze procedure, which is a surgical ablation requiring cardiopulmonary bypass.[6] The maze procedure is performed for some types of atrial flutter (a right atrial maze procedure) or atrial fibrillation (a biatrial maze procedure). Incisions or cryolesions created with an application of cold via a cryoprobe or radiofrequency ablation are placed in one or both atria to create one pathway for the cardiac impulse to travel from the sinoatrial (or sinus) node to the atrioventricular node. This prevents the circular movement of electrical impulses seen in atrial flutter or fibrillation. After the maze procedure, nurses will typically find a patient in a junctional rhythm—that is, a rhythm arising from the atrioventricular junction—requiring a temporary pacemaker for a couple of days.

Patients who have had a transcatheter pulmonary valve implant need to be monitored for fracture of the valve stent—a possible break in the stent, often from compression of the stent or the wringing motion of the right ventricular outflow tract during systole—which can compress the valve. The U.S. Melody Valve Trial showed that at three years a median 60% of patients who had received the valve were free of stent fractures.[7] Stent fracture is monitored by chest X-ray (frontal and lateral views). It's more common when the valve is implanted within a severely obstructed conduit or directly behind the anterior chest wall;

Figure 1. The Normal Heart



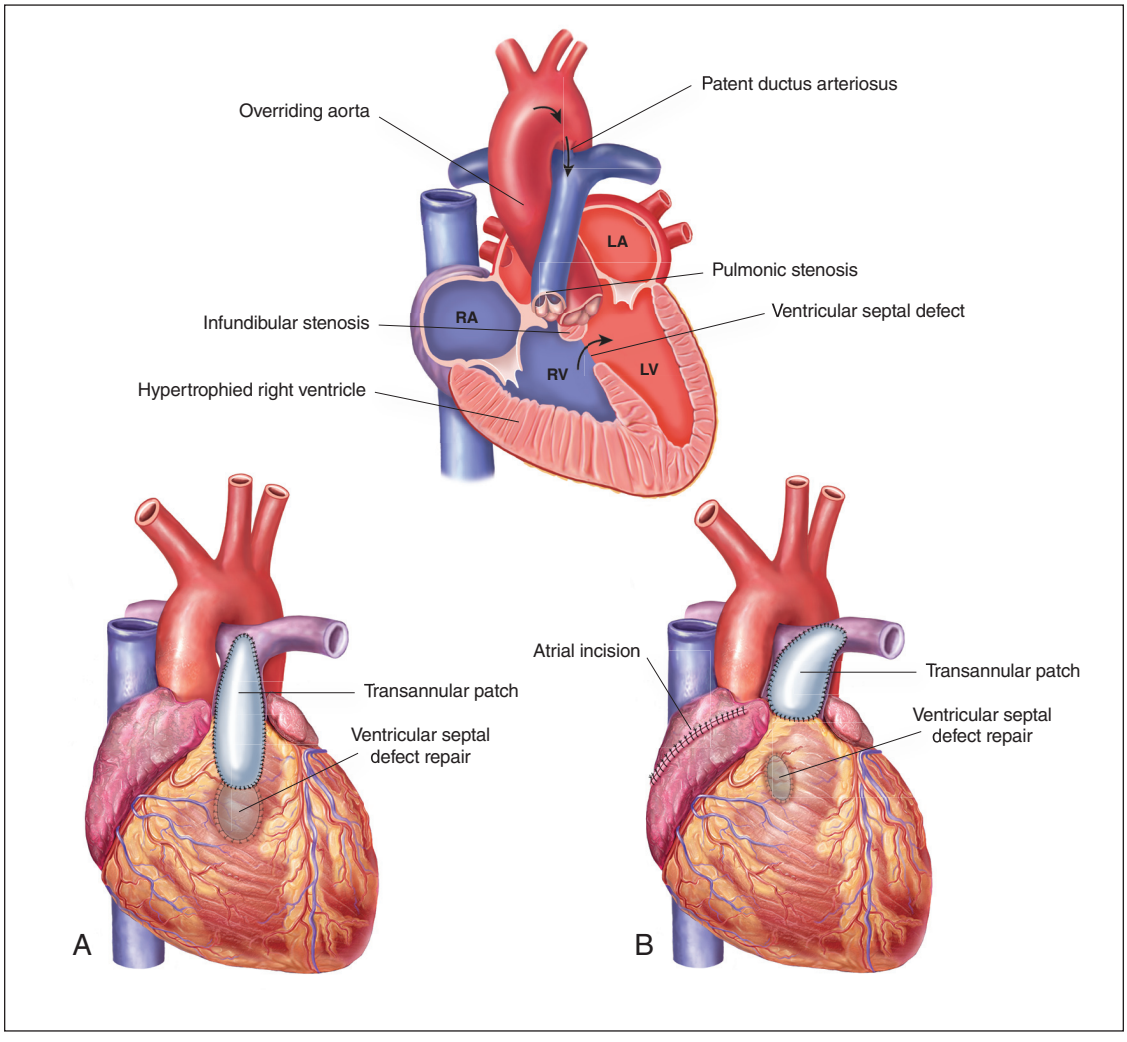
Illustrations by Anne Rains.

it's less common when the valve is implanted within a bioprosthetic valve or prestenting is performed (prestenting involves placing a rigid stent in the right ventricular outflow tract and then placing the stented valve within it).[7]

Patients with tetralogy of Fallot who have pulmonary valve replacement require infective endocarditis prophylaxis before undergoing dental and surgical procedures for the rest of their lives.[8] This includes patients with transcatheter pulmonary valves, which also increase risk of infective endocarditis.[9] After surgical valve repairs or aortic surgery where a graft is implanted, infective endocarditis prophylaxis is needed for six months, until the repair has endothelialized (become covered with tissue).[8]

Ventricular tachycardia may also occur as the right ventricle dilates. One easily identified predictor of ventricular tachycardia in a patient with tetralogy of Fallot is a QRS duration of 180 msec or greater on electrocardiography.[10] The PACES–HRS

Figure 2. Tetralogy of Fallot and Two Repairs



Components of tetralogy of Fallot include ventricular septal defect (VSD), an aorta that overrides both the left and right ventricles and receives blood from both, pulmonic stenosis (valvar, subvalvar, or both), and compensatory right ventricular hypertrophy. Transventricular repair with a transannular patch (A) is via a ventricular incision. The VSD is closed and a patch across the pulmonary valve annulus is used to enlarge the right ventricular outflow tract. Transatrial repair (B) is via an atrial incision. The VSD is closed working through the tricuspid valve (the septal leaflet may require detachment to do the repair and then is reattached). The pulmonary valve is split open via a pulmonary artery incision, and a patch is applied to the right ventricular outflow tract to permit adequate egress of blood to the lungs. LA = left atrium; LV = left ventricle; RA = right atrium; RV = right ventricle.

guidelines recommend surgical ablation guided by electrophysiologic mapping for adults with tetralogy of Fallot undergoing reoperation after an episode of monomorphic ventricular tachycardia (Class IIa, Level B).[5]

Aortic dilatation can occur over time and result in aortic valve regurgitation,[11] likely due to aortopathy and increased blood flow through the aorta before tetralogy repair, but the mechanisms are not completely understood.[12] All patients with tetralogy of Fallot should undergo regular screening to check for aortic dilatation, as well as right ventricle size and function, preferably by magnetic resonance imaging (MRI), every two or three years.[2] Aortic dissection is rare but can occur with aortic diameters exceeding 70 mm.[11] In severe cases of aortic regurgitation, an aortic-valve–sparing replacement of the ascending aorta may be needed. Replacing the dilated aortic root with a smaller graft reduces the traction on the aortic valve leaflets and decreases aortic regurgitation. Common signs and symptoms of aortic regurgitation include a diastolic murmur loudest at the left-upper to midsternal border, a wide pulse pressure with a low diastolic blood pressure, water-hammer pulses (pulses that are rapidly rising and falling), and bounding carotid pulses. Current guidelines recommend replacement of the aortic root when it is 55 mm or larger.[13]

TRANSPOSITION OF THE GREAT ARTERIES

Transposition of the great arteries is a cyanotic defect in which the aorta and pulmonary artery are connected to the wrong ventricle (see Figure 3). It accounts for about 1.8% of CHD in adults.[1] There may or may not be a VSD present.

Mustard and Senning procedures. Those born with transposition of the great arteries before the 1980s were repaired with a Mustard[14] or Senning[15] procedure (see Figure 4A). Both are atrial switch procedures, in which baffles (constructed tunnels) "switch" blood from the vena cavae into the left atrium and from the lungs into the right atrium. Baffles are formed in the Mustard procedure using the pericardium and in the Senning procedure using atrial wall tissue.

The problem with atrial switch procedures is that the right ventricle (rather than the left) remains the systemic ventricle—that is, the ventricle pumping blood to the body, a task it was not made to perform. The right ventricle is designed to pump at low pressures to the lungs, not at high pressures to the body. Likewise, the tricuspid valve at the inlet to the right ventricle was designed for low rather than high pressures. As a result of pumping blood to the body for decades, the right ventricle typically starts to fail and the tricuspid valve to leak, often in the third decade of life.[2, 16] Therefore, nurses should monitor these patients for signs and symptoms of heart failure, such as elevated blood levels of brain natriuretic peptide, dyspnea, and crackles heard in the lungs.

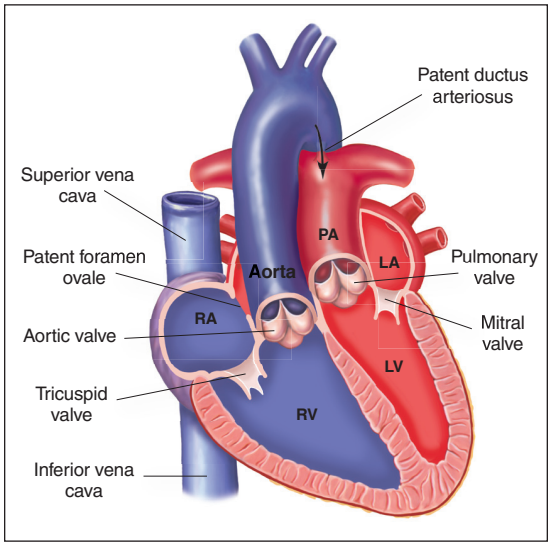
As the right ventricle fails, ventricular tachycardia and sudden death can occur. A recent study showed a 9% incidence of sudden death and about 7% of patients needing an implantable cardioverter–defibrillator (ICD).[16] One marker of sustained ventricular tachycardia and potential sudden death in a patient who has had an atrial switch is a QRS duration of more than 140 msec on the 12-lead electrocardiogram (ECG).[16] Nurses may see these patients undergoing an electrophysiology study to investigate an arrhythmia or a catheter ablation for an arrhythmia. They should closely examine patients for heart failure when there is a history of CHD and arrhythmia.

Unfortunately, there are few proven therapies for systemic right ventricular failure, unlike for left ventricular failure. Most studies of potential therapies such as angiotensin-converting enzyme inhibitors, angiotensin receptor blockers, β-blockers, and combined α- and β-blockers such as carvedilol are very small and equivocal in their findings.[17-21] Small studies suggest that resynchronization pacing may be helpful in the systemic right ventricle.[22] Heart transplantation is often needed in atrial switch patients as they age and the right ventricle fails. In fact, in a recent study of patients with CHD who had a heart transplant, 22% had a previous Mustard or Senning procedure.[23]

The atrial switch procedures involve long atrial suture lines to create the baffles. The suture lines run close to the sinus node, the heart's normal pacemaker. As a result, injury to the sinus node or to the sinus node artery can occur, causing loss of sinus rhythm—either at the time of surgery or over time—due to scarring from the atrial suture lines. Twenty percent to 50% of patients who had the Mustard procedure will develop sinus node dysfunction within 10 to 20 years.[16] Nurses should monitor for the loss of P waves on the ECG, indicating junctional rhythm, or for an abnormal or a varying shape of the P waves, indicating atrial ectopic rhythm (rhythm originating from somewhere in the right atrium other than the sinus node).

Supraventricular tachyarrhythmias (fast heart rhythms initiated in the atria or atrioventricular junction) are also common in patients who have had an atrial switch procedure, and have an incidence of 44% in early adulthood[16]; palpitations or presyncope may be early signs. These patients should have a Holter monitor or Zio patch placed to assess for arrhythmias. The PACES–HRS guidelines recommend periodic Holter monitoring in patients with previous atrial switch repair.[5] Nurses should use caution when giving medications that can lower heart rate—β-blockers and nondihydropyridine

Figure 3. Transposition of the Great Arteries



Blood from the superior and inferior vena cavae drains into the right atrium and through the tricuspid valve into a systemic right ventricle, which ejects the blood to the aorta (deoxygenated blood goes to the body). Oxygenated blood returning to the left atrium goes through the mitral valve into the left ventricle, which ejects the blood through the pulmonary valve into the pulmonary artery (oxygenated blood recirculates to the lungs only). There must be a communication between the two sides of the heart (such as a patent ductus arteriosus, as shown here; an atrial septal defect; or a ventricular septal defect) to permit survival. LA = left atrium; LV = left ventricle; PA = pulmonary artery; RA = right atrium; RV = right ventricle.

calcium channel blockers such as diltiazem can produce junctional rhythm as an adverse effect—and monitor heart rate and rhythm closely (new prescriptions for moderate or large starting doses should be questioned). Rapid supraventricular rhythms should be treated promptly, because a patient with systemic right ventricular failure may not tolerate these arrhythmias well. Nurses also need to ensure that patients with atrial flutter and fibrillation receive therapeutic anticoagulation within 48 hours to prevent embolic stroke.[5]

Permanent pacemakers are frequently needed in atrial switch patients as they age to correct sinus node dysfunction. A 2009 study showed that 23% of patients who had a Mustard procedure had subsequent pacemaker implantation.[16] Transvenous pacemaker wires (that is, wires threaded through the venous system into the superior vena cava) can significantly obstruct the superior baffle, and therefore may be placed instead on the outside of the heart (as epicardial leads).

Stenosis or leakage in the baffles that redirect blood occurs in 12% of atrial switch patients at 15 years.[24] Typically the baffle problems develop slowly over years, and therefore nurses may see only subtle signs such as increased fatigue or decreased exercise tolerance. Potential baffle problems are best evaluated by MRI or, if there are contraindications to MRI, a computed tomographic (CT) scan. Baffle stenosis can be treated with stenting in the cardiac catheterization laboratory, where baffle leaks, if significant, can also be treated with an occluder designed for use with atrial septal defects.

Arterial switch has been performed in those born with transposition of the great arteries since the mid-1980s. This procedure involves switching the aorta and pulmonary artery to reconnect the left ventricle to the aorta and the right ventricle to the pulmonary artery, with the coronary arteries removed and then reimplanted into the neo (new) aorta (see Figure 4B).[25] This has the advantage of making the left ventricle the systemic ventricle.

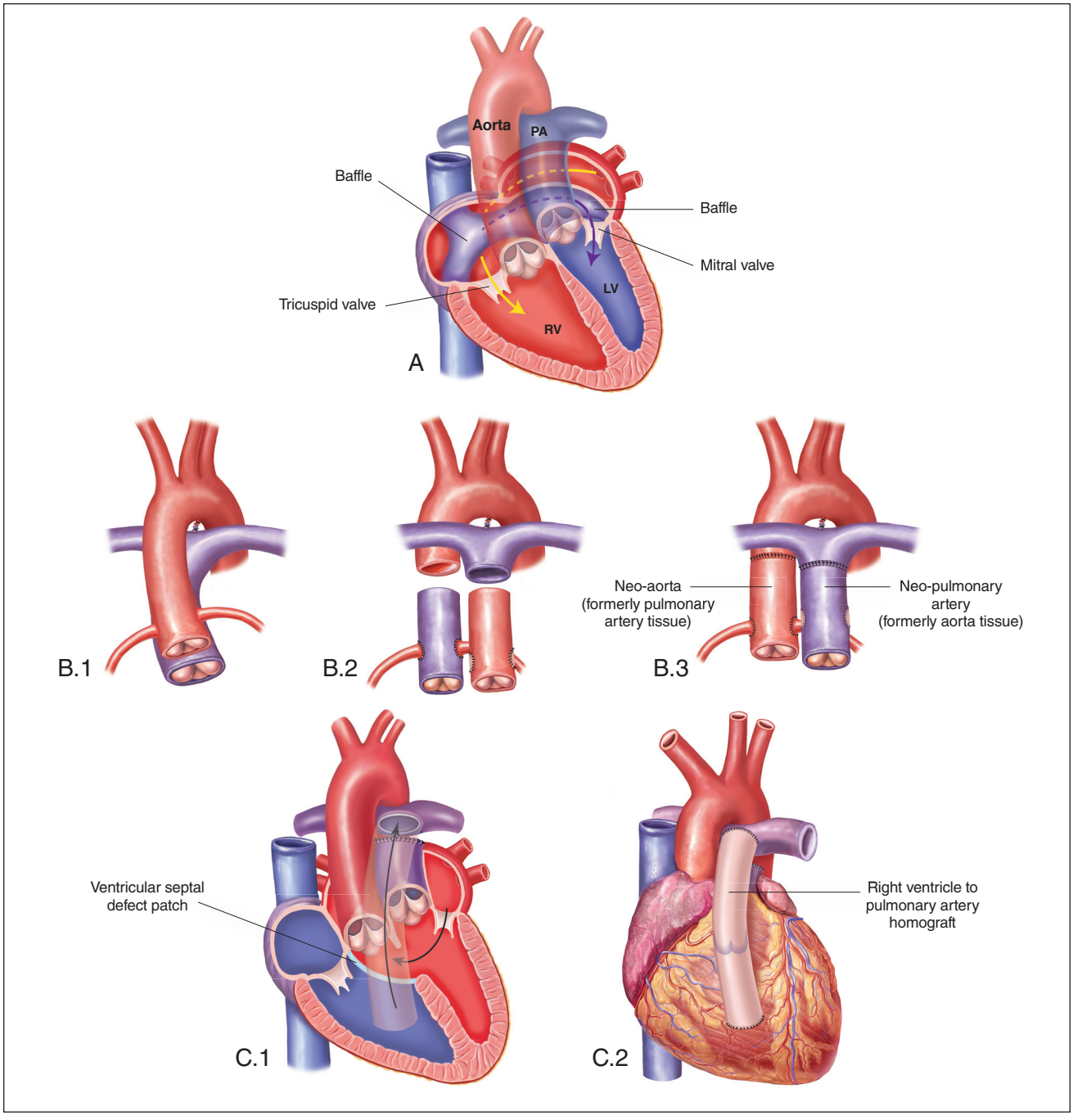
Sudden death can occur after the arterial switch. This may be associated with denervation (cutting of nerves) around the aorta during the operation.[26] Poor coronary blood flow due to reimplantation of the coronary arteries in the new aortic root can cause myocardial ischemia (usually seen soon after surgery but also in the long term). There may be no chest pain because of denervation from the procedure.

Regular ECGs should be performed to assess for abnormalities associated with myocardial ischemia or myocardial infarction, such as inverted T waves, ST-segment changes, or new Q waves. Supraventricular tachycardia and atrioventricular blocks are the most common arrhythmias seen over the long term.[27,28] Therefore, nurses should be alert to complaints of palpitations, presyncope, syncope, or an irregular pulse. If these occur, the patient should be assessed with a Holter monitor or a Zio patch.

Pulmonary artery stenosis at the site of the surgical anastomosis is the most common reason for reoperation after arterial switch, and has a prevalence of 21%.[29] Indicators of pulmonary artery stenosis may include dyspnea on exertion, decreased exercise tolerance, and a systolic murmur heard best at the left-upper sternal border and radiating to the back between the scapulae.[30]

The aortic root progressively dilates over time after arterial switch—66% of patients in one study showed enlargement that progressed over time[31]—and this may result in aortic valve regurgitation.[32] Signs and symptoms include water-hammer pulses, a wide pulse pressure often due to low diastolic blood pressure, bounding carotid pulses, a diastolic murmur heard best at the left-upper to midsternal border radiating to the apex or the carotid arteries or both, and left ventricular hypertrophy on the 12-lead ECG.

Figure 4. Repair of Transposition of the Great Arteries



In the Mustard and Senning procedures (A), blood from the vena cavae enters the right atrium and is baffled to the left atrium, where it goes through the mitral valve into the left ventricle, exiting through the pulmonary valve into the pulmonary artery. Blood returning to the left atrium from the lungs is baffled to the right atrium, where it goes through the tricuspid valve into the right ventricle, exiting via the aortic valve into the aorta. In the arterial switch procedure, the coronary arteries (B.1) are removed from the aorta and reimplanted into the pulmonary artery, and the aorta and pulmonary artery are transected (B.2) and transposed (B.3). This permits blood from the right ventricle to go to the lungs and blood from the left ventricle to go to the aorta. In the Rastelli procedure, the ventricular septal defect is closed with a patch that tunnels blood from the left ventricle into the aorta, and a conduit is inserted from the right ventricle to the pulmonary artery (C.1); an exterior view of the conduit is also shown (C.2). LV = left ventricle; PA = pulmonary artery; RV = right ventricle.

Aortic-valve–sparing replacement of the ascending aorta may be necessary to treat aortic dilatation and subsequent aortic regurgitation,[33] although few patients have needed this intervention.[31]

Rastelli procedure. When transposition of the great arteries with a VSD and subaortic obstruction is present, the Rastelli procedure[34] is performed (see Figure 4C). This involves closing the VSD with a patch that tunnels blood from the left ventricle to the aorta. A conduit is then inserted from the right ventricle to the pulmonary artery. Blood enters the right atrium from the vena cavae, goes through the tricuspid valve and into the right ventricle and out the conduit from the right ventricle to the pulmonary artery. Blood returning from the lungs to the left atrium goes through the mitral valve into the left ventricle and then into the aorta.

Long-term issues after the Rastelli procedure include the need for conduit replacement or stenting as the patient grows and needs a larger conduit or the conduit fails over time. In fact, at 10 years after the Rastelli procedure only about 25% of patients are free from surgical or transcatheter intervention for obstruction of the right ventricular outflow tract.[35]

There can be right ventricular failure and tricuspid regurgitation as the conduit fails, generally by calcification causing the valve within the conduit to become stenotic. Nurses should monitor for dyspnea on exertion, decreased exercise tolerance, and a systolic murmur heard best along the left-upper sternal border. If tricuspid regurgitation is present, there may also be a systolic murmur along the left-lower sternal border, elevated jugular venous pressure, peripheral edema, and ascites.

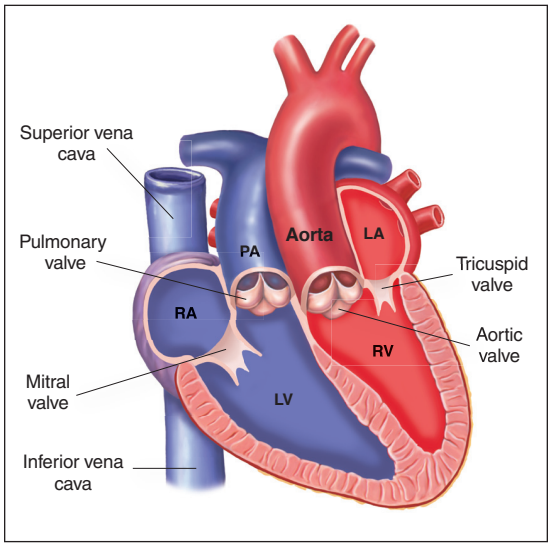
There can also be residual VSD and left ventricular outflow tract obstruction along the tunneled conduit that can require reoperation. Signs and symptoms of left ventricular outflow tract obstruction include dyspnea on exertion, lung crackles, and decreasing exercise tolerance. Residual VSD does not often create symptoms other than a systolic murmur at the left sternal border, unless there is large left-to-right blood flow across the defect. Atrial and ventricular arrhythmias can also occur,[36] and so nurses should question patients about palpitations, presyncope, and syncope, as well as feel or listen for an irregular pulse or heart rhythm. Because the Rastelli procedure involves implantation of a valved conduit, patients must be educated on the necessity for infective endocarditis prophylaxis for the rest of their lives.[8]

CONGENITALLY CORRECTED TRANSPOSITION OF THE GREAT ARTERIES

Congenitally corrected transposition of the great arteries, sometimes referred to as L-transposition of the great arteries, accounts for less than 1% of all CHD.[37] In this defect the great arteries are reversed, as they are in transposition of the great arteries, but here there is also a reversal of the ventricles (see Figure 5). This condition is referred to as "congenitally corrected" because a person born with it can survive into adulthood with no repair (the systemic and pulmonary circulations are connected without the need for intracardiac shunts). The problem, though, is that the right ventricle remains the systemic ventricle, the one pumping blood to the body, rather than the left. As can occur with transposition of the great arteries, the right ventricle is likely to fail as the person ages.

Many adults with congenitally corrected transposition of the great arteries have not had surgery to repair the circulation (to have the left ventricle become the systemic ventricle). A tricuspid valve repair or replacement may be performed if the tricuspid valve becomes regurgitant from the systemic pressure. Some people with congenitally corrected transposition of the great arteries also have Ebstein's anomaly, a rare defect in which the tricuspid valve is displaced downward into the right ventricle, the tricuspid valve leaflets are plastered to the right ventricle's wall, and the anterior tricuspid valve leaflet is large and sail-like.

Figure 5. Congenitally Corrected Transposition of the Great Arteries



Because the ventricles, as well as the great arteries, are reversed, blood from the vena cavae enters the right atrium and goes through the mitral valve into the left ventricle, exiting via the pulmonary valve into the pulmonary artery. Blood returning from the lungs to the left atrium goes through the tricuspid valve into the right ventricle, exiting via the aortic valve into the aorta. LA = left atrium; LV = left ventricle; PA = pulmonary artery; RA = right atrium; RV = right ventricle.

The upper part of the right ventricle is functionally part of the right atrium, creating a giant right atrium and a small right ventricle. This results in tricuspid regurgitation. If it is moderate to severe, a systolic murmur may be heard at the left-lower sternal border; the nurse may also note increased jugular venous pressure, ankle edema, and ascites. About 70% of people with congenitally corrected transposition of the great arteries have a VSD,[2] which may have been surgically closed. (See Part 1 of this article, published in January, for more on VSD closure.)

harder to overcome the pressure exerted by the band. When the left ventricle has pumped at systemic pressures for a few months (generally after several band tightenings over one to two years), an arterial switch procedure can be performed. Results of this procedure tend to be poorer when it's performed in late adolescence or adulthood.[41] (Some patients, because of subpulmonic obstruction, retain the ability to pump at systemic pressures from the left ventricle and may not need to retrain the left ventricle before arterial switching.)

This condition is referred to as 'congenitally corrected' because a person born with it can survive into adulthood with no repair, but the right (systemic) ventricle is likely to fail as the person ages.

Anytime after birth, patients with congenitally corrected transposition of the great arteries are at risk for complete heart block at a rate of about 2% per year[38] and requiring insertion of a permanent pacemaker. Pacemaker insertion and other electrophysiologic procedures in patients with this and other complex CHD should be performed by an electrophysiologist familiar with CHD anatomy to avoid complications.[2] Heart block occurs in congenitally corrected transposition of the great arteries because of dual atrioventricular nodes, meaning that an extra, abnormally placed atrioventricular node connects with the bundle of His.[39] Therefore, because of the potential for complete heart block in patients with congenitally corrected transposition of the great arteries—unless they have a pacemaker—nurses should use caution in giving them atrioventricular node–blocking drugs such as digoxin, β-blockers, or nondihydropyridine calcium channel blockers.

Young patients with congenitally corrected transposition of the great arteries may have undergone anatomic repair—a combination of either an atrial switch (such as a Mustard or Senning procedure) and an arterial switch (known as a double switch) or a Rastelli switch, which combines a Mustard or Senning procedure and a Rastelli procedure. The Rastelli switch was first described in 1990 by Ilbawi and colleagues[40] and is sometimes called an Ilbawi procedure. It should be performed in the first month or so of life, before the left ventricle becomes deconditioned from pumping at systemic pressures; otherwise, treatment to recondition the left ventricle must be undertaken first.

Reconditioning the left ventricle to pump at systemic pressures involves placing a band around the pulmonary artery so that the left ventricle pumps

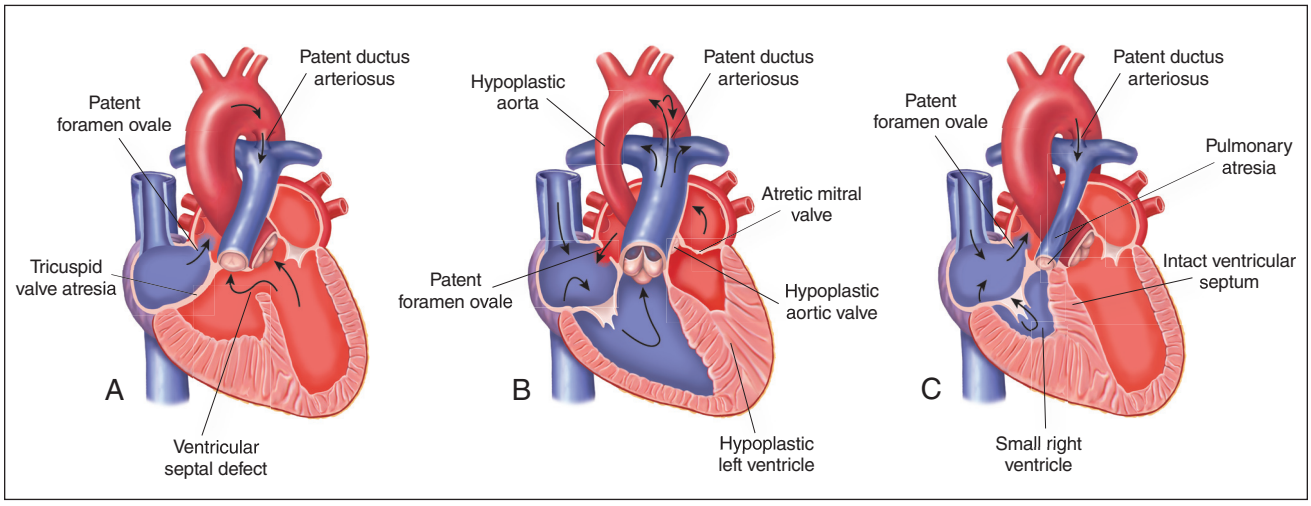
SINGLE-VENTRICLE HEART DEFECTS

Some CHD is associated with just a single functional ventricle; one ventricle may be so small that it contributes little to the pumping action of the heart. This type of CHD is repaired in children through a Glenn procedure—a bidirectional cavopulmonary anastomosis, in which a shunt is made between the superior vena cava and the pulmonary artery—followed by a Fontan procedure. The Fontan procedure is performed for many single-ventricle defects, including but not limited to tricuspid atresia, hypoplastic heart syndrome, and pulmonary atresia with intact ventricular septum (see Figure 6). Tricuspid atresia and pulmonary atresia with intact ventricular septum are both cyanotic CHDs.

There are four main variants of the Fontan procedure, three of which create a single-ventricle circulation (see Figure 7). In the atriopulmonary Fontan, blood flows passively through a surgically created connection between the right atrium and the pulmonary artery.[42] In the lateral tunnel Fontan[43] or the extracardiac Fontan,[44] blood returns from the vena cavae directly to the lungs and flows passively through the lungs without the benefit of a ventricle to pump this blood back to the left atrium (nonpulsatile flow). Venous hypertension throughout the body occurs from this passive blood flow to the lungs and in turn can cause leg varicosities, esophageal varices, portal hypertension, liver fibrosis or cirrhosis, protein-losing enteropathy from the intestine, pleural effusions, and plastic bronchitis (protein leaking into airways).[45] As a result of liver cirrhosis, hepatocellular carcinoma can occur.[46]

Nurses should monitor for evidence of venous hypertension and its complications. For example, about 60% of patients who have had a Fontan procedure

Figure 6. Common Indications for the Fontan Procedure



In tricuspid atresia (A), there is no connection between the right atrium and the right ventricle, which is generally small (although size varies). Blood flows from the right atrium across the patent foramen ovale or atrial septal defect to the left atrium and through the mitral valve to the left ventricle, where it can exit via the aorta and patent ductus arteriosus or through the ventricular septal defect to the pulmonary artery. In hypoplastic left heart syndrome (B), blood enters the right atrium, goes through the tricuspid valve into the systemic right ventricle and then through the pulmonary valve to the pulmonary artery. Blood crosses the patent ductus arteriosus to retrogradely perfuse the aortic arch and coronary arteries. Most or all of the blood entering the left atrium from the lungs crosses the patent foramen ovale or atrial septal defect, as there is either a small mitral valve (mitral valve hypoplasia) or no functional mitral valve (mitral atresia). There is also a hypoplastic (diminutive) left ventricle, and either aortic valve atresia or stenosis. The ascending aorta is very small. In pulmonary atresia (C), the connection between the right ventricle and the pulmonary artery is functionally absent. The right ventricle is small. There may be fistulas between the right ventricle and the coronary arteries (not shown).

have chronic venous insufficiency.[47] Patients with varicose veins may benefit from compression hosiery to prevent further venous insufficiency. The nurse may observe prominent veins on the chest wall or abdomen. Portal hypertension causes esophageal varices that may bleed. Therefore, extreme caution should be taken when placing a nasogastric tube or echocardiography probe into the esophagus as massive bleeding could result. Screening for varices before such procedures is recommended. Monitor the patient for bloody vomit or dark stools. Portal hypertension can also result in ascites and peripheral edema.

Nurses can assess for pleural effusion by listening for decreased air entry to the lower lobes of the lungs (dull to percussion). The patient may also experience dyspnea at rest or with exercise. Treatment may include diuretics or pleurocentesis for a large effusion. To screen for liver fibrosis in the Fontan patient, the nurse should monitor liver function tests and check for hepatomegaly (the liver would not be palpable below the costal margin unless enlarged). The liver manufactures clotting factors that may be decreased with liver fibrosis; therefore, the nurse should monitor for bleeding, especially after any surgical procedure. Fontan patients with liver dysfunction who had surgery before 1990, when routine screening of blood products for hepatitis C began, should undergo screening if they have not previously done so.

Another manifestation of high venous pressure in the Fontan patient is plastic bronchitis. Signs and symptoms include cough or bronchial casts—obstructive airway secretions—often accompanied by wheezing and hypoxemia. Acetylcysteine is sometimes used as a therapy to attempt to break up casts that plug the airway, but bronchoscopy is often needed to remove them.[48]

Venous hypertension can cause protein loss from the gastrointestinal tract (referred to as protein-losing enteropathy). This causes diarrhea, cachexia, and ascites. Patients with these symptoms should have their serum albumin level tested and a stool specimen evaluated for the α_1-antitrypsin level (high levels indicate protein-losing enteropathy).[49]

There may be an open fenestration (an artificially created connection similar to a patent foramen ovale) between the Fontan circuit and the left heart that allows mixing of oxygenated and deoxygenated blood. When pressures in the Fontan circuit rise, shunting of blood to the left heart can occur. With an open fenestration, the oxygen saturation level may be

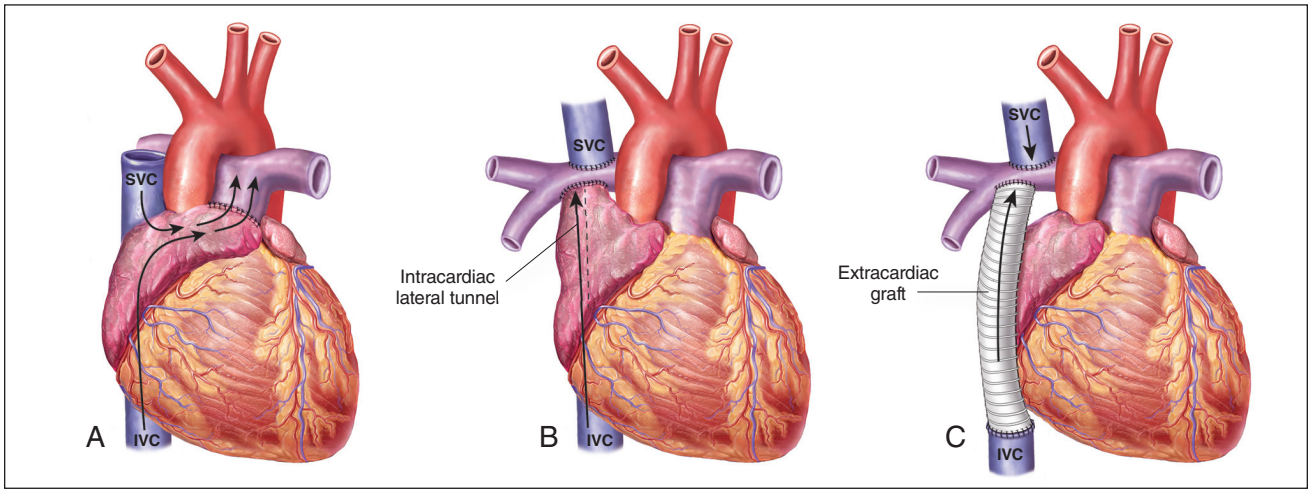
below 90% if there is bidirectional or right-to-left shunting.[50] Patients who have had an extracardiac Fontan procedure and whose coronary sinus normally drains to the left side of the heart have oxygen saturation levels of about 94%.[51] (The coronary sinus contains desaturated blood from the coronary veins; when this blood mixes with fully oxygenated blood on the left side of the heart, the oxygen saturation decreases.) The American Heart Association (AHA) guidelines for the prevention and treatment of thrombosis in CHD state that in patients who've had a fenestrated Fontan procedure, warfarin anticoagulation is reasonable until further data on the efficacy of warfarin for this indication are available.[52] Patients with fenestrations also need to practice infective endocarditis prophylaxis[8] owing to the turbulent blood flow intracardiac shunting creates.

Arrhythmias are common in patients with a Fontan circulation, particularly those involving sinus node dysfunction, intraatrial reentry tachycardia, and ventricular tachycardia.[53] Therefore, the PACES–HRS guidelines recommend periodic Holter monitoring during long-term follow-up (Class IIa, Level B).[5] Intraatrial reentry tachycardia is a slow atrial flutter (an atrial rate of 100 to 150 flutter waves per minute) that can be misinterpreted as sinus tachycardia because it typically conducts in a 2:1 fashion.[54] In other words, for every two flutter waves in the atria, only one will be conducted through the atrioventricular node to the ventricle. Therefore, the heart rate is half the atrial rate. This arrhythmia is often poorly tolerated in the Fontan patient, who has a single-ventricle circulation, and should be treated promptly with either drug or electrical cardioversion. The precautions taken for sinus node dysfunction in patients with transposition of the great arteries also apply to patients who have had the Fontan procedure. Many are on antiarrhythmic medications, have ablations, or require a permanent pacemaker or an ICD for atrial or ventricular arrhythmias or both.

Those with right-atrium-to-pulmonary-artery Fontan connections are especially prone to thrombus formation in the right atrium because of the passive blood flow and enlarged right atrium. It occurs in 3% to 16% of Fontan patients.[52] Pulmonary embolism can occur as a result.[55] A nurse should suspect pulmonary embolism in a patient with a right-atrium-to-pulmonary-artery Fontan connection who develops dyspnea, hypoxemia, or chest pain. A spiral CT scan can be performed to make the diagnosis. Patients with right atrial thrombus or pulmonary embolus will require anticoagulation per the AHA guidelines, with an international normalized ratio goal of between 2 and 3.[52] The passive blood flow through the lungs is also impeded if significant positive pressures are used to ventilate a patient. Therefore, avoiding positive end-expiratory pressure on the ventilator is ideal. Early extubation also mitigates the effects of positive-pressure ventilation, which can impair cardiac output.

The single ventricle also fails over time, especially if it is a systemic right ventricle. Many patients who had the older style Fontan (that is, the atriopulmonary connection) are now being converted to newer forms of the procedure with surgical ablation and

Figure 7. Three Variants of the Fontan Procedure



In the atriopulmonary Fontan (A), the right atrial appendage is anastomosed to the pulmonary artery. In the lateral tunnel Fontan (B), the superior and inferior vena cavae are connected directly to the pulmonary arteries, using the lateral wall of the right atrium to form the tunnel. In the extracardiac Fontan (C), the superior and inferior vena cavae are connected to the pulmonary arteries by a synthetic conduit. IVC = inferior vena cava; SVC = superior vena cava.

epicardial pacemaker implantation.[56] Still, many eventually need heart transplantation.

OTHER ISSUES COMMON TO ALL CHD DEFECTS

Pregnancy. All women with repaired CHD defects who're considering pregnancy should be encouraged to talk with a cardiologist, especially if they have arrhythmias, heart failure, mechanical valves, left ventricular outflow obstruction, or other symptoms. Often the pregnancy can proceed without significant problems if the hemodynamics are good and medications that may cause fetal harm are avoided.

Genetic syndromes commonly associated with CHD include Down, Turner, Noonan, William, Holt–Oram, and 22q11.2-deletion syndromes.[57] Adults with CHD may not have undergone genetic screening. Referral to a geneticist for screening may identify genetic syndromes that put patients at increased risk for certain types of complications. For example, 22q11.2-deletion syndrome can cause hypocalcemia, which can lead to hypotension and impaired T-cell function and raise the risk of infection. Knowledge of the potential complications allows patients to plan for future monitoring and to assess the risk of genetic transmission of the syndrome or CHD to offspring. A good online reference about genetic conditions associated with CHD is *GeneReviews* from the National Institutes of Health, at www.ncbi.nlm.nih.gov/books/NBK1116.

Those with CHD have an increased risk of having a child with CHD. The recurrence rate depends on the type of CHD and ranges from 0.6% to 8%[58]; it also depends on whether a genetic syndrome is present. Drenthen and colleagues provide an overview of the reported literature on obstetric and maternal cardiac complications associated with CHD.[58] Having fetal echocardiography at around 20 weeks' gestation can assist in detecting potential fetal problems. Delivery can then be planned at a center that specializes in handling infants with CHD as well as the potential complications that can arise in a mother with CHD.

CHD guidelines. When uncertain what to look for in a patient with CHD, the reference to start with should always be the latest adult CHD guidelines from the American College of Cardiology/AHA, which can be found at http://circ.ahajournals.org/content/118/23/e714.full.pdf.[2] There are also guidelines from Canada[13] and Europe.[37]

Resource. To access a series of videos and interactive three-dimensional animations on various heart defects, download the Heartpedia app, available on iTunes. ▼

For more than 50 additional continuing nursing education activities on cardiovascular topics, go to www.nursingcenter.com/ce.

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