Long-Term Outcomes After Repair of Congenital Heart Defects: Part 1

A review of six congenital defects, and the repairs and potential complications of each. Part 1 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 both cardiac and noncardiac settings. Here, in part 1, the author reviews six defects: bicuspid aortic valve, atrial septal defect, ventricular septal defect, atrioventricular septal defect, coarctation of the aorta, and pulmonic stenosis.

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

It's estimated that in the next few years one in 150 adults could have congenital heart disease (CHD).[1] And two-thirds of those who have CHD are adults, according to a recent Canadian study.[2] It is now known that problems can occur years after CHD repair and that ongoing cardiology follow-up can prevent or detect serious complications. But many patients whose CHD was repaired in childhood have not had regular cardiology follow-up. Indeed, many patients with CHD or with complications of CHD repair come to medical attention only by way of noncardiac care settings.

To provide high-quality care for these patients, in cardiac and noncardiac settings alike, nurses need to understand that long-term consequences can be related either to the CHD itself or to the specific repair done.

This two-part article will review the anatomy, physiology, and approach to the repair of 10 common CHD lesions (also known as defects or anomalies): bicuspid aortic valve, atrial septal defect, ventricular septal defect, atrioventricular septal defect, coarctation of the aorta, pulmonic stenosis, tetralogy of Fallot, transposition of the great arteries, congenitally corrected transposition of the great arteries, and single ventricle defect. Long-term outcomes of the treatments will be reviewed, identifying implications for nursing care. Part 1 provides an overview of six of the 10 conditions. For reference, Figure 1 depicts the normal heart.

BICUSPID AORTIC VALVE

The most common CHD, affecting 1% to 2% of the population,[3] bicuspid aortic valve is a condition

in which a patient has either two aortic leaflets or three leaflets with two fused together (see Figure 2A). Bicuspid valves often calcify and become stenotic (tight) from childhood to middle adulthood; they may also become regurgitant (insufficient or leaky). (There can be many causes of aortic stenosis and regurgitation, but this discussion is limited to bicuspid valves as the primary congenital cause.) Patients who have bicuspid valves are at risk for infective endocarditis and aortic dissection or rupture.

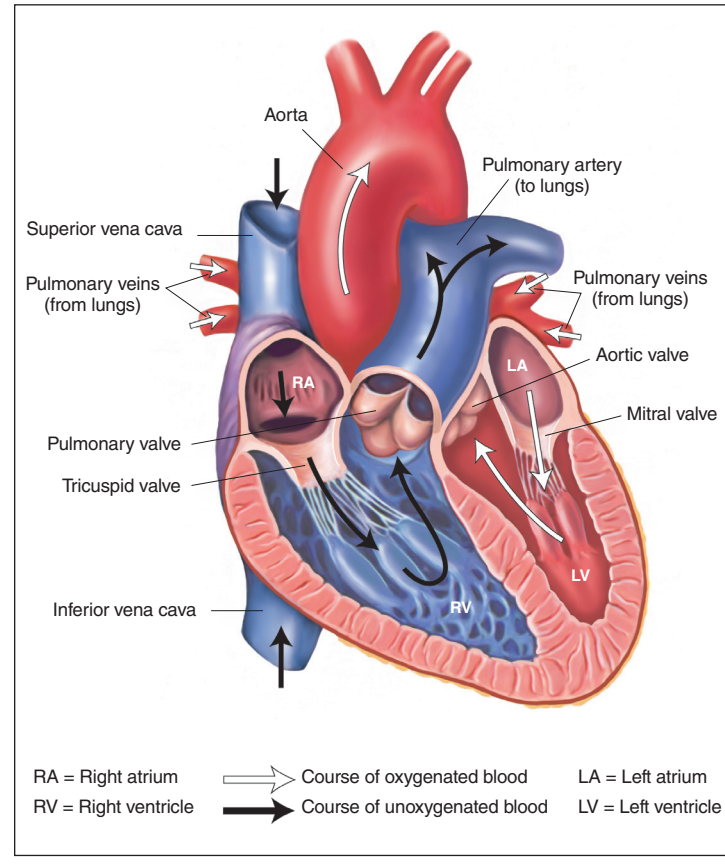
Treatments of stenotic bicuspid aortic valve may include balloon aortic valvuloplasty, in which the stenotic aortic valve is opened by a balloon, or surgical aortic valvotomy, in which the leaflets are cut apart. Balloon aortic valvuloplasty is less useful for aortic stenosis in adults than in children because valves tend to calcify with age. Surgical treatment options for the bicuspid aortic valve include valve repair (if this can be accomplished with reasonable durability over time), valve replacement with a bioprosthetic or mechanical valve, or a Ross procedure.

Lesions at the valve level (valvar or annular lesions) are not the only pathology of the aortic valve that may require intervention: lesions may also appear below the valve (subannular or subvalvar lesions) or above the valve (supravalvar or supraannular lesions), which are beyond the scope of this article. Dilatation of the aorta beyond the valvar stenosis (so-called poststenotic dilatation) can occur, creating an aneurysm; if severe enough, it may require replacing the ascending aorta.

The Ross procedure involves excision of the bicuspid aortic valve, replacement with the patient's own pulmonary valve, and implantation of a pulmonary homograft (a human-cadaver donor valve, also known as an allograft) (see Figure 2B). The coronary arteries must then be reimplanted into the neo (new) aorta (above the patient's own pulmonary valve, which is now in the aortic position).

Outcomes of the Ross procedure for regurgitant valves have been poorer than for stenotic valves.[4] Over time, the pulmonary homograft may require replacement due to stenosis or regurgitation. The patient may report decreased exercise tolerance and dyspnea on exertion when the homograft is significantly diseased. The ascending aorta in bicuspid aortic valve may also dilate due to abnormalities in the medial aortic layer and elastin fragmentation,[3] possibly resulting from smooth muscle cell death in the aorta.[5] This cell death can continue after the Ross procedure; dilatation of the new aortic root can cause aortic regurgitation (insufficiency), requiring reoperation.[6,7] If aortic regurgitation is present, the nurse will assess for wide pulse pressure, low diastolic blood pressure, water-hammer pulse (rapidly rising and falling), and a diastolic murmur best detected at the left-upper to midsternal border. The guidelines for thoracic aortic disease recommend replacing the ascending aorta in bicuspid aortic valve when the aorta measures 4 to 5 cm or when the rate of growth is more than 0.5 cm per year.[8] An average-height adult has an aortic diameter of around 4 cm.

Figure 1. The Normal Heart



Illustrations by Anne Rains.

A further concern with the Ross procedure is the risk of infective endocarditis; almost 20% of patients who had the Ross procedure needed reoperation for endocarditis in a recent large, multisite study.[9] Therefore, careful assessment for fever, night sweats, unexplained rashes, nail-bed hemorrhages, and malaise is important. Patient education on the signs and symptoms of infective endocarditis is critical so that blood cultures can be performed, and treatment begun, before severe complications develop. Patients at risk for infective endocarditis also need education on its prevention.[10] For a patient handout on infective endocarditis prophylaxis from the American Heart Association (AHA), go to http://bit.ly/1yqs3v8. The

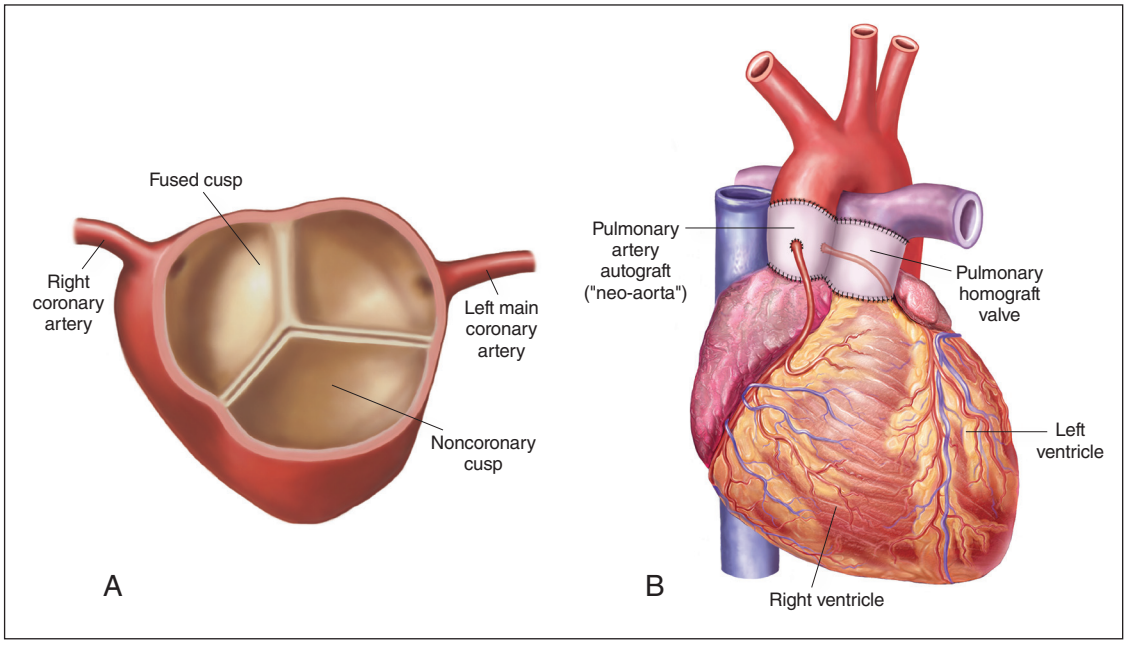
AHA recommends antibiotic prophylaxis before dental and other surgical procedures in those at highest risk for infective endocarditis; also, patients should be informed of the risks posed by tattoos and piercings.[11]

Bioprosthetic valve replacement involves replacing the aortic valve with a porcine (pig) valve or with one created from bovine (calf) or equine (horse) pericardium. When the bioprosthetic valve fails (typically, in 10 to 15 years) a transcatheter aortic valve can be implanted inside the failed bioprosthesis (in what is called a valve-in-valve procedure). Women in their childbearing years typically do not consider a mechanical valve replacement if they wish to become pregnant because of the risks posed by anticoagulation in pregnancy. Rather, they often elect to undergo a bioprosthetic valve replacement or the Ross procedure. When they no longer want to get pregnant or when the bioprosthetic valve fails, they may undergo either a transcatheter valve implantation or a mechanical valve replacement. In general, recovery is much faster from a transcatheter valve procedure because it involves an endovascular or otherwise minimally invasive procedure.

Mechanical valve replacement requires lifelong anticoagulation with warfarin and regular monitoring of international normalized ratio (INR), either at a laboratory or at home. Nurses may be involved with patient education for in-home INR-monitoring devices. If patients have insurance coverage or can afford a device, home INR monitoring is encouraged; weekly self-monitoring has been shown to result in a therapeutic INR range more often than monthly testing in an anticoagulation clinic, as well as in higher patient satisfaction and an improved quality of life.[12] The AHA guidelines for prevention and treatment of thrombosis in CHD recommend INR monitoring (a Class IIa, Level B, recommendation, meaning that the procedure is recommended, but some data in a single randomized trial or in nonrandomized studies were conflicting), and suggest that two home INR monitoring comparisons be made with a laboratory INR at the start of monitoring and every six to 12 months thereafter (a Class I, Level B, recommendation, meaning that the procedure should be performed, and that data were based on a single randomized trial or multiple nonrandomized studies).[13]

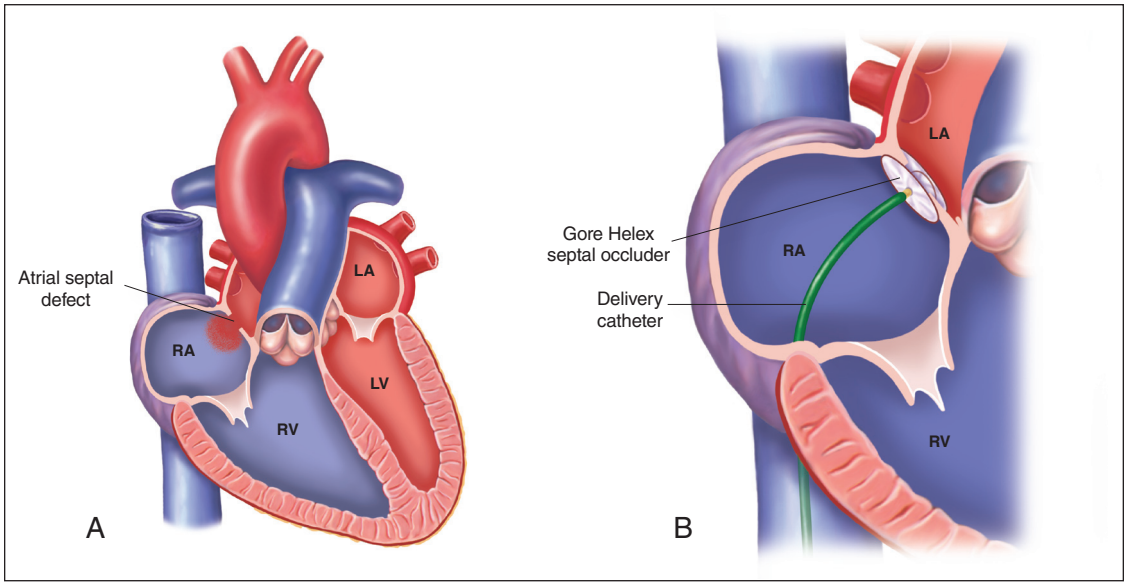
Nurses have an important role to play in educating patients on the use of warfarin, specifically in how they should maintain stable intake of vitamin K to avoid the need for frequent dosage adjustments. Patients should also report new medications to their prescriber, because many drugs, particularly antibiotics, can alter the INR. Patient education on avoiding interruption of warfarin therapy without bridging

Figure 2. Bicuspid Aortic Valve and the Ross Procedure



A valve with two leaflets (A) is caused by fusion of the right and left coronary cusps. The area of fusion is known as a raphe. In the repair (B), the patient's own pulmonary valve is implanted in the aortic position. A homograft is implanted in the pulmonary position and the coronary arteries are reimplanted into the new aorta.

Figure 3. Secundum Atrial Septal Defect and Transcatheter Occlusion



Secundum atrial septal defect is located in the center of the atrial septum (A). Blood usually shunts across the defect from the left atrium to the right atrium. The Gore Helex septal occluder is shown in a partially deployed position across the atrial septum (B). LA = left atrium; LV = left ventricle; RA = right atrium; RV = right ventricle.

with low-molecular-weight heparin or unfractionated heparin is also needed to avoid valve thrombosis.

Any woman with a mechanical valve who becomes pregnant should immediately report this to her warfarin prescriber. Anticoagulation to prevent valve thrombosis can be difficult to maintain during pregnancy and will require frequent monitoring of anticoagulation status. Discussion about possible warfarin embryopathy (birth defects due to warfarin exposure in the first trimester) should also take place.

Patients on warfarin may see advertisements for new anticoagulants that do not require INR monitoring. Patients curious about these newer anticoagulants (dabigatran [Pradaxa], rivaroxaban [Xarelto], and apixaban [Eliquis]) should be advised that they are not approved for use with mechanical valves. One trial of dabigatran and mechanical heart valves was stopped early because more thromboembolic and bleeding events occurred in the dabigatran group than in the warfarin group.[14] Also, no studies to date on these drugs have involved patients with CHD.[13] Until there is further evidence from randomized controlled studies that the newer anticoagulants are safe for use with mechanical valves, they should not be used.

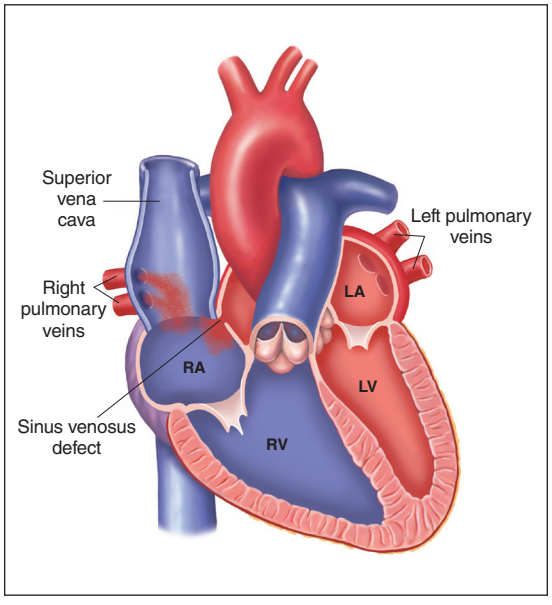
ATRIAL SEPTAL DEFECTS

An abnormally persistent opening between the atria, known as atrial septal defect (ASD), accounts for about 20% of CHD in adults.[15] It can occur alone or as a part of more complex CHD. There are four types of ASD[1]:

- secundum ASD, located in the center of the atrial septum and constituting about 75% of ASDs
- primum ASD or partial atrioventricular septal defect, constituting about 15% to 20% of ASDs, located low in the atrial septum and usually having a cleft in the anterior leaflet of the left atrioventricular valve (functional mitral valve)
- sinus venosus ASD, located high in the right atrium near the superior vena cava or, less often, low near the inferior vena cava, constituting about 5% to 10% of ASDs; can be associated with partial anomalous pulmonary venous connection (in which some but not all pulmonary veins drain to somewhere other than the posterior left atrium, often to the junction of the superior vena cava and right atrium)
- coronary sinus ASD, making up less than 1% of ASDs, involving a defective roof of the coronary sinus and often associated with a left-sided superior vena cava

ASDs permit the shunting of blood from the left heart to the right heart, unless pressures increase in the right heart and shunting occurs from right to left (the latter usually results from a cough or Valsalva maneuver). Many adults with ASD have never been diagnosed and often present with arrhythmias.

Figure 4. Sinus Venosus Atrial Septal Defect with Partial Anomalous Pulmonary Venous Connection



A sinus venosus defect (shown high near the superior vena cava) permits shunting of blood from the left to the right side of the heart. The right pulmonary veins are draining into the superior vena cava just above the right atrium rather than into the posterior left atrium. The left pulmonary veins drain normally into the posterior left atrium. LA = left atrium; LV = left ventricle; RA = right atrium; RV = right ventricle.

Secundum ASD (see Figure 3) can be repaired by transcatheter occlusion, the use of a closure device delivered percutaneously by a catheter, as long as the ASD is not too large and there is adequate tissue around the hole to anchor the device. The alternative is surgical patching, performed as an open-heart procedure requiring cardiopulmonary bypass and using a piece of autologous (the patient's own) pericardium. Transcatheter occlusion generally requires a 24-hour hospital stay, whereas surgical patching often requires several days of hospitalization. Transcatheter occlusion has been widely available for about 10 to 15 years; before then, surgical patching was the only option for ASD repair.

Sinus venosus ASD (see Figure 4) requires surgical closure, including repair of partial anomalous pulmonary venous connection, if present. One study of 236 children found that 90% of sinus venosus ASDs with partial anomalous venous connections involved the right pulmonary veins, and about 74% involved the right pulmonary veins draining into the superior vena cava.[16] In repairing this most common type of partial anomalous pulmonary venous connection, the abnormally draining pulmonary veins are redirected via a pericardial patch through the sinus venosus defect into the left atrium using one of a variety of surgical techniques. A patch to enlarge the superior vena cava may also be needed.[16, 17] Long-term outcomes after such repair are excellent, with a low incidence of pulmonary vein stenosis or other complications; repair of inferior sinus venosus defects and left-sided partial anomalous pulmonary venous connection are less common and more complicated and have poorer outcomes.[16]

There are two commercially available ASD closure devices used in the United States, the Gore Helex septal occluder (shown in Figure 3B) and the Amplatzer septal occluder (shown at http://links.lww.com/AJN/A63). The devices have a double disk that deploys on either side of the atrial septum and sandwiches the rim around the defect in order to anchor it. Fibers in the devices allow tissue to grow over and into the device. A balloon-sizing procedure assesses the size of the defect and determines the correct-size device. A catheter containing the collapsed device is threaded from the femoral vein into the right atrium, across the ASD, and into the left atrium; the device is pulled back snugly against the atrial septal rim, and the remaining disk is deployed into the right atrium. (To see a video of ASD device placement, click the video icon on the iPad edition of this article.) In children, low-dose aspirin therapy is recommended for at least six months after the procedure, until the device is endothelialized (covered with tissue). In adolescents and adults, low-dose aspirin and another antiplatelet agent (usually clopidogrel [Plavix]) are recommended for three to six months.[13]

Complications of transcatheter ASD closure include erosion of the atrial wall or the aorta due to pressure the device may exert on these structures. In October 2013, the Food and Drug Administration (FDA) issued a warning of tissue erosion with the Amplatzer device that can cause "life-threatening emergencies that require immediate surgery."[18] Erosion is a rare complication that results in cardiac tamponade and has been reported only with the Amplatzer device. The patient will report chest pain, dyspnea, and dizziness. Patients should be told to call 911 if they have such symptoms. There can be residual leaking around or through these devices, although the leak generally closes within days to months.[19] Periodic echocardiography to assess device position and tissue erosion is warranted. Patients who have had transcatheter closure or surgical closure of ASD should use infective endocarditis prophylaxis for six months, per the AHA guidelines.[10]

Patients who have an unrepaired ASD or a residual leak across or around the ASD occluder should

continue to practice infective endocarditis prophylaxis; the device will not properly endothelialize with a jet of blood going from the left to the right atrium.[10] As well, the nurse should debubble IV and hemodynamic monitoring lines before connecting them to a patient with residual interatrial shunting; this will prevent embolization and possible stroke. Hydrophobic air filters designed for either IV or hemodynamic monitoring lines can be used to prevent air emboli.[20] These filters are inserted at the distal end of the IV tubing, just prior to the patient connection, to eliminate air from the IV line. They must be primed with IV fluid before insertion. A transcatheter device closure of ASD may prevent access across the atrial septum or block the ability to ablate in cases of arrhythmia.

The long-term outcomes of transcatheter closure of ASD are unknown. To date, follow-up to a median of 10 years in surgical repair and three years in transcatheter repair has reported comparable death rates, risk of postprocedure atrial arrhythmias (about 9%), and stroke.[21,22] Functional capacity also appears similar between the two closure methods.[22] The right heart remodels to a smaller size, lessening the risk of atrial and ventricular arrhythmias.[23]

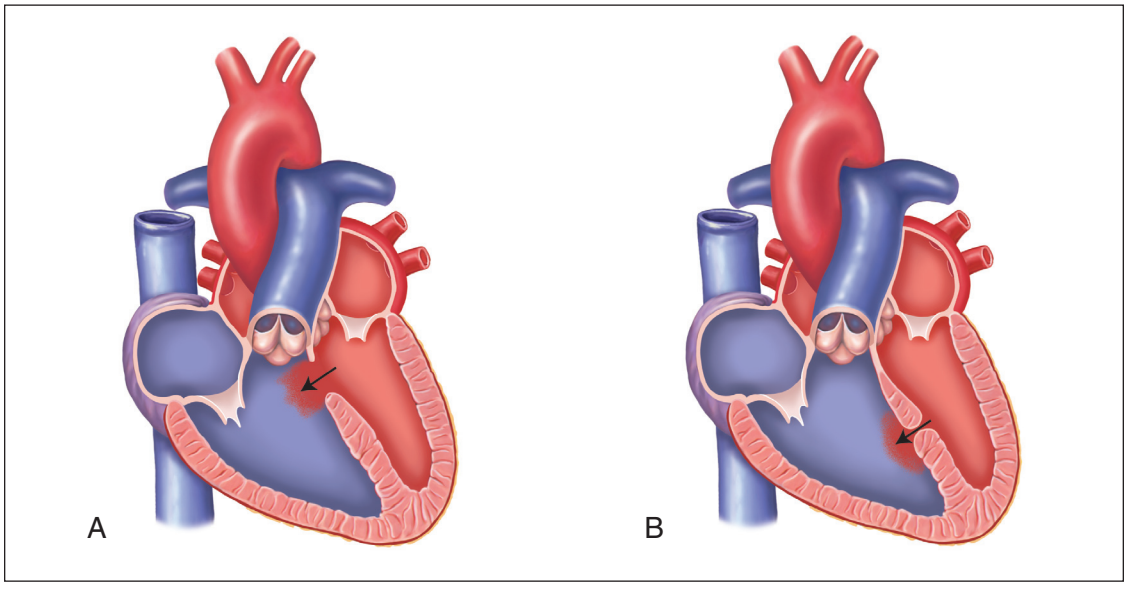
Several studies demonstrate that atrial arrhythmias are more common after surgical repair of ASD at an older age.[24-26] Increased incidence of atrial arrhythmias has been noted after transcatheter closure in adults as well.[22] Therefore, the nurse should ask patients about palpitations, presyncope, or syncope. For patients who are not currently hospitalized and on a cardiac monitor, a Holter monitor (a 24-to-72-hour ambulatory cardiac monitor) or Zio Patch can be used to detect arrhythmias. The Zio Patch is a 14-day form of continuous electrocardiographic monitoring that showed a higher rate of arrhythmia detection than Holter monitoring in one study.[27] It consists of a patch attached to the chest with adhesive and can tolerate some water exposure, unlike the Holter monitor.

VENTRICULAR SEPTAL DEFECTS

Ventricular septal defect (VSD) makes up about 20% of CHD in adults.[15] This section addresses VSD that occurs as a sole defect rather than as part of a more complex CHD or as a consequence of myocardial infarction. The two most common types of VSD are membranous (also known as perimembranous, accounting for about 75% of VSDs) and muscular (accounting for 10% to 15% of VSDs).[1] Other less common types of VSD make up the remainder. VSDs are associated with Down syndrome.

Membranous VSDs occur in the upper, thinner, membrane-like portion of the ventricular septum, with blood shunting from the left to the right ventricle (see Figure 5A). Depending on the size of the defect, the extra blood volume can overload the right heart and lungs, resulting in right-heart failure. The right coronary leaflet of the aortic valve can prolapse

Figure 5. Membranous and Muscular Ventricular Septal Defects



The membranous ventricular septal defect (A) is located high in the ventricular septum underneath the aortic valve. The right coronary leaflet of the aortic valve can prolapse into the defect, causing aortic regurgitation. The muscular ventricular septal defect (B) is located lower in the muscular septum. Multiple defects may be seen.

into the VSD, causing partial closure of the VSD and aortic regurgitation that overloads the left ventricle with blood that may cause left-heart failure.

Muscular VSDs are located in the lower, thicker, more muscular portion of the ventricular septum (see Figure 5B). Often, multiple muscular VSDs are present. They are likely to close spontaneously during the first few years of life and may not require closure if they're of little hemodynamic significance.[1]

repair if it is prolapsing and causing aortic regurgitation. Surgical closure of a VSD requires several days of hospitalization. Infective endocarditis prophylaxis is recommended for six months after transcatheter closure or surgical repair, unless there is a residual VSD, when it should be continued indefinitely.[10]

Long-term issues after either surgical or transcatheter VSD closure can include residual shunting across the ventricular septum. It can be difficult to

Currently, only muscular ventricular septal defects can be closed by transcatheter methods in the United States.

Currently, only muscular VSDs can be closed by transcatheter methods in the United States (devices to close membranous VSDs are in use in other countries but are not currently approved by the FDA for that use here). A recent study compared the transcatheter occluder and surgery for closure of membranous VSD in children and found no differences in the rates of closure or adverse events.[28] But the transcatheter device can be used only in patients weighing more than 6 kg (about 13 lbs.), and often membranous VSD requires closure because of heart failure before a child reaches that weight. There is also concern about trapping the aortic valve or tricuspid or mitral valve leaflets in the devices and causing valvar regurgitation and complete atrioventricular block because the devices put pressure on the septum close to the left and right bundle branches.

Transcatheter closure of muscular VSD is similar to that of ASD, except that the devices used in the former have more distance between the disks to accommodate sandwiching against the thicker ventricular septum (to see an Amplatzer muscular VSD occluder, go to http://links.lww.com/AJN/A63). Patients who have had transcatheter VSD closure take aspirin for six months after the procedure or longer if there is a residual VSD after device closure.[13] Adults undergoing this procedure may be prescribed a second antiplatelet agent such as clopidogrel. Transcatheter VSD closure usually requires a 24-hour hospitalization.

Before the last decade, closure of muscular VSD was performed surgically using a patch or sutures (for a very small VSD). To close membranous VSD, the leaflet of the tricuspid valve attached to the ventricular septum may be detached to repair the defect and then reattached. The aortic valve may require

see multiple muscular VSDs; they often traverse the ventricular septum obliquely and lie so deeply in the ventricle that they can be missed in surgery.[29] When residual shunting occurs, blood can shunt from the high-pressure left ventricle to the right ventricle, and it can reverse from right to left during coughing and the Valsalva maneuver. Therefore, the nurse must take precautions to prevent air embolization to the left heart and brain (as described earlier).

There can be residual intraventricular conduction problems after surgical VSD repair that can result in heart block. Membranous VSD patches are placed close to the atrioventricular node of the heart. One study found that the rate of pacemaker implantation in the immediate postoperative period or later was lower than 1%; older patients may have had a second- and third-degree heart block.[30] Hence, nurses may see patients who had a VSD repair years prior having to undergo pacemaker generator replacement. Because the VSD patch is placed near the right bundle branch and slows conduction through it, right bundle branch block is commonly seen on the 12-lead electrocardiogram (a widened QRS complex that is triphasic with inverted T waves in lead V_1).

Many years ago, VSD was repaired by an incision in the ventricle called a ventriculotomy, a procedure associated with long-term scarring and ventricular tachycardia.[31, 32] Although rare in the patient with VSD, syncope should be considered a consequence of arrhythmia; a Holter monitor or Zio Patch should be applied and an electrophysiology study performed to determine the inducibility of ventricular tachycardia. An implantable cardioverter–defibrillator is indicated if ventricular tachycardia is present or the patient has survived a sudden death event.

When the aortic valve has been repaired because of prolapse with a membranous VSD, the valve can become regurgitant again over time, and the nurse should expect to hear a diastolic murmur along the left-upper to midsternal border. Water-hammer pulses may be present, as well as a wide pulse pressure with a low diastolic blood pressure.[33] Nurses should be alert for signs and symptoms of left-heart failure, such as lung crackles and dyspnea on exertion. Brain natriuretic peptide levels can be monitored; an elevation indicates heart failure.

ATRIOVENTRICULAR SEPTAL DEFECTS

Atrioventricular septal defect (AVSD; also known as atrioventricular canal defect) makes up about 2.5% of CHD in adults.[15] The two major types of AVSD are partial (sometimes also called primum ASD) and complete. Here I will discuss AVSDs that are balanced (with equal-size left and right ventricles). Unbalanced AVSDs undergo repair via a single ventricle pathway ending with the Fontan procedure (which is described in Part 2).

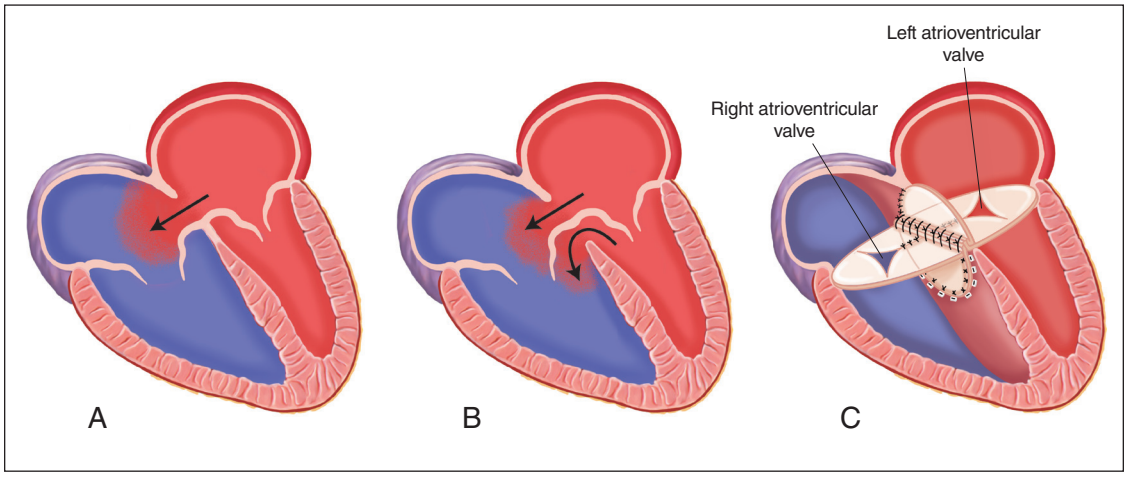
Partial AVSD is composed of an ASD in the lowest part of the atrial septum (see Figure 6A), near the tricuspid and mitral valves (or, less often, in the upper ventricular septum). The left atrioventricular valve (the mitral valve) is usually abnormal, generally with a cleft in the anterior leaflet.[34] The cleft usually causes mitral regurgitation (blood leaking backward from the left ventricle to the left atrium).

Complete AVSD has an ASD low in the atrial septum and a VSD high in the ventricular septum, as well as a combined five-leaflet atrioventricular valve rather than a separate tricuspid and mitral valve (see Figure 6B). Complete AVSD is more often seen in people with Down syndrome and partial AVSD in people without Down syndrome.[1]

Repair of a partial AVSD involves patching the septal defect and suturing the cleft in the mitral valve. Repair of a complete AVSD involves patching both atrial and ventricular septal defects with either a single- or a double-patch technique. The complete atrioventricular valve is separated into two valve orifices by suturing together the middle (bridging) leaflets (see Figure 6C).[34]

Long-term problems after AVSD repair include complete heart block that may require a permanent pacemaker and residual AVSD, which requires the same embolic and infective endocarditis prophylaxis as described for ASD and VSD. The repaired mitral valve may leak again years after repair, in which case a new repair of the valve may be indicated; if that's not possible, the mitral valve is replaced in about 7% to 10% of patients.[35] With longstanding significant left atrioventricular (mitral) valve regurgitation, the left atrium enlarges and predisposes the patient to atrial fibrillation. Therefore, the nurse should monitor for atrial tachyarrhythmias, irregular heart rates, or palpitations. Patients with atrial fibrillation sustained for more than 48 hours require therapeutic anticoagulation[36] to prevent left atrial thrombi that could cause a stroke. Also, dyspnea on exertion is common in patients with left atrioventricular valve regurgitation. Mitral regurgitation produces a "blowing" systolic murmur at the apex of the heart that radiates to the left axilla.

Figure 6. Partial and Complete Atrioventricular Septal Defects and Patching Repair



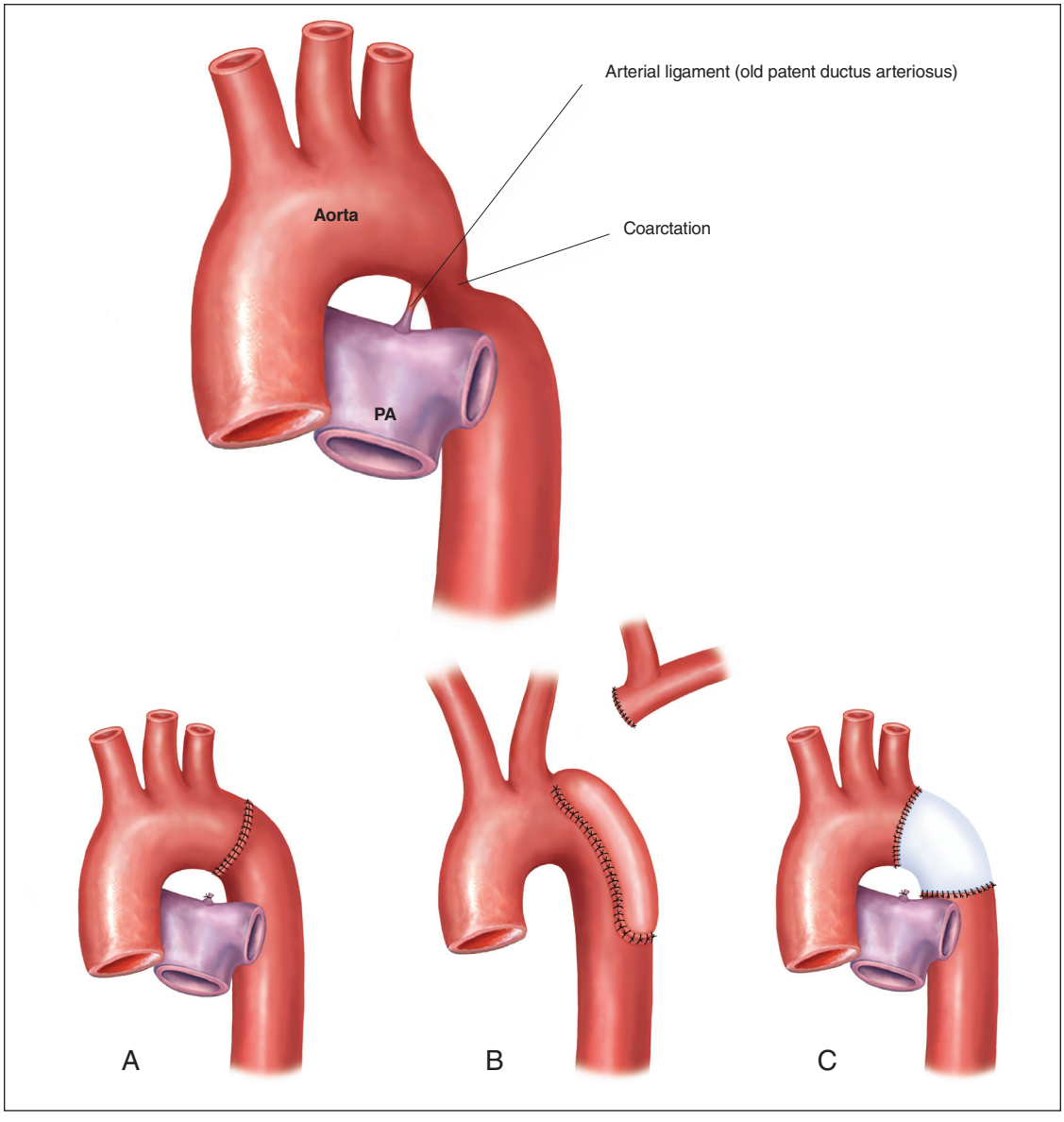
In partial atrioventricular septal defect (AVSD), there is a shunt at the atrial level and separate tricuspid and mitral valves (A). In complete AVSD, there is shunting at both atrial and ventricular levels and one combined five-leaflet atrioventricular valve (B). In a repair of complete AVSD, the atrial and ventricular septal defects are patched, and two valve orifices are created from the combined single valve by suturing the leaflets together (C).

COARCTATION OF THE AORTA

A narrowing of the aorta that obstructs distal blood flow, coarctation of the aorta (see Figure 7) represents about 8.4% of CHD.[15] One study found that 60% of those with coarctation also had a bicuspid aortic valve, 14% a hypoplastic (small) aortic arch, 13% a VSD, and 8% mitral valve abnormalities; a total of 83% of people with coarctation had at least one other cardiovascular abnormality.[37]

Depending on the location and extent of the defect, coarctation has been repaired by a number of surgical procedures over the years, most often end-to-end anastomosis, subclavian flap arterioplasty, and interposition graft. In end-to-end anastomosis

Figure 7. Coarctation of the Aorta and Three Repairs



Constriction in the aortic diameter is most often seen at the site of the former ductus arteriosus (juxtaductal, as illustrated), but it can also appear before the duct (preductal) or after the duct (postductal). In end-to-end anastomosis (A), the coarctation is resected and the proximal and distal aorta are anastomosed. In subclavian flap arterioplasty (B), the left subclavian artery is ligated and transposed to enlarge the aorta. In interposition grafting (C), the coarctation is resected and replaced with a graft. PA = pulmonary artery.

(see Figure 7A), the area of coarctation is resected and the proximal and distal segments of the aorta are sutured together. Subclavian flap arterioplasty (see Figure 7B) disconnects the left subclavian artery from the left-arm circulation and flaps this tissue down to use in enlarging the area of coarctation on the aorta.[38] As a result, a patient with this type of repair will not have a palpable left radial pulse. Therefore, nurses should not use the left arm for blood pressure measurement as it will not reflect central aortic pressure. In interposition graft repair, the coarctation is resected and replaced with a synthetic aortic graft (see Figure 7C).

Long-term problems after coarctation repair include recoarctation (recurrent narrowing) of the aorta. The AHA guidelines for reintervention suggest that pressure gradients (differences in systolic blood pressures between the arms and legs) of more than 20 mmHg should be assessed to determine whether angioplasty or aortic stenting is indicated.[39] Therefore, nurses should obtain four-limb blood pressure measurements to ascertain the pressure gradient, excluding left-arm measurement in cases of a subclavian flap arterioplasty. Recoarctation can also be treated by aortic stenting in the cardiac catheterization laboratory. Aortic stenting requires six months of infective endocarditis prophylaxis.[26]

Hypertension is a common long-term problem of coarctation. Patients with coarctation are often on at least one antihypertensive, and it's important that nurses encourage medication adherence and regular blood pressure monitoring. Although there is a higher incidence of coronary artery disease (CAD) in adults with coarctation than in adults with many other CHDs,[40] these patients' higher risk of CAD may be due to hypertension and other traditional cardiac risk factors rather than to the coarctation itself.[41] More research is needed to understand the precise mechanism.

Coarctation of the aorta is associated with a 10% prevalence of intracranial berry aneurysms,[42] small, round aneurysms on the cerebral arteries that look like berries hanging from the artery and can rupture, causing a hemorrhagic stroke. Many adults with coarctation have never had magnetic resonance imaging or computed tomographic scanning of the brain to screen for aneurysms. Any unusual headaches or changes in consciousness level in patients who have had coarctation require a search for possible intracranial aneurysms.

PULMONIC STENOSIS

Pulmonic stenosis, a narrowing at the pulmonary valve, can also appear immediately above (supravalvar) or below (subvalvar) the valve or as a combination of these. It represents about 14.4% of CHD in adults[15] and often results from fusion of the valve leaflets, giving the valve a dome-shaped appearance (see Figure 8). Here, discussion is limited to valvar pulmonic stenosis.

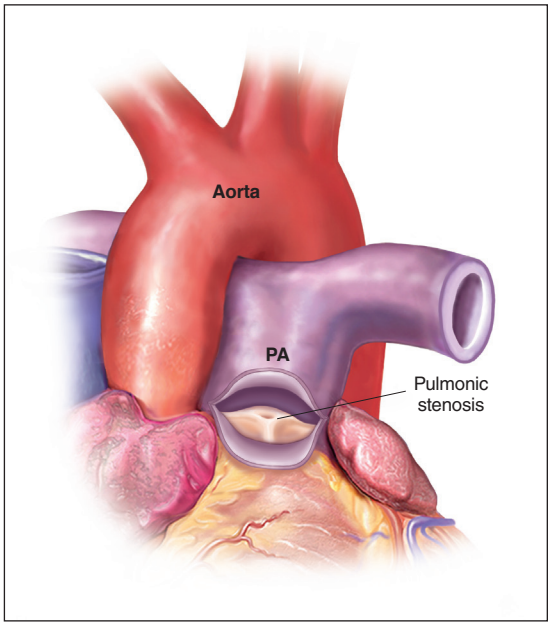
Mild and moderate pulmonic stenosis is well tolerated and may not be diagnosed in childhood. Those with known pulmonic stenosis are monitored with echocardiography. When it becomes severe enough (more than 40 mmHg peak-to-peak gradient[39] or more than 60 mmHg peak, or more than 40 mmHg mean gradient [pressure difference before and after the valve] via echocardiography in an asymptomatic patient, or lower gradients in symptomatic patients), a balloon valvuloplasty is undertaken to split the valve open and to permit better egress of blood from the right ventricle to the pulmonary arteries. Extremely dysplastic (malformed) valves may require surgical valvuloplasty. Patients treated for pulmonic stenosis before the mid-1980s may have had a surgical valvotomy to split the valve leaflets before balloon valvuloplasty was commonly performed.

Coarctation of the aorta is associated with a 10% prevalence of intracranial berry aneurysms.

Cuypers and colleagues summarized the long-term outcomes of patients who had undergone surgical valvotomy or balloon valvuloplasty.[43] Of the six surgical studies, which had up to a mean 33 years of follow-up, reintervention rates ranged from 2.6% to 52.8%, with more interventions needed for pulmonary regurgitation than for recurrent pulmonic stenosis. Most studies found that about 40% of patients had moderate to severe pulmonary regurgitation after surgery, but many may have been conducted at a time when concern about residual stenosis was greater than concern about regurgitation. Time to reintervention ranged from a mean of two to 34 years after surgery. In comparison, the 17 studies on balloon valvuloplasty had a mean follow-up of two to 10 years, with reintervention rates of 2.5% to 30%, the majority for residual pulmonic stenosis. Longer-term survival was 89% to 100% in all studies, regardless of treatment, demonstrating that those with pulmonic stenosis do quite well over time.

Those with residual pulmonary regurgitation after repair require regular echocardiographic and clinical

Figure 8. Pulmonic Stenosis



The pulmonary valve is dome shaped and narrowed, making it difficult for the right ventricle to eject blood to the lungs. PA = pulmonary artery.

monitoring over the years. If regurgitation becomes severe, with right ventricular dilatation or symptoms such as dyspnea, fatigue, chest pain, ankle edema, or ascites, then pulmonary valve replacement (either transcatheter or surgical) may be required (this will be discussed further in Part 2). On auscultation, the nurse will hear a diastolic murmur at the left-upper sternal border if there is pulmonic regurgitation. Ventricular arrhythmias may occur if there is right ventricular dilatation. Therefore, the nurse should look for symptoms such as palpitations, presyncope, or syncope in patients at risk. These patients should be investigated with a Holter monitor or Zio Patch to determine the severity of the arrhythmia.

Tricuspid regurgitation can also occur with right ventricular dilatation. The nurse should hear a systolic murmur at the left-lower sternal border if tricuspid regurgitation is moderate to severe. Significant tricuspid regurgitation can accompany atrial arrhythmia, such as atrial flutter. Again, the nurse would monitor for palpitations and for any other symptoms occurring with palpitations, such as presyncope. As with ventricular tachycardia, investigation with a Holter monitor or Zio Patch is indicated. Catheter ablation, radiofrequency energy applied to the circuit that perpetuates the arrhythmia, can be performed in the cardiac catheterization laboratory. If surgery is undertaken, a maze procedure, requiring cardiopulmonary bypass, can be performed to ablate the atrial flutter or fibrillation.[44] The maze procedure involves creating a single pathway for the atrial impulse to get from the sinoatrial node (the heart's pacemaker) to the atrioventricular node, thus avoiding circus movement of atrial impulses that can initiate and sustain atrial arrhythmias. This pathway is created by incisions or surgical ablation or a combination of the two. Reintervention with balloon valvuloplasty should be considered when the peak pressure gradient across the pulmonary valve is greater than 60 mmHg or the mean gradient is greater than 40 mmHg in an asymptomatic patient, or the pressure gradient is greater than 50 mmHg or the mean gradient is greater than 30 mmHg in a symptomatic patient.[1] The nurse can detect pulmonic stenosis by auscultating a systolic murmur at the left-upper sternal border. Those with moderate to severe pulmonic stenosis should be advised to avoid competitive sports and isometric exercise such as weight lifting until the condition is treated.[1] There are a number of genetic syndromes associated with valvar pulmonic stenosis, and so asking about a family history of pulmonic stenosis is advisable. Assessment by a geneticist is valuable in screening and in optimizing treatment in patients who have one of these genetic syndromes.

Next month, we will review tetralogy of Fallot, transposition of the great arteries, congenitally corrected transposition of the great arteries, and single ventricle defects. ▼

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