This review, the second of two parts, describes the repair of aortic arch anomalies, left-to-right shunts, valvular disease, tetralogy of Fallot, and truncus arteriosus. Cardiac transplantation is also discussed. Advances in the surgical management of congenital heart disease have led to improved patient survival and quality of life. Improvements in technology in computed tomography and magnetic resonance imaging have resulted in increasing utilization of cross-sectional imaging in these patients. Perioperative care necessitates that radiologists have a basic understanding of the surgical treatment and the resultant postoperative anatomy. Because many patients with treated congenital heart disease are being followed up into the 4th and 5th decades of life, this is information that will fall within the domain of all radiologists who interpret cross-sectional images of the thorax.

© RSNA, 2008

From the Division of Pediatric Radiology, Department of Radiology (A.M.G., L.T.D., G.S.B.) and Division of Cardiothoracic Surgery, Department of Surgery (J.J.J.), Duke University Health Systems, 1905 McGovern-Davison Children’s Health Center, Box 3808, Durham, NC 27710. Received November 9, 2006; revision requested January 8, 2007; revision received February 13; accepted March 19; final version accepted July 3; final review by A.M.G. February 1, 2008. Address correspondence to A.M.G. (e-mail: ana.gaca@duke.edu).

© RSNA, 2008
A s surgical techniques and patient survival continue to improve, pa-
tients with repaired congenital heart disease are being followed into
their 4th and 5th decades of life. While the care of these complex patients is
centered on echocardiography and cardiac angiography, improvements in multi-
detector computed tomography (CT) and magnetic resonance (MR) technol-
ogy have resulted in more imaging of these patients falling under the purview
of radiologists. To be part of the care
team of cardiologists and cardiothoracic surgeons seeing patients with congenital heart disease, radiologists must under-
stand the basic anatomy and physiology of these patients before and after surgical
repair.

What follows is the second of a two-
part review of surgery for the treatment of congenital heart disease. While part
1 covered median sternotomy and its
complications, palliative procedures, and complex repairs (1), part 2 will de-
scribe the repair of aortic arch anomalies, left-to-right shunts, valvular dis-
ease, tetralogy of Fallot (TOF), and truncus arteriosus. A brief section on
cardiac transplantation is also included. This is by no means exhaustive; it is,
rather, a brief overview that may serve as the starting point for a basic under-
standing of patients with congenital heart disease.

Aortic Arch Anomalies

Repair of Coarctation of the Aorta

Coarctation of the aorta is a congenital
narrowing, or stenosis, of the aorta. It
typically occurs just distal to the origin
of the left subclavian artery, at the level
at which the ductus arteriosus inserts
into the aorta, but it may occur at any
level of the aorta. Coarctation in the
typical location is thought to result ei-
ther from low flow through the aorta in
utero, often as a result of intracardiac
defects, or from extension of ductal tis-
sue into the wall of the aorta and its
subsequent contraction, producing a
posterior shelf in the lumen of the aorta.
The level of coarctation, degree of ste-
nosis, and presence of associated car-
diac abnormalities affect the develop-
ment of collateral vessels to bypass the
stenotic aorta.

If the coarctation occurs proximal to
the ductus arteriosus, blood flow is pro-
vided to the distal aorta via the patent
ductus. Few early collateral vessels de-
velop, the patient is ductal dependent,
and the condition typically manifests
shortly after birth. If the coarctation is
distal to the ductus arteriosus, which
occurs much more commonly, extensive
collateral vessels must form to provide
distal blood flow. Collateral vessels are
generally not as well formed when the
patient has an associated cardiac lesion.
Coarctation is often associated with other cardiac defects, including
ventricular septal defect (VSD), bicus-
pid aortic valve, and single-ventricle le-
sions such as hypoplastic left heart syn-
drome. Coarctation is also associated
with certain genetic syndromes, partic-
ularly Turner syndrome (2). The radi-
ographic and MR imaging findings of co-
arctation of the aorta have been well
documented (3–8) and will not be ad-
dressed here. Current treatment op-
tions for coarctation of the aorta include
surgical repair, balloon angioplasty, and
stent placement.

Surgical repair of coarctation of the
aorta was initially attempted by Clar-
ence Crafoord in 1944. In this original
surgery, the coarctation was resected,
and the aorta was repaired with a direct
end-to-end anastomosis (9). Coarctation
repair is typically performed via a left
thoracotomy. In some situations, how-
ever, such as when an intracardiac de-
fect is also being repaired or there is
hypoplasia of the transverse aortic arch,
the repair is performed via a median
sternotomy with cardiopulmonary by-
pass.

Surgical repair of coarctation of the
aorta is performed by dividing the
coarctation segment is excised, the two
ends of the aorta would be too far apart
for direct anastomosis. In 1957, Vosschulte (11) introduced the concept of repair with a prosthetic patch that can be used in this situation. In patch aortoplasty, a longitudinal incision is made along the aorta laterally from the origin of the left subclavian artery through the coarctation to the proximal descending aorta. An elliptic patch of prosthetic material is sutured in place, with the widest portion of the patch at the level of the coarctation (Fig 2). Dacron was initially used for repair; however pseudoaneurysms often developed after repair with these patches. Polytetrafluoroethylene (PTFE), less frequently associated with pseudoaneurysm formation, is the current patch of choice. These pseudoaneurysms can be seen up to 2 decades after repair (12–15). Patients may also be at substantial risk of true aneurysm formation if the posterior shelf within the aorta was excised.

An alternative to a prosthetic patch is the subclavian artery patch repair. In 1966, John Waldhausen and colleagues proposed an alternative procedure in which the left subclavian artery is used to augment the aortic lumen (113). This procedure involves ligation of the left subclavian artery at the origin of the vertebral artery. The artery is then incised along its lateral margin, and the incision is extended down the aortic isthmus, through the coarctation, and into the proximal descending aorta. The diaphragm of the coarctation is resected, and the now flattened left subclavian artery flap is folded down into the incision made in the aorta, augmenting the aortic diameter. The left vertebral artery is often ligated to prevent left subclavian steal phenomenon (Fig 3). The primary benefit of this procedure is the ability to repair a long-segment coarctation without prosthetic material. Complications of the Waldhausen procedure include compromised flow to the left upper extremity, often with impaired growth in older children. This procedure rarely results in serious ischemia (16).

Young infants and neonates often have severe aortic arch hypoplasia, as well as coarctation. In these children, the classic resection with end-to-end anastomosis would not completely relieve the obstruction. A technique referred to as the extended end-to-end anastomosis was developed. In this procedure, usually performed through a left thoracotomy, the entire aortic arch and all cephalic vessels are mobilized and the patent ductus arteriosus (PDA) is ligated and divided. A longitudinal incision is then made along the undersurface of the aortic arch to the level of the origin of the left carotid artery, and another longitudinal counterincision is made along the lateral aspect of the descending aorta. When the ends of the aorta are anastomosed, the result is an oblique anastomosis, with a larger diameter of the repaired aorta (Fig 4) (17). Benefits of this extended repair include the complete resection of the stenosis, repair of transverse arch hypoplasia, and ability to avoid the use of prosthetic patch material, which has no growth potential. This method of repair is associated with a lower incidence of recoarctation (residual or recurrent stenosis), compared with the incidence after a simple end-to-end anastomosis, subclavian flap, or aortoplasty with a synthetic patch in patients with aortic arch hypoplasia (17).

Acute complications associated with coarctation repair include residual coarctation, hypertension, recurrent laryngeal and phrenic nerve palsy, and spinal cord ischemia with resultant paraplegia or paraparesis. In the longer term, complications include aortic aneurysm, recurrent coarctation, and hypertension. Despite an appropriate decrease in blood pressure in the immediate postoperative period, the incidence of late hypertension increases dramatically when surgical repair is undertaken after 1 year of age. To balance this finding, however, there is an increased risk of recoarctation when surgical correction is performed before 1 year of age, and the results of some studies suggest increased mortality when repair is undertaken in the 1st year of life, although this risk has diminished in the current era (18–20). Aneurysm formation at the site of repair has been described with most surgical procedures for repair of coarctation, with an incidence of approximately 5%, but is most common with patch aortoplasty (reported incidence, 9%–30%) (14,21).

The imaging of postoperative complications of coarctation repair generally involve echocardiography and chest radiography as screening tools, with MR imaging or CT angiography reserved for more complete evaluation of abnormal findings. Radiographic findings of aneurysm include a soft-tissue mass in the region of the aortic isthmus (14). MR imaging is well suited for evaluation of the postoperative aorta. Its multiplanar capability allows evaluation of the complete aorta, even if tortuous and poorly visualized on the traditional sagittal-oblique view. MR imaging is contraindicated in patients who have a metallic prosthesis, including pacemakers and metallic endovascular stents. Metallic endovascular stents, even if MR compatible, produce a bloom artifact that obscures the areas of interest (22). CT of the aorta provides some of the multiplanar capabilities of MR imaging, with similar resolution. CT is not, however, affected by metallic artifact to the same extent as MR, and, because...
CT is a faster examination than MR imaging, may be better tolerated by younger patients. Therefore, CT is the preferred examination for evaluation of the complete postrepair stent-treated aorta.

CT and MR imaging are comparable for the detection of recoarctation, aneurysm, dissection, or wall irregularity. Hager et al (23), however, have shown that MR and CT are not interchangeable, with differences in measurements between MR and CT images. Some investigators (24) have advocated the use of MR imaging as a screening tool for the complications of coarctation repair, most notably for aneurysm detection.

Balloon angioplasty.—Balloon angioplasty is often used in the treatment of recurrent coarctation, with good results. This procedure is considered safe because of supportive postsurgical scar tissue at the site of dilation. Balloon angioplasty as a treatment option for native (previously untreated) coarctation of the aorta was first introduced in 1982, and its use remains controversial to this day (25). Balloon angioplasty is generally considered a safe alternative to surgery for adolescents and adults with native coarctation and is often the first line of therapy. Studies have, however, shown a higher incidence of recoarctation and aneurysm formation (15, 26–31).

The complications of balloon angioplasty, whether for native coarctation or recoarctation, are similar (28). Excluding arterial access–site injury, most early complications of balloon angioplasty, including aortic intimal tears and flaps and cerebrovascular accidents, are rare, occurring in fewer than 2% of procedures (28,32). Aneurysm formation at the site of dilation is both an early and a late complication in the dilation of coarctation and has been described in patients who had undergone angioplasty more than 5 years previously (29). Early recoarctation has been described in patients undergoing balloon angioplasty of both native coarctation and recoarctation (26–28,32). As with surgical correction, a late complication of balloon angioplasty is the development of hypertension in 30%–40% of patients (26).

Stent placement.—The newest of the treatment modalities for coarctation is stent placement. This procedure has generally been reserved for patients who have recoarctation after previous surgical repair or balloon angioplasty, who have unfavorable anatomy for balloon angioplasty (such as long-segment narrowing), or who are at high risk for surgical repair (33). An added potential benefit to stent implantation is the treatment of any aneurysms present at the time of catheterization (34).

The use of stents is generally avoided in small children owing to the relatively large size of the delivery system and the eventual need for repeat dilation as the child grows. As with surgical repair and balloon angioplasty, some degree of aortic renarrowing has been described with stent implantation (35,36). The most im-

Figure 2:
Illustrations show patch aortoplasty for repair of coarctation of aorta. (a) Incision site for patch repair (dashed line). (b) Incision has been performed prior to patch placement. (c) Patch repair of coarctation.
Important complications of stent placement include acute rupture or extensive dissection of the aorta (37,38). Additional complications include stent fracture, incomplete stent expansion, stent migration, and thromboembolic events. Aneurysm formation has been described in up to 11% of patients as a late complication (35,39). Long-term follow-up is still required to more completely evaluate the outcome of this endovascular treatment for coarctation.

At our institution, the treatment of coarctation tends to follow these general guidelines: (a) Neonates and small children undergo primary surgical repair. (b) Children near adult size can undergo balloon angioplasty. Because the aorta is large in these patients, if a stent is necessary it can be dilated to an adult diameter. (c) Adult coarctation is generally approached on a case-by-case basis. Recoarctation after initial surgical repair is usually treated with angioplasty.

**Interrupted Aortic Arch Repair**

An interrupted aortic arch is defined as complete discontinuity of the aortic lumen between the ascending and descending aorta. Flow to the distal aorta is generally maintained through a PDA (40). The majority of interrupted aortic arches are associated with a VSD, and most have some degree of mild aortic and subaortic stenosis. As a ductal-dependent lesion, patients with interrupted aortic arch are initially treated with prostaglandin therapy to maintain duct patency until surgical repair (41). Because these infants tend to develop cardiac failure in the 1st month of life and 80% of untreated infants die within the 1st month of life, surgical correction is performed at the time of diagnosis (42).

Historically, interrupted aortic arch had been approached as a two-stage repair. The initial procedure was via a left thoracotomy, with repair of the interrupted aortic arch. This could involve the use of a prosthetic graft from the ascending to the descending aorta or a direct anastomosis after mobilization of the aorta to avoid tension on the anastomosis. A pulmonary artery (PA) band was also usually placed to decrease pulmonary blood flow in patients with a VSD. The
patient underwent a later second procedure, generally via a median sternotomy, to remove the PA band, with reconstruction of the PA as needed and repair of the VSD. This approach mandated two separate procedures and committed the patient to further risks of additional arch reconstruction surgery.

In the current era, most neonates with interrupted aortic arch and VSD undergo a single primary repair performed via a median sternotomy with cardiopulmonary bypass. The aortic arch is reconstructed with an anastomosis between the end of the descending aorta and the side of the ascending aorta. This anastomosis may be augmented with an anteriorly placed prosthetic patch. The VSD is closed through the tricuspid valve with prosthetic material (Fig 5) (40,42).

Early postoperative mortality rates range from 10% to 50% (43–47). Immediate surgical complications from interrupted aortic arch repair include bleeding, renal failure, seizures, phrenic nerve injury, chylothorax, and arrhythmias (43). A late complication of repair includes residual or recurrent obstruction at the site of aortic repair in approximately 30% of patients. These patients can generally be treated with balloon angioplasty (43,48–50). Bronchial compression, right or left, is another complication seen after repair of an interrupted aortic arch; it is possibly related to excess tension at the site of anastomosis (47,50).

As with imaging of repaired coarctation of the aorta, both CT angiography and MR angiography are useful for evaluation of a stenosis at the site of prior repair, as well as for detection of aneurysm formation. Persistent atelectasis on chest radiographs following repair should raise suspicion for bronchial compression or phrenic nerve palsy. This could be more completely evaluated with CT angiography and fluoroscopy.

**Left-to-Right Shunt Repairs**

### Total Anomalous Pulmonary Venous Connection Repair

Total anomalous pulmonary venous connection (TAPVC) is a defect in the normal connection of the embryologic pulmonary venous buds to the posterior aspect of the developing left atrium. The result is pulmonary venous return to the right atrium via systemic venous connections. TAPVC is classified according to the site of connection of the anomalous draining vein(s): supracardiac, most commonly to the left brachiocephalic vein from a left vertical vein but also to the right or left superior vena cava or azygos vein; cardiac, drainage to the coronary sinus or right atrium; infracardiac, usually but not always below the diaphragm (inferior vena cava, portal vein, hepatic veins, or ductus venosus); and mixed, to more than one level. If the anomalous connection of the draining vein is obstructed, the patient tends to present as a neonate with pulmonary edema and hypoxemia (2). Some degree of obstruction is always present when the drainage is infracardiac (51). A patent foramen ovale or an atrial septal defect (ASD) is always present in these patients, who are dependent on these lesions for systemic blood supply and survival. TAPVC may be associated with more complex anomalies, including heterotaxy syndromes (2).

With supracardiac TAPVC, the anomalous draining veins tend to form a common venous chamber, or confluence, behind the heart. With supracardiac TAPVC, this common venous confluence drains via the left vertical vein to the left brachiocephalic vein, superior vena cava, or azygos vein. With infracardiac TAPVC, this common venous confluence drains via a descending vertical vein, generally terminating below the diaphragm. Cardiac TAPVC drains via the coronary sinus directly into the right atrium.

The goal of surgical correction of TAPVC is the same regardless of the site of anomalous drainage: to re-establish a normal connection of the pulmonary venous confluence to the left atrium. Anomalous systemic venous connections and intracardiac shunts are generally closed at the same time. Repair may be performed by using an intracardiac approach, in which the pulmonary venous confluence is repaired through the right atrium, or an extracardiac approach, in which the venous confluence is anastomosed directly to the left atrium. When the anomalous drainage is cardiac (via the coronary sinus to the right atrium), the surgical technique generally involves an incision in the common wall shared by the coronary sinus and the left atrium, allowing the coronary si-
nus to drain into the left side of the heart. The entire defect is then closed with a pericardial patch much like an ASD would be repaired (51,52). For the repair of supracardiac TAPVC, an incision is made along the posterior wall of the left atrium and along the anterior wall of the common venous chamber, allowing free drainage to the left side of the heart (Figs 6, 7) (51,53).

Early complications of TAPVC repair are usually related to increased pulmonary vascular resistance and pulmonary hypertension. The major postoperative complication of repair of TAPVC is the development of late postrepair pulmonary venous stenosis, which occurs in up to 10% of repairs. Surgical correction of this complication is associated with 30%–45% early mortalities (52–54).

A relatively new procedure, the “sutureless” technique, was originally designed for correction of this postrepair pulmonary venous stenosis but has subsequently been used for primary TAPVC, with preliminary results suggesting decreases in mortality and risk of repeat surgery (55,56). A transverse incision is made in the posterior wall of the left atrium, extending to the interatrial septum. The atrial–common venous chamber anastomosis created at the initial repair is completely excised, along with all stenotic venous tissue. The edges of the atrial incision are then sutured to the pericardium rather than to the walls of the pulmonary veins. This procedure essentially produces a controlled bleed from the pulmonary veins into a new chamber bounded by pericardium, with the blood then passing into the left atrium, eliminating the need for direct suturing of the pulmonary veins. If performed for recurrent pulmonary vein stenosis, pericardial adhesions present after surgery help maintain the integrity of this space (57–59).

The incidence of early mortality associated with primary repair of TAPVC has decreased to approximately 15% (52–54). The presence of obstructed TAPVC is associated with higher perioperative mortality. Diffuse pulmonary lymphangiectasia is also present in some infants with TAPVC and is also associated with higher mortality (53,60–64).

**ASD Repair**

ASDs are generally categorized according to their location, which is related to the complex embryology of the right atrium. This embryology is covered in detail in numerous excellent resources (2,65,66) and will not be discussed here, with the exception of one feature. The primum ASD is located low in the septal wall, and while it can be seen in isolation, it is frequently associated with abnormalities of the atrioventricular valves and will be discussed in the section dealing with the atrioventricular septal defects (AVSDs). The secundum
ASD, located more superiorly in the septal wall, can range from a tiny perforation to complete absence of the septum primum and is the most common type of ASD. Two less common types of ASD include the sinus venosus ASD and the coronary sinus ASD. The sinus venosus ASD, commonly located just inferior to the junction of the superior vena cava and the right atrium, is associated with anomalous pulmonary venous drainage in 80%–90% of cases, usually involving the right lung (2,66).

Patients with an ASD are generally asymptomatic and are usually diagnosed incidentally because of a murmur found at routine physical examination. Occasionally, children will present in infancy with a large ASD, with signs of right ventricular volume overload and congestive heart failure. Mild cyanosis may be present in some children with a large defect and increased pulmonary vascular resistance or when associated with other lesions such as an Epstein anomaly or pulmonary stenosis. Some secundum ASDs will close spontaneously. If the ASD has not closed in the 1st year of life, however, there is a 90% chance it will never close. Sinus venosus ASDs do not close spontaneously. Repair is usually undertaken electively between 2 and 4 years of age. In the presence of important symptoms, ASD repair may be performed in infancy (66).

Secundum ASD.—Surgical repair of a secundum ASD is the standard method of treatment. This requires open cardiac repair with cardiopulmonary bypass. Many different incisions and approaches have been used, usually to minimize trauma and improve cosmesis, including partial lower sternotomy, inframammary anterior thoracotomy and bilateral inframammary clamshell incisions. If the ASD is small and there is adequate septal tissue, the defect can be sutured closed. If the ASD is too large for primary repair, a pericardial or prosthetic patch can be used for closure. Occlusion of a small secundum ASD can be performed percutaneously with a clamshell-shaped prosthetic device delivered across the ASD through the femoral vein. These techniques eliminate the need for incisions and open cardiac repair, but the rate of complete closure is slightly less than that with surgery, and the long-term outcome of these devices within the heart is currently unknown. The position of the device is confirmed with angiography, echocardiography, and chest radiography (67,68).

Sinus venosus ASD.—Imaging with cardiac MR imaging, CT angiography, or conventional angiography may be needed prior to surgical repair to exclude the presence of anomalous pulmonary venous drainage of the right pulmonary veins to the superior or inferior vena cava (scimitar syndrome).

Surgical repair is indicated in all patients with sinus venosus ASD. These defects are repaired with open cardiac surgery via a median sternotomy. If there are no anomalous draining veins, the ASD can be closed with a pericardial patch via the right atrium. If there is insertion of an anomalous pulmonary vein high on the superior vena cava, this can be repaired with either (a) a longitudinal incision on the lateral aspect of the right atrium and placement of a bafflemike patch in the floor of the superior vena cava–right atrium junction (b) with the Warder procedure, in which the superior vena cava is divided just superior to the most cephalic anomalous vein. A pericardial patch is placed, creating a baffle in the right atrium that directs blood from the anomalous veins and proximal superior vena cava via the baffle through the ASD into the left atrium. Then the distal-most part of the superior vena cava (with systemic venous return) is rerouted to the right atrium via the right atrial appendage (66,68).

Coronary sinus ASD.—Open surgical repair of a coronary sinus ASD is similar to the repair of a secundum ASD and is usually performed by placing a pericardial patch over the orifice of the coronary sinus. A persistent left superior vena cava draining into the coronary sinus must be excluded prior to this type of repair because an obligatory right-to-left shunt from the left superior vena cava to the right atrium will result if it is not excluded (66).

Complications following ASD repair are typical of those for any open heart procedure, with a slightly greater risk of pericardial effusion and postpericardiotomy syndrome (idiopathic febrile illness with pericardial and pleural inflammation after open cardiac surgery). Complications of percutaneous closure include device embolization (up to weeks after placement), thrombus formation on the device, and arrhythmias. While some studies (67,68,70–72) have found higher complication rates in surgical patients, these were minor and did not affect patient care.

VSD Repair

As with the atrial septum, the embryologic formation of the ventricular septum is a complex process involving the fusion of multiple cardiac components. This discussion is beyond the scope of this review. The location of a VSD depends on failure of one of these components to form or fuse appropriately, resulting in communication between the ventricles (65,66). Numerous classification systems have been devised for VSDs according to various criteria and the system described by the Congenital Heart Surgery Nomenclature and Database Project in 2000 (73).

Small VSDs may not result in clinical symptoms. These patients are closely watched, as some small lesions will close with time. The exception to this is the supracristal-type VSD. These defects are typically large and, if left unrepaired, may result in aortic insufficiency. Owing to left-to-right shunting at the ventricular level, larger VSDs cause increased pulmonary blood flow and symptoms of congestive heart failure. Because the degree of shunting is dependent on downstream vascular resistance, most infants will not become symptomatic until pulmonary vascular resistance falls.

Surgical closure is indicated when medical management of congestive heart failure fails. For large unrestricted VSDs, this typically occurs in the first 3–6 months of life. If left untreated, a large VSD with unrestricted pulmonary blood flow will ultimately result in pulmonary vascular disease. After the development of Eisenmenger syndrome,
Repair of Congenital Heart Disease

Gaca et al

Volume 248: Number 1—July 2008

Closing the VSD is contraindicated, because right ventricular failure will ensue. Heart-lung transplantation or lung transplantation with VSD repair may ultimately be needed in these complex patients (51).

Surgical repair of a VSD is performed through a median sternotomy and usually through a right atrial incision, but it can be accomplished through an incision in the ventricle or PA, depending on the location of the defect. Repair is always performed with a patch, with special attention paid to the course of the conduction tissue in the heart. Complications from surgery include injury to conduction tissue and heart block, residual ventricular defects, and tricuspid valve insufficiency (74). The presence of multiple VSDs is associated with increased risk for early mortality, usually related to difficulty in adequate exposure and incomplete closure (66,75).

AVSD Repair

AVSD, also known as atrioventricular canal defect and endocardial cushion defect, encompasses a spectrum of anomalies that all arise from a common embryologic pathway. The inferior atrial septum, the superior ventricular septum, the septal leaflet of the tricuspid valve, and the anterior leaflet of the mitral valve all develop from the endocardial cushion tissue. AVSDs are typically divided into three categories: complete, partial, and transitional. A complete AVSD consists of a large VSD immediately below the plane of the atrioventricular valves, a large ASD immediately above the plane of the atrioventricular valves, and a common atrioventricular valve orifice. A partial AVSD is missing a component of the complete AVSD, usually the VSD. This is also referred to as a primum ASD. A transitional or intermediate AVSD is similar to a complete AVSD, but the leaflets of the common atrioventricular valve are fixed to the ventricular septum, resulting in a small VSD component (66). AVSD is the most common cardiac anomaly seen in patients with Down syndrome (one-half of cardiac defects) (76).

Patients with AVSD have left-to-right shunting, which causes increased pulmonary blood flow. Over a long period of time, this results in increased pulmonary vascular resistance and irreversible pulmonary hypertension (Eisenmenger syndrome). Surgical correction is indicated if medical management fails or when the child reaches 4–6 months of age. The best age for repair is dependent on surgeon preference but is generally younger than 6 months of age. For children with a partial AVSD in which the majority of shunting is across an ASD, surgery can be deferred until the toddler years, since there is less risk of pulmonary vascular disease with supraventricular valve shunts (66). PA banding may be used as palliation in patients with a complex AVSD, to control pulmonary blood flow until complete correction is performed.

With a partial AVSD (septum primum ASD and cleft mitral valve), repair is performed through a right atrial incision. The cleft between the superior and inferior portion of the left-sided atrioventricular valve is sutured together to create a bi-leaflet (mitral) valve, and the ASD is repaired with a patch (usually pericardium). The atrioventricular node and the bundle of His are at risk of injury during this repair. If a small VSD is present, as in a transitional AVSD, simple suture repair of the VSD is usually all that is needed, and the remainder of the defect is repaired as for a partial AVSD (66).

Several techniques are used for the repair of a complete AVSD. Most commonly, a two-patch technique is used. In this procedure, the VSD is usually repaired with prosthetic material and the atrioventricular valve tissue is sandwiched between the ASD patch, usually pericardium, and the VSD patch. While there is some debate as to whether to close the cleft in the left atrioventricular valve, most surgeons prefer to do this in an effort to prevent late left atrioventricular valve failure (66).

Mortality associated with repair of a complete AVSD by means of all techniques has decreased to less than 5%. Repeat operation is most commonly performed for mitral valve regurgitation, which is seen in up to 10% of patients. Complete heart block is also possible in nearly 5% of patients (77–81).

PDA Repair

In the normal-term infant, the ductus arteriosus generally closes within the first 24 hours after birth. A PDA represents the failure to transition from fetal to postnatal circulation and is seen in fewer than 1% of term infants but in nearly half of preterm infants weighing less than 1500 g (66,82). In 1939, Robert E. Gross and John P. Hubbard described the first successful surgical ligation of a PDA (83).

In patients with isolated PDA, several options are now available for the treatment: prostaglandin synthetase inhibitors, percutaneous catheter occlusion, video-assisted thoracoscopic surgery (or VATS) ligation, or open surgical ligation (via thoracotomy or, rarely, sternotomy) (84). The option chosen depends on the age of the patient, the presence of other cardiac lesions requiring repair, and the presence of symptoms.

In preterm neonates, a PDA is initially managed medically with fluid restriction, diuretics, and prostaglandin synthetase inhibitors (indomethacin). Surgical ligation is indicated if medical management fails in symptomatic patients (85). Earlier gestational age and lower birth weight are factors found to increase risk of failure of nonsurgical management (86). When surgery is necessary, these infants can be treated with PDA ligation by means of left lateral thoracotomy or video-assisted thoracoscopic surgery (84). Repair is accomplished with either simple ligation by means of a metal clip or ligature or with ligation and division of the ductus.

Larger infants, children, and adults can be treated with either percutaneous occlusion or surgical ligation (84). Percutaneous transcatheter occlusion can be performed with access via the femoral vein or artery, and the PDA is occluded by using either coils or an occlusion device or plug. Complications from percutaneous occlusion include persistent flow across the PDA (27% at 6 weeks after the procedure; 10%–20% at 6 months), which can be associated
with hemolysis (87). If the occluding device extends too far into the lumen of the PA or aorta, it can cause flow disturbance and narrowing. Because of the size of the introducer sheaths, percutaneous occlusion generally cannot be performed in infants weighing less than 5 kg (87). Percutaneous occlusion may also be limited in patients with a short wide PDA because the coils cannot be securely seated (88). The coils or occluding device have also been known to migrate or embolize, usually into the pulmonary circulation (84,87). As with other procedures involving percutaneous access, there is a risk of injury to the femoral artery or vein.

For PDA ligation by means of video-assisted thoracoscopic surgery, several small thoracostomies are created to allow placement of the scope, the retractors, and the device for applying the clip. When compared with the muscle division and rib spreading of a traditional thoracotomy, the minimally invasive video-assisted thoracoscopic surgery approach is believed to minimize the risk of postthoracotomy pain syndromes, scoliosis, rib deformity, and shoulder muscle dysfunction. Complications from either video-assisted thoracoscopic surgery or traditional surgical ligation include phrenic or recurrent laryngeal nerve injury and incomplete occlusion of the ductus (84,87).

Valvular Disease

Aortic Valve

Valvular aortic stenosis is one form of left ventricular outflow tract obstruction. Children with aortic valve stenosis often have a unicuspid (fusion of two or more commissures) or bicuspid (fusion of one commissure) aortic valve. Aortic valve stenosis may be found in association with other cardiac defects such as coarctation of the aorta, interrupted aortic arch, Shone complex, and mitral stenosis and is often asymptomatic. A minority of patients will have aortic stenosis in the first few weeks of life. These patients usually have a unicuspid aortic valve and severe or critical obstruction. In some neonates, aortic stenosis is so severe that a PDA is necessary to ensure adequate systemic perfusion. In these children, either surgical valvotomy or balloon valvotomy is necessary to allow forward flow from the left ventricle and to allow PDA closure. Children with a bicuspid aortic valve are usually asymptomatic, and may remain so well into adulthood. As these patients age, however, the valve leaflets may thicken and the valve may become stenotic, insufficient, or both.

The aortic valve may also be primarily insufficient. Children with primary insufficiency may have a bicuspid or tricuspid valve and some have connective tissue abnormalities such as Marfan syndrome or Ehlers-Danlos syndrome.

Aortic valve repair may be possible in a minority of patients, but valve replacement is usually required. In children, options for valve replacement are limited by small patient size and the desire to avoid use of anticoagulants. There are currently no suitable mechanical or bioprosthetic valves for infants and children. Preferred aortic valve prostheses for children are the aortic valved allograft or, more commonly, the pulmonary valve (Ross procedure). In the Ross procedure, the normal pulmonary valve is moved to the aortic position, and the pulmonary valve is replaced with a pulmonary allograft. This allows growth of the newly created aortic valve and eliminates the need for anticoagulation (66,89).

Mitral Valve

Mitral valve stenosis can occur in isolation but is more commonly associated with other left heart defects. One common association is Shone complex, an association of multiple left heart obstructive lesions, including mitral stenosis, supra–mitral valve ring, hypoplastic left ventricle, aortic valve stenosis, and coarctation of the aorta. Congenital mitral valve stenosis is often associated with an abnormal sub–mitral valve apparatus, including parachute mitral valve, mitral valve arcade, and single papillary muscle. Surgical repair of congenital mitral stenosis typically involves division or elongation and separation of the chordae and commissurotomy. In the case of a supra–mitral valve ring, simple resection of this abnormal fibrotic tissue may relieve the obstruction. In some situations, the fibrotic ring may involve the mitral leaflets, necessitating repair or replacement of the mitral valve. When mitral stenosis is associated with other left heart obstruction that precludes biventricular repair, single-ventricle palliation may be necessary. While results of balloon dilation of rheumatic mitral valves are encouraging, dilation of congenital mitral stenosis is less often successful (66,90).

Mitrail valve regurgitation is much more common than mitral valve stenosis and has many causes, including defects in the valve leaflets (e.g., cleft or dysplastic leaflets), abnormal chordae or papillary muscles, or a dilated valve annulus. Medical therapy, consisting of afterload reduction and anticongestive therapy, should be undertaken initially. Surgical therapy is indicated if medical therapy fails and is directed at the mitral valve abnormalities. This usually involves closure of clefts in the anterior leaflet of the mitral valve or simple commissuroplasty techniques. If annuloplasty is necessary, use of prosthetic material is avoided in children who are expected to grow. Valve replacement is reserved for those in whom other treatment has failed or who are not suitable candidates for valve repair (66,90,91).

TOF Repair

While the classic lesions of TOF include pulmonary stenosis, VSD, overriding aorta, and right ventricular hypertrophy, the essence of the lesion is a VSD and right ventricular outflow tract (RVOT) obstruction that limits pulmonary blood flow. When RVOT obstruction is severe, there is increased right-to-left shunting, resulting in increased cyanosis (blue tetralogy). When the obstruction is less severe, the shunting is predominantly left-to-right (pink tetralogy). Some patients have complete obstruction of the RVOT and their condition is referred to as TOF with pulmonary atresia. This defect represents a spectrum from simple atresia of the pulmonary valve with intact main and
branch PAs to absence of any normal PAs, with blood flow to the lungs coming from aortopulmonary collateral vessels. In these neonates with pulmonary atresia, pulmonary blood flow is dependent on the PDA. Prostaglandin therapy is mandated shortly after birth to maintain patency of the PDA. Patients with substantial collateral vessels can be relatively free of symptoms. These patients may develop signs of congestive heart failure with increasing collateral flow or may, over time, develop pulmonary hypertension from pulmonary blood supply at arterial pressures.

The first repair of TOF was performed more than 50 years ago by C. Walton Lillehei at the University of Minnesota (92). Since then, surgeons have achieved excellent outcomes for the past 25 years. Originally, pulmonary blood flow was augmented with a modified Blalock-Taussig shunt, and definitive repair was undertaken when the child was older. More recently, primary complete repair at 3–6 months of age is favored by most programs, with great success. Primary definitive repair prevents multiple scars and anesthesias and potential distortion of the PAs by the shunt. Both strategies, however, are acceptable (93).

**TOF with pulmonary stenosis.**—The goals for definitive TOF repair consist of closing the VSD and relieving the RVOT obstruction (94,95). Early techniques for repair of TOF included transannular repair. This involved closing the VSD through a large right ventricular incision and, usually, pulmonary valvectomy (Fig 8) (92). With this repair, the patient is left with a large section of dyskinetic right ventricle and with free pulmonary valve insufficiency. This has resulted in a large number of patients requiring subsequent pulmonary valve replacement (94,96).

Currently, a premium is placed on efforts to spare the pulmonary valve and avoid a large ventriculotomy. Often, the VSD can be closed via the right atrium and the RVOT obstruction, by means of a simple pulmonary valvotomy with limited or no transannular incision. This is referred to as a transatrial repair (Fig 8) (96,97). An anomalous left anterior descending coronary artery arising from the right coronary artery may cross the infundibulum of the right ventricle. A right ventricular incision should be not be performed in these patients, to prevent injury to the coronary artery. In this scenario, a right ventricle-to-PA conduit may be required to relieve RVOT obstruction (98–101).

Although TOF repair restores the normal two-ventricle physiology, the patient may have pulmonary insufficiency or residual pulmonary stenosis and a hypertrophied right ventricle. This will eventually result in severe right ventricular dysfunction and heart failure in the majority of patients. In these cases, placement of a pulmonary valve will relieve heart failure symptoms and improve right ventricular hemodynamics (97).

**TOF with pulmonary atresia.**—TOF with pulmonary atresia (which some classify together with pulmonary atresia with VSD) can range from simple atre-
sia of the valve to complete absence of the pulmonary arteries, with pulmonary blood flow via aortopulmonary collateral vessels. These collaterals provide a spectrum of blood supply to the lungs that ranges from direct connections to the PAs, to supply at the arteriolar or capillary level, to the formation of a discrete and separate blood supply to the lungs without communication with the PAs. Those infants with simple valve atresia have few collateral vessels and are ductal dependent; they present with severe cyanosis when the PDA closes. Patients with substantial collateral vessels can be relatively free of symptoms. These patients may develop signs of congestive heart failure, with increasing collateral flow, or over time may develop pulmonary hypertension from pulmonary blood supply at arterial pressures. The goal of surgical repair of TOF with pulmonary atresia is to reestablish continuity between the right heart and the PAs, which is essential for PA and alveolar development (66).

In patients with TOF and pulmonary valvular atresia, surgical repair is undertaken in the neonatal period. The VSD is closed via a right ventriculotomy. By using the same ventriculotomy, a right ventricle–to-PA conduit is placed, traditionally by using a valved aortic or pulmonary homograft (66). These patients will eventually outgrow their conduits and require repeat surgery (95).

For patients with TOF with hypoplastic PAs and prominent collateral vessels, surgical repair is usually staged. Many surgical approaches and strategies have been proposed, but the underlying goal is to restore antegrade right ventricle–to-PA blood flow and eventually close the intracardiac shunts. This may be accomplished with a modified Blalock-Taussig shunt or with unifocalization of the PAs, the surgical procedure in which aortopulmonary collateral vessels are separated from the aorta at their origin and are anastomosed to the reconstructed PA. Depending on the size of the reconstructed PAs, the VSD may or may not be closed at the initial operation. These patients require follow-up, since the collateral vessels and PAs tend to become stenotic and may require multiple repeated operations to replace the right ventricle–to-PA conduit and relieve pulmonary artery stenosis (99).

In addition to cardiac catheterization, noninvasive imaging is necessary in most patients with TOF and pulmonary atresia with VSD. With the development of faster techniques, both MR imaging and CT angiography can be used for the evaluation of the anatomic structures and to evaluate many aspects of these patients, including PA anatomy, additional sources of pulmonary blood flow, pulmonary valve and tricuspid valve function, and both right and left ventricular function and viability. In some situations, MR imaging may be useful to help determine differential blood flow to the lungs.

### Truncus Arteriosus Repair

Truncus arteriosus is a cardiac defect resulting from failure of septation of the conotruncus into a separate aorta and PA. The result is a single vessel arising from the heart, which supplies the systemic, pulmonary, and coronary blood flow, with at least one PA arising from this truncal valve. There is a single truncal valve, which may have a variable number of leaflets that may be stenotic or regurgitant. In more than 80% of the patients, a large VSD is also present (66).

Neonates with truncus arteriosus are generally asymptomatic while PA pressures remain elevated. Because both the pulmonary and the systemic blood flow arise from a single ventricular outlet, however, patients tend to develop excessive pulmonary blood flow and congestive heart failure as PA pressures decrease in the 1st weeks of life. The increased pulmonary blood flow can cause pulmonary hypertension if not addressed in the first 3–6 months of life. Patients may occasionally develop retrograde flow in the abdominal aorta during diastole, causing steal phenomenon from the abdominal visceras (66).

Truncus arteriosus was initially palliated with PA banding to limit pulmonary blood flow and prevent pulmonary hypertension. In 1968, McGoon et al (102) described the first surgical repair of truncus arteriosus. Via a median sternotomy, the PAs were separated from the aorta, and a valved homograft was placed from the right ventricle to the PAs. The VSD was closed at the same time (Fig 9). This technique, which is very similar to the Rastelli procedure, remains the predominant method of repair of truncus arteriosus, with typical use of a valved aortic or pulmonary homograft. Patients with abnormalities of the truncal valve may require repair or replacement of the valve. Children with successful repair of truncus arteriosus will require repeat operation to replace the right ventricle–to-PA conduit (103,104).

### Cardiac Transplantation

Cardiac transplantation is indicated for a broad range of clinical problems in children. Primary indications include irreparable congenital heart disease, failed repairs, and cardiomyopathy (105). The first successful infant heart transplant was performed by Leonard Bailey in 1985. After this, cardiac transplantation became a common occurrence in patients with hypoplastic left heart syndrome. However, as hypoplastic left heart syndrome surgery (Norwood procedure) has become more successful, cardiac transplantation in infants is now generally reserved for those patients who have more complex defects or in whom traditional repairs have failed. Cardiac transplantation is often performed in older children because of cardiomyopathy. Other indications include intractable arrhythmias and some cardiac tumors (eg, rhabdomyoma).

Current transplantation technique involves resection of the recipient heart, with a cuff of left atrium left behind at the insertion of the pulmonary veins. The great vessels are divided just above their respective valves. The right atrium can be partially resected, leaving behind the posterior wall with the superior and inferior venae cavae, or it can be resected entirely, dividing the superior and inferior vena cavae before their entry into the heart.
The implantation process is simply a series of anastomoses (Fig 10).

Infection is a notable cause of morbidity and mortality in pediatric patients after transplantation. In the 1st month following transplantation, bacterial and fungal infection are most common, resulting in pulmonary and sternal wound infections (106). Transplantation patients are particularly vulnerable to viral infections in the 6 months after surgery, particularly cytomegalovirus and Epstein-Barr virus. Both are associated with increased risk of organ rejection. Epstein-Barr virus infection increases the risk of posttransplantation lymphoproliferative disorder (PTLD).

PTLD includes a spectrum of disorders ranging from mononucleosis-type illness to B-cell lymphoma, which are associated with Epstein-Barr virus infection after solid organ transplant. Because children are less likely than adults to have been exposed to Epstein-Barr virus prior to transplantation, they are at higher risk of primary Epstein-Barr virus infection after transplantation. Patients are particularly at risk for developing PTLD when the Epstein-Barr virus infection occurs within the first few months after transplantation. This primary infection could be the result of transmission from a seropositive donor to a seronegative recipient (107,108). PTLD after cardiac transplantation rarely involves the heart, while it commonly involves the lungs in patients receiving a lung or heart-lung transplant. PTLD also commonly involves the gastrointestinal tract but can also cause cutaneous nodules and central nervous system disease (105, 108,109). Chest radiographs may suggest the diagnosis, but CT provides the most comprehensive information about PTLD. Pulmonary findings may include nodules, airspace disease, pleural and pericardial effusions, and mediastinal adenopathy. Abdominal findings include adenopathy, as well as involvement of the liver, kidneys, and bowel (107–109).

Rejection is typically diagnosed by means of surveillance endomyocardial biopsies. Noninvasive techniques, such as radionuclide scanning and echocardiography, may be supportive but are not definitive. Chronic rejection is associated with the development of graft coronary artery disease, which is the leading cause of death among late survivors of cardiac transplantation. Graft coronary artery disease differs from typical atherosclerotic coronary artery disease in that it involves long segments of the coronary
artery and is more difficult to treat with traditional angioplasty or bypass techniques (51,105). In addition, because the heart is denervated and patients may be completely asymptomatic, routine coronary angiography is mandatory (105,110). Evaluation of the coronary arteries with CT angiography has been promising in early investigations (111,112).

Conclusion

As survival of patients with congenital heart disease has improved, we have begun to see patients who are 30–40 years post repair. As these patients age, there has been increasing demand to evaluate the results (including complications) of these surgical procedures. While much of this evaluation has been focused on echocardiography, the evolution of CT and MR imaging has provided new diagnostic tools. Radiologists who interpret cross-sectional images of the thorax should have a basic understanding of the surgical treatment and resultant postoperative anatomy of these complex patients. This understanding will result in more precise image acquisition and meaningful accurate interpretation.

References

23. Mendelson AM, Lloyd TR, Crowley DC, Sandhu SK, Kocsik KC, Beeckman RH 3rd. Late follow-up of balloon angioplasty in chil-


67. Mashwan WE, King SB, Jacobs WC,


104. Behrendt DM, Dick M 3rd. Truncus re-


