Cardiovascular MR Imaging of Conotruncal Anomalies

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Conotruncal anomalies are congenital heart defects that result from abnormal formation and septation of the outflow tracts of the heart and great vessels. The major conotruncal anomalies include tetralogy of Fallot, transposition of the great arteries, double-outlet right ventricle, truncus arteriosus, and interrupted aortic arch. Cardiovascular magnetic resonance (MR) imaging is an important modality for the evaluation of patients with these defects. Major advances in cardiovascular MR imaging equipment and techniques allow precise delineation of the cardiovascular anatomy and accurate quantitative assessment of ventricular function and blood flow. The data provided by cardiovascular MR imaging are useful for treatment planning and posttreatment monitoring, supplement information obtained with echocardiography, and in many cases obviate cardiac catheterization. Supplemental material available at http://radiographics.rsna.org/lookup/suppl/doi:10.1148/rg.304095158/-/DC1.

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Abbreviations: DORV = double-outlet right ventricle, LVOT = left ventricular outflow tract, RVOT = right ventricular outflow tract, SSFP = steady-state free precession, 3D = three-dimensional, TOF = tetralogy of Fallot, 2D = two-dimensional

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Introduction
Conotruncal anomalies are a group of congenital heart defects that involve the outflow tracts of the heart and the great vessels. The outflow tract of the embryonic univentricular heart, also known as the conotruncus, begins as a common outlet but undergoes a complex, highly choreographed sequence of events that results in the creation of separate left and right ventricular outflow tracts (LVOT and RVOT, respectively) as well as the aorta and main pulmonary artery (1). In utero developmental abnormalities of the embryonic conus arteriosus and truncus arteriosus may affect the ventricular outflow tracts, ascending thoracic aorta, aortic arch, and pulmonary arteries. The major conotruncal anomalies are tetralogy of Fallot (TOF), transposition of the great arteries, double-outlet right ventricle (DORV), truncus arteriosus, and interrupted aortic arch.

Cardiovascular magnetic resonance (MR) imaging has emerged over the past decade as an alternative, complementary, and frequently superior imaging modality for the evaluation of patients with congenital heart defects, including conotruncal anomalies. In particular, cardiovascular MR imaging is the technique of choice for postoperative evaluations. Major advances in MR imaging hardware and software—including improvements in coil design, magnetic gradients, pulse sequences, and image reconstruction—allow rapid, high-resolution imaging of complex anatomic structures and accurate assessment of physiologic function. Cardiovascular MR imaging overcomes many of the limitations of echocardiography (eg, restricted acoustic windows), computed tomography (eg, exposure to ionizing radiation, lack of functional information), and cardiac catheterization (eg, ionizing radiation exposure, morbidity, and high cost). Transthoracic echocardiography often suffices for initial diagnosis and preoperative planning in infants; however, cardiovascular MR imaging is useful in selected cases to evaluate regions of the extracardiac vascular anatomy that are not readily visualized at echocardiography.

The article describes and illustrates the applications of cardiovascular MR imaging for evaluating major conotruncal anomalies, including TOF, dextro- (D-) and levo- (L-) transposition of the great arteries, DORV, truncus arteriosus, and interrupted aortic arch. The anatomy and surgical repair of these lesions are discussed, and cardiovascular MR imaging techniques, protocols, and pre- and postoperative appearances are described.

Overview of Cardiovascular MR Imaging Techniques
A wide array of MR imaging sequences and techniques are available for detailed assessments of the cardiovascular anatomy and function in patients with known or suspected congenital heart defects, including conotruncal anomalies (2,3). The cardiovascular MR imaging examination begins with a review of available clinical and surgical history and imaging data for each patient. The goal is to tailor the examination to answer the specific questions posed by the referring cardiologist and perform the examination as efficiently as possible so as to keep the imaging time at a minimum. Most patients younger than 8 years who are referred for cardiac MR imaging require either general anesthesia with endotracheal intubation for breath-hold acquisitions or intravenous deep sedation and modification of the MR imaging sequences for free-breathing acquisitions. Dedicated phased-array multiple-element cardiac coils enabling parallel imaging are typically used for MR imaging in older children and adults. Multiple-element knee and head coils may be used with good results in infants and younger children.

The first sequence usually employed in the MR imaging protocol is a non-cardiac-gated steady-state free precession (SSFp) localization sequence applied in the coronal, axial, and sagittal planes to cover the chest and upper abdomen. These localization sequences provide an overview of the visceral and cardiac situs and cardiovascular anatomy. They are the starting point on which the rest of the examination is based; they are used to generate the variable imaging planes needed for the type of cardiac disease to be evaluated. More precise morphologic and functional information then is acquired with various high-spatial-resolution sequences, which are described below. Many MR imaging systems are now equipped with real-time interactive gradient-echo sequences that can be applied without patient breath holding to quickly construct all of the imaging planes that will be needed for a given examination protocol. The imaging geometry can be labeled and stored for quick access later in the
examination, before the acquisition of higher-spatial-resolution breath-hold cine SSFP images of the heart for the evaluation of cardiovascular anatomy and function.

Cine imaging of cardiac function and blood flow is a primary component of the cardiovascular MR imaging protocol for evaluation of patients in whom the presence of a congenital heart defect is suspected. Assessments of global and regional wall motion, as well as accurate quantification of ventricular mass, stroke volume, chamber volumes, and ejection fraction, are reliant on cine MR imaging. The current standard cine MR imaging sequence is an electrocardiographically gated two-dimensional (2D) SSFP sequence. This sequence provides the advantages of good contrast between the blood pool and myocardium and relatively short acquisition times. To avoid respiratory motion, patient breath holding is usually necessary; however, MR data acquisition times can be shortened with the use of parallel imaging techniques. Signal averaging can be performed to compensate for respiratory motion during free-breathing acquisitions. A series of short-axis 2D SSFP images is obtained from the cardiac apex to the base to allow reliable quantitative estimations of ventricular function and chamber volume. Sequential monitoring of ventricular function and dimensions is important for follow-up of previously diagnosed congenital heart defects. Cardiovascular MR imaging is recognized as the reference standard for the assessment of left and right ventricular volumes and mass because it has been shown to be accurate, highly reproducible, and free of geometric assumptions (4). This is especially true for assessments of the right ventricle, the chamber that is most often involved in cardiac dysfunction due to congenital heart defects and the one that is most often affected by surgical repair (5).

Multisection 2D SSFP “bright-blood” imaging provides important dynamic information about the anatomy of the heart and great vessels in patients with congenital heart defects. Static high-resolution anatomic images of the heart and great vessels can be obtained with “black-blood” spin-echo–based techniques with a double inversion recovery pulse sequence for suppression of the signal from blood. Black-blood spin-echo MR imaging techniques have the advantage of being less susceptible to artifact from the metallic implanted devices that are commonly seen in patients with congenital heart defects, such as stents, coils, occluder devices, clips, and sternal wires.

The recently developed technique of isotropic three-dimensional (3D) SSFP imaging allows reliable assessment of the complex cardiac anatomy, including the cardiac chambers, as well as extracardiac structures such as the coronary arteries, in patients with complex congenital heart defects (6). Instead of a series of 2D nonisotropic acquisitions, possibly with different angulations, a single operator-independent acquisition provides all the necessary information. The isotropic data allow arbitrary reconstruction of images in any plane desired, without any loss of spatial resolution. The image data acquisition is timed to coincide with the relatively quiescent phase of end diastole to avoid the effects of cardiac motion, and navigator-echo respiratory gating is performed to avoid respiration-related motion artifacts. This 3D SSFP technique obviates the administration of a gadolinium-based contrast material. Instead, a spectrally selective fat saturation pulse and a T2 preparatory pulse are used to increase the contrast among blood, fat, and myocardium.

Contrast-enhanced MR angiography allows a detailed study of the thoracic vasculature. A rapid non–cardiac-gated breath-hold 3D spoiled gradient-echo acquisition is performed during the intravenous bolus injection of a gadolinium-based contrast agent. Multiplanar reformatting of the acquired 3D image data set can be performed to generate 2D images in any orientation. Maximum intensity projection and volume rendering can be performed to display the image data in three dimensions, thus facilitating the visualization of overlapping anatomic structures. In patients in whom the administration of gadolinium is contraindicated, unenhanced MR angiography can be performed by using a 3D SSFP imaging technique, as described in the preceding paragraph (7).

Velocity-encoded phase-contrast MR imaging, which allows flow velocity mapping and flow quantification in thoracic blood vessels and across heart valves, also has various possible applications in patients with congenital heart defects. Specifically, phase-contrast imaging may be performed to quantify intracardiac and extracardiac shunts (ie, measure the ratio of pulmonary flow to
systemic flow \[Q_p/Q_s\] ratio), measure differential pulmonary flow, determine valvular regurgitant fractions, and estimate the pressure gradient in the presence of stenosis. The pressure gradient may be estimated in millimeters of mercury by using the modified Bernoulli equation 4 \(\cdot v^2\), where \(v\) is the peak velocity in meters per second. In vivo and in vitro studies have shown that blood flow measurements obtained with velocity-encoded MR imaging are both accurate and reproducible (8,9).

Delayed myocardial enhancement imaging, a sensitive and specific technique for depicting myocardial infarction, is also useful for detecting myocardial fibrosis in patients with congenital heart defects. The gadolinium-based MR imaging contrast agents in clinical use are T1 shortening, extravascular, and extracellular. Infarcted and fibrotic regions provide a larger distribution volume for these agents than does normal myocardium. As a result, within 10–15 minutes after intravenous injection of the contrast agent, a greater tissue concentration of the agent is visible in infarcted or fibrosed myocardium than in viable tissue. The application of a T1-weighted inversion-recovery sequence in which the inversion time is set to null the signal from viable myocardium produces images in which infarcted and fibrotic tissues appear bright because of their shorter T1. There is growing evidence to support the use of delayed myocardial enhancement imaging in adult patients with congenital heart defects for predicting posttreatment outcomes.

In the following sections, examples of pre- and postoperative MR imaging protocols for the evaluation of each conotruncal anomaly are outlined. It should be borne in mind that modifications in these protocols may be necessary to address anatomic and functional abnormalities specific to individual patients. Online (real-time) review of the initial image data should be performed to allow the detection of previously unsuspected abnormalities and appropriate adjustment of the acquisition protocol.

**Tetralogy of Fallot**

TOF is the most common cyanotic cardiac defect, with an annual incidence of approximately 1300 cases, or 3.26 per 10,000 live births, in the United States (10). The primary problem in TOF is underdevelopment of the pulmonary infundibulum (11). The classic tetrad of manifestations—pulmonary outflow tract stenosis or atresia, ventricular septal defect, aortic override (4), and right ventricular hypertrophy—are secondary to hypoplasia of the pulmonary infundibulum (Fig 1). The surgical anatomy of TOF has been well described (12). There is anterior malalignment with anterior deviation of the conal septum and infundibular hypoplasia, which constitute the anatomic subpulmonary obstruction. Infundibular stenosis may be complicated...
by a more proximal intracavitary obstruction produced by hypertrophied septal and parietal muscle bundles. The pulmonary valve is often abnormal, with a variably hypoplastic annulus and thickened, fused, and doming leaflets producing valvular stenosis. Frequently there is right-sided obstruction at multiple levels in TOF. The main or branch pulmonary arteries may be hypoplastic or atretic and may be affected by discrete peripheral stenosis (Fig 2). A substantial percentage of patients with TOF have atresia instead of stenosis of the pulmonary valve, with no physiologic antegrade pulmonary blood flow. In these patients, a spectrum of manifestations of hypoplasia or atresia of the central pulmonary arteries may be seen. Pulmonary blood flow may come from a patent ductus arteriosus or aortopulmonary collateral arteries. In a rare anatomic variant, the pulmonary valve leaflets are not stenotic or atretic but are completely absent; free pulmonary regurgitation, pronounced right ventricular dilatation, and marked dilatation of the main and branch pulmonary arteries are characteristic features of this variant (Fig 3). The ventricular septal defect is typically large, unrestrictive, and subaortic, involving the membranous septum. The extent of the aortic override is variable, and the overriding aortic root is typically enlarged. About 25% of patients have a right aortic arch, most often with mirror-image branching. As many as 5% of patients have an important coronary anomaly.
in which part or all of the territory of the left anterior descending artery is supplied by a large branch of the right coronary artery that crosses over the RVOT (Fig 4). This anatomic configuration may complicate or preclude infundibulotomy, which is part of the standard surgical procedure for repair of TOF, and may mandate the use of an alternative approach.

Current management of TOF consists of early single-stage reconstructive surgery, which is usually performed during the 1st year of life, often in the first 6 months (13,14). Staged reconstruction is required if there is significant hypoplasia of the central pulmonary arteries.

In patients with TOF and pulmonary stenosis, the goals of definitive repair are to close the ventricular septal defect and relieve the RVOT obstruction. Both transatrial-transpulmonary and transventricular surgical approaches are described in the literature. The transventricular approach previously was standard; however, the transatrial-transpulmonary approach currently is favored because it avoids a right ventricular incision and helps preserve right ventricular structure and function (15). In the transventricular approach, a longitudinal incision is made in the free wall of the infundibulum. If the annulus is hypoplastic, the incision extends distally across it. The ventricular septal defect is visualized and repaired through the infundibulotomy, and any obstructing right ventricular muscle bundles are resected. Thickened and obstructing pulmonary valve leaflets also are resected. An outflow tract patch is applied to augment the circumference of the RVOT. If a transannular incision is used, the patch continues into the main pulmonary artery and to one or both pulmonary artery origins if they are narrow. In the transatrial-transpulmonary approach, subpulmonary resection and ventricular septal defect closure are performed through a right atrial incision. If a pulmonary valvotomy or valve ring widening is necessary, it is achieved through a pulmonary arteriotomy.

Management of a patient with TOF and pulmonary atresia is contingent on the individual anatomy, particularly the extent of atresia, severity of pulmonary artery hypoplasia, and degree of collateralization. It is possible in some patients to perform surgical repair with a single procedure, but in many patients a staged approach is required (16,17). The goals are to restore antegrade flow from the right ventricle to the pulmonary artery, minimize competing collateral flow, and eventually close the intracardiac shunts. It may be necessary to perform coil embolization of excessive aortopulmonary collateral vessels that deliver competing blood flow to the lungs. Long-segment atresia involving much of the main pulmonary artery may require the creation of a right ventricle–pulmonary artery homograft conduit. If the native pulmonary arteries supply only a limited number of bronchopulmonary segments, it is important to recruit additional segments supplied solely by collateral vessels for inclusion in the reconstructed pulmonary arterial tree. This is accomplished with an approach known as unifocalization, in which collateral vessels are detached from the aorta and anastomosed to the central pulmonary artery confluence. Depending on the size of the reconstructed pulmonary arteries, the ventricular
septal defect may or may not be closed during the initial surgical procedure. Patients with TOF and pulmonary atresia typically require one or more additional procedures as the anatomy continues to grow and mature; surgically created conduits between the right ventricle and pulmonary artery have a fixed diameter and must be replaced or enlarged when they become restrictive.

Outcomes after complete repair of TOF are generally excellent, with early mortality of less than 2% and with survival of nearly 90% at 20 years after surgery (18). Postoperative complications that require follow-up include residual pulmonary regurgitation; right ventricular dilatation and dysfunction due to pulmonary regurgitation, possibly with associated tricuspid regurgitation; residual RVOT obstruction, branch pulmonary artery stenosis, or hypoplasia; sustained ventricular tachycardia, atrioventricular block, atrial flutter, and atrial fibrillation, alone or in combination; ventricular septal defect patch leak; residual aortopulmonary collateral vessels; and aortic root dilatation (19).

Most patients who undergo surgical repair of TOF have residual hemodynamic abnormalities, primarily pulmonary insufficiency, which leads to progressive right ventricular dilatation and systolic dysfunction. Cardiac rhythm disturbances and sudden cardiac death contribute to postoperative morbidity and mortality.

Preoperative MR Imaging

Transthoracic echocardiography is the imaging modality of choice for initial diagnosis and preoperative assessment of unrepaired TOF in infants. Cardiovascular MR imaging is more informative when acoustic windows are poor, particularly for the evaluation of extracardiac anatomy in some older patients and, rarely, in infants. The primary goal of the cardiovascular MR imaging examination in patients with unrepaired TOF is to delineate the anatomy of the pulmonary vascular bed and identify sources of pulmonary blood flow, including aortopulmonary collateral vessels and ductus arteriosus. Gadolinium-enhanced 3D MR angiography is robust and efficient, and in comparison with conventional angiography, it has been shown to have high accuracy in depicting all sources of pulmonary blood in patients with complex pulmonary stenosis or atresia (20). MR angiography allows the identification of the size of the central pulmonary artery and branch pulmonary arteries, assessment of their continuity with the RVOT and the distance between any discontinuous vessels, and detection of aortopulmonary collateral vessels (Fig 5), and thus may obviate cardiac catheterization. A free-breathing whole-heart 3D SSFP or black-blood fast spin-echo sequence may be applied to allow evaluation of the proximal coronary arteries when they cannot be evaluated with other techniques. Attention should be paid to the exclusion of a major coronary artery crossing the RVOT.

A standard cardiovascular MR imaging protocol for evaluating a patient with unrepaired TOF and pulmonary atresia includes the following: localization imaging in three planes;
two-chamber (left-heart), four-chamber, and, optionally, short-axis (ventricular function) cine SSFP imaging of the heart; axial black-blood imaging of the pulmonary and proximal coronary arteries; gadolinium-enhanced 3D MR angiography; and whole-heart isotropic 3D SSFP imaging (Table E1, A [online]).

Postoperative MR Imaging
The main role of cardiovascular MR imaging in patients with TOF is in the assessment of postoperative complications. Accurate quantification of right and left ventricular dimensions and pulmonary regurgitation and clear definition of RVOT anatomy are particularly important when deciding the type and timing of further intervention. Surgical or transcatheter pulmonary valve replacement is indicated for symptomatic patients with severe pulmonary regurgitation and moderate to severe right ventricular dysfunction (19). To maintain adequate right ventricular contractility in these patients, pulmonary valve implantation should be performed before the right ventricular function deteriorates. Right ventricular volumes were successfully reduced to normal values when pulmonary valve replacement was performed before the right ventricular end-diastolic volume reached 160 mL/m² and the right ventricular end-systolic volume reached 82 mL/m² as measured at cardiovascular MR imaging (21). Residual RVOT obstruction (valvular or subvalvular) may require surgery. Branch pulmonary artery stenosis contributing to right ventricular dysfunction may be treated with balloon angioplasty. Progressive aortic root dilatation and aortic regurgitation may develop in some patients and should be sought and evaluated at regular intervals (19).

Findings of right ventricular dilatation and dysfunction at cardiovascular MR imaging are predictive of adverse outcomes such as right heart failure, major arrhythmic events, and death (22,23) (Fig 6a). The degree of pulmonary regurgitation measured at velocity-encoded MR imaging is closely associated with the degree of right ventricular dilatation (24–26) (Fig 6b, Movie 1 [online]). The presence of pulmonary artery end-diastolic forward flow at velocity-encoded MR imaging correlates with more severe pulmonary regurgitation and poorer exercise performance at mid- to long-term follow-up evaluations (27). Left ventricular systolic dysfunction in long-term survivors is independently associated with impaired clinical status and, as a result of right ventricle–left ventricle interaction, is correlated with right ventricular dysfunction (22). The presence of regional dysfunction such as that produced by aneurysms in the RVOT (Fig 7a, Movie 2 [online]), which are relatively common after transannular patch repair, has an adverse effect on the global right ventricular function, left ventricular systolic function, and ex-
Exercise capacity after TOF repair (28,29). Recently, delayed enhancement at surgical sites (Fig 7b) and elsewhere in the heart, including the left ventricle, was shown to correlate with adverse outcomes, including ventricular dysfunction, exercise intolerance, and clinical arrhythmia (30,31). In conjunction with clinical findings and electrophysiologic data, information derived from cardiovascular MR imaging about the presence and severity of a pulmonary regurgitant fraction, the dimensions and function of the right and left ventricles, the presence and extent of an RVOT aneurysm, and the presence of branch pulmonary artery stenosis is used to decide the timing of further intervention in patients after TOF repair.

The goals of cardiovascular MR imaging after surgical repair of TOF include quantitative assessment of the right and left ventricular volumes, stroke volume, and ejection fraction; anatomic evaluation of the RVOT, pulmonary arteries, aorta, and aortopulmonary collateral vessels; quantification of pulmonary regurgitation, tricuspid regurgitation, cardiac output, and pulmonary flow-to-systemic flow ratio; and identification of regions of myocardial scarring. A standard cardiovascular MR imaging protocol for evaluating patients after a TOF repair includes localization in three planes; two-chamber (left), four-chamber, and short-axis cine SSFP imaging of the heart; oblique sagittal and, possibly, oblique coronal cine SSFP imaging of the RVOT; cine SSFP imaging in an orthogonal axial plane centered on the pulmonary arteries; three-chamber and oblique coronal cine SSFP imaging of the LVOT; axial black-blood imaging of the pulmonary arteries if a metallic artifact is seen on cine SSFP images; velocity-encoded MR imaging in a plane perpendicular to the main pulmonary artery (and branch pulmonary arteries, for measurement of differential flow), aortic root, and the atioventricular valves; gadolinium-enhanced 3D MR angiography; and delayed myocardial enhancement imaging along both the short and the long axes (Table E1, B [online]).

A baseline study should be performed in late childhood, when sedation is no longer necessary (8–10 years of age). Serial follow-up MR imaging evaluations should be performed every 1–4 years thereafter, depending on the initial findings and clinical symptoms.

**D-Loop Transposition of the Great Arteries**

Transposition of the great arteries is defined by discordant connections between the ventricles and the great arteries: The aorta arises from the right ventricle, and the pulmonary artery arises...
Figure 8. Postoperative appearance of d-transposition of the great arteries after a Mustard procedure with creation of a superior vena cava (SVC) baffle. (a) Oblique coronal image from 3D SSFP MR imaging shows ventriculoarterial discordance, with the aorta (A) arising from the morphologic right ventricle (R) and the pulmonary artery (P) arising from the morphologic left ventricle (L). (b) Maximum intensity projection image from gadolinium-enhanced 3D MR angiography shows patency of the baffle directing blood flow from the pulmonary vein (PV) to the right ventricle (RV). A = aorta. (c) Oblique coronal image from 3D SSFP MR imaging shows distal stenosis (arrow) of the superior vena cava (SVC) baffle at the opening to the left atrium (LA). Atrioventricular concordance, with the left atrium opening into the left ventricle (LV), also is depicted. (Case courtesy of P. Beerbaum, Evelina Children’s Hospital, King’s College, London, England.)

from the left ventricle. The most common type of transposition of the great arteries is a d-loop, an anatomic arrangement summarized by the annotation “S,D,D,” in which S stands for visceroatrial situs solitus (the normal morphologic arrangement, with the right atrium located to the right of the morphologic left atrium, the inferior vena cava to the right of the aorta, the liver on the right, and the spleen on the left), the first D stands for a d-loop (location of the morphologic right ventricle on the right side and the morphologic left ventricle on the left side), and the second D stands for d-malposition of the aortic valve in relation to the pulmonary valve. In d-malposition, the aortic valve is situated to the right of and anterior to the pulmonary valve, and the great arteries are parallel instead of crossing as they do in the normal heart.

d-loop transposition of the great arteries is the second most common cyanotic congenital heart condition diagnosed in the 1st year of life, with an incidence of 315 per million live births (10).

As a result of atrioventricular concordance and ventriculoarterial discordance, systemic venous blood passes through the right heart to the aorta, whereas pulmonary venous blood passes through the left heart to the lungs. Survival is dependent on the existence of a communication between the parallel pulmonary and systemic circulations (ie, a patent ductus arteriosus, ventricular septal defect, or atrial septal defect). A ventricular septal defect is present at birth in 50% of patients with a d-loop transposition of the great arteries. Other associated anomalies include pulmonary stenosis, pulmonary atresia, overriding or straddling atrioventricular valves, aortic coarctation, interrupted aortic arch, and right ventricular hypoplasia.

Until the mid 1980s, this defect was surgically corrected with an atrial-level switch (Mustard or Senning) procedure. In both the Mustard and the Senning variants of this procedure, the systemic and pulmonary venous blood is redirected within the atria so that the pulmonary venous blood reaches the tricuspid valve, right ventricle, and aorta, whereas the systemic venous blood reaches the mitral valve, left ventricle, and pulmonary arteries. In the Mustard procedure, pericardium is used to redirect blood flow, whereas in the Senning procedure native atrial tissue is used (32). Beginning in the mid 1980s, the arterial switch (Jatene) procedure largely replaced the atrial switch procedure, and it remains the current
standard of care (32,33). In the arterial switch procedure, the coronary arteries are excised from the aorta with a small cuff of the aortic wall attached. Next, the ascending aorta and the pulmonary artery are transected and switched. Before the vessels are anastomosed, the pulmonary artery is relocated anterior to the aorta. This relocation of the neopulmonary artery, known as the LeCompte maneuver, was added to the procedure in 1981. The LeCompte maneuver involves threading the ascending aorta behind the bifurcation of the pulmonary artery to maximize the length of the aorta and thus reduce the risk of coronary artery kinking or compression (34). Small openings are created in the neo-aorta near its origin from the left ventricle, and the coronary arteries are implanted. Because of complications related to pulmonary stenosis with direct anastomosis of the pulmonary artery, the pulmonary artery is now generally reconstructed by using pericardial patch augmentation. The arterial switch procedure has the advantage of establishing the left ventricle as the systemic ventricle, producing physiologically and anatomically normal circulation.

Although intraatrial repair has been superseded by the arterial switch procedure, a sizable population has undergone either a Senning or a Mustard procedure. The most common early structural complications included baffle obstruction, which most commonly affects the superior limb of the baffle at the junction of the right atrium with the superior vena cava (Fig 8) (19). Baffle leaks occur in an estimated 25% of patients. Most such leaks are small, but many pose a risk of paradoxical embolus, particularly in the presence of atrial arrhythmia and an endocardial pacemaker. Pulmonary venous obstruction also may occur but is less common. Subpulmonary stenosis and pulmonary stenosis may occur, in part because of the abnormal geometry of the left ventricle, which becomes distorted and compressed by the enlarged right ventricle. The most important long-term complication after the creation of an atrial baffle is failure of the systemic right ventricle with resultant tricuspid regurgitation (Fig 9, Movie 3 [online]). Important but less common complications include pulmonary arterial hypertension, residual ventricular septal defect, dynamic subpulmonary stenosis, and a host of conduction and rhythm disturbances that might necessitate implantation of a permanent pacemaker or result in sudden death.

Complications that may be seen at imaging after an arterial switch procedure include RVOT obstruction, main pulmonary artery and branch
pulmonary artery obstruction, aortic root dilatation with aortic regurgitation, and coronary artery stenosis (19). The main late complications are RVOT and branch pulmonary artery obstruction. The most frequent types of obstruction are (a) discrete narrowing at the suture line of the anastomosis and (b) long-segment obstruction in the pulmonary arteries (especially the right pulmonary artery [Fig 10]) because they are draped across the aorta behind the sternum (35). Pulmonary stenosis is the most frequent reason for repeat surgery in long-term survivors of the arterial switch procedure (36). RVOT–branch pulmonary artery narrowing may be associated with pulmonary regurgitation and right ventricular dysfunction. A less common complication of the arterial switch procedure is coronary ostial stenosis and ischemia secondary to reimplantation. Most coronary complications result in early postoperative mortality; however, a small number of patients have experienced a later postoperative coronary event (37). After the great arteries are reversed in the arterial switch procedure, the native pulmonary valve fulfills the function of a neo–aortic valve. Neo–aortic valve regurgitation and neo–aortic root dilatation (Fig 11) may develop over time (38).

In cases in which d-transposition of the great arteries is associated with a ventricular septal defect and subpulmonary stenosis, the Rastelli procedure is preferred. This procedure consists of a patch closure of the ventricular septal defect to the aortic valve and placement of a conduit between the right ventricle and the pulmonary arteries (32). Complications that have been recognized after a Rastelli procedure include
Figure 12. Postoperative complications after a Rastelli procedure for treatment of d-transposition of the great arteries with subaortic ventricular septal defect and subpulmonary stenosis in a 17-month-old infant. A = aorta, LV = left ventricle, PA = pulmonary artery, RV = right ventricle. (a) Oblique coronal SSFP MR image shows a ventricular septal defect patch (black arrowheads) and a postoperative LVOT aneurysm (white arrowheads). (b, c) Oblique sagittal (b) and axial (c) images from gadolinium-enhanced 3D MR angiography depict severe stenosis (arrowheads) of the distal part of the right ventricle–pulmonary artery conduit. (d) Volume-rendered image from 3D MR angiography not only helps confirm the finding of a distal conduit stenosis (arrowhead) but also shows a large aneurysm arising from the left ventricle.

RVOT and pulmonary conduit obstruction (Fig 12), superimposed right ventricular failure, and tricuspid regurgitation (19). Other possible complications include LVOT obstruction caused by an intraventricular baffle, arrhythmia due to atriotomy or ventriculotomy, residual ventricular septal defect, myocardial hypertrophy, chamber enlargement, aortic root dilatation, and aortic valve regurgitation. Currently, transthoracic echocardiography is the imaging modality of choice for the preoperative diagnosis and assessment of d-transposition of the great arteries. The main role of MR imaging is in the diagnosis of postoperative complications, particularly those that develop as the child grows older.
Post–Atrial Switch MR Imaging
The goals of cardiovascular MR imaging after an atrial switch procedure include evaluations of the ventricles for size and function, pulmonary and systemic venous pathways for baffle leak and stenosis, atrioventricular valves for regurgitation, and outflow tracts for obstruction. Delayed gadolinium-enhanced MR imaging can be used to detect myocardial fibrosis in the systemic right ventricle, a finding that is associated with right ventricular dysfunction, poor exercise tolerance, arrhythmia, and progressive clinical deterioration (39). The response of the systemic right ventricle to stress induced by the administration of dobutamine or by exercise can be assessed at MR imaging; submaximal responses were found during such stress tests in patients who had undergone the atrial switch procedure, in comparison with the responses in controls (40–42). Further data are required before this technique can be used in routine clinical practice.

A standard cardiovascular MR imaging protocol for evaluating patients after an atrial switch procedure includes the following: localization imaging in three planes; four-chamber or axial cine SSFP imaging in multiple contiguous sections from the level of the diaphragm to the level of the aortic arch (to allow dynamic evaluation of the pulmonary venous baffles, qualitative assessment of ventricular and atrioventricular valve function, and assessment of the great arteries); oblique coronal cine SSFP imaging in multiple contiguous sections parallel to the superior and inferior venae cavae (to allow detection of a systemic venous baffle obstruction or leak); short-axis cine SSFP imaging for quantitative assessment of right and left ventricular function; velocity-encoded MR imaging of the aortic root, main pulmonary artery, and atrioventricular valves; gadolinium-enhanced 3D MR angiography; whole-heart isotropic 3D SSFP imaging; and delayed myocardial enhancement imaging along the short and long axes (Table E2, A [online]). After a baseline investigation, MR imaging may be repeated every 2–4 years for serial monitoring of ventricular function and systemic and pulmonary venous baffles.

Post–Arterial Switch MR Imaging
Because the RVOT and the branch pulmonary arteries are positioned immediately behind the sternum after the arterial switch procedure, transthoracic echocardiography is a poor method for evaluating obstructive lesions in these vessels. The anatomic and functional significance of such obstructions can be more accurately assessed with MR imaging by using a combination of techniques including spin-echo and SSFP imaging, gadolinium-enhanced 3D MR angiography, and velocity-encoded MR imaging (43,44). In patients in whom the presence of coronary ischemia is suspected, conventional coronary angiography is probably the imaging modality of choice (19). However, coronary MR angiography is useful for the noninvasive investigation of proximal segments of the coronary arteries. Other possible roles for MR imaging include the evaluation of myocardial perfusion during pharmacologic stress testing with adenosine or dobutamine and the assessment of myocardial viability (detection of scarring), but these uses are still under investigation.

A cardiovascular MR imaging protocol for post–arterial switch examinations includes the following: axial cine SSFP imaging in contiguous sections from the diaphragm to the transverse arch (to allow dynamic visualization of the great vessels, including the branch pulmonary arteries, and qualitative assessment of ventricular and atrioventricular valve function); oblique sagittal and oblique coronal cine SSFP imaging in planes parallel to the RVOT and LVOT (ideally, two orthogonal views each for the RVOT and LVOT); two-chamber (left), four-chamber, and short-axis cine SSFP imaging of the heart; velocity-encoded imaging of the aortic root and the main and branch pulmonary arteries; gadolinium-enhanced 3D angiography; whole-heart isotropic 3D SSFP imaging; and delayed myocardial enhancement imaging in short- and long-axis planes (Table E2, B [online]).

Depending on the initial findings and clinical symptoms, the MR imaging examination may be repeated at regular follow-up intervals.

1-Loop Transposition of the Great Arteries
The second most common type of transposition of the great arteries is congenitally corrected transposition, or 1-loop transposition of the great arteries. The segmental anatomy can be described
in shorthand as “S,L,L,” with S signifying the presence of situs solitus, the first L signifying an L-loop (with the morphologic right ventricle positioned to the left of the morphologic left ventricle), and the second L signifying L-malposition of the aortic valve in relation to the pulmonary valve. Congenitally corrected transposition of the great arteries is rare and is characterized by atrioventricular and ventriculoarterial discordance (Fig 13a, 13b). The two outflow tracts are parallel, and the ventricular septum lies in a more anteroposterior position. The aorta is usually anterior to and left of the pulmonary artery (Fig 13c). Congenitally corrected transposition is also known as physiologically corrected transposition because the systemic venous return reaches the pulmonary circulation through the right-sided left ventricle, and the pulmonary venous return reaches the aorta through the left-sided right ventricle. The condition is not cyanotic and may be discovered incidentally in asymptomatic patients. However, most patients have an associated cardiac lesion such as a ventricular septal defect, tricuspid valve abnormality (eg, Ebstein anomaly), or pulmonary stenosis (45). Even without an associated abnormality, most patients with congenitally corrected transposition develop systemic ventricular failure over time because of the inability of the right ventricle to cope with the systemic overload (46). Tricuspid valvular (systemic atrioventricular) regurgitation is strongly associated with right ventricular dysfunction and congestive heart failure, but whether the regurgitation is a cause or an effect is subject to speculation. In general, cases with no associated structural problem or only a minor structural problem can be managed medically; the role of MR imaging in these cases is mainly in the assessment of systemic right ventricle function and associated tricuspid regurgitation. Systemic atrioventricular valve replacement is often required in adult patients (45).
In patients with a clinically significant associated lesion, surgical repair is performed. In the classic surgical procedures, the tricuspid valve is repaired or replaced and the ventricular septal defect is closed or a left ventricle–pulmonary artery conduit is created, with the morphologic right ventricle remaining the systemic ventricle. However, late failure of the systemic right ventricle after such procedures has led to the increasing use of combined techniques for anatomic surgical repair, such as the combined atrial and arterial switch (double switch) procedure and, in the presence of an associated ventricular septal defect, the combined Rastelli and atrial switch procedure (45,47).

Double-Outlet Right Ventricle

DORV is said to be present when both great arteries arise predominantly from the right ventricle (Fig 14, Movie 4 [online]). The incidence of DORV is estimated at 127 per million live births (10).

DORV is not a single cardiac anomaly; rather, the term is used to describe an aberrant position of the great arteries in association with various cardiac anomalies in which physiologic function...
is similar to that in ventricular septal defect, TOF, transposition of the great arteries, presence of a single ventricle, or atrioventricular atresia. In most patients the aorta and pulmonary artery are side by side, with the former on the right (d-malposition) (48). Continuity between the mitral valve and the adjacent semilunar valve is absent, a finding that is thought to represent the sine qua non for this diagnosis. The conal musculature is usually seen inferior to both great arteries (Fig 15). A ventricular septal defect is almost always present; its location in relation to the semilunar valves may be subaortic (50% of cases) (a), subpulmonary (30% of cases) (b), uncommitted or remote (c), or doubly committed (d) in relation to the semilunar valves. Arrows indicate the direction of blood flow from the left ventricle to the LVOT and RVOT. A = anterior limb of the septomarginal trabecula, Ao = aorta, IS = infundibular septum, P = posterior limb of the septomarginal trabecula, PA = pulmonary artery, RA = right atrium, RV = right ventricle, SMT = septomarginal trabecula.

Figure 16. Diagrams show the four types of DORV. The location of the ventricular septal defect with regard to the semilunar valves significantly affects physiologic function and determines the classification and surgical management of cases of DORV. The ventricular septal defect may be subaortic (50% of cases) (a), subpulmonary (30% of cases) (b), uncommitted or remote (c), or doubly committed (d) in relation to the semilunar valves. Arrows indicate the direction of blood flow from the left ventricle to the LVOT and RVOT. A = anterior limb of the septomarginal trabecula, Ao = aorta, IS = infundibular septum, P = posterior limb of the septomarginal trabecula, PA = pulmonary artery, RA = right atrium, RV = right ventricle, SMT = septomarginal trabecula.

circulation may be obstructed at the aortic valve or in a subaortic location. Coarctation of the aorta is frequently found in patients with a subpulmonary ventricular septal defect, particularly when there is stenosis or atresia of the aortic valve. Coronary artery anomalies also may be encountered, including origin of the right coronary artery from the left main coronary artery (Fig 14); duplication of the left anterior descending coronary artery; and anomalous origin of the left anterior descending coronary artery from the right coronary artery (a variant that is associated with a subaortic ventricular septal defect and pulmonary stenosis).

The relationship between the ventricular septal defect and the great arteries, the relative outflow obstruction, the size of the right ventricular and left ventricular sinuses, and the anatomy of the atrioventricular valves determine the physiologic function, clinical course, and surgical management of DORV. The relationship between the ventricular septal defect and the outflow tracts is particularly important and provides a basis for the classification of cases of DORV (Fig 16). Common anatomic-physiologic variants include the following (listed in order of decreasing...
frequency): (a) a TOF-like variant consisting of DORV with a subaortic ventricular septal defect and pulmonary stenosis; (b) a variant resembling transposition of the great arteries, consisting of DORV with a subpulmonary ventricular septal defect (Taussig-Bing anomaly); (c) a variant resembling ventricular septal defect, consisting of DORV with a subaortic ventricular septal defect but without pulmonary stenosis; and (d) a variant resembling a univentricular heart, consisting of DORV with mitral atresia, an unbalanced atrioventricular canal, or severe hypoplasia of one of the ventricular sinuses (50).

Biventricular repair that leaves the patient with functioning right and left ventricles is the goal, when possible. The orientation of the ventricular septal defect is critical for selecting the appropriate surgical approach to avoid obstructing a newly created subaortic or subpulmonary outflow channel.

Repair of DORV in the presence of a subaortic ventricular septal defect depends on the proximity of the ventricular septal defect to the aorta. In most cases, an intraventricular tunnel constructed from a Gore-Tex fabric patch may be used as a baffle to direct blood flow from the left ventricle through the ventricular septal defect to the aorta (51). If the ventricular septal defect is small, obstruction of the tunnel may be avoided by enlarging the defect. RVOT obstruction may necessitate surgical resection or the creation of a patch or conduit similar to those created in surgical repair of TOF. Most such repairs are performed in the 1st year of life. Repair of DORV with a subpulmonary ventricular septal defect (Taussig-Bing anomaly) is now accomplished with an arterial switch procedure and a ventricular septal defect patch (52). Associated aortic coarctation or hypoplasia is repaired at the same time. Repair of DORV with a doubly committed ventricular septal defect often resembles the repair of a large ventricular septal defect. At times, the ventricular septal defect may be more closely related to the pulmonary artery, and an arterial switch may be required; alternatively, it may be more closely related to the aorta, and the creation of an intraventricular tunnel (baffle) may be preferred. Repair of DORV in a patient with a remote ventricular septal defect can be especially complex. However, such lesions are rare, and most patients undergo single-ventricle palliative surgery with pulmonary banding as an infant and a Fontan procedure (total cavopulmonary connection) later, in early childhood. In a few patients, biventricular repair may be accomplished with the construction of a long tunnel, resection of infundibular muscle, and an arterial switch procedure. Severe hypoplasia or absence of one of the ventricular sinuses, substantial straddling of an atrioventricular valve, or significant atresia of the mitral valve also may necessitate single-ventricle palliative surgery with a subsequent Fontan procedure.

Late complications after surgical repair of DORV vary with the individual anatomy and physiologic function, as well as the type of surgical procedure. Subaortic stenosis may develop after a baffle is placed to direct blood flow from the left ventricle to the aorta (53). After a TOF-type repair of DORV in the presence of a pulmonary outflow tract obstruction, chronic pulmonary regurgitation or right ventricular dilatation and dysfunction may develop. Those who undergo an arterial switch procedure may have similar complications, as described in the preceding section: RVOT obstruction, coronary artery obstruction, and aortic root dilatation. An aortic arch obstruction may arise after surgical repair of coarctation or interruption.
Figure 18. Postoperative appearance of DORV repair in a 37-year-old man. The preoperative physiologic function resembled that in TOF with a subaortic ventricular septal defect and pulmonary stenosis. (a) Axial SSFP MR image shows an intraventricular tunnel (T) created to channel blood flow from the left ventricle (LV) to the aorta (A) through the ventricular septal defect. (b) Oblique sagittal SSFP MR image shows bulging of the thin anterior wall of the right ventricle at the level of the RVOT patch (arrow). The marked right ventricular dilatation is due to severe pulmonary regurgitation.

Preoperative MR Imaging
In most cases, echocardiography is sufficient for diagnosis and surgical planning in infants and newborns with DORV. However, when the findings at that initial imaging examination are inconclusive, cardiovascular MR imaging may play an important role because it accurately depicts the anatomy of the ventricular septal defect, its relationship to the semilunar valves, the morphologic structure and patency of the outflow tracts, and the state of the extracardiac vascular anatomy. Findings at preoperative black-blood spin-echo MR imaging of the ventricular septal defect correlate well with surgical findings and are predictive of the type of repair needed (54). Any coexistent anomalies of the aortic arch, pulmonary arteries, pulmonary veins (Fig 17), or aortopulmonary collateral vessels in infants with DORV are adequately depicted with gadolinium-enhanced 3D MR angiography. The intracardiac anatomy, ventricular function, and outflow tract dynamics may be assessed with a combination of dynamic 2D cine SSFP and black-blood double inversion recovery imaging techniques. Free-breathing electrocardiographically triggered navigator-gated isotropic 3D SSFP sequences provide volume data that can be reformatted in multiple planes to allow accurate assessment of both the intracardiac and the extracardiac anatomy, including the origin and course of the coronary arteries (6).

Postoperative MR Imaging
The role of cardiovascular MR imaging after DORV repair increases as patients grow older and acoustic windows become more limited. Follow-up imaging should be performed to evaluate the functional status of both ventricles and to identify any residual valvular or subvalvular stenosis or insufficiency (especially subaortic or subpulmonary obstruction if a tunnel-type surgical procedure was performed), residual ventricular septal defect, residual coarctation or recoarctation, and RVOT or conduit stenosis or regurgitation (Fig 18). The specific cardiovascular MR imaging protocol is tailored to the type of surgery; a protocol like that used for patients with a TOF repair or surgically corrected transposition of the great arteries may be applied, as described earlier. Evaluation of the LVOT and RVOT in two orthogonal views is important to rule out insufficiency, because dephasing jets may be eccentric and may be missed if only one view is obtained. Axial multisection cine SSFP imaging from the apex of the heart to the pulmonary artery bifurcation is useful for evaluating the intraventricular tunnel that leads from the left ventricle to the aorta, determining the state of the pulmonary arteries, and identifying a ventricular septal defect leak.
Truncus Arteriosus

Truncus arteriosus is an uncommon conotruncal anomaly, with a reported incidence of 94 per million live births (10). It is characterized by a single arterial vessel that originates from the heart, overrides the ventricular septum, and supplies the systemic, pulmonary, and coronary circulation, all from the proximal ascending vessel. An association with DiGeorge syndrome and chromosome 22q11 deletion is well recognized (55).

The classification of truncus arteriosus relies on the branching pattern of the pulmonary artery. The original classification system, which was devised by Collett and Edwards (56), was later modified by Van Praagh (57) (Fig 19).

The single arterial trunk is larger in diameter than the normal aorta at a comparable age. It is positioned above the ventricular septum, being dominant over either ventricle or shared equally by both. The truncal valve is often abnormal, with thickened and deformed leaflets that are variably stenotic or, more often, incompetent. Structurally, the valve is most often tricommissural, with a bicommissural structure being the second most common variant. The conal septum is usually absent, and a malalignment ventricular septal defect is present in almost all patients. An interrupted aortic arch is a common associated lesion (11%–14% of patients), as are abnormalities of the mitral valve, coronary arteries, and pulmonary venous connections (57,58).

Truncus arteriosus is usually diagnosed early, and surgical repair is undertaken within the first few weeks after birth. The ventricular septal defect is closed with a patch so that the truncal valve is aligned, with the left ventricle becoming the neo-aortic valve. The pulmonary arteries are detached from the arterial trunk and connected to the right ventricle by a valved homograft (14). Aortic arch interruption or coarctation is repaired at the same time. Mild truncal valve stenosis and minor regurgitation are left alone. Prosthetic valve placement at this age is not often possible, but valvuloplasty may be beneficial in some (59). The main complications after truncal repair are right ventricle–pulmonary artery conduit...
steno
calization imaging in three planes; two-chamber,
four-chamber, and short-axis cine SSFP imaging; sagittal oblique cine SSFP imaging of the RVOT; three-chamber and oblique coronal cine SSFP imaging of the LVOT; axial SSFP imaging of the pulmonary arteries, or if a metallic artifact (stent) is present, black-blood imaging of the pulmonary arteries; cine SSFP imaging in an oblique sagittal plane centered on the aortic arch in a patient with a previous repair; gadolinium-enhanced 3D angiography; and velocity-encoded imaging of the aortic root, main pulmonary artery, branch pulmonary arteries, and atrioventricular valves (Table E3 [online]).

**Interrupted Aortic Arch**

An interrupted aortic arch is defined by luminal discontinuity between the ascending and descending portions of the thoracic aorta (63). The discontinuity may be complete, or it may be spanned by an atretic fibrous band.Interrupted aortic arch is a rare condition; it is found in approximately 1% of infants with a critical congenital heart defect (63). An underlying genetic cause is thought to be responsible for many cases: A chromosome 22q11.2 deletion is identifiable in approximately 50% of patients with an interrupted aortic arch, and 42% of patients with DiGeorge syndrome have an interrupted aortic arch (63,64).

If an interrupted aortic arch is left untreated, death usually follows after physiologic closure of the ductus arteriosus (median age, 4–10 days) (65). Hypoperfusion-related complications, including acute renal failure and metabolic acidosis, develop after the ductus arteriosus closes (63). If there is extensive collateral flow, the patient may survive longer and the interrupted aortic arch may be diagnosed later in life; however, such occurrences are rare (63,66).

Occurrences of interrupted aortic arch are classified according to the location of the interruption (67) (Fig 22). In type A interrupted aortic arch, the interruption is located just beyond the left subclavian artery; in type B, it is between the left common carotid and left subclavian arteries; and in type C, it is between the innominate (brachiocephalic) and left common carotid arteries. Type B interrupted aortic arch is considered the most common variant, and type C is the least commonly observed. In their review of
95 cases of interrupted aortic arch, Schreiber et al (68) classified 13% as type A, 84% as type B, and 3% as type C.

An interrupted aortic arch rarely occurs in isolation; it almost always is found with one or more other congenital cardiovascular anomalies. A patent ductus arteriosus, which provides blood flow to the descending thoracic and abdominal aorta, is observed in nearly all cases. A ventricular septal defect is identified in approximately 90% of patients with an interrupted aortic arch (69). Other congenital defects that limit blood flow to the aortic arch also may be seen; these include aortic stenosis, bicuspid aortic valve, truncus arteriosus, and aortopulmonary window (63,64,69). Most patients with an interrupted aortic arch have some degree of aortic or subaortic stenosis. An aberrant right subclavian artery, typically arising from the right side of the proximal descending thoracic aorta, near the origin of the left subclavian artery, is another frequent finding in patients with an interrupted aortic arch.

 Interruption of the aortic arch is initially managed with intravenous prostaglandin to ensure continued patency of the ductus arteriosus. Surgical repair should be undertaken as soon as possible after the appropriate diagnostic imaging examinations. Generally, in all types of interruption, the aortic arch structures can be mobilized and a primary anastomosis of the proximal and distal segments of the arch can be carried out (14). Artificial conduits or homografts are rarely necessary to bridge the defect. In the preferred single-stage repair procedure, any associated cardiovascular lesions are corrected at the same time as the aortic interruption. On occasion, a two-stage approach may be preferred; however, this approach is associated with higher early mortality (70). Long-term postoperative survival has improved as surgical techniques have been refined; survival now approaches 85% at 12 years after repair of an interrupted aortic arch (71).

Postoperative complications that require follow-up include residual or recurrent arch obstruction, aneurysm formation at the surgical site, residual obstruction of the LVOT, ventricular septal defect patch leak, and left ventricular hypertrophy. In addition, associated congenital cardiac anomalies corrected at the time of surgery may require follow-up evaluation.

**Preoperative MR Imaging**

Detailed planning is imperative before surgical repair of an interrupted aortic arch is undertaken; the exact sites of interruption and of any associated congenital cardiovascular anomalies must be defined. Important anatomic considerations include the location and length of the aortic arch defect, size and appearance of the aorta proximal and distal to the interruption, branching pattern of the great vessels, location and patency of the ductus arteriosus, and presence of other congenital cardiovascular defects. Echocardiography is generally regarded as the first-line imaging modality for the preoperative work-up (72).

MR imaging is useful if the anatomy is not clearly delineated at echocardiography (73). MR imaging can accurately define cardiac, aortic, and great vessel anatomy as well as provide information pertaining to cardiac chamber and valve function. Various unenhanced MR imaging sequences may be useful, but gadolinium-enhanced 3D angiography provides particularly valuable information (Fig 23, Movie 5 [online]).

Various MR imaging features may be observed in the presence of an interrupted aortic arch (73). The most specific imaging finding is nonvisualization of a portion of the arch. The vascular
Figure 23. Preoperative appearance of interrupted aortic arch in a neonate. (a) Volume-rendered image from gadolinium-enhanced 3D MR angiography shows interruption of the aortic arch between the left common carotid artery (LCCA) and the left subclavian artery (LSA), a finding indicative of type B interrupted aortic arch. A = aorta, IA = innominate artery, MPA = main pulmonary artery, PDA = patent ductus arteriosus, RCCA = right common carotid artery, RSA = right subclavian artery. (b) Maximum intensity projection image from the same angiographic acquisition as a shows a persistent embryonic artery (EA) that communicates between the left common carotid artery and left subclavian artery. A patent ductus arteriosus (PDA) supplies blood flow to the descending thoracic aorta (DA). A = ascending aorta.

defect should be identified on multiple images obtained with different sequences and in different planes to increase diagnostic specificity. An interrupted aortic arch also may be indicated by the appearance of a complete thoracic vascular arch on a single true sagittal image. Although this feature mimics the normal aortic arch, it represents a patent ductus arteriosus; the normal arch is obliquely oriented within the thorax and thus should not be visible on a single sagittal image. The vascular arch formed by a patent ductus arteriosus also appears flattened in comparison with a normal aortic arch, and the ascending thoracic aorta may appear diminished in caliber because of decreased blood flow.

Postoperative MR Imaging
Because patients with surgical repair of an interrupted aortic arch are living longer, posttreatment imaging has increasing importance. MR imaging can be used to evaluate both pediatric and adult patients for a variety of postoperative complications, including anastomotic narrowing (Fig 24) and aneurysmal dilatation. Associated cardiovascular abnormalities such as altered left ventricular size and function, LVOT obstruction, residual ventricular septal defect, aortic valve stenosis or regurgitation, and other anomalies also should be evaluated, depending on the preoperative anatomy and surgical correction. These objectives can be achieved by using the following protocol: localization imaging in three
planes; two-chamber, four-chamber, and short-axis cine SSFP imaging; three-chamber and oblique coronal cine SSFP imaging of the LVOT; oblique sagittal cine SSFP imaging, and optionally, black-blood imaging, of the aortic arch; gadolinium-enhanced 3D MR angiography; and velocity-encoded imaging in planes perpendicular to the ascending and descending aorta and orthogonal to any site of residual aortic arch narrowing (Table E4 [online]).

Measures of the hemodynamic severity of residual or recurrent aortic arch narrowing include the body-surface-area–adjusted cross-sectional area of the aortic arch (obtained at gadolinium-enhanced 3D MR angiography) and the heart rate–adjusted mean deceleration rate in the descending aorta (obtained with velocity-encoded phase-contrast MR imaging), as described by Nielsen et al (74). Another approach to assessing the severity of aortic arch narrowing is to measure the peak coarctation jet velocity on velocity-encoded phase-contrast MR images and estimate the pressure gradient by using the modified Bernoulli equation described earlier (75).

Conclusions
Cardiovascular MR imaging provides morphologic and functional information that may be decisive in the treatment of patients with conotruncal anomalies. In neonates and infants who are scheduled to undergo surgical repair, echocardiography usually suffices for preoperative planning. However, cardiovascular MR imaging may be used as an adjunct, to clarify details of the intracardiac and extracardiac anatomy that are not well depicted with echocardiography. Given recent advances in pediatric cardiovascular surgery, catheter-based interventional therapies, intensive care, and medical management, many children with complex congenital heart defects including conotruncal anomalies are surviving into adulthood. Most of these patients have residual hemodynamic abnormalities that require serial imaging examinations and, often, further intervention. The greatest clinical utility of cardiovascular MR imaging is in this older postoperative group, for whom it may be considered a first-line imaging technique. Familiarity with the surgical procedures and imaging appearances is essential for accurate preoperative and postoperative assessment.

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Cardiovascular MR Imaging of Conotruncal Anomalies

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Page 1070
The major conotruncal anomalies are tetralogy of Fallot (TOF), transposition of the great arteries, double-outlet right ventricle (DORV), truncus arteriosus, and interrupted aortic arch.

Page 1075
The primary goal of the cardiovascular MR imaging examination in patients with unrepaired TOF is to delineate the anatomy of the pulmonary vascular bed and identify sources of pulmonary blood flow, including aortopulmonary collateral vessels and ductus arteriosus.

Page 1077
The goals of cardiovascular MR imaging after surgical repair of TOF include quantitative assessment of the right and left ventricular volumes, stroke volume, and ejection fraction; anatomic evaluation of the RVOT, pulmonary arteries, aorta, and aortopulmonary collateral vessels; quantification of pulmonary regurgitation, tricuspid regurgitation, cardiac output, and pulmonary flow–to–systemic flow ratio; and identification of regions of myocardial scarring.

Page 1082
The goals of cardiovascular MR imaging after an atrial switch procedure include evaluations of the ventricles for size and function, pulmonary and systemic venous pathways for baffle leak and stenosis, atrioventricular valves for regurgitation, and outflow tracts for obstruction.

Page 1082
The main objectives of cardiovascular MR imaging after an arterial switch procedure include quantitative evaluation of the function of the right and left ventricles; evaluation of the outflow tracts for obstruction and valvular insufficiency; and evaluation of the great vessels, especially for stenotic lesions in the pulmonary arteries and dilatation of the aortic root.