Approximately 1% of newborn infants have congenital heart disease. With current advances in cardiac surgery, >85% of these infants are expected to reach adulthood. It is currently estimated that there are close to 1 million adults with congenital heart disease in the United States.1 Many or most of these patients will require lifelong medical care.

It is important to be able to recognize the appearance of congenital anomalies on computed tomography (CT), as they may be discovered incidentally, eg, when the patient comes to the emergency room with chest pain, arrhythmias, or other symptoms. In other cases, they may be detected on CT scans requested to evaluate postoperative anatomy or suspected complications.2,3

Scanning protocol

The scanning protocol we use to study congenital heart disease is similar to that for pulmonary embolus (Table 1). With our 16-slice scanner, we use a detector collimation of 0.75 mm, and with our 64-slice scanner, we use a detector collimation of 0.6 mm. Slice thickness is 1 mm, and pitch is 1 to 1.5. We image in either the cranial-caudal direction or the reverse, and reconstruct 3-dimensional (3D) images in 1- to 2-mm sections.

Contrast administration consists of 75 to 100 mL of nonionic contrast medium, 350 mgI/mL, injected at 3 to 4 mL/sec. We use bolus tracking to determine the optimal scan delay, defining a region of interest and triggering image acquisition when contrast enhancement reaches 100 to 120 HU.

The region of interest varies from one type of study to another. If we are imaging the aorta—eg, in a patient with surgical shunts or coarctation of the aorta—we trigger image acquisition off contrast enhancement in the ascending aorta. If we are interested in examining the pulmonary arteries, we trigger scanning off the main pulmonary artery or the right or left pulmonary artery. The approach is more complicated in the case of intracardiac shunts. If we suspect a right-to-left shunt, we trigger scanning off the right atrium, and if we suspect a left-to-right shunt, we trigger scanning off the left atrium.

When viewing images, we derive a great deal of information from the axial planes. We can see the valves and valve leaflets, the right and left atria, and the ventricles, and can evaluate the morphology of the intraventricular septum.

We also use postprocessing techniques, particularly when we are interested in examining the great vessels, outflow tracts, and the ventricular wall. We first create 2-dimensional (2D) multiplanar reconstructions, because they are easy to do at the scanner console and provide information very quickly. We also do more sophisticated reconstructions at the workstation, including 3-dimensional (3D) volume renderings and maximum-intensity projections.

Obstructive lesions

In patients with adult congenital heart disease, the clinical indications for CT fall into 2 general categories: Obstructive lesions, including coarctation of the aortic and valvular stenosis, and intracardiac malformations, including simple shunts and complex cyanotic heart disease.

The adult form of coarctation of the aorta occurs distal to the left subclavian artery (Figure 1). The diameter of the aortic arch is usually normal in adults (as compared with infants, in whom the arch is often hypoplastic). In adults, it is common to observe collateral vessels. Often they are unilateral, arise from the intercostal and internal mammary arteries, and are seen between the third and eighth ribs. Collateral vessels are uncommon in the infantile form of coarctation.

There are several ways to repair coarctation of the aorta—stenting, angioplasty, and patch aortoplasty, among others. The complications of coarctation repair include restenosis and pseudoaneurysm (Figure 2). Virtually all invasive techniques carry these risks.4

Valvular stenosis is another form of obstructive lesion. Aortic stenosis is usually associated with a bicuspid valve,
which is prone to degeneration and calcification. In patients with bicuspid aortic valve, calcification is common by age 30. This diagnosis is often made incidentally—eg, the patient comes to the emergency room with chest pain, and we observe a thickened, calcified bicuspid valve on CT. More sophisticated image reconstruction enables assessment of the degree of dilatation of the aortic root, a finding that plays a role in determining management.

Pulmonic stenosis is often identified on a chest radiograph. It is the most common form of right-sided obstruction and
usually results from fusion of the valve leaflets. This leads to a dome-shaped valve with a small opening. The valve is sometimes dysplastic with thickened leaflets but no fusion of the cusps. Most patients are asymptomatic. In severe cases, patients may have right heart failure. Typically, patients are then assessed with echocardiography and may go to cardiac catheterization before surgery.

Typical CT findings of valvular pulmonic stenosis are dilated main and left pulmonary arteries and a more normal caliber right pulmonary artery (Figure 3).

**Intracardiac shunts**

Intracardiac shunt lesions are more complex than obstructive lesions and include atrial septal and ventricular septal defects.

There are 3 types of atrial septal defects (ASDs), 2 of which are seen in adults and a third that is more common in neonates (Figure 4). Ostium secundum is the most common form of ASD. It occurs in the mid-septum and is usually isolated. Typically, the right and left pulmonary arteries are dilated. The patient may also present with hypertension.

Sinus venosus occurs high in the septum near the junction with the superior vena cava. It is often associated with partial anomalous pulmonary venous return. Ostium primum, which we typically see in the neonates, occurs in the low atrial septum near the atrioventricular valves. It is usually associated with a cleft in the mitral valve, which causes mitral regurgitation and sometimes a left-ventricle-to-right-atrium shunt.

There are 2 common methods of ASD repair. Traditionally, septal patch grafting has been used to isolate the right and left sides of the heart. This method is still used for large defects. A more recent alternative is percutaneous repair using a
catheter-mounted occluder. The Amplatzer Septal Occluder (AGA Medical Corp., Golden Valley, MN), for example, is composed of 2 nitinol wire discs, one larger than the other, with a neck in between. The larger disc is placed in the left atrium and the smaller disc in the right atrium. When the 2 discs are opened and sandwiched together, the ASD is blocked.

Ventricular septal defects (VSDs) usually occur in the muscular septum between the ventricles (Figure 5) or in the membranous part of the septum, in the subaortic or subpulmonic regions. Membranous VSDs usually occur in patients with more complex heart disease, such as tetralogy of Fallot.

**Postoperative complex heart disease**

Most patients with complex congenital heart disease who live to adulthood have undergone surgery, whether insertion of a palliative shunt or a complete repair. Therefore, although we do see primary disease in adults, we more typically see the results of surgical correction.  

Effective evaluation of congenital heart disease depends on knowing both the anatomy of these complex lesions and the clinical question that sends the patient to CT. It is important, therefore, that we collaborate closely with our clinical colleagues.
Figure 6 shows a young-adult patient with tetralogy of Fallot who received only a palliative shunt as a child and did not return as planned for definitive surgery. Because of cyanosis, the patient underwent CT imaging, which not only demonstrated the patency of the shunt, but also showed the underlying anatomic abnormalities, including pulmonary stenosis, narrowing of the outflow tract, right ventricular hypertrophy, and a VSD between the right and the left ventricles. A calcified aortic valve was detected incidentally.

Transposition of the great vessels causes the systemic blood to circulate to the right atrium and ventricle and out the aorta, never reaching the pulmonary circulation. Oxygenated blood circulates in parallel from the pulmonary veins to the left atrium and ventricle, and out the pulmonary artery. To survive, all patients must have surgery as neonates, initially to allow communication at the atrial level, which enables blood flow to the pulmonary system. Later, patients undergo definitive surgery.

In the past, definitive surgery consisted of an intracardiac atrial switch without change in the external anatomy. With a Mustard or Senning procedure, the aorta continues to come off the right ventricle, and the
pulmonary artery off the left ventricle. Systemic blood flows from the superior and inferior venae cavae into the right atrium, where a baffle shunts blood to the left ventricle and out the pulmonary artery. Oxygenated blood is then shunted to the right ventricle and out the aorta.

Now that patients with transposition of the great vessels are living longer, we are observing complications with the intracardiac shunt (Figure 7). Baffle leaks are characterized by back flow into the right heart and can result in cyanosis. Stenosis of the intracardiac shunt may be accompanied by development of large azygous collateral vessels to divert blood away from the site of stenosis.

For the last 5 to 10 years, the Jatene procedure has been used to correct transposition of the great vessels. This procedure consists of an extracardiac switch of the great vessels. The aorta is reconnected to the left ventricle, and the pulmonary artery to the right ventricle. The pulmonary artery is pulled up, and both the right and left pulmonary arteries straddle the aorta, positioning the aorta to the left of the pulmonary artery. To date, we have not observed long-term complications in patients who have undergone the Jatene procedure.

In levo-transposition of the great vessels, the aorta is to the left of the pulmonary artery, and the right and left ventricles are inverted. Levo-transposition may be discovered incidentally, when the patient presents to the emergency room with chest pain and undergoes a CT examination for suspected pulmonary embolism or aortic dissection.

Tricuspid atresia is characterized by a fatty bar between the right atrium and the right ventricle. The right atrium is enlarged and the right ventricle is small. The left heart is normal. These lesions are not reparable by intracardiac surgery; rather, an extracardiac technique is used.

The Fontan procedure was initially used to shunt blood from the right atrium directly to the pulmonary artery in patients with tricuspid atresia. The right atrium cannot accommodate a large volume of blood on a chronic basis, however, and many patients eventually went into heart failure. Arteriovenous malformations were another common complication. Today, the total caval pulmonary shunt is used to repair tricuspid atresia. In this approach,
the superior and inferior venae cavae are directly anastomosed to the pulmonary artery, and blood bypasses the right side of the heart.

Conclusion

CT evaluation of congenital heart disease in adults is becoming increasingly common, as we develop both better diagnostic methods and better treatment options. Today, >85% of neonates with congenital heart disease are expected to reach adulthood.

These cases are complicated, and they require knowledge of both anatomy and the clinical implications of imaging findings. Armed with this knowledge, we can diagnose congenital heart disease in adults and evaluate the long-term outcome after surgical repair.

REFERENCES


Discussion

ELLIOT K. FISHMAN, MD: Thanks very much, Marilyn. As you said, there’s so much more to cardiac imaging than coronary arteries—whether it is congenital heart disease in pediatric patients or congenital heart disease in adults that you do so much of, but also identifying disease in adult patients. Are there any specific questions for Marilyn?

STEPHAN ACHENBACH, MD: I have a question. One of the critical issues in many of those patients is the function of the right ventricle, especially with the transpositions after all the correction you do, that the right ventricle doesn’t bear the load, and the right ventricle functional assessment is very difficult by echo. You showed us only the morphologic imaging. Do you use the functional assessment also?

Marilyn J. SIEGEL, MD: We’re starting to do functional assessment. However, to date, most patients have been referred to us for evaluation of residual anatomic defects following palliative or complete surgical repair. Patients are referred by the cardiologist because of an equivocal echocardiogram. CT is done to obtain anatomic information. The necessary functional information can usually be acquired by echocardiography.

ACHENBACH: How do you read those studies? Do you bring in the pediatric cardiologists, the adult cardiologists, and the radiologists to read it all together?

SIEGEL: No. The adult cardiologist and the radiologist review the CT studies in adult patients, and the pediatric cardiologist and the radiologist review the studies in children.

ACHENBACH: You avoid the two and one situation: two cardiologists.

SIEGEL: We avoid the two cardiologist versus one radiologist scenario.

FISHMAN: Our experience has been that the pediatric cardiologists especially are becoming interested in CT now. For years, we didn’t hear much from them, but now we see more referrals. Whenever the echo is problematic or there’s some question, or if they’re going to redo a complicated case, where there’s worry about some graft or shunt being occluded or narrowed, we’re seeing much more interest in CT. We do 5 to 10 cases a week, and in the past we basically did none.

SIEGEL: I agree with Elliot. The cardiologists believe that they can get useful anatomic information about grafts or shunts using CT. CT is excellent at evaluating surgical shunts. It is faster than MR and provides the needed anatomic detail about graft occlusion or stenosis. Functional information is usually not an issue in these instances.

ACHENBACH: But I’ve noticed sometimes that the pediatric cardiologists are very disappointed when we have to tell them that we cannot provide flow, and, we cannot tell them anything about pressure because that’s what they get from the echo pressure gradients and those kinds of things. Sometimes they’re disappointed when we don’t get that information, which is very important to them.

SIEGEL: It depends on what information the cardiologist wants. You have to select the imaging study based on the clinical question. If the question is anatomy, CT usually suffices. If functional information is needed, MRI is probably a better study.

J. JEFFREY CARR, MD, MSCE: We’ve done a lot with cardiac MR in congenital heart disease, and we’ve started doing more adult congenital heart disease with our cardiac CT. Working with our cardiothoracic surgeons, one of the things I think they were most impressed with was not only showing the chamber anatomy, but many of these cases also have coronary artery anomalies. I don’t know what your experience is, but it really impresses the surgeons to be able to lay out the anomalous coronary anatomy for their surgical planning.

SIEGEL: You’re absolutely right, because many of these anomalies don’t occur in isolation. Other anomalies, particularly those involving the origin of the coronary arteries, are not uncommon. CT is an excellent way of showing the coronary anatomy for surgical planning.

MICHAEL POON, MD: For the adults, in general, would you recommend the contrast run at a slower rate without saline chase?

SIEGEL: The rate of contrast administration should be similar to that used for a routine PE study. It doesn’t need to be slower. We don’t use a saline chase.

POON: Because we do that for coronary, to wash out the right ventricles. In your case, you probably want the right ventricle.

SIEGEL: Yes, we want to see the right ventricle. In some cases, we need to see both sides of the heart. In these cases, we do dual-phase imaging. For instance, in patients with total cav pulmonary shunts, the anastomosis of the superior
vena cava to the pulmonary artery is seen well on arterial phase imaging. The inferior vena cava anastomosis is seen better on venous-phase imaging.

**FISHMAN:** One issue we found with the pediatric population, particularly with some of the shunts that are very slow-filling, is that if you go too early, you’re wondering, “Is this shunt occluded, or it’s just not opacified?” We have been doing a lot of dual-phase imaging.

**SIEGEL:** Yes.

**FISHMAN:** It seems there’s no way to really presume in advance what the right timing is. We’ve been going at something like the equivalent of 25 and 45, trying to get the best of both worlds.

**CARR:** We do the dual-phase routinely now. I use the low dose prospectively triggered for the delayed phase, although it doesn’t have the spatial resolution of helical, retrospectively gated cardiac protocol. But if you’re just looking at shunts or when different chambers are filling, you can get by with a really low-dose scan and do multiphase imaging. We sometimes do it precontrast, too, so we’ll do the low-dose, prospectively triggered scan up front; that way if there’s calcification or valvular disease, then we do more of an arterial phase, and then immediately do the delayed low-dose scan. If you think about that, that’s ≥8 mSv, so you’re right at the 10-mSv dose for the whole scan.

**SIEGEL:** We do the noncontrast scan in many instances, but not in all cases. The noncontrast scans help in identifying calcification. They also should be performed in patients with stents. In the latter population, the unenhanced scans help to identify contrast extravasation.

**RICHARD D. WHITE, MD:** Do you see MR being redefined by CT with congenital cases?

**SIEGEL:** Yes. I do see it being redefined. If anatomy is the clinical question, CT can replace MRI.