Contrast Use in Cardiac CTA Applications

CT evaluation of adult congenital heart disease: A practical approach

Cardiac CT is increasingly used to evaluate patients with chest pain. As the volume of studies grows, so does the likelihood of discovering clinically significant congenital heart disease in adult patients.

Chip Gilkeson, MD

Dr. Gilkeson is an Associate Professor of Radiology, Case Western Reserve University School of Medicine, and Section Chief, Cardiothoracic Imaging, University Hospitals of Cleveland, Cleveland, OH.

Nearly 1 million people in the United States have documented congenital heart disease, while another 100,000 to 400,000 people have an undiagnosed congenital abnormality. Traditionally, invasive angiography was used to evaluate patients with congenital heart disease. Today, physicians increasingly turn to noninvasive methods, such as echocardiography, magnetic resonance imaging (MRI), and, more recently, multislice computed tomography (CT).

The recognition of congenital heart disease on cardiac CT is becoming increasingly common, as the volume of such studies increases. A typical patient comes to the emergency room with chest pain or dyspnea and leaves with a diagnosis not of myocardial infarction, pulmonary embolism, or lung cancer, but, instead, of a previously undiagnosed congenital heart disease. Common congenital abnormalities visualized by cardiac CT include intracardiac shunts and anomalies of the aorta, pulmonary arteries, pulmonary veins, and coronary arteries. It is critical for those who interpret cardiac CT studies to be prepared to accurately recognize and report findings of congenital heart disease, not only for the purposes of diagnosis but also for treatment planning.

Role of CT

It is useful to compare the relative strengths and weakness of noninvasive imaging modalities in the evaluation of congenital heart disease. Echocardiography is the mainstay. Its strengths include an absence of radiation, the ability to evaluate cardiac structure and function, and the ability to perform hemodynamic assessment. Echocardiography is limited, however, in the evaluation of certain portions of the aorta (particularly the ascending aorta and the transverse arch), the distal pulmonary arteries, the right ventricle, and the pulmonary veins. These are relative “blind spots” for the echocardiographer.

MRI, unhindered by “blind spots,” is excellent for the depiction of cardiac anatomy. Like echocardiography, it eliminates radiation exposure and enables assessment of cardiac function and quantification of blood flow.

Multislice CT has important strengths in comparison with each of these imaging modalities. Unlike echocardiography, CT provides excellent anatomic information, ably identifying the aorta, right ventricle, pulmonary arteries, and pulmonary veins. Moreover, CT is faster to perform than echocardiography and, therefore, reduces sedation time in pediatric patients.

CT can be superior to MRI in important ways. First, it is readily available, which reduces delays in the scheduling of imaging examinations. A reduction in imaging time is another comparative advantage of CT, particularly in the sick patient. Significant tracheobronchial pathology is often associated in patients with congenital heart disease. Superior 3-dimensional evaluation of airway pathology with multislice CT is a significant additional advantage when compared with echocardiography or MRI.

Several CT protocols can be used with 16- and 64-slice scanners in the evaluation of congenital heart disease. In general, slice thickness is 1 to 2 mm. At our institution, contrast administration consists of 50 to 125 mL of ioversol 350 mgI/mL, injected at 4 mL/sec. A 50-mL saline flush follows the contrast injection when coronary artery pathology is of particular concern.

At University Hospitals of Cleveland, we use automatic bolus tracking to determine the optimal timing of image acquisition, a method that our technologists find helpful. We use retrospective electrocardiographic (ECG) gating under certain circumstances, based on consultation with the referring physician. For example, we always gate coronary CT angiographic (CTA) studies and, occasionally, examinations of aortic valve pathology. We do not usually gate studies of the pulmonary arteries or veins.

Aortic anomalies

The most common aortic anomalies include a double aortic arch, a left aortic...
arch with an aberrant right subclavian artery, a right aortic arch with an aberrant left subclavian artery, a coarctation of the aorta, and a bicuspid or dysplastic aortic valve.

Adults with an aberrant subclavian artery are at increased risk for intra-arterial plaque and aneurysm formation. Often, the patient presents with a large mediastinal mass, as shown in Figure 1. This 73-year-old patient was referred for evaluation of a heavily calcified mediastinal mass. On contrast-enhanced CT, the mass proved to be an aneurysmal dilatation of an aberrant right subclavian artery.

Such patients are also at increased risk for stroke. Figure 2 shows an 84-year-old woman who presented with hoarseness and dysphagia. A large mediastinal mass, which was initially suspected to be lung cancer, was found on CTA to be an extensively diseased and ulcerated aberrant right subclavian artery, complicated by a large mobile thrombus.

A double aortic arch is often observed in patients who undergo CT after coming to the emergency room with chest pain. It is particularly common in elderly patients with unusual mediastinal abnormalities, such as a widened mediastinum or a mediastinal adenopathy. In patients with a double aortic arch, the right arch is usually higher and dominant, and the left arch is smaller (Figure 3). It is important to distinguish such findings from other causes of mediastinal mass. For example, in a patient with pre-existing lung cancer, an incorrect diagnosis of mediastinal adenopathy would markedly affect disease staging and, potentially, the patient's therapeutic options.

CT nicely visualizes coarctation of the aorta, typically revealing both the aortic abnormality and the presence of collateral vessels and enlarged internal mammary arteries (Figure 4). CT is useful not only in the initial diagnosis of coarctation but also in the noninvasive follow-up of patients who have undergone surgical repair. It is important to recognize that while aortic coarctation classically presents in the young patient, it may...
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**FIGURE 3.** A double aortic arch in a 78-year-old woman who came to the emergency room with chest pain. CT shows (A) a right aortic arch that is larger than and superior to (B) the left aortic arch.

**FIGURE 4.** A 27-year-old man with chest pain and mediastinal widening. CT reveals a focal coarctation of the thoracic aorta (arrow) with dilatation of the thoracic aortic arch.

**FIGURE 5.** A quadricuspid aortic valve in a patient who presented with chest pain. CT also shows an enlarged aortic root and aortic insufficiency (arrow).
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unexpectedly occur in older patients with chest pain.

Congenital aortic valve disease is increasingly detected on ECG-gated multislice CTA. Information on aortic valve anatomy and function is always available on coronary CTA studies, and it is important to be able to recognize and report such incidental findings as bicuspid valve, dysplastic aortic valve, and other forms of congenital aortic valve disease, as they might otherwise go unrecognized.

Figure 5 shows an unusual case of quadricuspid aortic valve in a patient who presented with chest pain and an enlarged aortic root. Patients with this rare condition often have aortic insufficiency, as was shown on this study.

**Figure 6.** (A and B) Bicuspid aortic valve disease (arrows) in 2 men in their 40s.

**Figure 7.** A 77-year-old woman who came to the emergency room with chest pain and underwent CTA to rule out pulmonary embolism. (A) The chest X-ray shows lateralization of blood flow, with marked enlargement of the main and left pulmonary arteries. (B) CT reveals an enlarged left pulmonary artery (arrow) and a normal-sized right pulmonary artery, with (C) thickened pulmonic valve leaflets (arrow) that are consistent with congenital pulmonary valvular stenosis.

**Figure 8.** In a patient who presented with both a stroke and pulmonary embolism, CT reveals a large mobile thrombus situated in the patent foramen ovale (arrow).

**Figure 9.** A previously undiagnosed muscular ventricular septal defect (arrow) in a 46-year-old woman who underwent coronary CTA after developing chest pain.
Surgeons appreciate the imaging power of CT and its advanced postprocessing techniques for evaluation of aortic valve disease. Bicuspid aortic valve is the most common congenital heart lesion. With the enhanced imaging capabilities associated with ECG gating, bicuspid aortic valve is now clearly diagnosed with CT (Figure 6).

CT has the power to detect both supra-valvular and sub-valvular aortic stenosis, not only in children but also in previously undiagnosed adults who develop congestive heart failure, dyspnea, or other symptoms. Electrocardiographic gating also enables the visualization of rare congenital lesions, such as a subaortic web, which previously might have gone undetected.

Pulmonary artery anomalies

Pulmonary artery anomalies include pulmonary artery agenesis, pulmonary artery stenosis (including tetralogy of Fallot and Williams syndrome), and pulmonary valvular stenosis (including congenital valvular stenosis and postoperative stenosis). Pulmonary artery anomalies are routinely visualized on CTA, particularly on studies performed for the diagnosis of pulmonary embolism in patients with chest pain.

Figure 7 shows a 77-year-old woman who came to the emergency room with chest pain and underwent CTA to rule out pulmonary embolism. The chest X-ray showed lateralization of blood flow, with marked enlargement of the main and left pulmonary arteries. CT revealed an enlarged left pulmonary artery and a normal-sized right pulmonary artery, with congenital pulmonary valvular stenosis and thickening of the pulmonary valve. CT offers valuable information for surgical planning, particularly the relationship of the coronary arteries to the stenotic valve.

In patients with tetralogy of Fallot, CT readily depicts right ventricular outflow tract stenosis. It is also useful in the non-invasive follow-up of patients who have undergone surgical repair, confirming patency of a Blalock-Taussig shunt, for example, or revealing recurrent outflow tract stenosis. In the past, follow-up would have been accomplished through invasive angiography.

Atrioventricular septal defects

Atrioventricular septal defects include atrial septal defect (ASD), ventricular septal defect (VSD), patent ductus arteriosus, and partial anomalous venous defect.

CT often provides the first diagnosis of ASD, typically in a patient who comes to the emergency room with chest pain. CT often reveals communication between the right and left atria. In some cases, the addition of ECG gating further enhances visualization of these important interatrial communications. Figure 8 shows a patient who presented with both a stroke and pulmonary embolism. CT revealed a large mobile thrombus lodged within the patient’s patent foramen ovale.

CT is equally adept at visualizing ventricular septal defects. Figure 9 shows a previously undiagnosed muscular VSD in a patient with chest pain who underwent coronary CTA.

Partial anomalous pulmonary venous return

Partial anomalous pulmonary venous return is a challenging diagnosis to make, as this congenital anomaly presents in several different ways. The anomalous venous return may be to the right atrium, the superior vena cava, or brachiocephalic vein, for example. The clinical presentation includes right ventricular dilatation and, in some cases, an associated sinus venosus ASD. Echocardiography can be limited in visualization of the drainage patterns of the pulmonary veins, while multislice CT imaging clearly defines pulmonary venous anatomy and can play an important role in the evaluation of the patient with an enlarged right heart.

Figure 10 shows a patient who was evaluated by echocardiography for right ventricular hypertrophy. CT confirmed enlargement of the right heart. Volume-rendered images reveal the partial anomalous pulmonary venous drainage from the right upper lobe veins to the brachiocephalic vein.
Coronary artery anomalies

The subject of coronary artery anomalies is too vast to be discussed in detail here. It is important to note, however, that CT is often performed to evaluate coronary artery anomalies in children and teenagers with syncope and chest pain, particularly athletes. It is important in this population to pay very close attention to radiation dose and to use dose-modulation software.

Conclusion

A large number of congenital heart defects go undiagnosed during early childhood. Multislice CT is playing an increasingly important role in identifying and characterizing congenital heart disease in adults, often revealing abnormalities on contrast-enhanced CTA after the patient comes to the emergency room with chest pain. It is important for those who interpret cardiothoracic CT studies to be able to recognize and report on significant congenital heart defects in adults.

References


Discussion

ELLIOT K. FISHMAN, MD: Thanks very much, Chip, for a great talk. I have a couple of questions. In a number of the cases, you showed surprise findings, mediastinal mass, etc., but in other cases, I know your referred cardiologic CT studies were looking for congenital heart disease. Do you typically gate those patients? How do you do those studies from a protocol perspective?

CHIP GILKESON, MD: I think the gating part is certainly important, and the decision about gating is certainly in consultation with the referring surgeons and cardiologists. What is the other information that you need? If there are any issues about any kind of anomalies, any relationship of the coronary arteries to the defects, are there any anomalous coronaries? Certainly, we gate those. Pulmonary artery evaluations we don’t gate. Pulmonary venous anomalies we usually don’t gate. Occasionally, some bicuspid and some of the aortic valve pathology we’ll gate. But, we do not routinely do it just for radiation issues. It’s a much more tailored approach to the question. It is really important to answer.

FISHMAN: You don’t really worry about heart rate either?

GILKESON: With these patients, certainly; for heart rate with non-gated, certainly not. When we do gate, we certainly try to get the heart rate down to <65 bpm if we’re going to do gated studies.

STEPHAN ACHENBACH, MD: When you showed some of these cases coming from the emergency room, it seemed that you had a pretty good filling of the right and of the left heart. So was there some suspicion maybe—a cardiac murmur or something—that told you to prolong the injection of contrast? My second question is if you had a specific goal, for instance if you know you’re looking for an ASD, what do you do as far as contrast is concerned? Is it necessary for an ASD to have contrast both in the left and in the right?

GILKESON: Right. There are certainly some contrast issues. If there is really complex congenital heart disease, since the right ventricle is a very important ventricle, we always make sure that is still optimally opacified. We don’t do a saline flush in those patients, for instance, to clear the right heart. We usually also take out the injection time and often the volumes are a little bit higher. Routinely, 60% to 75% of these patients get up to 100 mL to make sure their right ventricle is increasingly opacified. For ASDs, ASD can still be a hard diagnosis. Talking about contrast, there may be some opportunities. There is certainly MR literature now looking at first-pass MR contrast for the evaluation of ASD. We’ve had a couple of patients in whom there’s a question about a small ASD. If you do a small, very limited scan through the areas of suspected ASD, you do a Valsalva technique, and inject a low dose of contrast. You often can at least diagnose the right heart from the ASD by looking at opacified contrast going from the right heart to the left heart in those patients.

ACHENBACH: Of course, you cannot rely on actually seeing the septum, even in a normal case. You might not see the septum, so it’s hard to diagnose with confidence the absence of the septum. So I think it is difficult.

GILKESON: Those were examples of obviously gross ASDs. What’s interesting, though, is that all those examples were previously undiagnosed. There are a lot of people out there in whom the pulmonary hypertension and shortness of breath leads to their first presentation of a very large ASD that had been missed.

ACHENBACH: You showed a case with the small ventricular septal defect coming from the ER. That would be something that you hear in a patient; a small VSD typically causes a loud noise. It’s surprising that this was not suspected.

GILKESON: But it was not.

SAMUEL WANN, MD, MACC: Those are spectacular pictures and it was a very nice presentation. For which of those anomalies would you consider CT to be the first diagnostic test of choice, as opposed to echo, MR, or an EKG, for that matter? Almost always, ASDs have an abnormal electrocardiogram. It’s not very specific, but it’s almost always abnormal. When would you recommend that your referring doctors first do a CTA for congenital heart disease? Do you do it as the first test off the block?

GILKESON: Certainly, if there is a suspicion of congenital heart disease, an echocardiogram is always the first test off the block. It’s really an important evaluation of function. Certainly, their sensitivity for small shunts with echo and Doppler is always going to be better. There’s dynamic information that’s certainly valuable first. Those cases I showed were some of the unsuspected patients we received from the ER. But, for example, with partial anomalous veins, those are patients for whom the echo is done first by cardiologist and are very, very complete. If there is a specific question, it can be “I’m not seeing certain things. I’m seeing a right heart that’s too big, but I can’t explain that. You know,
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I’m not quite seeing the rest of those veins. I’m seeing some turbulence up in the ascending aorta. Do you see any kind of anomaly or any kind of stenosis?” So, CT is not usually the first test off the block. I think usually that most of the CT studies come after an evaluation with a cardiologist with echocardiograms.

WANN: When would you recommend a CTA as opposed to an MRA in the same scenario? Or is CT better?

GILKESON: In each situation, I think certainly with coronary anomalies, you really have to get the full course of coronary arteries and, for that, the CT is better than the MRA. For most of the pulmonary artery issues, it’s just a matter of sedation times, availability, and if they need an answer now. Often, that triggers a CT; many times it is a convenience issue. Certainly there’s still not quite the dynamic function of MR. Certainly in a lot of these patients with congenital heart disease, there are issues of mental retardation, and there are issues about the safety of sedation with MR. Often because of those issues, we go to CT.

ACHENBACH: You showed several very impressive cases coming from the emergency room. The question, of course, is, do you have a patient who has chest pain, and then you find an ASD? For example, I think you showed 2 cases. Well, the ASD probably will not explain the chest pain, so is something else going on with the patient?

GILKESON: You’re absolutely right. Again, as we see more and more patients through the ER now, where chest pain is often the question. CT is such a common exam right now that often, for a lot of the symptoms, sometimes CTs are questionable as a screening exam, and the chest pain may be musculoskeletal, it may be pleuritic. But if you’re going to start to screen your patients for a lot of those things, you’re going to start to diagnose unusual conditions. I think you’re right, in that patient the ASD is not the cause of the acute chest pain, but it is important to recognize it.

MATTHEW BUDOFF, MD: From our perspective, we do a lot of congenital studies, as well, although probably not as many as Chip. But we get a lot of borderline or questionable echoes, and they get referred straight to CT. If you’re thinking about temporal resolution or slice thickness, CT has an advantage, so for things that are moving and small, CT will be better than MR. For larger structures that are moving, MR would be better because you get the functional data as well, so there’s definitely advantages to both. Ask Chip about balancing in a young patient or in a child. There are radiation issues of CT versus the intubation issues of MR. I think they’re both significant in an infant, or in young child in whom radiation might be really an issue. Do you choose, or is it local expertise? How do you do that?

GILKESON: It’s really with very close consultation about what other risks there are for sedation and intubation. Another evaluation that we didn’t talk about, but that is really important for our docs, is the evaluation of the airway for a lot of these anomalies. Certainly, in that situation, CT is definitely the superior modality. To look at the airway, we have a lot of kids who come in with congenital heart disease with stridor or wheeze. How do we best evaluate both the airway pathology and the vascular pathology? CT wins out. Often in those cases (which used to be diagnosed as diagnostic bronchoscopy that didn’t show anything), virtual CT bronchoscopy can avoid a lot of those issues. I think that the airway is also an issue and is, again, another advantage to CT.

FISHMAN: That’s our experience too. You basically go for the pediatric cardiologist, starting with echo, and often ending with echo. If it needs to go further, because of the sedation issues, and because of the spatial resolution of CT, and, as you say, because you kill many birds with one stone, we routinely use the virtual bronchoscopies and the vascular mapping. So for patients with prior surgery and patients in whom we need to evaluate vascular anomalies, we’re routinely doing CTs with 2-second scans, as opposed to sedation. It just makes all the difference in the world.

BUDOFF: We haven’t done a pediatric cardiac cath in a long time now because there are not issues of “is there a 30% or 50% stenosis in the coronary,” so the pediatric cardiac surgeons often get enough answers either from echo alone or echo plus CT. So they stop at that point, and we’ve been doing a lot less cardiac caths in our pediatric population than we used to, to make that definitive diagnosis.

GILKESON: You’re right. Clearly, every field has significant points of anxiety—chest pain in the ER is a point of anxiety. For the cardiologists, chest pain in the athlete is the point of anxiety and is really significant. So, is there any concern for an anomaly? For a significant bridge, they go to CT.