Advanced Cardiac Imaging for Complex Adult Congenital Heart Diseases

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ABSTRACT: The population of patients with adult congenital heart disease has grown and is currently estimated to include approximately 1 million people in the United States. Cardiologists and imagers frequently encounter complex patients who have undergone multiple prior operations and interventions. A myriad of imaging tests are currently available, including echocardiography, cardiovascular magnetic resonance imaging, and computed tomography, all of which collectively provide invaluable information on cardiac anatomy and hemodynamics. Advanced imaging plays a role in diagnosis and preprocedural planning and also determines the need and frequency of follow-up. This article provides a contemporary review of the current role of cardiac imaging in patients with complex congenital heart disease.

INTRODUCTION

Adult congenital heart disease (ACHD) is the most common birth defect, with an incidence of 1%.[1] Cardiac imaging is a fundamental tool in evaluating and following patients with ACHD. With a growing number of patients diagnosed with ACHD, cardiologists and imagers frequently encounter complex patients with multiple prior operations and interventions. In this regard, the images obtained from echocardiography, cardiovascular magnetic resonance imaging, and computed tomography are invaluable, providing critical information on cardiac anatomy and hemodynamics. In addition, imaging remains an essential tool to identify late complications of complex surgical treatments that are typically performed in the early years of life and to plan interventions. A myriad of imaging applications, ranging from high-resolution volumetric analysis in complex geometries to 3-dimensional (3D) reconstruction of the vasculature, has made cardiac magnetic resonance imaging the cornerstone of ACHD evaluation. This high demand has created a significant need for specialists in highly advanced cardiac imaging centers who understand the strengths, limitations, and pitfalls of each cardiac imaging modality. It is important to choose the proper diagnostic test that can address the clinical question and adapt the technology to the patient's needs. In this review, we focus on the burgeoning role of commonly used advanced imaging modalities and discuss the pros and cons of each for evaluating patients with ACHD.

IMAGING MODALITIES FOR EVALUATING ADULT CONGENITAL HEART DISEASE

Interpreting cardiac imaging studies of patients with ACHD can be challenging when the anatomical diagnosis is incomplete (ie, due to unknown prior surgical repair). In these cases, a segmental approach called the Van Praagh classification system is widely used to facilitate communication among specialists involved in the patient's care.[2] This system involves three steps: establishing the site of the viscera and atria, defining the morphological features of both ventricles and atrioventricular connections, and identifying the vascular status by the position of the aorta and pulmonary artery. The information obtained from this anatomical imaging data provides the basis for study analysis and interpretation.

ECHOCARDIOGRAPHY

With its wide availability and low cost, echocardiography is usually the first-line imaging modality in diagnosing ACHD and is a mandatory step prior to undertaking advanced imaging.[3] By far, echocardiography provides the highest temporal resolution (~ 10 msec) among all other noninvasive imaging modalities. Furthermore, the use of color Doppler allows assessment of intracardiac blood flow and direction, thereby facilitating estimation of transvalvular gradients. However, this modality is heavily dependent on operator experience and can have very limited acoustic windows in patients with a high body mass index or obstructive pulmonary disease. Also, assessment of adaptive responses and detection of long-term complications require cross-sectional imaging, and echocardiographic assessment of right ventricular (RV) function—which is uniquely at risk in ACHD patients—has shown poor correlation to cardiac magnetic resonance (CMR). By correcting for geometric distortions, 3D echocardiography overcomes some of these limitations despite the fact that it has been shown to underestimate ventricular volumes compared with CMR.[1]

Recent advances in echocardiography with the addition of speckle tracking and strain analysis have produced fewer

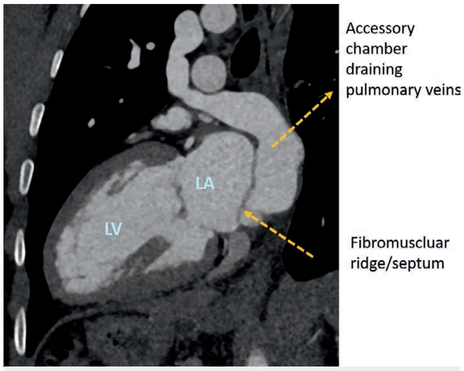

Figure 1.
Two-chamber sagittal view on cardiac computed tomography showing septal ridge separating the left atrium into two chambers, one draining the pulmonary veins and the other one connecting to the mitral inflow, consistent with Cor triatriatum sinister.

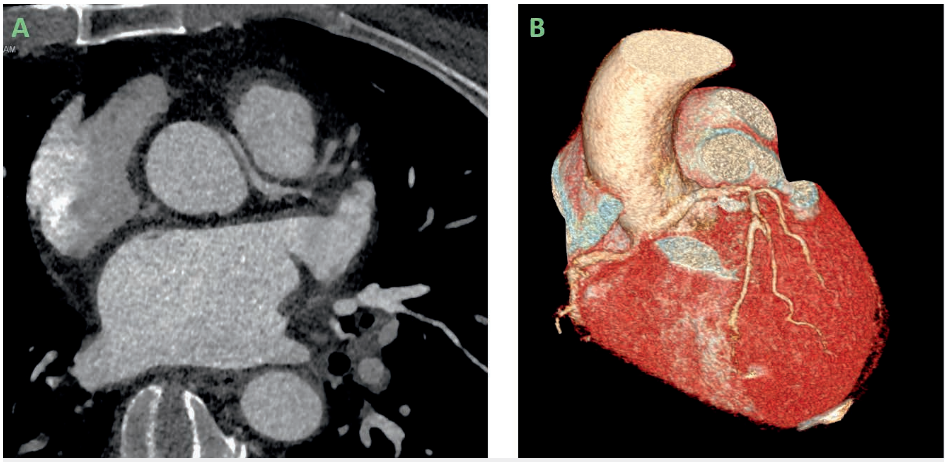

Figure 2.
(A) Cardiac computed tomography axial image and (B) volume-rendered 3-dimensional image showing anomalous origin of left main from the right coronary cusp. There is no evidence of malignant features including lack of intera-arterial or intramural course.

load-dependent indices for contractility. However, there currently is no standardization in imaging acquisition, and normal values are lacking.[4]

CARDIAC COMPUTED TOMOGRAPHY

In the past several years, cardiac computed tomography (CCT) has become highly regarded as a fast, reliable anatomic imaging tool for simple and complex ACHD cases. It provides excellent 3D anatomical imaging of the cardiac chambers and vascular structures with very high contrast resolution (Figure 1).[5] CCT has the highest spatial resolution (~ 0.5 mm) for assessing coronary artery anomalies (Figure 2) and is also a reliable alternative to invasive coronary angiography for assessing coronary anatomy in patients at risk for ostial stenosis after coronary reimplantation procedures (ie, Ross and Jatene procedures). In addition, CCT is the ideal test for detailed vascular analysis of collateral circulation (Figure 3), pulmonary venous anatomy, and major aortopulmonary collateral arteries.[4] It can provide excellent intracardiac anatomical evaluation of valvular defects, detect atrial or ventricular septal defects (Figure 4), and determine atrioventricular and ventriculoarterial connections (Figure 5). In patients who have contraindications for CMR, CCT can provide a reliable diagnostic alternative for assessing ventricular size and function in a single breath-hold examination and faster data segmentation.

One drawback of routine CCT use is the need for iodinated contrast media and the cumulative radiation dose in patients undergoing serial imaging, which in epidemiological studies is associated with an increased relative risk of cancer.[5] For young patients, this still remains a significant concern.[6] As a result, CCT has become a second-line anatomical testing tool after CMR for patients with ACHD. In addition, diagnostic motion-free imaging requires electrocardiographic gating and regular heart rhythm; hence,

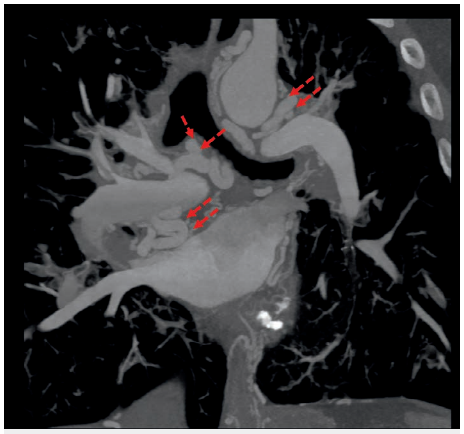

Figure 3.
Cardiac computed tomography, coronal image showing extensive mediastinal and peribronchial collateral circulation in a patient with coronary artery to pulmonary artery fistula.

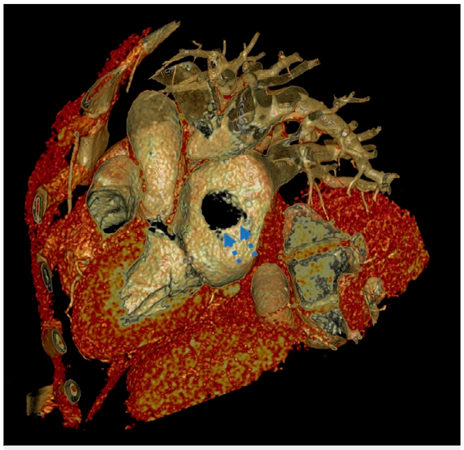

Figure 4.
Volume-rendered imaging with cardiac computed tomography and 3-dimensional depiction of secundum atrial septal defect (arrows).

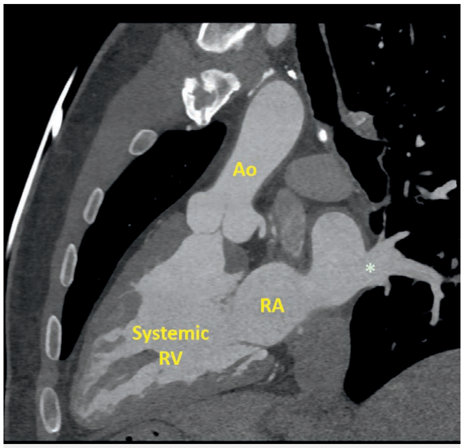

Figure 5.
Three-chamber sagittal view on cardiac computed tomography showing a patient with D-transposition of the great arteries after arterial switch surgery. The morphologic right ventricle is connected to the aorta, hence it is the systemic ventricle. The pulmonary veins were baffled to the right atrium (asterisk).

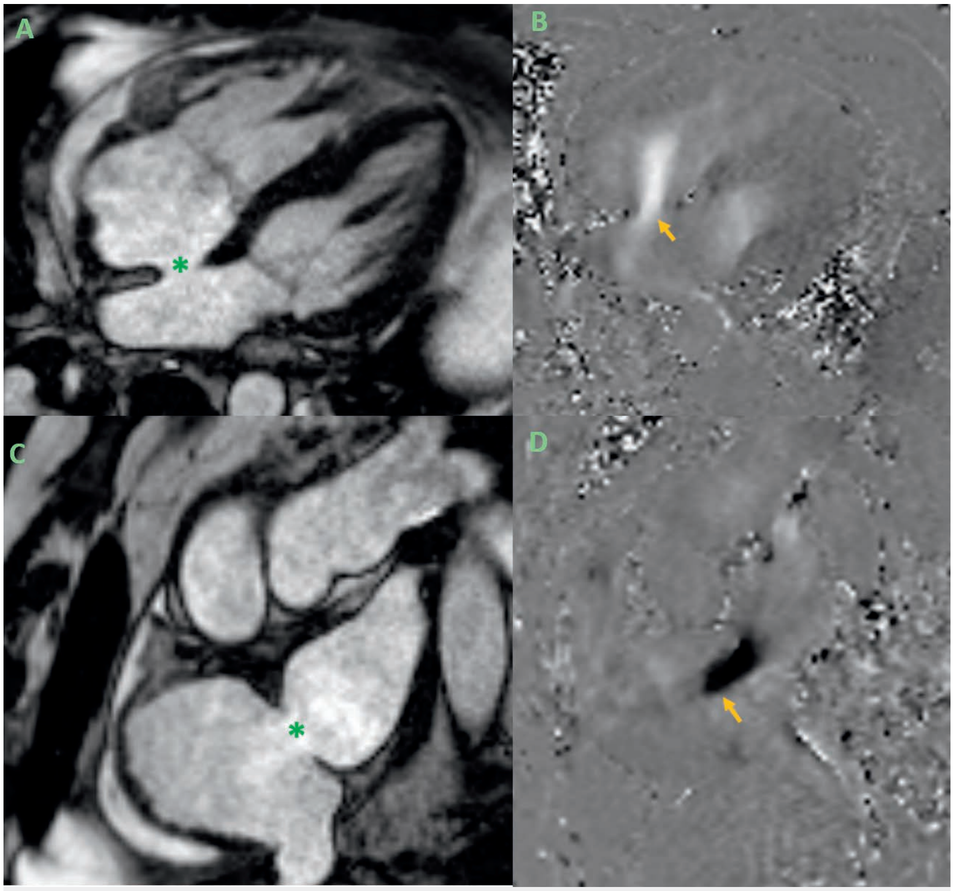

Figure 6.
(A, B) Steady-state free precession (SSFP) cardiac magnetic resonance imaging (CMR) sequence, 4-chamber view, showing large secundum atrial septal defect (ASD, asterisk, 2.0 × 1.5 cm). (B) Corresponding phase-contrast imaging at same plane showing left-to-right shunting across the defect. (C, D) SSFP CMR sequence, sagittal view, again showing ASD with corresponding phase-contrast imaging at same plane. (D) There was a dilated right ventricle (RV), with an RV end-diastolic volume of 261 mL, and normal RV systolic function, with an RV ejection fraction of 52%. The net Qp:Qs in this case was 1.5:1.

patients with arrhythmias or atrial fibrillation can produce a degraded imaging quality. However, with the new-generation scanners such as dual-source CT and large volume coverage scanners, the whole heart can be imaged in 1 to 2 seconds, or within a single beat, with a very low (~ 1 millisievert) effective radiation dose.[7]

CARDIAC MAGNETIC RESONANCE IMAGING

CMR has had a long history of being the one-stop technique for comprehensive evaluation of patients with ACHD. As a highly reproducible, radiation-free cardiac imaging tool that offers an unrestricted field of view, CMR is the cornerstone for volumetric assessment, biventricular functional evaluation, and detailed anatomical data for complex pathologies.[6,7] Despite the long acquisition time and lack of hemodynamics, the 2018 American Heart Association/American College of Cardiology (AHA/ACC) Guideline for the Management of Adults With Congenital Heart Disease recommends CMR as the preferred follow-up technique for patients with ACHD who need serial imaging studies—for example, patients at risk for right ventricular enlargement and dysfunction (class I level of evidence B).[8-10]

Cine Functional Data

Using steady-state free precession (SSFP) cine CMR (the power horse of cardiac imaging), short-axis bright blood images covering both ventricles are acquired at certain slice thickness (6-8 mm); this allows accurate quantification of volumes, mass, and biventricular ejection fraction irrespective of the geometry. CMR has the limitless option of prescribing imaging planes in any direction and is crucial for the segmental approach when the anatomy is unknown.

Phase-Contrast Imaging

Phase-contrast CMR imaging acquires short-axis planes of vessels and allows flow to be calculated as a product of area and pixel velocity.[1] This is currently the main tool to quantify pulmonary and

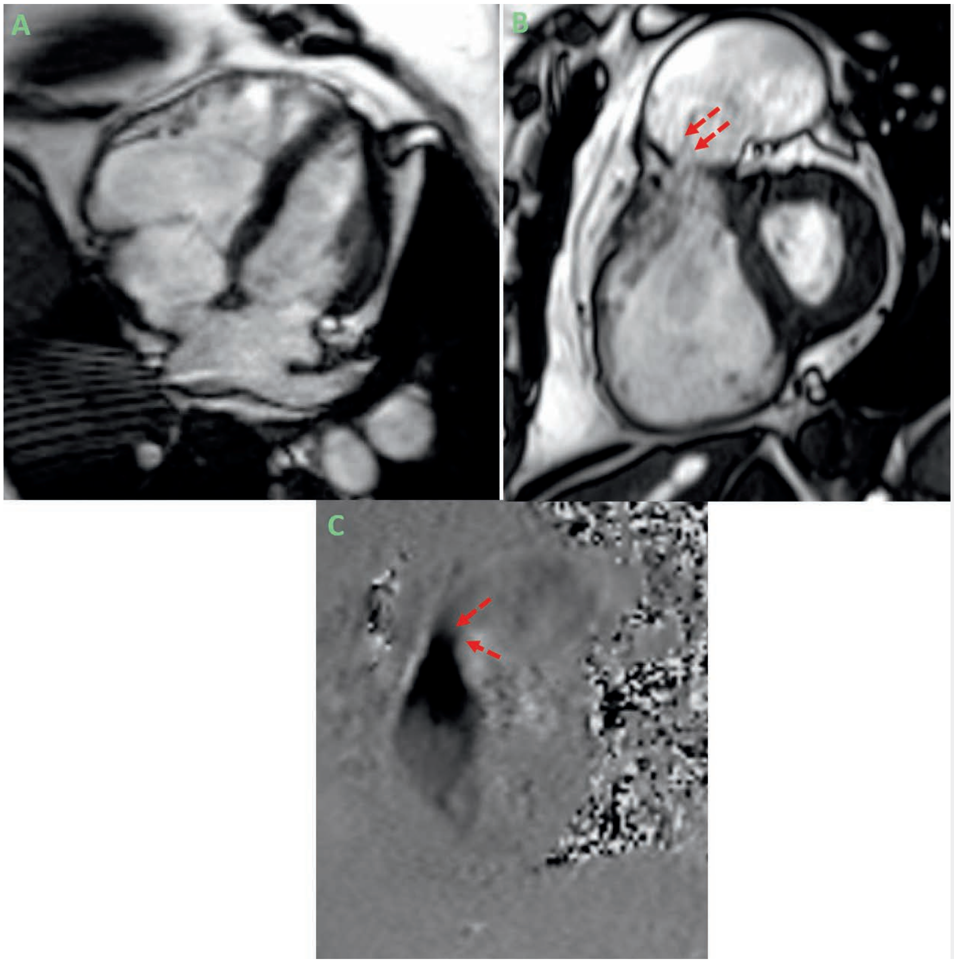

Figure 7.
A patient with repaired tetralogy of Fallot/pulmonic stenosis with pulmonic valvotomy and transannular patch presented with worsening shortness of breath. (A) Cine steady-state free precession (SSFP) computed magnetic resonance imaging (CMR) sequence 4-chamber view showing severe right ventricular (RV) enlargement (RV end diastolic volume of 165 mL/m²). (B) Cine SSFP CMR sequence sagittal oblique view at the pulmonic valve level showing transannular patch. (C) Phase-contrast CMR imaging at the same location as (B) in diastole showing pulmonic regurgitation (quantitatively, the regurgitant volume was 132 mL and regurgitant fraction was 78%).

aortic forward flow and intracardiac shunts (Figure 6) and detect valvular regurgitation. Pulmonic valve regurgitation is a common residual lesion in patients with repaired tetralogy of Fallot. According to the new AHA/ACC guidelines,[10] patients with symptomatic moderate or greater pulmonary regurgitation or evidence of RV dilation (RV end-diastolic volume index ≥ 160 mL/m² or RV end-systolic volume index ≥ 80 mL/m²) and asymptomatic patients with mild or moderate RV or LV systolic dysfunction and moderate or severe pulmonic regurgitation meet guideline recommendations for pulmonic valve replacement surgery (Figure 7).

Scar Imaging

The use of intravenous gadolinium contrast has permitted tissue characterization at the myocardial level (ie, fibrosis, inflammation).[11] The presence of late gadolinium enhancement of the right ventricular insertion points is commonly seen in patients with congenital heart disease; it is a consequence of chronic RV pressure and volume overload and has been adversely related to RV function.[4]

THREE-DIMENSIONAL CONTRAST-ENHANCED ANGIOGRAPHY

A special sequential CMR using electrocardiographic gating and a respiratory navigator to mitigate motion artifacts can provide whole-heart 3D imaging for vascular anatomy. These provide higher signal-to-noise ratio and near isotropic voxel resolution for 3D multiplanar reformatting assessment (Figure 8).[12] Another advantage of vascular magnetic resonance imaging is the ability to assess arterial and venous structures simultaneously, regardless of the use of a contrast agent, in a relatively short acquisition time (less than 30 seconds). Recent advances in technology have permitted faster data acquisition using acceleration methods such as parallel imaging, with a whole 3D data set obtained in less than 5 minutes.[13] In a study using time-resolved angiography, Kozerke and Tsao assessed pulmonary perfusion by inspecting the peak signal enhancement and calculated cardiopulmonary transit time, which showed a strong association with pulmonary vascular resistance in patients with pulmonary hypertension.[13]

Finally, a novel time-resolved 4D flow CMR is able to capture flow data from

Figure 8.
Sagittal view, 3-dimensional magnetic resonance angiography of the aorta showing severe coarctation (A, arrows) with post-stenotic dilation of the descending aorta. In addition, there is extensive collateral circulation in the chest originating from the descending thoracic aorta (B, arrows).

both intra- and extracardiac structures as a single 3D volumetric acquisition, and flow can be quantified using any desired plane and region of interest.[14] The downside to widespread adoption of this technology is related to long reconstruction times and the need for specialized software.

ALTERNATIVE MODALITIES

CMR imaging is prone to artifacts in patients with metallic objects, mechanical valves, baffles, and conduits, and this can inherently alter the image quality. Hence, CCT can be an alternative for better visualization of the above-mentioned anatomic structures.[8] Limitations for use of CMR include relatively higher cost, long acquisition times, restricted access, and relative contraindications in patients with cardiac implantable electronic devices or claustrophobia. In addition, unlike CCT, where images are obtained in axial stacks and then reconstructed, CMR relies heavily on operator experience, and a deep understanding of the anatomy and physiology are needed for proper imaging plane acquisition.[14]

In summary, the wealth of data obtained from advanced cardiac imaging requires detailed understanding of the strengths and limitations of each technique along with in-depth knowledge of complications that arise from surgical repair. With a growing number of patients with ACHD, a multidisciplinary approach is key in coordinating care of this unique population.

Conflict of Interest Disclosure:
The authors have completed and submitted the *Methodist DeBakey Cardiovascular Journal* Conflict of Interest Statement and none were reported.

KEY POINTS

- Patients with adult congenital heart disease need lifelong clinical follow-up typically involving serial multimodality imaging, which is helpful to monitor for late complications.
- There is a need to integrate advanced cardiac imaging modalities—such as computed tomography and cardiac resonance magnetic imaging (MRI)—into a diagnostic workup to address clinical questions.
- Cardiac MRI is the first-line choice in the clinical workup when diagnostic information from transthoracic echocardiography is insufficient.
- Careful consideration related to the patient's medical conditions (arrhythmia, claustrophobia, and renal status), age, compliance, and radiation exposure in addition to operator availability and expertise will alter the choice of imaging modality used.

Keywords:
adult congenital heart disease, cardiac computed tomography, cardiac magnetic resonance imaging, three-dimensional contrast-enhanced angiography

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