

Cardiac CT in the Diagnosis and Postoperative Assessment of Congenital Heart Disease

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Abstract Patients with complex congenital heart disease require serial diagnostic evaluation throughout their lives. Although echocardiography and cardiac MRI are the primary modes of evaluation in the current era, cardiac computed tomography (CT) plays an increasingly important role for specific indications. The high temporal and spatial resolution of the most modern CT scanners used for cardiac imaging allows for rapid, high quality image acquisition. The decreased need for sedation and anesthesia combined with recent developments markedly reducing radiation exposure make it an ideal imaging modality for certain indications in congenital heart disease. This article will briefly review the current use of noninvasive diagnostics in congenital heart disease, focusing on the use of CT for the most common congenital heart lesions referred for surgical intervention. When used appropriately, CT angiography (CTA) can provide critical information necessary to care for patients with congenital heart disease who require evaluation in addition to echocardiography.

Keywords Congenital heart disease · Pediatrics · Advanced imaging · Cardiac CT · Diagnosis · Postoperative · Preoperative

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Introduction

Congenital heart disease is the most common congenital anomaly. There have been tremendous advances in surgical and catheter-based intervention since the first ductal ligation in 1937, successful shunt for cyanotic heart disease in 1945, and intra-cardiac repair of an atrial septal defect in 1952. The surgical survival for all forms of congenital heart disease is approximately 95 % in the current era [1]. The majority of patients with even the most complex congenital heart disease now survive to adulthood, and the median age of this population is continually rising [2]. Most patients with congenital heart disease require serial evaluation of their cardiac status throughout their lives. The combination of a growing population of patients with palliated congenital heart disease and a current and expected future shortage of pediatric subspecialists requires that radiologists and adult cardiac imagers develop a proficiency in imaging and understanding of this patient population.

Use of Imaging Modalities in Congenital Heart Disease

When palliative surgical procedures became common in the 1950s and 1960s, cardiac catheterization was the only available diagnostic modality. Catheterization requires central vascular access and sedation or anesthesia for all cases and there is relatively high contrast loads and radiation exposure [3]. It is difficult to opacify different structures simultaneously and invasive angiograms provide 2-dimensional imaging planes, often requiring multiple different injections to properly outline the object of interest. Since the introduction of newer imaging modalities, catheterization is primarily used for intervention, or when pressure measurements are required for clinical decision-making.

Echocardiography has been in routine use since the 1980's and remains the most commonly used imaging modality for

congenital heart disease due to ease of use, wide availability, and excellent diagnostic accuracy for most indications when used by experienced providers [4]. In our practice, echocardiography accounts for 96 % of diagnostic studies. Limitations of echocardiography include degradation of acoustic windows with surgical scarring and obesity, poor vascular-airway definition, poor definition of extra cardiac structures, and poor visualization of distal pulmonary arterial and collateral vessel anatomy. In addition, there is poor reproducibility of right and single ventricular function, poor reproducibility of valvular regurgitation, and poor quantification of intracardiac shunting [5]. When imaging is required after echocardiography, a thorough evaluation of the risk vs benefit of each modality for the specific indication will determine the best test for the patient. For MRI and CT, the anesthesia risk of MRI for very young or critically ill patients and the radiation risk of CT are the most important considerations. In addition, the specific use of an advanced cardiac diagnostic test will depend on the availability of both the technology and the skilled personnel at each institution. As advanced CT technology becomes widely available, cardiac CT will play an increasingly important role in the diagnosis and management of congenital heart disease.

Cardiac MRI overcomes many of the limitations of echocardiography. Because of the lack of ionizing radiation, MRI is the modality first considered, and used most commonly, when imaging is required after echocardiography. It allows reproducible quantification of single and right ventricular systolic function, valvular regurgitant volumes and shunt fraction, and excellent definition of systemic and pulmonary venous and arterial anatomy [6]. Three-dimensional noncontrast sequences based on tissue differentiation as well as gadolinium based angiography provide 3D visualization of complex cardiac structures [7]. Limitations of cardiac MRI include incompatibility with pacemakers and defibrillators, artifact from certain types of metallic coils and implants, requirement for breath holding for most image sequences, and long imaging times that necessitate anesthesia for most patients less than 8 years of age. Most complete MRI scans require 45–70 minutes of scanning [8, 9]. For patients who are critically ill on ventilator support in the ICU, transition to MRI compatible anesthesia equipment is required. MRI with anesthesia has a relatively high relative risk for an adverse event [10, 11]. MRI is also technically challenging in the very smallest patients with the highest heart rates due to limitations in spatial resolution, temporal resolution, and poor signal to noise and contrast to noise ratios.

Cardiac CT has become increasingly used in the assessment of cardiac disease in since the introduction of multidetector CT technology. Recent technical advances in

CT technology have resulted in submillimeter spatial resolution and temporal resolution as low as 75 msec. Image acquisition is as fast as a single heart beat for volumetric scanners, is approximately 0.25 seconds for high pitch scanners to cover the typical scan range of a pediatric thorax, or is a single breath hold for ECG gated functional imaging. These advances combined with radiation dose reduction techniques make cardiac CT an excellent modality for certain congenital cardiac indications [12, 13].

Cardiac CT is particularly useful for:

- (1) Determination of valvular regurgitation, shunt fraction, or estimation of ventricular function in patients with complex congenital heart disease who are contraindicated for Cardiac MRI, or who are unlikely to have a diagnostic MRI due to artifacts.
- (2) Evaluation of complex cardiac anatomy in very young patients who are high risk for adverse event with anesthesia and who could undergo a CT scan with no or limited sedation or anesthesia.
- (3) Critically ill patients who are less likely to tolerate the long imaging times required for cardiac MRI.
- (4) Coronary artery imaging for evaluation of anomaly, Kawasaki aneurysm, or after surgical reimplantation, or surgical manipulation (s/p Ross procedure, Nikaidoh procedure, arterial switch operation).
- (5) Concurrent evaluation of cardiac and extra cardiac structures including airway anatomy and lung parenchyma, or evaluation of sternal anatomy prior to repeat surgical intervention.
- (6) Highly complex congenital heart disease where a high resolution 3D or 4D dataset aids in surgical planning, particularly when there is abnormal cardiac situs combined with pulmonary venous anomalies, systemic venous anomalies, and abnormal relationship of the atrioventricular valves to the outflow tracts.
- (7) Evaluation of prosthetic valve function or of perivalvular leak.

Figure 1 shows our current institutional use of advanced diagnostics in our congenital heart disease population. Our MRI volume is 2–3 fold higher than our CT volume. Despite the considerable growth of our use of cardiac CT, it accounts for approximately 1 % of our diagnostic studies in this patient population.

Risks of Cardiac CT in the Recent Era

The risk of advanced diagnostics in patients with congenital heart disease is inversely related to their hemodynamic stability, size and age of the patient, and the complexity of their heart disease. The smallest patients with the most complex disease, particularly those who are critically ill, are at highest risk for adverse event with any imaging study.

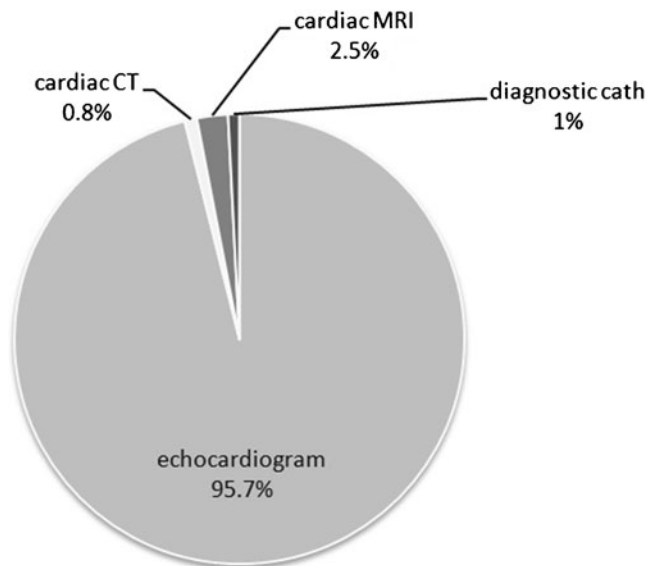


Fig. 1 The figure above illustrates the use of cardiac diagnostics for patients with congenital heart disease at a single institution over 2 years. The total number of diagnostic studies performed per year in this patient population is between 11,000 and 12,000

Radiation Exposure

Radiation exposure is one of the major concerns regarding CT scanning and cardiac catheterization in children. Children are more sensitive to radiation exposure and also have a longer life span over which any adverse effect may manifest. Newer generation CT scanners significantly reduce radiation dose compared with previous generation scanners, and there are many studies that estimate doses less than 1 mSv for many CHD applications even when calculated using a 16 cm phantom size and using age adjusted chest conversion factors [14, 15]. *CT has been shown to deliver 10–15 fold less radiation than cardiac catheterization using current technology [16•]. High doses are still delivered in very large patients or in patients with fast or highly irregular heart rhythms who need an ECG gated scan [17, 18].

Measurement of Radiation Dose in Pediatric CT

Most adult practitioners calculate estimated radiation dose from a cardiac CT scan by multiplying the scan DLP by a standard chest conversion factor. Calculating age and size specific radiation dose for CT scanning in pediatric patients is more complicated, and multiple methods are proposed [19, 20]. The most common is converting a 32 cm phantom to a 16 cm phantom DLP estimate, and further adjusting this by an age adjusted chest conversion factor to determine dose in millisievert (mSv) [21]. The phantom and age conversions combined give a dose estimate that is approximately 7 times higher than for the same scan output delivered to a larger and older patient [22]. The various methods used to

determine radiation dose from CT scanning complicate scanner, sequence, and inter-modality comparisons. In addition, these age and size adjustments are typically not performed for other imaging modalities that use radiation such as cardiac catheterization, fluoroscopy for chest Xray. To compare scan modes and sequences, comparison of DLP for similar size phantoms may be the most reproducible until consensus on this topic is achieved.

The following are general guidelines for reducing scanner output and radiation dose in pediatric cardiac scans. Furthermore, we use aggressive beta blockage if coronary artery imaging is needed to allow use of the highest pitch scan sequence with minimal diastolic padding.

- (1) Use the minimal scanner output (kv and mAs) required for a diagnostic scan. We use 80 kv to 60 kg for 0.6 mm collimator, and to 80 kg if using a 1.2 collimator. 70 kv voltage tubes are also now available on some scanner platforms and could further reduce dose for the smallest patients.
- (2) Use weight based mAs, with additional online tube current modulation based on localizer images. Additionally, reduce mA 20 %–30 % prospectively when iterative reconstruction is available.
- (3) Use the highest pitch scan mode that is diagnostic to minimize both image acquisition time and radiation dose.
- (4) For ECG gated scans, use prospective ECG triggering whenever possible, and use minimal padding (systolic and diastolic or systolic intervals only for high heart rates if coronary artery detail is needed), diastolic interval only for heart rate <60 bpm or at higher heart rates when coronary detail is not required.
- (5) Use 1.2 collimator with prospective 20 %–30 % decrease in mA if fine detail is not needed.
- (6) Limit the scan range to area of interest.
- (7) Use a long contrast injection with image acquisition at the end of the injection to acquire venous and arterial anatomy in the same scan when both are needed (we do second scan on less than 10 % of patients).

Anesthesia Considerations

*Young patients with congenital heart disease in anesthesia class 3 or above are at highest risk for an adverse event with anesthesia [23, 24•]. In a study of 12,000 pediatric anesthetics, the risk of cardiac arrest was higher outside the operating room, and highest in those with single ventricle heart disease. Cardiac CT scans can now be performed without sedation, or with minimal sedation for most indications. Studies have shown very little motion artifact using these techniques [25]. For patients who do require anesthesia for cooperation with a breath hold in the length of

anesthesia for MDCT will be much shorter than with MRI or catheterization due to the rapid data acquisition of current generation MDCT scanners. For these indications, a single scan sequence obtained over 1 breath hold will acquire the entire 3 or 4D dataset.

*There is an evolving body of evidence that anesthesia exposure in young patients may adversely affect long term cognitive and behavioral outcomes, particularly those exposed to multiple anesthetics or exposed younger than 2 years of age [26, 27, 28•, 29–33].

IV and Contrast Exposure

Most pediatric cardiac CT scans are performed using a power injector through a peripheral IV or a central line in postoperative patients. The use of power injections in pediatrics is well described and safe [34]. The rate of contrast reaction in the pediatric age range is low and increases with age [35, 36].

Use of CT and Examples for Common Indications

Cardiac CT has been well described to define congenital heart disease. CT is particularly useful for anomalous pulmonary veins, vascular rings, aortic arch pathology, and pulmonary arterial or lung parenchymal pathology [37–41]. Figure 2A shows the images from a patient with an absent left pulmonary artery who also had left lung agenesis. The scan was performed free breathing without sedation, scan DLP 5. Figure 2B is a posterior view of a patient with an AV canal defect and recurrent left lung collapse. The picture shows the vascular airway interaction with compression of the left bronchus between the enlarged pulmonary artery and the aorta. A LeCompte maneuver was performed at the time of AV canal repair.

Planning for Bypass Cannulation and Sternal Entry or Closure

Sternal re-entry can be aided by CT angiography. It readily shows the sternum and the relationship to underlying cardiac and vascular structures. Figure 3A and B show a large pseudoaneurysm from the aortic root in a patient who developed endocarditis after truncal valve repair and VSD closure. Defining the location of the pseudoaneurysm allowed safe sternal re-entry. The central veins were additionally found to be occluded, which changed the plan for venous cannulation. For patients with an open sternum following surgical repair, CT can identify residual lesions that may be addressed at that time of sternal closure. For these critically ill patients in particular the fast acquisition time can limit possible anesthesia adverse events and time

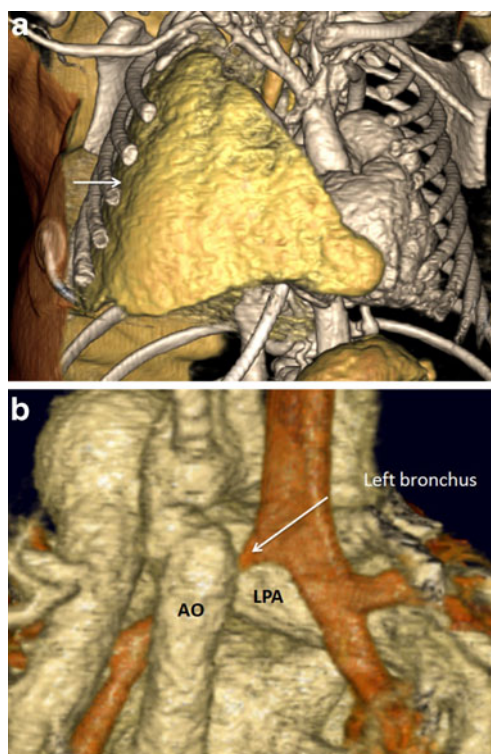


Fig. 2 **a**, Enlarged right lung (arrow) with left lung and left pulmonary artery agenesis in a 2-day old. The heart is shifted leftward. The scan was performed free breathing without sedation, scan DLP 4. **b**, This posterior view shows left bronchial compression between an enlarged left pulmonary artery and the aorta. The patient has an AV canal defect with high output congestive heart failure

away from the intensive care unit. Figure 4 shows images obtained in a 3-day-old with an open sternum and mediastinal chest tubes after repair of total anomalous pulmonary venous return, with a patent vertical vein to the superior vena cava. The time from the first localizer to the helical scan was 3 minutes, helical scan image acquisition was 0.25 seconds, and scan DLP was 9. There was no change in the ICU ventilator management or equipment in preparation for, or during acquisition of the scan. The vertical vein was ligated at the time of delayed sternal closure.

Single Ventricle Heart Disease

Patients with single ventricle heart disease require advanced imaging for interstage evaluation because echocardiography provides for a complete vascular evaluation in only a minority of patients [42]. The traditional modality used for interstage evaluation is cardiac catheterization, which is no longer necessary in low risk patients given the high complication rate without improvement in surgical outcome compared with non-invasive evaluation [43]. MRI has been proposed for interstage evaluation prior to the cavopulmonary anastomosis (Glenn) but has not been widely adopted [44]. Metallic implants decrease

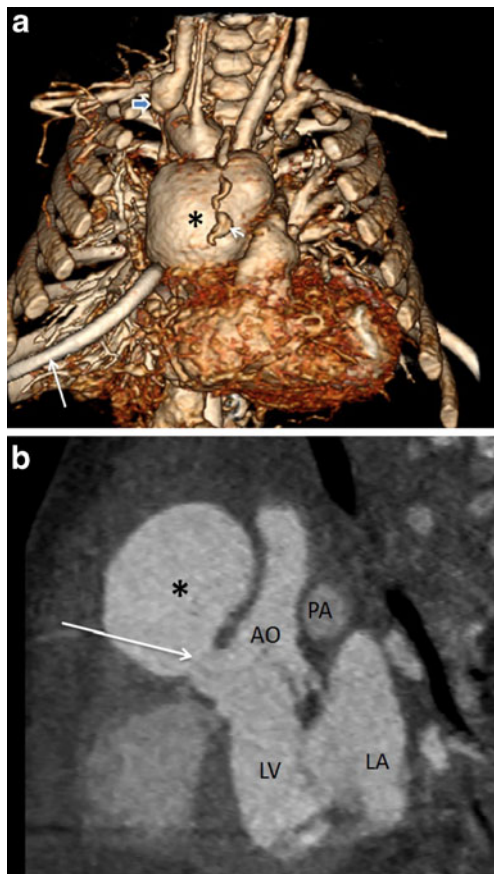


Fig. 3 **a**, The asterisk shows a large truncal valve pseudoaneurysm and the relationship to the sternum (short arrow). A right sided chest tube is seen (long arrow), as are an occluded right subclavian vein, proximal superior vena cava (fat arrow), and innominate vein. The left jugular vein drains via collaterals to the inferior vena cava. **b**, This sagittal view shows a large pseudoaneurysm (asterisk) from the left ventricular outflow tract and proximal truncal root in a patient with a history of truncus arteriosus after VSD closure and truncal valve repair. The truncal cusp is seen (arrow) with aneurysm extending from both above and below the valve leaflet

the diagnostic quality of MRI for these patients additionally [45]. Cardiac CTA is an alternate modality for assessment of anatomy prior to Glenn cavopulmonary anastomosis with excellent visualization of aortopulmonary shunt or Sano shunt anatomy, pulmonary anatomy, and aortic arch anatomy. High pitch CTA can be obtained free breathing and with minimal or no sedation in this age range. Evaluation of the postoperative Glenn cavopulmonary anastomosis is reliably seen by CTA. Visualization of venous collaterals causing cyanosis after the Glenn procedure is also seen. Some now propose Fontan completion with noninvasive interstage evaluation [46]. Fontan opacification by CT imaging is frequently difficult to achieve and the venous filling is likely dependent on cardiac output. We use either a 2 phase injection with late image acquisition or a delayed scan for this indication to increase the chance of opacifying the inferior vena cava and Fontan pathway. CT imaging has also been proposed as a method of detecting

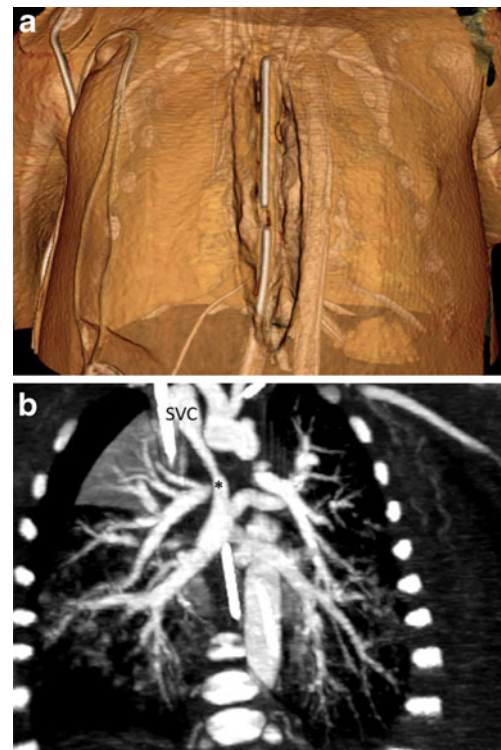


Fig. 4 **a**, Two-day old has an open sternum with a mediastinal chest tube after repair of total anomalous pulmonary venous return. Sick patients can be imaged with CTA even prior to sternal closure. **b**, This coronal view shows a patent vertical vein (asterisk) to the superior vena cava. The vertical vein was ligated at the time of delayed sternal closure

pulmonary embolism and plastic bronchitis in single ventricle patients [47–49]. Figure 5 A H show examples of interstage single ventricle palliation through the third stage Fontan.

Tetralogy of Fallot

Advanced imaging plays a crucial role in the long-term management of patients with Tetralogy of Fallot. This includes defining optimal timing for pulmonary valve replacement for patients with significant pulmonary insufficiency, and the assessment the branch pulmonary arteries and collateral anatomy for all stages of repair in those with pulmonary atresia [50]. Most all centers index right ventricular end diastolic and end systolic volumes to body surface area along with assessment of ejection fraction to determine the need for repeat intervention on the right ventricular outflow tract in the presence of pulmonary insufficiency [50, 51]. An ECG gated CT scan can be used to obtain this same functional information for patients with repair of Tetralogy of Fallot who are contraindicated for MRI or who have imaging artifacts [52]. CT evaluation of tetralogy of Fallot has been shown to be highly specific for native pulmonary artery anatomy, aorto-

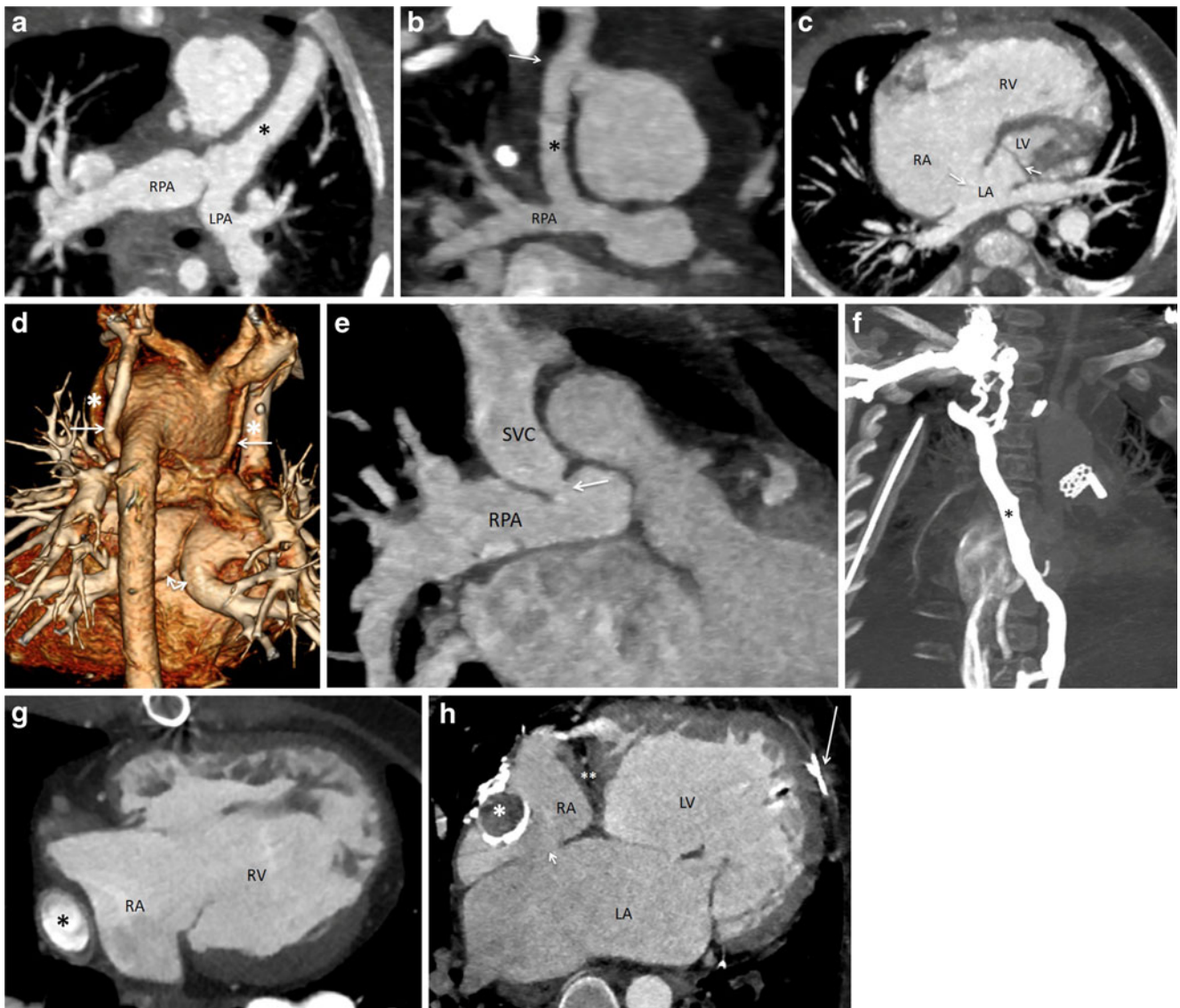


Fig. 5 **a**, This is an axial view of the distal Sano conduit (asterisk) to the branch pulmonary arteries in a patient with palliated hypoplastic left heart syndrome after a stage 1 procedure. **b**, This figure shows the length of a modified Blalock Taussig shunt from the innominate artery to the mid-right pulmonary artery in a patient after stage 1 single ventricle palliation. **c**, This axial view shows the small mitral valve (arrow) and left ventricle in a patient with hypoplastic left heart syndrome. The atrial septum is patent in the area of prior septectomy. **d**, This posterior view shows bilateral superior vena cava (asterisk), bilateral modified Blalock Taussig shunts (arrows) to the branch pulmonary arteries, ipsilateral pulmonary venous drainage (short arrows), and a reconstructed aorta prior to the Glenn procedure. **e**, This shows the superior vena cava to right pulmonary artery anastomosis in a patient after second stage single ventricle palliation with a

cavopulmonary anastomosis (Glenn). There is a narrowing at the site of anastomosis (arrow). **f**, A right arm injection (coronal MIP) is performed to evaluate the cause of cyanosis in a patient after the Glenn procedure. The right internal jugular vein and right subclavian vein are occluded with decompression through the azygous vein (asterisk) to the inferior vena cava. **g**, This axial 2D image shows an extracardiac Fontan conduit (asterisk) in a patient with hypoplastic syndrome (mitral and aortic atresia). The patient also has ventricular dysfunction and elevated pulmonary artery pressures. Opacification of the extracardiac conduit can be difficult given that it carries unopacified IVC blood following an arm injection. **h**, This patient with tricuspid valve atresia (double asterisk) has an interatrial Fontan (asterisk). Note the epicardial pacer lead (long arrow) and the patent atrial communication (short arrow)

pulmonary collateral anatomy, postoperative shunt anatomy and coronary artery assessment [53–61]. A 2006 study directly compared cardiac catheterization and CT angiography in a small number of patients with pulmonary artery atresia and found complete agreement between modalities for definition

of pulmonary artery and aorto-pulmonary collateral anatomy [62]. One hundred patients with Tetralogy of Fallot were evaluated by CT preoperatively and there was 100 % sensitivity and specificity for coronary artery anomalies compared with surgical findings in patients with a median age of

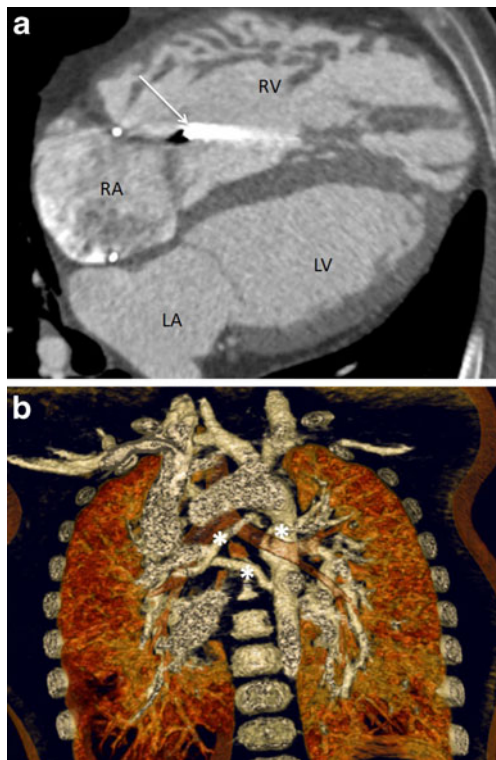


Fig. 6 **a**, This adult patient has a transvenous pacer (arrow) after repair of Tetralogy of Fallot. Note the right atrial and right ventricular enlargement, even after replacement of the pulmonary valve for severe pulmonary insufficiency. **b**, This 3D coronal VRT shows the collateral vessels (asterisk) arising from the descending aorta in a patient with Tetralogy of Fallot, pulmonary atresia with aortopulmonary collateral vessels

6.8 months [53]. In patients who have had placement of a RVOT conduit, transcatheter valve replacement is now an option when repeat intervention is needed. Defining the dimensions of the proximal and distal conduit, the branch pulmonary arteries, anomalous muscle bundles in the right ventricular outflow tract that could affect catheter course and complete definition of the coronary arteries is required prior to

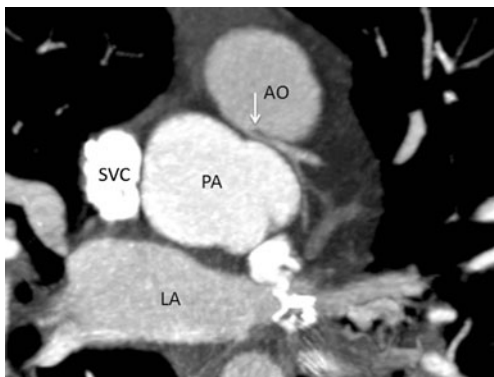


Fig. 7 The arrow shows a left coronary artery arising from the right facing aortic sinus in a patient with d-TGA who had an atrial switch procedure. The origin of the artery is small, and the course is interarterial between the aorta and pulmonary artery

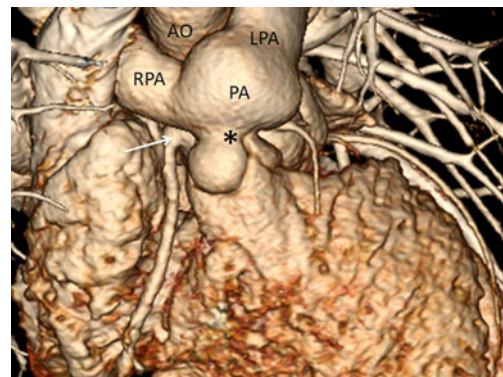


Fig. 8 This 3D VRT shows supravalvular pulmonary stenosis (asterisk) in a patient after the arterial switch with LeCompte maneuver for d-TGA. Note the position of the main and branch pulmonary arteries anterior to the aorta, and a patent right coronary artery (arrow) after reimplantation at the time of arterial switch

catheterization or surgical intervention. Figure 6A shows an enlarged right ventricle in a pacemaker dependent patient with Tetralogy of Fallot. Figure 6B shows aorto-pulmonary collateral arteries in a neonate with pulmonary atresia and the relationship of the collateral vessels to the bronchus that helps guide future surgical dissection.

Transposition Complexes

Atrial Switch

Patients with transposition of the great arteries primarily underwent an atrial switch operation until the technical details of the coronary transfer allowed for survival after the arterial switch operation in 1980s. The systemic venous baffles, pulmonary venous baffles, and biventricular systolic function are well seen by CT imaging when using a biventricular injection protocol. Valvular regurgitant fraction may be calculated by assessing stroke volume differences. Figure 7 shows transposition of the great arteries with

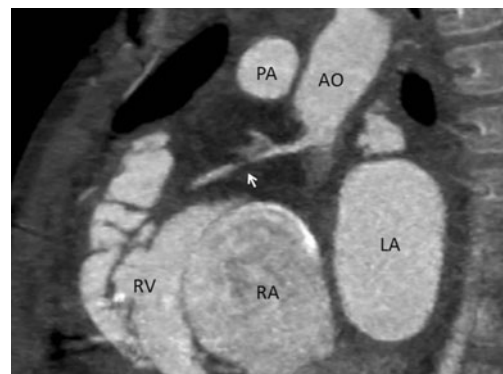


Fig. 9 A severe right coronary artery stenosis is present (arrow) in a patient after the Nikaidoh procedure (subsequently proven by invasive coronary angiography)



Fig. 10 A narrowed pulmonary venous baffle is present (asterisk) anterior to the left ventricle in a patient after a double switch procedure

an anomalous and interarterial left main coronary artery in a patient who presented with a ventricular fibrillation arrest 15 years after an atrial baffle procedure.

Arterial Switch/Rastelli Procedure/Nikaidoh Procedure

Since the 1980s, the arterial switch operation is the procedure of choice for patients with transposition of the great arteries. The neo-pulmonary root, neo-aortic root, and coronary arteries all subsequently need long-term follow-up with serial evaluation. The mid-term results for the arterial switch are good, but a certain percentage of patients will develop coronary artery problems, particularly if there was a high risk coronary artery pattern such as a commissural malalignment complicating coronary transfer, surgical concern regarding the coronary anastomosis, or a difficult postoperative course [63]. Additionally supravalvular pulmonary stenosis is relatively common at the site of the neo-pulmonary anastomosis (Fig. 8). It is recommended that all patients with reimplanted coronary arteries undergo angiography either prior to adulthood or if symptomatic [1]. In patients with transposition of the great vessels with pulmonary stenosis, the pulmonary conduit placed during the Rastelli operation also requires repeat evaluation to determine the timing of eventual replacement. The Nikaidoh procedure is a newer surgical approach for d-TGA with pulmonary stenosis, consisting of aortic translocation with reconstruction of the right ventricular outflow tract. Figure 9 shows right coronary artery stenosis in a 3 year old following the Nikaidoh procedure, confirmed at the time of therapeutic coronary angioplasty.

Double Switch

In patients with physiologically corrected transposition (also called L-TGA, (S,L,L) (I,D,D), a concomitant atrial baffle and arterial switch are needed to restore normal cardiac physiology. Pre-operative evaluation for this lesion includes visualization of the coronary arteries, great artery relationships, and the

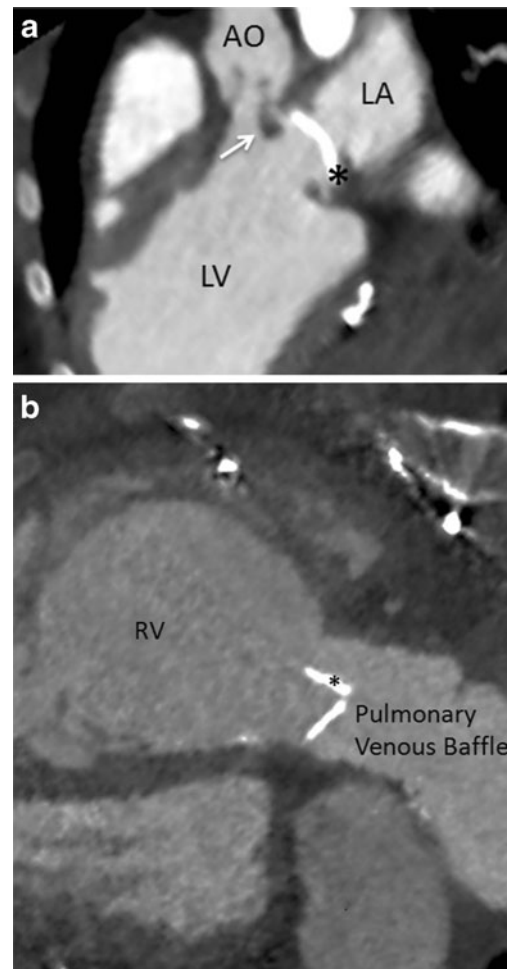


Fig. 11 a, A subaortic membrane (arrow) extends from the undersurface of the mechanical mitral valve (asterisk) to the coronary cusp. b, Note the abnormal position of the mechanical tricuspid valve leaflet (asterisk) in ventricular diastole on this ECG triggered scan

relationship of the VSD to the right ventricular outflow tract for visualization of baffles. Some patients have a pulmonary artery band placed to increase right ventricular afterload prior

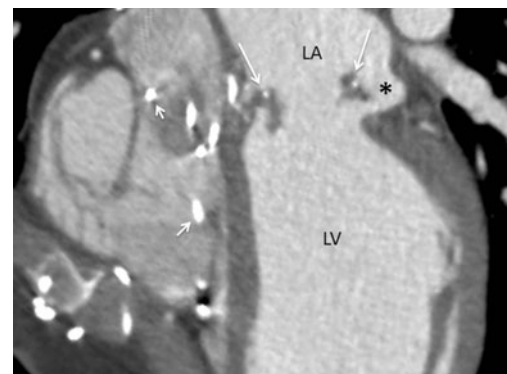


Fig. 12 A mitral bioprosthetic valve (arrows) is present with a large perivalvular leak (asterisk). Pacer wire artifacts (short arrows) are seen in the right atrium and ventricle

to a double switch to increase left ventricular afterload. An ECG gated functional scan allows visualization of the interventricular septal configuration and also quantification of left ventricular myocardial mass. Figure 10 shows a narrowed pulmonary venous baffle to the anterior left ventricle in a patient after the double switch procedure.

Prosthetic Valves

Many pediatric patients develop a progressive gradient through a mechanical valve. This may be a result of pannus formation with limitation of valve leaflet mobility, patient size mismatch with growth, or thrombus.

For bileaflet mechanical valves, the end systolic and end diastolic angles can be determined from an ECG gated scan. In patients requiring valve replacement, definition of coronary artery relationship to the AV valve can be determined. Figure 11A shows a prosthetic mitral valve with progressive left ventricular outflow tract gradient thought secondary to the valve position after catheterization. CT showed tissue extending from the valve onto the aortic valve that could be removed lowering the LVOT gradient postoperatively, without replacing the mitral valve. Figure 11B shows a diastolic image of an obstructed prosthetic tricuspid valve that partially responded to thrombolytic therapy.

Perivalvular leaks can also be visualized and regurgitant volume quantified. Figure 12 shows the location of a large mitral perivalvular leak in a patient with severe mitral regurgitation and left ventricular enlargement following valve replacement.

Coronary Artery Abnormalities

Coronary artery imaging is recommended in pediatric patients with symptomatic anomalous coronary arteries with reimplanted coronary arteries, and prior to right ventricular outflow tract intervention [1, 64–66]. Coronary CTA is considered superior to conventional angiography in delineating the ostial origin and proximal path of an anomalous coronary artery. Catheter manipulation may mechanically alter the coronary ostium and the intrinsic 3D noninvasive nature of CT allows simultaneous visualization of the coronary artery, aorta and pulmonary artery [67, 68]. CT evaluation of coronary artery course is possible even in young patients [69]. Recently, the proximal course and length of the intramural segment of an anomalous right coronary artery from the left facing sinus may help risk stratify these lesions [70, 71]. In a study directly comparing cardiac CT angiography and catheterization in patients after the arterial switch operation, there was complete agreement regarding the detection of a significant coronary stenosis [72].

Conclusions

The combination of improved temporal and spatial resolution, rapid image acquisition, and radiation dose reduction techniques makes CT an ideal imaging modality for certain indications in patients with congenital heart disease. It is critical to pay careful attention to all of the details of scan acquisition and radiation dose reduction techniques in order to maintain diagnostic image quality while minimizing risk. As this patient population grows and modern CT technology becomes more accessible, it will play an increasingly important role in the diagnosis and management of congenital heart lesions.

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