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Review Article

CT imaging in congenital heart disease: An approach to imaging and interpreting complex lesions after surgical intervention for tetralogy of Fallot, transposition of the great arteries, and single ventricle heart disease

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ABSTRACT

Echocardiography and cardiac magnetic resonance imaging are the most commonly performed diagnostic studies in patients with congenital heart disease. A small percentage of patients with congenital heart disease will be referred to cardiac CT subsequent to echocardiography when magnetic resonance imaging is insufficient, contraindicated, or considered high risk. The most common complex lesions referred for CT at our institution are tetralogy of Fallot, transposition complexes, and single ventricle heart disease. This review discusses the most common surgical procedures performed in these patients and the technical considerations for optimal image acquisition on the basis of the prior procedure and the individual patient history. Cardiac CT can provide the functional and anatomic information required for decision making in complex congenital heart disease. Image interpretation is aided by knowledge of the common approaches to operative repair and the residual hemodynamic abnormalities. Acquisition and interpretation that is both individualized to the patient's underlying disease and the specific clinical question is likely to maintain diagnostic accuracy while decreasing the potential risk of cardiac CT.

1. Introduction

CT has become a standard clinical test for many indications in adult cardiology. Because of concerns about radiation, it has been introduced cautiously in the pediatric and young adult cardiac patient population. Technical advances have allowed for both high-quality images and a marked decrease in the radiation dose of cardiac CT. For select indications, cardiac CT may provide better information with a lower risk than other diagnostic modalities.

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This article describes our approach to the use of cardiac CT for patients who have undergone previous repair or palliation of complex congenital heart disease (CHD). All CHD patients referred for CT imaging after intervention will have undergone previous testing and will have known disease that requires further definition of anatomy or physiology for clinical management.

1.1. *Use of imaging modalities in CHD*

Most patients with congenital heart lesions survive to adulthood and require serial cardiac evaluation.[1,2] When surgical intervention became common for congenital heart lesions, catheterization was the only available diagnostic modality. Echocardiography quickly replaced cardiac catheterization for most indications, and most patients with CHD are managed with this modality alone.[3] The limitations of echocardiography include degradation of acoustic windows with surgical scarring and obesity, poor definition of the vascular–airway relationship, and poor visualization of thoracic vasculature. In addition, right ventricular (RV) and single ventricular systolic function, valvular regurgitation, and shunt fraction calculation are not reproducibly quantified by echocardiography.[4]

Cardiac magnetic resonance imaging (MRI) overcomes many of these limitations but is incompatible with most current generation pacemakers and defibrillators, and artifact from certain metallic coils and implants degrade image quality. The relatively long imaging time and requirement for breathholding for most sequences also limits its use in certain patients.[5–8] For the patient with CHD who needs imaging subsequent to echocardiography and when MRI is contraindicated or considered high risk, cardiac CT is now the modality most often used for anatomic and physiological evaluation at our institution (Table 1). Cardiac catheterization is primarily used for patients who need invasive pressure measurement for clinical management.

1.2. *Cardiac CT in CHD*

Early CT scanners provided limited value for cardiac applications in young patients. The latest multidetector row CT technology produces isotropic submillimeter spatial resolution, improved temporal resolution, and rapid image acquisition with large coverage. As a result, cardiac CT scans even in the youngest patients with high heart rates are diagnostic and have lessened or eliminated the requirement for sedation or anesthesia. Newer CT technology has decreased radiation dose compared with previous generation scanners, and image processing techniques allow further reduction in dose without loss of image quality. Each CT scan must be tailored to the patient and clinical question to decrease diagnostic risk (Table 2).

Table 1 – When to consider CT for complex congenital heart disease.

Contraindications to MRI (pacemaker, defibrillator)
Poor MRI image quality because of metallic artifact or device
Evaluation of stent anatomy and integrity (particularly small vessel and stent)
Evaluation of prosthetic valve function or paravalvular leak when other modalities are insufficient
Adult patient who needs coronary artery evaluation in addition to definition of complex anatomy
Young patient if considered high risk for adverse event with anesthesia required for MRI and a CT can be performed with minimal or no sedation
Older patient with claustrophobia unable to cooperate with an MRI
Patient requiring a CT for evaluation of extracardiac anatomy (lung parenchyma or airway, skeletal abnormality) Obesity, unable to have cardiac MRI performed with available scanner

MRI, magnetic resonance imaging.

2. An approach to interpreting CT in palliated CHD

A disease-based approach to the performance and interpretation of the cardiac CT examination may be the most useful for the patient with CHD. Understanding the initial anatomy, surgical procedures performed, and the common short- and long-term complications is essential for successful imaging. The most common complex diseases referred for CT evaluation in our repaired or palliated CHD population are tetralogy of Fallot (TOF), transposition of the great arteries (TGA), and single ventricle heart disease.

3. Tetralogy of Fallot

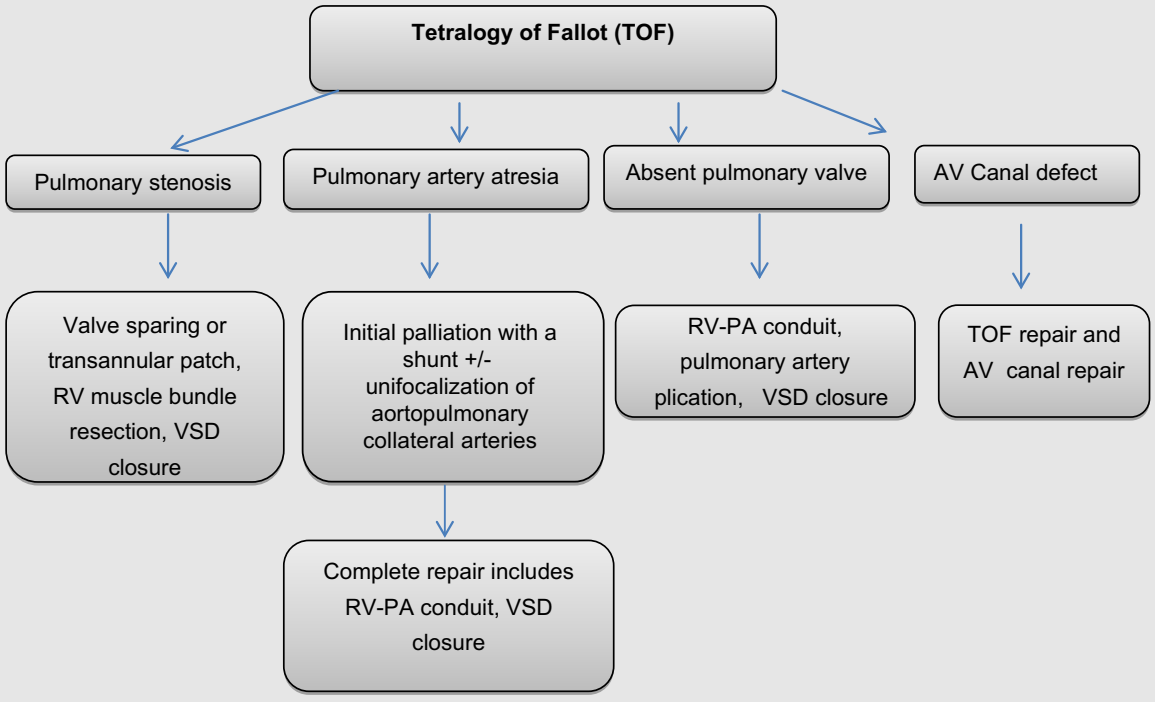
TOF consists of 4 distinct clinical entities, with differing anatomic features, surgical approaches, and long-term outcomes (Table 3). The classic form of TOF is the most common and includes a large ventricular septal defect (VSD) and subvalvar and valvar pulmonary stenosis. Less common is TOF with pulmonary artery (PA) atresia and aortopulmonary collaterals, and rarely TOF with absent pulmonary valve or TOF with an atrioventricular (AV) canal defect. Before initial surgical intervention, TOF with pulmonary stenosis rarely requires

Table 2 – Dose-reduction techniques for CT in congenital heart disease.

Perform a clinically indicated CT scan only when risk is less, or the information superior, to other imaging modalities
Adjust contrast injection and image acquisition to allow 1 scan to obtain all information needed (no “routine” delayed venous scan)
Limit the scan range to the area of interest
Adjust scanner output for patient size
Use automated exposure algorithms for tube current and tube potential adjustment
If iterative reconstruction is available, prospectively reduce scanner output by at least 30%
Use wider collimator and decrease dose by additional 20% to 30% if fine detail is not needed
Use prospective ECG triggering when possible for ECG-gated scans
Use the minimal padding necessary for coronary imaging on the basis of heart rate
For coronary artery imaging, consider β-blockade to lower the heart rate if this allows prospective ECG-triggered techniques to be used

ECG, electrocardiogram.

Table 3 – Tetralogy of Fallot: Common interventions and residual hemodynamic lesions.



Type of tetralogy	Common residual hemodynamic lesions
TOF with pulmonary stenosis	Pulmonary stenosis, pulmonary insufficiency, RV dilation and dysfunction, aortic root dilation
TOF with pulmonary artery atresia	Right ventricle–PA conduit, branch PA or distal PA stenosis, conduit insufficiency, RV dilation and dysfunction, aortic root dilation
TOF with absent pulmonary valve	Right ventricle–PA conduit stenosis or insufficiency, airway abnormalities, PA dilation, RV dilation and dysfunction, aortic root dilation
TOF with AV canal defect	See tetralogy, right- or left-sided AV valve regurgitation, LVOTO

Suggested scan modifications

- Biventricular injection protocol.
- Increase scan range to include proximal PAs for TOF with PS, increase further if history of pulmonary atresia and continued pulmonary hypertension to evaluate distal pulmonary bed.
- Retrospectively ECG-gated scan with pulsed radiation, minimal padding, and multiphase reconstruction for calculation of biventricular ejection fraction, end-diastolic and end-systolic dimensions.
- If severe pulmonary insufficiency documented on echocardiography, use stroke volume differences between ventricles to estimate regurgitant fraction.

AV, atrioventricular; LVOTO, left ventricular outflow tract obstruction; PA, pulmonary artery; RV, right ventricular; TOF, tetralogy of Fallot; TGA, ventricular septal defect.

imaging beyond echocardiography. Advanced imaging is required for patients with PA atresia to determine native PA and aortopulmonary collateral anatomy and for interstage evaluation[9–16] (Fig. 1). CT can also be used to define the common associated problem of a coronary anomaly, which is particularly important if it crosses the RV outflow tract (RVOT) and affects the surgical approach.[9] In patients with TOF and absent pulmonary valve, a ductus arteriosus is usually not present in utero, and all blood flow is required to flow through the lungs, with a high resistance. The PAs often dilate in response and can cause bronchial compression. In rare cases the lungs may not develop properly. For patients with significant PA dilation and respiratory symptoms, imaging is required to assess the airway for extrinsic bronchial compression, intrinsic airway pathology, or pulmonary hypoplasia. PA plication is often performed at the time of the initial intervention and the PAs may remain dilated after intervention.

3.1. *Common interventions for TOF*

Complete repair of TOF with pulmonary stenosis includes VSD closure and relief of pulmonary stenosis with muscle bundle resection, valve sparing patch, pulmonary valvotomy, or a transannular patch. Complete repair of TOF with PA atresia may be preceded by palliative shunts and “unifocalization” of aortopulmonary collaterals into a common vessel. The timing of VSD closure in pulmonary atresia depends on the growth of the reconstructed PAs and resulting RV pressures. Residual and recurrent proximal and distal PA stenosis may result in RV hypertension after VSD closure. Catheterization procedures

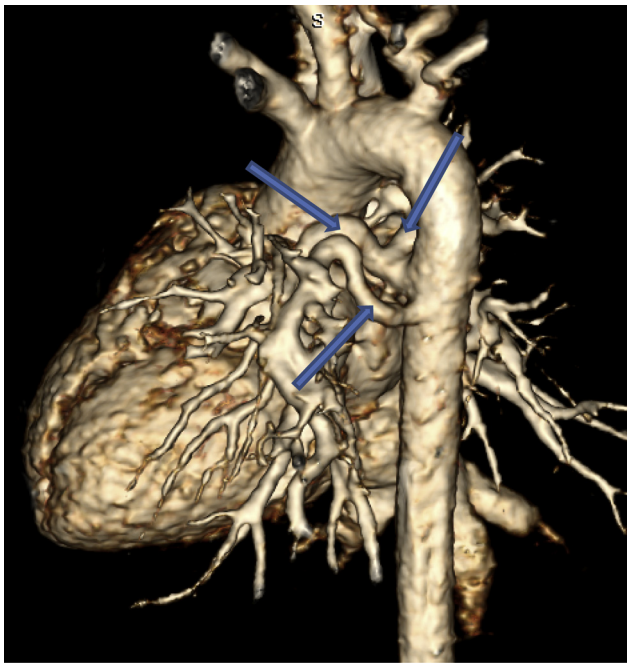


Fig. 1 – This 3-dimensional reconstruction shows a posterior view of aortopulmonary collaterals from the proximal descending aorta (*arrows*) in a neonate with tetralogy of Fallot and pulmonary artery atresia.

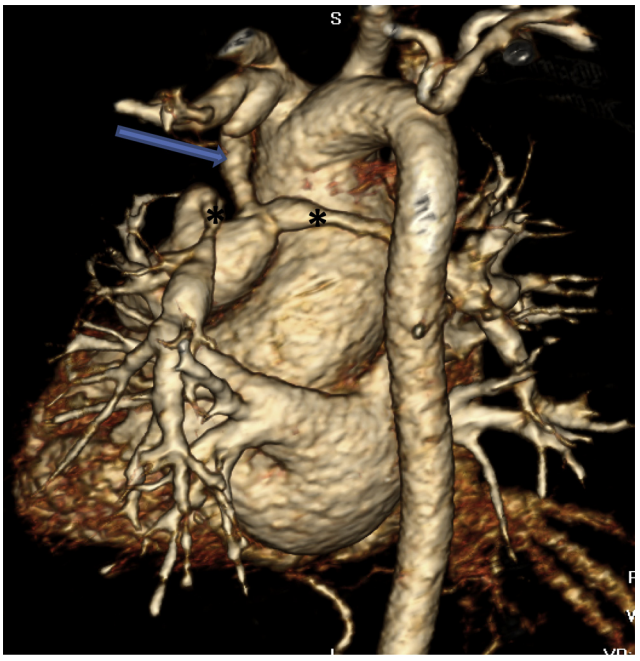


Fig. 2 – This 3-dimensional reconstruction shows the posterior view of bilateral branch pulmonary artery stenosis (*) after aortopulmonary shunt (*arrow*) from the base of a left-sided innominate artery. The patient has tetralogy of Fallot with severe pulmonary stenosis. Initial palliation with a shunt was performed because of the small size of the native pulmonary arteries.

with pulmonary balloon angioplasty or stent placement or both are common in these patients before and after complete repair with VSD closure. In TOF with absent pulmonary valve, PA plication to relieve airway compression is often performed at the time of VSD closure in patients with respiratory symptoms. For both TOF with pulmonary atresia and TOF with absent pulmonary valve, a right ventricle–PA conduit is placed to the branch PAs. For patients with residual distal PA stenosis and RV hypertension an atrial communication may be left at the time of surgery to allow right-to-left atrial shunting. Patients with TOF and AV canal defect usually undergo simultaneous repair of both lesions and are at risk of postoperative left ventricular (LV) outflow obstruction from left-sided AV valve tissue.

3.2. *Postoperative imaging*

Almost all patients will have some degree of pulmonary stenosis or pulmonary insufficiency after the initial repair of TOF. Advanced imaging plays a crucial role in the long-term management of this patient population. This includes defining the optimal timing for pulmonary valve replacement for patients with significant pulmonary insufficiency after valve-sparing repair or transannular patch. In addition, it is useful for assessment of the branch PAs and unifocalized collaterals for all stages of repair in patients with PA atresia[17–19] (Figs. 2 and 3). Most centers use MRI to index RV end-diastolic and end-systolic volumes to body surface area and to assess ejection fraction to determine the timing for repeat intervention in the presence of pulmonary insufficiency.[17,18] When MRI is contraindicated or considered high risk for patients, an electrocardiogram (ECG)-gated CT scan can be used to obtain this important functional information that allows comparison of left and RV stroke volumes to calculate a pulmonary regurgitant fraction.[20]

In patients who have a right ventricle–PA conduit, transcatheter valve replacement is now an option when repeat intervention is needed. Cardiac CT angiography (CTA) is used to define the dimensions of the proximal and distal conduit, the branch PAs, anomalous muscle bundles in the RVOT that could affect catheter course, and the coronary arteries. Defining the relationship of the coronary arteries to the right ventricle–PA conduit is essential for judging whether percutaneous placement of a prosthetic pulmonary valve has potential to compress an adjacent coronary artery. In these cases, surgical conduit replacement is recommended. In addition, aortic root dilation is common in patients with repaired TOF and should be evaluated.

3.2.1. *Imaging considerations*

A biventricular injection protocol can be used to opacify the right and left heart simultaneously. A retrospectively ECG-gated scan performed with pulsed radiation and multiphase reconstruction can be used to evaluate ventricular function and to quantify valvular regurgitation by measuring stroke volume differences (Fig. 4). The scan range should include the proximal PAs for all patients, and the entire lung field may be included if there is significant

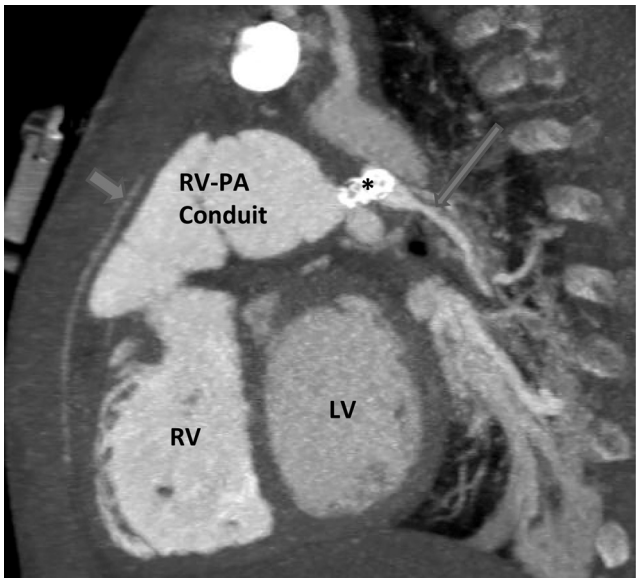


Fig. 3 – This 2-dimensional sagittal maximum intensity projection shows a mildly enlarged but hypertrophied right ventricle in a patient with tetralogy of Fallot who has undergone right ventricle-to-pulmonary artery conduit placement with subsequent stenting of the proximal left pulmonary artery (*). Note the conduit is adherent to the sternum anteriorly (*short arrow*) and the hypoplastic distal left pulmonary artery (*long arrow*).

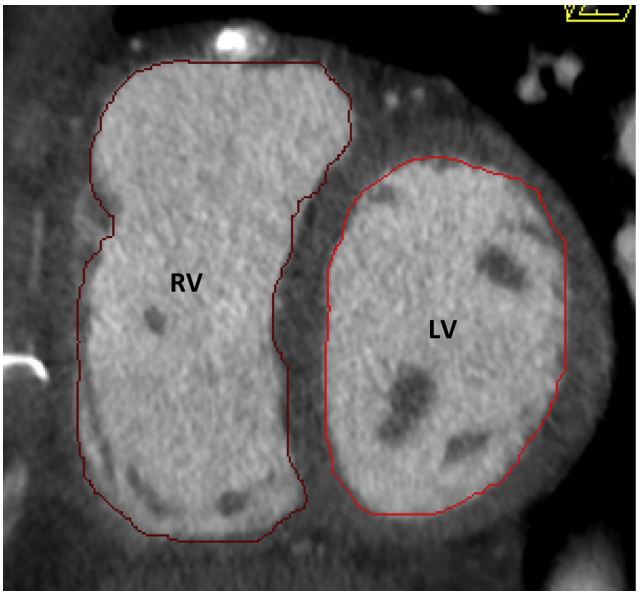


Fig. 4 – This short-axis reconstruction shows the right (*red outline*) and left (*orange outline*) ventricular end-diastolic contours used to help determine stroke volume differences and calculate the pulmonary regurgitant fraction. (Color version of this figure is available online.)

residual airway abnormality or a suspicion of distal pulmonary pathology.

4. Transposition complexes

TGA is one of the most common cyanotic lesions that presents in the neonatal period (Table 4). Transposition complexes are sometimes separated into simple transposition, complex transposition, and corrected transposition. Compared with other lesions, patients with transposition complexes are prone to sinus node dysfunction and heart block and are more likely to have pacer wires in place, which become a contraindication to MRI.

Simple transposition refers to isolated ventricular–arterial discordance with roughly equivalent valve dimensions, normally functioning valves and a "usual" coronary artery pattern. The most common position of the great arteries is dextro-TGA (d-TGA), with an aortic valve anterior and rightward of the PA. TGA often has an associated VSD.

4.1. *Surgical intervention for d-TGA*

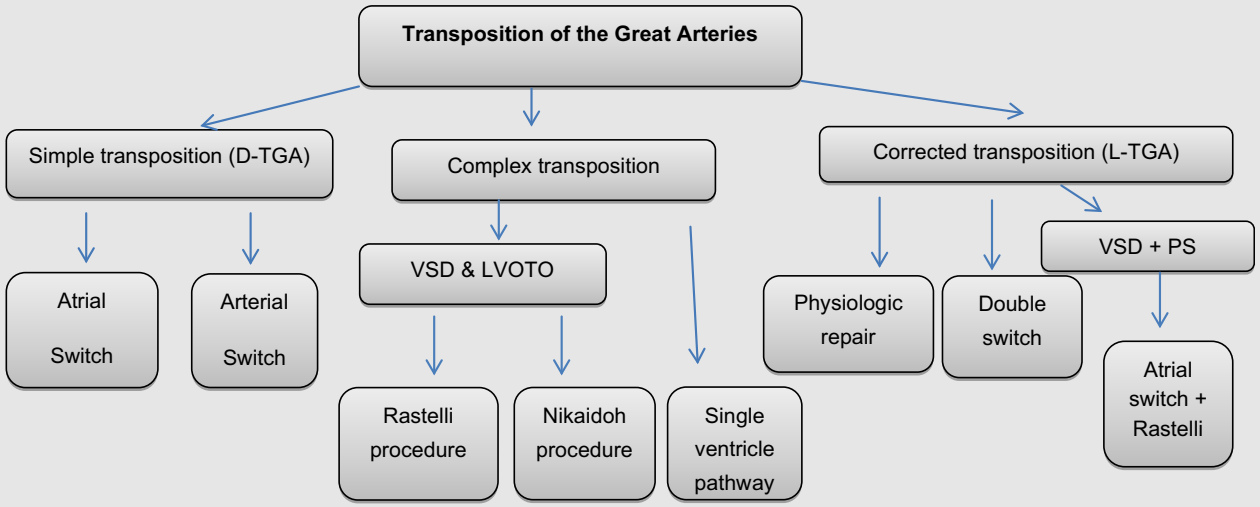
The historical approach for surgical correction of d-TGA was an atrial switch. This diverts systemic venous blood to the morphologic left ventricle then to the transposed PA. The pulmonary venous blood is directed to the morphologic right ventricle that ejects against systemic ventricular afterload to the transposed aorta.[21] Successful atrial baffle procedures were described with the use of flaps of atrial tissue by Senning[22] in 1959 and autologous pericardium by Mustard et al[23] in 1964. In 1975 the first successful arterial switch for d-TGA was described by Jatene et al[24] and has been universally adopted. The arterial switch consists of transecting the proximal aorta and PA and "switching" the great arteries by anastomosing the valve root to the appropriate great artery and re-implanting the coronary arteries to the neo-aortic root. The Lecompte maneuver is performed as part of the arterial switch procedure and repositions the neo-pulmonary root anterior to the reconstructed aorta (Fig. 5). Early surgical outcomes are excellent when the semilunar valves are of equivalent size, the valve commissures are aligned, and the coronary artery pattern is usual. If VSD closure is performed at the time of arterial switch operation, surgical survival is somewhat less favorable.

4.2. *Postoperative imaging*

After an atrial switch, the systemic venous baffles, pulmonary venous baffles, and biventricular systolic function are well seen by cardiac CTA (Figs. 6–8). Declining RV (systemic) function and tricuspid regurgitation are common after the atrial switch. Many patients develop sinus node dysfunction, heart block, and arrhythmias and require pacemaker placement. Pacer leads will be placed across the systemic venous baffle and into the LV (venous) apex.

After an arterial switch procedure, supravalvar stenosis of the reconstructed PA and branch PAs is common (Fig. 9). Coronary artery compromise at the site of re-anastomosis is uncommon in transposition with the normal coronary artery pattern. Patients who had complex operative courses at the time of arterial switch or patients with a high-risk coronary pattern are at highest risk of coronary compromise. It is recommended that the re-implanted coronary arteries be

Table 4 – Transposition complexes: Common interventions and residual hemodynamic lesions.



Procedure	Potential residual hemodynamic lesions
Atrial switch	Systemic or pulmonary venous baffle narrowing or occlusion RV failure (systemic ventricle) Tricuspid regurgitation (quantify by stroke volume differences between ventricles) RVOT obstruction from ventricular septal shift Atrial arrhythmias or sinus node dysfunction
Arterial switch	Neo-pulmonary root or branch PA stenosis, pulmonary insufficiency Neo-aortic root dilation, stenosis, or insufficiency Stenosis of re-implanted coronary arteries
Rastelli	Obstruction of the right ventricle–PA conduit or branch PAs, pulmonary insufficiency Obstruction of left ventricle-aortic pathway in the area of TGA patch
Nikaidoh	Native RVOT or RVOT conduit stenosis or insufficiency Right coronary artery stenosis because of distortion

LVOTO, left ventricular outflow tract obstruction; PA, pulmonary artery; PS, pulmonary stenosis; RV, right ventricular; RVOT, right ventricular outflow tract; TGA, ventricular septal defect.

Suggested scan modifications

- Biventricular injection protocol.
- Manual scan trigger with extended monitoring if systemic venous baffle obstruction suspected.
- Extend scan range to include systemic venous and pulmonary venous baffles, neo-aortic root and neo-pulmonary root or conduit.
- Consider β-blockade to decrease heart rate if re-implanted coronary arteries require evaluation.

studied by angiography at least once in adulthood, or if a patient is symptomatic.[25,26] Cardiac CTA is excellent for evaluating the neo-pulmonary root, neo-aortic root (Fig. 10), and re-implanted coronary arteries.

4.3. *Complex transposition (complex d-TGA)*

Surgical repair for TGA is technically more difficult when there is a concomitant TGA, coronary artery variation, either aortic or pulmonary stenosis or atresia, commissural non-alignment of the great arteries that can make coronary transfer difficult, or malposition of both great arteries from one ventricle (double outlet right ventricle). These lesions are considered complex transposition.

4.4. *Surgical procedures for complex transposition*

When pulmonary stenosis or atresia is present, the valve can be oversewn and the VSD is baffled to the aorta via the right ventricle, and a conduit is placed to the distal PA (Rastelli procedure).[27–29] The Rastelli procedure is commonly performed for d-TGA with VSD and pulmonary atresia or for double outlet right ventricle with pulmonary stenosis or pulmonary atresia (Fig. 11). A 3-dimensional or 4-dimensional data set can aid in surgical planning for patients who will undergo complex VSD baffling to a malposed aorta or who require VSD enlargement to relieve LV outflow tract (LVOT) obstruction at the site of the previously placed VSD patch. Resection of the outlet septum or enlargement of the VSD can reduce the chance of LV outflow obstruction with VSD closure but has a relatively high risk of heart block.[28] Another approach to relieve potential LVOT obstruction from a VSD patch (or from the VSD in single ventricle heart disease with malposed great vessels and an aorta from an outlet chamber) is to reconstruct the ascending aorta and main PA into a single outflow tract (Damus-Kaye-Stancel procedure) (Fig. 12). In this case the VSD patch will be placed through the right ventricle to the reconstructed great artery, and a pulmonary conduit is

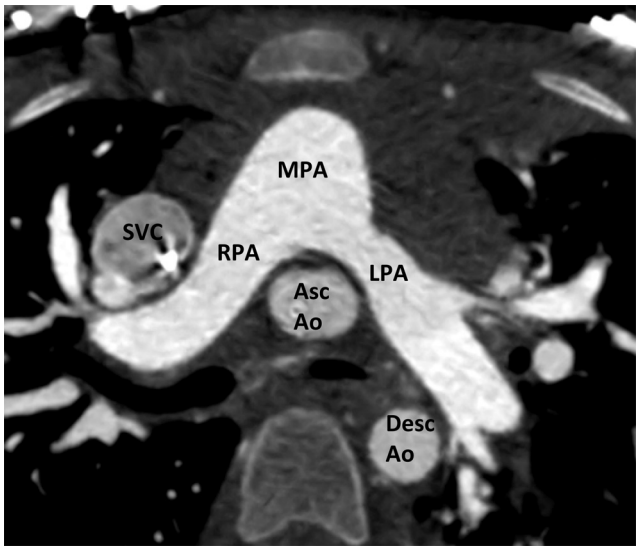


Fig. 5 – This 2-dimensional axial image shows the usual position of the neo-pulmonary root and branch pulmonary arteries after the LeCompte maneuver performed during the arterial switch operation. The main pulmonary artery is anterior to the aorta, with the right and left pulmonary arteries along the side of the aorta.

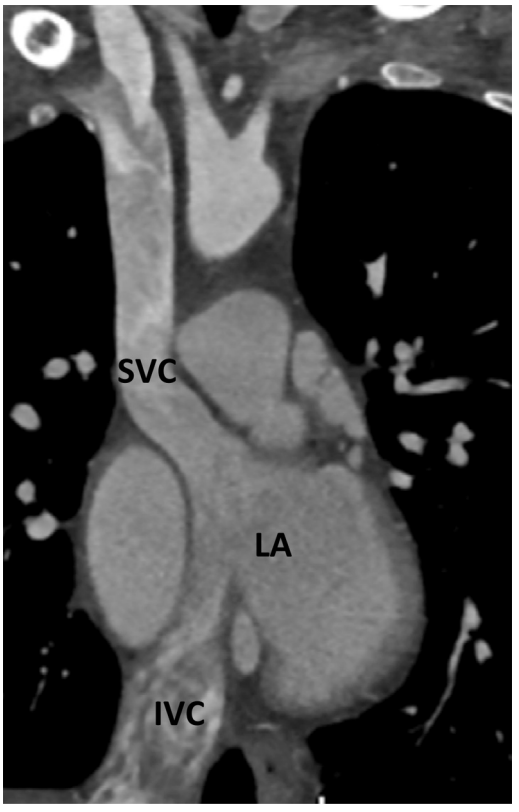


Fig. 6 – This coronal 2-dimensional image shows a patent superior vena cava and inferior vena cava baffle to the left atrium in an adult patient after the atrial switch procedure.

placed from the right ventricle to the branch PAs (Fig. 13). For patients with a large VSD, it is critical to preoperatively estimate the amount of right ventricle taken up by the VSD patch. If the right ventricle size is predicted to be inadequate with VSD closure, a Glenn procedure (superior vena cava [SVC]–PA anastomosis) can be performed at the time of the VSD closure. A more recent procedure for complex transposition with VSD and pulmonary stenosis is the Nikaidoh procedure. In this procedure the aorta is translocated and moved medially (closer to the left ventricle), the VSD is closed, and the RVOT is either reconstructed or patched, or a homograft is placed.

4.5. *Postoperative imaging*

Complete postoperative evaluation of complex transposition includes assessment of the RVOT and branch PAs, assessment of the left ventricle to aortic pathway, assessment of the proximal aorta if reconstructed, and evaluation of the coronary arteries if they were manipulated as part of the procedure. Patients with initial anatomy of a small outlet portion of the VSD are at highest risk of LVOT obstruction after VSD closure. Medial translocation of the aorta in the Nikaidoh procedure puts the right coronary artery at risk long term[30,31] (Fig. 14). Transcatheter placement of a pulmonary conduit is an option after previous conduit placement, and attention to the coronary artery position relative to the RVOT is essential to avoid coronary compression during transcatheter valve deployment (similar to TOF described earlier). Patients who have retained pulmonary valve leaflets with PA ligation (earlier surgical technique) at the time of a Rastelli procedure are at risk of systemic emboli, and this finding should be specifically mentioned to the referring cardiologist.

4.6. *Corrected transposition, physiologically corrected transposition, levo transposition*

Corrected transposition is characterized by the presence of both AV and ventricular–arterial discordance. This is the rare complex lesion that may have initial presentation in adulthood. Corrected transposition may be present in situs solitus or in situs inversus and is commonly referred to as physiologically corrected transposition or levo-TGA.[32] The right atrium is connected to the mitral valve and left ventricle, which connects to the PA. The pulmonary venous atrium connects to a tricuspid valve and right ventricle, which then connects to the aorta. The anatomic right ventricle faces systemic pressure and eventually develops systolic failure or tricuspid regurgitation in most patients[33,34] (Fig. 15).

4.7. *Surgical intervention*

In patients without systemic RV dysfunction, significant tricuspid regurgitation, or other cardiac defects, the timing and type of repair is controversial. Options include no intervention or physiological repair to correct associated abnormalities (atrial septal defect/VSD closure, pulmonary stenosis relief, tricuspid valve surgery). The double switch procedure, the Senning-Rastelli, or the Fontan procedure have also been performed for palliation of corrected transposition.[33] The double switch consists of a concomitant atrial

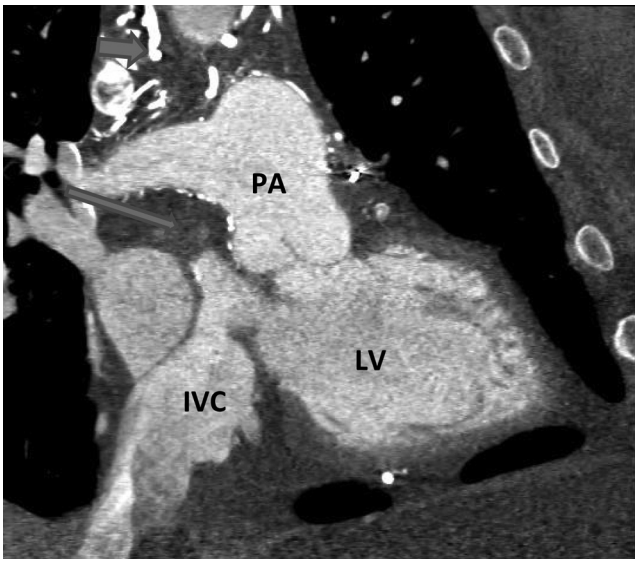


Fig. 7 – This is a 2-dimensional coronal image of an obstructed superior vena cava baffle (*long arrow*) after the atrial switch procedure. Note the contrast in the superior mediastinum and the contrast in the inferior vena cava from draining collaterals.

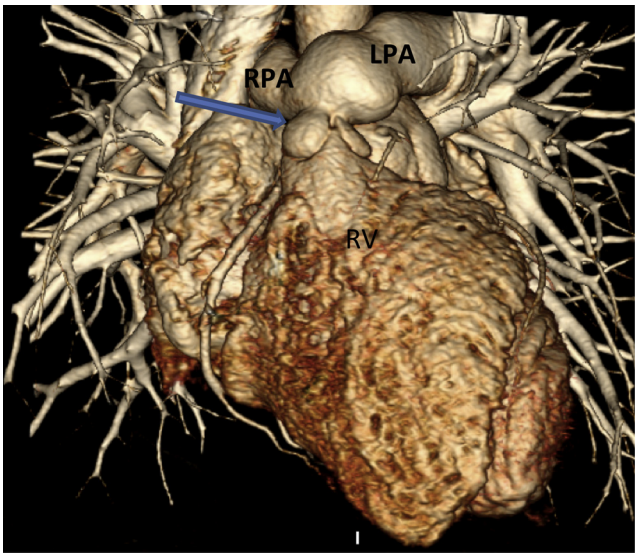


Fig. 9 – This is a 3-dimensional reconstruction of the great arteries after the arterial switch operation. Note the supravalvar pulmonary stenosis in the neo-pulmonary root (*long arrow*).

baffle and arterial switch. It is commonly performed in 2 stages when no pulmonary stenosis or VSD results in LV systolic pressure. The first stage consists of PA banding to increase subpulmonary LV afterload and wall thickness. The second stage consists of PA band removal, arterial switch, and atrial switch.[34–36] Preoperative evaluation for this possible intervention includes visualization of the coronary arteries and great artery relationships, quantification of tricuspid regurgitation when present, or assessment of LV myocardial function and mass after PA banding. Similar to isolated AV discordance, the performance of a Rastelli type of procedure for the arterial switch portion of the operation will depend on

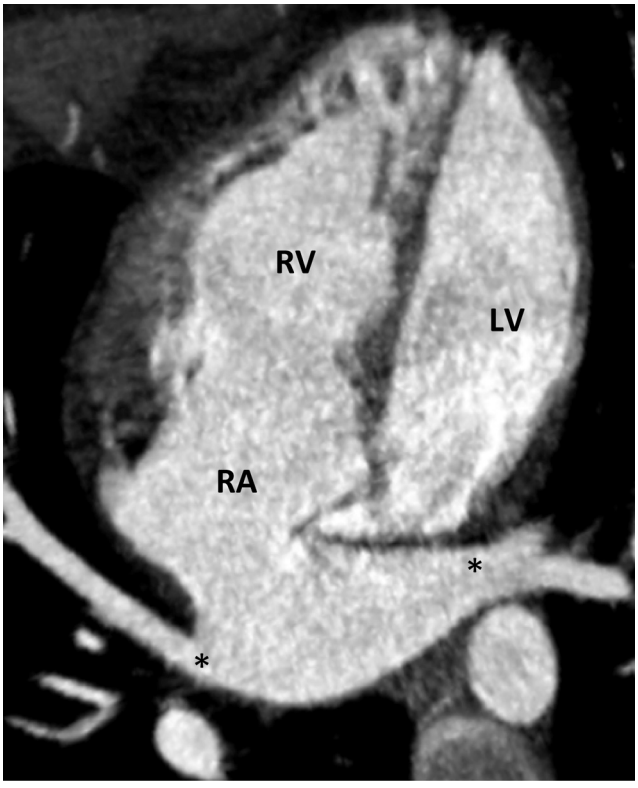


Fig. 8 – A 2-dimensionl 4-chamber view shows a patent pulmonary venous baffle to the right atrium. Note the right ventricular hypertrophy and the thin-walled subpulmonary left ventricle. Pulmonary veins are noted by an *asterisk*.

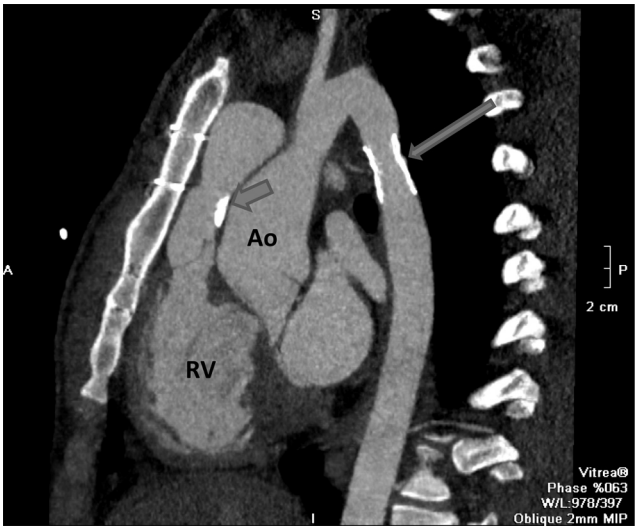


Fig. 10 – This is a 2-dimensional sagittal image in a patient after the arterial switch operation. Note the right ventricular outflow tract directly anterior to the aorta with calcification in the area of prior patch augmentation (*short arrow*). The neo-aortic root is dilated, and a stent is seen in the proximal descending aorta after angioplasty and stent placement for concomitant aortic coarctation (*long arrow*).

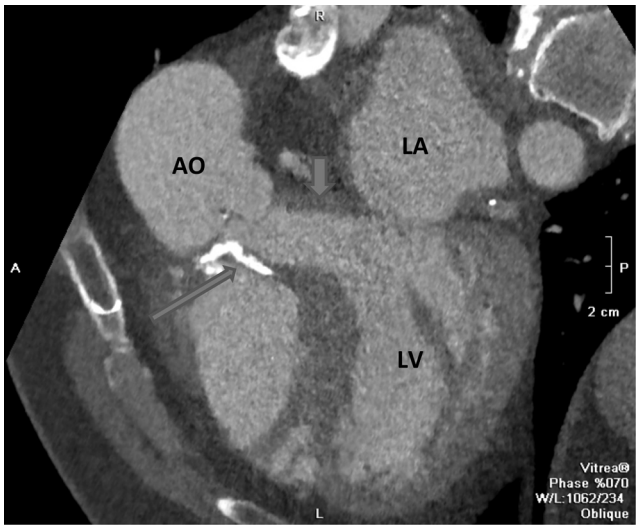


Fig. 11 – This 2-dimensional image shows the left ventricle-to-aortic pathway after the Rastelli procedure. Note the calcified ventricular septal defect patch (*long arrow*) and the lack of aortomitral continuity (*short arrow*), consistent with double outlet right ventricle.

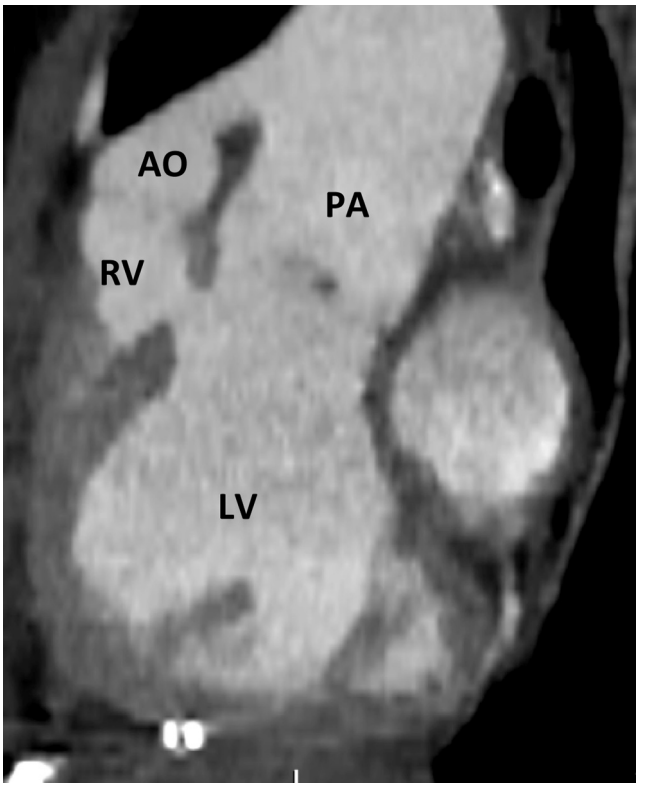


Fig. 12 – This is a 2-dimensional image of a proximal aortopulmonary anastomosis (Damus-Kaye-Stancel procedure) in a patient with a malposed aorta coming from the small right ventricular outlet chamber.

the suitability of the PA to function as a neo-aortic valve and the geometry of the LV outflow relationship.

4.8. *Postoperative imaging*

All aspects of both the atrial and arterial switch, or the atrial switch and Rastelli procedure need to be evaluated (see earlier descriptions). In these complex cases, the systemic and pulmonary venous baffles, biventricular systolic function, biventricular outflow tracts, and coronary arteries need to be evaluated.

4.9. *Imaging considerations*

All forms of TGA need complete evaluation of both the right and left heart with the use of a biventricular injection protocol. A retrospectively ECG-gated scan with pulsed radiation may be used to determine biventricular systolic function, with calculation of valvular regurgitant fraction from stroke volume differences. After the arterial switch operation, β-blockade may be helpful to decrease heart rate and to optimize coronary artery imaging. The scan range will depend on the area of concern, but it usually includes the proximal neo-pulmonary root, neo-aortic root, and branch PAs.

5. Single ventricle physiology

Single ventricle heart disease (Table 5) is associated with a variety of cardiac lesions such as tricuspid atresia, mitral atresia, double inlet left ventricle, unbalanced AV canal defect, and hypoplastic left heart syndrome.[37] In the current era, most patients who undergo surgical palliation for single ventricular physiology are expected to reach adulthood.[37]

5.1. *Surgical palliation for single ventricle heart disease*

The hemodynamic goal of single ventricle palliation is to allow passive flow of systemic venous blood to the lungs and to allow the single ventricle to pump oxygenated pulmonary

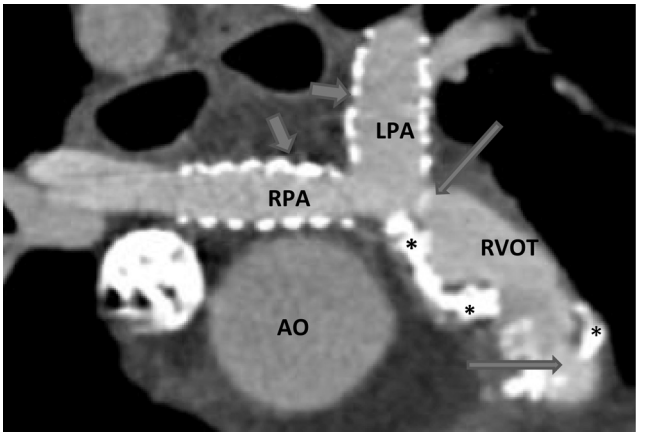


Fig. 13 – This 2-dimensional image shows stents in the proximal pulmonary arteries (*short arrows*) and narrowing at the proximal and distal end (*long arrows*) of a calcified (*) right ventricle-to-pulmonary artery conduit.

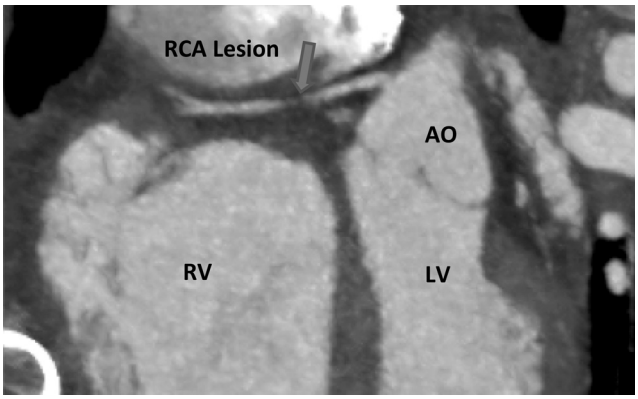


Fig. 14 – A lesion (*arrow*) in the proximal portion of the right coronary artery is seen after the Nikaidoh procedure.

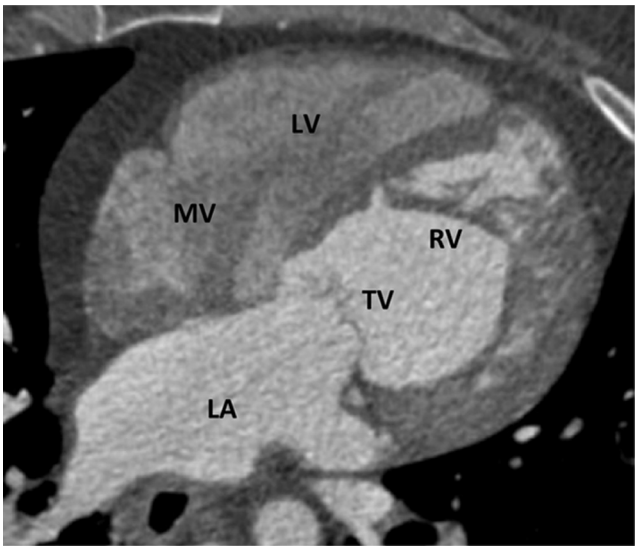


Fig. 15 – The left atrium empties into a hypertrophied systemic right ventricle through the tricuspid vale in a patient with physiologically corrected transposition.

venous blood to the body. Surgical and catheter-based palliation typically occurs in 3 stages. The first stage usually consists of aortic arch reconstruction, atrial septectomy, and an aortopulmonary shunt (Fig. 16) or right ventricle–PA conduit (Sano procedure; Fig. 17) for hypoplastic left heart syndrome. For patients with hypoplastic right-sided structures, an atrial septectomy and aortopulmonary shunt are usually placed, with ligation of the PA if there is unrestricted pulmonary blood flow. The second stage of surgical palliation is similar for all lesions and consists of shunt or conduit takedown, and anastomosis of the SVC to the PA. This procedure is commonly called the Glenn procedure or the superior cavopulmonary connection (Fig. 18). The third stage of palliation is called the Fontan procedure and consists of re-routing the inferior vena cava (IVC) flow to the PAs, effectively allowing passive venous flow to the lungs and separation of the venous and systemic circulation. There have been many modifications of the Fontan procedure since its initial description. The 2 types commonly used in the current era are the lateral tunnel Fontan procedure that baffles the IVC to the PAs through the atrium (Fig. 19) and the extracardiac Fontan procedure that uses a Gore-Tex (Gore) tube from the IVC to the pulmonary circulation (Fig. 20).

5.2. *Postoperative imaging in patients with single ventricle heart disease*

Patients with single ventricle physiology require serial diagnostic evaluation of anatomic connections, ventricular function, and valvular complications throughout their lives.[38–40] PA narrowing, decreased ventricular function, and valvular regurgitation are all predictors of poor long-term outcome.[41] Echocardiography is insufficient for interstage and post-Fontan evaluation of single ventricle heart disease.[42] Cardiac catheterization is the historical standard for interstage evaluation, but it may no longer be necessary in low-risk patients with good systolic function and no valvular regurgitation.[7,43] A completely noninvasive evaluation with the use of MRI has been proposed by some centers, but many patients have contraindications to this modality.[44] In a study of 137 patients with single ventricle heart disease followed for a median of 5.6 years, 12% had pacemaker placement, and interventional catheterizations were performed in 52% of patients, often with the placement of metal devices that degrade MRI image quality.[45,46] Late complications of single ventricle palliation are common and include hepatic failure, heart failure, protein-losing enteropathy, and plastic bronchitis. The long-term outcome for adults with palliated single ventricle heart disease is not known. This is the first generation in whom most survive into adulthood.

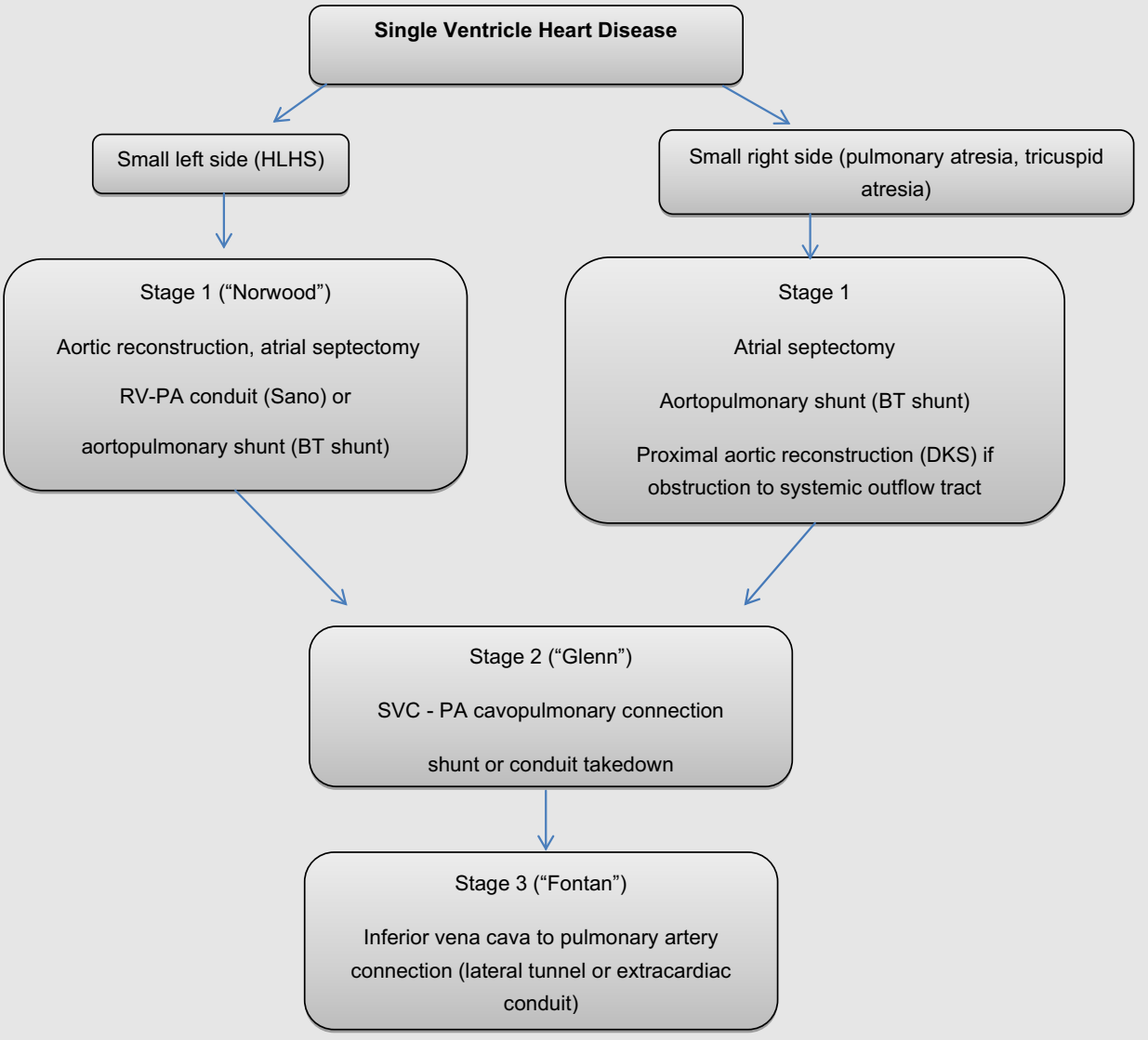
Cardiac CTA has been described for interstage evaluation of single ventricle heart disease but has not yet been described in a large series of patients. Venous collaterals that cause cyanosis after the Glenn procedure are well seen by CTA with the use of a delayed scan.

Fontan opacification by CT imaging is frequently difficult to achieve (Fig. 21). The venous filling depends on cardiac output, and differences in contrast timing are based on the type of Fontan procedure, the presence of valvular regurgitation or ventricular dysfunction, and elevated end-diastolic and PA pressures. In older types of atriopulmonary Fontan procedures, contrast will go to the atrial portion of the Fontan pathway before circulating to the PAs (Fig. 22). CTA is a useful method for detecting pulmonary embolism and plastic bronchitis in patients with single ventricle heart disease.[47–49] Branch PA narrowing, Fontan pathway narrowing, and ventricular function can be assessed with CT. Valvular regurgitation cannot be estimated by CT, given that only one ventricle is present, preventing a comparison of stroke volume differences. Fenestration closure devices are well visualized with CTA (Figs. 19 and 21), as is thrombus on a delayed scan.

5.3. *Imaging considerations*

Because Fontan opacification can be difficult, we use a 2-phase injection protocol to opacify the IVC and Fontan pathway with

Table 5 – Single ventricle heart disease: Common interventions and residual hemodynamic lesions.



Procedure	Common residual hemodynamic lesions
Norwood	Shunt, conduit, branch PA stenosis or clot
	Distal aortic reconstruction to descending aorta narrowing (coarctation)
	Systemic venous occlusion or collaterals, and aortic/bronchial collaterals
	Single ventricle dysfunction or valvular regurgitation (regurgitation cannot be determined from CT)
Glenn	SVC to PA narrowing, branch PA stenosis or clot
	Systemic venous occlusion collaterals and aortic/bronchial collaterals
	Single ventricle dysfunction or valvular regurgitation (regurgitation cannot be determined from CT)
Fontan	SVC to PA, IVC to PA, or branch PA narrowing
	Atrial dilation (most commonly in older atriopulmonary types of Fontan procedure)
	Systemic venous occlusion, collaterals, or aortic bronchial collaterals
	Clot within the Fontan system, pulmonary embolism
	Ventricular to aortic obstruction for complex transposition with aorta from an outlet chamber
	Ventricular dysfunction or valvular regurgitation (regurgitation cannot be determined from CT)
	Recurrent arch obstruction
	Fenestration or leak in lateral tunnel Fontan pathway

(*continued on next page*)

Table 5 (*continued*)

	Hepatic disease Plastic bronchitis Atrial Arrhythmia Protein losing enteropathy
Suggested scan modifications	• Automatic trigger in the aorta, or manual trigger; extend monitoring sequence because aortic opacification will be later than normal because of passive flow into lungs from an upper extremity injection. • Extend scan range to include the superior vena cava and the inferior vena cava before Fontan insertion. • Fontan visualization (visualization of inferior portion of pathway is difficult). ○ Give 50% contrast before arterial imaging so venous and arterial structures are opacified on a single acquisition, or perform late venous acquisition (allow at least 1 minute between contrast administration and venous image acquisition). ○ Plan for additional delay in venous opacification if poor single ventricular function, significant AV valve regurgitation, or atriopulmonary Fontan connection. • Restate trigger function scan of aortic opacification.

AV, atrioventricular; BT, Blalock-Taussig; DKS, Damus-Kaye-Stancel; HLHS, hypoplastic left heart syndrome; IVC, inferior vena cava; PA, pulmonary artery; SVC, superior vena cava.

the arterial image acquisition or a delayed scan that will allow for venous recirculation. We divide the contrast load and give approximately half before imaging, and the remainder of the contrast is given as a bolus with image acquisition timed to aortic opacification. Dual injections in both upper and lower extremities have been described, but this results in swirling of contrast and unopacified hepatic venous flow into the Fontan circuit, making the diagnosis of thrombus difficult. It is not recommended in our opinion.[50] An atriopulmonary Fontan pathway tends to opacify later than a direct SVC–PA connection, and image acquisition in these patients may be delayed an additional 20 to 30 seconds. Ventricular function can be quantified with a retrospective ECG-gated scan with multiphase reconstruction. In addition, the SVC flow sometimes is directed primarily to one PA. Because the contralateral PA may then be best seen on a delayed scan, we routinely use a delayed scan for this indication. Two scans of the Fontan pathway performed at different time points can be helpful to differentiate clot from the hepatic venous unopacified flow seen on the early scan.

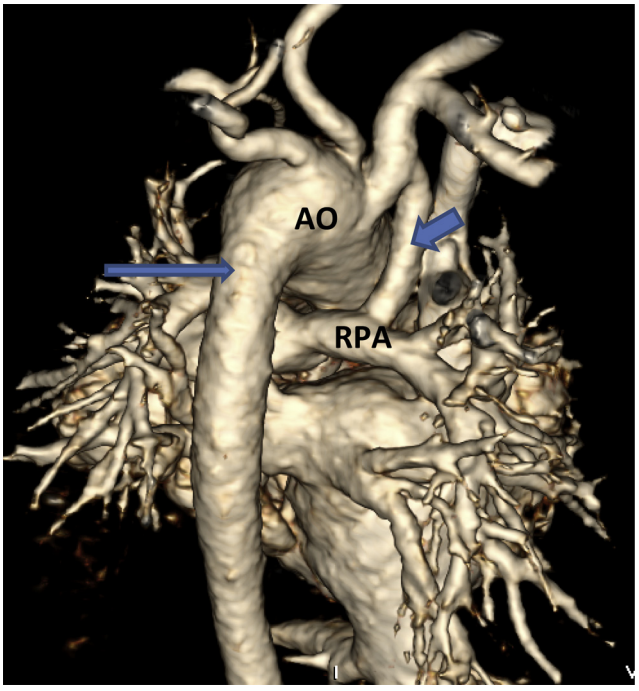


Fig. 16 – This posterior view shows an aortopulmonary shunt (*short arrow*) arising from the base of the innominate artery and inserting into the right pulmonary artery. Note the distal Norwood anastomosis to the proximal descending aorta is widely patent (*long arrow*).

6. An approach to image acquisition in the patient with CHD

6.1. *Vascular access*

Peripheral intravenous (IV), percutaneous indwelling central venous catheter line, umbilical venous line, central line, and indwelling venous catheters have been used for contrast

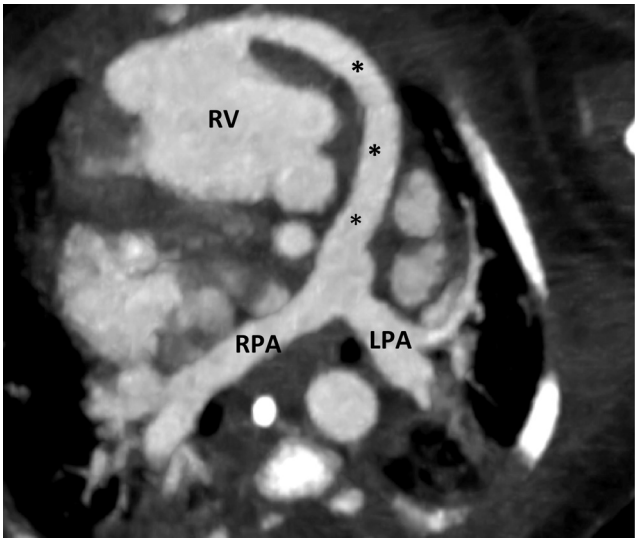


Fig. 17 – This 2-dimensional axial image shows a right ventricle-to-pulmonary artery conduit (Sano procedure; **) from the anterior right ventricle to the branch pulmonary arteries in a patient after first-stage single ventricle palliation.

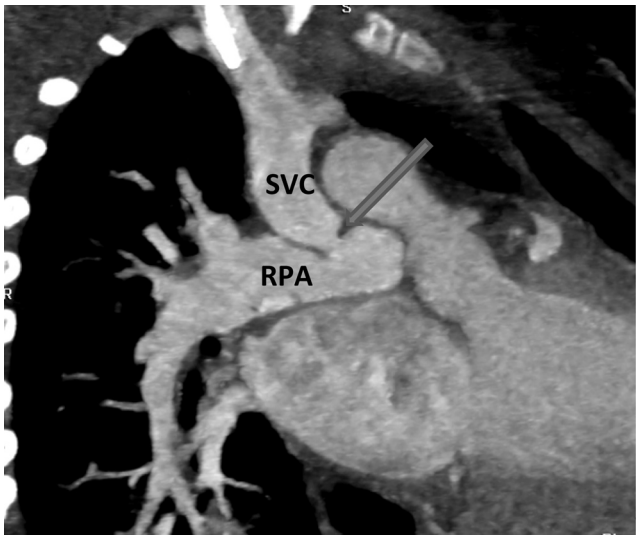


Fig. 18 – This 2-dimensional coronal oblique projection shows narrowing at the superior vena cava-to-pulmonary artery anastomosis (*arrow*) after the Glenn procedure performed for second-stage palliation for single ventricle hart disease.

injection for congenital cardiac CT scans.[51,52] Because of the high incidence of intracardiac shunting in patients with complex CHD, particular care to avoid any air bubbles in the injection is important, because it may result in pulmonary or systemic arterial embolus.

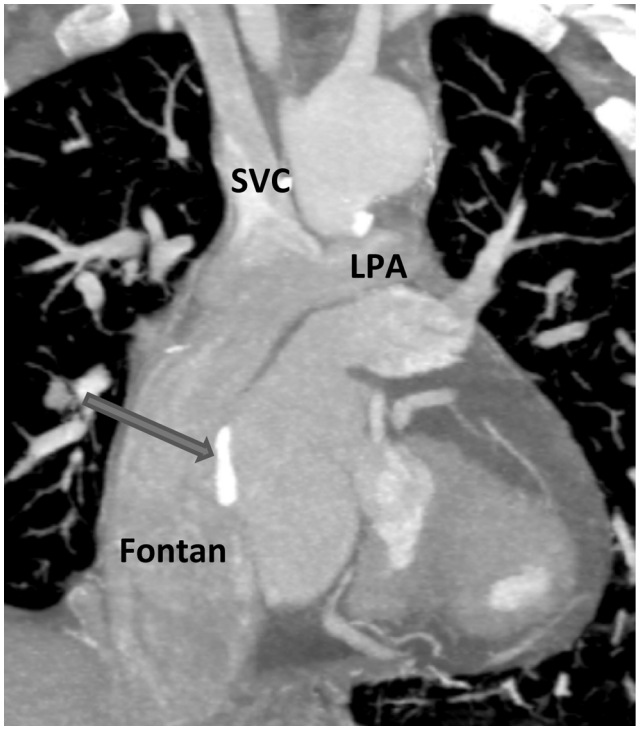


Fig. 19 – This coronal projection shows the superior vena cava and lateral tunnel Fontan connection to the branch pulmonary arteries. Note the calcified fenestration closure patch (*arrow*).

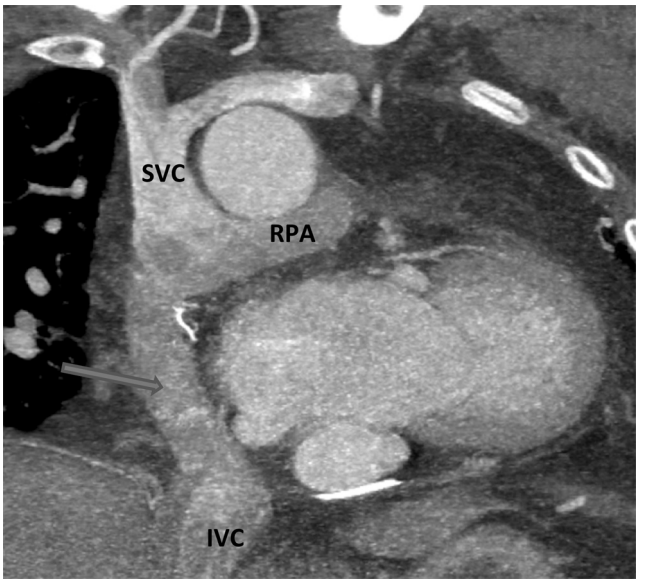


Fig. 20 – This 2-dimensional image shows late venous opacification of an extracardiac Fontan pathway (*arrow*) in an adult patient.

6.2. *IV placement*

The optimal site for peripheral IV placement will vary by cardiac lesion and indication. If systemic venous return is normal, and the indication is pulmonary or arterial angiography, the IV catheter may be placed in any extremity. For patients with anomalous systemic venous drainage such as an interrupted IVC or bilateral SVC, IV placement may need to be in a certain location to optimize contrast opacification in the structures of interest. If a venous occlusion is present, injection may result in cardiac opacification via collateral vessels which makes the timing of image acquisition difficult. In addition, thrombus or occlusion can be difficult to diagnose

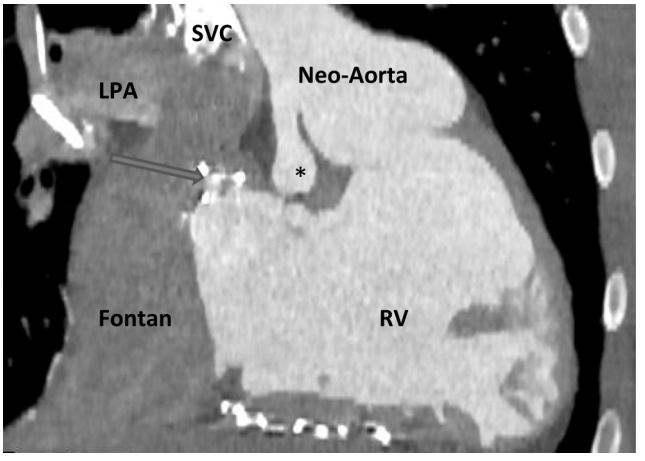


Fig. 21 – This arterial acquisition shows the superior vena cava filling from an upper extremity injection and an unopacified lateral tunnel Fontan pathway. A Fontan closure device is seen (*arrow*) and the reconstructed neo-aorta.

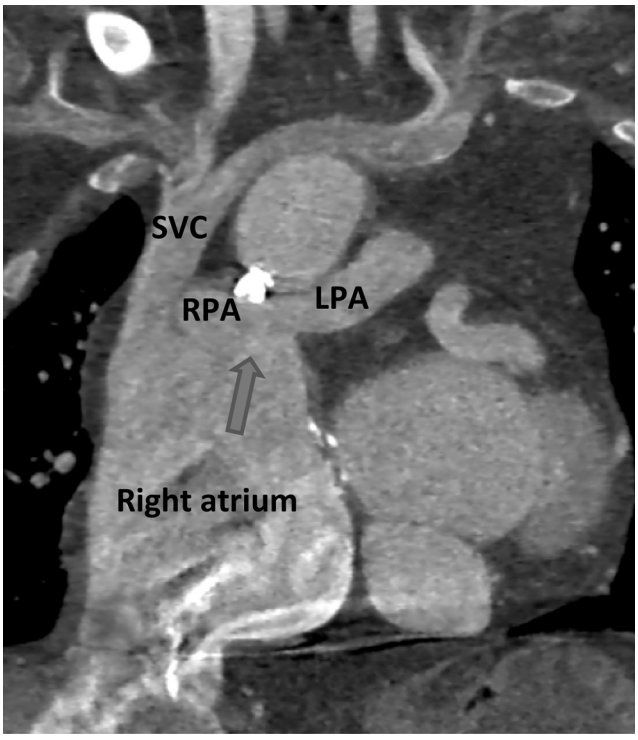


Fig. 22 – This image shows an atriopulmonary Fontan pathway with a patent connection between the superior aspect of the atrium and the main pulmonary artery (*arrow*). The very proximal right pulmonary artery and proximal-to-mid left pulmonary artery are seen in this projection.

in the presence of contrast swirling or beam hardening artifact. If venous occlusion is suspected, recirculation is preferred and the extremity draining the suspected site of venous occlusion should not be used for injection.

6.3. *IV gauge*

Power injectors are safely used in pediatric patients with IV gauge from 24 to 18, depending on patient size. The rate of injection and the pounds per square inch (psi) should be adjusted for the IV gauge. A 22-gauge IV catheter or larger is preferred, but safe injection through a 24-gauge catheter is reported for neonates at low flow rates of 1 to 1.5 mL/s and <100 psi.[52] When using a power injector through a small-gauge IV catheter, a saline test injection with careful observation of the injection site and psi value should be used before contrast injection. A pressure-limited injection to a maximum of 25 psi has been used safely in central catheters with acceptable opacification in patients <30 kg. Unfortunately, many power injectors have a lower pressure limit of 50 psi, which is higher than recommended for most indwelling catheters.[51]

6.4. *Injection protocols*

The total contrast volume used for pediatric CT angiography is typically 1 to 2 mL/kg until the adult size and contrast rates are achieved. The time between scan initiation and image acquisition is as long as 4 seconds for the highest pitch scan modes. Estimation of where the contrast will be in 4 seconds (10 heartbeats at a heart rate of 150 beats/min) can be problematic. Mixing contrast and saline to lengthen the injection increases the chance of optimal contrast injection at the time of image acquisition.

A biventricular injection protocol (2-phase contrast injection with saline flush) is most commonly used for pulmonary and aortic angiography or when stroke volume differences will be used to determine regurgitant fraction or to quantify shunting. Approximately half the injection is given at the usual rate for patient size and IV gauge, and the remainder is given at a slower rate so the right heart will maintain opacification during image acquisition. When venous and arterial anatomies are both required, we give a percentage of the contrast early and time the image acquisition to the aortic injection. This usually results in simultaneous venous and arterial opacification in a single acquisition. For patients with intracardiac mixing, a longer and slower injection with image acquisition at the end of injection often allows venous and arterial opacifications on the same scan.

6.5. *Scan trigger*

6.5.1. *Bolus tracking*

Bolus tracking is reliable for pulmonary and arterial angiography when systemic and pulmonary venous return is normal, and there is no intracardiac shunting.

6.5.2. *Timing bolus*

A timing bolus can be used when abnormal systemic or pulmonary venous return or intracardiac shunting makes optimal scan acquisition difficult to predict. This method uses some of the total contrast available for the angiogram and results in additional radiation exposure, and so is not often used in young patients.

6.5.3. *Manual*

The scan may be manually triggered on the basis of the visual estimate of optimal contrast in the region of interest on a monitoring sequence. This is a reliable method of scan acquisition when there are multiple levels of shunting or when unexpected venous anomalies or venous occlusion might preclude precise contrast timing.

6.6. *Scan sequence*

6.6.1. *Anatomic imaging*

Only a single data set is needed for an anatomic survey. For dual-source scanners, a prospectively ECG-triggered high-pitch mode should be used, and for volumetric scanners the entire scan range should be captured in a single heartbeat. On older generation scanners nongated modes may be used, but systolic motion or respiratory artifact may be present.

6.6.2. *Functional imaging*

For estimation of ventricular function or valvular regurgitation with the use of stroke volume differences, a retrospectively ECG-gated or prospectively ECG-triggered scan

with pulsed radiation and minimal padding can be used for multiphase reconstruction. If fine detail is not needed, the collimator may be increased to allow for a further decrease in dose. The scan data should be reconstructed in a short-axis stack to provide accurate biventricular volume measurement.

If both functional imaging and an anatomic survey are needed, the approach will depend on the scanner available. A functional scan with a limited range may be performed, followed by a high-pitch or volumetric scan with an extended scan range on the newest generation scanners. On older scanner platforms an extended scan range ECG-triggered scan with multiphase reconstruction may give less dose than a limited scan range function study followed by an anatomic scan. The choice will vary, depending on the technology available at each institution.

7. Conclusion

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 CHD who need advanced imaging when MRI is considered contraindicated, high risk, or unlikely to be diagnostic because of metal artifacts. Meticulous attention to all of the details of scan acquisition and radiation dose-reduction techniques is required to maintain diagnostic image quality while minimizing risk. As this patient population grows and modern technology becomes more accessible, CTA will play an increasingly important role in the diagnosis and management of congenital heart lesions.

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