American College of Radiology
ACR Appropriateness Criteria®
Congenital or Acquired Heart Disease

Variant 1: **Child or adult. Repaired tetralogy of Fallot or pulmonary valve stenosis with concern for pulmonary valve dysfunction or branch pulmonary artery stenosis. Incomplete or inadequate assessment of cardiovascular morphology and function after transthoracic echocardiography. Next imaging study.**

Procedure	Appropriateness Category	Relative Radiation Level
Radiography chest	Usually Appropriate	☢
MRA chest without and with IV contrast	Usually Appropriate	O
MRI heart function and morphology without and with IV contrast	Usually Appropriate	O
MRI heart function and morphology without IV contrast	Usually Appropriate	O
CTA chest with IV contrast	Usually Appropriate	☢☢☢☢
CT heart function and morphology with IV contrast	Usually Appropriate	☢☢☢☢
US echocardiography transesophageal	May Be Appropriate	O
Arteriography coronary with ventriculography	May Be Appropriate	☢☢☢☢
Arteriography pulmonary	May Be Appropriate	☢☢☢☢
MRA chest without IV contrast	May Be Appropriate	O
CTA coronary arteries with IV contrast	May Be Appropriate	☢☢☢☢
Perfusion scan lung	May Be Appropriate	☢☢☢☢
MRA abdomen without and with IV contrast	Usually Not Appropriate	O
MRA abdomen without IV contrast	Usually Not Appropriate	O
MRA neck without and with IV contrast	Usually Not Appropriate	O
MRA neck without IV contrast	Usually Not Appropriate	O
MRI heart function with stress without and with IV contrast	Usually Not Appropriate	O
MRI heart function with stress without IV contrast	Usually Not Appropriate	O
FDG-PET/CT heart	Usually Not Appropriate	☢☢☢☢
SPECT or SPECT/CT MPI rest and stress	Usually Not Appropriate	☢☢☢☢☢

<u>Variant 2:</u> **Child or adult. Transposition of the great arteries after atrial switch. Incomplete or inadequate assessment of cardiovascular morphology and function after transthoracic echocardiography. Next imaging study.**

Procedure	Appropriateness Category	Relative Radiation Level
MRA chest without and with IV contrast	Usually Appropriate	O
MRA chest without IV contrast	Usually Appropriate	O
MRI heart function and morphology without and with IV contrast	Usually Appropriate	O
MRI heart function and morphology without IV contrast	Usually Appropriate	O
CTA chest with IV contrast	Usually Appropriate	☢☢☢☢
CT heart function and morphology with IV contrast	Usually Appropriate	☢☢☢☢
US echocardiography transesophageal	May Be Appropriate	O
Radiography chest	May Be Appropriate	☢
Arteriography pulmonary	May Be Appropriate	☢☢☢☢
CTA coronary arteries with IV contrast	May Be Appropriate	☢☢☢☢
Arteriography coronary with ventriculography	Usually Not Appropriate	☢☢☢☢
MRA abdomen without and with IV contrast	Usually Not Appropriate	O
MRA abdomen without IV contrast	Usually Not Appropriate	O
MRA neck without and with IV contrast	Usually Not Appropriate	O
MRA neck without IV contrast	Usually Not Appropriate	O
MRI heart function with stress without and with IV contrast	Usually Not Appropriate	O
MRI heart function with stress without IV contrast	Usually Not Appropriate	O
FDG-PET/CT heart	Usually Not Appropriate	☢☢☢☢
SPECT or SPECT/CT MPI rest and stress	Usually Not Appropriate	☢☢☢☢☢

Variant 3: **Child or adult. Transposition of the great arteries after arterial switch. Incomplete or inadequate assessment of cardiovascular morphology and function after transthoracic echocardiography. Next imaging study.**

Procedure	Appropriateness Category	Relative Radiation Level
Radiography chest	Usually Appropriate	☢
MRA chest without and with IV contrast	Usually Appropriate	O
MRA chest without IV contrast	Usually Appropriate	O
MRI heart function and morphology without and with IV contrast	Usually Appropriate	O
MRI heart function and morphology without IV contrast	Usually Appropriate	O
MRI heart function with stress without and with IV contrast	Usually Appropriate	O
CTA chest with IV contrast	Usually Appropriate	☢☢☢☢
CT heart function and morphology with IV contrast	Usually Appropriate	☢☢☢☢
MRI heart function with stress without IV contrast	May Be Appropriate (Disagreement)	O
CTA coronary arteries with IV contrast	May Be Appropriate	☢☢☢☢
SPECT or SPECT/CT MPI rest and stress	May Be Appropriate	☢☢☢☢☢
US echocardiography transesophageal	Usually Not Appropriate	O
Arteriography coronary with ventriculography	Usually Not Appropriate	☢☢☢☢
Arteriography pulmonary	Usually Not Appropriate	☢☢☢☢
MRA abdomen without and with IV contrast	Usually Not Appropriate	O
MRA abdomen without IV contrast	Usually Not Appropriate	O
MRA neck without and with IV contrast	Usually Not Appropriate	O
MRA neck without IV contrast	Usually Not Appropriate	O
FDG-PET/CT heart	Usually Not Appropriate	☢☢☢☢

Variant 4: **Child. Suspected or confirmed congenital or acquired coronary artery abnormality. Incomplete or inadequate assessment of coronary morphology after transthoracic echocardiography. Next imaging study.**

Procedure	Appropriateness Category	Relative Radiation Level
MRA chest without and with IV contrast	Usually Appropriate	O
MRA chest without IV contrast	Usually Appropriate	O
MRI heart function and morphology without and with IV contrast	Usually Appropriate	O
MRI heart function and morphology without IV contrast	Usually Appropriate	O
CTA coronary arteries with IV contrast	Usually Appropriate	☢☢☢☢
Arteriography coronary with ventriculography	May Be Appropriate	☢☢☢☢
MRA abdomen without and with IV contrast	May Be Appropriate	O
MRI heart function with stress without and with IV contrast	May Be Appropriate	O
MRI heart function with stress without IV contrast	May Be Appropriate	O
CTA chest with IV contrast	May Be Appropriate	☢☢☢☢
CT heart function and morphology with IV contrast	May Be Appropriate	☢☢☢☢
SPECT or SPECT/CT MPI rest and stress	May Be Appropriate (Disagreement)	☢☢☢☢☢
US echocardiography transesophageal	Usually Not Appropriate	O
Radiography chest	Usually Not Appropriate	☢
Arteriography pulmonary	Usually Not Appropriate	☢☢☢☢
MRA abdomen without IV contrast	Usually Not Appropriate	O
MRA neck without and with IV contrast	Usually Not Appropriate	O
MRA neck without IV contrast	Usually Not Appropriate	O
FDG-PET/CT heart	Usually Not Appropriate	☢☢☢☢

Variant 5: **Child. Known single ventricle physiology. Preoperative evaluation for stage 2 single ventricle palliation. Incomplete or inadequate assessment of cardiovascular morphology and function after transthoracic echocardiography. Next imaging study.**

Procedure	Appropriateness Category	Relative Radiation Level
Arteriography coronary with ventriculography	Usually Appropriate	☢☢☢☢
Arteriography pulmonary	Usually Appropriate	☢☢☢☢
MRA chest without and with IV contrast	Usually Appropriate	O
MRA chest without IV contrast	Usually Appropriate	O
MRI heart function and morphology without and with IV contrast	Usually Appropriate	O
MRI heart function and morphology without IV contrast	Usually Appropriate	O
CTA chest with IV contrast	Usually Appropriate	☢☢☢☢
CT heart function and morphology with IV contrast	Usually Appropriate	☢☢☢☢
US echocardiography transesophageal	May Be Appropriate	O
Radiography chest	Usually Not Appropriate	☢
MRA abdomen without and with IV contrast	Usually Not Appropriate	O
MRA abdomen without IV contrast	Usually Not Appropriate	O
MRA neck without and with IV contrast	Usually Not Appropriate	O
MRA neck without IV contrast	Usually Not Appropriate	O
MRI heart function with stress without and with IV contrast	Usually Not Appropriate	O
MRI heart function with stress without IV contrast	Usually Not Appropriate	O
CTA coronary arteries with IV contrast	Usually Not Appropriate	☢☢☢☢
FDG-PET/CT heart	Usually Not Appropriate	☢☢☢☢
SPECT or SPECT/CT MPI rest and stress	Usually Not Appropriate	☢☢☢☢☢

Variant 6: **Child. Known single ventricle physiology. Preoperative evaluation for stage 3 single ventricle palliation (total cavopulmonary connection). Incomplete or inadequate assessment of cardiovascular morphology and function after transthoracic echocardiography. Next imaging study.**

Procedure	Appropriateness Category	Relative Radiation Level
Arteriography coronary with ventriculography	Usually Appropriate	☢☢☢☢
Arteriography pulmonary	Usually Appropriate	☢☢☢☢
MRA chest without and with IV contrast	Usually Appropriate	O
MRI heart function and morphology without and with IV contrast	Usually Appropriate	O
MRI heart function and morphology without IV contrast	Usually Appropriate	O
CTA chest with IV contrast	Usually Appropriate	☢☢☢☢
CT heart function and morphology with IV contrast	Usually Appropriate	☢☢☢☢
US echocardiography transesophageal	May Be Appropriate	O
Radiography chest	May Be Appropriate	☢
MRA chest without IV contrast	May Be Appropriate	O
MRA abdomen without and with IV contrast	Usually Not Appropriate	O
MRA abdomen without IV contrast	Usually Not Appropriate	O
MRA neck without and with IV contrast	Usually Not Appropriate	O
MRA neck without IV contrast	Usually Not Appropriate	O
MRI heart function with stress without and with IV contrast	Usually Not Appropriate	O
MRI heart function with stress without IV contrast	Usually Not Appropriate	O
CTA coronary arteries with IV contrast	Usually Not Appropriate	☢☢☢☢
FDG-PET/CT heart	Usually Not Appropriate	☢☢☢☢
SPECT or SPECT/CT MPI rest and stress	Usually Not Appropriate	☢☢☢☢☢

Variant 7: **Child or adult. Known single ventricle physiology. Postoperative evaluation after stage 3 single ventricle palliation (total cavopulmonary connection). Incomplete or inadequate assessment of cardiovascular morphology and function after transthoracic echocardiography. Next imaging study.**

Procedure	Appropriateness Category	Relative Radiation Level
MRA chest without and with IV contrast	Usually Appropriate	O
MRA chest without IV contrast	Usually Appropriate	O
MRI heart function and morphology without and with IV contrast	Usually Appropriate	O
MRI heart function and morphology without IV contrast	Usually Appropriate	O
CTA chest with IV contrast	Usually Appropriate	☢☢☢☢
CT heart function and morphology with IV contrast	Usually Appropriate	☢☢☢☢
US echocardiography transesophageal	May Be Appropriate	O
Radiography chest	May Be Appropriate	☢
Arteriography coronary with ventriculography	May Be Appropriate	☢☢☢☢
Arteriography pulmonary	May Be Appropriate	☢☢☢☢
CTA coronary arteries with IV contrast	May Be Appropriate	☢☢☢☢
MRA abdomen without and with IV contrast	Usually Not Appropriate	O
MRA abdomen without IV contrast	Usually Not Appropriate	O
MRA neck without and with IV contrast	Usually Not Appropriate	O
MRA neck without IV contrast	Usually Not Appropriate	O
MRI heart function with stress without and with IV contrast	Usually Not Appropriate	O
MRI heart function with stress without IV contrast	Usually Not Appropriate	O
FDG-PET/CT heart	Usually Not Appropriate	☢☢☢☢
SPECT or SPECT/CT MPI rest and stress	Usually Not Appropriate	☢☢☢☢☢

Variant 8: **Child or adult. Known or suspected anomalous pulmonary venous return with inadequate evaluation after transthoracic echocardiography. Next imaging study.**

Procedure	Appropriateness Category	Relative Radiation Level
MRA chest without and with IV contrast	Usually Appropriate	O
MRA chest without IV contrast	Usually Appropriate	O
MRI heart function and morphology without and with IV contrast	Usually Appropriate	O
MRI heart function and morphology without IV contrast	Usually Appropriate	O
CTA chest with IV contrast	Usually Appropriate	☢☢☢☢
MRA abdomen without and with IV contrast	May Be Appropriate	O
MRA abdomen without IV contrast	May Be Appropriate	O
CT heart function and morphology with IV contrast	May Be Appropriate	☢☢☢☢
US echocardiography transesophageal	Usually Not Appropriate	O
Radiography chest	Usually Not Appropriate	☢
Arteriography coronary with ventriculography	Usually Not Appropriate	☢☢☢☢
Arteriography pulmonary	Usually Not Appropriate	☢☢☢☢
MRA neck without and with IV contrast	Usually Not Appropriate	O
MRA neck without IV contrast	Usually Not Appropriate	O
MRI heart function with stress without and with IV contrast	Usually Not Appropriate	O
MRI heart function with stress without IV contrast	Usually Not Appropriate	O
CTA coronary arteries with IV contrast	Usually Not Appropriate	☢☢☢☢
FDG-PET/CT heart	Usually Not Appropriate	☢☢☢☢
SPECT or SPECT/CT MPI rest and stress	Usually Not Appropriate	☢☢☢☢☢

Variant 9: **Child or adult. Suspected aortic coarctation with inadequate evaluation after transthoracic echocardiography. Next imaging study.**

Procedure	Appropriateness Category	Relative Radiation Level
MRA chest without and with IV contrast	Usually Appropriate	O
MRA chest without IV contrast	Usually Appropriate	O
MRI heart function and morphology without and with IV contrast	Usually Appropriate	O
MRI heart function and morphology without IV contrast	Usually Appropriate	O
CTA chest with IV contrast	Usually Appropriate	☢☢☢☢
Arteriography coronary with ventriculography	May Be Appropriate	☢☢☢☢
CT heart function and morphology with IV contrast	May Be Appropriate (Disagreement)	☢☢☢☢
US echocardiography transesophageal	Usually Not Appropriate	O
Radiography chest	Usually Not Appropriate	☢
Arteriography pulmonary	Usually Not Appropriate	☢☢☢☢
MRA abdomen without and with IV contrast	Usually Not Appropriate	O
MRA abdomen without IV contrast	Usually Not Appropriate	O
MRA neck without and with IV contrast	Usually Not Appropriate	O
MRA neck without IV contrast	Usually Not Appropriate	O
MRI heart function with stress without and with IV contrast	Usually Not Appropriate	O
MRI heart function with stress without IV contrast	Usually Not Appropriate	O
CTA coronary arteries with IV contrast	Usually Not Appropriate	☢☢☢☢
FDG-PET/CT heart	Usually Not Appropriate	☢☢☢☢
SPECT or SPECT/CT MPI rest and stress	Usually Not Appropriate	☢☢☢☢☢

Variant 10: **Child. Known aortopathy or connective tissue disease. Surveillance of the aorta after inadequate or incomplete evaluation by transthoracic echocardiography. Next imaging study.**

Procedure	Appropriateness Category	Relative Radiation Level
MRA abdomen without and with IV contrast	Usually Appropriate	O
MRA abdomen without IV contrast	Usually Appropriate	O
MRA chest without and with IV contrast	Usually Appropriate	O
MRA chest without IV contrast	Usually Appropriate	O
MRA neck without and with IV contrast	Usually Appropriate	O
MRA neck without IV contrast	Usually Appropriate	O
CTA chest with IV contrast	Usually Appropriate	☢☢☢☢
US echocardiography transesophageal	May Be Appropriate	O
MRI heart function and morphology without and with IV contrast	May Be Appropriate	O
MRI heart function and morphology without IV contrast	May Be Appropriate	O
CTA coronary arteries with IV contrast	May Be Appropriate	☢☢☢☢
Radiography chest	Usually Not Appropriate	☢
Arteriography coronary with ventriculography	Usually Not Appropriate	☢☢☢☢
Arteriography pulmonary	Usually Not Appropriate	☢☢☢☢
MRI heart function with stress without and with IV contrast	Usually Not Appropriate	O
MRI heart function with stress without IV contrast	Usually Not Appropriate	O
CT heart function and morphology with IV contrast	Usually Not Appropriate	☢☢☢☢
FDG-PET/CT heart	Usually Not Appropriate	☢☢☢☢
SPECT or SPECT/CT MPI rest and stress	Usually Not Appropriate	☢☢☢☢☢

CONGENITAL OR ACQUIRED HEART DISEASE-CHILD

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Summary of Literature Review

Introduction/Background

Congenital and acquired pediatric heart disease is a large and diverse field with an overall prevalence estimated at 6 to 13 per 1,000 live births [1,2]. Congenital heart disease (CHD) can be broadly divided into acyanotic and cyanotic conditions. The 3 most common conditions are all acyanotic; bicuspid aortic valve, ventricular septal defects (VSD), and atrial septal defects. Common acquired cardiovascular conditions in children include aortopathy, cardiomyopathy, myocarditis, and vasculitis. Advanced imaging plays an important complementary role to transthoracic echocardiography (TTE) in the initial evaluation of congenital and acquired cardiovascular disease in children. Advances in treatment [2,3] have resulted in much longer survival of patients with CHD [4]. This longer survival has resulted in postsurgical patients often requiring imaging follow-up to look for secondary complications of corrective procedures like ventricular dysfunction, valvular dysfunction, and secondary vascular compromise. This document covers variants representing some of the most common indications for advanced imaging of childhood onset cardiovascular disease.

Special Imaging Considerations

For the purposes of distinguishing between CT and CT angiography (CTA), ACR Appropriateness Criteria topics use the definition in the ACR–NASCI–SIR–SPR Practice Parameter for the Performance and Interpretation of Body Computed Tomography Angiography (CTA) [5]:

> *"CTA uses a thin-section CT acquisition that is timed to coincide with peak arterial or venous enhancement. The resultant volumetric dataset is interpreted using primary transverse reconstructions as well as multiplanar reformations and 3-D renderings."*

All elements are essential: 1) timing, 2) reconstructions/reformats, and 3) 3-D renderings. Standard CTs with contrast also include timing issues and reconstructions/reformats. Only in CTA, however, is 3-D rendering a **required** element. This corresponds to the definitions that the CMS has applied to the Current Procedural Terminology codes.

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Discussion of Procedures by Variant

Variant 1: Child or adult. Repaired tetralogy of Fallot or pulmonary valve stenosis with concern for pulmonary valve dysfunction or branch pulmonary artery stenosis. Incomplete or inadequate assessment of cardiovascular morphology and function after transthoracic echocardiography. Next imaging study.

Tetralogy of Fallot (TOF) is the most common type of cyanotic CHD. The hallmarks of TOF are infundibular right ventricular outflow tract (RVOT) obstruction and VSD resulting in shunting of deoxygenated blood into the aorta. Definitive treatment of TOF is surgical repair of the RVOT obstruction and VSD. The surgical correction of RVOT includes resection of infundibular or subinfundibular muscles and pulmonary valvotomy to augment the outflow tract and, if needed, applying a transannular patch across the RVOT. Despite the increasing popularity of various valve sparing approaches, ventriculotomy and transannular patch repair remains the most commonly used approach in current practice. The integrity of the pulmonary valve is disrupted in an attempt to relieve the RVOT obstruction, and approximately 40% to 85% of patients go on to develop pulmonary regurgitation (PR) 5 to 10 years after repair [6-8]. Chronic PR leads to progressive RV dilatation and dysfunction, which is a risk factor for ventricular arrhythmias and may result in right heart failure and sudden death. Unilateral or bilateral branch pulmonary artery stenosis or hypoplasia may be present in TOF and may affect pulmonary development or worsen RV compliance. Poor outcomes may be prevented by timely pulmonary valve replacement (PVR) and pulmonary artery rehabilitation by surgical or percutaneous approaches, which should be performed before irreversible RV remodeling occurs but ideally after the patient's growth is completed to decrease the need for repeated interventions. Determining when to perform PVR is not straightforward, and imaging provides vital assessment of PR severity and cardiac hemodynamic status for planning. Because open surgical and percutaneous approaches to PVR exist, imaging is also essential to determine treatment candidacy for each approach, indications for treatment, and treatment outcome.

Arteriography Coronary with Ventriculography

Catheter angiography is a useful modality for the evaluation of the coronary arteries and measurement of atrial and ventricular pressure. It provides direct measurements of RVOT gradients, dynamic assessment of regurgitant flow, and hemodynamic assessment of diastolic dysfunction [9]. Coronary angiography is performed in patients with suspected anomalous coronary artery to delineate the course of the coronary artery before interventions in the RVOT [10,11]. This could be important in the setting of percutaneous intervention, because one concern is coronary artery compression with inflation of a balloon-expandable stent in the RVOT when performing PVR percutaneously.

Arteriography Pulmonary

Cardiac catheterization is performed in patients only when alternative measures cannot accurately assess the RV and pulmonary artery anatomy or hemodynamics noninvasively. However, angiographic evaluation of PR severity is complicated by the fact that catheter position across the PV may influence the angiographic severity. It allows for simultaneous catheter-guided therapeutic interventions such as pulmonary artery balloon dilatation and stent placement and PVR [12].

CT Heart Function and Morphology

A common deficiency of TTE imaging in repaired TOF is imaging of the branch pulmonary arteries. Multidetector CT provides accurate assessment of the anatomy of the branch pulmonary arteries, including within stented segments. It provides detailed anatomic evaluation of the heart and RVOT as well as other vascular abnormalities, such as pulmonary artery aneurysms or pseudoaneurysms, associated coronary anomalies, or aortic dilation [13,14]. Performing electrocardiogram (ECG) gating permits dynamic assessment of the RVOT for percutaneous pulmonary valve planning, and measurement of RV and left ventricular (LV) volume and function. Multiplanar reformats allow for precise measurement of the cross-sectional area of the RVOT and valvular annulus, which are essential for selection of the appropriate prosthetic valve, and to screen for conduit calcification and kinking. before repeat surgery to replace the conduit or valve, CT is also helpful to determine safety of sternal re-entry.

CTA Chest

CTA without ECG gating or with prospective ECG gating may be used in the setting of TOF to avoid the need for sedation for morphological assessment of the branch pulmonary arteries, including within stented segments. It is also helpful to screen for right ventricle to pulmonary artery conduit calcification and stenosis and to determine safety of sternal re-entry before surgery.

CTA Coronary Arteries

CTA technique can be used to study accurate coronary artery anatomy and exclude anomalous coronary artery crossing the RVOT before RVOT intervention or reoperation. Particularly, the relationship of the left or right coronary artery to the pulmonary outflow tract and pulmonary valve is important to assess because this may be compressed during percutaneous valve placement, conduit dilation, or sternal re-entry.

FDG-PET/CT Heart

There is no relevant literature to support the use of fluorine-18-2-fluoro-2-deoxy-D-glucose (FDG)-PET/CT heart in the evaluation of repaired TOF.

MRA Abdomen

There is no relevant literature to support the use of MR angiography (MRA) abdomen in the evaluation of repaired TOF.

MRA Chest

MRA of the chest is routinely performed along with MRI heart for function and morphology for delineation of the extracardiac vasculature, including the branch pulmonary arteries, aorta, and remaining mediastinal vasculature.

MRA Neck

There is no relevant literature to support the use of MRA neck in the evaluation of repaired TOF.

MRI Heart Function and Morphology

MRI heart function and morphology is an accurate imaging modality for delineating the cardiac anatomy and extracardiac vascular structures, quantifying ventricular size and function, and assessing PR by means of velocity-encoded cine sequences. MR provides more precise and reproducible quantification of ventricular volumes, myocardial mass, and function than TTE [15], thereby allowing comprehensive assessment of cardiovascular morphology and physiology. Cardiac MR (CMR) provides excellent assessment of ventricular size and function, valve regurgitation and stenosis, ratio of pulmonary and systemic flows, and myocardial fibrosis or remodeling. ECG-gated free-breathing cine phase-contrast CMR is a useful method for the evaluation of velocity, volume, and the pattern of blood flow that would allow accurate mapping and measurement of both systolic and diastolic pulmonary artery flow and calculation of PV regurgitation fraction [16]. PR fraction is calculated as retrograde flow volume divided by antegrade flow volume. RV volume calculation using CMR correlates well with volume as measured by angiography. Myocardial late gadolinium enhancement has been used to assess myocardial fibrosis related to areas of surgical resection as well as patch placement and may be of prognostic value for the risk of arrhythmia in these patients [17].

MRI Heart Function with Stress

There is no relevant literature to support the use of MR stress perfusion imaging in the setting of repaired TOF.

Radiography Chest

The chest radiograph in TOF patients with severe PR often demonstrates increased cardiothoracic ratio on frontal view and retrosternal fullness on the lateral view due to cardiomegaly secondary to RV volume overload. The chest radiograph may also demonstrate dilated pulmonary trunk and may demonstrate dilatation of the ascending aorta [9]. In the posttreatment setting, radiography is essential to monitor location and integrity of stents in the pulmonary arteries, screen for RVOT pseudoaneurysm after conduit placement, and screen for asymmetry in the size and branching pattern of the parenchymal pulmonary arteries, which may indicate the presence of proximal vessel hypoplasia or stenosis.

Perfusion Scan Lung

The use of pulmonary perfusion scintigraphy allows for the assessment of differential pulmonary blood flow in patients with TOF and branch pulmonary artery stenosis or hypoplasia [12].

SPECT/CT MPI Rest and Stress

Radionuclide angiography may be performed to assess LV function but has been replaced by MRI in this setting. Its use to assess RV function is limited by the confounding effect of counts from other chambers. In addition, its resolution is poor compared with other imaging methods and, therefore, has been of limited use in this setting.

US Echocardiography Transesophageal
Transesophageal echocardiography (TEE) is considered a useful technique for intraoperative assessment of adequacy of outflow tract repair in terms of ruling out any residual obstruction or valve regurgitation as a consequence of surgery. TEE offers the advantages of permitting visualization of the operative procedure in real time and providing guidance for the surgeon [6,18] . The routine use of TEE during follow-up after surgery for patients with TOF is not established.

TEE is also useful in certain patients to guide interventional procedures or evaluate valve anatomy when transthoracic imaging is challenging, or when infectious endocarditis is suspected [12].

Variant 2: Child or adult. Transposition of the great arteries after atrial switch. Incomplete or inadequate assessment of cardiovascular morphology and function after transthoracic echocardiography. Next imaging study.

Transposition of the great arteries (TGA) is a cardiac congenital defect characterized by discordant ventriculoarterial connections, in which the aorta arises above the morphological RV, and the pulmonary artery arises above the morphological LV. In TGA, the aorta usually is anterior to the PA, but it can be beside or behind the PA.

The prevalence of TGA is 4.7 per 10,000 live births [19]. TGA accounts for 3% of all CHD and 20% of cyanotic heart disease. There are 2 major types of surgery performed for D-TGA: the atrial switch procedures (Mustard and Senning procedures) and the arterial switch procedures (Jatene procedure). In the atrial switch procedure, intra-atrial venous baffles redirect the systemic and pulmonary venous blood returns to the appropriate atria restoring circulating blood flow. These baffles are created by either using in situ tissue from the right atrial wall and interatrial septum (Senning procedure) or with autologous or synthetic material (Mustard procedure). Ventriculoseptal defects, if present, are also closed.

Patients with Mustard or Senning type repair are now typically adult patients. The most common complications following the atrial switch procedures are intra-atrial venous baffle stenoses and leaks. Baffle stenoses typically occur at the superior limb of the baffle where the superior vena cava meets the right atrium. Baffle obstructions and leaks are estimated to occur in approximately 25% of patients [20]. The most important and relatively common postoperative complication is hypertrophy and decreased function of the systemic RV. Less common complications include pulmonary arterial hypertension, residual VSD, subpulmonary stenosis, and arrhythmias.

Arteriography Coronary with Ventriculography
Since the coronary arteries are not surgically manipulated during an atrial switch procedure, there is no defined role for coronary arteriography in this setting. In patients with complex coronary anatomy in the setting of TGA, angiography may be helpful to demonstrate dynamic compression and angulation of the coronary arteries in patients with an interarterial course or in those with a muscular bridge [21], but this role has been superseded by CTA in most centers.

Arteriography Pulmonary
Pulmonary arteriography may be helpful if hemodynamic assessment of pulmonary artery pressure and resistance is required. Pulmonary arteriography is performed during interventions such as branch pulmonary artery balloon dilation and stent placement, to assess for baffle leaks, or narrowing of the systemic or pulmonary venous pathways [21].

CT Heart Function and Morphology
CT is an alternative imaging modality to provide incremental information to echocardiography. CT functional assessment is a safer option to CMR in patients who are hemodynamically unstable. It is the most useful modality if anatomic restenosis is suspected in the setting of stents.

CTA Chest
CTA is the most useful modality if anatomic restenosis of pulmonary arteries is suspected in the setting of stents or presence of a metal susceptibility artifact that may hamper the use of MRA for this purpose.

CTA Coronary Arteries
Coronary CTA would be helpful when combined with cardiac CTA if repeat surgery is being considered given the increased rate of coronary anomalies.

FDG-PET/CT Heart
There is no relevant literature to support the use of FDG-PET/CT heart in the evaluation of TGA after atrial switch.

MRA Abdomen
There is no relevant literature to support the use of MRA abdomen in the evaluation of TGA after atrial switch.

MRA Chest
MRA chest provides important information regarding extracardiac structures such as the branch pulmonary arteries, systemic and pulmonary veins, and superior and inferior limb baffles and is used in conjunction with CMR for morphology and function to provide comprehensive evaluation of the cardiovascular system after treatment for TGA.

MRA Neck
There is no relevant literature to support the use of MRA neck in the evaluation of TGA after atrial switch.

MRI Heart Function and Morphology
CMR yields accurate and reproducible data regarding ventricular size and function, especially follow-up of systemic RV status and intracardiac baffles.

CMR is considered the optimal imaging modality in determining timing and method of reintervention in a patient's status after atrial switch repair for D-TGA, providing accurate morphologic, functional, and physiological data. Delayed enhancement techniques have been applied to the hypertrophied systemic RV after atrial switch in adult patients, with the presence of myocardial scarring reported to correlate with progressive ventricular dysfunction and clinical deterioration [22].

MRI Heart Function with Stress
CMR is a the most useful imaging modality to screen for myocardial ischemia, but this is uncommon after atrial switch for TGA because the coronary arteries are not manipulated during surgery.

Radiography Chest
Radiography of the chest can be used for preliminary assessment of stent position, pacemaker generator and lead position, and as needed during the course of follow-up.

SPECT or SPECT/CT MPI Rest and Stress
There is no relevant literature to support the use of single-photon emission CT (SPECT)/CT myocardial perfusion imaging (MPI) rest and stress in the evaluation of TGA after atrial switch.

US Echocardiography Transesophageal
TEE is useful in patients with poor transthoracic acoustic windows, during intraoperative imaging, and in patients (usually adolescents) who require cardioversion for arrhythmia. It is typically used to assess the adequacy of intraoperative repair TTE and may be more sensitive to detect baffle leaks and baffle obstruction. TEE is also helpful to guide catheter-based treatment of pathway obstruction such as balloon dilation, stent placement, or device closure of baffle leaks.

Variant 3: Child or adult. Transposition of the great arteries after arterial switch. Incomplete or inadequate assessment of cardiovascular morphology and function after transthoracic echocardiography. Next imaging study.

The arterial switch operation (ASO) has superseded the Mustard and Senning procedures as the most common surgery performed in D-TGA. In the ASO (Jatene procedure), the ascending aorta and the main pulmonary artery are transected above their valve leaflets and moved to their correct circulatory position (Lecompte maneuver). The coronary arteries are then transplanted from the native aortic root to the neoaortic root (which is the native pulmonary trunk). Septal defects, if present, are surgically closed.

Well-known sequelae after an ASO include obstruction of the coronary arteries with related myocardial ischemia, LV dysfunction, narrowing of the pulmonary artery side branches, RVOT obstruction, neoaortic valve insufficiency, and dilatation of the aortic root. RVOT and pulmonary artery stenoses may lead to RV dysfunction necessitating corrective procedures. Supravalvular pulmonary stenosis, particularly in the side branches of the pulmonary arteries, occurs early after an ASO. Dilatation and insufficiency of the neoaortic root usually develops over time. Coronary abnormalities can be present in 8% of asymptomatic individuals on 1- to 20-year follow-up and, although uncommon, can cause sudden death [23]. Noninvasive imaging of the postoperative anatomy is integral to the management of patients who have undergone an ASO. A combination of imaging tests and exercise testing are performed to screen for complications and to assess severity and the need for intervention and the type of intervention.

Arteriography Coronary with Ventriculography
The coronary artery course has a particularly high variability in patients with TGA. After an ASO, many patients require repeated imaging of their reimplanted coronary arteries. Cardiac catheterization and ventriculography after ASO is reserved for only complex cases in whom incremental information is needed following advanced imaging. Angiography is performed to assess for coronary artery stenosis after coronary artery reimplantation or during interventional procedures such as branch pulmonary artery balloon dilation and stent placement. In patients with complex coronary anatomy, angiography is useful in demonstrating dynamic compression and angulation of the coronary arteries in patients with an interarterial course or in those with a muscular bridge [21].

Arteriography Pulmonary
Pulmonary arteriography is performed during interventions such as branch pulmonary artery balloon dilation and stent placement.

CT Heart Function and Morphology
CT is an alternative imaging modality to provide incremental information to echocardiography. CT functional assessment is more useful over CMR in patients who are hemodynamically unstable. It is the most useful modality if anatomic restenosis is suspected in the setting of stents.

CTA Chest
CTA of the chest is useful to study airway compromise, which may happen related to the aortic root or ascending aortic dilatation after ASO. A dynamic CTA may be helpful in this setting to distinguish intrinsic from extrinsic causes of airway obstruction. CTA is the most useful modality if anatomic restenosis of pulmonary arteries is suspected in the setting of stents or presence of a metal susceptibility artifact that may hamper the use of MRA for this purpose.

CTA Coronary Arteries
Obstructed coronary arteries occur in 8% of survivors and is a common cause of morbidity and mortality after the ASO [23]. ECG-triggered CTA is currently the most useful modality for the primary evaluation of coronary arteries. Its high spatial and temporal resolution allows for the reliable visualization of coronary arteries and screening for potential stenosis, usually without the use of sedation [19,24]. CT coronary angiography may also be performed as an alternative to coronary catheterization when SPECT/CT MPI shows perfusion deficit or CMR stress perfusion imaging shows ischemia or wall motion abnormality in a vascular distribution [25].

FDG-PET/CT Heart
FDG-PET/CT can be used as the alternative method to SPECT/CT MPI for assessing viability when myocardial ischemia is suspected or to assess blood flow to the branch pulmonary arteries after the ASO. FDG-PET/CT can have confirmatory and/or incremental value to TEE and CTA in diagnosing prosthetic endocarditis after RV-PA conduit placement, particularly when anatomic imaging findings are inconclusive or equivocal.

MRA Abdomen
There is no relevant literature to support the use of MRA abdomen in the evaluation of TGA after arterial switch.

MRA Chest
MRA chest provides important information regarding extracardiac structures such as the branch pulmonary arteries, ascending aorta, and the aortic arc, and is used in conjunction with CMR for morphology and function to provide comprehensive evaluation of the cardiovascular system after treatment for TGA.

MRA Neck
There is no relevant literature to support the use of MRA neck in the evaluation of TGA after arterial switch.

MRI Heart Function and Morphology
CMR yields accurate and reproducible data regarding both LV and RV size and function, differential pulmonary blood flow, neoaortic root diameter and regurgitant blood flow, coronary artery diameter, myocardial perfusion, and myocardial fibrosis. CMR provides important information regarding myocardial performance and viability, as well as quantitative assessment of valvar function and accurate evaluation of conduits [21]. If the echocardiographic

assessment of ventricular parameters or the severity of valve regurgitation is in question, CMR can resolve this uncertainty [26]. The quantitative data on CMR can be validated by comparing the main pulmonary artery flow to the sum of the branch pulmonary artery flows. CMR using 3-D steady-state free precession navigator respiratory gated sequences can noninvasively evaluate the coronary arteries looking for ostial stenoses or proximal kinking and screen for myocardial injury using perfusion and viability sequences [27,28].

MRI Heart Function with Stress
CMR is the most useful imaging modality to screen for myocardial ischemia [28]. CMR coronary angiography can provide good resolution images of coronary lumen especially in adolescents and larger children, but it is inferior in comparison with CTA in young children [26]. Myocardial stress perfusion MRI can also be done when there is a concern for ischemia following arterial switch procedure [28,29].

Radiography Chest
Radiography of the chest can be used for the preliminary assessment of stent position, pacemaker and lead position, and as needed during the course of follow-up.

SPECT or SPECT/CT MPI Rest and Stress
Coronary reimplantation during ASO increases the risk of long-term ischemic complications in the postoperative period. SPECT/CT MPI is used when patient symptoms or ECG findings are concerning for myocardial ischemia [30].

US Echocardiography Transesophageal
TEE is useful in patients with poor transthoracic acoustic windows, during intraoperative imaging, and in patients (usually adolescents) who require cardioversion for arrhythmia. It is typically used to assess the adequacy of intraoperative repair. If postoperative access to the transthoracic window is limited, TEE can be used to assess the coronary artery origins and flow patterns in the early postoperative period.

Variant 4: Child. Suspected or confirmed congenital or acquired coronary artery abnormality. Incomplete or inadequate assessment of coronary morphology after transthoracic echocardiography. Next imaging study.

Anomalous aortic origin of a coronary artery (AAOCA) that courses between the 2 great vessels is a rare congenital cardiac anomaly that may carry an increased risk for myocardial ischemia and sudden cardiac death in young patients [31]. It is further characterized by 1 of 5 course subtypes: interarterial, subpulmonic, prepulmonic, retroaortic, or retrocardiac. The prevalence of anomalous coronary arteries noted in coronary CTA series is 0.99% to 5.8% [32]. Most patients with AAOCA remain undiagnosed because of a lack of symptoms, but a minority become symptomatic and experience adverse cardiac events. It is important to delineate the coronary anatomy accurately, including the ostial status, coronary branching pattern, and presence and length of an intramural segment, because narrowing of the ostium and proximal coronary artery with exertion has been proposed as a possible mechanism of sudden death in patients with interarterial, intramural coronary course. Other congenital coronary anomalies that may be clinically significant include anomalous left coronary artery from the pulmonary artery and intraseptal or intramyocardial course of the coronary artery.

The most commonly acquired coronary artery disease in children is secondary to Kawasaki disease. It is an acute vasculitis primarily affecting children <5 years of age. It affects the medium-sized vessels, including the coronary arteries, and may lead to the formation of coronary artery aneurysms throughout the body that may be complicated by coronary artery aneurysms immediately after the acute phase or by stenotic lesions several years after the acute episode.

Arteriography Coronary with Ventriculography
Coronary angiography is traditionally reserved for interventional procedures or when noninvasive diagnostic imaging with CT or MRI is inconclusive. It can correctly identify all types of anomalous coronary arteries. However, it lacks optimum 3-D information and the ability to image soft tissue, making it difficult to demonstrate the spatial relationship among the coronary artery course, myocardium, and great vessels.

Coronary angiography is essential to clarify the extent of the vulnerable territory of the anomalous vessel (ie, dominant versus nondominant right coronary artery, single coronary, etc).

Fractional flow reserve may offer an adjunct to determine the functional significance of AAOCA narrowing or intramyocardial course of the coronary artery, although it is unproven in these settings.

In Kawasaki disease, to predict the progress of the disease and determine appropriate treatment and follow-up protocols, it is essential to understand the size and shape of the aneurysms in the early phase. Therefore, traditionally, invasive angiography is performed after the acute stage but, since the advent of CT and MRA invasive coronary angiography, has become second line [33].

Intravascular Ultrasound (IVUS): IVUS affords precise sequential cross-sectional imaging of the coronary lumen and coronary wall thickness with a discriminating power, based on superior spatial resolution. It is particularly useful in stenosis of the anomalous vessel in its intramural course.

Arteriography Pulmonary
There is no relevant literature to support the use of pulmonary arteriography in the evaluation of suspected or confirmed congenital or acquired coronary artery abnormality.

CT Heart Function and Morphology
Functional assessment with CT may be considered in the setting of Kawasaki disease for assessment of the consequences of coronary artery compromise.

CTA Chest
CTA of the chest is rarely helpful for assessment of other vascular involvement in the setting of Kawasaki disease, with axillary artery aneurysms being a common manifestation in the chest. MRA of the chest and abdomen is more useful to CTA due to its ability to interrogate a wider vascular territory for screening for aneurysms in the setting of Kawasaki disease [34].

CTA Coronary Arteries
CTA of coronary arteries is currently the most useful imaging modality for evaluating coronary arteries. Virtual angioscopic view of coronary CTA helps to evaluate anatomic high-risk features, such as the slit-like morphology of the ostium [35]. CT can identify other anatomic high-risk features, such as acute take-off angle, luminal shape, and proximal narrowing of the anomalous coronaries related to the intramural and interarterial course [35]. Coronary CTA, overall, provides a more accurate depiction of the entire anomalous vessel including the distal coronary segments. Coronary CTA is also useful in the setting of Kawasaki disease to screen for the presence and extent of coronary involvement by ectasia, aneurysm, or stenosis and to diagnose and follow-up the presence of mural thrombus in larger aneurysms.

FDG-PET/CT Heart
There is no relevant literature to support the use of FDG-PET/CT in the evaluation of suspected or confirmed congenital or acquired coronary artery abnormality.

MRA Abdomen
MRA abdomen is used to screen for extracardiac involvement in Kawasaki disease, with iliac aneurysms being a common manifestation of the disease.

MRA Chest
MRA chest allows 3-D reconstruction visualization of coronary artery origin and proximal course in AAOCA and the presence of coronary aneurysms in Kawasaki disease, but its spatial resolution is inferior to that of coronary CTA. MRA using 3-D steady-state free precession or 3-D fast gradient echo techniques can be used to evaluate the proximal coronary in AAOCA or the coronary tree in Kawasaki disease. MRA may also be performed with intravenous (IV) contrast to assess the systemic arterial tree for evidence of extracardiac aneurysms in Kawasaki disease [36-38].

MRI Heart Function and Morphology
CMR offers a wealth of additional relevant information including valvular function, ventricular function, regional contractility, and myocardial viability, all of which could be important considerations during the preoperative evaluation or postoperative follow-up in patients who undergo surgical repair of coronary artery.

MRI Heart Function with Stress
Pharmacological stress CMR cine and MPI can be performed to assess the functional relevance of a coronary anomaly [36,39].

Radiography Chest
There is no relevant literature to support the use of chest radiography in the evaluation of suspected or confirmed congenital or acquired coronary artery abnormality.

SPECT or SPECT/CT MPI Rest and Stress
Nuclear cardiac imaging modalities play an important role in assessing myocardial perfusion, thus allowing for assessment of the functional relevance of any acquired coronary artery. SPECT MPI and PET MPI may unmask ischemia in asymptomatic and symptomatic patients with anomalous coronary artery [40]. The panel disagreed on SPECT or SPECT/CT MPI rest and stress to screen for myocardial ischemia as the next imaging study after TEE, although its use was supported by expert opinion in centers with pediatric experience with coronary ischemia.

US Echocardiography Transesophageal
TEE may be useful to visualize coronary artery anatomy perioperatively when the thoracic window is limited. However, TEE is not useful for routine imaging of AAOCA or Kawasaki disease.

MRA Neck
There is no relevant literature to support the use of MRA neck in the evaluation of suspected or confirmed congenital or acquired coronary artery abnormality.

Variant 5: Child. Known single ventricle physiology. Preoperative evaluation for stage 2 single ventricle palliation. Incomplete or inadequate assessment of cardiovascular morphology and function after transthoracic echocardiography. Next imaging study.

Single ventricle lesions are congenital heart anomalies in which one of the 2 ventricular chambers is either absent or so severely hypoplastic that a biventricular repair is impossible. Typically, the atrioventricular valve associated with the absent or hypoplastic ventricle is also absent or hypoplastic. The arterial outflow valve associated with the absent/hypoplastic ventricle is also frequently affected. Single ventricle congenital heart defects cover a wide spectrum including the following: hypoplastic left heart syndrome, tricuspid atresia, unbalanced atrioventricular, pulmonary atresia with intact ventricular septum, Ebstein anomaly, and double outlet left ventricle.

The repair of this entity involves 3-staged reconstruction to establish a circulatory system wherein all the deoxygenated blood flows directly into the lungs, bypassing the heart, while the oxygenated blood goes to the functional single ventricle to be pumped out to the systemic circulation. Stage 1 of this repair involves optimizing the pulmonary arterial blood flow, sometimes with a systemic-pulmonary arterial shunt or pulmonary artery band, and is performed shortly after birth. This procedure allows postoperative recovery and growth until stage 2 palliation can be performed. The stage 2 procedure, otherwise known as superior cavopulmonary connection, involves establishing a connection between the systemic venous return from the upper half of the body and the pulmonary arteries to allow deoxygenated blood flow into the pulmonary circulation. Some procedures in stage 2 are bidirectional Glenn, hemi-Fontan, and Kawashima operations. This procedure is typically performed within 6 months of birth.

The primary goals of the imaging evaluation before the stage 2 operation include an assessment for obstruction in the pulmonary arteries and veins, the atrial septal defect, ventricular outflow tract, and aortic arch; for the degree and mechanism of valve regurgitation; and ventricular size and function. Another role of imaging could be to decide if the ventricular volumes and functions still support the patient proceeding down the single ventricle pathway or if the patient is a candidate for a 1.5-ventricle or 2-ventricle repair. A crucial role of imaging in this setting is to demonstrate the pulmonary arterial anatomy, including the distal branches, to evaluate for anatomic obstruction that may impact the Glenn circulation. Knowledge of branch pulmonary artery anatomy adds significantly to the surgical planning process. A complete anatomic assessment of the branch pulmonary arteries to detect stenoses should be performed in all patients undergoing stage 2 repair.

Arteriography Coronary with Ventriculography
Direct cardiac catheterization and angiography has historically been the standard of care for assessing hemodynamic and anatomic suitability before stage 2 repair. Its routine use is decreasing since the advent of advanced CMR and CT methods. The use of catheter angiography is supported in the literature [41]; for example, in patients with pulmonary atresia and intact ventricular septum, the diagnosis of RV dependent coronary circulation is established by RV and selective coronary angiography and has an important impact upon surgical approach.

One of the advantages of cardiac catheterization before bidirectional Glenn operation is that anomalies such as systemic-to-pulmonary collateral vessels and aortic coarctation can be palliated using transcatheter techniques. It particularly pertinent for those who have undergone prior stage 1 palliation with aortic arch reconstruction, which is associated with a 10% to 15% incidence of recurrent coarctation at the distal end of the arch reconstruction [42].

Arteriography Pulmonary
Cardiac catheterization is useful for the imaging evaluation before stage 2 repair to assess pulmonary arterial anatomy before surgical repair [41]. Routine cardiac catheterization is less frequently performed before undertaking stage 2 repair, but it has a useful role in the assessment of hemodynamic suitability to adapt to this type of circulation by directly measuring pulmonary vascular resistance.

CT Heart Function and Morphology
CT heart function and morphology can be useful for therapeutic decision-making regarding 2-ventricle versus single ventricle repair. This technique is frequently used to supplement morphological information related to the heart, coronaries, and extracardiac vasculature. Performance of cardiac CT for pre–stage 2 evaluation has the potential to supplement echocardiographic data sufficiently that catheterization would not be needed. The disadvantages of CT versus CMR in this setting are reduced functional information regarding blood flow and ventricular function [43].

CTA Chest
CTA chest is a useful modality and provides accurate means of evaluation of pulmonary arterial anatomy and pulmonary and systemic venous status before stage 2 repair. Thrombosis of the systemic-to-pulmonary arterial shunt may occur in 6% to 17% of cases and constitutes a surgical emergency [44]. CTA can be used as the confirmatory test especially if echocardiography is inconclusive.

CTA Coronary Arteries
There is no relevant literature to support the use of CTA coronary arteries in the evaluation of pre–stage 2 single ventricle physiology.

MRA Abdomen
There is no relevant literature to support the use of MRA abdomen in the evaluation of known single ventricle physiology.

MRA Chest
MRA chest is an excellent imaging technique for the evaluation of pulmonary arterial anatomy before stage 2 repair. It has the advantage that it can be easily combined with MRI heart function and morphology for a complete evaluation of the single ventricle patient. MRA has great diagnostic utility compared with echocardiography and is accurate in comparison to catheter angiography for the diagnosis of pulmonary artery stenoses.

MRA Neck
There is no relevant literature to support the use of MRA neck in the evaluation of known single ventricle physiology.

MRI Heart Function and Morphology
CMR allows for the quantification of ventricular volume and function, with abnormal ventricular morphology [42]. This can be useful in decisions regarding stage 1, 1.5, or 2 ventricle repair.

MRI Heart Function with Stress
There is no relevant literature to support the use of MRI heart function with stress in the evaluation of known single ventricle physiology.

Radiography Chest
Other than evaluation of patent ductus arteriosus or aortopulmonary shunt stent positioning, there is no relevant literature to support the use of radiography chest in the evaluation of known single ventricle physiology.

SPECT or SPECT/CT MPI Rest and Stress
There is no relevant literature to support the use of SPECT or SPECT/CT MPI rest and stress in the evaluation of known single ventricle physiology.

FDG-PET/CT Heart
There is no relevant literature to support the use of FDG-PET/CT heart in the evaluation of known single ventricle physiology.

US Echocardiography Transesophageal
TEE may provide useful information before, during, or after surgery for stage 2 palliation regarding pulmonary arterial or pulmonary venous flow patterns, severity of atrioventricular valve regurgitation, atrial shunt, and cardiac thrombus.

Variant 6: Child. Known single ventricle physiology. Preoperative evaluation for stage 3 single ventricle palliation (total cavopulmonary connection). Incomplete or inadequate assessment of cardiovascular morphology and function after transthoracic echocardiography. Next imaging study.

The Fontan operation for single ventricle patients is the procedure in which a conduit (the total cavopulmonary connection) is placed to channel the remaining systemic venous return (including the hepatic veins) into the pulmonary arteries. The aim of the diagnostic evaluation before Fontan is to identify those few patients in whom the Fontan operation should not be performed and those who require additional intervention before or at the time of Fontan. The main consideration before Fontan procedure is elevated pulmonary artery pressure, which determines the prognosis during the postoperative course. However, with the modern stage 2 repair techniques, pulmonary resistance is seldom an important issue at the pre-Fontan stage.

Arteriography Coronary with Ventriculography
Cardiac catheterizations are done when elevated pulmonary artery pressure or elevated ventricular filling pressure is suspected. Cardiac catheterization also provides an opportunity for interventional procedures before the Fontan operation consisted of embolization of aortopulmonary collateral vessels [45].

Other important indications for cardiac catheterization include severe atrioventricular valve regurgitation semiquantitatively evaluated by means of echo color Doppler, suspected pulmonary vein stenosis at echo or CMR, suspected pulmonary pathway or Glenn anastomosis obstruction, or suspected significant venovenous collaterals at MRA [46].

Arteriography Pulmonary
Pulmonary arteriography is performed after echocardiography when there is a suspicion of branch pulmonary artery stenosis because smaller pulmonary arteries are not optimally seen on echocardiography.

CT Heart Function and Morphology
CT heart function and morphology is frequently used to supplement morphological information related to the heart, coronaries, and extracardiac vasculature but can provide information on ventricular function as well. Performance of cardiac CT for pre–stage 3 evaluation has the potential to supplement echocardiographic data sufficiently so that catheterization would not be needed in low-risk patients [47].

CTA Chest
Pulmonary embolism and thrombosis are known complications in patients undergoing single ventricle palliation. CTA is an effective method for diagnosis of pulmonary embolism in patients presenting with symptoms suspicious for pulmonary embolism [48].

CTA Coronary Arteries
There is no relevant literature to support the use of CTA coronary arteries in the evaluation of known single ventricle physiology.

MRA Abdomen
There is no relevant literature to support the use of MRA abdomen in the evaluation of pre–stage 3 single ventricle physiology.

MRI Heart Function and Morphology
Routine preoperative imaging before Fontan procedure is obtained using CMR for anatomy and to determine the current hemodynamics and physiology [49,50]. CMR is a comprehensive modality for accurate assessment of ventricular function, pulmonary artery, pulmonary venous return, cavopulmonary anastomosis, aortic arch, valves, and any valvular regurgitation.

MRA Chest
MRA chest provides good assessment of branch pulmonary arteries, their caliber, and presence of thrombus within but often fails to visualize very small aortopulmonary collaterals that may require preoperative embolization.

MRA Neck
There is no relevant literature to support the use of MRA neck in the evaluation of known single ventricle physiology.

MRI Heart Function with Stress
There is no relevant literature to support the use of MRI heart function with stress in the evaluation of known single ventricle physiology.

Radiography Chest
Other than evaluation of device or stent positioning, there is no relevant literature to support the use of radiography chest in the evaluation of known single ventricle physiology.

SPECT or SPECT/CT MPI Rest and Stress
There is no relevant literature to support the use of SPECT or SPECT/CT MPI rest and stress in the evaluation of known single ventricle physiology.

FDG-PET/CT Heart
There is no relevant literature to support the use of FDG-PET/CT heart in the evaluation of known single ventricle physiology.

US Echocardiography Transesophageal
TEE may provide useful information before, during, or after surgery for stage 3 palliation regarding pulmonary arterial or pulmonary venous flow patterns, severity of atrioventricular valve regurgitation, atrial shunt, and cardiac thrombus.

Variant 7: Child or adult. Known single ventricle physiology. Postoperative evaluation after stage 3 single ventricle palliation (total cavopulmonary connection). Incomplete or inadequate assessment of cardiovascular morphology and function after transthoracic echocardiography. Next imaging study.

Stage 3 repair for single ventricle palliation involves baffling of all systemic venous drainage directly to the pulmonary arteries, known as total cavopulmonary connection or Fontan operation. This is typically performed between 2 to 4 years of age and most commonly consists of either a lateral tunnel intracardiac baffle or an extracardiac conduit, with or without a fenestration. The Fontan operation also ensures that hepatic blood flow is directed into the lungs via the pulmonary arteries, thus preventing the formation of pulmonary arteriovenous malformations. Today, the estimates of 20-year survival for survivors of the Fontan procedure vary between 61% and 85% [51].

Early diagnosis and management of complications associated with Fontan physiology hold the most promise for improving longevity. Imaging after stage 3 repair can assess anatomy, ventricular and valve function, blood flow, and myocardial fibrosis. Anatomic considerations include the presence of thrombus in the circuit, the patency of the extracardiac conduit, stenoses within the caval veins and pulmonary arteries, and the size and location of aortopulmonary collaterals and venovenous collaterals. Considerations for assessment of flow include quantification of the degree of aortopulmonary and venovenous collateralization, the relative flow within the 2 branch pulmonary arteries, and valvular regurgitation. Considerations for assessment of ventricular function and morphology include screening for systolic and diastolic dysfunction, ventricular type and shape, and myocardial characterization.

Arteriography Coronary with Ventriculography
Clinical indications for this examination include investigating symptoms associated with decreased ventricular function such as unexplained volume retention, fatigue, exercise limitation, and cyanosis. Cardiac catheterization with coronary angiography can evaluate coronary artery anatomy, and ventriculography can be used to estimate the function of the single ventricle and valvular flow. Cardiac catheterization is also helpful to assess the presence and severity of aortopulmonary, systemic-pulmonary venous, and systemic venovenous collaterals and also provides access to perform embolization of clinically significant collaterals. Cardiac catheterization is also suggested in the

assessment of patients with failing Fontan physiology and features of Fontan-associated liver disease. Imaging of a patent fenestration is seen well with direct angiography on cardiac catheterization [52].

Arteriography Pulmonary

Cardiac catheterization for pulmonary angiography can be performed when pulmonary artery stenosis is detected on echocardiography or MRI and pulmonary artery angioplasty is indicated [52].

CT Heart Function and Morphology

CT heart function and morphology is indicated when MRI is limited by the presence of significant metallic susceptibility artifact obscuring the heart or due to the presence of MR-incompatible pacemakers/devices.

CTA Chest

CTA is useful for assessment of anatomy after stage 3 repair. Anatomic considerations include the presence of thrombus in the circuit, the patency of the extracardiac conduit, stenoses within the caval veins and pulmonary arteries, and the size and location of aortopulmonary collaterals and venovenous collaterals [52,53]. After Fontan surgery, imaging of coronaries, systemic arteries, and Fontan circuit require different timing and dose considerations, and dedicated contrast injection and scanning protocols designed for the Fontan circulation are needed to optimize the study [48].

CTA Coronary Arteries

CTA of the coronary arteries is useful and allows for high spatial resolution evaluation of coronary artery anatomy in patients after stage 3 repair [52].

FDG-PET/CT Heart

There is no role for routine performance of FDG-PET imaging after stage 3 palliation in single ventricle. FDG-PET/CT of the heart can be performed in the setting of changes of clinical status that might indicate underlying cardiac dysfunction.

MRA Abdomen

There is no relevant literature to support the use of MRA abdomen after stage 3 repair.

MRA Chest

MRA chest can provide accurate anatomic characterization of stage 3 repair. Anatomic considerations include the presence of thrombus in the circuit, the patency of the extracardiac conduit, stenoses within the caval veins and pulmonary arteries, and the size and location of aortopulmonary collaterals and venovenous collaterals [52,53]. Gadolinium-enhanced CMR angiography has been demonstrated to be more accurate in the diagnosis of a collateral vessels in comparison to catheter angiography and offers promise to plan transcatheter interventions in those who prove to have clinically significant collateral vessels. Of note, MRA chest has lower spatial resolution compared with CTA chest for small vessels such as the coronary arteries [52].

MRA Neck

There is no relevant literature to support the use of MRA neck after stage 3 repair.

MRI Heart Function and Morphology

CMR is used regularly in addition to echocardiography for long-term monitoring of Fontan patients. MRI for heart function and morphology is helpful in assessment of anatomy, ventricular and valve function, flows, and myocardial fibrosis. It allows for estimating ventricular function, quantifying valvular regurgitation, quantifying pulmonary and systemic blood flow, and aortopulmonary collaterals [54]. Late gadolinium enhancement CMR is used to detect myocardial fibrosis and infarction [55].

Phase-contrast MRA flow measurements in the branch pulmonary arteries are useful for the quantification of differential pulmonary blood flow in the Fontan patient compared with nuclear scintigraphy, due to venous streaming effects on caval flows [53]. Four-dimensional flow, with or without the use of blood pool contrast agents, has been used to simplify, accelerate, and abbreviate the MR scanning protocol in these patients [56].

MRI Heart Function with Stress

There is no role for routine performance of vasodilator stress perfusion after stage 3 palliation in single ventricle. There is some evidence that vasodilator stress perfusion MRI can be used to assess latent diastolic dysfunction after stage 3 repair [57,58].

Radiography Chest
There is no relevant literature to support the routine use of chest radiography after stage 3 palliation in single ventricle. However, chest radiography may be used to screen for the location of devices and metallic implants that are placed in this patient population.

SPECT or SPECT/CT MPI Rest and Stress
There is no role for routine performance of vasodilator stress perfusion after stage 3 palliation in single ventricle. SPECT or SPECT/CT MPI rest and stress tests are used in the setting of changes of clinical status that might indicate underlying cardiac dysfunction.

US Echocardiography Transesophageal
TEE can be used for comprehensive evaluation of the heart and central vascular anatomy, ventricular and valvular function, and flows in areas that are difficult to assess by TTE. One area in which it is better than TTE is in the detection of intracardiac thrombus.

Variant 8: Child or adult. Known or suspected anomalous pulmonary venous return with inadequate evaluation after transthoracic echocardiography. Next imaging study.

Total anomalous pulmonary venous connection (TAPVC) refers to the condition in which all of the pulmonary veins drain into the right atrium, either directly or indirectly through other systemic pathways. TAPVC is rare, with prevalence estimated at approximately 0.01% [59]. The contexts of imaging in TAPVC management are to define the surgical anatomy, when one or more pulmonary veins are not visualized by TTE, a TTE showing right-sided cardiac chamber dilation with unclear delineation of the pulmonary venous anatomy, when obstruction is suspected, or when there is an associated abnormality like a varix that is not characterized adequately by TTE. There is also an important role for imaging in the postoperative period after TAPVC repair to screen for recurrent obstruction.

Partial anomalous pulmonary venous connection (PAPVC) is where one or more pulmonary veins drains into the right atrium, either directly or indirectly. PAPVC is more common with a prevalence at approximately 1% [59]. PAPVC has a few common variants. The most common form is where the left upper pulmonary vein connects to the left brachiocephalic vein into the superior vena cava. Another type is where the right upper pulmonary veins connect directly into the superior vena cava, which is commonly associated with other left to right shunts, including atrial septal and superior sinus venosus defects. Another unique variant of PAPVC is Scimitar syndrome, in which part or all of the right pulmonary veins connect to the inferior vena cava through the diaphragm. The right lung is often hypoplastic and/or has anomalous bronchial or arterial anatomy in addition to the PAPVC. The ultimate management of PAPVC is usually surgical correction, but unlike TAPVC, this does not always happen shortly after birth due to delayed diagnosis or mild symptoms. Advanced imaging in diagnosis of PAPVC is usually requested in the setting in which a TTE shows right-sided cardiac chamber dilation with unclear delineation of the pulmonary venous anatomy. The role of imaging in PAPVC management is to confirm the diagnosis, define the surgical anatomy, and quantify the shunt to help decide on need for and timing of intervention.

Arteriography Coronary with Ventriculography
Cardiac catheterization was traditionally the imaging modality used for the diagnosis of TAPVC and PAPVC; however, it has mostly been replaced by less invasive modalities [60]. Cardiac catheterization can be used to demonstrate the anatomy and quantify the degree of shunting using oximetry. However, cardiac catheterization has been shown by some studies to be less accurate in the setting of PAPVC [61]. Occasionally, simultaneous depiction of systemic and pulmonary vascular systems may be difficult in catheter angiography due to overlapping views of adjacent vascular structures. Moreover, it carries high risk of mortality in obstructive TAPVC. The main role for a cardiac catheterization is now in the rare patient with TAPVC who requires interventional cardiac palliation before complete repair, such as atrial septostomy or stent placement [62,63].

Arteriography Pulmonary
There is no relevant literature to support the use of pulmonary arteriography in the evaluation of known or suspected anomalous pulmonary venous return with inadequate evaluation after TTE.

CT Heart Function and Morphology
CT heart function and morphology is indicated when MRI is limited by the presence of significant metallic susceptibility artifact obscuring the heart or due to the presence of MR-incompatible pacemakers/devices.

CTA Chest
CTA chest is useful for the anatomical definition of anomalous pulmonary venous drainage anatomy. CTA chest can also be used in the critical setting of possible obstructed TAPVC to define the level and degree of obstruction, especially in children with infracardiac and mixed TAPVC [64,65]. In obstructed infradiaphragmatic TAPVC, CTA chest can correctly depict the drainage site of the common pulmonary vein, stenosis of the vertical vein, and the course of the atypical vessel into the systemic vein, especially outside the usual echocardiographic windows [66].

In other types of TAPVC and PAPVC, CTA chest has excellent spatial resolution and can help define the complete anomalous drainage pathways of the pulmonary veins. CTA chest has the added advantage of being an excellent modality for evaluating the pulmonary parenchyma to look for associated pulmonary and bronchovascular abnormalities (eg, pulmonary sequestration, hypoplastic lung, abnormal bronchial branching) [64,67,68].

CTA Coronary Arteries
There is no relevant literature to support the use of CTA coronary arteries in the evaluation of known or suspected anomalous pulmonary venous return with inadequate evaluation after TTE.

MRA Abdomen
Vascular imaging of the abdomen may be helpful in the cases of infracardiac TAPVC or PAPVC to demonstrate the drainage of pulmonary vein into hepatic vein or portal vein. It may be needed to assess the resulting complications such as portal venous system aneurysm formation and collaterals.

MRA Chest
MRA chest is excellent at identifying the anatomy, specifically the anomalous course and connections of the pulmonary veins [68,69]. A small case study showed that MRA chest was more accurate than both echocardiography and cardiac catheterization in accurately identifying the anomalous veins in 7 patients with TAPVC [70].

MRI Heart Function and Morphology
MRI heart function and morphology is complementary to MRA chest and can be performed during the same imaging session. MRI heart function and morphology is useful in cases of PAPVC, because it is the ideal noninvasive modality to quantify the shunt amount by means flow rate assessment, ventricular muscle masses, and ventricular volumes and function [68,71]. The shunt quantification is usually performed using phase-contrast imaging through the aorta and pulmonary vein.

MRI Heart Function with Stress
There is no relevant literature to support the use of MRI heart function with stress in the evaluation of known or suspected anomalous pulmonary venous return with inadequate evaluation after TTE.

MRA Neck
There is no relevant literature to support the use of MRA neck in the evaluation of known or suspected anomalous pulmonary venous return with inadequate evaluation after TTE.

Radiography Chest
Chest radiography may demonstrate additional findings such as associated dextrocardia, signs of vascular congestion, and pulmonary edema, which may point towards obstructed TAPVC [65].

In PAPVC, the abnormal vein is almost never identified on chest radiograph except in cases of “scimitar syndrome” in which anomalous draining vein may be seen as a tubular structure paralleling the right heart border.

FDG-PET/CT Heart
There is no relevant literature to support the use of FDG-PET/CT heart in the evaluation of known or suspected anomalous pulmonary venous return with inadequate evaluation after TTE.

SPECT or SPECT/CT MPI Rest and Stress
There is no relevant literature to support the use of SPECT or SPECT/CT MPI rest and stress in the evaluation of known or suspected anomalous pulmonary venous return with inadequate evaluation after TTE.

US Echocardiography Transesophageal
TEE can be used in cases of PAPVC in which TTE does not completely delineate the partial veins and cardiac chamber enlargement [72,73]. However, the procedure is more invasive than CT or MRI, and it can suffer from

airways obscuring some of the acoustic windows, especially for anomalous venous drainage that occurs superior to the right atrium. TEE is also less accurate than MRI for some pulmonary vein anatomy and shunt quantification [72].

TEE has just recently been shown to be safe in patients with TAPVC, but only after median sternotomy [74]. There is no published role for TEE as the most useful imaging modality after an insufficient TTE with suspicion for TAPVC.

Variant 9: Child or adult. Suspected aortic coarctation with inadequate evaluation after transthoracic echocardiography. Next imaging study.

Coarctation of aorta (CoA) refers to discrete or diffuse narrowing in the aorta causing obstruction to the flow of blood. It accounts for 6% to 8% of all CHDs [75]. Aortic coarctation can be more commonly associated with other cardiac lesions such as bicuspid aortic valve, aortic arch hypoplasia, aberrant aortic branch vessels, and other intracardiac and conotruncal defects. This variant will focus on imaging recommendations for aortic coarctation and associated aortic arch pathology. If a patient has associated intracardiac or conotruncal abnormalities, please refer to those variants for recommendations on how to image those concurrent diseases. Aortic coarctation is managed by surgical treatment, transcatheter balloon angioplasty, and transcatheter stent placement.

Arteriography Coronary with Ventriculography

Before advances in echocardiography, cardiac catheterization was the mainstay for making the diagnosis of CoA. The pressure gradient across the coarctation segment and the collateral vascularity can be accurately assessed with direct angiography. In the current era, it is primarily used when as catheter-based investigation such as balloon angioplasty or stent insertion are being considered [76].

Arteriography Pulmonary

There is no relevant literature to support the use of pulmonary arteriography in the evaluation of known or suspected aortic coarctation.

CT Heart Function and Morphology

There is no relevant literature to support the use of CT heart function and morphology in the evaluation of suspected aortic coarctation, although its use was supported in adults by expert opinion in situations in which TEE windows are inadequate. The panel agreed that CT for function is not useful for a child in this clinical scenario.

CTA Chest

CTA chest is a complementary imaging modality in patients with suspected aortic coarctation after echocardiography. In patients with suboptimal arch imaging with echocardiography, CTA chest works as a great tool to assist with surgical planning [77-79]. Aortic coarctation is often associated with a hypoplastic aortic arch, and CTA chest is helpful in identifying the severity of arch hypoplasia, the length of the hypoplasia, and the relative origins of the aortic arch branch vessels in relation to the coarctation and arch hypoplasia [79].

CTA Coronary Arteries

There is no relevant literature to support the use of CTA coronary arteries in the evaluation of suspected aortic coarctation.

MRA Abdomen

There is no relevant literature to support the use of MRA abdomen in the evaluation of suspected aortic coarctation.

MRA Chest

MRA chest can be used as the second line of imaging after echocardiography in young children who do not require sedation and adolescents [79,80]. One advantage of MRA chest is that it can be combined with MRI heart function and morphology to quantify the degree of collaterals around the coarctation, more accurately estimate pressure gradients across the coarctation, and assess associated cardiac and conotruncal abnormalities [81,82].

MRI Heart Function and Morphology

MRI heart function and morphology can quantify the degree of collaterals around the coarctation. It can also be easily combined with an MRA chest to provide more comprehensive evaluation in patients with aortic coractation and associated cardiac and conotruncal abnormalities [75].

MRI Heart Function with Stress
There is no relevant literature to support the additional use of MRI heart function with stress in the evaluation of suspected aortic coarctation.

MRA Neck
There is no relevant literature to support the use of MRA neck in the evaluation of suspected aortic coarctation.

Radiography Chest
Radiography chest has limited clinical utility in diagnosis. Conventional signs on the chest radiograph may be subtle and not always easy to appreciate but may provide clues about the presence of an aortic coarctation.

SPECT or SPECT/CT MPI Rest and Stress
There is no relevant literature to support the use of SPECT or SPECT/CT MPI rest and stress in the evaluation of suspected aortic coarctation.

FDG-PET/CT Heart
There is no relevant literature to support the use of FDG-PET/CT heart in the evaluation of suspected aortic coarctation.

US Echocardiography Transesophageal
TEE is used for intraoperative imaging when other cardiac abnormalities are present in conjunction with CoA. The role of TEE for Doppler examination is limited because the ultrasound beam is almost perpendicular to the line of blood flow, leading to inaccuracies in velocity estimation.

Variant 10: Child. Known aortopathy or connective tissue disease. Surveillance of the aorta after inadequate or incomplete evaluation by transthoracic echocardiography. Next imaging study.

Dilatation of the aortic root and the ascending aorta is a key finding in connective tissue disorders, such as Marfan syndrome, Loeys-Dietzs syndrome, Ehlers-Danlos syndrome (especially the vascular form), and Turner syndrome. It is also a frequent finding in patients with bicuspid aortic valve and conotruncal abnormalities such as TOF, pulmonary atresia with VSD, or truncus arteriosus. The aorta is a part of the aortoventricular complex, comprising the systemic ventricle, the aortic valve, the aortic root, and the aortic vascular wall. Each component of this complex may influence the other components, thus introducing a dysfunction at multiple levels, often defined as "aortopathy."

Common manifestation of Marfan syndrome include annuloaortic ectasia with or without aortic valve insufficiency, aortic dissection, aortic aneurysm, pulmonary artery dilatation, and mitral valve prolapse. Aortic root aneurysms are present in up to 98% of patients with Loeys-Dietz syndrome, with thoracic aortic dissection being the leading cause of death (67%), followed by abdominal aortic dissection. Arterial tortuosity is a distinguishing feature of Loeys-Dietz syndrome, and there is a propensity for involvement of the extracranial carotid arteries and vertebral arteries [83]. Other cardiovascular abnormalities described in Loeys-Dietz syndrome include coronary artery aneurysms, pulmonary artery aneurysms, and aneurysms of the ductus arteriosus.

Ehlers-Danlos syndrome also predisposes to aortic dissections. Nearly 25% of people with bicuspid aortic valve may experience severe aortic valve dysfunction, ascending aortic aneurysm, cardiac death, hospital admission for heart failure, and aortic dissection or rupture [84].

Children with aortopathy require regular imaging surveillance to measure the aortic diameter, status of the aortic valve, and cardiac functional status. Current surgical mandate is based on aortic diameter cutoffs to decide the time of surgical intervention [85,86]. The guidelines from the American Heart Association provide thresholds for prophylactic replacement of the aorta, based primarily on aortic diameter or rapid aortic enlargement [87,88].

Arteriography Coronary with Ventriculography
Coronary artery ectasia can be an associated finding in many connective tissue diseases like bicuspid aortic valve [89], Marfan syndrome, and systemic lupus erythematosus. Coronary arteriography has been useful for imaging, but in current practice, it is used only if coronary CTA or coronary MRA provide insufficient information.

Arteriography Pulmonary
Pulmonary arteriography is seldom required for diagnosis or surveillance of pulmonary artery dilatation which can occur in Marfan syndrome.

CT Heart Function and Morphology

Although LV volume and ejection fraction can be assessed with retrospectively gated CT, LV volumes determined by CT are significantly higher than those measured with cine MRI and ejection fractions determined by using CT are significantly lower than those measured with cine MRI when β-blockers are administered before CT, because β-blockers significantly alter LV function [90].

CTA Chest

CTA chest may be used for imaging surveillance to measure the aortic diameter, and to screen for involvement of the arterial tree in the chest. CTA is especially useful in the emergent setting. ECG gating is critical for accurate assessment of the aortic root.

CTA Coronary Arteries

CTA is the most useful modality for the evaluation of coronary ectasia or aneurysms, which are often seen in the setting of connective tissue diseases. The diagnostic utility of CTA is very useful compared to coronary MRA in terms of spatial resolution and shorter scan duration.

FDG-PET/CT Heart

There is no relevant literature to support the use of FDG-PET/CT heart in the evaluation of suspected aortopathy or connective tissue disorder.

MRA Chest

MRA is the most useful modality for aortic assessment when TTE is suboptimal for assessment of aortopathy in pediatric patients due to its ability to screen multiple territories with the same contrast injection, including the head, neck, chest, and abdomen. To avoid multiple administrations of gadolinium-based contrast agents, follow-up imaging of the aortic root may be performed with noncontrast MRA techniques like cine gradient echo and navigator respiratory gated 3-D steady-state free precession. MRA may offer further valuable information, when complemented with phase-contrast flow data in bicuspid aortic valve, about the possibility of associated valvular regurgitation and alterations in shear stress that may predispose to progressive dilation [91-93].

MRA Neck

MRA neck is frequently performed at the same setting as MRA chest to screen for extracardiac vascular involvement in connective tissue disease and in all cases of aortopathy to measure vertebral artery tortuosity, which is used as a prognostic marker to determine risk of aortic dissection [94].

MRA Abdomen

MRA abdomen is frequently performed at the same setting as MRA chest at initial assessment to screen for extracardiac vascular involvement in connective tissue disease. MRA abdomen is also useful for regular follow-up of vessel involvement from connective tissue diseases [86].

MRI Heart Function and Morphology

MRI heart can be used to measure aortic valve regurgitant fraction and LV function, in patients with aortic root dilation that involves the aortic valve annulus, and in patients with coexisting aortic or mitral (rare in children) valve disease [95,96]. A small study shows some evidence that MRI could be useful for assessing connective tissue–related cardiomyopathy [97].

MRI Heart Function with Stress

There is no relevant literature to support the use of MR heart function with stress in the evaluation of aortopathy.

Radiography Chest

Chest radiography can be used to evaluate for emphysematous bullae and resultant pneumothoraces in the setting of connective tissue disorders [98]. However, chest radiography has no role in aortic surveillance.

SPECT or SPECT/CT MPI Rest and Stress

There is no relevant literature to support the use of SPECT or SPECT/CT MPI rest and stress in the evaluation of pediatric patients with aortopathy.

US Echocardiography Transesophageal

TEE can be supplementary modality for assessment of distal ascending aorta especially when TTE is suboptimal, but it is less useful than MRI or CT.

Summary of Recommendations

- **Variant 1**: Radiography chest, MRA chest without and with IV contrast, MRI heart function and morphology without and with IV contrast, MRI heart function and morphology without IV contrast, CTA chest with IV contrast, and CT heart function and morphology with IV contrast are usually appropriate for the next imaging study of a child or adult with repaired TOF or pulmonary valve stenosis with concern for pulmonary valve dysfunction or branch pulmonary artery stenosis and incomplete or inadequate assessment of cardiovascular morphology and function after TTE. These procedures may be complementary (ie, more than one procedure is ordered as a set or simultaneously in which each procedure provides unique clinical information to effectively manage the patient's care) in some settings. Radiography provides a global assessment of pulmonary blood flow, and location/integrity of stents and devices, MRA and CTA provide accurate vascular information, and MRI and CT for morphology and function provide comprehensive assessment of morphology, function and flow in the setting of an inadequate TTE. MRI also offers the ability to perform differential pulmonary blood flow quantification, and CT provides information on stent patency. Usually, MRA chest without and with IV contrast and CTA chest with IV contrast are equivalent alternatives (ie, only one procedure will be ordered to provide the clinical information to effectively manage the patient's care) whereas MRI heart function and morphology without and with IV contrast, MRI heart function and morphology without IV contrast, and CT heart function and morphology with IV contrast are generally equivalent alternatives.

- **Variant 2**: MRA chest without and with IV contrast, MRA chest without IV contrast, MRI heart function and morphology without and with IV contrast, MRI heart function and morphology without IV contrast, CTA chest with IV contrast, and CT heart function and morphology with IV contrast are usually appropriate for the next imaging study of a child or adult with TGA after atrial switch and incomplete or inadequate assessment of cardiovascular morphology and function after TTE. MRA chest without and with IV contrast, MRA chest without IV contrast, and CTA chest with IV contrast are equivalent alternatives (ie, usually only one procedure will be ordered to provide the clinical information to effectively manage the patient's care), whereas MRI heart function and morphology without and with IV contrast, MRI heart function and morphology without IV contrast, and CT heart function and morphology with IV contrast are generally equivalent alternatives.

- **Variant 3**: Radiography chest, MRA chest without and with IV contrast, MRA chest without IV contrast, MRI heart function and morphology without and with IV contrast, MRI heart function and morphology without IV contrast, MRI heart function with stress without and with IV contrast, CTA chest with IV contrast, and CT heart function and morphology with IV contrast are usually appropriate for the next imaging study of a child or adult with TGA after arterial switch and incomplete or inadequate assessment of cardiovascular morphology and function after TTE. MRI heart function with stress without and with IV contrast is used to screen for inducible myocardial ischemia and viability assessment. Usually, MRA chest without and with IV contrast, MRA chest without IV contrast, and CTA chest with IV contrast are equivalent alternatives (ie, usually only one procedure will be ordered to provide the clinical information to effectively manage the patient's care), whereas MRI heart function and morphology without and with IV contrast, MRI heart function and morphology without IV contrast, and CT heart function and morphology with IV contrast are generally equivalent alternatives. The panel did not agree on recommending MRI heart function with stress without IV contrast for the next imaging study of a child or adult with TGA after arterial switch and incomplete or inadequate assessment of cardiovascular morphology and function after TTE. There is insufficient medical literature to conclude whether or not these patients would benefit from MRI heart function with stress without IV contrast for this clinical scenario. This procedure in this patient population is controversial but may be appropriate.

- **Variant 4**: MRA chest without and with IV contrast, MRA chest without IV contrast, MRI heart function and morphology without and with IV contrast, MRI heart function and morphology without IV contrast, and CTA coronary arteries with IV contrast are usually appropriate for the next imaging study of a child with suspected or confirmed congenital or acquired coronary artery abnormality with incomplete or inadequate assessment of coronary morphology after TTE. MRA chest without and with IV contrast and MRA chest without IV contrast are equivalent alternatives (ie, only one procedure will be ordered to provide the clinical information to effectively manage the patient's care). MRI heart function and morphology without and with IV contrast provides information in myocardial viability and function, whereas CTA coronary arteries with IV contrast provides dynamic high-resolution assessment of coronary morphology across the cardiac cycle, and the procedures are complementary (ie, more than one procedure is ordered as a set or simultaneously in which each procedure provides unique clinical information to effectively manage the patient's care) to each other. The panel did not agree on recommending SPECT or SPECT or SPECT/CT MPI rest and stress for the next imaging study of a child with suspected or confirmed congenital or acquired coronary artery abnormality with incomplete or inadequate assessment of coronary morphology after TTE. There is insufficient medical literature to conclude whether or not these patients would benefit from SPECT or SPECT or SPECT/CT MPI rest and stress for this clinical scenario. This procedure in this patient population is controversial but may be appropriate.

- **Variant 5**: Arteriography coronary with ventriculography, arteriography pulmonary, MRA chest without and with IV contrast, MRA chest without IV contrast, MRI heart function and morphology without and with IV contrast, MRI heart function and morphology without IV contrast, CTA chest with IV contrast, and CT heart function and morphology with IV contrast are usually appropriate for the next imaging study of a child with known single ventricle physiology and preoperative evaluation for stage 2 single ventricle palliation and incomplete or inadequate assessment of cardiovascular morphology and function after TTE. These procedures are usually equivalent alternatives (ie, usually only one procedure will be ordered to provide the clinical information to effectively manage the patient's care).
- **Variant 6**: Arteriography coronary with ventriculography, arteriography pulmonary, MRA chest without and with IV contrast, MRI heart function and morphology without and with IV contrast, MRI heart function and morphology without IV contrast, CTA chest with IV contrast, and CT heart function and morphology with IV contrast are usually appropriate for the next imaging study of a child with known single ventricle physiology and preoperative evaluation for stage 3 single ventricle palliation (total cavopulmonary connection) and incomplete or inadequate assessment of cardiovascular morphology and function after TTE. These procedures are usually equivalent alternatives (ie, usually only one procedure will be ordered to provide the clinical information to effectively manage the patient's care).
- **Variant 7**: MRA chest without and with IV contrast, MRA chest without IV contrast, MRI heart function and morphology without and with IV contrast, MRI heart function and morphology without IV contrast, CTA chest with IV contrast, and CT heart function and morphology with IV contrast are usually appropriate for the next imaging study of a child or adult with known single ventricle physiology and postoperative evaluation after stage 3 single ventricle palliation (total cavopulmonary connection) and incomplete or inadequate assessment of cardiovascular morphology and function after TTE. These procedures are usually equivalent alternatives (ie, usually only one procedure will be ordered to provide the clinical information to effectively manage the patient's care).
- **Variant 8**: MRA chest without and with IV contrast, MRA chest without IV contrast, MRI heart function and morphology without and with IV contrast, MRI heart function and morphology without IV contrast, and CTA chest with IV contrast are usually appropriate for the next imaging study of a child or adult with known or suspected anomalous pulmonary venous return with inadequate evaluation after TTE. MRA chest without and with IV contrast, MRA chest without IV contrast, and CTA chest with IV contrast are equivalent alternatives (ie, usually only one procedure will be ordered to provide the clinical information to effectively manage the patient's care). MRI heart function and morphology without IV contrast provides additional information on ventricular size and function and Qp/Qs (pulmonary to systemic flow ratio) assessment.
- **Variant 9**: MRA chest without and with IV contrast, MRA chest without IV contrast, MRI heart function and morphology without and with IV contrast, MRI heart function and morphology without IV contrast, and CTA chest with IV contrast are usually appropriate for the next imaging study of a child or adult with suspected aortic coarctation with inadequate evaluation after TTE. MRA chest without and with IV contrast, MRA chest without IV contrast, and CTA chest with IV contrast are equivalent alternatives (ie, usually only one procedure will be ordered to provide the clinical information to effectively manage the patient's care). MRI heart function and morphology without IV contrast provides additional information on ventricular size and function and myocardial mass. The panel did not agree on recommending CT heart function and morphology with IV contrast for the next imaging study of a child or adult with suspected aortic coarctation with inadequate evaluation after TTE. There is insufficient medical literature to conclude whether or not these patients would benefit from CT heart function and morphology with IV contrast for this clinical scenario. This procedure in children is controversial but may be appropriate in adults.

- **Variant 10**: MRA abdomen without and with IV contrast, MRA abdomen with IV contrast, MRA chest without and with IV contrast, MRA chest without IV contrast, MRA neck without and with IV contrast, MRA neck without IV contrast, and CTA chest with IV contrast are usually appropriate for the next imaging study of a child with known aortopathy or connective tissue disease and surveillance of the aorta after inadequate or incomplete evaluation by TTE. Although MRA and CTA in a single region (neck, chest, or abdomen) may be equivalent alternatives (ie, only one procedure will be ordered to provide the clinical information to effectively manage the patient's care), it is common to require screening of multiple body regions in this setting.

Supporting Documents

The evidence table, literature search, and appendix for this topic are available at https://acsearch.acr.org/list. The appendix includes the strength of evidence assessment and the final rating round tabulations for each recommendation.

For additional information on the Appropriateness Criteria methodology and other supporting documents go to www.acr.org/ac.

Appropriateness Category Names and Definitions

Appropriateness Category Name	Appropriateness Rating	Appropriateness Category Definition
Usually Appropriate	7, 8, or 9	The imaging procedure or treatment is indicated in the specified clinical scenarios at a favorable risk-benefit ratio for patients.
May Be Appropriate	4, 5, or 6	The imaging procedure or treatment may be indicated in the specified clinical scenarios as an alternative to imaging procedures or treatments with a more favorable risk-benefit ratio, or the risk-benefit ratio for patients is equivocal.
May Be Appropriate (Disagreement)	5	The individual ratings are too dispersed from the panel median. The different label provides transparency regarding the panel's recommendation. "May be appropriate" is the rating category and a rating of 5 is assigned.
Usually Not Appropriate	1, 2, or 3	The imaging procedure or treatment is unlikely to be indicated in the specified clinical scenarios, or the risk-benefit ratio for patients is likely to be unfavorable.

Relative Radiation Level Information

Potential adverse health effects associated with radiation exposure are an important factor to consider when selecting the appropriate imaging procedure. Because there is a wide range of radiation exposures associated with different diagnostic procedures, a relative radiation level (RRL) indication has been included for each imaging examination. The RRLs are based on effective dose, which is a radiation dose quantity that is used to estimate population total radiation risk associated with an imaging procedure. Patients in the pediatric age group are at inherently higher risk from exposure, because of both organ sensitivity and longer life expectancy (relevant to the long latency that appears to accompany radiation exposure). For these reasons, the RRL dose estimate ranges for pediatric examinations are lower as compared with those specified for adults (see Table below). Additional information regarding radiation dose assessment for imaging examinations can be found in the ACR Appropriateness Criteria® Radiation Dose Assessment Introduction document [99].

Relative Radiation Level Designations		
Relative Radiation Level*	**Adult Effective Dose Estimate Range**	**Pediatric Effective Dose Estimate Range**
O	0 mSv	0 mSv
☢	<0.1 mSv	<0.03 mSv
☢☢	0.1-1 mSv	0.03-0.3 mSv
☢☢☢	1-10 mSv	0.3-3 mSv
☢☢☢☢	10-30 mSv	3-10 mSv
☢☢☢☢☢	30-100 mSv	10-30 mSv
*RRL assignments for some of the examinations cannot be made, because the actual patient doses in these procedures vary as a function of a number of factors (eg, region of the body exposed to ionizing radiation, the imaging guidance that is used). The RRLs for these examinations are designated as "Varies."		

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