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APPROPRIATE USE CRITERIA

ACC/AHA/ASE/HRS/ISACHD/SCAI/SCCT/SCMR/SOPE 2020 Appropriate Use Criteria for Multimodality Imaging During the Follow-Up Care of Patients With Congenital Heart Disease

A Report of the American College of Cardiology Solution Set Oversight Committee and Appropriate Use Criteria Task Force, American Heart Association, American Society of Echocardiography, Heart Rhythm Society, International Society for Adult Congenital Heart Disease, Society for Cardiovascular Angiography and Interventions, Society of Cardiovascular Computed Tomography, Society for Cardiovascular Magnetic Resonance, and Society of Pediatric Echocardiography

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ABSTRACT

The American College of Cardiology (ACC) collaborated with the American Heart Association, American Society of Echocardiography, Heart Rhythm Society, International Society for Adult Congenital Heart Disease, Society for Cardiovascular Angiography and Interventions, Society of Cardiovascular Computed Tomography, Society for Cardiovascular Magnetic Resonance, and the Society of Pediatric Echocardiography to develop Appropriate Use Criteria (AUC) for multimodality imaging during the follow-up care of patients with congenital heart disease (CHD). This is the first AUC to address cardiac imaging in adult and pediatric patients with established CHD.

A number of common patient scenarios (also termed "indications") and associated assumptions and definitions were developed using guidelines, clinical trial data, and expert opinion in the field of CHD (1). The indications relate primarily to evaluation before and after cardiac

surgery or catheter-based intervention, and they address routine surveillance as well as evaluation of new-onset signs or symptoms. The writing group developed 324 clinical indications, which they separated into 19 tables according to the type of cardiac lesion. Noninvasive cardiac imaging modalities that could potentially be used for these indications were incorporated into the tables, resulting in a total of 1,035 unique scenarios. These scenarios were presented to a separate, independent panel for rating, with each being scored on a scale of 1 to 9, with 1 to 3 categorized as "Rarely Appropriate," 4 to 6 as "May Be Appropriate," and 7 to 9 as "Appropriate." Forty-four percent of the scenarios were rated as Appropriate, 39% as May Be Appropriate, and 17% as Rarely Appropriate.

This AUC document will provide guidance to clinicians in the care of patients with established CHD by identifying the reasonable imaging modality options available for evaluation and surveillance of such patients. It will also serve as an educational and quality improvement tool to identify patterns of care and reduce the number of Rarely Appropriate tests in clinical practice.

ABBREVIATIONS

AR = aortic regurgitation

AS = aortic stenosis

ASD = atrial septal defect

AV = atrioventricular

AVSD = atrioventricular septal defect

ccTGA = congenitally corrected transposition of the great arteries

CCT = cardiovascular computed tomography

CHD = congenital heart disease

CMR = cardiovascular magnetic resonance

DORV = double outlet right ventricle

ES = Eisenmenger syndrome

LV = left ventricle

LVOT = left ventricular outflow tract

LV-to-PA = left ventricle to pulmonary artery

MR = mitral regurgitation

MS = mitral stenosis

MVP = mitral valve prolapse

PA = pulmonary artery

PA/IVS = pulmonary atresia with intact ventricular septum

PAPVC = partial anomalous pulmonary venous connection

PDA = patent ductus arteriosus

PFO = patent foramen ovale

PH = pulmonary hypertension

PR = pulmonary regurgitation

PS = pulmonary stenosis

PVR = pulmonary valve replacement

RV = right ventricle

RVOT = right ventricular outflow tract

RV-to-PA = right ventricle to pulmonary artery

TA = truncus arteriosus

TAPVC = total anomalous pulmonary venous connection

TEE = transesophageal echocardiogram

TGA = transposition of the great arteries

TOF = tetralogy of Fallot

TR = tricuspid regurgitation

TTE = transthoracic echocardiogram

VSD = ventricular septal defect

PREFACE

The ACC has a long history of developing documents (e.g., decision pathways, health policy statements, appropriate use criteria) to provide members with guidance on both clinical and nonclinical topics relevant to cardiovascular care. In most circumstances, these documents have been created to complement clinical practice guidelines and to inform clinicians about areas where evidence may be new and evolving or where sufficient data may be more limited. In spite of this, numerous care gaps continue to exist, highlighting the need for more streamlined and efficient processes to implement best practices in service to improved patient care.

Central to the ACC's strategic plan is the generation of "actionable knowledge"—a concept that places emphasis on making clinical information easier to consume, share, integrate, and update. To this end, the ACC has evolved from developing isolated documents to the development of integrated "solution sets." Solution sets are groups of closely related activities, policy, mobile applications, decision support, and other tools necessary to transform care and/or improve heart health. Solution sets address key questions facing care teams and attempt to provide practical guidance to be applied at the point of care. They use both established and emerging methods to disseminate information for cardiovascular conditions and their related management. The success of the solution sets rests firmly on their ability to have a measurable impact on the delivery of care. Because solution sets reflect current evidence and ongoing gaps in care, the associated tools will be refined over time to best match member needs.

AUC represent a key component of solution sets. They consist of common clinical scenarios associated with a given disease state and ratings that help to define when it is reasonable to perform testing and, importantly, when it is not. The methodology for AUC is grounded in appointing

content development work groups, which create patient scenarios, and independent rating panels, which employ a modified Delphi process to rate the relevant options for testing and intervention as Appropriate, May Be Appropriate, or Rarely Appropriate. AUC should not replace clinician judgment and practice experience, but should function as a tool to improve patient care and health outcomes in a cost-effective manner.

Ty J. Gluckman, MD, FACC
Chair, ACC Solution Set Oversight Committee

1. INTRODUCTION

Significant advances in the diagnosis and treatment of patients with CHD have resulted in improved survival and outcomes (2,3). However, many patients require lifelong follow-up to monitor sequelae following cardiac surgery or catheter-based intervention, or the development of complications such as valvular and ventricular dysfunction (4). Noninvasive cardiac imaging plays a key role in the diagnosis and follow-up of these patients. While transthoracic echocardiography (TTE) remains the cornerstone of cardiac imaging in patients with CHD, other imaging modalities, such as transesophageal echocardiography (TEE), cardiovascular magnetic resonance (CMR), cardiovascular computed tomography (CCT), and stress imaging, also play an important role in anatomic and functional assessment (5–9). Although these tests can all be used for patients with CHD, there is a paucity of lesion-specific guidance for clinicians to utilize as decision support tools (1,10,11). Furthermore, there is significant variability in the frequency with which these imaging modalities are used during follow-up.

AUC have been established to guide clinicians in the use of imaging modalities in clinical practice with an overall goal of improving patient care and outcomes in a cost-effective manner. The first pediatric cardiology AUC document focused on the initial outpatient TTE evaluation of patients age ≤18 years without established heart disease (12). It did not address evaluation of children and adults with CHD or the use of cardiac imaging modalities other than TTE. Other AUC documents focused on cardiovascular conditions in adult patients have addressed very few indications related to CHD (13–15). Therefore, the purpose of this document is to delineate the appropriate use of various cardiac imaging modalities for surveillance and evaluation of patients with established CHD. The scope of this document has been purposefully limited to the more common scenarios encountered in routine practice.

2. METHODS

To begin the AUC process, a writing group of multidisciplinary experts was formed to identify and categorize common clinical scenarios for pediatric and adult patients with CHD. This group of representatives from several cardiovascular subspecialty societies and ACC councils consisted of pediatric and adult CHD experts, including cardiac imagers with expertise in specific cardiac imaging modalities. The goal of the writing group was to choose common patient scenarios experienced in clinical practice and to categorize these scenarios on the basis of specific cardiac lesions, giving due consideration to patient age, clinical course, and surgical and catheter-based interventions. The writing group focused on identifying the most typical situations encountered in routine practice to avoid making the document excessively long.

Once the indications were drafted, they were critiqued by numerous external reviewers representing a variety of cardiovascular subspecialty societies and ACC councils. After the writing group incorporated this initial feedback, the indications were sent to an independent rating panel comprising additional experts specializing in CHD. Also provided to the rating panelists was a Guideline Mapping and References document, where the indications were mapped to relevant guidelines, clinical trials, and other key references in the field (see Guideline Mapping and References). Of note, the 2018 AHA/ACC Guideline for the Management of Adults with Congenital Heart Disease was used in the Guideline Mapping exercise, but when the indication and/or frequency of imaging was unavailable in this guideline, then the 2008 Guideline for the Management of Adults with Congenital Heart Disease was used (1,16).

Next, the rating panelists were tasked with scoring the clinical scenarios from 1 through 9, with 1 to 3 classified as "Rarely Appropriate care," 4 to 6 representing "May Be Appropriate care," and 7 to 9 classified as "Appropriate care." Rating panel members conducted this scoring via an electronic survey platform, and the median score from the 17 panelists was calculated for each scenario. Next, the panelists, several writing group representatives, and a moderator gathered for an in-person rating panel meeting, where robust discussion of each indication ensued, and feedback was given to the writing group representatives. The writing group then took this input and completed further vetting of the clinical scenarios, before sending the document back to the rating panel for an additional round of electronic scoring. When some of the scores came back in misalignment with guideline recommendations and other evidence, the writing group elaborated and provided further evidence to clarify the clinical scenarios. This additional evidence was then offered to the rating panelists and a final round of scoring commenced (see Final Deidentified AUC Scores). These multiple rounds of review and revision by independent groups ensured that numerous clinician viewpoints were heard and considered.

A detailed description of the methods used for rating the clinical scenarios can be found in previous AUC methodology

publications, including the ACC Appropriate Use Criteria Methodology: 2018 Update (17). Briefly, this process combines evidence-based medicine and practice experience and engages a rating panel in a modified Delphi exercise. For the scoring, care was taken to provide the rating panel with objective, unbiased information, including guidelines and key references in the field (see Guideline Mapping and References). Other steps of the modified Delphi process are convening a formal writing group with diverse expertise in the treatment of CHD, circulating the indications for external review before sending the indications to the rating panel, and establishing a moderator for facilitating panel interaction at the face-to-face meeting.

In scoring the clinical scenarios, the rating panel was asked to assess whether the different modality options for each indication should be categorized as Appropriate, May Be Appropriate, or Rarely Appropriate. It should be emphasized that each modality option was not ranked in comparison with the others or based on physician preference but was instead considered on its own merits and reasonableness for the given clinical scenario. When the panelists scored the indications, they were provided the following definition of appropriate use.

An appropriate test is one for which the potential benefits, in terms of survival or health outcomes (symptoms, functional status, and/or quality of life), exceed the potential negative consequences.

Median Score 7-9: *Appropriate care* for a specific indication (test **is** generally acceptable and **is** a reasonable approach for the indication).

An appropriate option for management of patients in this population due to benefits generally outweighing risks; an effective option for individual care plans, although not always necessary depending on physician judgment and patient-specific preferences (i.e., test is generally acceptable and is generally reasonable for the indication).

Median Score 4-6: *May Be Appropriate care* for specific indication (test **may** be generally acceptable and **may** be a reasonable approach for the indication). May Be Appropriate also implies that more research and/or patient information is needed to classify the indication definitively.

At times an appropriate option for management of patients in this population due to variable evidence or agreement regarding the benefit/risk ratio, potential benefit based on practice experience in the absence of published evidence, and/or variability in the population. Effectiveness for individual care must be determined by a patient's physician in consultation with the patient and on the basis of additional clinical variables and judgment along with patient preferences (i.e., test may be acceptable and may be reasonable for the indication).

Median Score 1-3: *Rarely Appropriate care* for specific indication (test **is not** generally acceptable and **is not** a reasonable approach for the indication).

Rarely an appropriate option for management of patients in this population due to the lack of a clear benefit/risk advantage; rarely an effective option for individual care plans. Exceptions should have documentation of the clinical reasons for proceeding with this care option (i.e., test is not generally acceptable and is not generally reasonable for the indication).

The scenarios included in this document are based on our current understanding of patient outcomes plus the potential benefits compared with risks of the imaging strategies involved. Each patient should be treated individually in accordance with their own particular needs. It is expected that clinicians will occasionally care for patients with unique conditions that could result in the use of a Rarely Appropriate test. When this occurs, clinicians should document the specific situation and patient characteristics leading to this decision. Thus, the AUC document should not be used as a deterrent for treating the patient or denial of reimbursement. While a Rarely Appropriate or May Be Appropriate designation should not prevent a test from being performed, an Appropriate designation is also not a requirement that a given test be performed. The AUC are offered to guide patient care but should not be considered a substitute for sound clinical judgement and practice experience.

3. ASSUMPTIONS AND SCOPE

1. This document addresses only the follow-up of patients with established CHD using various cardiovascular imaging modalities. It is assumed that a complete anatomic cardiac diagnosis has been established by a qualified clinician using 1 or more modalities. The initial evaluation by TTE prompted by signs and symptoms suggesting CHD has been addressed in the 2014 AUC for Initial Transthoracic Echocardiography in Outpatient Pediatric Cardiology (12) and is not included in this document, nor are scenarios in critical care settings that may require frequent imaging.
2. The goal of this document is to determine the modalities that may be reasonable for a specific indication and not to designate the single best modality or the rank order of modalities. For each indication, the rating reflects whether the imaging modality is reasonable for the patient according to the appropriate use definition. As such, more than 1 imaging modality or even all modalities may have the same rating for a given clinical indication. Although in clinical practice the choice of initial imaging modality may alter the appropriateness of the other modalities, this was not taken into account when rating individual imaging modalities. If an imaging modality was deemed to be highly unlikely to be used in the given

scenario, it was not rated at all and is represented by gray shading in the tables.

3. The range of indications for cardiovascular imaging of CHD is quite large; therefore, the indications included are purposefully broad to cover an array of cardiovascular diseases, signs, and symptoms, and to account for the ordering physician's best judgment as to the presence of cardiovascular abnormalities. Indications related to surveillance imaging following complete repair of simple lesions without sequelae do not address duration of follow-up. This does not imply indefinite follow-up in such cases, and clinicians should base this decision on available guidelines. The indications in this document primarily address a single heart lesion and its common associations; therefore, this document does *not* include:
   - Combinations of CHD such as truncus arteriosus with an interrupted aortic arch or tetralogy of Fallot with an atrioventricular canal defect
   - Rare CHD or case scenarios, rare surgical or catheter-based interventions, and intraprocedural imaging (e.g., intracardiac echocardiography or transesophageal echocardiogram guidance during cardiac surgery or catheter-based procedures)
   - Cardiomyopathies, myocarditis, myocardial bridge, cardiac tumors, acquired heart disease, infective endocarditis in the absence of CHD, or cardiac involvement with systemic disorders (e.g., systemic lupus erythematosus)
   - Evaluation before consideration of cardiac transplantation, electrophysiology procedure, placement of a pacemaker and/or implanted cardioverter defibrillator or ventricular assist device, noncardiac surgery, sports participation, or military recruitment
   - Evaluation before consideration of pregnancy, during pregnancy, or in the immediate postpartum period in women with CHD.
   - Patients who were lost to follow-up and need to have their diagnoses re-established
   - Clinical scenarios that are unlikely to exist in current practice in the United States
4. Although this document examines the use of various cardiovascular imaging modalities in specific scenarios, it does not address the specific techniques associated with each modality. The following conditions are assumed:
   - A comprehensive TTE examination may include 2-dimensional, 3-dimensional, M-mode, and strain imaging; color, spectral, and tissue Doppler imaging; and use of contrast agents for opacification, when applicable.
   - The "stress imaging" modality includes echocardiography and myocardial perfusion imaging (single-photon emission computed tomography, positron emission tomography, or magnetic resonance imaging [MRI]) performed with exercise or pharmacological stress designed to provoke ischemia. Stress imaging may also refer to echocardiography performed with exercise or dobutamine to assess the severity of valvular or outflow tract obstruction or changes in Doppler-derived RV pressure estimates. These tests should be considered under "stress imaging" rather than the underlying modality (e.g., echocardiography or MRI).
   - Other diagnostic tests, such as electrocardiography and chest x-ray, and imaging of noncardiovascular structures (e.g., transcranial Doppler or hepatic imaging) are not addressed in this document.
5. Use of imaging modalities may be influenced by local availability of technology and qualified professional staff. Especially for newer modalities, technology and professional capabilities may vary by institution. The most current technology and best practices are used to determine the appropriateness of indications for each modality. For example, while rating the appropriateness of CCT, current-generation scanners that use significantly lower radiation dose have been considered (18). The level of appropriateness does not consider issues of local availability or expertise for a given modality.
6. X-ray angiography during cardiac catheterization has been excluded from this document due to the focus on noninvasive imaging modalities.
7. When assessment of a patient by a particular modality is known to have limitations (e.g., poor acoustic windows for TTE, contrast agent allergy for CCT, or implanted device-related image artifact for CMR), alternative modalities should be considered and may take precedence on the basis of the clinician's judgment (19).
8. Comprehensive risk assessment is assumed throughout the document when rating imaging modalities. This includes consideration of the combined and cumulative risks of anesthesia, vascular access, contrast agents, and radiation exposure (20–22). Anesthesia risk includes both the procedural risk of adverse events and the potential for adverse long-term neurodevelopmental outcomes (20,23–25). Vascular access risk for noninvasive imaging is related to peripheral intravenous catheter insertion, which has been shown to be safe in patients of all ages, with a low rate of complications such as extravasation. Contrast agent risk includes allergic reaction or renal toxicity from iodinated agents and nephrogenic systemic fibrosis and the undefined risk of neuronal deposition with gadolinium-based agents (21,26). Radiation exposure in children has been linked to later development of cancer (27,28). Patients with CHD have an

increased risk of developing cancer compared with healthy individuals (29). Radiation dose estimates from CCT scans in children are highly variable in the current era and are related to the scanner platforms and the aggressiveness of radiation dose optimization (18). For CMR, an implanted electronic device (pacemaker or defibrillator) must be thoroughly evaluated to ensure that it does not pose a significant safety risk. The comprehensive risk profile for the imaging modalities should be carefully factored into decisions regarding each clinical scenario.

9. This document refers to patients as infants, children, or adults. Adolescents are not specifically mentioned; however, indications that include CMR and CCT for adults may be applied to compliant adolescents.
10. It is assumed that a comprehensive history and physical examination have been performed by a qualified clinician and that the signs and symptoms accurately represent the current patient status. It is also assumed that the tests are ordered by clinicians knowledgeable in CHD, and the tests are performed and interpreted by qualified personnel in a facility compliant with national standards for the relevant modality.
11. Clinicians should use their best judgment and available clinical practice guidelines to determine the clinical significance and severity of cardiovascular diagnoses before classifying the appropriateness of the indications. Clear documentation of the reason for ordering the test should be included in the medical record. If the reason for a test can be assigned to more than 1 indication, the justification should be classified under the most clinically significant indication.
12. Indications related to routine surveillance have a time range specified for testing. This does not imply that an imaging modality rated Appropriate is mandatory to perform during every follow-up visit. It also does not imply that the test should be performed exactly within the specified time interval, as there could be clinical or social circumstances precluding performance of the test within this time period. The clinician and the patient should have the flexibility to obtain the test at a time that is reasonably close to the specified time period. For some indications, a broad time range for surveillance has been presented to cover a wider spectrum of symptomatic patients (e.g., heart failure). It is assumed that clinicians will use their best judgment in such cases and weigh the risks and benefits of various modalities before ordering a given test.
13. Cost is considered implicitly in the appropriate use determination. Clinical benefits should always be considered first, and costs should be considered in relationship to these benefits in order to convey net value.

4. DEFINITIONS

Anatomic and surgical nomenclature are primarily adapted from the International Pediatric and Congenital Cardiac Code (30).

Adult: A person older than 18 years.

Aortic regurgitation (AR): Congenital or acquired cardiovascular malformation of the aortic valve allowing retrograde flow into the ventricle (30).

Aortic stenosis (AS): A congenital or acquired cardiovascular malformation of the aortic valve in which there is narrowing or obstruction of flow (30).

Arrhythmia: Documented irregular and/or abnormal heart rate or rhythm (12).

Arterial switch operation: An operation used for transposition of the great arteries that involves translocation of the aorta from its attachment to the RV and of the pulmonary artery (PA) from the left ventricle (LV) and reattachment of the great arteries to the contralateral ventricles with reimplantation of the coronary arteries into the neoaorta. This results in the LV supporting the systemic circulation and the RV supporting the pulmonary circulation. A LeCompte procedure is often performed, which involves translocation of the PA confluence anterior to the ascending aorta.

Asymptomatic: Lacking characteristic signs and/or symptoms for a given condition.

Atrial septal defect (ASD): A congenital cardiac malformation in which there is a hole or pathway between the atrial chambers (30).

- *Small ASD*: Likely to be hemodynamically insignificant.
- ≥ *Moderate ASD*: Likely to be hemodynamically significant.

Atrial switch operation: An operation that redirects systemic and pulmonary venous return to the contralateral ventricle.

Atrioventricular septal defect (AVSD) or *atrioventricular canal defect*: This cardiac defect is subcategorized into complete, transitional, and partial AVSD (30).

- *Complete AVSD:* A congenital cardiac malformation in which both atria connect to a common atrioventricular valve, which characteristically has 4 or 5 leaflets, including superior and inferior bridging leaflets, and a single annulus. There is an interatrial communication just above the atrioventricular valve and an interventricular communication just below it.
- *Partial AVSD*: A congenital cardiac malformation comprising an interatrial communication just above the atrioventricular valve, no interventricular communication just below the atrioventricular valve, separate right and left atrioventricular valvular orifices, and varying degrees of malformation of the left-sided component of the common atrioventricular valve.

- *Transitional AVSD*: A variant of complete AVSD with an interatrial communication immediately above the atrioventricular valve, and a restrictive interventricular communication immediately below the atrioventricular valve.

Cardiovascular computed tomography (CCT): Computed tomography imaging of the heart using electrocardiographic gating or electrocardiographic triggering. Coronary CT angiography refers to contrast enhanced tomographic visualization of the coronary arteries. For this document, the general term "CCT" will be used and will include imaging of the cardiac structures, vasculature, or coronary arteries. No differentiation will be made for gating techniques, triggering, or use of contrast (31).

Cardiovascular magnetic resonance: Magnetic resonance imaging of the heart and blood vessels. In the context of this document, imaging with concurrent intravascular catheterization is not included. Stress protocols including MRI are considered separately under "stress imaging."

Child: A person age 1 to 18 years (32).

Coarctation of the aorta: A congenital cardiovascular malformation in which there is a luminal narrowing most commonly of the junction between the aortic arch and the descending aorta (30).

Congenitally corrected transposition of the great arteries (ccTGA) or L-loop TGA: A congenital cardiovascular malformation of atrioventricular and ventriculoarterial discordance, in which the right atrium connects to the LV, the left atrium connects to the RV, the RV connects to the aorta, and the LV connects to the pulmonary trunk (30).

Coronary anomaly: Coronary anomalies include congenital anomalous origin and/or course of the coronary arteries, and coronary arteriovenous or coronary-cameral fistulae in the setting of normal conotruncal anatomy. It excludes myocardial bridges, coronary artery reimplantation after Ross procedure and arterial switch operation, and coronary anomalies related to supravalvular aortic stenosis.

Double-chambered RV: A congenital cardiac malformation in which the RV is divided into 2 chambers—1 inferior that includes the inlet and trabecular portions of the RV, and 1 superior that includes the trabecular portion and infundibulum.

Double outlet right ventricle (DORV): A congenital cardiovascular malformation in which both great arteries arise entirely or predominantly from the RV (30).

Double switch procedure: An operation typically performed in patients with congenitally corrected transposition of the great arteries that results in anatomic correction of the atrial-ventricular and ventricular-great arterial relationships so that the LV supports the systemic circulation. It includes an arterial switch operation and an atrial switch operation (1).

Ebstein anomaly: A congenital cardiac malformation of the tricuspid valve and RV that is characterized by apical displacement of the functional annulus, usually involving the septal and inferior (posterior) leaflets (30).

Eisenmenger syndrome (ES): Pulmonary arterial hypertension in the setting of CHD with elevation of pulmonary vascular resistance, resulting in bidirectional or right-to-left shunting through a systemic-to-PA connection or septal defect and cyanosis (33).

Evaluation: The use of an imaging test prompted by concern for structural or functional changes since the time of last follow-up. This contrasts with surveillance, in which a test is ordered following a certain prespecified time period.

Fluoroscopy: Imaging technique that uses x-rays to obtain real-time moving images.

Fontan procedure: A palliative procedure for patients with single-ventricle circulation that involves diversion of systemic venous return directly to the PA, usually without the interposition of a subpulmonary ventricle.

Glenn procedure: Direct anastomosis of the superior vena cava to a branch pulmonary artery.

Heart failure: A clinical and pathophysiological syndrome that results from ventricular dysfunction, volume, or pressure overload, whether alone or in combination (34).

Infant: A person age less than 1 year (32).

Interrupted aortic arch: A congenital cardiovascular malformation in which there is no luminal continuity between the ascending and descending aorta (30).

Lung scan: A nuclear scan to measure circulation (perfusion) in all areas of the lungs (35).

Mild, moderate, or severe valvular disease: Severity of the lesion is determined by the clinician on the basis of existing clinical practice guidelines and other clinical policy recommendations (36-40).

Mitral regurgitation (MR): A congenital or acquired cardiac finding of retrograde flow through the mitral valve (30).

Mitral stenosis (MS): A congenital or acquired cardiac malformation of narrowing or stricture of the orifice of the mitral valve (obstruction to flow) (30).

Mitral valve prolapse (MVP): A congenital or acquired cardiac malformation of the mitral valve in which 1 or both leaflets move to the atrial side of the plane of the annulus in systole (30).

Partial anomalous pulmonary venous connection (PAPVC): A congenital cardiovascular malformation in which 1 or more (but not all) of the pulmonary veins connect anomalously to the right atrium or to 1 or more of its venous tributaries and the remaining pulmonary veins connect to the left atrium (30).

Patent ductus arteriosus (PDA): A congenital cardiovascular finding in which the arterial duct (ductus arteriosus) is open beyond the normal age of spontaneous closure (30).

- *Trivial and silent PDA*: Hemodynamically insignificant; murmur not heard on auscultation.

- *Small and audible PDA*: Likely to be hemodynamically insignificant; murmur heard on auscultation.
- ≥*Moderate PDA*: Likely to be hemodynamically significant.

Patent foramen ovale (PFO): A congenital cardiovascular finding in which there is a small interatrial communication (or potential communication) confined to the region of the fossa ovalis without deficiency of the septum primum or septum secundum (30).

Patient: An individual of any age.

Postoperative: Within 30 days of surgical intervention.

Postprocedural: Within 30 days of surgical or catheter-based interventions.

Procedural: Surgical or catheter-based interventions.

Pulmonary atresia with intact ventricular septum: A congenital cardiovascular malformation in which there is no opening between the RV and the pulmonary trunk, no ventricular level communication, and normally aligned great arteries (30).

Pulmonary hypertension (PH): A resting mean PA pressure ≥25 mm Hg (33).

Pulmonary stenosis (PS): A congenital cardiovascular malformation of the pulmonary valve in which there is narrowing or stricture (obstruction to flow) (30).

Pulmonary regurgitation (PR): Congenital or acquired cardiovascular malformation of the pulmonary valve allowing retrograde flow into the ventricle (30).

Rastelli procedure: An operation for transposition of the great arteries, ventricular septal defect, and (sub)pulmonary stenosis. The operation involves redirection of ventricular outflows; an intracardiac baffle tunnels the LV to the aorta, and an external conduit connects the RV and the PA (41).

Ross procedure: A method of aortic valve replacement that involves autograft transplantation of the pulmonary valve, annulus, and main PA into the aortic position with reimplantation of the coronary ostia into the neoaorta. The RV outflow tract is usually reconstructed with a valved conduit (1).

Secundum atrial septal defect: A congenital cardiac malformation in which there is an interatrial communication confined to the region of the oval fossa (fossa ovalis), most commonly owing to a deficiency of the primary atrial septum (septum primum), but deficiency of the septum secundum (superior interatrial fold) may also contribute (30).

Single-ventricle heart disease: A spectrum of congenital cardiac malformations in which one of the ventricular chambers is hypoplastic (underdeveloped) or the ventricular chambers are not readily partitioned to allow one ventricle to pump to the systemic circulation and another to the pulmonary circulation (30).

Sinus venosus defect: A congenital cardiovascular malformation in which there is a caval vein (vena cava) and/or pulmonary vein (or veins) that overrides the atrial septum, producing an interatrial or anomalous venoatrial communication (30).

Stage I single-ventricle palliation: A surgical or catheter-based intervention performed to provide adequate pulmonary or systemic blood flow involving a systemic-to-PA shunt; PDA stent; Norwood procedure with a systemic-PA shunt or a RV-to-PA conduit; PA band; or a hybrid procedure that includes PA bands and a PDA stent.

Stress imaging: Echocardiography and myocardial perfusion imaging (single photon emission computed tomography, positron emission tomography, or MRI) performed with exercise or pharmacological stress designed to provoke ischemia. Stress imaging may also refer to echocardiography performed with exercise or dobutamine to assess the severity of valvular or outflow tract obstruction or changes in Doppler-derived RV pressure estimates.

Subvalvular aortic stenosis: A congenital cardiovascular malformation associated with narrowing within the outflow tract below the aortic valve (30).

Supravalvular aortic stenosis: A congenital cardiovascular malformation associated with narrowing of the aorta at the level of the sinotubular junction. This narrowing may extend into the ascending aorta (30).

Surveillance: A test being considered for a periodic evaluation because a certain period of time has elapsed, and clinically important changes may be detected in the absence of concerning findings on physical examination and history.

Symptomatic: Demonstrating characteristic signs and/or symptoms of a given disease (14).

Tetralogy of Fallot (TOF): A combination of congenital cardiac malformations resulting from anterosuperior deviation of the conal (outlet) septum or its fibrous remnant leading to narrowing or atresia of the pulmonary outflow, a malaligned ventricular septal defect, aortic override of the ventricular septal crest, and RV hypertrophy.

Total anomalous pulmonary venous connection (TAPVC): A congenital cardiovascular malformation in which none of the pulmonary veins connect to the left atrium (30).

Transesophageal echocardiography (TEE): Ultrasound imaging via the esophagus to assess cardiac structures and valvular and ventricular function. It may include 2- and 3-dimensional imaging, color and spectral Doppler imaging, and use of contrast agents for opacification.

Transposition of the great arteries (TGA) or D-loop TGA: A congenital cardiovascular malformation of ventriculoarterial discordance, in which the RV connects to the aorta and the LV connects to the pulmonary trunk (30).

Transthoracic echocardiography (TTE): Noninvasive ultrasound imaging to assess cardiac structures and

valvular and ventricular function. It may include 2-dimensional, 3-dimensional, M-mode, and strain imaging; color, spectral, and tissue Doppler imaging; and use of contrast agents for opacification.

Transthoracic echocardiography with contrast (TTE + contrast): The use of agitated saline as a contrast agent to determine the presence or flow characteristics of a shunt during transthoracic echocardiography.

Tricuspid regurgitation (TR): A congenital or acquired cardiac finding in which there is retrograde flow through the tricuspid valve (30).

Tricuspid valve dysplasia: A congenital cardiac malformation of the tricuspid valve, commonly consisting of leaflet thickening and restricted mobility with normally hinged leaflets.

Truncus arteriosus (TA): A congenital cardiovascular malformation in which a single arterial trunk arises from the heart, giving origin sequentially to the coronary arteries, 1 or more pulmonary arteries, and the systemic arterial circulation (30).

Valvular dysfunction: Valvular stenosis and/or regurgitation.

Ventricular dysfunction: Impaired systolic and/or diastolic function of the RV, LV, or both.

Ventricular septal defect (VSD): A congenital cardiac malformation in which there is a hole or pathway between the ventricular chambers (30).

- *Small VSD*: Likely to be hemodynamically insignificant.
- *Moderate or large VSD*: Likely to be hemodynamically significant.

Z-score: The number of standard deviations a value is from the mean value in a distribution. The echocardiographic z-score is the number of standard deviations a specific value is from the mean in the distribution relative to age or body surface area in the normal population (42).

5. MULTIMODALITY IMAGING IN CONGENITAL HEART DISEASE: APPROPRIATE USE CRITERIA (BY INDICATION)

The final ratings for multimodality imaging in CHD are listed by indication in **Tables 1 to 19**. The final score for each indication reflects the median score of the 17 rating panel members and has been labeled as Appropriate/A (median score 7 to 9), May Be Appropriate/M (median score 4 to 6), or Rarely Appropriate/R (median score 1 to 3). In the tables, the final score for each indication is shown in parentheses next to the AUC rating of A, M, or R. Before each table, considerations that went into construction of the indications are discussed, such as lesions and procedures that were included or excluded. Reference is made to other tables or AUC documents to avoid redundancy.

When reading through the tables and indications, the following points should be considered:

- In general, TTE is the most common imaging modality used during routine follow-up of patients with CHD. Other imaging modalities are considered for specific clinical indications or for intermittent serial follow-up unless acoustic windows for TTE are suboptimal for diagnostic purpose. If an imaging modality was deemed to be highly unlikely to be used in a given scenario, it was not rated at all and is represented by gray shading in the tables.
- The indication "evaluation due to change in clinical status and/or new concerning signs or symptoms" is meant to address a broad array of presentations and complications that could occur sooner than the timing for routine surveillance.
- The indication "routine postprocedural evaluation" refers to an evaluation performed within 30 days of the surgical or catheter-based procedure and assumes that the procedure was performed without significant complications and that the patient is following a typical postprocedural clinical course. Intraprocedural assessment using TEE to assist cardiac surgery or catheter-based procedures and immediate postoperative assessment in a critical care setting are not included under this scenario.
- Indications were grouped together if the type or frequency of imaging modalities was similar and grouped separately if there were specific clinical practice guidelines or other clinical policy recommendations available for postprocedure surveillance.
- Indications addressing ES/pulmonary arterial hypertension related to unrepaired, large systemic to PA shunts are presented in **Table 6** rather than in the tables for individual cardiac lesions.
- When the frequency of imaging and type of imaging modalities were unique for specific age ranges, separate indications were included.

Table 1 Considerations

This table addresses PFO, ASDs (including secundum and sinus venosus types), common atrium, and PAPVC. Primum ASD is considered under **Table 3** (Atrioventricular Septal Defects). Coronary sinus ASDs have not been addressed because they are rare. Surveillance of only asymptomatic patients with ASDs is considered, because it is assumed that a symptomatic patient will be referred for closure of the ASD. Scenarios in which patients with ASDs are managed medically due to other complicating factors, such as prematurity and chronic lung disease, are not considered here. Indications related to pulmonary hypertension (PH) in repaired or unrepaired patients with ASD are addressed in **Table 6** (Pulmonary

TABLE 1 PFO, ASD, and PAPVC

		AUC Score				
Patent Foramen Ovale		**TTE**	**TTE + Contrast**	**TEE**	**CMR**	**CCT**
1.	Routine surveillance of an asymptomatic patient with a PFO	R (1)	R (1)	R (1)		
Atrial Septal Defects and Partial Anomalous Pulmonary Venous Connection						
Unrepaired		**TTE**	**TTE + Contrast**	**TEE**	**CMR**	**CCT**
2.	Routine surveillance (1–2 years) in an asymptomatic patient with a small ASD or PAPVC involving a single pulmonary vein	M (4)				
3.	Routine surveillance (3–5 years) in an asymptomatic patient with a small ASD or PAPVC involving a single pulmonary vein	A (7)				
4.	Routine surveillance (1–2 years) in an asymptomatic patient with ≥ moderate ASD or PAPVC involving >1 pulmonary vein	A (8)			M (4)	M (4)
5.	Evaluation due to change in clinical status and/or new concerning signs or symptoms	A (9)	M (5)	M (5)	M (6)	R (3)
6.	Evaluation to determine the method of closure of isolated secundum ASD	A (9)	M (4)	A (7)	M (5)	R (3)
7.	Evaluation prior to planned repair of sinus venosus defect and/or PAPVC	A (9)	M (4)	A (7)	A (8)	A (7)
Postprocedural: Surgical or Catheter-Based		**TTE**	**TTE + Contrast**	**TEE**	**CMR**	**CCT**
8.	Routine postprocedural evaluation (within 30 days)	A (9)	M (5)		R (2)	R (1)
9.	Evaluation due to change in clinical status and/or new concerning signs or symptoms	A (9)	M (6)	A (8)	A (7)	A (7)
10.	Routine surveillance within 1 week following device closure of ASD in an asymptomatic patient with no or mild sequelae	A (9)	R (3)			
11.	Routine surveillance at 1 month following device closure of ASD in an asymptomatic patient with no or mild sequelae	A (9)	R (3)			
12.	Routine surveillance at 3–6 months following device closure of ASD in an asymptomatic patient with no or mild sequelae	A (9)	R (3)			
13.	Routine surveillance at 1 year following device closure of ASD in an asymptomatic patient with no or mild sequelae	A (9)	R (3)			
14.	Routine surveillance (2–5 years) after the first year following device closure of ASD in an asymptomatic patient with no or mild sequelae	A (8)	R (2)			
15.	Routine surveillance within a year following surgical ASD closure or PAPVC repair in an asymptomatic patient with no or mild sequelae	A (9)	R (2)			
16.	Routine surveillance (annually) after the first year following surgical ASD closure or PAPVC repair in an asymptomatic patient with no or mild sequelae	M (6)	R (2)		R (3)	R (2)
17.	Routine surveillance (2–5 years) after the first year following surgical ASD closure or PAPVC repair in an asymptomatic patient with no or mild sequelae	A (9)	R (2)		M (4)	M (4)
18.	Routine surveillance (3–12 months) following surgical or device closure of ASD in a patient with significant residual shunt, valvular or ventricular dysfunction, arrhythmias, and/or pulmonary hypertension	A (9)	M (4)	M (5)	M (5)	M (4)
19.	Routine surveillance (3–12 months) following repair of PAPVC in a patient with systemic or pulmonary venous obstruction, valvular or ventricular dysfunction, arrhythmias, and/or pulmonary hypertension	A (9)	M (5)	M (5)	M (5)	M (5)

A = Appropriate; ASD = atrial septal defects; AUC = Appropriate Use Criteria; CCT = cardiovascular computed tomography; CMR = cardiovascular magnetic resonance; M = May Be Appropriate; PAPVC = partial anomalous pulmonary venous connection; PFO = patent foramen ovale; R = Rarely Appropriate; TEE = transesophageal echocardiogram; TTE = transthoracic echocardiogram.

Hypertension Associated With CHD). The postprocedural section also applies to patients with Ebstein anomaly who undergo ASD device closure. In the postprocedural section, evaluation due to change in clinical status and/or new concerning signs or symptoms includes complications such as significant residual shunt, device migration, thrombosis or erosion, systemic or pulmonary venous obstruction, valvular lesions, ventricular dysfunction, arrhythmias, and PH. Scenarios related to surveillance imaging following complete repair without sequelae do not address duration of follow-up (indications 16 and 17). This does not imply indefinite follow-up in such cases, and clinicians should base follow-up decisions on available guidelines. Indications related to evaluation of patients with transient ischemic attacks or strokes with suspected atrial level shunt, and preprocedural and intraprocedural evaluation for closure of PFO or ASD have been addressed in the previous AUC document on Multimodality Imaging in Nonvalvular Heart Disease (43).

Table 1 Results and Discussion

Routine surveillance of an asymptomatic patient with a PFO using TTE or TEE was rated Rarely Appropriate.

Evaluation of a symptomatic patient with a PFO including preprocedural and intraprocedural guidance for closure of a PFO is addressed in the 2019 AUC for Multimodality Imaging in Nonvalvular Heart Disease (43). Use of TTE for routine surveillance of a small ASD and single anomalous pulmonary vein at 3 to 5 years, and for larger ASDs or more than 1 anomalous pulmonary vein at 1 to 2 years, was rated Appropriate. While TTE and TEE were rated Appropriate for evaluation prior to closure of secundum ASD, CMR and CCT were also rated Appropriate prior to planned repair of sinus venosus ASD and PAPVC because these modalities are known to provide superior imaging of pulmonary venous anatomy (8,44). Indications 10 to 13 address routine surveillance in an asymptomatic patient within 1 week, at 1 month, 3 to 6 months, and 1 year following device closure of a secundum ASD. Indication 14 addresses routine surveillance in an asymptomatic patient 2 to 5 years after the first year following device closure of a secundum ASD. Use of TTE for all of these indications was rated Appropriate, but use of TTE + contrast was rated Rarely Appropriate (1). Contrary to the rating in this document, use of TTE + contrast was rated Appropriate in the 2019 AUC for Multimodality Imaging in Nonvalvular Heart Disease for 6-month routine follow-up after ASD/PFO device closure for position and integrity of the device (43). If there are significant residual lesions or other complications following ASD closure or PAPVC repair, then routine surveillance with TTE at a higher frequency of 3 to 12 months was rated Appropriate depending on the level of clinical concern, and TTE + contrast was rated May Be Appropriate.

Table 2 Considerations

This table addresses isolated VSDs, including type 1 (subarterial/supracristal/conal), type 2 (perimembranous/conoventricular), type 3 (inlet), and type 4 (muscular) (45). Gerbode defects (LV to right atrial shunts) are not included. Distinction is made between isolated small muscular VSDs and other types of VSDs, as complications such as aortic valve prolapse, subaortic membrane, and double chambered RV may be associated with the latter. Long-term interval surveillance of symptomatic patients is not considered because it is assumed that they will undergo VSD closure. VSDs associated with other cardiac defects, such as AVSD and TOF, are included in the tables for those specific lesions (**Tables 3 and 14**, respectively).

TABLE 2 Ventricular Septal Defects

		AUC Score			
Unrepaired		**TTE**	**TEE**	**CMR**	**CCT**
20.	Routine surveillance (1–2 years) in an asymptomatic child with a small muscular VSD	R (3)			
21.	Routine surveillance (3–5 years) in an asymptomatic child with a small muscular VSD	A (7)			
22.	Routine surveillance (3–5 years) in an asymptomatic adult with a small muscular VSD	A (7)			
23.	Routine surveillance (1–2 years) in an asymptomatic child with a small VSD in a location other than muscular septum	A (7)			
24.	Routine surveillance (3–5 years) in an asymptomatic adult with a small VSD in a location other than muscular septum	A (8)	M (4)	R (3)	
25.	Routine surveillance (1–3 months) in an infant with ≥ moderate VSD on medical management	A (9)			
26.	Evaluation due to change in clinical status and/or new concerning signs or symptoms	A (9)	M (6)	M (6)	M (4)
27.	Evaluation prior to planned repair	A (9)	M (6)	M (6)	M (4)
Postprocedural: Surgical or Catheter-Based		**TTE**	**TEE**	**CMR**	**CCT**
28.	Routine postprocedural evaluation (within 30 days)	A (9)			
29.	Evaluation due to change in clinical status and/or new concerning signs or symptoms	A (9)	M (6)	M (6)	M (6)
30.	Routine surveillance within a year following surgical or device VSD closure in an asymptomatic patient with no or mild sequelae	A (8)			
31.	Routine surveillance (2–3 years) after the first year following device closure of VSD in an asymptomatic patient with no or mild sequelae	A (9)			
32.	Routine surveillance (annually) after the first year following surgical VSD closure in an asymptomatic patient with no or mild sequelae	M (5)			
33.	Routine surveillance (2–3 years) after the first year following surgical VSD closure in an asymptomatic patient with no or mild sequelae	A (8)			
34.	Routine surveillance (2–3 years) following surgical or device closure in a patient with small residual shunt, ≤ mild valvular dysfunction, no ventricular dysfunction, arrhythmias, or pulmonary hypertension	A (9)	R (3)	R (3)	R (3)
35.	Routine surveillance (3–12 months) following surgical or device closure in a patient with significant residual shunt, valvular or ventricular dysfunction, arrhythmias, and/or pulmonary hypertension	A (9)	M (5)	M (5)	M (4)

A = Appropriate; AUC = Appropriate Use Criteria; CCT = cardiovascular computed tomography; CMR = cardiovascular magnetic resonance; M = May Be Appropriate; R = Rarely Appropriate; TEE = transesophageal echocardiogram; TTE = transthoracic echocardiogram; VSD = ventricular septal defects.

Indications related to surveillance are based on a patient's symptoms and the hemodynamic significance of cardiac lesions. Unrepaired VSD with ES is addressed in Table 6 (PH Associated With CHD). In the postprocedural section, evaluation due to change in clinical status and/or new concerning signs or symptoms includes complications such as significant residual shunt, device migration, thrombosis or erosion, valvular lesions, ventricular dysfunction, development of double-chambered RV or subaortic membrane, arrhythmias, and PH. Scenarios related to surveillance imaging of small muscular VSDs (indications 20 to 22) and those following complete repair without sequelae (indications 31 to 33) do not address duration of follow-up. This does not imply indefinite follow-up in such cases, and clinicians should base this decision on available guidelines.

Table 2 Results and Discussion

Routine surveillance of small muscular VSDs with TTE was rated Rarely Appropriate at 1 to 2 years and Appropriate at a 3- to 5-year interval. Routine surveillance using TTE every 1 to 2 years in a child and 3 to 5 years in an adult with a small VSD in a location other than muscular septum was rated Appropriate owing to the possible development of prolapse of the aortic valve leaflet into the VSD, double chambered RV, and subaortic membrane. TEE at 3- to 5-year intervals was rated May Be Appropriate in adults and CMR was rated Rarely Appropriate. In symptomatic children with significant shunts who are medically managed, more frequent use of TTE at a 1- to 3-month interval was rated Appropriate. Following a surgical or catheter-based intervention, surveillance using TTE within the first year was deemed Appropriate. Following that, if there are no or mild sequelae, then surveillance every 2 to 3 years with TTE was rated Appropriate. If there are significant residual lesions, surveillance every 3 to 12 months with TTE was rated Appropriate. CCT and CMR were rated Rarely Appropriate for those with no or mild residual sequelae but May Be Appropriate for those with significant sequelae.

Table 3 Considerations

AVSD, also known as atrioventricular canal defects, are categorized as complete, transitional, and partial (46). This table addresses balanced and isolated AVSD and does not address AVSD associated with TOF, unbalanced AVSD, or an inlet VSD associated with an LV to right atrial shunt (Gerbode defect). Partial and transitional AVSD are separated from complete AVSD for indications related to surveillance due to the need for more frequent surveillance in those with complete AVSD. Unrepaired defects with ES are addressed in Table 6 (ES and PH Associated With CHD). In the unrepaired section, evaluation due to change in clinical status or new concerning signs or symptoms includes heart failure or progression of valvular regurgitation. In the postoperative section, it includes significant residual shunt, valvular or ventricular dysfunction, left ventricular outflow tract (LVOT) obstruction, arrhythmias, and PH.

TABLE 3 Atrioventricular Septal Defects

		AUC Score			
Unrepaired: Partial/Transitional		**TTE**	**TEE**	**CMR**	**CCT**
36.	Routine surveillance (3–6 months) in an asymptomatic infant	A (9)			
37.	Routine surveillance (1–2 years) in an asymptomatic child	A (9)			
Unrepaired: Complete		**TTE**	**TEE**	**CMR**	**CCT**
38.	Routine surveillance (1–3 months) in an infant	A (9)			
Unrepaired: All Types		**TTE**	**TEE**	**CMR**	**CCT**
39.	Evaluation due to change in clinical status and/or new concerning signs or symptoms	A (9)	M (6)	M (5)	M (4)
40.	Evaluation prior to planned repair	A (9)	M (5)	M (5)	M (4)
Postoperative		**TTE**	**TEE**	**CMR**	**CCT**
41.	Routine postprocedural evaluation (within 30 days)	A (9)			
42.	Evaluation due to change in clinical status and/or new concerning signs or symptoms	A (9)	M (6)	M (6)	M (5)
43.	Routine surveillance within a year after AVSD repair in an asymptomatic patient with no or mild sequelae	A (9)			
44.	Routine surveillance (1–3 years) after the first year following repair in an asymptomatic patient with no or mild sequelae	A (9)			
45.	Routine surveillance (3–12 months) in a patient with significant residual shunt, valvular or ventricular dysfunction, LVOT obstruction, arrhythmias, and/or pulmonary hypertension	A (9)	M (6)	M (6)	M (4)
46.	Routine surveillance (3–12 months) in a patient with heart failure symptoms	A (9)		M (6)	M (4)

A = Appropriate; AUC = Appropriate Use Criteria; AVSD = atrioventricular septal defects; CCT = cardiovascular computed tomography; CMR = cardiovascular magnetic resonance; LVOT = left ventricular outflow tract; M = May Be Appropriate; TEE = transesophageal echocardiogram; TTE = transthoracic echocardiogram.

Table 3 Results and Discussion

TTE was rated Appropriate for routine surveillance of unrepaired partial or transitional AVSD at a 3- to 6-month interval in an infant and a 1- to 2-year interval in a child. For complete AVSD, routine surveillance every 1–3 months in an infant was rated Appropriate. For evaluation due to change in clinical status and/or new concerning signs or symptoms in all forms of both repaired and unrepaired AVSD, TTE was rated Appropriate, while TEE, CMR, and CCT were all rated May Be Appropriate. For routine surveillance in patients with significant residual problems, use of TTE at 3 to 12 months was rated Appropriate, and TEE, CCT, and CMR were rated May Be Appropriate.

Table 4 Considerations

This table does not address infants with PDAs who are being managed in the neonatal intensive care unit or have complicating factors of prematurity and chronic lung disease. Indications related to PH in repaired or unrepaired patients with PDA are addressed in **Table 6** (PH Associated With CHD). Indication 58 is based on the ACC/AHA 2008 Guidelines for the Management of Adults with Congenital Heart Disease (1), which, because of the lack of long-term data, recommend follow-up approximately every 5 years for patients who have undergone device PDA closure. In the postprocedural section, evaluation due to change in clinical status and/or new concerning signs or symptoms includes complications such as significant residual shunt, device migration, coarctation of the aorta, aortic obstruction by a device, and left PA stenosis. This table does not have an indication for surveillance of an audible, residual PDA, because it is assumed that these patients will be referred for repeat closure.

Table 4 Results and Discussion

Routine surveillance of a patient with a trivial, silent PDA was rated Rarely Appropriate because this is an extremely low-risk lesion that does not require treatment. Use of TTE for routine surveillance of an infant or child with a small, audible PDA until closure was rated Appropriate. While TTE was rated Appropriate for surveillance every 3 to 5 years in an adult with a small PDA, CMR and CCT were rated Rarely Appropriate in this circumstance. In the first 2 years after PDA closure, either surgically or with a device, annual TTE was rated Appropriate. After 2 years following surgical closure, TTE was rated Rarely Appropriate. However, TTE every 5 years was rated Appropriate for surveillance of patients after successful device closure, even with no or mild sequelae.

TABLE 4 Patent Ductus Arteriosus

		AUC Score				
Unrepaired		**TTE**		**CMR**		**CCT**
47.	Routine surveillance (3–5 years) in an asymptomatic patient with a trivial, silent PDA	R (3)				
48.	Routine surveillance (3–6 months) in an infant with ≥ moderate PDA	A (9)		R (2)		R (2)
49.	Routine surveillance (3–6 months) in an infant with a small, audible PDA until closure	A (7)				
50.	Routine surveillance (1–2 years) in an infant or child with a small, audible PDA until closure	A (8)				
51.	Routine surveillance (3–5 years) in an adult with a small PDA	A (9)		R (3)		R (2)
52.	Evaluation due to change in clinical status and/or new concerning signs or symptoms	A (9)		M (6)		M (5)
53.	Evaluation prior to planned repair	A (9)		M (5)		M (5)
Postprocedural: Surgical or Catheter-Based		**TTE**	**TEE**	**CMR**	**CCT**	**Lung Scan**
54.	Routine postprocedural evaluation (within 30 days)	A (9)				
55.	Evaluation due to change in clinical status and/or new concerning signs or symptoms	A (9)	M (4)	M (5)	M (5)	M (4)
56.	Routine surveillance (annually) within 2 years following PDA closure in an asymptomatic patient with no or mild sequelae	A (8)				
57.	Routine surveillance (5 years) after the first 2 years following surgical closure in an asymptomatic patient with no or mild sequelae	R (3)				
58.	Routine surveillance (5 years) after the first 2 years following device closure in an asymptomatic patient with no or mild sequelae	A (7)				
59.	Routine surveillance (1–2 years) in a patient with postprocedural left pulmonary artery stenosis	A (9)		M (6)	M (6)	A (7)
60.	Routine surveillance (1–2 years) in a patient with postprocedural aortic obstruction	A (9)		A (7)	A (7)	

A = Appropriate; AUC = Appropriate Use Criteria; CCT = cardiovascular computed tomography; CMR = cardiovascular magnetic resonance; M = May Be Appropriate; PDA = patent ductus arteriosus; R = Rarely Appropriate; TEE = transesophageal echocardiogram; TTE = transthoracic echocardiogram.

TABLE 5 Total Anomalous Pulmonary Venous Connection

	AUC Score					
Unrepaired	**TTE**		**TEE**		**CMR**	**CCT**
61. Evaluation due to change in clinical status and/or new concerning signs or symptoms	A (9)		M (6)		A (7)	A (7)
62. Evaluation prior to planned repair	A (9)		M (6)		A (7)	A (7)
Postoperative	**TTE**	**TEE**	**CMR**	**CCT**	**Stress Imaging**	**Lung Scan**
63. Routine postprocedural evaluation (within 30 days)	A (9)		R (3)	R (3)		
64. Evaluation due to change in clinical status and/or new concerning signs or symptoms	A (9)	A (7)	A (7)	A (7)	R (3)	A (7)
65. Routine surveillance (3–6 months) in an asymptomatic infant with no or mild sequelae	A (8)					
66. Routine surveillance (1–2 years) in an asymptomatic child with no or mild sequelae	A (8)					
67. Routine surveillance (3–5 years) in an asymptomatic adult with no or mild sequelae			M (6)	M (5)		

A = Appropriate; AUC = Appropriate Use Criteria; CCT = cardiovascular computed tomography; CMR = cardiovascular magnetic resonance; M = May Be Appropriate; R = Rarely Appropriate; TAPVC = total anomalous pulmonary venous connection; TEE = transesophageal echocardiogram; TTE = transthoracic echocardiogram.

Table 5 Considerations

This table includes clinical scenarios related to total anomalous pulmonary venous connection both before and after operative repair. It is not applicable when a patient has other coexisting major CHD, such as conotruncal defects or heterotaxy syndrome. Postoperative evaluation scenarios may include concerns for pulmonary vein stenosis or ventricular dysfunction. In the postoperative scenarios for asymptomatic patients, age is considered because this impacts the frequency of imaging surveillance and modalities under consideration.

Table 5 Results and Discussion

In the unrepaired patient with TAPVC, TTE, CMR, and CCT were rated Appropriate for evaluation prior to planned repair, and Appropriate for evaluation due to a change in clinical status or new concerning signs and/or symptoms. In the early postoperative period, TTE was rated Appropriate for routine postprocedural evaluation, but CMR and CCT were rated Rarely Appropriate. In the repaired patient with TAPVC with new clinical signs and/or symptoms, TTE, TEE, CMR, CCT, and lung scan were rated Appropriate. Indications 65 to 67 address routine surveillance following repair of TAPVC at 3 to 6 months in an asymptomatic infant, 1 to 2 years in an asymptomatic child, and 3 to 5 years in an adult with no or mild sequalae, respectively. Use of TTE was rated Appropriate for indications 65 and 66 and CMR and CCT were rated May Be Appropriate for indication 67 (47). In an adult with no or mild sequalae, CMR and CCT were rated May Be Appropriate.

Table 6 Considerations

This table includes the following clinical scenarios related to patients with suspicion of PH in the setting of CHD: ES/pulmonary arterial hypertension related to unrepaired, large systemic to PA shunts (e.g., ASD, VSD, AVSD, PDA, aortopulmonary window, truncus, transposition of the great arteries with a nonrestrictive VSD); late postoperative pulmonary arterial hypertension in the setting of repaired CHD; and postcapillary PH associated with CHD (especially after repair of VSD or congenital MS). This table does not include idiopathic PH, pulmonary vein stenosis, pulmonary veno-occlusive disease, or immediate post-operative PH in the setting of repaired CHD. Although noninvasive assessment may provide useful morphological information on the central and segmental pulmonary vessels in PH, cardiac catheterization remains the mainstay of PH assessment in the initiation and follow-up of pharmacological therapy. This table also does not include nuclear medicine ventilation/perfusion scans, which may be appropriate for patients with suspected chronic pulmonary thromboembolism.

Table 6 Results and Discussion

TTE was rated Appropriate for the evaluation of a patient with ES. CMR was rated Appropriate for initial evaluation of ES or due to a change in clinical status, and May Be Appropriate for an evaluation due to a change in PH therapy. CCT and stress imaging were rated May Be Appropriate in the evaluation of a patient with ES. In the routine surveillance of patients with ES, TTE every 6 to 12 months was rated Appropriate in a stable child and adult. This is supported by the 2015 AHA/ATS Pediatric Pulmonary Hypertension Guidelines and the 2015 ESC/ERS Guidelines for the Diagnosis and Treatment of Pulmonary Hypertension (33,48). TTE was rated Rarely Appropriate every 3 months in a stable adult with ES. CMR and CCT were rated Rarely Appropriate in the routine surveillance of a stable child or adult with ES except in indication 74, in which CMR was rated May Be Appropriate every 6 to 12 months in a stable adult with ES.

TTE and CMR were rated Appropriate in the evaluation of a patient with PH associated with CHD. CCT and stress

TABLE 6 ES and Pulmonary Hypertension Associated With CHD

		AUC Score			
Eisenmenger Syndrome (ES)		**TTE**	**CMR**	**CCT**	**Stress Imaging**
68.	Initial evaluation with suspicion of ES	A (9)	A (7)	M (6)	M (4)
69.	Evaluation due to change in clinical status and/or new concerning signs or symptoms in a patient with ES	A (9)	A (7)	M (6)	M (5)
70.	Evaluation due to change in pulmonary arterial hypertension-targeted therapy in a patient with ES	A (9)	A (7)	M (5)	M (5)
71.	Routine surveillance (3 months) in a stable child with ES	M (6)	R (2)	R (2)	
72.	Routine surveillance (6–12 months) in a stable child with ES	A (9)	R (3)	R (3)	
73.	Routine surveillance (3 months) in a stable adult with ES	R (3)	R (3)	R (2)	
74.	Routine surveillance (6–12 months) in a stable adult with ES	A (9)	M (4)	R (3)	
Pulmonary Hypertension (PH) Associated With CHD		**TTE**	**CMR**	**CCT**	**Stress Imaging**
75.	Initial evaluation with suspicion of pulmonary hypertension following CHD surgery	A (9)	A (7)	M (6)	M (5)
76.	Evaluation due to change in clinical status and/or new concerning signs or symptoms in a patient with postoperative PH	A (9)	A (7)	M (6)	M (5)
77.	Evaluation due to change in pulmonary arterial hypertension-targeted therapy in a patient with postoperative PH	A (9)	A (7)	M (6)	M (5)
78.	Routine surveillance (3 months) in a stable child with postoperative PH	A (7)	R (3)	R (2)	
79.	Routine surveillance (6–12 months) in a stable child with post-operative PH	A (9)	R (3)	R (3)	
80.	Routine surveillance (3 months) in a stable adult with postoperative PH	M (5)	R (3)	R (2)	
81.	Routine surveillance (6–12 months) in a stable adult postoperative PH	A (9)	M (5)	R (3)	

A = Appropriate; AUC = Appropriate Use Criteria; CCT = cardiovascular computed tomography; CHD = congenital heart disease; CMR = cardiovascular magnetic resonance; ES = Eisenmenger syndrome; M = May Be Appropriate; PH = pulmonary hypertension; R = Rarely Appropriate; TTE = transthoracic echocardiogram.

imaging were rated May Be Appropriate in similar indications for the evaluation of a patient with PH associated with CHD. In the routine surveillance of patients with PH associated with CHD, TTE every 6 to 12 months was rated Appropriate in a stable child and adult. This is supported as well by the 2015 AHA/ATS Pediatric Pulmonary Hypertension Guidelines and the 2015 ESC/ERS Guidelines for the Diagnosis and Treatment of Pulmonary Hypertension (33,48). TTE was also rated Appropriate every 3 months in a stable child with post-operative PH and May Be Appropriate every 3 months in a stable adult. CMR and CCT every 3 months were rated Rarely Appropriate in a stable child or adult with post-operative PH, whereas CMR every 6 to 12 months was rated May Be Appropriate in a stable adult with post-operative PH.

Table 7 Considerations

This table addresses key components of cardiac imaging in patients with unrepaired Ebstein anomaly or tricuspid valve dysplasia as well as those who have undergone surgical repair. While ASDs are commonly encountered with Ebstein anomaly, only indication 87 focuses on noninvasive evaluation of the ASD before device closure. Otherwise, the postprocedural evaluation of patients with ASD is provided in Table 1 (PFO, ASD, and PAPVC). Evaluation due to change in clinical status or symptoms such as heart failure, cyanosis, or atrial arrhythmias, may prompt an evaluation that investigates changes in biventricular systolic function, residual intracardiac shunt, or valvular function. Indication 95 is dedicated to the surveillance of heart failure and atrial arrhythmias, which may include assessment of biventricular volumes and function to guide medical therapy. The post-procedural section includes tricuspid valve repair or replacement and ASD device closure, although indications specific to ASD device closure are covered in Table 1.

Table 7 Results and Discussion

TTE was rated Appropriate in the routine surveillance of an asymptomatic infant, child, or adult with Ebstein anomaly and mild TR. In indications 83 and 85, TTE + contrast was rated May Be Appropriate. For routine surveillance (3 to 5 years) in the asymptomatic adult with mild TR, CMR was rated May Be Appropriate and CCT was rated Rarely Appropriate. CMR may provide complimentary information regarding tricuspid valve anatomy and biventricular systolic function. For the evaluation due to change in clinical status, TTE, TTE + contrast, TEE, and CMR were rated Appropriate, and CCT and stress imaging were rated May Be Appropriate. In the evaluation for transcatheter ASD closure, TTE and TEE were rated Appropriate, and TTE + contrast, CMR, and CCT were rated May Be Appropriate. In the evaluation prior to surgical repair, TTE and CMR were rated Appropriate; TTE +

TABLE 7 Ebstein Anomaly and Tricuspid Valve Dysplasia

	AUC Score					
Unrepaired	**TTE**	**TTE + Contrast**	**TEE**	**CMR**	**CCT**	**Stress Imaging**
82. Routine surveillance (1–2 years) in an asymptomatic infant or child with mild TR	A (9)			R (3)	R (3)	
83. Routine surveillance (3–5 years) in an asymptomatic adult with mild TR	A (9)	M (5)	M (4)	M (5)	R (3)	
84. Routine surveillance (3–6 months) in an asymptomatic infant with ≥ moderate TR without hypoxemia	A (9)					
85. Routine surveillance (6–12 months) in an asymptomatic patient with ≥moderate TR and previously stable RV size and/or function without hypoxemia	A (9)	M (4)	M (4)	M (5)	R (3)	
86. Evaluation due to change in clinical status and/or new concerning signs and symptoms	A (9)	A (7)	A (7)	A (7)	M (6)	M (5)
87. Evaluation of an ASD for device closure in a patient with mild or moderate TR, RV enlargement, and no hypoxemia	A (9)	M (6)	A (8)	M (6)	M (5)	
88. Evaluation prior to planned repair	A (9)	M (6)	M (6)	A (7)	M (4)	R (3)
Postprocedural: Surgical or Catheter-Based	**TTE**	**TTE + Contrast**	**TEE**	**CMR**	**CCT**	**Stress Imaging**
89. Routine postprocedural evaluation (within 30 days)	A (9)	M (6)				
90. Evaluation due to change in clinical status and/or new concerning signs or symptoms	A (9)	A (7)	A (7)	A (7)	M (6)	M (5)
91. Routine surveillance (1–2 years) in an asymptomatic patient with no or mild sequelae	A (9)		R (3)	R (3)	R (2)	
92. Routine surveillance (3–5 years) in an asymptomatic patient with no or mild sequelae			M (4)	M (5)	M (4)	
93. Routine surveillance (6–12 months) in an asymptomatic child with valvular or ventricular dysfunction or arrhythmias	A (9)			M (4)	R (3)	
94. Routine surveillance (1–2 years) in an asymptomatic adult with valvular or ventricular dysfunction or arrhythmias	A (9)		M (5)	M (5)	R (3)	
95. Routine surveillance (3–12 months) in a patient with symptoms of heart failure and/or atrial arrhythmias	A (9)			M (5)	R (3)	

A = Appropriate; AUC = Appropriate Use Criteria; ASD = atrial septal defect; CCT = cardiovascular computed tomography; CMR = cardiovascular magnetic resonance; M = May Be Appropriate; R = Rarely Appropriate; RV = right ventricle; TEE = transesophageal echocardiogram; TR = tricuspid regurgitation; TTE = transthoracic echocardiogram.

contrast, TEE, and CCT were rated May Be Appropriate; and stress imaging was rated Rarely Appropriate. TTE in the routine postoperative assessment was rated Appropriate and TTE + contrast was rated May Be Appropriate. In the final indication of this table, routine surveillance (3 to 12 months) in a patient with symptoms of heart failure or atrial arrhythmias, TTE was rated Appropriate, CMR May Be Appropriate, and CCT Rarely Appropriate.

Table 8 Considerations

This table includes clinical scenarios related to pulmonary valve stenosis, both before and after surgical or catheter-based procedures. Evaluation may be performed due to concerns for PA dilation, ventricular hypertrophy, dilation, and dysfunction. One and a half ventricle repair (superior cavopulmonary anastomosis with RV-to-PA connection) has not been specifically addressed in this document. The procedures (balloon valvuloplasty and operative pulmonary valve replacement) are grouped together because their subsequent surveillance issues are similar. Table 14 (Tetralogy of Fallot) addresses surveillance and evaluation of transcatheter pulmonary valve replacement (PVR) patients.

Table 8 Results and Discussion

In the asymptomatic patient with unrepaired PS, TTE was rated Appropriate for routine surveillance at the frequencies provided in indications 96 to 100. The frequency of imaging with TTE is driven by the age of the patient, with more frequent imaging in the younger age groups. CMR and CCT were rated Rarely Appropriate for the routine surveillance of patients with mild PS and infants with any degree of PS; however, they were rated May Be Appropriate for a child or adult with moderate or severe PS, and CMR was rated Appropriate for those with PA dilation. Prior to planned repair and for evaluation due to a change in clinical status or new concerning signs and/or symptoms, TTE was rated Appropriate, and TEE, CMR, and CCT were rated May Be Appropriate. For postprocedural evaluation due to change in clinical status and/or new concerning signs or symptoms, TTE was rated Appropriate, and TEE, CMR, CCT, and stress imaging were rated May Be Appropriate. For postprocedural routine surveillance in patients of any age with or without symptoms or sequelae, TTE was rated Appropriate. Postprocedural routine surveillance with CMR was rated Appropriate or May Be Appropriate at the specified

TABLE 8 Pulmonary Stenosis

		AUC Score				
Unrepaired		**TTE**	**TEE**	**CMR**	**CCT**	**Stress Imaging**
96.	Routine surveillance (3–6 months) in an asymptomatic infant with mild PS	A (8)				
97.	Routine surveillance (1–2 years) in an asymptomatic child with mild PS	A (8)				
98.	Routine surveillance (3–5 years) in an asymptomatic adult with mild PS	A (9)		R (3)	R (3)	
99.	Routine surveillance (3–6 months) in an asymptomatic infant with ≥ moderate PS	A (9)		R (3)	R (3)	
100.	Routine surveillance (1–2 years) in an asymptomatic child or adult with ≥moderate PS	A (9)		M (5)	M (4)	
101.	Routine surveillance (3–5 years) in an asymptomatic adult with PS and pulmonary artery dilation			A (7)	M (6)	
102.	Evaluation due to change in clinical status and/or new concerning signs or symptoms	A (9)		M (6)	M (6)	M (5)
103.	Evaluation prior to planned repair	A (9)	M (6)	M (6)	M (6)	
Postprocedural: Surgical or Catheter-Based		**TTE**	**TEE**	**CMR**	**CCT**	**Stress Imaging**
104.	Routine postprocedural evaluation (within 30 days)	A (9)				
105.	Evaluation due to change in clinical status and/or new concerning signs or symptoms	A (9)	M (5)	M (6)	M (5)	M (4)
106.	Routine surveillance (1–2 years) in an asymptomatic child with no or mild sequelae	A (9)		R (3)	R (3)	
107.	Routine surveillance (3–5 years) in an asymptomatic adult with no or mild sequelae	A (9)		M (4)	R (3)	
108.	Routine surveillance (6–12 months) in an asymptomatic child with moderate or severe sequelae	A (9)		M (5)	M (4)	
109.	Routine surveillance (1–3 years) in an asymptomatic adult with moderate or severe sequelae	A (9)		A (7)	M (6)	M (5)
110.	Routine surveillance (3–12 months) in a patient with heart failure symptoms	A (9)		M (5)	M (4)	

A = Appropriate; AUC = Appropriate Use Criteria; CCT = cardiovascular computed tomography; CMR = cardiovascular magnetic resonance; M = May Be Appropriate; PS = pulmonary stenosis; R = Rarely Appropriate; TEE = transesophageal echocardiogram; TTE = transthoracic echocardiogram.

timeframe in an asymptomatic child with moderate or severe sequelae, an asymptomatic adult regardless of sequelae, and in patients of all ages with heart failure symptoms. Postprocedural routine surveillance with CCT was rated May Be Appropriate at the specified timeframe in a child or adult with moderate or severe sequelae, and in the setting of heart failure symptoms.

Table 9 Considerations

This table includes clinical scenarios related to pulmonary atresia with intact ventricular septum (PA/IVS) in anticipation of an eventual biventricular repair or patients after a biventricular repair (transcatheter pulmonary valve perforation and dilation or surgical RV outflow tract reconstruction). Single-ventricle palliation for PA/IVS is addressed in Table 19 (Single-Ventricle Heart Disease). One and one-half ventricle repair (superior cavopulmonary anastomosis with RV-to-PA connection) has not been specifically addressed in this document. Refer to Table 14 (Tetralogy of Fallot) for surveillance and evaluation after pulmonary valve replacement. Patients with PA/IVS who have undergone biventricular repair can have tricuspid and pulmonary valve dysfunction and RV diastolic dysfunction; these are grouped together in the indications.

Table 9 Results and Discussion

In the unrepaired patient, in patients following palliative procedures, or for evaluation prior to planned repair, TTE was rated Appropriate, and TEE, CMR, CCT, and lung scan were rated May Be Appropriate. In patients who have undergone palliative surgery, TTE was rated Appropriate for early postprocedural evaluation, routine surveillance in asymptomatic patients, and evaluation of a change in clinical status or new concerning signs or symptoms. TEE, CMR, CCT, stress imaging, and lung scan were rated May Be Appropriate for the evaluation of change in clinical status or new concerning signs and symptoms. In patients who have undergone complete repair for PA/IVS, TTE was rated Appropriate for all indications; the frequency of imaging is determined by the severity of the sequelae and the age of the patient. CMR was rated Appropriate or May Be Appropriate for all indications other than routine postprocedural evaluation and routine surveillance in an asymptomatic infant. CCT was generally rated slightly lower than CMR for the same indications. Lung scan was rated May Be Appropriate for routine postprocedural evaluation, evaluation due to change in clinical status and/or new concerning signs or symptoms, and routine surveillance in an asymptomatic child and adult with moderate or severe sequelae.

TABLE 9 Pulmonary Atresia With Intact Ventricular Septum

	AUC Score					
Unrepaired	**TTE**	**TEE**	**CMR**	**CCT**	**Stress Imaging**	**Lung Scan**
111. Evaluation prior to planned repair	A (9)	M (6)	M (6)	M (6)		M (5)
Postprocedural: Palliation	**TTE**	**TEE**	**CMR**	**CCT**	**Stress Imaging**	**Lung Scan**
112. Routine postprocedural evaluation (within 30 days)	A (9)					M (4)
113. Routine surveillance (1–3 months) in an asymptomatic patient	A (9)					
114. Evaluation due to change in clinical status and/or new concerning signs or symptoms	A (9)	M (6)	M (6)	M (6)	M (5)	M (5)
115. Evaluation prior to planned repair	A (9)	M (6)	M (6)	M (6)		M (4)
Postprocedural: Complete Repair	**TTE**	**TEE**	**CMR**	**CCT**	**Stress Imaging**	**Lung Scan**
116. Routine postprocedural evaluation (within 30 days)	A (9)					M (5)
117. Evaluation due to a change in clinical status and/or new concerning signs or symptoms	A (9)	M (5)	M (6)	M (6)	M (4)	M (5)
118. Routine surveillance (3–6 months) in an asymptomatic infant	A (9)		R (3)	R (3)		R (3)
119. Routine surveillance (1–2 years) in an asymptomatic child with no or mild sequelae	A (9)		M (4)	R (3)		R (3)
120. Routine surveillance (2–3 years) in an asymptomatic adult with no or mild sequelae	A (9)		M (5)	M (4)		R (3)
121. Routine surveillance (6–12 months) in an asymptomatic child with ≥moderate sequelae	A (9)		M (5)	M (4)		M (4)
122. Routine surveillance (1–3 years) in an asymptomatic adult with ≥moderate sequelae	A (9)	M (6)	A (7)	M (5)		M (4)
123. Routine surveillance (3–12 months) in a patient with heart failure symptoms	A (9)		M (5)	M (4)		

A = Appropriate; AUC = Appropriate Use Criteria; CCT = cardiovascular computed tomography; CMR = cardiovascular magnetic resonance; M = May Be Appropriate; PA/IVS = pulmonary atresia with intact ventricular septum; R = Rarely Appropriate; TEE = transesophageal echocardiogram; TTE = transthoracic echocardiogram.

Table 10 Considerations

This table addresses clinical scenarios related to congenital MS, MR, and MVP in infants and children. The 2017 AUC for Multimodality Imaging in Valvular Heart Disease addresses mitral valve disease in adult patients (14). This table does not address surveillance and evaluation of bioprosthetic and mechanical mitral valves in adults, as this is also covered in the aforementioned 2017 AUC document (14). Rare lesions, such as isolated mitral atresia, have not been addressed, and AV septal defects are addressed in Table 3 (Atrioventricular Septal Defects). Patient age and severity of the lesion have been given due consideration in most scenarios, as these are important factors impacting the frequency of surveillance. Scenarios related to surveillance of severe, symptomatic lesions are not considered, as it is assumed that these patients will undergo surgical or catheter-based intervention. The postprocedural section includes surgical repair, surgical valve replacement (bioprosthetic or mechanical), and balloon mitral valvuloplasty. Transcatheter mitral valve replacement and repair are not covered in this AUC document. Separate scenarios were not created for postprocedural valvular stenosis, regurgitation, or mixed lesions, as the imaging needs are similar, and it would have resulted in an exceedingly long list of possible scenarios. In the postprocedural section, evaluation due to change in clinical status and/or new concerning signs or symptoms includes complications such as suspected PH and concern for repaired or replaced mitral valve dysfunction.

Table 10 Results and Discussion

In the unrepaired infant and child with congenital MS, TTE was rated Appropriate for surveillance, with the interval based on the age of the patient and severity of the MS. Stress imaging was rated May Be Appropriate in children with moderate MS, in infants and children with a change in clinical status and/or new concerning signs or symptoms, and in infants and children being evaluated for repair. CMR and CCT were rated May Be Appropriate in those with a change in clinical status and/or new concerning signs or symptoms and in those being evaluated for repair, while TEE was rated Appropriate in those circumstances.

In the unrepaired infant and child with congenital MR, including those with MVP, TTE was rated Appropriate for surveillance, with the interval based on the age of the patient and the severity of the MR. In the asymptomatic child with MVP and only mild MR, TTE was rated Appropriate for surveillance (3 to 5 years) but May Be Appropriate every 1 to 2 years. In addition to TTE, CMR was rated May Be Appropriate for routine surveillance every 6 to 12 months in a child with ≥ moderate MR. For evaluation due to change in clinical status and/or new concerning signs or symptoms, TEE, CMR, CCT, and stress

TABLE 10 Mitral Valve Disease

	AUC Score				
Unrepaired: Congenital Mitral Stenosis (MS)	**TTE**	**TEE**	**CMR**	**CCT**	**Stress Imaging**
124. Routine surveillance (1–4 weeks) in an infant <3 months with any degree of MS	A (8)				
125. Routine surveillance (3–6 months) in an infant ≥3 months with mild MS	A (8)				
126. Routine surveillance (1–3 months) in an infant ≥3 months with ≥moderate MS	A (9)				
127. Routine surveillance (1–2 years) in an asymptomatic child with mild MS	A (9)				
128. Routine surveillance (3–12 months) in an asymptomatic child with ≥moderate MS	A (9)				M (4)
129. Evaluation due to change in clinical status and/or new concerning signs or symptoms	A (9)	A (7)	M (5)	M (4)	M (5)
130. Evaluation prior to planned repair	A (9)	A (7)	M (5)	M (5)	M (4)
Unrepaired: Congenital Mitral Regurgitation (MR) including Mitral Valve Prolapse (MVP)	**TTE**	**TEE**	**CMR**	**CCT**	**Stress Imaging**
131. Routine surveillance (6–12 months) in an asymptomatic infant with mild MR	A (9)				
132. Routine surveillance (1–3 months) in an asymptomatic infant with ≥moderate MR	A (9)				
133. Routine surveillance (2–5 years) in a child with mild MR, normal LV size and systolic function	A (9)		R (3)	R (3)	
134. Routine surveillance (6–12 months) in a child with ≥moderate MR	A (9)		M (4)	R (3)	
135. Routine surveillance (1–2 years) in an asymptomatic child with MVP and mild MR	M (5)				
136. Routine surveillance (3–5 years) in an asymptomatic child with MVP and mild MR	A (9)				
137. Evaluation due to change in clinical status and/or new concerning signs or symptoms	A (9)	M (6)	M (6)	M (5)	M (5)
138. Evaluation prior to planned repair	A (9)	A (7)	M (5)	M (5)	R (3)

Postprocedural: Surgical or Catheter-Based	**TTE**	**TEE**	**CMR**	**CCT**	**Stress Imaging**	**Fluoro**
139. Routine postprocedural evaluation (within 30 days)	A (9)					
140. Evaluation in an infant or child due to change in clinical status and/or new concerning signs or symptoms	A (9)	A (7)	M (5)	M (5)	M (4)	A (7)
141. Routine surveillance (3–6 months) in an infant with mild MS or MR, and no LV dysfunction	A (9)					
142. Routine surveillance (1–3 months) in an infant with ≥moderate MS or MR, dilated LV, and no LV dysfunction	A (9)	R (3)	R (3)	R (3)		
143. Routine surveillance (1–2 years) in a child with mild MS or MR, and no LV dysfunction	A (9)					
144. Routine surveillance (3–12 months) in a child with ≥moderate MS or MR, dilated LV, and no LV dysfunction	A (9)	R (3)	R (3)	R (3)	R (3)	
145. Routine surveillance (annually) in a child with normal prosthetic mitral valve function and no LV dysfunction	A (9)					
146. Routine surveillance (3–12 months) in a child with prosthetic mitral valve or ventricular dysfunction, and/or arrhythmias	A (9)	M (5)	M (5)	M (4)		

A = Appropriate; AUC = Appropriate Use Criteria; CCT = cardiovascular computed tomography; CMR = cardiovascular magnetic resonance; Fluoro = fluoroscopy; LV = left ventricle; M = May Be Appropriate; MR = mitral regurgitation; MS = mitral stenosis; MVP = mitral valve prolapse; R = Rarely Appropriate; TEE = transesophageal echocardiogram; TTE = transthoracic echocardiogram.

imaging were rated May Be Appropriate. In the evaluation prior to planned repair, however, TEE was rated Appropriate and stress imaging was Rarely Appropriate, with CMR and CCT rated May Be Appropriate.

TTE was rated Appropriate for evaluation and surveillance after surgical or catheter-based repair, with the frequency based on the severity of the sequelae and age of the patient. TEE, CMR, and CCT were rated Rarely Appropriate, unless there was a change in clinical status, new concerning signs or symptoms, prosthetic mitral valve dysfunction, ventricular dysfunction, and/or arrhythmias in a child. Fluoro was rated Appropriate for evaluation due to change in clinical status and/or new concerning signs or symptoms in a patient with a prosthetic mitral valve.

Table 11 Considerations

This table includes clinical scenarios related to subvalvular, valvular, and supravalvular aortic disease. The scenarios related to subvalvular and supravalvular lesions include pediatric and adult patients, whereas those for valvular lesions are only for pediatric patients because the 2017 AUC for Multimodality Imaging in Valvular Heart Disease has addressed valvular lesions in adults (14). Separate scenarios were created for supravalvular AS because assessment of coronary arteries in these patients

may impact appropriateness. Surveillance of severe lesions in pediatric patients was not considered because it was assumed that these patients will undergo surgical or catheter-based intervention. For similar reasons, scenarios related to > mild AR with subvalvular lesions are not considered. Isolated valvular stenosis, regurgitation, and mixed lesions were grouped together if the imaging needs were similar and to avoid creating an exhaustive list of scenarios if each one was further stratified on the basis of the age and severity of the lesion. Rare lesions, including aortic valve atresia, LV to aortic tunnel, and sinus of Valsalva aneurysm, were not addressed. In addition, syndromic aortic dilation, such as in Marfan syndrome, other connective tissue disorders, and Turner syndrome, have been excluded, but aortic dilation related to bicuspid aortic valve in pediatric patients has been included. Aortic dilation in adults has been addressed in the 2017 AUC for Multimodality Imaging in Valvular Heart Disease (14).

The postprocedural section for surgical or catheter-based interventions includes surgical repair or replacement, the Ross procedure (excluding conduit-related indications that are addressed in **Table 14**, Tetralogy of Fallot), and balloon aortic valvuloplasty. Transcatheter aortic valve replacement has not been included because the current experience in pediatric patients is limited and this procedure has been addressed for adult patients in the 2017 AUC for Multimodality Imaging in Valvular Heart Disease (14). Indications 166 to 168 are specific to aortic sinus/ascending aortic dilation in pediatric patients and apply to the postprocedural section as well. In unrepaired and repaired patients, depending on the type of lesion and intervention, the indication for evaluation due to change in clinical status and/or new concerning signs or symptoms may include heart failure symptoms, concerns for progressive LV dilation or dysfunction, worsening stenosis or regurgitation (including prosthetic valve dysfunction), and coronary insufficiency or aneurysm.

TABLE 11 LVOT Lesions

		AUC Score				
Unrepaired: Subvalvular Aortic Stenosis (AS)		**TTE**	**TEE**	**CMR**	**CCT**	**Stress Imaging**
147.	Routine surveillance (1–3 months) in an infant with any degree of subvalvular AS and ≤mild AR	A (9)				
148.	Routine surveillance (1–2 years) in a child or adult with mild subvalvular AS and no AR	A (9)				
149.	Routine surveillance (6–12 months) in a child or adult with ≥ moderate subvalvular AS and/or ≤mild AR	A (9)	M (4)	M (4)	R (3)	R (3)
150.	Routine surveillance (3–5 years) in an asymptomatic adult with ≥ moderate subvalvular AS				M (4)	M (5)
151.	Evaluation due to change in clinical status and/or new concerning signs or symptoms	A (9)	M (6)	A (7)	M (6)	M (6)
152.	Evaluation prior to planned repair	A (9)	A (7)	A (7)	M (6)	M (4)
Postoperative		**TTE**	**TEE**	**CMR**	**CCT**	**Stress Imaging**
153.	Routine postoperative evaluation (within 30 days)	A (9)				
154.	Evaluation due to change in clinical status and/or new concerning signs or symptoms	A (9)	A (7)	M (6)	M (6)	M (5)
155.	Routine surveillance (3–6 months) in an infant with ≤ mild stenosis and/or AR	A (9)				
156.	Routine surveillance (1–3 months) in an infant with ≥ moderate stenosis and/or AR	A (9)				
157.	Routine surveillance (1–2 years) in a child or adult with ≤ mild stenosis and/or AR	A (9)	R (3)	R (3)	R (3)	R (3)
158.	Routine surveillance (6–12 months) in a child or adult with ≥ moderate stenosis and/or AR	A (9)	M (5)	M (5)	M (4)	M (4)
159.	Routine surveillance (3–12 months) in an adult with heart failure symptoms or ≥ moderate stenosis and/or AR	A (9)	M (6)	M (6)	M (5)	M (5)
Unrepaired: Aortic Valve Stenosis and/or Regurgitation*		**TTE**	**TEE**	**CMR**	**CCT**	**Stress Imaging**
160.	Routine surveillance (1–4 weeks) in an infant (<3 months old) with any degree of AS and/or AR not requiring neonatal surgery	A (9)				
161.	Routine surveillance (3–6 months) in an infant (3–12 months old) with mild AS and/or mild AR	A (9)				
162.	Routine surveillance (1–3 months) in an infant (3-12 months old) with ≥moderate AS and/or ≥moderate AR	A (9)				
163.	Routine surveillance (6 months) in an asymptomatic child with mild AS and/or mild AR without aortic dilation	R (3)				
164.	Routine surveillance (1–2 years) in an asymptomatic child with mild AS and/or mild AR without aortic dilation	A (9)		R (3)	R (3)	

Continued on the next page

TABLE 11 Continued

Unrepaired: Aortic Valve Stenosis and/or Regurgitation*	TTE	TEE	CMR	CCT	Stress Imaging
165. Routine surveillance (6–12 months) in an asymptomatic child with ≥ moderate AS and/or ≥ moderate AR	A (9)		M (5)	M (4)	M (4)
166. Routine surveillance (3–5 years) in a child with a bicuspid aortic valve with trivial or mild valvular dysfunction with no aortic sinus and/or ascending aortic dilation	A (9)		M (5)	M (4)	
167. Routine surveillance (2–3 years) in a child with aortic sinus and/or ascending aortic dilation with stable z-scores	A (9)		A (7)	M (6)	
168. Routine surveillance (6–12 months) in a child with aortic sinus and/or ascending aortic dilation with increasing z-scores	A (9)		A (8)	A (7)	
169. Evaluation due to change in clinical status and/or new concerning signs or symptoms	A (9)	M (6)	A (7)	A (7)	M (6)
170. Evaluation prior to planned repair	A (9)	A (7)	A (7)	A (7)	R (3)

*This part of the table does not include indications for adults.

Postprocedural: Surgical or Catheter-Based*	TTE	TEE	CMR	CCT	Stress Imaging	Fluoro
171. Routine postprocedural evaluation (within 30 days)	A (9)					
172. Evaluation due to change in clinical status and/or new concerning signs or symptoms	A (9)	A (7)	A (7)	A (7)	M (6)	A (7)
173. Routine surveillance (3–6 months) in an infant following neonatal intervention with ≤mild AS and/or AR and no LV dysfunction	A (9)					
174. Routine surveillance (1–3 months) in an infant following neonatal intervention with ≥moderate AS and/or regurgitation, and/or LV dysfunction	A (9)					
175. Routine surveillance (1–2 years) in a child with ≤mild AS and/or AR following repair or normal prosthetic valve function	A (9)					
176. Routine surveillance (6–12 months) in a child with ≥moderate AS or AR	A (9)	M (5)	M (5)	M (5)	M (4)	
177. Routine surveillance (3–12 months) in a child with heart failure symptoms and/or ventricular dysfunction	A (9)		M (6)	M (5)	M (5)	

*This part of the table does not include indications for adults.

Unrepaired: Supravalvular Aortic Stenosis (AS)	TTE	TEE	CMR	CCT	Stress Imaging
178. Routine surveillance (3–6 months) in an infant with any degree of supravalvular AS	A (9)				
179. Routine surveillance (1–2 years) in an asymptomatic child or adult with mild supravalvular AS	A (9)				
180. Routine surveillance (6–12 months) in an asymptomatic child or adult with moderate supravalvular AS	A (9)				
181. Routine surveillance (2–5 years) in an asymptomatic adult with moderate supravalvular AS		M (5)	M (6)	M (5)	M (5)
182. Evaluation due to change in clinical status and/or new concerning signs or symptoms	A (9)	M (6)	A (7)	A (7)	M (6)
183. Evaluation prior to planned repair	A (9)	M (6)	A (7)	A (8)	
Postoperative	**TTE**	**TEE**	**CMR**	**CCT**	**Stress Imaging**
184. Routine postoperative evaluation (within 30 days)	A (9)				
185. Evaluation due to change in clinical status and/or new concerning signs or symptoms	A (9)	M (6)	A (7)	A (7)	A (7)
186. Routine surveillance (2–5 years) in a patient with no or mild supravalvular AS	A (9)	R (3)	M (4)	M (4)	R (3)
187. Routine surveillance (6–12 months) in a patient with ≥moderate supravalvular AS	A (9)	M (4)	M (5)	M (5)	M (4)

A = Appropriate; AR = aortic regurgitation; AS = aortic stenosis; AUC = Appropriate Use Criteria; CCT = cardiovascular computed tomography; CMR = cardiovascular magnetic resonance; Fluoro = fluoroscopy; LV = left ventricle; LVOT = left ventricular outflow tract; M = May Be Appropriate; R = Rarely Appropriate; TEE = transesophageal echocardiogram; TTE = transthoracic echocardiogram.

Table 11 Results and Discussion

In patients with unrepaired subvalvular AS, TTE was rated Appropriate for all indications. TEE and CMR were rated May Be Appropriate for routine surveillance every 6-12 months in those with ≥ moderate stenosis. CCT and stress imaging were rated Rarely Appropriate every 6 to 12 months but May Be Appropriate every 3 to 5 years. For postoperative evaluation, TTE for all indications and TEE for evaluation due to change in status and new concerning signs or symptoms were rated Appropriate. TEE, CMR, CCT, and stress imaging were rated May Be Appropriate for those with ≥ moderate stenosis and/or AR or heart failure symptoms.

For unrepaired aortic valve disease, TTE was rated Appropriate for all indications, except for routine surveillance every 6 months in an asymptomatic child with mild AS and/or AR with no aortic dilation, where it was rated as Rarely Appropriate. CMR and CCT were rated Rarely Appropriate for routine surveillance every 1 to 2 years in those with mild disease but rated May Be Appropriate for those with ≥ moderate disease at a 6- to 12-month interval. For those with no aortic dilation,

routine surveillance every 3 to 5 years with CMR and CCT was rated May Be Appropriate, but both were rated Appropriate at a 6- to 12-month interval for those with increasing z-scores of the aorta. In general, these ratings are concordant with those for similar indications in the 2017 AUC for adults with valvular heart disease (14). Following surgical or catheter-based intervention, TTE was rated Appropriate for all indications. TEE, CMR, CCT, and fluoro were also rated Appropriate for evaluation due to change in clinical status, but stress imaging was rated May Be Appropriate. CMR, CCT, and stress imaging were rated May Be Appropriate for surveillance in those with ≥ moderate disease at a 6- to 12-month interval or for those with heart failure symptoms.

In those with unrepaired supravalvular AS, TTE was rated Appropriate for all indications. TEE, CMR, CCT, and stress imaging were rated May Be Appropriate at a 2- to 5-year interval for routine surveillance of asymptomatic adults with moderate supravalvular AS. CMR and CCT were rated Appropriate following a change in clinical status as well as prior to planned repair owing to the frequent involvement of coronary arteries with this lesion. TTE following surgical repair was rated Appropriate for all scenarios, and CMR, CCT, and stress imaging were rated Appropriate only if there is a need to evaluate owing to change in clinical status. Stress imaging was rated Rarely Appropriate for routine surveillance in those with no or mild residual supravalvular AS but May Be Appropriate in those with ≥ moderate disease at a 6- to 12-month interval.

Table 12 Considerations

This table includes clinical scenarios related to aortic coarctation and interrupted aortic arch before and after operative repair, angioplasty, or aortic stent placement. This table does not include evaluation in infants with suspected coarctation and a patent ductus arteriosus, in whom more frequent imaging may be needed. Imaging for bicuspid aortic valve is included in **Table 11** (LVOT Lesions). In the postprocedural evaluation scenarios, the concerns may include residual or recurrent aortic obstruction, aortic aneurysm, in-stent stenosis, stent fracture, and endoleak. In the postprocedural scenarios for asymptomatic patients, age is considered as this impacts the frequency of imaging surveillance and the modalities under consideration.

Table 12 Results and Discussion

In the asymptomatic patient with unrepaired mild aortic coarctation, TTE was rated Appropriate for routine surveillance, with more frequent imaging at a younger age. For the same indications, CMR and CCT were rated Appropriate for routine surveillance every 3 to 5 years but Rarely Appropriate at more frequent intervals (1 to 2

TABLE 12 Aortic Coarctation and Interrupted Aortic Arch

		AUC Score				
Unrepaired		**TTE**	**TEE**	**CMR**	**CCT**	**Stress Imaging**
188.	Routine surveillance (3–6 months) in an infant with mild aortic coarctation in the absence of a PDA	A (9)				
189.	Routine surveillance (1–2 years) in a child or adult with mild aortic coarctation	A (9)		R (3)	R (3)	
190.	Routine surveillance (3–5 years) in a child or adult with mild aortic coarctation			A (8)	A (7)	
191.	Evaluation due to change in clinical status and/or new concerning signs or symptoms	A (9)	M (4)	A (8)	A (7)	M (6)
192.	Evaluation prior to planned repair	A (9)		A (8)	A (8)	
Postprocedural: Surgical or Catheter-Based		**TTE**	**TEE**	**CMR**	**CCT**	**Fluoro**
193.	Routine postprocedural evaluation (within 30 days)	A (9)		R (3)	R (3)	
194.	Evaluation due to change in clinical status and/or new concerning signs or symptoms	A (9)	M (4)	A (7)	A (7)	M (5)
195.	Routine surveillance (3–6 months) within the first year following surgical or catheter-based intervention in an asymptomatic patient with no or mild sequelae	A (9)				
196.	Routine surveillance (6–12 months) within the first year following catheter-based intervention in an asymptomatic patient with no or mild sequelae			M (5)	M (6)	
197.	Routine surveillance (6 months) after the first year following surgical or catheter-based intervention in an asymptomatic patient with no or mild sequelae	A (9)				
198.	Routine surveillance (1–2 years) after the first year following surgical or catheter-based intervention in an asymptomatic patient with no or mild sequelae	A (9)		M (5)	M (4)	
199.	Routine surveillance (3–5 years) in an asymptomatic patient to evaluate for aortic arch aneurysms, in-stent stenosis, stent fracture, or endoleak			A (8)	A (8)	M (4)
200.	Routine surveillance (3–12 months) in a patient with heart failure symptoms	A (9)		M (6)	M (5)	

A = Appropriate; AUC = Appropriate Use Criteria; CCT = cardiovascular computed tomography; CMR = cardiovascular magnetic resonance; M = May Be Appropriate; PDA = patent ductus arteriosus; R = Rarely Appropriate; TEE = transesophageal echocardiogram; TTE = transthoracic echocardiogram.

years). TTE, CMR, and CCT were all rated Appropriate prior to planned repair and for evaluation due to a change in clinical status or new concerning signs and/or symptoms. For routine postprocedural evaluation, TTE was rated Appropriate, and CMR and CCT were rated Rarely Appropriate. For evaluation due to change in clinical status and/or new concerning signs or symptoms, TTE, CMR, and CCT were rated Appropriate, and TEE and fluoro were rated May Be Appropriate. Postprocedural routine surveillance by TTE was rated Appropriate for all indications. CMR and CCT were rated May Be Appropriate for routine surveillance every 1 to 2 years after the first year following intervention in an asymptomatic patient with no or mild sequelae, and for routine surveillance every 3 to 12 months in a patient with heart failure symptoms; they were rated Appropriate for routine surveillance every 3 to 5 years in an asymptomatic patient to evaluate for aortic arch aneurysms, in-stent stenosis, stent fracture, or endoleak.

Table 13 Considerations

Congenital coronary anomalies include abnormal origin of a coronary artery from the PA, anomalous aortic origin of a coronary artery from a different aortic sinus of Valsalva (left coronary artery from the right sinus of Valsalva or right coronary artery from the left sinus of Valsalva), coronary arteriovenous fistula, and coronary artery ostial atresia, all in the setting of normal conotruncal anatomy. A coronary artery anomaly in the setting of other CHDs is beyond the scope of this table. In addition, isolated abnormalities in coronary artery branching (without an interarterial course) are also not included because these findings usually do not have clinical significance. Patients with anomalous origin of a coronary artery from the PA, anomalous origin of the left coronary artery from the right sinus of Valsalva, and coronary artery ostial atresia almost always undergo surgical repair shortly after diagnosis. Therefore, AUC scenarios for evaluation or surveillance after these diagnoses have been made are not included in this table.

The literature in the setting of anomalous aortic origin of a coronary artery and sudden death remains limited to observational studies. Therefore, the decision to pursue intervention or observation is beyond the scope of this document. Recommendations for follow-up after intervention are based on expert consensus opinion and the best available evidence. Interventions for abnormal origin of a coronary artery include reimplantation, bypass graft, Takeuchi repair (baffling a coronary artery within the pulmonary root to the aorta), unroofing of an intramural coronary artery segment, coronary artery translocation, and PA translocation. Interventions for coronary arteriovenous fistulae include transcatheter coil or device occlusion, as well as surgical ligation. The most common intervention for coronary artery ostial atresia is a bypass graft. Postoperative issues related to reimplanted

TABLE 13 Coronary Anomalies

		AUC Score			
Unrepaired		**TTE**	**CMR**	**CCT**	**Stress Imaging**
201.	Routine surveillance (annually) in an asymptomatic patient with anomalous right coronary artery from the left aortic sinus	R (3)			R (3)
202.	Routine surveillance (2–5 years) in an asymptomatic patient with anomalous right coronary artery from the left aortic sinus	A (7)	R (3)	R (3)	A (7)
203.	Routine surveillance (annually) in an asymptomatic patient with small coronary fistula	R (3)			R (3)
204.	Routine surveillance (2–5 years) in an asymptomatic patient with small coronary fistula	A (8)	R (3)	R (3)	R (3)
205.	Routine surveillance (1–2 years) in an asymptomatic patient with moderate or large coronary fistula	A (9)	M (5)	M (5)	M (5)
206.	Evaluation due to change in clinical status and/or new concerning signs or symptoms	A (9)	A (7)	A (7)	A (8)
207.	Evaluation prior to planned repair	A (9)	A (7)	A (7)	M (6)
Postprocedural: Surgical or Catheter-Based		**TTE**	**CMR**	**CCT**	**Stress Imaging**
208.	Routine post-procedural evaluation (within 30 days)	A (9)	M (4)	M (5)	
209.	Evaluation due to change in clinical status and/or new concerning signs or symptoms	A (9)	A (7)	A (7)	A (8)
210.	Evaluation within 1 year after surgery or catheter-based intervention with no or mild sequelae	A (9)	M (6)	M (6)	M (4)
211.	Routine surveillance (1–3 months) within the first year following repair	A (7)	R (3)	R (3)	R (3)
212.	Routine surveillance (3–6 months) in an infant with or without ventricular or valvular dysfunction	A (9)	R (3)	R (3)	R (3)
213.	Routine surveillance (3–6 months) in a child or adult with ventricular or valvular dysfunction	A (9)	M (5)	M (5)	M (5)
214.	Routine surveillance (annually) with no or mild sequelae	A (7)	R (3)	R (3)	R (3)
215.	Routine surveillance (2–5 years) with no or mild sequelae		M (5)	M (5)	M (6)

A = Appropriate; AUC = Appropriate Use Criteria; CCT = cardiovascular computed tomography; CMR = cardiovascular magnetic resonance; M = May Be Appropriate; R = Rarely Appropriate; TTE = transthoracic echocardiogram.

coronary arteries after an arterial switch procedure for transposition of the great arteries or a Ross procedure for aortic valve disease are not included in this table.

Table 13 Results and Discussion

In an asymptomatic patient with anomalous right coronary artery from the left aortic sinus, routine surveillance every 2 to 5 years by TTE and stress imaging were rated Appropriate, whereas routine annual surveillance by TTE and stress imaging and routine surveillance every 2 to 5 years by CMR and CCT were rated Rarely Appropriate. Aside from stress imaging, the appropriateness of various tests was similar for an asymptomatic patient with a small coronary fistula: Routine surveillance every 2 to 5 years by TTE was rated Appropriate, and routine annual surveillance by stress imaging was rated Rarely Appropriate. In an asymptomatic patient with a moderate or large coronary fistula, routine surveillance every 1 to 2 years by TTE was rated Appropriate, and routine surveillance every 1 to 2 years by CMR, CCT, and stress imaging were rated May Be Appropriate. TTE, CMR, and CCT were rated Appropriate in the evaluation of a patient with a coronary anomaly, when there is a change in clinical status and/or new concerning signs and symptoms, and in a patient prior to planned repair. Stress imaging was rated Appropriate for evaluation due to change in clinical status and/or new concerning signs or symptoms and May Be Appropriate for evaluation prior to planned repair.

After surgical or catheter-based intervention, TTE was rated Appropriate and CMR and CCT were rated May Be Appropriate in routine evaluation within 30 days after the intervention. TTE, CMR, CCT, and stress imaging were rated Appropriate if there was a change in clinical status and/or new concerning signs or symptoms. TTE was rated Appropriate for evaluation within 1 year after the intervention with no or mild sequelae, while CMR and CCT were rated May Be Appropriate. TTE was also rated Appropriate in routine surveillance every 1 to 3 months for all patients within 1 year after the intervention, every 3 to 6 months for infants with or without ventricular or valvular dysfunction, every 3 to 6 months for children and adults with ventricular or valvular dysfunction, and annually for all patients with no or mild sequelae. CMR, CCT, and stress imaging were rated May Be Appropriate in routine surveillance every 3 to 6 months for children and adults with ventricular or valvular dysfunction and every 2 to 5 years for all patients with no or mild sequelae, and they were rated Rarely Appropriate in routine surveillance every 1 to 3 months for all patients within 1 year after the intervention, every 3 to 6 months for all infants with or without ventricular or valvular dysfunction, and annually for all patients with no or mild sequelae.

Table 14 Considerations

This table addresses patients with TOF and is divided into 3 sections based on the current status of repair: pre-repair, following initial repair, and following subsequent interventions. It includes patients with the more common form of TOF with pulmonary stenosis and the more severe form of TOF with pulmonary atresia (TOF/PA), recognizing that the more severe form of TOF may require more frequent imaging (within the range of times provided). Patients with TOF/PA with major aortopulmonary collateral arteries as a primary source of pulmonary blood flow are not included in this document. Surveillance following palliative shunts and stents is included in the unrepaired section of this table because these patients have not undergone complete initial repair. The post-initial repair scenarios include concerns for ventricular dilation and dysfunction, valvular dysfunction, RV outflow tract (RVOT) obstruction, residual shunt, stent fracture, and deteriorating exercise capacity. The postprocedural indications have been combined for surgical or transcatheter PVR when the frequency of imaging and type of imaging modalities used are similar but are separated when there are specific clinical policy recommendations available for either. Indication 222 is for surveillance following intervention and includes patients with PR of any severity.

Table 14 Results and Discussion

In the asymptomatic patient with unrepaired TOF, or following palliation with a shunt, RVOT or PDA stent, TTE was rated Appropriate for routine surveillance every 1 to 3 months. TTE was also rated Appropriate prior to planned repair or for evaluation due to a change in clinical status or new concerning signs and/or symptoms, while CMR and CCT were rated May Be Appropriate in these scenarios.

Following complete repair, TTE was rated Appropriate for routine postprocedural evaluation. For an evaluation with a change in clinical status and/or new concerning signs or symptoms, TTE and CMR were rated Appropriate, and TEE, CCT, stress imaging and lung scan were rated May Be Appropriate. In an asymptomatic patient with no or mild sequelae or PR of any severity, annual routine surveillance with TTE was rated Appropriate, whereas CMR, CCT and lung scan were rated Rarely Appropriate. For routine surveillance every 2 to 3 years in a patient with PR and preserved ventricular function, CMR was rated Appropriate; however, CCT was rated May Be Appropriate and lung scan Rarely Appropriate.

Prior to planned PVR, TTE, CMR, and CCT were rated Appropriate, while TEE was rated May Be Appropriate and lung scan Rarely Appropriate. In the immediate post-PVR evaluation period, TTE was rated Appropriate, and CMR, CCT, fluoro, and lung scan were Rarely Appropriate. In the

TABLE 14 Tetralogy of Fallot (TOF)

	AUC Score						
Unrepaired	**TTE**	**TEE**		**CMR**		**CCT**	
216. Routine surveillance (1–3 months) in an infant before complete repair	A (9)						
217. Routine surveillance (1–3 months) in an infant following valvuloplasty, PDA and/or RVOT stenting, or shunt placement before complete repair	A (9)						
218. Evaluation due to change in clinical status and/or new concerning signs or symptoms	A (9)			M (5)		M (5)	
219. Evaluation prior to planned repair	A (9)	R (2)		M (5)		M (5)	
Postoperative: Initial Repair	**TTE**	**TEE**	**CMR**	**CCT**	**Stress Imaging**		**Lung Scan**
220. Routine postoperative evaluation (within 30 days)	A (9)						
221. Evaluation due to change in clinical status and/or new concerning signs or symptoms	A (9)	M (5)	A (7)	M (6)	M (5)		M (5)
222. Routine surveillance (annually) in an asymptomatic patient with no or mild sequelae or PR of any severity	A (9)		R (3)	R (3)			R (3)
223. Routine surveillance (6–12 months) in a patient with valvular dysfunction other than pulmonary valve, RVOT obstruction, branch pulmonary artery stenosis, arrhythmias, or presence of an RV-to-PA conduit	A (9)	M (4)	M (5)	M (5)			M (5)
224. Routine surveillance (2–3 years) in a patient with PR and preserved ventricular function			A (7)	M (6)			R (3)
225. Routine surveillance (3–12 months) in a patient with heart failure symptoms	A (9)		M (5)	M (4)			R (3)
226. Evaluation prior to planned pulmonary valve replacement (percutaneous or surgical)	A (9)	M (5)	A (8)	A (8)			R (3)
Postprocedural: Surgical or Catheter-based Pulmonary Valve Replacement	**TTE**	**TEE**	**CMR**	**CCT**	**Stress Imaging**	**Fluoro**	**Lung Scan**
227. Routine postprocedural evaluation (within 30 days)	A (9)		R (3)	R (3)		R (3)	R (3)
228. Evaluation due to change in clinical status and/or new concerning signs or symptoms	A (9)	M (5)	A (8)	A (7)	M (6)	M (6)	M (6)
229. Evaluation at 1 year following transcatheter or surgical pulmonary valve replacement	A (9)		M (6)	M (6)		M (5)	
230. Routine surveillance at 1 and 6 month(s) in an asymptomatic patient following transcatheter pulmonary valve replacement	A (9)		R (3)	R (3)			
231. Routine surveillance (annually) in an asymptomatic patient following transcatheter pulmonary valve replacement	A (9)		R (3)	R (3)			
232. Routine surveillance (annually) in an asymptomatic patient with no or mild sequelae	A (9)		R (3)	R (3)			R (3)
233. Routine surveillance (6–12 months) in a patient with RV-to-PA conduit dysfunction, valvular or ventricular dysfunction, branch pulmonary artery stenosis, or arrhythmias	A (9)	R (3)	M (5)	M (4)			M (4)
234. Routine surveillance (2–3 years) in an asymptomatic patient with no or mild sequelae			A (7)	M (6)			M (5)
235. Routine surveillance (2–3 years) in a patient with valvular or ventricular dysfunction, RVOT obstruction, branch pulmonary artery stenosis, arrhythmias, or presence of an RV-to-PA conduit	A (9)	M (4)	A (8)	A (7)			M (6)
236. Routine surveillance (3–12 months) in a patient with heart failure symptoms	A (9)		M (5)	M (5)			

A = Appropriate; AUC = Appropriate Use Criteria; CCT = cardiovascular computed tomography; CMR = cardiovascular magnetic resonance; Fluoro = fluoroscopy; M = May Be Appropriate; PDA = patent ductus arteriosus; PR = pulmonary regurgitation; R = Rarely Appropriate; RV-to-PA = right ventricle to pulmonary artery; RVOT = right ventricular outflow tract; TEE = transesophageal echocardiogram; TOF = tetralogy of Fallot; TTE = transthoracic echocardiogram.

evaluation of a TOF patient with PVR and a change in clinical status and/or new concerning signs or symptoms, TTE, CMR, and CCT were rated Appropriate, and TEE, stress imaging, fluoro, and lung scan were rated May Be Appropriate. For evaluation at 1 year following PVR, TTE was rated Appropriate and CMR, CCT, and fluoro were rated May Be Appropriate. Following either surgical or transcatheter PVR, TTE was rated Appropriate in all scenarios at the designated frequency. For annual surveillance in a patient with no or mild sequalae, other imaging modalities were rated Rarely Appropriate. For surveillance every 2-3 years in this setting, CMR was rated Appropriate and CCT and lung scan were rated May Be Appropriate. In patients with RV-to-PA conduit dysfunction, valvular or ventricular dysfunction, branch pulmonary artery stenosis, or arrhythmias, CMR and CCT were rated Appropriate every 2 to 3 years and May Be Appropriate for routine surveillance every 6 to 12 months. For routine surveillance every 3 to 12 months in an adult with repaired TOF with or without PVR and heart failure symptoms, TTE was rated Appropriate and CMR and CCT were rated May Be Appropriate.

Table 15 Considerations

Table 15 addresses clinical scenarios related to patients with DORV. Most patients with DORV undergo surgical

intervention shortly after the diagnosis is made. However, some patients are able to maintain adequate cardiac output and tissue oxygen delivery with balanced systemic and pulmonary circulations when the anatomy involves some degree of pulmonary stenosis and an unobstructed aorta, thus allowing for delayed surgical intervention. Consequently, imaging studies may be considered for these patients after the initial diagnosis is made and before any corrective surgical intervention.

Postoperative evaluation of these patients assumes that the anatomic substrate is amenable to a biventricular repair. DORV variants that require single-ventricle palliation are not included in this section. In addition, DORV involves a wide spectrum of anatomic arrangements, resulting in variable physiological presentations. Surgical intervention is therefore determined by the predominant preoperative physiology and includes repairs for any of these anatomic substrates (ventricular septal defect closure baffling the LV to the aorta, TOF-type repair, and arterial switch procedures with ventricular septal defect closure baffling the LV to the native pulmonary/neoaortic valve). Potential complications after biventricular repair include ventricular dilation and/or dysfunction, left or RV outflow tract obstruction, and residual shunt lesion. Postoperative evaluation must include these possible sequelae, particularly for the patient with unexplained symptoms or deteriorating exercise capacity. Occasionally, repair will involve placement of a conduit or homograft from the RV to the PA. Because most of these patients will require replacement or additional intervention over time, surveillance of these patients has been included among other patients with ventricular outflow tract obstruction. Refer to Table 14 (Tetralogy of Fallot) for surveillance and evaluation of transcatheter PVR patients.

Table 15 Results and Discussion

TTE was rated Appropriate for all the scenarios in the setting of an unrepaired DORV. In evaluation due to a change in clinical status, new concerning signs or symptoms, and prior to planned repair of DORV, TEE was rated May Be Appropriate and CMR and CCT were rated Appropriate. In postoperative DORV, evaluation and routine surveillance with TTE was rated Appropriate in all scenarios. CMR and CCT were rated Appropriate for an evaluation due to change in clinical status and/or new concerning signs or symptoms. CMR was rated Appropriate, while CCT was rated May Be Appropriate for routine surveillance of the postoperative DORV every 3 to 5 years. Stress imaging was rated May Be Appropriate in all the scenarios. Finally, TEE and lung scan were rated Rarely Appropriate in the routine surveillance of an asymptomatic postoperative DORV every 3 to 5 years.

TABLE 15 Double Outlet Right Ventricle (DORV)

		AUC Score					
Unrepaired		**TTE**	**TEE**	**CMR**	**CCT**		
237.	Routine surveillance (1–3 months) in an infant with balanced systemic and pulmonary circulation	A (9)					
238.	Routine surveillance (3–6 months) in a child with balanced systemic and pulmonary circulation	A (9)					
239.	Evaluation due to change in clinical status and/or new concerning signs or symptoms	A (9)	M (4)	A (7)	A (7)		
240.	Evaluation prior to planned repair	A (9)	M (4)	A (7)	A (7)		
Postoperative		**TTE**	**TEE**	**CMR**	**CCT**	**Stress Imaging**	**Lung Scan**
241.	Routine postprocedural evaluation (within 30 days)	A (9)					
242.	Evaluation due to change in clinical status and/or new concerning signs or symptoms	A (9)	M (6)	A (7)	A (7)	M (5)	M (4)
243.	Routine surveillance (6 months) within a year following repair in an asymptomatic infant or child with no or mild sequelae	A (9)					
244.	Routine surveillance (1–2 years) in an asymptomatic patient with no or mild sequelae	A (9)					
245.	Routine surveillance (3–12 months) in a patient with valvular or ventricular dysfunction, right or left ventricular outflow tract obstruction, branch pulmonary artery stenosis, arrhythmias, or presence of an RV-to-PA conduit	A (9)	M (4)	M (5)	M (5)	M (4)	M (4)
246.	Routine surveillance (3–5 years) in an asymptomatic patient with no or mild sequelae		R (3)	A (7)	M (6)	M (5)	R (3)
247.	Routine surveillance (3–12 months) in a patient with heart failure symptoms	A (9)		M (5)	M (5)		

A = Appropriate; AUC = Appropriate Use Criteria; CCT = cardiovascular computed tomography; CMR = cardiovascular magnetic resonance; DORV = double outlet right ventricle; M = May Be Appropriate; R = Rarely Appropriate; RV-to-PA = right ventricle to pulmonary artery; TEE = transesophageal echocardiogram; TTE = transthoracic echocardiogram.

Table 16 Considerations

This table addresses clinical scenarios associated with TGA. Most patients with TGA undergo surgical intervention shortly after the diagnosis is made. However, some extenuating circumstances may preclude immediate surgical intervention, and the patient's clinical status may be temporarily stable in the setting of adequate mixing of arterial and venous blood. Consequently, imaging

TABLE 16 D-Loop Transposition of the Great Arteries (D-Loop TGA)

	AUC Score					
Unrepaired	**TTE**		**CMR**		**CCT**	
248. Evaluation due to change in clinical status and/or new concerning signs or symptoms	A (9)		M (4)		M (4)	
249. Evaluation prior to planned repair	A (9)		M (6)		M (6)	
Postoperative: Arterial Switch Operation	**TTE**	**TEE**	**CMR**	**CCT**	**Stress Imaging**	**Lung Scan**
250. Routine postoperative evaluation (within 30 days)	A (9)					
251. Evaluation due to change in clinical status and/or new concerning signs or symptoms	A (9)	M (6)	A (7)	A (7)	M (6)	M (5)
252. Evaluation for coronary imaging in an asymptomatic patient			A (8)	A (9)	A (7)	
253. Routine surveillance (1–3 months) in an asymptomatic infant with moderate sequelae	A (9)					
254. Routine surveillance (3–6 months) in an asymptomatic infant with no or mild sequelae	A (9)					
255. Routine surveillance (3–12 months) in an asymptomatic child or adult with ≥moderate valvular or ventricular dysfunction, right or left ventricular outflow tract obstruction, branch pulmonary artery stenosis, or arrhythmias	A (9)		M (6)	M (6)	M (5)	M (4)
256. Routine surveillance (1–2 years) in an asymptomatic child or adult with no or mild sequelae	A (9)		M (4)	M (4)	R (3)	R (3)
257. Routine surveillance (3–5 years) in an asymptomatic patient		M (4)	A (7)	A (7)	M (6)	M (5)
258. Routine surveillance (1–2 years) in a patient with dilated neoaortic root with increasing Z scores, or neoaortic regurgitation	A (9)	R (3)	A (8)	A (7)		
259. Routine surveillance (3–12 months) in a patient with heart failure symptoms	A (9)		M (5)	M (5)	M (4)	
Postoperative: Rastelli	**TTE**	**TEE**	**CMR**	**CCT**	**Stress Imaging**	**Lung Scan**
260. Routine postoperative evaluation (within 30 days)	A (9)					
261. Evaluation due to change in clinical status and/or new concerning signs or symptoms	A (9)	M (5)	A (7)	A (7)	M (6)	M (6)
262. Routine surveillance (3–6 months) within the first year following repair	A (9)					
263. Routine surveillance (6 months) after the first year following repair in an asymptomatic patient with no or mild sequelae	A (9)		R (3)	R (3)		
264. Routine surveillance (1–2 years) in an asymptomatic patient with no or mild sequelae	A (9)		M (5)	M (4)		
265. Routine surveillance (3–5 years) in an asymptomatic patient			A (7)	A (7)		
266. Routine surveillance (3–12 months) in a patient with ≥ moderate valvular dysfunction, LVOT obstruction, presence of an RV-to-PA conduit, branch pulmonary artery stenosis, or arrhythmias	A (9)		M (5)	M (5)	R (3)	M (4)
267. Routine surveillance (3–12 months) in a patient with heart failure symptoms	A (9)		M (5)	M (5)	M (5)	
Postoperative: Atrial Switch Operation	**TTE**	**TTE + Contrast**	**TEE**	**CMR**	**CCT**	**Stress Imaging**
268. Evaluation due to concerning signs or symptoms and/or change in clinical status	A (9)	M (6)	M (6)	A (7)	A (7)	M (6)
269. Routine surveillance (6 months) in an asymptomatic patient with no or mild sequelae	R (3)	R (3)		R (3)	R (3)	
270. Routine surveillance (1–2 years) in an asymptomatic patient with no or mild sequelae	A (9)	R (3)		R (3)	R (3)	
271. Routine surveillance (3–5 years) in an asymptomatic patient		R (3)		A (7)	A (7)	
272. Routine surveillance (3–12 months) in a patient with ≥ moderate systemic AV valve regurgitation, systemic RV dysfunction, LVOT obstruction, or arrhythmias	A (9)		M (5)	A (7)	A (7)	M (5)
273. Routine surveillance (3–12 months) in a patient with heart failure symptoms	A (9)			M (6)	M (6)	

A = Appropriate; AUC = Appropriate Use Criteria; CCT = cardiovascular computed tomography; CMR = cardiovascular magnetic resonance; LVOT = left ventricular outflow tract; M = May Be Appropriate; R = Rarely Appropriate; RV = right ventricle; RV-to-PA = right ventricle to pulmonary artery; TEE = transesophageal echocardiogram; TGA = transposition of the great arteries; TTE = transthoracic echocardiogram.

studies may be considered for these patients after the initial diagnosis is made and while awaiting surgical intervention.

Postoperative evaluation of these patients is determined in large part by the type of surgical intervention that is performed, and these include the arterial switch procedure, the Rastelli procedure with baffling of the LV through the ventricular septal defect to the aorta (thereby closing the defect) and establishment of a connection from the RV to the PA (usually with a conduit or homograft), and the atrial switch operation. Indications for imaging after the Nikaidoh procedure (which involves translocating the aorta to the LV, closing the ventricular septal defect, and establishing a connection from the RV to the PA with a conduit or homograft) are not included in this document.

The most common sequelae after the arterial switch procedure include coronary artery stenosis, branch pulmonary artery stenosis, neoaortic valve regurgitation, and neoaortic root dilation. The most common sequelae after the Rastelli procedure include a residual VSD and obstruction along the right or left ventricular outflow tracts. Surveillance for patients with a clinically significant degree of any of these sequelae is generally similar, and these indications have been combined in the table. The most common sequelae after an atrial switch operation include baffle leak or obstruction, systemic RV dysfunction, systemic atrioventricular valve regurgitation, atrial arrhythmias, LVOT obstruction, and PH. These patients also have a higher incidence of pacemaker or implantable defibrillator placement, which may affect the decision regarding CMR versus CCT. Surveillance for patients with a clinically significant degree of these sequelae is generally similar; therefore, these indications have been combined in the table. Please refer to **Table 14** (Tetralogy of Fallot) for surveillance and evaluation of patients with transcatheter PVR.

Table 16 Results and Discussion

TTE was rated Appropriate, and CMR and CCT were rated May Be Appropriate for all scenarios in the setting of an unrepaired D-loop TGA. Evaluation and routine surveillance with TTE after an arterial switch operation was rated Appropriate in all scenarios. CMR, CCT, and stress imaging were all rated Appropriate in the evaluation of coronaries in an asymptomatic patient after an arterial switch operation. This is supported by the 2008 and 2018 Guidelines for the Management of Adults With Congenital Heart Disease (1,16). Routine surveillance with CMR and CCT of patients after arterial switch operation was rated Appropriate every 3 to 5 years and May Be Appropriate for all other scenarios. CMR and CCT were rated Appropriate every 1 to 2 years for evaluation of a dilated neoaortic root with increased Z scores, or neoaortic regurgitation. The use of routine surveillance with stress imaging and lung scan was rated May Be Appropriate in most scenarios except for surveillance every 1 to 2 years in an asymptomatic patient with no or mild sequelae, where it was rated Rarely Appropriate.

Evaluation and routine surveillance with TTE was rated Appropriate in all scenarios in patients after a Rastelli procedure. CMR and CCT were rated Appropriate for evaluation due to change in clinical status and routine surveillance every 3 to 5 years, and May Be Appropriate for routine surveillance every 3 to 12 months in patients with heart failure and every 3 to 12 months in patients with ≥ moderate residual sequelae. Stress imaging was rated May Be Appropriate for an evaluation due to a change in clinical status, and routine surveillance (3 to 12 months) for heart failure symptoms, but Rarely Appropriate every 3 to 12 months in a patient with ≥ moderate residual sequelae. Lung scan was rated May Be Appropriate in those with a change in clinical status or for patients with ≥ moderate residual sequelae.

Evaluation and routine surveillance with TTE was rated Appropriate in all scenarios in patients after an atrial switch operation except in the setting of routine surveillance every 6 months, which was rated Rarely Appropriate. CMR and CCT were rated Appropriate in the evaluation due to concerning signs or symptoms, in routine surveillance every 3 to 5 years, and in a patient with ≥ moderate residual sequelae. TTE + contrast was rated May Be Appropriate in the evaluation due to concerning signs or symptoms but rated as Rarely Appropriate in routine surveillance. Stress imaging was rated May Be Appropriate in the evaluation due to concerning signs or symptoms, and in routine surveillance in a patient with ≥ moderate residual sequelae.

Table 17 Considerations

This table includes clinical scenarios related to ccTGA, also known as L-loop transposition of the great arteries. The most commonly associated defects are VSDs, PS, and tricuspid valve or systemic atrioventricular (AV) valve anomalies. Patients with unrepaired ccTGA with VSD and/or PS are not considered in this table. Systemic AV valve anomalies can range from an Ebstein-like anomaly to functional systemic AV valve regurgitation in the setting of RV dilation. The first part of this table focuses on patients with unrepaired ccTGA, including patients with varying severity of systemic AV valve regurgitation.

Scenarios focus on the presence or absence of systemic RV dysfunction rather than the severity of systemic RV dysfunction.

A separate section of this table is dedicated to patients who have undergone an anatomic repair, including atrial switch (Mustard/Senning) with arterial switch, and atrial switch with Rastelli operation. This postprocedural section includes evaluation for common complications after these repairs but is not an exhaustive list of scenarios. These complications include ventricular dilation, valvular or ventricular dysfunction, left or RV outflow tract obstruction, and systemic or pulmonary venous pathway obstruction or baffle leak. Patients after atrial switch operation have a higher incidence of pacemakers or implantable defibrillators that may affect the decision regarding CMR versus CCT. Complications after the arterial switch procedure, such as neoaortic dilation, neoaortic regurgitation, and coronary artery stenosis, are not included here but can be found in Table 16 (D-Loop TGA).

TABLE 17 Congenitally Corrected Transposition of the Great Arteries (ccTGA)

	AUC Score				
Unrepaired	**TTE**	**TEE**	**CMR**	**CCT**	**Stress Imaging**
274. Evaluation due to change in clinical status and/or new concerning signs or symptoms	A (9)	M (6)	A (7)	A (7)	M (6)
275. Routine surveillance (3–6 months) in an asymptomatic infant	A (9)				
276. Routine surveillance (1–2 years) in a patient with <moderate systemic AV valve regurgitation	A (9)	R (3)	R (3)	R (3)	R (3)
277. Routine surveillance (6–12 months) in a patient with ≥moderate systemic AV valve regurgitation	A (9)	M (5)	M (6)	M (5)	M (5)
278. Routine surveillance (3–5 years) in an asymptomatic patient	A (9)	M (5)	A (7)	A (7)	M (4)
279. Routine surveillance (3–12 months) in a patient with heart failure symptoms	A (9)		M (5)	M (5)	
280. Evaluation prior to planned repair	A (9)	A (7)	A (8)	A (8)	

Postoperative: Anatomic Repair	**TTE**	**TTE + Contrast**	**TEE**	**CMR**	**CCT**	**Stress Imaging**
281. Routine post-operative evaluation (within 30 days)	A (9)	M (5)				
282. Evaluation due to change in clinical status and/or new concerning signs or symptoms	A (9)	M (5)	M (5)	A (7)	A (7)	M (6)
283. Routine surveillance (3–6 months) within a year following repair in an asymptomatic patient with no or mild sequelae	A (9)					
284. Routine surveillance (1–2 years) after the first year following repair in an asymptomatic patient with no or mild sequelae	A (9)			M (5)	M (5)	M (5)
285. Routine surveillance (6–12 months) in a patient with valvular or ventricular dysfunction, right or left ventricular outflow tract obstruction, or presence of a RV-to-PA conduit	A (9)		R (3)	A (7)	A (7)	M (5)
286. Routine surveillance (3–5 years) in an asymptomatic patient			M (5)	A (7)	A (7)	M (5)
287. Routine surveillance (3–12 months) in a patient with heart failure symptoms	A (9)			M (6)	M (6)	

Postoperative: Physiological Repair With VSD Closure and/or LV-to-PA Conduit	**TTE**	**TEE**	**CMR**	**CCT**	**Stress Imaging**
288. Routine postoperative evaluation (within 30 days)	A (9)				
289. Evaluation due to change in clinical status and/or new concerning signs or symptoms	A (9)	M (6)	A (7)	A (7)	M (6)
290. Routine surveillance (3–6 months) within a year following repair in an asymptomatic patient with no or mild sequelae	A (9)				
291. Routine surveillance (1–2 years) in an asymptomatic patient with no or mild sequelae	A (9)	M (5)	R (3)	R (3)	R (3)
292. Routine surveillance (3–5 years) in an asymptomatic patient with no or mild sequelae	A (9)	M (5)	A (7)	A (7)	M (5)
293. Routine surveillance (3–12 months) in a patient with ≥moderate systemic AV valve regurgitation, systemic RV dysfunction, and/or LV-to-PA conduit dysfunction	A (9)	M (5)	A (7)	A (7)	M (5)
294. Routine surveillance (3–12 months) in a patient with heart failure symptoms	A (9)		M (6)	M (6)	

A = Appropriate; AUC = Appropriate Use Criteria; AV = atrioventricular; CCT = cardiovascular computed tomography; ccTGA = congenitally corrected transposition of the great arteries; CMR = cardiovascular magnetic resonance; LV-to-PA = left ventricle to pulmonary artery; M = May Be Appropriate; R = Rarely Appropriate; RV = right ventricle; RV-to-PA = right ventricle to pulmonary artery; TEE = transesophageal echocardiogram; TTE = transthoracic echocardiogram.

The last section of this table includes patients who have undergone a physiological repair (biventricular repair without addressing the AV and ventriculoarterial discordance such as VSD closure and/or subpulmonic LV-to-PA conduit). This postprocedural section focuses on common complications after such a repair, including residual VSD, subpulmonic LV-to-PA conduit stenosis, systemic AV valve regurgitation, and systemic RV dysfunction. Systemic AV valve repair and replacement are not included in this table because they are covered in the 2017 AUC for Multimodality Imaging in Valvular Heart Disease (14). Finally, three of the scenarios are dedicated to the surveillance of systemic RV dysfunction every 3 to 5 years by CMR or CCT in an asymptomatic patient with either unrepaired ccTGA or ccTGA after physiological repair.

Table 17 Results and Discussion

TTE was rated Appropriate for all scenarios in the setting of unrepaired ccTGA. CMR and CCT were rated Appropriate for the evaluation due to change in clinical status, and rated May Be Appropriate for routine surveillance every 3 to 12 months in patients with heart failure and every 6 to 12 months in patients with ≥ moderate systemic AV valve regurgitation. CMR and CCT were rated Appropriate in routine surveillance every 3 to 5 years of an asymptomatic patient, and all modalities were rated Appropriate prior to planned ccTGA repair.

After anatomic repair of ccTGA, TTE was rated Appropriate for all scenarios. CMR and CCT were rated Appropriate for an evaluation due to change in clinical status and May Be Appropriate for routine surveillance every 1 to 2 years and every 3 to 12 months in patients with heart failure symptoms. CMR and CCT were rated Appropriate for routine surveillance every 6 to 12 months in patients with residual sequelae and for routine surveillance every 3 to 5 years in asymptomatic patients. TTE + contrast was rated May Be Appropriate postoperatively and for evaluation due to change in clinical status, and stress imaging was rated May Be Appropriate for all of the scenarios.

After physiological repair of ccTGA, TTE was rated Appropriate for all scenarios. CMR and CCT were rated Appropriate for an evaluation due to change in clinical status and for routine surveillance every 3 to 5 years in asymptomatic patients and every 3 to 12 months in patients with moderate residual sequelae. CMR and CCT were rated Rarely Appropriate for routine surveillance every 1 to 2 years and May Be Appropriate every 3 to 12 months in patients with heart failure symptoms. Stress imaging was rated May Be Appropriate for all the scenarios except for routine surveillance every 1 to 2 years in an asymptomatic patient with no or mild sequelae, where it was rated Rarely Appropriate.

Table 18 Considerations

This table addresses key components of cardiac imaging in patients with unrepaired TA and those who have undergone repair. Indications related to the dilated truncal root with or without truncal valve regurgitation and interrupted aortic arch have been addressed in Table 11 (LVOT Lesions) and Table 12 (Aortic Coarctation and Interrupted Aortic Arch). The second section of this table focuses on the postoperative patient with TA. Follow-up imaging in the postoperative period may include surveillance or evaluation for potential complications such as biventricular dysfunction, RV-to-PA conduit obstruction, truncal valve stenosis or regurgitation, branch pulmonary artery stenosis, and aortic arch obstruction. Surveillance and evaluation of patients with transcatheter PVR is addressed in Table 14 (Tetralogy of Fallot).

Table 18 Results and Discussion

In an unrepaired patient, TTE, CMR, and CCT were rated Appropriate for evaluation prior to planned repair or due to change in clinical status. Postoperatively, TTE was rated Appropriate for the routine postprocedural evaluation within 30 days. In the evaluation due to change in clinical status, TTE, CMR, and CCT were rated Appropriate while TEE, stress imaging, and lung scan were rated May Be Appropriate. TTE was rated Appropriate for routine surveillance of the asymptomatic patient with no or mild sequelae at 1 to 3 months within the first year following surgery and at 6 to 12 months thereafter, whereas TEE was rated Rarely Appropriate for this indication. TEE, CMR, CCT, and stress imaging were rated May Be Appropriate for routine surveillance (3 to 5 years) in both asymptomatic children and adults with no or mild sequelae. This provides an array of imaging modalities during this time interval that may be suitable for asymptomatic patients of varied ages. In asymptomatic children or adults with ≥ moderate truncal stenosis and/or regurgitation, TTE was rated Appropriate for routine surveillance (3 to 6 months), while all other imaging modalities for this indication were rated May Be Appropriate. For routine surveillance (1 to 2 years) in asymptomatic children or adults with ≥ moderate truncal stenosis and/or regurgitation, CMR and CCT were rated Appropriate, and TEE and stress imaging were rated May Be Appropriate. TTE was rated Appropriate for routine surveillance (3 to 12 months) in patients with known residual VSD, presence of RV-to-PA conduit obstruction, or branch pulmonary artery obstruction. TEE, CMR, CCT, and lung scan were rated May Be Appropriate in these scenarios. In the final scenario of this table (routine

TABLE 18 Truncus Arteriosus

		AUC Score					
Unrepaired		**TTE**		**CMR**		**CCT**	
295.	Evaluation due to change in clinical status and/or new concerning signs or symptoms	A (9)		A (7)		A (7)	
296.	Evaluation prior to planned repair	A (9)		A (7)		A (7)	
Postoperative		**TTE**	**TEE**	**CMR**	**CCT**	**Stress Imaging**	**Lung Scan**
297.	Routine postprocedural evaluation (within 30 days)	A (9)					
298.	Evaluation due to change in clinical status and/or new concerning signs or symptoms	A (9)	M (5)	A (7)	A (7)	M (5)	M (5)
299.	Routine surveillance (1-3 months) within the first year following repair in an asymptomatic patient	A (9)					
300.	Routine surveillance (6-12 months) after the first year following repair in an asymptomatic child or adult with no or mild sequelae	A (9)	R (3)				
301.	Routine surveillance (3-5 years) in an asymptomatic child or adult with no or mild sequelae		M (4)	M (6)	M (5)	M (5)	
302.	Routine surveillance (3-6 months) in an asymptomatic child or adult with ≥moderate truncal stenosis and/or regurgitation	A (9)	M (4)	M (5)	M (5)	M (4)	
303.	Routine surveillance (1-2 years) in an asymptomatic child or adult with ≥moderate truncal stenosis and/or regurgitation		M (6)	A (7)	A (7)	M (5)	
304.	Routine surveillance (3-12 months) in a patient with known residual VSD, presence of an RV-to-PA conduit, or branch pulmonary artery obstruction	A (9)	M (4)	M (6)	M (6)		M (5)
305.	Routine surveillance (3-12 months) in a patient with heart failure symptoms	A (9)		M (6)	M (5)		

A = Appropriate; AUC = Appropriate Use Criteria; CCT = cardiovascular computed tomography; CMR = cardiovascular magnetic resonance; M = May Be Appropriate; R = Rarely Appropriate; RV-to-PA conduit = right ventricle to pulmonary artery conduit; TA = truncus arteriosus; TEE = transesophageal echocardiogram; TTE = transthoracic echocardiogram; VSD = ventricular septal defect.

surveillance of a patient with symptoms of heart failure), TTE was rated Appropriate, and CMR and CCT were rated May Be Appropriate.

Table 19 Considerations

This table addresses the various clinical scenarios encountered in single-ventricle heart disease. Single-ventricle heart disease includes a wide spectrum of underlying cardiac defects such as hypoplastic left heart syndrome, double-inlet LV, double-inlet RV, mitral atresia, tricuspid atresia, unbalanced AVSD, and forms of PA/IVS not amenable to biventricular repair.

Stage 1 palliation includes systemic-to-PA shunt, PDA stent, Norwood procedure with a systemic-PA shunt or an RV-to-PA conduit, PA band, or a hybrid procedure including PA bands and a PDA stent. Stage 2 includes superior cavopulmonary anastomosis (classic or bidirectional Glenn procedure) or Kawashima procedure in those with interrupted inferior vena cava, and stage 3 includes total cavopulmonary anastomosis (Fontan procedure). Palliation consisting of a one and one-half ventricle repair (superior cavopulmonary anastomosis with RV-to-PA connection) is not included, nor are scenarios related to unrepaired single-ventricle disease in older patients. Patients with single-ventricle physiology understandably have a complex clinical course; therefore, only the common scenarios and imaging modalities have been included in this document. Novel interventions and imaging that are currently limited to a few centers, such as that for lymphatics, have not been included. Although the use of lung scan following single-ventricle palliation is controversial, it has been included for rating given its continued used in some centers (49-51). Stress imaging has been included for some scenarios following stage 3 palliation. Depending on the stage of palliation, evaluation due to change in clinical status and/or new concerning signs or symptoms may include increasing cyanosis due to concern for shunt, PDA stent, or cavopulmonary anastomosis stenosis or thrombosis; tight PA band; branch pulmonary artery stenosis; collaterals; restrictive atrial septum; valvular dysfunction; ventricular dysfunction; pulmonary venous obstruction; arch obstruction; and arrhythmias. In addition, following stage 3 palliation, evaluation may be prompted to assess fenestration status, lateral tunnel leaks, protein-losing enteropathy, and plastic bronchitis. This document does not address imaging of other organ systems such as the liver or brain following Fontan palliation.

Table 19 Results and Discussion

TTE was rated Appropriate for all indications at the frequencies listed in the scenarios. TTE + contrast was rated May Be Appropriate for evaluation due to change in status following stage 2 or 3 palliation, and prior to planned stage 3 palliation, especially if there is concern for pulmonary arteriovenous malformations or Fontan baffle leak. TEE has been rated May Be Appropriate prior to planned stage 2 or 3 palliation, for evaluation due to

TABLE 19 Single-Ventricle Heart Disease

	AUC Score				
Unrepaired	**TTE**	**TEE**	**CMR**	**CCT**	**Lung Scan**
306. Routine surveillance (1–4 weeks) in a patient with balanced systemic and pulmonary circulation not requiring neonatal surgery	A (9)				
307. Evaluation due to change in clinical status and/or new concerning signs or symptoms	A (9)	R (3)	M (6)	M (6)	M (4)
308. Evaluation prior to planned surgical palliation	A (9)	R (3)	A (7)	A (7)	M (4)
Postprocedural: Surgical and/or Catheter-Based (Stage 1 Palliation)	**TTE**	**TEE**	**CMR**	**CCT**	**Lung Scan**
309. Routine post-procedural evaluation (within 30 days)	A (9)		R (3)	R (3)	
310. Evaluation due to change in clinical status and/or new concerning signs or symptoms	A (9)	M (4)	A (7)	A (7)	M (5)
311. Routine surveillance (1–4 weeks) in an asymptomatic infant	A (9)				
312. Evaluation prior to planned stage 2 palliation	A (9)	M (4)	A (7)	A (7)	M (4)

Postoperative: Stage 2 Palliation	**TTE**	**TTE + Contrast**	**TEE**	**CMR**	**CCT**	**Lung Scan**
313. Routine postoperative evaluation (within 30 days)	A (9)		R (3)	R (3)	R (3)	
314. Evaluation due to change in clinical status and/or new concerning signs or symptoms	A (9)	M (6)	M (4)	A (8)	A (7)	R (3)
315. Routine surveillance (1–6 months) in an asymptomatic infant or child	A (9)					
316. Routine surveillance (1–2 years) in an asymptomatic adult	A (9)			A (7)	M (6)	
317. Evaluation prior to planned stage 3 palliation	A (9)	M (5)	M (4)	A (8)	A (7)	R (2)

Postoperative: Stage 3 Palliation	**TTE**	**TTE + Contrast**	**TEE**	**CMR**	**CCT**	**Stress Imaging**
318. Routine postoperative evaluation (within 30 days)	A (9)	R (3)	R (3)	R (3)	R (3)	
319. Evaluation due to change in clinical status and/or new concerning signs or symptoms	A (9)	M (6)	A (7)	A (7)	A (7)	M (5)
320. Routine surveillance (3–6 months) within a year following stage 3 palliation in an asymptomatic patient	A (9)					
321. Routine surveillance (6–12 months) after the first year following stage 3 palliation in an asymptomatic patient	A (9)		R (3)	R (3)	R (3)	R (3)
322. Routine surveillance (3–5 years) in an asymptomatic patient	A (9)		M (5)	A (8)	A (7)	M (5)
323. Routine surveillance (3–12 months) in a patient with valvular or ventricular dysfunction, arrhythmias, or other cardiac complications	A (9)			M (6)	M (6)	M (4)
324. Routine surveillance (3–12 months) in a patient with heart failure symptoms	A (9)			M (6)	M (6)	

A = Appropriate; AUC = Appropriate Use Criteria; CCT = cardiovascular computed tomography; CMR = cardiovascular magnetic resonance; M = May Be Appropriate; R = Rarely Appropriate; TEE = transesophageal echocardiogram; TTE = transthoracic echocardiogram.

change in clinical status or new concerning signs or symptoms after any intervention, and for assessment every 3 to 5 years after stage 3 palliation. CMR and CCT were rated Appropriate prior to planned surgical repair at each stage of surgical palliation, for evaluation due to change in status or new symptoms after each stage of palliation, and for routine surveillance every 3 to 5 years after stage 3 palliation. Lung scan was rated May Be Appropriate for evaluation prior to planned repair, due to change in status in the unrepaired patient, or following stage 1 palliation, but it was rated Rarely Appropriate after stage 2 palliation. Stress imaging was considered only after stage 3 palliation and was rated May Be Appropriate for evaluation due to change in status and for routine surveillance every 3 to 5 years in an asymptomatic patient or at 3 to 12 months in those with complications depending on the level of clinical concern.

6. DISCUSSION

This AUC effort was initiated to address the use of cardiac imaging in children and adults with established CHD. Unlike earlier AUC documents that focused on a single imaging modality (12), this document is aligned with the more recent AUC documents (14,15), which include multiple imaging modalities that are part of the armamentarium available to clinicians taking care of these patients. The final ratings for each indication presented in this document reflect the median score of the rating panelists and are based on clinical practice guidelines, when available, or expert opinion. With 324 clinical indications and up to 7 imaging modalities offered per indication, there were 1,035 unique scenarios. Of these scenarios, 44% were rated Appropriate, 39% May Be Appropriate, and 17% Rarely Appropriate.

6.1. Trends and Themes in Scoring

The scenarios in this document are presented on the basis of the sequence of clinical events expected with each CHD, beginning with unrepaired lesions, moving on to surgical and procedural planning, and then addressing follow-up after such interventions. Some broad indications were common to most of the tables in the document. One such indication included evaluation due to change in clinical status and/or new concerning signs or symptoms in the unrepaired and postprocedural section of the table for each CHD. In general, most imaging modalities were rated Appropriate or May Be Appropriate for this indication—likely in an effort to include the entire spectrum of clinical scenarios that could emerge outside of the routine surveillance period. Another such common indication was evaluation prior to planned repair, where TTE was rated Appropriate for all CHD lesions and CMR and CCT were rated Appropriate in those lesions with complex intracardiac anatomy or involvement of the systemic and pulmonary veins, pulmonary arteries, and aortic arch. The general trends that emerged for specific imaging modalities are presented in the following text.

6.1.1. TTE/TEE

TTE was rated as Appropriate in most scenarios involving the evaluation of or routine surveillance for patients with CHD. Scenarios in which it was deemed May Be Appropriate include routine surveillance for an asymptomatic patient with a small ASD or PAPVC involving a single pulmonary vein, MVP and mild MR, mild AS and/or mild AR, or small coronary fistula. It was also rated May Be Appropriate for asymptomatic patients after surgical ASD closure, PAPVC repair, surgical or catheter-based PDA closure, and atrial switch procedure with no or mild sequelae, and for stable adults with ES or postoperative PH. It was rated Rarely Appropriate in routine surveillance for an asymptomatic patient with a PFO, with anomalous right coronary artery from the left aortic sinus, with a small muscular VSD, with a trivial silent PDA, and after the first 2 years following surgical PDA closure with no or mild sequelae.

TTE + contrast was rated May Be Appropriate for the evaluation of a patient before and after repair of an ASD or PAPVC who presents with a change in clinical status and/or new concerning signs or symptoms; of a patient with an isolated secundum ASD to determine the method of closure; of a patient with a sinus venosus defect and/or PAPVC prior to repair; and for routine surveillance after ASD closure or PAPVC repair with significant residual sequelae, ventricular or valvular dysfunction, and/or PH. It was rated Rarely Appropriate in routine surveillance for a patient with an isolated PFO or an asymptomatic patient after ASD device or surgical closure or PAPVC repair with no or mild sequelae. TTE + contrast was also rated Appropriate for the evaluation of a patient before and after Ebstein anomaly or tricuspid valve dysplasia repair who presents with a change in clinical status and/or concerning signs and symptoms and May Be Appropriate for most of the other scenarios in the unrepaired patient and in the routine postprocedural evaluation within 30 days after repair. TTE + contrast was rated May Be Appropriate in the setting of a change in clinical status and/or concerning signs and symptoms for TGA after atrial switch operation and ccTGA after anatomic repair. Last, TTE + contrast was rated May Be Appropriate for the evaluation of a single-ventricle patient after stage 2 and 3 palliation who presents with a change in clinical status and/or new concerning signs or symptoms and a patient after stage 2 palliation in preparation for stage 3 palliation. It was rated Rarely Appropriate in routine postoperative evaluation within 30 days of the single-ventricle patient after stage 3 palliation.

TEE was rated Appropriate or May Be Appropriate in most scenarios prior to planned repair or where a patient presents with a change in clinical status and/or new concerning signs or symptoms before or after surgical repair. Similarly, it was deemed Appropriate or May Be Appropriate in most scenarios involving routine surveillance of adults and some children with moderate or severe disease or sequelae. It was rated Rarely Appropriate in most scenarios involving routine surveillance of patients over a specified time period, particularly in the setting of mild disease or sequelae.

6.1.2. CMR

For CMR, higher ratings were generally found in more complex conditions. For example, routine surveillance by CMR every 3 to 5 years was deemed Appropriate in repaired DORV, D-loop TGA, ccTGA, and single-ventricle heart disease following stage 2 and 3 palliation, and every 2 to 3 years in repaired TOF in asymptomatic patients with no or mild sequalae. Moreover, indications with a change in clinical status and/or new concerning signs or symptoms or preprocedure evaluation were often rated Appropriate. These trends likely reflect CMR's ability to provide a 3-dimensional evaluation of intracardiac and vascular anatomy and to comprehensively assess valve and ventricular function. CMR was also rated highly in scenarios involving adult patients, likely owing to the diminished utility of TTE in larger patients, particularly for assessment of the thoracic vasculature. The key role of CMR in the management of adults with CHD has also been

recognized in published clinical practice guidelines and other clinical policy recommendations (16,52,53). CMR was Appropriate in conditions involving the RV (e.g., TOF, Ebstein anomaly, ES) as its retrosternal position and complex shape may limit evaluation by TTE (54–57). CMR had a lower rating than TTE for many indications involving routine surveillance in asymptomatic infants and children with mild disease. This may be because CMR's higher costs, limited availability, and potential need for sedation did not seem warranted under these circumstances. CMR was rated higher or similar to CCT for all indications. When rated higher than CCT, this was likely because CMR does not incur radiation exposure and has superior blood flow measurement and tissue characterization capabilities. Evaluation of the pulmonary-to-systemic blood flow ratio (58), valve regurgitation, and myocardial fibrosis by CMR may have a significant impact on patient management (59–63).

6.1.3. CCT

Higher ratings for CCT were generally found in more complex conditions and when definitive coronary imaging or complex vascular anatomy assessment is needed. CCT was rated high for indications related to complex CHD lesions, in adult patients, for preprocedural evaluation, and for evaluation due to change in clinical status. CCT received lower ratings for routine surveillance at relatively short intervals in asymptomatic patients with stable cardiac status or in patients with mild disease or minimal hemodynamic sequelae after intervention. CCT and CMR were rated equivalent for many indications, but CCT received lower ratings for serial evaluation and when definitive coronary imaging is not required. For example, CCT is rated more highly after arterial switch or for new symptoms when there has been surgical coronary manipulation than for lesions where only coronary course and relationship to the cardiac and thoracic structures need to be assessed. The main limitation for CCT remains its use of ionizing radiation, which is an important consideration for all CHD patients requiring serial diagnostic imaging over a lifetime. Newer-generation CCT scanners offer submillimeter, isotropic spatial resolution with improved temporal resolution, rapid image acquisition, and significantly lower radiation doses than does older technology (64). These advances lower the anesthesia and radiation risk and improve the diagnostic utility of CCT in CHD patients. For indications for which CCT was rated Rarely Appropriate, CCT may still be a reasonable alternative for specific patients if CMR is considered Appropriate but unlikely to be diagnostic (metallic artifact) or is contraindicated (pacemaker-dependent patient) when equivalent information can be obtained. CMR and CCT have been shown to be equivalent for quantification of ventricular function if a scanner with appropriate temporal resolution is used (65,66).

6.1.4. Stress Imaging

Stress imaging is typically used for the detection of coronary artery disease, assessment of the area of myocardium at risk in known coronary disease, and risk stratification after coronary revascularization. Stress imaging can also be used to evaluate changes in cardiac hemodynamics, including valve and subvalvular gradients (AS, subaortic stenosis, MS, PS, and prosthetic valves), aortic coarctation, and RV pressures. The use of stress imaging was rated Appropriate and May Be Appropriate in most of the clinical scenarios involving coronary anomalies and transposition of the great arteries following an arterial switch operation. Clinical practice guidelines have supported the role of stress imaging for detection of ischemia in these patients (1,16). Stress testing for evaluation due to change in clinical status and/or new concerning signs or symptoms was rated May Be Appropriate in all the scenarios. The use of stress testing for routine surveillance was rated May Be Appropriate in most scenarios involving patients who were asymptomatic with $\geq$ moderate residual sequelae except for those with $>$ moderate subvalvular AS and/or $<$ mild AR, when it was rated Rarely Appropriate. Stress imaging was rated May Be Appropriate in routine surveillance (2 to 3 years and 3 to 5 years) in an asymptomatic patient with no or mild sequelae in scenarios with DORV, D-Loop TGA, ccTGA, TA, and single-ventricle heart disease after stage 3 palliation. Stress imaging was generally rated Rarely Appropriate in routine surveillance (1 to 2 years) in an asymptomatic child or adult with no or mild residual sequelae.

6.1.5. Lung Scan

A lung perfusion scan is a noninvasive nuclear imaging study used in children and adults with congenital and acquired pulmonary vascular abnormalities to determine the relative regional pulmonary perfusion. These studies are used to determine when PA intervention should be performed. Intervention on the pulmonary arteries is indicated when the relative flow discrepancy between the 2 lungs is 35%/65% or worse (67). Due to this reason, lung perfusion scanning has been rated May Be Appropriate in patients following repair or palliation with single-ventricle heart disease, TA, TGA, DORV, TOF, PA/IVS, and PDA, when evaluation is needed owing to change in clinical status and/or new concerning signs or symptoms or when branch pulmonary artery stenosis is suspected or

known. It is rated Appropriate in postoperative TAPVC patients with a change in clinical status and/or concerning signs or symptoms and in patients with PDA post-procedure with known left PA stenosis.

6.1.6. Fluoroscopy

When mechanical valve leaflet dysfunction is suspected on echocardiogram, fluoroscopy may be used to evaluate leaflet motion. Nevertheless, it cannot assess the underlying etiology of the valve obstruction, such as differentiating between thrombosis and pannus (68,69). Fluoroscopy was rated Appropriate when a patient with a mechanical mitral or aortic valve has a change in clinical status and/or new concerning signs or symptoms. Fluoroscopy is also used to evaluate for stent fracture and was rated May Be Appropriate when there is a change in clinical status, there are new concerning signs or symptoms, or for routine surveillance in aortic stent and transcatheter pulmonary valve patients.

6.1.7. Multimodality Imaging in Adults With CHD

In light of the growing number of adults with CHD, it is important to recognize specific indications unique to this population. For adults with CHD of simple complexity such as a small ASD, PAPVC involving a single pulmonary vein, a small VSD, and a small PDA, TTE was rated Appropriate during routine surveillance every 3 to 5 years. TTE was rated Appropriate for routine surveillance in asymptomatic adults with no or mild sequelae following ASD closure, PAPVC repair, VSD closure, AVSD repair, and TAPVC. As lesion complexity increased, additional imaging modalities received higher appropriateness ratings. In the asymptomatic adult with D-loop TGA after arterial switch operation, Rastelli, or atrial switch operation; ccTGA after anatomic or physiological repair; and single ventricle after stage 3 palliation with no or mild sequelae, CMR and CCT were rated Appropriate for routine postoperative surveillance at 3 to 5 years. In the asymptomatic adult with repaired TOF with no or mild sequalae, CMR was rated Appropriate every 2 to 3 years. In the adult with CHD and clinical heart failure, periodic imaging with TTE, CMR, and CCT were often rated Appropriate for evaluating valvular and ventricular function.

6.2. Use of AUC to Improve Care

This AUC document has been designed to provide guidance for utilizing various imaging modalities to clinicians caring for patients with CHD. While the newer 2018 AHA/ACC Guidelines for the Management of Adults With Congenital Heart Disease were primarily used for this document, the 2008 Guidelines for the Management of Adults with Congenital Heart Disease were also referred to if they provided specific follow-up intervals for imaging (1,16). Currently, there is a dearth of clinical practice guidelines related to cardiac imaging in CHD. In the few that exist, there is a lack of guidance on frequency of imaging in the broad array of clinical scenarios faced by clinicians in their routine practice (1,10,11,16). This AUC document addresses this deficiency by including clinical scenarios from the time of diagnosis to after interventions and by providing a multimodality perspective. With the significant advances in imaging technology and accessibility, clinicians will need to remain mindful of resource utilization and the risks versus benefits when selecting a diagnostic pathway. This AUC document will serve not only as a clinical guide, but also as a tool to improve quality of care and reduce practice variation across physicians and centers.

As with the prior AUC documents, it is important to emphasize that an Appropriate rating does not mean that a given procedure has to be performed; nor does a Rarely Appropriate rating mean a procedure should never be performed in a patient who fits the scenario(s) listed in this document (12,15). Rather, a procedure with an Appropriate rating should be seen as an option that would be reasonable to perform if the information obtained may be useful in managing the patient. Similarly, for a Rarely Appropriate rating, there may be additional clinical circumstances that dictate the need for testing. It is recommended that such circumstances be documented clearly by the ordering clinician. It is also important to recognize that the categories of May Be Appropriate and Rarely Appropriate should not be considered as grounds for denial of insurance coverage or reimbursement for a particular imaging study, as clinician judgment is essential for determining what individual imaging test is best for a specific patient. This document can be used to identify areas of excessive use of cardiac imaging for indications rated Rarely Appropriate. Similar to previous efforts using the 2014 AUC for Initial Transthoracic Echocardiography in Outpatient Pediatric Cardiology, this document can be used for educational intervention to improve appropriateness of cardiac imaging during follow-up care of patients with CHD (70-72).

7. CONCLUSIONS

This AUC report serves as a guide to physicians taking care of children and adults with CHD. Of the broad array of clinical scenarios covered in this document, 44% were rated Appropriate, 39% May Be Appropriate, and 17% Rarely Appropriate. These ratings are based on currently available technology, resources, and evidence-based medicine, which may change over time and dictate future revisions of this document. Implementation

studies using this document may help improve appropriate use and reduce practice variation for noninvasive cardiac imaging modalities utilized during the follow-up care of patients with CHD.

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KEY WORDS ACC Appropriate Use Criteria, congenital heart disease, multimodality imaging

APPENDIX A. RELATIONSHIPS WITH INDUSTRY (RWI) AND OTHER ENTITIES

Appropriate Use Criteria for Multimodality Imaging During the Follow-Up Care of Patients With Congenital Heart Disease: Members of the Writing Group, Rating Panel, External Reviewers, and Solution Set Oversight Committee—Relationships With Industry and Other Entities (Relevant)

The ACC and the Solution Set Oversight Committee continue to focus considerable attention on avoiding real or perceived relationships with industry (RWI) and other entities that might affect the rating of a test/procedure. The ACC maintains a database that tracks all relevant relationships for ACC members and persons who participate in ACC activities, including the development of AUC. A table of relevant disclosures by the writing group, rating panel, external reviewers, and Solution Set Oversight Committee can be found below. In addition, to ensure complete transparency, a full list of disclosure information—including relationships not pertinent to this document—is available as an Online Appendix.

A more specific RWI policy applies to the Writing Group and Rating Panel of AUC documents:

- Writing Group: AUC Writing Groups must be chaired (or co-chaired) by a person with no relevant RWI. Although Writing Group members play an important role in the development of the final published document for a given set of AUC, they do not have any role in the AUC rating process and therefore have limited impact on how the documents will guide clinical care. Accordingly, RWI restrictions are not applied to Writing Group members, other than the Chair (or Co-Chair).
- Rating Panel: To avoid the potential for bias in the actual indication rating, fewer than 50% of Rating Panel members may have relevant RWI. AUC documents utilize a modified Delphi method as outlined in the RAND Appropriateness Criteria Method paper and the ACC AUC Methodology paper. This method utilizes a 2-step process: Step 1) writing committee members develop a list of typical clinical scenarios/indications; Step 2) technical panel members review and rate the individual clinical scenarios. The RAND Delphi method allows for the contribution of a wide range of viewpoints while minimizing and controlling bias through an independent rating panel, a review of score dispersion, use of a median score as the final rating, and a highly structured process for determining recommendations. As such, all rating panel members, even those with RWI, are allowed to rate as part of the technical panel modified Delphi process.

APPENDIX A. CONTINUED

Participant	Employment	Representing	Consultant	Speakers Bureau	Ownership/ Partnership/ Principal	Personal Research	Institutional, Organizational, or Other Financial Benefit	Expert Witness
			Writing Group					
Ritu Sachdeva, MBBS *(Co-Chair)*	Sibley Heart Center Cardiology, Children's Healthcare of Atlanta, Cardiovascular Imaging Research Core—Medical Director; Emory University School of Medicine, Division of Pediatric Cardiology—Professor of Pediatrics	ACC	None	None	None	None	None	None
Anne Marie Valente, MD *(Co-Chair)*	Boston Children's Hospital and Brigham & Women's Hospital, Boston Adult Congenital Heart Disease Program—Section Chief; Harvard Medical School—Associate Professor of Medicine & Pediatrics	SOPE	None	None	None	None	■ Higgins Family Noninvasive Research Fund ■ Dunlevie Foundation ■ Lerner Research Award ■ Sarah Marie Liamos Fund	None
Aimee K. Armstrong, MD	Nationwide Children's Hospital, The Heart Center, Cardiac Catheterization & Interventional Therapies—Director; Ohio State University College of Medicine—Professor of Pediatrics	SCAI	■ Abbott* ■ B. Braun Interventional Systems ■ Edwards Lifesciences* ■ Medtronic*	None	None	■ Abbott† ■ Edwards Lifesciences† ■ Medtronic† ■ PFM Medical† ■ Siemens Medical Solutions USA†	None	None
Stephen C. Cook, MD	Spectrum Health Helen DeVos Children's Hospital, Adult Congenital Heart Disease Center—Director; Michigan State University College of Human Medicine, Pediatrics and Human Development—Clinical Associate Professor	ACC	None	None	None	None	None	None
B. Kelly Han, MD	Minneapolis Heart Institute and Children's Minnesota, Advanced Congenital Cardiac Imaging—Director	SCCT	■ Edwards Lifesciences	None	None	■ Minneapolis Heart Institute Foundation*	None	None
Leo Lopez, MD	Lucile Packard Children's Hospital Stanford—Medical Director of Echocardiography; Stanford University School of Medicine—Clinical Professor of Pediatrics	ASE	None	None	None	None	None	None
George K. Lui, MD	Lucile Packard Children's Hospital and Stanford Health Care Collaboration, The Adult Congenital Heart Program at Stanford—Medical Director; Stanford University School of Medicine, Division of Cardiovascular Medicine & Pediatric Cardiology—Clinical Associate Professor of Medicine & Pediatrics	ASE	None	None	None	None	None	None
Sarah S. Pickard, MD	Boston Children's Hospital, Department of Cardiology; Harvard Medical School—Instructor of Pediatrics	ACC	None	None	None	None	None	None

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APPENDIX A. CONTINUED

Participant	Employment	Representing	Consultant	Speakers Bureau	Ownership/ Partnership/ Principal	Personal Research	Institutional, Organizational, or Other Financial Benefit	Expert Witness
Andrew J. Powell, MD	Boston Children's Hospital, Department of Cardiology, Cardiac Imaging Division—Chief; Harvard Medical School—Professor of Pediatrics	SCMR	None	None	None	None	None	None
			Rating Panel					
Nicole M. Bhave, MD, *Moderator*	University of Michigan and VA Ann Arbor Healthcare System, Division of Cardiovascular Medicine, Internal Medicine—Assistant Professor	ACC	None	None	None	■ Medtronic	None	None
Jeanne M. Baffa, MD	Nemours Alfred I. duPont Hospital for Children, The Nemours Cardiac Center, Division of Pediatric Cardiology—Director of Echocardiography, Pediatric Cardiology Fellowship Program Director, and Emeritus Professor	ACC	■ Merck Manual	None	None	None	None	■ Third party, Prozac and congenital heart disease, 2011
Puja Banka, MD	Boston Children's Hospital—Director of Cardiac CT; Harvard Medical School—Associate Professor of Pediatrics; Merck Research Laboratories, Cardiovascular Disease, Clinical Research—Director	SCCT	■ Zogenix*	None	None	None	None	None
Scott B. Cohen, MD	Medical College of Wisconsin, Departments of Internal Medicine and Pediatrics, Divisions of Adult Cardiovascular Medicine and Pediatric Cardiology—Associate Professor	ACC	None	None	None	None	None	None
Julie S. Glickstein, MD	Columbia University Irving Medical Center, Department of Pediatrics, Division of Pediatric Cardiology—Professor; Morgan Stanley Children's Hospital of New York Presbyterian, Pediatric Cardiology Fellowship Program—Director	AAP	None	None	None	None	None	None
Joshua P. Kanter, MD	Children's National Health System, Division of Pediatric Cardiology—Director of Interventional Cardiology; George Washington University School of Medicine—Associate Professor of Pediatrics	ACC	None	None	None	None	None	None
Ronald J. Kanter, MD	Nicklaus Children's Hospital, Division of Cardiology, Electrophysiology—Director	ACC	None	None	None	None	None	None
Yuli Y. Kim, MD	Penn Medicine and the Children's Hospital of Philadelphia, Philadelphia Adult Congenital Heart Center—Medical Director and Assistant Professor of Medicine	ACC	None	None	None	None	None	None
Alaina K. Kipps, MD	Lucile Packard Children's Hospital at Stanford, Acute Care Pediatric Cardiology—Medical Director; Stanford School of Medicine, Division of Pediatric Cardiology—Clinical Associate Professor of Pediatrics	ACC	None	None	None	None	None	None
Larry A. Latson, MD	Joe DiMaggio Children's Hospital and Memorial Healthcare System, Congenital Cardiac Catheterization—Director, and Adult Congenital Heart Disease—Co-Director	SCAI	None	None	None	None	None	None
Jeannette P. Lin, MD	UCLA Medical Center—Associate Clinical Professor of Medicine	ACC	None	None	None	None	None	None

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APPENDIX A. CONTINUED

Participant	Employment	Representing	Consultant	Speakers Bureau	Ownership/ Partnership/ Principal	Personal Research	Institutional, Organizational, or Other Financial Benefit	Expert Witness
David A. Parra, MD	Monroe Carell Jr. Children's Hospital at Vanderbilt, Division of Pediatric Cardiology—Director of Non-Invasive Imaging; Vanderbilt University—Associate Professor of Pediatrics	SCMR	None	None	None	None	None	None
Fred H. Rodriguez III, MD	Sibley Heart Center Cardiology and Emory University School of Medicine, Division of Cardiology—Assistant Professor of Pediatrics and Internal Medicine	ACC	None	None	None	None	None	None
Elizabeth V. Saarel, MD	Cleveland Clinic, Pediatric Cardiology—Chair; Cleveland Clinic Lerner College of Medicine at Case Western Reserve University—Professor	AHA	None	None	None	None	None	None
Shubhika Srivastava, MBBS	Mount Sinai Medical Center, Children's Heart Center, Advanced Imaging Fellowship and Non-Invasive Imaging—Director; Icahn School of Medicine—Professor of Pediatrics	ASE	None	None	None	None	None	None
Elizabeth A. Stephenson, MD	The Hospital for Sick Children, Labatt Family Heart Centre, Electrophysiology—Section Head; University of Toronto—Professor of Paediatrics	HRS	None	None	None	None	None	None
Karen K. Stout, MD	University of Washington Medical Center, Cardiology Clinics—Associate Chief of Cardiology; University of Washington—Professor of Cardiology and Adjunct Professor of Pediatric Cardiology	ACC	None	None	None	None	None	None
Ali N. Zaidi, MD	Mount Sinai Cardiovascular Institute, Adult Congenital Heart Disease Center—Director; Mount Sinai Kravis Children's Hospital, Children's Heart Center—Director; Icahn School of Medicine, Internal Medicine and Pediatrics—Associate Professor	ACC	None	None	None	None	None	None
			External Reviewers					
Naser M. Ammash, MD	Mayo Clinic, Department of Cardiovascular Medicine—Professor of Medicine	ISACHD	None	None	None	None	■ ABIM (test writing committee) ■ UpToDate (writing chapters)	None
Luc M. Beauchesne, MD	University of Ottawa Heart Institute, Division of Cardiology, Adult Congenital Heart Disease Program—Director	ISACHD	None	None	None	None	None	None
Craig S. Broberg, MD	Oregon Health and Science University, Knight Cardiovascular Institute, Adult Congenital Heart Program—Director	ACC	None	None	None	■ Actelion† ■ American Heart Association* ■ NHLBI†	None	None
David W. Brown, MD	Boston Children's Hospital—Senior Associate in Cardiology; Harvard Medical School—Associate Professor in Pediatrics	ACC	None	None	None	None	None	None

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Participant	Employment	Representing	Consultant	Speakers Bureau	Ownership/ Partnership/ Principal	Personal Research	Institutional, Organizational, or Other Financial Benefit	Expert Witness
Luke J. Burchill, MD	University of Melbourne, Department of Medicine, Royal Melbourne Hospital Adult Congenital Heart Disease Program—Associate Professor	AHA	None	None	None	None	None	None
Joseph A. Camarda, MD	Lurie Children's Hospital of Chicago, Pediatric Cardiology—Clinical Practice Director; Northwestern University Feinberg School of Medicine—Assistant Professor	ACC	None	None	None	None	None	None
M. Jay Campbell, MD	Duke University—Associate Professor of Pediatrics	SCMR	None	None	None	None	None	None
Robert M. Campbell, MD	Sibley Heart Center Cardiology—Pediatric Cardiologist; Emory University School of Medicine—Professor of Pediatrics	ACC	None	None	None	None	None	None
Frank A. Cetta, Jr., MD	Mayo Clinic, Division of Pediatric Cardiology and Department of Cardiovascular Diseases—Professor of Pediatrics and Medicine	ACC	None	None	None	None	None	None
Curtis J. Daniels, MD	Nationwide Children's Hospital and The Ohio State University, Adult Congenital Heart Disease Program—Director	ACC	None	None	None	None	■ ABIM (ACHD subspecialty exam committee)	None
Nancy A. Drucker, MD	The University of Vermont Children's Hospital, Larner College of Medicine, Division of Pediatric Cardiology—Associate Professor of Pediatrics	ACC	None	None	None	None	None	None
Brian M. Fonseca, MD	Children's Hospital Colorado, The Heart Institute—Director of Cardiac MRI and Clinical Informatics; University of Colorado, Pediatric Cardiology—Associate Professor	SCMR	None	None	None	None	None	None
Daniel E. Forsha, MD	Ward Family Heart Center, Children's Mercy Kansas City—Assistant Professor	ASE	None	None	None	None	None	None
Ruchira F. Garg, MD	Cedars-Sinai Medical Center, Congenital Noninvasive Imaging—Director; Guerin Family Congenital Heart Program—Associate Director; Professor of Pediatrics	SCMR	None	None	None	None	None	None
Salil Ginde, MD	Children's Hospital of Wisconsin, Adult Congenital Heart Disease and Pediatric Cardiology; Medical College of Wisconsin, Departments of Medicine and Pediatrics—Associate Professor	ACC	None	None	None	None	None	None
Marye J. Gleva, MD	Washington University in St. Louis School of Medicine—Professor of Medicine	HRS	None	None	None	None	None	None
Michelle Z. Gurvitz, MD	Boston Children's Hospital—Attending Physician; Harvard Medical School—Assistant Professor of Pediatrics	ACC	None	None	None	■ National Institutes of Health*	■ Adult Congenital Heart Association† ■ Pediatric Congenital Heart Association†	None

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APPENDIX A. CONTINUED

Participant	Employment	Representing	Consultant	Speakers Bureau	Ownership/ Partnership/ Principal	Personal Research	Institutional, Organizational, or Other Financial Benefit	Expert Witness
Anthony M. Hlavacek, MD	Medical University of South Carolina, Pediatric Cardiology Inpatient Unit—Medical Director, and Professor of Pediatrics and Radiology	ACC	None	None	None	None	None	■ Plaintiff, cardiac tamponade, 2015 ■ Defendant, total anomalous pulmonary venous drainage, 2015 ■ Defendant, double outlet right ventricle, 2015
Daphne T. Hsu, MD	Children's Hospital at Montefiore, Pediatric Heart Center—Division Chief and Co-Director; Albert Einstein College of Medicine—Professor of Pediatrics	ACC	None	None	None	■ National Institutes of Health	None	None
Eric V. Krieger, MD	University of Washington Medical Center and Seattle Children's Hospital, Seattle Adult Congenital Heart Service—Director; University of Washington School of Medicine, Division of Cardiology—Associate Professor of Medicine	ISACHD	None	None	None	None	■ Canadian Heart Research Centre/ Actelion‡	None
Emy Kuriakose, MD	Spectrum Health Helen DeVos Children's Hospital, Congenital Cardiac Imaging—Section Chief	ACC	None	None	None	None	None	None
Sean M. Lang, MD	Cincinnati Children's Hospital Medical Center, Heart Institute—Medical Director of Coronary Artery Clinic, Associate Director of Cardiac MRI, and Assistant Professor of Pediatrics	ACC	None	None	None	None	None	None
Jimmy C. Lu, MD	University of Michigan Congenital Heart Center, Pediatric Cardiology—Associate Professor	ASE	None	None	None	None	None	None
Francois Marcotte, MD	Mayo Clinic Arizona, Adult Congenital Diseases Clinic—Director, and Senior Associate Consultant in Cardiovascular Diseases	AHA	None	None	None	None	None	None
Shiraz A. Maskatia, MD	Lucile Packard Children's Hospital, Fetal Cardiology—Associate Director; Stanford University School of Medicine, Division of Cardiology, Department of Pediatrics—Associate Professor	ACC	None	None	None	None	None	■ Plaintiff, infant mortality due to CHD, 2017
Laura M. Mercer-Rosa, MD	Children's Hospital of Philadelphia, University of Pennsylvania, Perelman School of Medicine—Assistant Professor of Pediatrics	ASE	None	None	None	■ National Institutes of Health* ■ Pulmonary Hypertension Association*	None	None
Mark D. Norris, MD	University of Michigan, Congenital Heart Center, Adult Congenital Heart Program—Co-Director, and Assistant Professor	ACC	None	None	None	None	None	None

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APPENDIX A. CONTINUED

Participant	Employment	Representing	Consultant	Speakers Bureau	Ownership/ Partnership/ Principal	Personal Research	Institutional, Organizational, or Other Financial Benefit	Expert Witness
Anitha Parthiban, MD	Children's Mercy Hospital, The Ward Family Heart Center, and University of Missouri School of Medicine at Kansas City—Associate Professor of Pediatrics	ACC	None	None	None	None	None	None
Sara L. Partington, MD	Hospital of the University of Pennsylvania—Assistant Professor of Clinical Medicine	ACC	None	None	None	None	None	None
Marie-France Poulin, MD	Beth Israel Deaconess Medical Center and Harvard Medical School, Interventional and Structural Cardiology, Structural Heart Clinical Services—Associate Director	SCAI	None	None	None	None	None	None
Saurabh Rajpal, MBBS, MD	Nationwide Children's Hospital, The Heart Center, Adult Congenital Heart Disease and Pulmonary Hypertension Program; The Ohio State University, Internal Medicine—Assistant Professor	ACC	None	None	None	None	None	None
Cynthia K. Rigsby, MD	Lurie Children's Hospital of Chicago, Department of Medical Imaging—Vice Chair for Research; Body Imaging—Division Head; Northwestern University Feinberg School of Medicine—Professor of Radiology and Pediatrics	ACC	None	None	None	None	None	None
Margaret M. Samyn, MD	Herma Heart Institute at the Children's Hospital of Wisconsin—Pediatric Cardiologist; Medical College of Wisconsin—Professor of Pediatric Cardiology	SCMR	None	None	None	None	■ Society for Cardiac Magnetic Resonance—Pediatric and Congenital Heart Disease Interest Group representative†	None
Michael D. Seckeler, MD	Banner University Medical Center in Tucson, Pediatric and Adult Congenital Cardiac Catheterization Laboratory—Director; University of Arizona—Associate Professor of Pediatrics (Cardiology)	ACC	None	None	None	None	None	None
Maully J. Shah, MBBS	The Children's Hospital of Philadelphia, Cardiac Electrophysiology—Director; University of Pennsylvania, Perelman School of Medicine—Professor of Pediatrics	HRS	None	None	None	None	■ Wiley U.K. (book royalty)†	■ Plaintiff, Paxil causing CHD, 2009*
Timothy C. Slesnick, MD	Sibley Heart Center Cardiology, Children's Healthcare of Atlanta, Emory University School of Medicine, Division of Pediatric Cardiology—Associate Professor of Pediatrics	ACC	None	None	None	None	None	None
Brian D. Soriano, MD	Seattle Children's Hospital and University of Washington School of Medicine—Associate Professor of Pediatrics and Adjunct Associate Professor of Radiology	ACC	None	None	None	None	None	■ Defendant, missed diagnosis, 2014

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Participant	Employment	Representing	Consultant	Speakers Bureau	Ownership/ Partnership/ Principal	Personal Research	Institutional, Organizational, or Other Financial Benefit	Expert Witness
Christopher J. Statile, MD	Cincinnati Children's Hospital Medical Center, The Heart Institute—Medical Director of Outpatient Services, and Associate Professor of Pediatrics	ACC	None	None	None	None	None	None
Corey A. Stiver, MD	Nationwide Children's Hospital—Pediatric Cardiologist; The Ohio State University College of Medicine—Assistant Professor of Pediatrics	ACC	None	None	None	None	None	None
Carolyn L. Taylor, MD	MUSC Pediatric Cardiology, Pediatric Echo Laboratory—Director, and Associate Professor of Pediatrics	ASE	None	None	None	None	None	■ Defendant, patent ductus arteriosus litigation, 2016*
Seda S. Tierney, MD	Stanford University, Noninvasive Imaging Laboratory—Director of Research, Director of Vascular Laboratory, and Associate Professor of Pediatrics	ACC	None	None	None	None	None	None
Solution Set Oversight Committee								
RWI and disclosure statements for members of the SSOC can be found here: https://www.acc.org/guidelines/about-guidelines-and-clinical-documents/guidelines-and-documents-task-forces.								

This table represents *relevant* relationships of participants with industry and other entities that were reported at the time this document was under development. The table does not necessarily reflect relationships with industry at the time of publication. A person has a *relevant* relationship IF: the relationship or interest relates to the same or similar subject matter, intellectual property or asset, topic, or issue addressed in the document; the company/entity (with whom the relationship exists) makes a drug, drug class, or device addressed in the document or makes a competing drug or device addressed in the document; or the person or a member of the person's household has a reasonable potential for financial, professional, or other personal gain or loss as a result of the issues/content addressed in the document.

A person is deemed to have a *significant* interest in a business if the interest represents ownership of ≥5% of the voting stock or share of the business entity, or ownership of ≥$5,000 of the fair market value of the business entity; or if funds received by the person from the business entity exceed 5% of the person's gross income for the previous year. Relationships in this table with no symbol are considered *modest* (less than significant under the preceding definition). Relationships that exist with *no financial benefit* are also included for the purpose of transparency. Please refer to http://www.acc.org/guidelines/about-guidelines-and-clinical-documents/relationships-with-industry-policy for definitions of disclosure categories or additional information about the ACC Disclosure Policy for Writing Committees.

*Significant relationship.

†No financial benefit.

‡Clinical trial enroller.

AAP = American Academy of Pediatrics; ABIM = American Board of Internal Medicine; ACC = American College of Cardiology; ACHD = Adult Congenital Heart Disease; AHA = American Heart Association; ASE = American Society of Echocardiography; AUC = Appropriate Use Criteria; CHD = Congenital Heart Disease; HRS = Heart Rhythm Society; ISACHD = International Society for Adult Congenital Heart Disease; NHLBI = National Heart, Lung, and Blood Institute; SCAI = Society for Cardiovascular Angiography and Interventions; SCCT = Society of Cardiovascular Computed Tomography; SCMR = Society for Cardiovascular Magnetic Resonance; SOPE = Society of Pediatric Echocardiography; SSOC = Solution Set Oversight Committee.