

**American College of Radiology
ACR Appropriateness Criteria®
Known or Suspected Congenital Heart Disease in the Adult**

Variant 1: Known or suspected congenital heart disease in the adult.

Radiologic Procedure	Rating	Comments	RRL*
X-ray chest	9	This is usually the first imaging procedure performed.	☼
US echocardiography transthoracic resting	9	This is usually one of the first procedures performed.	○
MRI heart function and morphology without and with IV contrast	9	This procedure is complementary to the transthoracic echocardiogram. It may be an alternative to the echocardiogram in patients with a poor acoustic window.	○
MRI heart function and morphology without IV contrast	8	This procedure is complementary to the transthoracic echocardiogram and may be performed as an alternative to MRI heart function without and with IV contrast. It may be an alternative to the echocardiogram in patients with a poor acoustic window.	○
US echocardiography transesophageal	7	This procedure is complementary to the transthoracic echocardiogram.	○
CT heart function and morphology with IV contrast	7	This procedure is complementary to the transthoracic echocardiogram and may be performed as an alternative to the MRI heart function and morphology examination. This procedure provides information about ventricular function and cardiac anatomy but, unlike MRI, does not provide information about flow.	☼☼☼☼
CTA coronary arteries with IV contrast	7	This procedure is complementary to the transthoracic echocardiogram and may be performed as an alternative to either CT heart function and morphology with IV contrast or MRI heart function and morphology, especially if coronary artery anatomy is in question. This study can be performed using techniques that provide either anatomy alone or ventricular function with anatomy. It does not provide information about flow.	☼☼☼
MRA chest without and with IV contrast	7	This procedure is complementary to the transthoracic echocardiogram and may be performed as an alternative to MRI heart function and morphology if only great-vessel anatomical information is needed and no information is needed about intracardiac anatomy, heart function, and flow. Occasionally, it may be complementary to MRI heart function and morphology without IV contrast.	○

CTA chest with IV contrast	6	This procedure is complementary to the transthoracic echocardiogram and may be performed as an alternative to CTA heart function and morphology if only great anatomical information is needed and no detailed information is needed about intracardiac anatomy, heart function, and flow.	☢☢☢
MRA chest without IV contrast	6	This procedure is complementary to the transthoracic echocardiogram and may be performed as an alternative to MRA chest without and with IV contrast or CTA chest with IV contrast.	○
Arteriography coronary with ventriculography	6	This procedure is complementary to the transthoracic echocardiogram, transesophageal echocardiogram, MRI heart function and morphology, CT heart function with morphology, CTA chest, and MRA chest. It may be an alternative to CTA coronary arteries.	☢☢☢
Nuclear medicine ventriculography	4		☢☢☢
Rating Scale: 1,2,3 Usually not appropriate; 4,5,6 May be appropriate; 7,8,9 Usually appropriate			*Relative Radiation Level

KNOWN OR SUSPECTED CONGENITAL HEART DISEASE IN THE ADULT

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Summary of Literature Review

Introduction/Background

Congenital heart disease (CHD) has been estimated to occur in approximately 0.4% to 1% of live births [1], with increasing incidence of CHD in the adult patient population [2] in part as a result of better patient survival. In particular, patients with more severe CHD (tetralogy of Fallot, truncus arteriosus, transposition complexes, endocardial cushion defects, and univentricular hearts) are living longer [3]. It is estimated that 1 to 1.3 million of all CHD patients in the United States are adults [3,4]. In addition, over 85% of infants with CHD are expected to reach adulthood [5]. As a result, annual hospital admissions of adult patients with CHD now account for 37% of all CHD hospital admissions [2], with more than half of adult patients with CHD having undergone surgery. Nearly all adults with known CHD require periodic imaging as a means of monitoring their disease process. All adult patients with suspected CHD require imaging as a means of definitive diagnosis.

Congenital heart lesions may become symptomatic at any time from birth until adulthood. Several common congenital heart defects often survive into adulthood. These include bicuspid aortic valve, congenital forms of mitral valve prolapse, aortic coarctation, atrial septal defect (ASD), pulmonary valve stenosis, patent ductus arteriosus, and tetralogy of Fallot [6]. Uncommon congenital cardiac defects that may present in adulthood include Ebstein anomaly, corrected transposition of the great vessels, pulmonary arteriovenous malformation, coronary artery anomalies, and sinus of Valsalva aneurysm [7,8]. Approximately 10% of patients with a common CHD survive undetected until adulthood [9]. The most common congenital heart defect in children, ventricular septal defect, may escape detection and present in adults either as a small, physiologically insignificant defect or as a large defect with Eisenmenger physiology. Anomalies of the great arteries such as complete transposition and total anomalous pulmonary venous drainage are usually symptomatic, whereas less significant anomalies such as a persistent left superior vena cava and many variants of the origin of the great vessels from the arch are often asymptomatic.

Imaging procedures for the diagnosis of known or suspected CHD in the adult include chest radiography, fluoroscopy, echocardiography (transthoracic and transesophageal), nuclear scintigraphy, cardiac-gated computed tomography (CT), magnetic resonance imaging (MRI), and cardiac catheterization and angiography. The physician trying to diagnose these often-complex conditions needs complete and reliable information that includes details about intracardiac anatomy, vascular anatomy, hemodynamics, and function. Adults with CHD also have acquired comorbid factors related to aging, such as hypertension, atherosclerosis, coronary artery occlusive disease, pulmonary disease, and renal disease, which may complicate their medical and/or surgical management. When comorbid factors are present and in some congenital conditions (eg, anomalous coronary artery, univentricular heart), the capability of the myocardium to meet metabolic and physiologic needs may be uncertain. In these instances, reference should be made to the ACR Appropriateness Criteria[®] related to the evaluation of myocardial ischemia: [Chest Pain-Possible Acute Coronary Syndrome](#) [10] and [Chronic Chest Pain—High Probability of Coronary Artery Disease](#) [11].

Chest radiography

The initial workup of adults with known or suspected CHD usually includes posterior anterior and lateral chest radiographs. Occasionally radiography will be the first study to alert the radiologist and the clinician to the

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possibility of a congenital cardiac defect or great-vessel anomaly. This simple and inexpensive examination remains a first-line test for patients with suspected CHD.

The chest radiograph quickly illustrates gross cardiac and mediastinal contours, pulmonary vascularity, pathologic calcification, and the presence of certain indwelling metallic devices. It also provides an assessment of cardiac size, cardiac configuration, and position of the aortic arch. The situs of the abdomen and thorax can usually be determined. Thoracic cage anomalies associated with CHD and postoperative changes may also be detected. The chest radiograph continues to be a useful tool for the initial assessment of the patient with surgically treated CHD. Nevertheless, the chest radiograph alone is insufficient as a means of definitive and full diagnosis of CHD [12].

Transthoracic echocardiography

Transthoracic echocardiography (TTE) remains a first-line imaging examination in adults with known or suspected CHD. This test has long been established as a clinically useful diagnostic modality for CHD in children, often eliminating the need for cardiac catheterization in uncomplicated lesions. Although adults present certain technical problems related to the need for lower-frequency transducers, limited acoustical windows, and postoperative changes, this examination provides a unique 2-D, real-time evaluation of the anatomic and hemodynamic relationships of intracardiac lesions.

TTE is widely available, reproducible, safe, and painless. As such it remains a valuable tool in the investigation of CHD.

Echocardiography using color flow Doppler is essential for evaluating blood flow as seen across an atrial defect or a ventricular septal defect or across a valve. Assessment of the valves (sclerosis, fusion, estimation of valve gradients) and an estimation of right ventricular (RV) systolic pressure can usually be achieved [13,14], although RV volumes, often followed to determine time of specific interventions, may be underestimated [15,16].

Saline contrast echocardiography (“bubble echo”) may be useful in verifying the presence of a shunt and in differentiating intracardiac from extracardiac shunts [17].

TTE has difficulty in consistently providing high-quality, clinically useful information in some adult patients with intracardiac defects. Imaging of the great vessels with TTE is difficult even in children and is even more problematic in adults who have poorer acoustical windows. In these situations, transesophageal echocardiography (TEE) and MRI have roles to play. Echocardiography also suffers from intraobserver variability in terms of examination reproducibility [18].

Current 2-D TTE is limited by a field of view of 90° and the need for the examiner to assimilate tomographic slices into a 3-D or 4-D diagnosis. The development of a real-time 3-D rotational acquisition format with dynamic volume rendering has allowed presentation of TTE in a 3-D display. In one study assessing hypoplastic left heart pediatric patients, 3-D echocardiography was found to be particularly useful when compared to 2-D echocardiography for evaluating ventricular volume, tricuspid valve regurgitation, tethering, and annulus configuration [19]. However, although 3-D echocardiography can provide more information than traditional 2-D techniques, it has been reported to be nondiagnostic in 27% to 48% [20] of adult patients secondary to inadequate quality caused by a variety of patient-related factors such as morbid obesity, narrow intercostal spaces, and severe pulmonary emphysema [21]. Three-dimensional echocardiography has also been reported to underestimate RV volumes in the adult patient [22].

Echocardiographic speckle tracking to assess ventricular strain and ventricular interaction has shown some utility in predicting patient outcomes [23]. In particular, echocardiographic-derived measurements of ventricular strain in Fontan patients has been shown to provide similar outcomes predictions as cardiac MRI-derived indexed ventricular end-diastolic volumes [24].

Transesophageal echocardiography

TEE has some clear advantages over TTE in adolescents and adults with CHD. TEE can provide a new or altered diagnosis (14%) or new information (56%) in adults with CHD [25]. New information obtained with TEE as compared to TTE includes identification of the atrial appendages and atrial septum, delineation of systemic and pulmonary venous connections, improved morphologic assessment of the atrioventricular junction and valves, improved definition of subaortic obstruction, improved definition of the ascending aorta and coronary arteries, and better evaluation of atrial baffle function and Fontan anatomy. TEE has been shown to be as accurate as CT for assessment of secundum ASD size and rim length in preparation for septal occluders [26]. Limitations of TEE include limited planes of view, poor visualization of specific regions (eg, apical-anterior septum and RV outflow tract), and blind areas created by masking of flow by implanted prosthetic material [25]. Areas that may be

difficult to visualize on TEE are the RV outflow tract, the pulmonary valve, the distal right pulmonary artery, and the proximal left pulmonary artery. With the development of 3-D TEE, these problem areas are better seen [27].

The standard TEE is an invasive examination that requires administration of a local anesthetic to the pharynx and intravenous (IV) midazolam in small doses. In large studies, it has been shown that the examination may be unsuccessful in up to 3% to 5% of patients because of their inability to tolerate the probe after intubation [28]. Another 4% to 5% of patients have the examination while under general anesthesia as part of an invasive or surgical procedure.

TEE is clearly operator dependent. In an area as complex as CHD, the examiner must be trained to interpret the findings in real time so that important information is not missed.

Radionuclide imaging

Although quantitation of cardiac shunts is feasible using technetium Tc-99m first-pass techniques, it is seldom used today, with cardiac MR flow quantification now the principal noninvasive method of intracardiac shunt assessment [29,30]. In some adult patients with abnormal pulmonary blood flow patterns, perfusion lung scanning may assist in the diagnosis, although this also has been largely replaced by pulmonary arterial MR flow quantification assessment [31]. Congenital anomalies of the coronary artery origins, notably anomalous origin of the coronary artery from the pulmonary artery and interarterial anomalous coronary artery, may result in myocardial ischemia and/or silent infarction, which can be identified using stress/rest radionuclide single-photon emission CT (SPECT) imaging. Stress/rest radionuclide SPECT imaging can also be used to evaluate myocardial perfusion and function of the systemic right ventricle in patients following repair of transposition of the great vessels, in which perfusion defects can commonly (54%) be seen on long-term follow-up [32].

Computed tomography angiography chest

Current CT scanners can evaluate the entire heart and great-vessel region with 3-D isovolumetric coverage [33-35]. Cardiac gating is not always necessary for assessment of great vessels and rings, but it is needed for evaluating intracardiac anatomy and coronary artery anomalies and to detect cardiac isomerism [36,37]. CT is advantageous in its ability to evaluate extracardiac anatomy such as the mediastinum and airway and sometimes eliminates the need for more invasive conventional tracheobronchoscopy. Compression of the airway may be present in CHD because of abnormal enlargement or an abnormal path of vasculature.

Computed tomography heart

Electrocardiogram gated CT of the heart that can contribute valuable information about intracardiac anatomy and the great vessels, including postoperative complications. CT of the heart has a short acquisition time, reducing the need for sedation [38]. Initially, patient exposure to radiation limited the use of CT in patients with CHD. However, variable pitch and low-kV techniques (80 kV) now permit acquisition with low-mSv radiation dosing in many patients [39,40] (See additional radiation dose reduction techniques below). Although visualization of the heart and great vessels on CT requires use of contrast media, adverse effects occur less often with low-osmolality agents [36,41,42].

Essentially all types of congenital cardiac malformations have been accurately described using CT of the heart [33-35,43-45]. CT of the heart has been used to calculate cardiac output, shunt flow, pulmonary to systemic flow ratios, ventricular volumes, ejection fraction, regurgitant volumes, and myocardial mass [39]. CT is limited in evaluation of the interatrial septum and membranous portion of the ventricular septum and is not currently used to assess flow patterns and turbulence.

Additionally, because of its high spatial resolution, CT is useful in assessing smaller and tortuous vessels as commonly seen in disorders such as total and partial anomalous pulmonary venous return and tetralogy of Fallot with pulmonary atresia [46,47]. CT can also be used as an alternative to MRI in patients with contraindications to MRI and in patients with metallic implants that may limit the use of MRI [48].

Computed tomography angiography coronary

Coronary CT angiography (CTA) is also electrocardiogram gated and performed with essentially the same CT acquisition technique as computed CT of the heart. Methods of postprocessing, however, include dedicated review and often multiplanar reformation of the coronary arteries. Although rare, the incidence of anomalous coronary arteries is increased in the congenital heart population. [49] Coronary CTA has been shown to be a high-resolution and high-image-quality assessment of anomalous coronary artery origin and course [50,51], especially to address questions of interarterial or aortic intramural course for purposes of surgical planning. Coronary CTA has also been shown to be useful for assessment of coronary artery fistula or malformation [52] and can assess for atherosclerosis and coronary artery stenosis in the older adult patient [53-55]. Because the coronary arteries are

highly susceptible to motion artifact, depending upon the type of scanner used, patients with higher heart rates (above 65 beats per minute) may benefit from the administration of an oral or IV beta-blocker to decrease their heart rate if there is no contraindication.

Recent advances in cardiac CT imaging technology allow for further radiation dose reduction in cardiac CTA examinations [56]; new and available dose-reducing techniques include prospective triggering [57-59], adaptive statistical iterative reconstruction [60], and high-pitch spiral acquisition [61]. However, these newer low-dose techniques may not be appropriate in all patients because of their dependency on a combination of factors, including heart rate, rhythm, and large body habitus. Thus, although these techniques are promising in terms of reducing patient radiation dose, there may be patients for whom these radiation dose techniques are not optimal, such as an obese elderly patient with an arrhythmia who might best benefit from retrospective gating in order to allow assessment of the coronary arteries at multiple phases of the cardiac cycle. In addition, not all scanners are capable of all radiation dose reduction techniques. In all cases, the imaging physician must select the appropriate combination of imaging parameters to acquire a diagnostic examination at a radiation dose that is as low as reasonably achievable. See also the section on Relative Radiation Level Information below.

Magnetic resonance imaging and magnetic resonance angiography

MRI is invaluable for evaluating CHD [43,62-70]. Without the concerns related to exposure to ionizing radiation, it can provide morphologic and functional information essential for detecting and managing CHD and information for surgical planning and predicting postoperative outcomes [71]. Traditional “black-blood” techniques (eg, fast spin-echo MRI and double inversion recovery fast spin echo) are useful for delineating basic cardiac and pericardiac anatomy. “Bright-blood” techniques, notably using cine steady-state free precession or gradient-recalled echo pulse sequences [72], can be used to demonstrate flow abnormalities (eg, a flow jet) related to lesions such as an interventricular or interatrial septal defect, valvular insufficiency, valvular stenosis, outflow obstruction, or aortic coarctation. Parallel imaging [62] and newer k-space schemes can shorten the acquisition times in most instances such that cine bright-blood imaging can be performed during a short breath hold. Bright-blood techniques also enable volumetric coverage of cardiac chambers for determining cardiac metrics such as ventricular volumes, ejection fractions, and myocardial mass and have been shown to be highly reproducible for both the right and left heart [73,74]. High-resolution cardiac-gated total coverage of the heart and great vessels can now be obtained using free-breathing methods, either with blood-pool gadolinium-based contrast enhancement or without contrast, using a high-native-contrast steady-state free precession sequence [75,76]. These highly accurate cardiac-gated MR angiography (MRA) techniques allow for isovolumetric assessment of the intracardiac anatomy, great vessels, and potential coronary anomalies and allow for multiplanar reformats and 3-D rendering of the heart, useful in both diagnosis and presurgical planning [76,77].

Phase-contrast techniques demonstrate directional blood flow information for improved identification of subtle intracardiac or extracardiac shunt lesions. Phase contrast [62,64] also allows quantification of blood flow (eg, calculation of the ratio of pulmonary to systemic blood flow [Qp/Qs]), regurgitant fractions, and pressure gradients across stenotic regions. Patients with highly complex congenital lesions, such as Fontan circuitry, can be assessed by MR phase-contrast methods to accurately determine systemic-to-pulmonary collateral flow, which can predict patient outcomes [78].

MRI has been used for diagnosing essentially all congenital heart and great-vessel abnormalities. It has been shown to have very high sensitivity and specificity for diagnosing common CHDs. Pulmonary and systemic venous anomalies and RV outflow obstructions are detected with high sensitivity. Vascular rings can also be accurately diagnosed without the need for invasive angiography. MRI can also be performed using 3-D techniques for high-spatial-resolution gadolinium-enhanced 3-D MRA [67,70] or to provide volumetric coverage of cardiac chambers [63-65,69]. Time-resolved MRA has been found to provide a very high diagnostic value (92% of diagnostic parameters assessed) that included thoracic vascular anatomy, sequential cardiac anatomy, and shunt detection with high sensitivity (93% to 100%) and high specificity (87% to 100%) [68].

Gradient-echo imaging acquisition viewed in a cine format facilitates physiologic measurements, including stroke volume, ejection fraction, and wall motion of both ventricles. Blood flow, valve gradients, shunt flow, regurgitant flow, and pulmonary flow can all be measured using velocity-encoded cine techniques [63,64,69,79,80].

MRI is ideally suited for evaluating adults with suspected or known CHD. Although claustrophobia in the gantry may require sedation in a few patients, the study is noninvasive, and image quality is not affected by body habitus. MRI can provide high-spatial-resolution images even in more complex CHD without the limitation of imaging “windows” or planes as experienced during echocardiography. MRI images can be obtained in essentially any plane for improved 3-D presentation of cardiac anatomy. MRI is useful as well in evaluating

postoperative patients with CHD, whether it involves a palliative procedure, a surgically created conduit, or reconstructed great vessels [63-67,70].

MRI, however, does have some contraindications and limitations. For instance, pacemakers are generally considered an exclusion for MRI, although MRI has been performed safely in patients with pacemakers under rigorously safe conditions [81] and using new MRI-conditional pacemakers [82]. The use of gadolinium chelate contrast agents may not be possible in patients with a known severe allergy to gadolinium or in patients with renal insufficiency [83]. Detection of calcification remains problematic for MRI, so adults with homografts or bioprosthetic valved conduits in whom the detection of calcification implies deterioration may not be optimally imaged. Motion and respiratory artifacts also may pose a problem on some examinations.

In terms of specific defects, MRI is probably not as accurate as color flow Doppler in visualizing small ventricular and atrial defects. Detection of valvular pathology is perhaps better achieved with TEE. Cardiac MRI studies require supervision and monitoring of the procedure by a physician who understands the clinical question and can acquire an appropriate and optimal imaging study. This is essential for consistency and reliable data.

3-D printing using cardiac magnetic resonance imaging and computed tomography data sets

Although no large trials or outcomes studies have been performed, case reports and small studies have demonstrated some utility in using high-resolution CT and blood-pool contrast-enhanced MR data sets to generate 3-D–printed reconstructions of the heart and great vessels for presurgical planning [84,85].

Transthoracic and Transesophageal Echocardiography versus Magnetic Resonance Imaging

Few prospective studies are available to compare TTE and TEE with MRI. Studies limited to specific congenital lesions (coarctation of the aorta, pulmonary valvular regurgitation and stenosis, and pulmonary artery anomalies) indicate that MRI gives a more reliable assessment of severity and is technically more successful than TTE in the adult patient [86-88]. Studies comparing TTE with MRI in the evaluation of patients who have had surgical correction or palliation of CHD indicate that MRI information is additive to that from TTE [24]. When TTE and MRI are compared for imaging a variety of congenital heart lesions, MRI is comparable to echocardiography in evaluating isolated intracardiac defects [89] and left ventricular function but more useful in diagnosing complex congenital lesions.

Echocardiography has also been shown to have good agreement with MRI in evaluating RV volumes, but echocardiography has been noted to have a much wider interobserver variation [18,90]. Similar agreement has been reported between echocardiographic and MRI measurements in patients with a functional single ventricle, with MRI showing better reproducibility than echocardiography in certain measurements, such as myocardial mass and diastolic volume [91]. In the evaluation of extracardiac ventriculopulmonary conduits and the right ventricle, MRI and echocardiography can often provide complementary and diagnostic information that, when in agreement, may obviate the need for cardiac catheterization [78,92].

Cardiac catheterization and angiocardiography

Cardiac catheterization has been the traditional gold standard for the diagnosis and management of CHD over the past 50 years. For the past 25 years, it has been increasingly supplemented by noninvasive diagnostic modalities—initially cardiac ultrasound and, more recently, CT scanning and MRI. Advances in these technologies have been logarithmic, and it is likely that in the coming decade both morphologic and functional assessments of this patient population will be increasingly accomplished noninvasively.

The 2008 American College of Cardiology/American Heart Association Guidelines for Adults with CHD suggest the use of diagnostic catheterization primarily to resolve specific issues related to surgical intervention (eg, preoperative evaluation of coronary arteries, assessment of pulmonary vascular disease and its response to vasoactive agents for planned surgical intervention, and/or heart or heart/lung transplantation) and as an adjunct to noninvasive assessment of morphologic and/or functional characteristics of complex CHD (eg, for delineating arterial and venous anatomy in patients with heterotaxy, patients who are candidates for a Fontan procedure, or patients who have had previous palliation in the form of a shunt) [93]. This guideline document notes that evaluation for possible interventional catheterization is an increasingly common indication for diagnostic catheterization [93]. Catheter intervention, for instance, is commonly sought as the treatment of choice for correcting valvular pulmonary stenosis or regurgitation [94], branch pulmonary stenosis, residual or recurrent aortic coarctation, and arteriovenous fistulae. Coil or device occlusion of lesions such as patent ductus or secundum ASD is another preferred intervention for treatment [95,96].

For many years, the purpose of cardiac catheterization and angiocardiography for CHD was to acquire pressure, oximetric, and morphologic data. Pressures defined gradients across stenosis and between cardiac chambers

connected by defects, as well as the severity of pulmonary hypertension. Oxygen saturations helped to define the volume of shunts. Morphologic data of simple and complex anomalies were obtained by cine angiograms using angulated views, contrast material, and radiation. For the most part, these studies are accomplished safely but with some morbidity (contrast reactions, renal failure, hematomas, arterial and venous injuries, radiation exposure, etc) and a small but definite mortality.

Although cardiac catheterization remains the gold standard in evaluating CHD, noninvasive methods increasingly limit the need for catheterization unless intervention is considered. Many simple congenital cardiac defects are now sent to surgery without catheterization [93]. Cardiac catheterization and angiocardiology complement these noninvasive techniques in the evaluation of adults with suspected CHD.

Summary of Recommendations

- The chest radiograph is a good initial assessment of the overall thorax in the adult with suspected or known CHD.
- TTE and MRI are frontline modalities for evaluating heart function and morphology and provide complementary information in diagnosing or assessing adult CHD.
- Heart CT (CT heart function and morphology with contrast) may be an alternative to MRI and TEE/TTE for evaluating heart function and anatomy. The high spatial resolution of CT is helpful in evaluating smaller and tortuous vessels commonly seen in some disorders. Heart CT does not provide information regarding vessel flow.
- Although noninvasive methods have reduced some of the need for cardiac catheterization, it remains an important adjunct to noninvasive imaging for hemodynamic measurements and for intervention.
- MRA of the chest and CTA of the chest provide anatomical information regarding the great vessels and their relationship to each other and the trachea and provide information on extracardiac anatomy.
- CTA of the coronary arteries is an important adjunctive modality for evaluating coronary anatomy and anomalies or atherosclerosis in the adult patient.

Summary of Evidence

Of the 96 references cited in the *ACR Appropriateness Criteria® Known or Suspected Congenital Heart Disease* document, 9 are categorized as therapeutic references including 1 well-designed study, 7 good-quality studies, and 1 quality study that may have design limitations. Additionally, 87 references are categorized as diagnostic references including 5 well-designed studies, 16 good-quality studies, and 32 quality studies that may have design limitations. There are 34 references that may not be useful as primary evidence.

The 96 references cited in the *ACR Appropriateness Criteria® Known or Suspected Congenital Heart Disease* document were published from 1991 to 2015.

Although there are references that report on studies with design limitations, 29 well-designed or good-quality studies provide good evidence.

Relative Radiation Level Information

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

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

Supporting Documents

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

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The ACR Committee on Appropriateness Criteria and its expert panels have developed criteria for determining appropriate imaging examinations for diagnosis and treatment of specified medical condition(s). These criteria are intended to guide radiologists, radiation oncologists and referring physicians in making decisions regarding radiologic imaging and treatment. Generally, the complexity and severity of a patient's clinical condition should dictate the selection of appropriate imaging procedures or treatments. Only those examinations generally used for evaluation of the patient's condition are ranked. Other imaging studies necessary to evaluate other co-existent diseases or other medical consequences of this condition are not considered in this document. The availability of equipment or personnel may influence the selection of appropriate imaging procedures or treatments. Imaging techniques classified as investigational by the FDA have not been considered in developing these criteria; however, study of new equipment and applications should be encouraged. The ultimate decision regarding the appropriateness of any specific radiologic examination or treatment must be made by the referring physician and radiologist in light of all the circumstances presented in an individual examination.