Complete Summary

GUIDELINE TITLE

Suspected congenital heart disease in the adult.

BIBLIOGRAPHIC SOURCE(S)

Ho VB, Yucel EK, Khan A, Haramati LB, Mammen L, Rozenshtein A, Rybicki FJ, Schoepf UJ, Stanford W, Stein B, Woodard PK, Jaff M, Expert Panel on Cardiovascular Imaging. Suspected congenital heart disease in the adult. [online publication]. Reston (VA): American College of Radiology (ACR); 2007. 8 p. [55 references]

GUIDELINE STATUS

This is the current release of the guideline.

It updates a previously published version: American College of Radiology (ACR), Expert Panel on Cardiovascular Imaging. Suspected congenital heart disease in the adult. Reston (VA): American College of Radiology (ACR); 2002. 6 p.(ACR appropriateness criteria). [38 references]

The appropriateness criteria are reviewed annually and updated by the panels as needed, depending on introduction of new and highly significant scientific evidence.

COMPLETE SUMMARY CONTENT

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SCOPE

DISEASE/CONDITION(S)

Congenital heart disease

GUIDELINE CATEGORY

Diagnosis
Evaluation

CLINICAL SPECIALTY

Cardiology
Family Practice
Internal Medicine
Nuclear Medicine
Radiology

INTENDED USERS

Health Plans
Hospitals
Managed Care Organizations
Physicians
Utilization Management

GUIDELINE OBJECTIVE(S)

To evaluate the appropriateness of initial radiologic examinations for adults with suspected congenital heart disease

TARGET POPULATION

Adults with suspected congenital heart disease

INTERVENTIONS AND PRACTICES CONSIDERED

1. X-ray, chest
2. Ultrasound (US) echocardiography transthoracic with Doppler
3. Magnetic resonance imaging (MRI) of the heart
4. US echocardiography transesophageal
5. Computed tomography angiogram (CTA), coronary arteries
6. Computed tomography (CT) of the heart with contrast
7. Magnetic resonance angiogram (MRA) of the chest
8. Invasive (INV) heart cardiac catheterization with angiocardiography
9. Nuclear medicine (NUC), scintigraphy of the heart
10. NUC shunt detection

MAJOR OUTCOMES CONSIDERED

Utility of radiologic examinations in differential diagnosis

METHODOLOGY

METHODS USED TO COLLECT/SELECT EVIDENCE

Searches of Electronic Databases

DESCRIPTION OF METHODS USED TO COLLECT/SELECT THE EVIDENCE

The guideline developer performed literature searches of recent peer-reviewed medical journals and the major applicable articles were identified and collected.

NUMBER OF SOURCE DOCUMENTS

Not stated

METHODS USED TO ASSESS THE QUALITY AND STRENGTH OF THE EVIDENCE

Weighting According to a Rating Scheme (Scheme Not Given)

RATING SCHEME FOR THE STRENGTH OF THE EVIDENCE

Not stated

METHODS USED TO ANALYZE THE EVIDENCE

Systematic Review with Evidence Tables

DESCRIPTION OF THE METHODS USED TO ANALYZE THE EVIDENCE

One or two topic leaders within a panel assume the responsibility of developing an evidence table for each clinical condition, based on analysis of the current literature. These tables serve as a basis for developing a narrative specific to each clinical condition.

METHODS USED TO FORMULATE THE RECOMMENDATIONS

Expert Consensus (Delphi)

DESCRIPTION OF METHODS USED TO FORMULATE THE RECOMMENDATIONS

Since data available from existing scientific studies are usually insufficient for meta-analysis, broad-based consensus techniques are needed for reaching agreement in the formulation of the appropriateness criteria. The American College of Radiology (ACR) Appropriateness Criteria panels use a modified Delphi technique to arrive at consensus. Serial surveys are conducted by distributing questionnaires to consolidate expert opinions within each panel. These questionnaires are distributed to the participants along with the evidence table and narrative as developed by the topic leader(s). Questionnaires are completed by participants in their own professional setting without influence of the other members. Voting is conducted using a scoring system from 1-9, indicating the least to the most appropriate imaging examination or therapeutic procedure. The survey results are collected, tabulated in anonymous fashion, and redistributed

after each round. A maximum of three rounds is conducted and opinions are unified to the highest degree possible. Eighty percent agreement is considered a consensus. This modified Delphi technique enables individual, unbiased expression, is economical, easy to understand, and relatively simple to conduct.

If consensus cannot be reached by the Delphi technique, the panel is convened and group consensus techniques are utilized. The strengths and weaknesses of each test or procedure are discussed and consensus reached whenever possible. If "No consensus" appears in the rating column, reasons for this decision are added to the comment sections.

RATING SCHEME FOR THE STRENGTH OF THE RECOMMENDATIONS

Not applicable

COST ANALYSIS

A formal cost analysis was not performed and published cost analyses were not reviewed.

METHOD OF GUIDELINE VALIDATION

Internal Peer Review

DESCRIPTION OF METHOD OF GUIDELINE VALIDATION

Criteria developed by the Expert Panels are reviewed by the American College of Radiology (ACR) Committee on Appropriateness Criteria.

RECOMMENDATIONS

MAJOR RECOMMENDATIONS

ACR Appropriateness Criteria®

Clinical Condition: Suspected Congenital Heart Disease in the Adult

Radiologic Procedure	Rating	Comments	RRL*
X-ray chest	9	Recommended in combination with TTE.	Min
US echocardiography transthoracic with Doppler	9	Recommended in combination with chest x-ray.	None
MRI heart	8	May be done as adjunct to TTE by trained operator if additional	None

Radiologic Procedure	Rating	Comments	RRL*
		information is required. See comments regarding contrast in text under "Anticipated Expectations."	
US echocardiography transesophageal	7	May be done as adjunct to TTE by trained operator if additional information is required.	None
CTA coronary arteries	7	Preferred technique for suspected coronary anomalies. Can also be used for evaluation of coronary artery disease.	High
CT heart with contrast	6	May be an alternative to MRI and TTE/TEE.	High
MRA chest	6	To evaluate associated vascular abnormalities. See comments regarding contrast in text under "Anticipated Expectations."	None
INV heart cardiac catheterization with angiocardiography	5	Adjunctive to noninvasive testing, for hemodynamic measurements or coronary artery status, or if other diagnostic information is required.	IP
NUC scintigraphy heart	4	May be used for perfusion in patients with suspected ischemic heart disease.	IP
NUC shunt detection	4	Alternative to MRI for shunt quantification.	Low
Rating Scale: 1=Least appropriate, 9=Most appropriate			***Relative Radiation Level**

Note: Abbreviations used in the tables are listed at the end of the "Major Recommendations" field.

Summary of Literature Review

Although patients with congenital heart disease (CHD) account for only a small percentage of adults with recognized heart disease, the number of adult patients in North America with CHD (ACHD) is increasing. A review of data from the National Center for Health Statistics and the Centers of Disease Control and Prevention showed a 39% decline in reported mortality from the CHD in the United States between 1979 and 1997. This decline is explained by 1) improvements in surgical treatment and survival, which is greater than 90% at 10 years, 2) the increasing numbers of immigrants, and 3) changes in the statistical methods used to calculate incidence of ACHD. The underestimation of these

figures is due in part to measuring the incidence of CHD presenting in infancy and childhood. However, at least 10% of patients diagnosed in ACHD clinics (in particular, those with secundum atrial septal defect, Ebstein's anomaly, and congenitally corrected transposition) are not diagnosed until adulthood. In addition to adults newly diagnosed with CHD, there is an increasing number of adults (estimated at >500,000) who have surgically treated CHD, with nearly 50% requiring two or more operations and 23%, requiring three or more. In Toronto, for example, a 269% expansion of outpatient workload in ACHD centers was noted between 1987 and 1997.

Adults with CHD also have acquired co-morbid factors, such as hypertension, atherosclerosis, coronary artery occlusive disease, pulmonary disease and renal disease, which may complicate their medical and/or surgical management.

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 (PDA), and tetralogy of Fallot. Uncommon congenital cardiac defects that may present in adulthood include Ebstein's anomaly, corrected transposition of the great vessels, pulmonary arteriovenous malformation, coronary artery anomalies, and sinus of Valsalva aneurysm.

The most common congenital heart defect in children, ventricular septal defect (VSD), 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 severe anomalies such as a persistent left superior vena cava and many anomalies of the origin of the great vessels from the arch are often asymptomatic.

Imaging procedures for the diagnosis of suspected CHD in the adult include plain chest film 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 intercardiac anatomy, vascular anatomy, hemodynamics, and function.

Chest Radiography

The initial work-up of adults with suspected congenital heart disease (CHD) usually includes a posteroanterior (PA) and lateral chest radiograph. Occasionally the radiograph will be the first study to alert the radiologist and the clinician to the 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 an invaluable tool for following the patient with surgically treated CHD.

Echocardiography

Transthoracic echocardiography (TTE) remains a first-line imaging examination in adults with 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-dimensional, real-time evaluation of the anatomic and hemodynamic relationships of intracardiac lesions.

Transthoracic echocardiography 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 particularly well suited for evaluating blood flow as seen across an atrial or a ventricular septal defect or across a valve. In postoperative patients, VSD patch defects can readily be visualized. Assessment of the valves (sclerosis, fusion, estimation of valve gradients) and determination of right ventricular systolic pressure can usually be achieved.

TTE, however, has difficulty in consistently providing high-quality clinically useful information in some adult patients with intracardiac defects. Imaging of the great vessel 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 exam reproducibility.

Current 2-dimensional TTE is limited by a field of view of 90 degrees and the need for the examiner to assimilate tomographic slices into a 3- or 4-dimensional diagnosis. The recent development of a rotational acquisition format with dynamic volume rendering has allowed presentation of TTE in a 3-dimensional display. In one study, 3-dimensional echocardiography was found to be particularly useful, when compared to 2-dimensional echocardiography, for evaluating mitral valve, aortoseptal continuity, and the intra-arterial septum. While 3D echocardiography can provide additional information over traditional 2D techniques, but has been reported to be nondiagnostic in up to 27% of patients secondary to inadequate quality from morbid obesity, narrow intercostal spaces, and severe pulmonary emphysema.

TEE has 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. 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. Limitations of TEE include limited planes of view, poor visualization of specific regions (e.g., apical-anterior

septum and right ventricular outflow tract), and blind areas created by masking of flow by implanted prosthetic material. Areas that may be difficult to visualize on TEE are the right ventricular outflow tract, the pulmonary valve, the distal right pulmonary artery, and the proximal left pulmonary artery. With the addition of the vertical axis in the newer probes, these problem areas, as well as the pulmonary veins, are better seen.

The standard TEE is an invasive examination that requires administration of a local anesthetic to the pharynx and intravenous midazolam in small doses. In large studies, it has been shown that the examination may be unsuccessful in up to 5% of patients due to their inability to tolerate the probe after intubation. Another 4%-5% of patients have the examination while under general anesthesia as part of invasive or surgical procedures. Although the risk of bacterial endocarditis from TEE is small, and prophylactic antibiotics are not routinely administered, endocarditis has been attributed to the procedure.

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

With improvements and refinements in echocardiographic technology, most adults attending an outpatient clinic undergo TTE and, when necessary, complementary TEE and MRI. Two-dimensional imaging is more challenging in this patient population because of large body size and often multiple previous surgical scars. The use of TEE intraoperatively is also increasing, and it has been shown to have a major impact on cardiac surgical procedures in 6%-9% of cases (e.g., that it is desirable or necessary for the patient to be put back onto cardiopulmonary bypass for revision of the cardiac procedure). Physicians interpreting these echocardiograms need to be experienced and have expertise in all aspects of CHD.

A high rate of diagnostic error in pediatric echocardiograms performed in community-based adult laboratories has been reported. One study found that 53% of patients of varying ages, from 1 day to 18 years, had either interpretive or technical errors that were of major or moderate importance. There is reason to believe that diagnostic errors occur even more frequently in older patients because image acquisition is typically more challenging.

Radionuclide Imaging

Although quantitation of cardiac shunts is feasible using technetium Tc 99m first-pass techniques, it is seldom used today. There are, however, a few selected uses for radionuclide imaging in evaluating adults with CHD. Left ventricular dysfunction is known to complicate certain long-standing congenital heart defects associated with right and left heart volume overload. Left ventricular radionuclide scintigraphy with ejection fraction calculation can be a useful noninvasive technique for evaluating these patients. Gated radionuclide scintigraphy can improve risk assessment in patient with single or systemic right ventricles by assessing heart failure and ventricular dysfunction. In some adult patients with abnormal pulmonary blood flow patterns related to conditions such as pulmonary artery agenesis, ventilation-perfusion lung scanning may assist in the diagnosis. 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 using stress/rest radionuclide SPECT (single photon emission computed tomography) 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.

Computed Tomography

Cardiac-gated CT and CT angiography (CTA) can contribute valuable information about congenital abnormalities of the coronary arteries and thoracic aorta, including the identification of vascular rings and postoperative complications such as pseudoaneurysm. The need for intravenous contrast material and the exposure to radiation, however, have limited their use in the pediatric population with CHD.

Current cardiac-gated CT scanners can evaluate the entire heart and great vessel region in a 3-dimensional matrix of CT information. Essentially all types of congenital cardiac malformations have been accurately described with this technique. Cardiac-gated CT has been used to calculate cardiac output, shunt flow, pulmonary-to-systemic flow ratios, ventricular volumes, ejection fraction, regurgitant volumes, and myocardial mass.

Magnetic Resonance Imaging

MRI is useful for evaluating CHD. Without the concerns related to exposure to ionizing radiation or the use of nephrotoxic iodinated contrast agents, it can provide morphologic and functional information essential for detecting and managing CHD. Traditional "black-blood" techniques (e.g., spin-echo MRI and double inversion recovery fast spin echo) are useful for delineating cardiac and pericardiac anatomy. "Bright-blood" techniques, notably using newer cine steady-state free-precession pulse sequences, can demonstrate flow abnormalities (e.g., a flow jet) related to lesions such as an interventricular or interatrial septal defect, valvular insufficiency, valvular stenosis, or coarctation. Defining the plane in which the jet velocity is maximal can be difficult with MRI. However, with improvements in software and real-time localization algorithms, this is becoming easier. Parallel imaging 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. Longer acquisitions as may be required for coronary magnetic resonance angiography (MRA) are typically performed using navigator respiratory gating methods.

Phase contrast techniques demonstrate directional blood flow information for improved identification of subtle intra- or extracardiac shunt lesions. Phase contrast also allows quantification of blood flow (e.g., estimation of the ratio of pulmonary to systemic blood flow [Qp/Qs]), regurgitant fractions, and pressure gradients across valves.

MRI has been used for diagnosing essentially all congenital heart and great-vessel abnormalities. Conventional spin-echo MRI has been shown to have very high

sensitivity and specificity for diagnosing common CHD. At a specificity of 90%, spin echo was found to have high sensitivity in diagnosing great-vessel relationships (100%), thoracic aortic abnormalities (94%), ASDs (91%), VSDs (100%), visceroatrial situs (100%), and the cardiac loop (100%). Pulmonary and systemic venous anomalies and right ventricular outflow obstructions are also detected with high sensitivity. Vascular rings can also be accurately diagnosed without the need for angiography. MRI can also be performed using 3D techniques for high spatial- resolution Gd-enhanced 3-D MRA, or to provide volumetric coverage of cardiac chambers. Time-resolved MRA was 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%-100%) and high specificity (87%-100%).

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.

MRI seems to be 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 plane as experienced during echocardiography. MRI images can be obtained in essentially any plane for improved 3-dimensional presentation of cardiac anatomy. MRI is useful as well in evaluating the postoperative patients with CHD, whether it is a palliative procedure, a surgically created conduit, or reconstructed great vessels.

Pharmacologic stress MRI using dobutamine has also been shown to be useful in evaluating systemic right ventricular function in patients with transposition of the great vessels. However, one group of researchers have shown significant differences in the responses between exercise stress and dobutamine stress MRI in patients with patients with intra-atrial correction of transposition of the great vessels, suggesting the need for further evaluations.

MRI, however, does have a few contraindications and limitations. For instance, pacemakers are generally considered an exclusion for MRI, although it has been performed safely in patients with pacemakers under rigorously safe conditions. The use of gadolinium (Gd) chelate contrast agents may not be possible in a patient with known severe allergy to Gd. 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.

Real-time MRI has recently been introduced and can facilitate evaluation in patients with dysrhythmias or limited breath-hold capacity. Current implementations have lower resolution than standard techniques.

Current cine studies represent a summation of acquisitions gated from the ECG. Any factor that affects ECG gating therefore may degrade the images, atrial fibrillation being the most common cause. Sternal wires, vascular clips, arterial

stents, biosynthetic valve rings, and mechanical valves can all interfere with the quality of the images and thereby limit the clinical value of the examination in some patients.

In terms of specific defects, MRI is probably not as accurate as color flow Doppler in visualizing small ventricular and atrial defects. Cardiac MR 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. Detection of thickened atrioventricular valves and the diagnosis of bicuspid or fused aortic valves are perhaps better achieved with TEE.

Future developments in MRI may optimize the acquisition of functional data and provide real-time visualization of cardiac structures, much as echocardiography does now.

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, subpulmonary and pulmonary artery anomalies) indicate that MRI gives a more reliable assessment of severity and is technically more successful than TTE. 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. In patients who have had palliative and corrective surgery for cyanotic heart disease, MRI and TEE are equivalent for demonstrating abnormalities of the right ventricular outflow tract, main pulmonary artery, and systemic-to-pulmonary shunts. MRI is superior in demonstrating abnormalities of the right and left pulmonary arteries. When TTE and MRI are compared in a variety of congenital heart lesions, MRI is comparable to echo in evaluating isolated intracardiac defects but more useful in diagnosing complex congenital lesions.

Echocardiography has also been shown to have good agreement with MRI in evaluating right ventricular volumes, but echocardiography has a much wider interobserver variation. 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.

When TEE and MRI have been evaluated prospectively in adults with CHD, TEE is shown to be superior in evaluating intracardiac anatomy; MRI is superior for extracardiac anatomy and is slightly better than TEE for hemodynamic and functional evaluation. Taken individually, the two modalities provided similar overall diagnostic information; however, when used in combination, they provide important complementary information in all diagnostic categories.

Cardiac Catheterization and Angiocardiography

Cardiac catheterization has been the diagnostic "gold standard" for CHD over the past 50 years. For the past 20 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.

In 2001, the Bethesda Conference Task Force 1, "The Changing Profile on Congenital Heart Disease in Adult Life," suggested the use of diagnostic catheterization primarily "for resolving specific issues concerning operative interventions including 1) the preoperative evaluation of coronary arteries; 2) the assessment of pulmonary vascular disease and its response to vasoactive agents for planned, traditional surgical intervention, and/or heart or heart/lung transplantation; and 3) as an adjunct to the noninvasive assessment of the morphologic and functional characteristics of many complex congenital lesions (e.g., delineation of arterial and venous anatomy, patients with heterotaxy, Fontan candidates, and patients who have had previous palliation in the form of a shunt.)" The group further suggested that only experienced and trained operators who maintain an adequate volume annually should perform such procedures. They further supported this suggestion by noting that evaluation for possible interventional catheterization is an increasingly common indication for diagnostic catheterization. Catheter intervention, for instance, is commonly sought as the treatment of choice for correcting valvular pulmonary stenosis, branch pulmonary stenosis, residual or recurrent aortic coarctation, and arteriovenous fistulae. Coil or device occlusion of lesions such as patent ductus or secundum atrial septal defect are other preferred interventions for treatment.

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 achieved by cine angiograms using angulated views, contrast material, and radiation. For the most part, these studies were 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 continues to be performed and is currently still considered by many to be 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. In the future, cardiac catheterization and angiocardiography may very well be reserved as a complement to these noninvasive techniques in the evaluation of adults with suspected CHD. However, until these less invasive studies provide an accurate depiction of the coronary arteries, the catheterization laboratory will continue to be involved in the assessment of this unique group of adult patients.

Anticipated Exceptions

Nephrogenic systemic fibrosis (NSF, also known as nephrogenic fibrosing dermopathy) was first identified in 1997 and has recently generated substantial concern among radiologists, referring doctors and lay people. Until the last few years, gadolinium-based MR contrast agents were widely believed to be almost

universally well tolerated, extremely safe and non-nephrotoxic, even when used in patients with impaired renal function. All available experience suggests that these agents remain generally very safe, but recently some patients with renal failure who have been exposed to gadolinium contrast agents (the percentage is unclear) have developed NSF, a syndrome that can be fatal. Further studies are necessary to determine what the exact relationships are between gadolinium-containing contrast agents, their specific components and stoichiometry, patient renal function and NSF. Current theory links the development of NSF to the administration of relatively high doses (e.g., >0.2 mM/kg) and to agents in which the gadolinium is least strongly chelated. The FDA has recently issued a "black box" warning concerning these contrast agents (http://www.fda.gov/cder/drug/InfoSheets/HCP/gcca_200705HCP.pdf.)

This warning recommends that, until further information is available, gadolinium contrast agents should not be administered to patients with either acute or significant chronic kidney disease (estimated GFR <30 mL/min/1.73m^2), recent liver or kidney transplant or hepato-renal syndrome, unless a risk-benefit assessment suggests that the benefit of administration in the particular patient clearly outweighs the potential risk(s).

Abbreviations

- CT, computed tomography
- CTA, computed tomography angiography
- INV, invasive
- IP, in progress
- Min, minimal
- MRA, magnetic resonance angiography
- MRI, magnetic resonance imaging
- NUC, nuclear imaging
- TEE, transesophageal echocardiography
- TTE, transthoracic echocardiography
- US, ultrasound

CLINICAL ALGORITHM(S)

None provided

EVIDENCE SUPPORTING THE RECOMMENDATIONS

TYPE OF EVIDENCE SUPPORTING THE RECOMMENDATIONS

The recommendations are based on analysis of the current literature and expert panel consensus.

BENEFITS/HARMS OF IMPLEMENTING THE GUIDELINE RECOMMENDATIONS

POTENTIAL BENEFITS

Appropriate selection of initial radiologic exam procedures for suspected congenital heart disease

POTENTIAL HARMS

- The relative radiation level is high for computed tomography (CT) angiography of the coronary arteries and CT of the heart with contrast.
- There is a small risk of bacterial endocarditis from transesophageal echocardiography (TEE).
- Sternal wires, vascular clips, biosynthetic valve rings, and mechanical valves can all interfere with the quality of magnetic resonance (MR) images and thereby limit their clinical value.
- Some patients with renal failure who have been exposed to gadolinium-based MR contrast agents (the percentage is unclear) have developed nephrogenic systemic fibrosis, a syndrome that can be fatal. Until further information is available, gadolinium contrast agents should not be administered to patients with either acute or significant chronic kidney disease (estimated GFR <30 mL/min/1.73m^2), recent liver or kidney transplant or hepato-renal syndrome, unless a risk-benefit assessment suggests that the benefit of administration in the particular patient clearly outweighs the potential risk(s)

CONTRAINDICATIONS

CONTRAINDICATIONS

- Pacemakers are generally considered an exclusion for magnetic resonance imaging (MRI), although it has been performed safely in patients with pacemakers under rigorously safe conditions.
- The use of gadolinium (Gd) chelate contrast agents may not be possible in a patient with known severe allergy to Gd.
- Detection of calcification is a problem for MRI, so adults with homografts or bioprosthetic valved conduits in whom the detection of calcification implies deterioration may not be optimally imaged.

QUALIFYING STATEMENTS

QUALIFYING STATEMENTS

An American College of Radiology (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 exams 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 U.S. Food and Drug Administration (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.

IMPLEMENTATION OF THE GUIDELINE

DESCRIPTION OF IMPLEMENTATION STRATEGY

An implementation strategy was not provided.

IMPLEMENTATION TOOLS

Personal Digital Assistant (PDA) Downloads

For information about availability, see the "Availability of Companion Documents" and "Patient Resources" fields below.

INSTITUTE OF MEDICINE (IOM) NATIONAL HEALTHCARE QUALITY REPORT CATEGORIES

IOM CARE NEED

Living with Illness

IOM DOMAIN

Effectiveness

IDENTIFYING INFORMATION AND AVAILABILITY

BIBLIOGRAPHIC SOURCE(S)

Ho VB, Yucel EK, Khan A, Haramati LB, Mammen L, Rozenshtein A, Rybicki FJ, Schoepf UJ, Stanford W, Stein B, Woodard PK, Jaff M, Expert Panel on Cardiovascular Imaging. Suspected congenital heart disease in the adult. [online publication]. Reston (VA): American College of Radiology (ACR); 2007. 8 p. [55 references]

ADAPTATION

Not applicable: The guideline was not adapted from another source.

DATE RELEASED

1998 (revised 2007)

GUIDELINE DEVELOPER(S)

American College of Radiology - Medical Specialty Society

SOURCE(S) OF FUNDING

The American College of Radiology (ACR) provided the funding and the resources for these ACR Appropriateness Criteria®.

GUIDELINE COMMITTEE

Committee on Appropriateness Criteria, Expert Panel on Cardiovascular Imaging

COMPOSITION OF GROUP THAT AUTHORED THE GUIDELINE

Panel Members: Vincent B. Ho, MD; E. Kent Yucel, MD; Arfa Khan, MD; Linda B. Haramati, MD; Leena Mammen, MD; Anna Rozenshtein, MD; Frank J. Rybicki, MD, PhD; U. Joseph Schoepf, MD; William Stanford, MD; Barry Stein, MD; Pamela K. Woodard, MD; Michael Jaff, MD

FINANCIAL DISCLOSURES/CONFLICTS OF INTEREST

Not stated

GUIDELINE STATUS

This is the current release of the guideline.

It updates a previously published version: American College of Radiology (ACR), Expert Panel on Cardiovascular Imaging. Suspected congenital heart disease in the adult. Reston (VA): American College of Radiology (ACR); 2002. 6 p.(ACR appropriateness criteria). [38 references]

The appropriateness criteria are reviewed annually and updated by the panels as needed, depending on introduction of new and highly significant scientific evidence.

GUIDELINE AVAILABILITY

Electronic copies: Available in Portable Document Format (PDF) from the American College of Radiology (ACR) Web site.

ACR Appropriateness Criteria® *Anytime*, *Anywhere*™ (PDA application). Available from the ACR Web site.

Print copies: Available from the American College of Radiology, 1891 Preston White Drive, Reston, VA 20191. Telephone: (703) 648-8900.

AVAILABILITY OF COMPANION DOCUMENTS

The following are available:

- ACR Appropriateness Criteria®. Background and development. Reston (VA): American College of Radiology; 2 p. Electronic copies: Available in Portable Document Format (PDF) from the American College of Radiology (ACR) Web site.
- ACR Appropriateness Criteria®. Relative radiation level information. Reston (VA): American College of Radiology; 2 p. Electronic copies: Available in Portable Document Format (PDF) from the American College of Radiology (ACR) Web site.

PATIENT RESOURCES

None available

NGC STATUS

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