Agency for Healthcare Research and Quality
Advancing Excellence in Health Care

NATIONAL GUIDELINE CLEARINGHOUSE

General

Guideline Title

ACR Appropriateness Criteria® pulmonary hypertension.

Bibliographic Source(s)

Brown K, Gutierrez AJ, Mohammed TL, Kirsch J, Chung JH, Dyer DS, Ginsburg ME, Heitkamp DE, Kanne JP, Kazerooni EA, Ketai LH, Parker JA, Ravenel JG, Saleh AG, Shah RD, Steiner RM, Suh RD, Expert Panel on Thoracic Imaging. ACR Appropriateness CriteriaÂ® pulmonary hypertension. [online publication]. Reston (VA): American College of Radiology (ACR); 2012. 10 p. [90 references]

Guideline Status

This is the current release of the guideline.

Recommendations

Major Recommendations

ACR Appropriateness Criteria®

Clinical Condition: Pulmonary Hypertension

Radiologic Procedure	Rating	Comments	RRL*
US echocardiography transthoracic resting	9	Catheterization and echocardiography are complementary examinations. Both should be performed. Echocardiography is typically performed before catheterization.	O
Right heart catheterization	9	Catheterization and echocardiography are complementary examinations. Both should be performed. Echocardiography is typically performed before catheterization.	
X-ray chest	8		
CT chest (noncoronary) with contrast	4,5,6 May be appropriate; 7,8,9 Usually appropriate		

Rating Scale: 1,2,3 Usually not appropriate; 4,5,6 May be appropriate; 7,8,9 Usually appropriate

*Relative Radiation Level

Radiologic Procedure	Rating	Comments	RRL*
Tc-99mV/Q scanning	7		
MRI heart function and morphology without contrast	6	May be performed with MRA.	O
MRI heart function and morphology without and with contrast	6	See statement regarding contrast in text under "Anticipated Exceptions."	O
MRA pulmonary arteries without and with contrast	6	See statement regarding contrast in text under "Anticipated Exceptions."	O
Arteriography pulmonary with right heart catheterization	6		
CT chest without contrast	5	If there is a concern for an occult ILD, HRCT may be appropriate.	
US echocardiography transesophageal	5		O
MRA pulmonary arteries without contrast	2		O
CT chest without and with contrast	1	CT chest with and without contrast does not always provide the same information as a CTA chest.	
Rating Scale: 1,2,3 Usually not appropriate; 4,5,6 May be appropriate; 7,8,9 Usually appropriate			*Relative Radiation Level

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

Summary of Literature Review

Introduction/Background

Pulmonary hypertension (PH) is a complex disorder and may be idiopathic or related to a variety of diseases. In patients with pulmonary arterial hypertension (PAH), progressive narrowing of the small pulmonary arteries and arterioles results in increased pulmonary vascular resistance, which may ultimately lead to right ventricular failure and death. Vasoconstriction, vascular-wall remodeling, and thrombosis in situ are factors increasing vascular resistance. Although different pathologic characteristics are seen in the diverse clinical PH groups, medial hypertrophy, intimal proliferation and fibrosis, and the presence of plexiform lesions are common features.

A series of global meetings has been critical in the evolution of understanding of PH. The first hemodynamic definition was proposed at the first World Symposium on Pulmonary Hypertension in 1973 in Geneva, Switzerland. It defined PAH as an increase in main pulmonary arterial pressure, with a mean pulmonary artery pressure (PAP) >25 mm Hg at rest or >30 mm Hg with exercise, in the presence of a pulmonary capillary wedge pressure ≤15 mm Hg. The Second World Symposium on Pulmonary Hypertension, held in Evian, France in 1998, resulted in the "Evian Classification," a clinical classification of PH comprising five major categories with similarities in pathophysiological mechanisms, clinical presentations, and therapeutic options. The 2003 World Symposium assessed and updated the Evian Classification and resulted in the Revised Clinical Classification of Pulmonary Hypertension. These modifications were: 1) the inclusion of a genetic classification, 2) discontinuing the use of the term "primary pulmonary hypertension," 3) reclassification of pulmonary veno-occlusive disease (PVOD) and pulmonary capillary hemangiomatosis (PCH), 4) update on the new risk factors for PAH, and 5) reassessment of the classification of congenital systemic-to-pulmonary shunts. Most recently, the Fourth World Symposium on Pulmonary Hypertension in 2008 in Dana Point, California, recommended a revised hemodynamic definition of PH as a mean PAP of ≥25 mm Hg without inclusion of exercise criterion, in view of evidence that the cut-off level of >30 mm Hg did not clearly differentiate PH from physiological response to exercise.

The five categories of PH include Group 1, PAH; Group 1′, PVOD and/or PCH; Group 2, PH associated with left heart diseases; Group 3, PH associated with lung respiratory disease and/or hypoxia; Group 4, PH due to chronic thromboembolic disease; and Group 5, PH with unclear and/or multifactorial mechanism (see Appendix 1 in the original guideline document).

Other classifications of PAH have been based on the histologic findings, such as precapillary and postcapillary etiologies and functional severity of symptoms (World Health Organization Functional Status). A genetic classification of PAH has also been proposed, as mutations in the bone morphogenetic protein receptor II (BMPR2) gene may be associated with 50% to 60% of cases with familial PAH and up to 26% of sporadic cases of primary PH.

Manifestations of PH may not be apparent until pulmonary vascular disease is advanced, with symptoms of PH attributable to impaired oxygen transport and reduced cardiac output. Exertional dyspnea is the most frequent presenting symptom, with other complaints including fatigue, weakness, angina, syncope, peripheral edema, and abdominal distension. Given the nonspecific signs and symptoms and the diverse group of diseases that comprise the PH spectrum, detection and characterization can be challenging. A careful history is critical to evaluate for risk factors for PH, including family history, history of drugs and toxins associated with PH, collagen vascular disease, human immunodeficiency virus (HIV), portal hypertension, congenital or left heart disease, and venous thromboembolic disease. Clinical evaluation includes pulmonary function tests and arterial blood gases to evaluate for lung disease, and routine biochemistry, hematology, thyroid function, and serological testing to evaluate for lung disease, liver disease, connective tissue disorders, and HIV. The diagnosis, accurate assessment of etiology and severity, prognosis, treatment response, and follow-up of PH can be achieved by using the diverse set of diagnostic examinations in a reasonable manner that is tailored for each specific patient, with a goal of early detection and accurate characterization so that an appropriate medical or surgical therapy can be instituted.

Overview of Imaging Modalities

Chest Radiography

Historical studies have demonstrated that the noninvasive chest radiograph is an appropriate study for evaluating patients at risk for PH and for the detection or confirmation of the diagnosis of PH. The chest radiograph may reveal evidence of diffuse lung disease such as interstitial lung disease (ILD) and emphysema or evidence of pulmonary venous hypertension. However, the chest radiograph is relatively insensitive for detecting mild PH. Often, the right descending pulmonary artery may be difficult to evaluate in the setting of right heart enlargement. In addition, most studies of chest radiography and PH are in the setting of chronic obstructive pulmonary disease (COPD) and mitral stenosis, and their applicability in patients with the World Health Organization (WHO) group idiopathic PAH (IPAH) is less clear.

In a study of patients with COPD, authors compared analysis of the pulmonary vasculature on chest radiographs with PAPs measured during right heart catheterization and demonstrated that combined increased right descending pulmonary artery and increased left pulmonary artery diameter permitted the correct diagnosis in 98% of patients with PAH secondary to COPD. In another study, mean PAP was measured in patients with moderate to severe COPD, and correlated with 1) hilar thoracic index, 2) diameter of the descending branch of the right pulmonary artery, 3) the hilar width, and 4) the cardiothoracic ratio. The authors found that PAP best correlated with the hilar thoracic index, and significantly correlated with the other three indexes, but the accuracy with which PAP was predicted was poor.

In a prospective study of IPAH, the chest radiograph demonstrated prominence of the main pulmonary artery in 90% of patients, enlarged hilar vessels in 80%, and decreased peripheral vessels in 51%. All three abnormalities were seen in 42%, and the presence of all three abnormalities was associated with a higher mean PA pressure and lower cardiac index. A normal chest radiograph was seen in only 6% of patients.

In patients with chronic thromboembolic PH (CTEPH), surgical thrombectomy may be curative. One study evaluated whether noninvasive examinations, including conventional radiography and chest computed tomography (CT) could predict the extent of PH and reversibility after surgery. A dilated pulmonary trunk was the most common radiographic abnormality, seen in 96% of patients, with dilatation of the right and left pulmonary arteries in 40% and 14%, respectively. The degree of PH did not closely correlate with any radiographic sign.

In clinical practice, although chest radiography is insensitive for the detection of mild-moderate PH, it is nonetheless recommended in the initial evaluation of adult patients with unexplained dyspnea or other symptoms of possible PH or the evaluation of patients at risk for PH.

Doppler Echocardiography

Echocardiography is noninvasive, widely available, reproducible, and relatively inexpensive. The lack of radiation exposure makes it a valuable examination for serial follow-up studies. Limitations of Doppler echocardiography include acoustic window restrictions (particularly in patients with underlying lung disease), limitations due to body habitus, and operator dependence.

Patients at high risk for development of PAH may benefit from screening using Doppler echocardiography. At-risk patients include: 1) individuals with known genetic-mutation-associated PAH or a first-degree relative with IPAH; 2) scleroderma spectrum of disease; 3) patients with congenital

heart disease and systemic-to-pulmonary shunts; or 4) patients with portal hypertension prior to liver transplant.

Doppler echocardiography allows estimation of right ventricular systolic pressure (RVSP) and pulmonary diastolic pressure. Most often, PAP is calculated by measuring the velocity of the tricuspid regurgitant jet. Doppler echocardiography is also useful for evaluating cardiac anatomy such as chamber enlargement, valvular function and morphology, left ventricular systolic and diastolic dysfunction, and the presence of pericardial effusion. An echocardiographic contrast or "bubble" study using agitated saline may be useful for detecting intracardiac shunts. Echocardiographic evaluation of the right ventricular myocardial performance index and tricuspid annular plane systolic excursion index should be measured concomitantly with mean PAP, as PAP in patients with advanced PAH may decrease with deterioration of right ventricular function.

Some authors have suggested that echocardiographic abnormalities such as pericardial effusion, right atrial enlargement, and septal displacement may reflect the severity of right heart failure and may predict poor outcome. Other investigators have found that tissue Doppler imaging may allow estimation of right ventricular filling pressures and predict cardiac events, functional status, and exercise capacity.

Ventilation-Perfusion Scans

The algorithm recommended by the American College of Cardiology Working Group calls for ventilation-perfusion (V/Q) scanning in all patients with unexplained PH. V/Q scans are particularly useful in determining CTEPH and differentiating CTEPH from other causes of PH. V/Q scanning demonstrated a sensitivity of 90% to 100% and specificity of 94% to 100% for differentiation between IPAH and CTEPH. A normal or low-probability scan essentially excludes the diagnosis of CTEPH with a sensitivity of 90% to 100% and a specificity of 94% to 100%. The V/Q scan may be normal in other causes of PH.

A group of researchers found that V/Q scintigraphy was more sensitive than multidetector CT pulmonary angiography (CTPA) in detecting chronic thromboembolic pulmonary disease amenable to surgery, with V/Q scans demonstrating a sensitivity of 96% to 97.4% and a specificity of 90% to 95% compared to a sensitivity of 51% and specificity of 99% for multidetector CTPA. However, more recent studies using 40- or 64-row scanners have demonstrated sensitivities and specificities of CTPA of 99% to 100% and 100%, respectively.

Right Heart Catheterization

Right heart catheterization is the gold standard for the diagnosis of PAH. At experienced institutions, it has morbidity and mortality rates of 1.1% and 0.055%, respectively. Right heart catheterization directly measures PAP and cardiac function. The vasoreactivity of pulmonary circulation should be tested using a short-acting drug and taking direct pressure measurements. Such tests may identify patients likely to respond to long-term therapy with calcium channel blockers. Subsequent catheterizations are then required to monitor treatment response.

Pulmonary Angiography

Catheter pulmonary angiography remains the standard of care for assessing operability in the patient with CTEPH. Traditional pulmonary angiography has been shown to be superior to CTPA in determining which patients will benefit from pulmonary thrombectomy, although other authors have demonstrated that CTPA is as reliable as digital subtraction angiography in the evaluation of CTEPH. Angiography and CTPA may be used for evaluating possible arteriovenous malformations, and CTPA, magnetic resonance imaging (MRI), and positron emission tomography (PET) may be used for evaluating vasculitis. Findings of CTEPH on angiography include webs or bands with or without stenotic dilatation, intimal irregularities, and abrupt narrowing or occlusion of segmental or larger vessels. Similar findings may be seen on CTPA and MRI.

Computed Tomography

Chest CT and CTPA may be useful in assessing the clinical classification of PAH. A number of findings on CT have been shown to be useful in evaluating possible PAH. A ratio of pulmonary artery diameter to aortic diameter of >1 has been shown to correlate with elevated mean PAP, although a ratio of <1 does not exclude PAH. In one study, a main pulmonary artery diameter of ≥29 mm was shown to have a sensitivity of 69% and specificity of 100% for predicting PAH. Main pulmonary artery diameter has been shown to be useful for detecting PAH in patients with advanced lung disease, with a sensitivity of 87% and specificity of 89%, with the additional finding that a segmental artery-to-bronchus ratio >1:1 increases specificity. However, more recent studies have shown that the sensitivity and specificity of main pulmonary artery diameter vary widely, depending on the etiology of the lung disease. Pericardial thickening or effusion is a frequent finding in patients with severe PH, with one study demonstrating small to moderate pericardial thickening or effusions in over half of patients with a mean PAP >35 mm Hg. Extrinsic compression of the left main coronary artery by a dilated main pulmonary artery, an uncommon finding in PAH, has been reported on CT.

High-resolution CT (HRCT) is indicated for evaluating diffuse lung disease and for screening of patients with chronic unexplained dyspnea (see the National Guideline Clearinghouse [NGC] summary of ACR Appropriateness Criteria® chronic dyspnea — suspected pulmonary origin and the ACR Appropriateness Criteria® dyspnea — suspected cardiac origin). Diffuse lung diseases associated with PH include ILD and emphysema, sarcoidosis, connective tissue disease and pulmonary Langerhans cell histiocytosis. In many cases, HRCT may obviate the need for open lung biopsy.

The diagnosis of several rare diseases associated with PAH may be suggested by HRCT. PVOD and PCH are uncommon disorders and demonstrate similar pathologic features, risk factors, and associated conditions and may represent spectrums of the same disease process. PVOD is characterized by extensive and diffuse obstructions of pulmonary veins and venules by intimal fibrosis. Clinical criteria include PAH, chronic edema, and a normal wedge pressure. Although studies in the literature are primarily case reports and small series, smooth interlobular septal thickening, diffuse multifocal regions of ground glass opacity, mosaic attenuation, pleural effusions, normal size pulmonary veins and left atrium, and enlarged central pulmonary arteries are features seen on CT. Adenopathy has also been reported. PCH is characterized by capillary proliferation and dilatation in the alveolar wall with obstruction of small venules. CT findings include small nodular opacities, thickened interlobular septa, and regions of ground glass opacification. Diffuse ill-defined centrilobular micronodules have been identified in a small series of patients with severe PAH. Of interest, pulmonary cholesterol granulomas have been seen in approximately 25% of patients with severe PAH at autopsy, and were found in histopathologic examination in a small series of patients reported by this study.

CTPA is increasingly used to evaluate thromboembolic disease, and it has become the standard of care at many institutions. Some authors have suggested that the greater expertise in interpretation of CT angiography (CTA) supports the use of CTA rather than V/Q scans for initial evaluation of patients with suspected CTEPH. Findings of chronic thromboembolic disease include eccentric pulmonary arterial filling defects, complete vessel occlusion, calcification within chronic thrombi, and enlarged bronchial and nonbronchial systemic arteries. Abnormalities of the lung parenchyma include mosaic attenuation, focal ground glass opacities, parenchymal scars, and bronchial abnormalities. Although nonspecific and identified in other causes of PH, mosaic perfusion secondary to regions of hypoperfusion and redistribution of blood flow within the arterial bed may be present. Evidence of airway obstruction with air trapping on expiratory images and cylindrical bronchiectasis has also been reported. Accuracy of CTPA is greatest for main and lobar arterial thrombi.

Other findings may be helpful in the evaluation of PH. Pulmonary neovascularity has been shown on CT in Eisenmenger syndrome as small tortuous intrapulmonary vessels. Other investigators have shown centrilobular nodules, mosaicism, neovascularity, and bronchial artery hypertrophy in patients with IPAH and Eisenmenger syndrome, with a higher number of enlarged bronchial arteries in patients with Eisenmenger syndrome. These findings did not help determine the cause of PAH, nor predict the severity of PAH.

Magnetic Resonance Imaging

Current MRI and magnetic resonance angiography (MRA) techniques can provide accurate, reproducible, and noninvasive assessment of the cardiovascular system. In addition, emerging techniques have proven to be promising in the assessment of pulmonary parenchymal and pulmonary perfusion abnormalities. Using current and new techniques, MRI/MRA can provide reproducible quantification of morphologic and functional changes occurring in PH that can aid in diagnosis, helps evaluate disease severity and prognosis, and provide a noninvasive alternative in evaluating treatment response.

MRI/MRA can identify the morphological changes seen in PH. Indirect signs of PH are similar to those seen on chest CT — for example, dilatation of the central pulmonary arteries and pruning of peripheral vasculature. Additionally, morphologic changes of the right heart are used as indirect signs of elevated right heart pressures and cardiac remodeling. Right ventricle remodeling is characterized by right ventricular dilatation, concentric right ventricular hypertrophy, increased right ventricular mass, increased right ventricular trabeculation, and in severe IPAH, right ventricular pressure overload causes the interventricular septum to flatten with a D-shaped configuration of the left ventricle on short-axis images.

MRI can also provide reproducible and noninvasive functional information to assess severity of disease and prognosis. Functional abnormalities seen in cardiac remodeling secondary to PH include right ventricular hypokinesis, paradoxical movement of the interventricular septum, right ventricular dysfunction (increased end-diastolic volume, reduced ejection fraction [EF], reduced cardiac index [CI], reduced stroke volume), and pulmonary and tricuspid insufficiency. Predictors of poor outcome determined by MRI include right ventricular dysfunction, right atrial dilatation, septal flattening/inversion, delayed enhancement of the right ventricle, left ventricular dysfunction, and extracardiac signs such as ascites, pericardial effusion, and lower-extremity edema.

Small studies have shown that MRI can also be used as a noninvasive method for obtaining functional information to monitor treatment response, and can be performed serially to monitor changes in right ventricular parameters as a response to medical therapy. By detecting increases in right ventricular stroke volume, reversion of the septal shift, and decreases in right ventricular mass, MRI can assess the long-term effects of vasodilator therapy.

Differentiating CTEPH from IPAH is critical because surgical thrombectomy may be curative in CTEPH patients. Small studies have shown that current and new MRI techniques are accurate and reliable adjuncts in differentiating IPAH from CTEPH. The combination of MRA and MR perfusion imaging can reliably rule in or out CTEPH while providing important functional and morphologic cardiac information. MRI can also determine if a patient is a surgical candidate and can assess treatment response.

MRI has proven to be essential for the diagnosis and characterization of congenital heart abnormalities. Specifically, MRI has become an important tool in detecting and quantifying cardiovascular shunt lesions while providing information on right heart function (Group 1.4.4. [see Appendix 1 in

the original guideline document]). Of utmost importance is the ability of MRI to detect and characterize shunts that are difficult to identify on echocardiography, including sinus venosus atrial septal defects, atrioventricular septal defects, and partial anomalous pulmonary venous return, with studies showing a high sensitivity (93%–100%) and specificity (87%–100%) for shunt detection.

The advantages of MRI include its lack of ionizing radiation and its ability to provide high spatial resolution images in any plane without the need of an imaging "window" as echocardiography does. The major contraindication to MRI is the presence of specific ferromagnetic and/or conducting implants such as cardiac pacemakers, although MRI has been performed safely in patients with pacemakers under rigorous safety conditions. Contraindications to intravenous gadolinium chelate contrast that is required for certain sequences include allergy to gadolinium or renal dysfunction.

Limitations of MRI include motion and respiratory artifacts that may degrade image quality particularly for certain motion sensitive sequences; long acquisition times; and the need for sedation in patients with claustrophobia. Given its high diagnostic sensitivity and specificity and lack of ionizing radiation, MRI may be used as an adjunct or provide a comprehensive alternative to current first-line or invasive examinations at many tertiary centers. This is particularly important for young patients for whom the risks from repeated radiation exposures are greater and for patients with significant comorbidities that result in greater risk from repeated right heart catheterizations.

Positron Emission Tomography (FDG-PET)

Fluorine-18-2-fluoro-2-deoxy-D-glucose (FDG) positron emission tomography (PET) allows in-vivo imaging of metabolic processes and is complementary to the structural/anatomic information provided by cross-sectional imaging modalities. FDG-PET is well established for the diagnosis and management of malignancy, but is becoming a valuable imaging modality for characterization and diagnosis of various inflammatory conditions. The use of FDG-PET in the evaluation of pulmonary hypertension is extremely limited, but it may be useful in distinguishing rare cross-sectional mimics of chronic thromboembolic pulmonary hypertension, including pulmonary artery sarcoma and medium to large vessel vasculitis (e.g., Takayasu arteritis), both of which will demonstrate increased FDG uptake. Differentiating CTEPH from these rare mimics is critical because of important treatment implications.

In addition, small studies have suggested that the metabolic information provided by FDG-PET in patients with pulmonary hypertension may be used to assess treatment response to medical therapy. A small study demonstrated increased FDG uptake in the lungs of patients with idiopathic pulmonary arterial hypertension that may be measured to assess response to targeted therapy. Another small study demonstrated that increased FDG uptake in the right ventricular myocardium correlated with the severity of right ventricular overload and that after treatment with epoprostenol the right ventricular FDG activity decreased, along with a decrease in pulmonary vascular resistance and right ventricular peak-systolic wall stress.

Summary

- Chest radiography is indicated in the diagnostic evaluation of PH, based on historical studies, its utility in the evaluation of patients with unexplained dyspnea, its low cost, and its low radiation dose. However, the sensitivity of the chest radiograph is inadequate for it to function as a screening test for patients at risk for PH, and the degree of PH does not correlate with radiographic findings.
- Doppler echocardiography is the screening test of choice for evaluating PH and the examination of choice for the follow-up of patients with PH.
- Right heart catheterization remains the gold standard for the diagnosis of PAH and directly measures PAP and cardiac function. Vasoreactivity of the pulmonary circulation may identify patients likely to respond to long-term therapy with calcium channel blockers.
- V/Q scan should be performed to evaluate for possible CTEPH. CTPA and MRI may also be used to evaluate for CTEPH.
- CT is a noninvasive method to evaluate diffuse lung disease, and pulmonary artery diameter and pulmonary artery to aorta ratios are easily determined on cross-sectional imaging. HRCT is recommended for further evaluation of patients with unexplained dyspnea and may aid in the clinical classification of PAH.
- Cardiopulmonary MRI is emerging as an important diagnostic modality in the complex evaluation and management of PH. MRI can provide a wide-range of morphological and functional cardiovascular information and is indicated in the evaluation of congenital heart disease and cardiovascular shunts as an etiology of PH. Larger studies are needed to corroborate the results from smaller studies suggesting that MRI is an effective noninvasive adjunct or alternative to current first-line or invasive examinations.
- The use of FDG-PET in evaluating pulmonary hypertension is extremely limited, but it may be useful in distinguishing rare mimics of CTEPH.

Anticipated Exceptions

Nephrogenic systemic fibrosis (NSF) is a disorder with a scleroderma-like presentation and a spectrum of manifestations that can range from limited clinical sequelae to fatality. It appears to be related to both underlying severe renal dysfunction and the administration of gadolinium-based contrast agents. It has occurred primarily in patients on dialysis, rarely in patients with very limited glomerular filtration rate (GFR) (i.e., <30 mL/min/1.73 m^2), and almost never in other patients. There is growing literature regarding NSF. Although some controversy and lack of clarity

remain, there is a consensus that it is advisable to avoid all gadolinium-based contrast agents in dialysis-dependent patients unless the possible benefits clearly outweigh the risk, and to limit the type and amount in patients with estimated GFR rates <30 mL/min/1.73 m^2. For more information, please see the American College of Radiology (ACR) Manual on Contrast Media (see the "Availability of Companion Documents" field).

Abbreviations

- CT, computed tomography
- CTA, computed tomography angiography
- HRCT, high-resolution CT
- ILD, interstitial lung disease
- MRA, magnetic resonance angiography
- MRI, magnetic resonance imaging
- Tc, technetium
- US, ultrasound
- V/Q, ventilation-perfusion

Relative Radiation Level Designations

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

Clinical Algorithm(s)

Algorithms were not developed from criteria guidelines.

Scope

Disease/Condition(s)

Pulmonary hypertension

Guideline Category

Diagnosis

Evaluation

Risk Assessment

Screening

Clinical Specialty

Cardiology

Family Practice

Internal Medicine

Pulmonary 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 patients with pulmonary hypertension

Target Population

Patients suspected of having or with pulmonary hypertension

Interventions and Practices Considered

1. Ultrasound (US) echocardiography
    - Transthoracic resting
    - Transesophageal
2. Right heart catheterization
3. X-ray chest
4. Computed tomography angiography (CTA) chest (noncoronary) with contrast
5. Technetium (Tc)-99m ventilation-perfusion (V/Q) scan lung
6. Magnetic resonance imaging (MRI) heart function and morphology
    - Without contrast
    - Without and with contrast
7. Magnetic resonance angiography (MRA) pulmonary arteries
    - Without and with contrast
    - Without contrast
8. Pulmonary arteriography with right heart catheterization
9. Computed tomography (CT) chest
    - Without contrast
    - Without and with contrast

Major Outcomes Considered

Methodology

Methods Used to Collect/Select the Evidence

Searches of Electronic Databases

Description of Methods Used to Collect/Select the Evidence

Literature Search Procedure

The Medline literature search is based on keywords provided by the topic author. The two general classes of keywords are those related to the condition (e.g., ankle pain, fever) and those that describe the diagnostic or therapeutic intervention of interest (e.g., mammography, MRI).

The search terms and parameters are manipulated to produce the most relevant, current evidence to address the American College of Radiology Appropriateness Criteria (ACR AC) topic being reviewed or developed. Combining the clinical conditions and diagnostic modalities or therapeutic procedures narrows the search to be relevant to the topic. Exploding the term "diagnostic imaging" captures relevant results for diagnostic topics.

The following criteria/limits are used in the searches:

1. Articles that have abstracts available and are concerned with humans.
2. Restrict the search to the year prior to the last topic update or in some cases the author of the topic may specify which year range to use in the search. For new topics, the year range is restricted to the last 5 years unless the topic author provides other instructions.
3. May restrict the search to Adults only or Pediatrics only.
4. Articles consisting of only summaries or case reports are often excluded from final results.

The search strategy may be revised to improve the output as needed.

Number of Source Documents

The total number of source documents identified as the result of the literature search is not known.

Methods Used to Assess the Quality and Strength of the Evidence

Weighting According to a Rating Scheme (Scheme Given)

Rating Scheme for the Strength of the Evidence

Strength of Evidence Key

Category 1 - The conclusions of the study are valid and strongly supported by study design, analysis, and results.

Category 2 - The conclusions of the study are likely valid, but study design does not permit certainty.

Category 3 - The conclusions of the study may be valid, but the evidence supporting the conclusions is inconclusive or equivocal.

Category 4 - The conclusions of the study may not be valid because the evidence may not be reliable given the study design or analysis.

Methods Used to Analyze the Evidence

Review of Published Meta-Analyses

Systematic Review with Evidence Tables

Description of the Methods Used to Analyze the Evidence

The topic author drafts or revises the narrative text summarizing the evidence found in the literature. American College of Radiology (ACR) staff draft an evidence table based on the analysis of the selected literature. These tables rate the strength of the evidence for all articles included in the narrative text.

The expert panel reviews the narrative text, evidence table, and the supporting literature for each of the topic-variant combinations and assigns an appropriateness rating for each procedure listed in the table. Each individual panel member forms his/her own opinion based on his/her interpretation of the available evidence.

More information about the evidence table development process can be found in the ACR Appropriateness Criteria® Evidence Table Development document (see the "Availability of Companion Documents" field).

Methods Used to Formulate the Recommendations

Expert Consensus (Delphi)

Description of Methods Used to Formulate the Recommendations

Modified Delphi Technique

The appropriateness ratings for each of the procedures included in the Appropriateness Criteria topics are determined using a modified Delphi methodology. A series of surveys are conducted to elicit each panelist's expert interpretation of the evidence, based on the available data, regarding the appropriateness of an imaging or therapeutic procedure for a specific clinical scenario. American College of Radiology (ACR) staff distributes surveys to the panelists along with the evidence table and narrative. Each panelist interprets the available evidence and rates each procedure. The surveys are completed by panelists without consulting other panelists. The ratings are a scale between 1 and 9, which is further divided into three categories: 1, 2, or 3 is defined as "usually not appropriate"; 4, 5, or 6 is defined as "may be appropriate"; and 7, 8, or 9 is defined as "usually appropriate." Each panel member assigns one rating for each procedure per survey round. The surveys are collected and the results are tabulated, de-identified and redistributed after each round. A maximum of three rounds are conducted. The modified Delphi technique enables each panelist to express individual interpretations of the evidence and his or her expert opinion without excessive bias from fellow panelists in a simple, standardized and economical process.

Consensus among the panel members must be achieved to determine the final rating for each procedure. Consensus is defined as eighty percent (80%) agreement within a rating category. The final rating is determined by the median of all the ratings once consensus has been reached. Up to three rating rounds are conducted to achieve consensus.

If consensus is not reached, the panel is convened by conference call. The strengths and weaknesses of each imaging procedure that has not reached consensus are discussed and a final rating is proposed. If the panelists on the call agree, the rating is accepted as the panel's consensus. The document is circulated to all the panelists to make the final determination. If consensus cannot be reached on the call or when the document is circulated, "No consensus" appears in the rating column and the 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.

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

Selection of appropriate radiologic imaging procedures for evaluation of patients with pulmonary hypertension

Potential Harms

- Limitations of Doppler echocardiography include acoustic window restrictions (particularly in patients with underlying lung disease), limitations due to body habitus, and operator dependence.
- At experienced institutions, right heart catheterization has morbidity and mortality rates of 1.1% and 0.055%, respectively.
- Limitations of MRI include motion and respiratory artifacts that may degrade image quality particularly for certain motion sensitive sequences, long acquisition times, and the need for sedation in patients with claustrophobia.

Gadolinium-based Contrast Agents

Nephrogenic systemic fibrosis (NSF) is a disorder with a scleroderma-like presentation and a spectrum of manifestations that can range from limited clinical sequelae to fatality. It appears to be related to both underlying severe renal dysfunction and the administration of gadolinium-based contrast agents. It has occurred primarily in patients on dialysis, rarely in patients with very limited glomerular filtration rate (GFR) (i.e., <30 mL/min/1.73 m^2), and almost never in other patients. Although some controversy and lack of clarity remain, there is a consensus that it is advisable to avoid all gadolinium-based contrast agents in dialysis-dependent patients unless the possible benefits clearly outweigh the risk, and to limit the type and amount in patients with estimated GFR rates <30 mL/min/1.73 m^2. For more information, please see the American College of Radiology (ACR) Manual on Contrast Media (see the "Availability of Companion Documents" field).

Relative Radiation Level (RRL)

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 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. Additional information regarding radiation dose assessment for imaging examinations can be found in the ACR Appropriateness Criteria® Radiation Dose Assessment Introduction document (see the "Availability of Companion Documents" field).

Contraindications

Contraindications

- The major contraindication to magnetic resonance imaging (MRI) is the presence of specific ferromagnetic and/or conducting implants such as cardiac pacemakers.
- Contraindications to intravenous gadolinium chelate contrast include allergy to gadolinium or renal dysfunction.

Qualifying Statements

Qualifying Statements

The 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 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 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.

Institute of Medicine (IOM) National Healthcare Quality Report Categories

IOM Care Need

Getting Better

Living with Illness

IOM Domain

Effectiveness

Identifying Information and Availability

Bibliographic Source(s)

Brown K, Gutierrez AJ, Mohammed TL, Kirsch J, Chung JH, Dyer DS, Ginsburg ME, Heitkamp DE, Kanne JP, Kazerooni EA, Ketai LH, Parker JA, Ravenel JG, Saleh AG, Shah RD, Steiner RM, Suh RD, Expert Panel on Thoracic Imaging. ACR Appropriateness CriteriaÂ®

pulmonary hypertension. [online publication]. Reston (VA): American College of Radiology (ACR); 2012. 10 p. [90 references]

Adaptation

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

Date Released

2012

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 Thoracic Imaging

Composition of Group That Authored the Guideline

Panel Members: Kathleen Brown, MD (*Principal Author*); Antonio J. Gutierrez, MD (*Research Author*); Tan-Lucien H. Mohammed, MD (*Panel Chair*); Jacobo Kirsch, MD (*Panel Vice-chair*); Jonathan H. Chung, MD; Debra Sue Dyer, MD; Mark E. Ginsburg, MD; Darel E. Heitkamp, MD; Jeffrey P. Kanne, MD; Ella A. Kazerooni, MD; Loren H. Ketai, MD; J. Anthony Parker, MD, PhD; James G. Ravenel, MD; Anthony G. Saleh, MD; Rakesh D. Shah, MD; Robert M. Steiner, MD; Robert D. Suh, MD

Financial Disclosures/Conflicts of Interest

Not stated

Guideline Status

This is the current release of the guideline.

Guideline Availability

Electronic copies: Available from the American College of Radiology (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®. Overview. 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®. Literature search process. Reston (VA): American College of Radiology; 1 p. Electronic copies: Available in PDF from the ACR Web site.
- ACR Appropriateness Criteria®. Evidence table development – diagnostic studies. Reston (VA): American College of Radiology; 2013 Nov. 3 p. Electronic copies: Available in PDF from the ACR Web site.
- ACR Appropriateness Criteria®. Radiation dose assessment introduction. Reston (VA): American College of Radiology; 2 p. Electronic copies: Available in PDF from the ACR Web site.
- ACR Appropriateness Criteria®. Manual on contrast media. Reston (VA): American College of Radiology; 90 p. Electronic copies: Available in PDF from the ACR Web site.
- ACR Appropriateness Criteria®. Procedure information. Reston (VA): American College of Radiology; 1 p. Electronic copies: Available in PDF from the ACR Web site.
- ACR Appropriateness Criteria® pulmonary hypertension. Evidence table. Reston (VA): American College of Radiology; 2012. 37 p. Electronic copies: Available from the ACR Web site.

Patient Resources

None available

NGC Status

This NGC summary was completed by ECRI Institute on May 9, 2013.

Copyright Statement

Instructions for downloading, use, and reproduction of the American College of Radiology (ACR) Appropriateness Criteria® may be found on the ACR Web site.

Disclaimer

NGC Disclaimer

The National Guideline Clearinghouseâ„¢ (NGC) does not develop, produce, approve, or endorse the guidelines represented on this site.

All guidelines summarized by NGC and hosted on our site are produced under the auspices of medical specialty societies, relevant professional associations, public or private organizations, other government agencies, health care organizations or plans, and similar entities.

Guidelines represented on the NGC Web site are submitted by guideline developers, and are screened solely to determine that they meet the NGC Inclusion Criteria which may be found at http://www.guideline.gov/about/inclusion-criteria.aspx.

NGC, AHRQ, and its contractor ECRI Institute make no warranties concerning the content or clinical efficacy or effectiveness of the clinical practice guidelines and related materials represented on this site. Moreover, the views and opinions of developers or authors of guidelines represented on this site do not necessarily state or reflect those of NGC, AHRQ, or its contractor ECRI Institute, and inclusion or hosting of guidelines in NGC may not be used for advertising or commercial endorsement purposes.

Readers with questions regarding guideline content are directed to contact the guideline developer.