American College of Radiology
ACR Appropriateness Criteria®
Suspected Pulmonary Hypertension

Variant 1: **Suspected 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
Catheterization right heart	9	Catheterization and echocardiography are complementary examinations. Both should be performed. Echocardiography is typically performed before catheterization.	☢☢
X-ray chest	9	X-ray chest is usually performed during the initial workup/screening of suspected pulmonary hypertension and is often the first test performed.	☢
CTA chest with IV contrast	8	This procedure is equivalent to CT chest with IV contrast. The examination choice of CTA chest with IV contrast or CT chest with IV contrast depends on institutional preference.	☢☢☢
Tc-99m V/Q scan lung	8	This procedure is the examination of choice to evaluate for CTEPH.	☢☢☢
CT chest with IV contrast	7	This procedure is equivalent to CTA chest with IV contrast. The examination choice of CTA chest with IV contrast or CT chest with IV contrast depends on institutional preference.	☢☢☢
MRI heart function and morphology without IV contrast	6		O
MRI heart function and morphology without and with IV contrast	6		O
MRA chest without and with IV contrast	5	CTA chest with IV contrast has better sensitivity for detection of CTEPH.	O
US echocardiography transesophageal	5		O
CT chest without IV contrast	4	If there is a concern for an occult interstitial lung disease, HRCT may be appropriate.	☢☢☢
Arteriography pulmonary with right heart catheterization	3	Noninvasive imaging is generally performed instead. This procedure can be performed when considering surgical or percutaneous embolectomy.	☢☢☢☢
MRA chest without IV contrast	2		O
FDG-PET/CT heart	2		☢☢☢☢
CT chest without and with IV contrast	1		☢☢☢
Rating Scale: **1,2,3 Usually not appropriate; 4,5,6 May be appropriate; 7,8,9 Usually appropriate**			***Relative Radiation Level**

SUSPECTED PULMONARY HYPERTENSION

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Summary of Literature Review

Introduction/Background

Pulmonary hypertension (PH), defined by a mean pulmonary arterial pressure ≥25 mm Hg at rest (measured at right heart catheterization [RHC]), may be idiopathic or may be related to a large variety of diseases. Normal mean pulmonary arterial pressure at rest is 14 to 20 mm Hg. Mean pulmonary arterial pressure between 21 and 24 mm Hg is of uncertain clinical significance but warrants close follow-up [1,2]. PH will lead to right ventricular failure and subsequent death if left untreated [1-4].

The term *pulmonary arterial hypertension* (PAH) is used to describe a population of PH patients who have precapillary PH (pulmonary artery wedge pressure ≤15 mm Hg and pulmonary vascular resistance >3 Wood units) in the absence of other causes of precapillary PH such as chronic thromboembolic pulmonary hypertension (CTEPH) [2].

A series of global meetings has been critical in the evolution of understanding PH, as well as in developing the clinical classification of PH. The first World Symposium on Pulmonary Hypertension was held in 1973 in Geneva, Switzerland [5]. Since 1973, several world symposia on PH have taken place (Evian, France, in 1998; Venice, Italy, in 2003; Dana Point, California, in 2008), resulting in various updates to the clinical classification [6-8]. The clinical classification was most recently updated at the fifth World Symposium on Pulmonary Hypertension, held in Nice, France, in 2013 [2,9].

The 2013 updated clinical classification of PH includes Group 1, PAH; Group 1′, pulmonary venoocclusive disease (PVOD) and/or pulmonary capillary hemangiomatosis (PCH); Group 1″, persistent PH of the newborn; Group 2, PH due to left heart disease; Group 3, PH due to lung diseases and/or hypoxia; Group 4, CTEPH and other pulmonary artery obstructions; and Group 5, PH with unclear and/or multifactorial mechanisms [2] (see Appendix 1).

The signs and symptoms of PH are nonspecific and may include fatigue, dyspnea, weakness, angina, peripheral edema, hepatomegaly, ascites, and syncope [10,11]. Because of the nonspecific symptoms as well as the large, diverse group of diseases that can cause PH, diagnosis 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 [11,12]. Clinical evaluation includes pulmonary function tests, arterial blood gases, routine biochemistry, hematology, thyroid function, and serological testing to evaluate for lung disease, liver disease, connective tissue disorders, and HIV [12]. Imaging examinations that may aid in the diagnosis of PH include chest radiography (CXR), ultrasound (US) echocardiography, ventilation/perfusion (V/Q) scans, computed tomography (CT), magnetic resonance imaging (MRI), RHC, pulmonary angiography, and fluorine-18-2-fluoro-2-deoxy-D-glucose positron emission tomography/CT (FDG-PET/CT).

Overview of Imaging Modalities

Chest radiography

CXR is often the first imaging test performed because the clinical presentation of PH is nonspecific. Multiple historic studies have demonstrated that CXR is an appropriate study in the initial evaluation of PH [13-19].

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Additionally, Miniati et al [20] showed high sensitivity (96.9%) and specificity (99.1%) for detection of PH by CXR. However, CXR is known to be insensitive in the detection of mild PH. Thus, a normal CXR does not exclude PH and further imaging evaluation should be pursued if there are persistent unexplained symptoms such as dyspnea or risk factors for PH [2,14,21,22].

Findings of PH on CXR include enlargement of the central pulmonary arteries, with or without rapid tapering (pruning), and right heart chamber enlargement [3,10,14,22,23]. A measurement of the right interlobar artery >15 mm in women (>16 mm in men) at the hilum is suggestive of PH [10]. In 1 prospective study on patients with PAH, the CXR demonstrated prominence of the main pulmonary artery in 90% of patients, enlarged hilar vessels in 80%, and decreased peripheral vessels in 51%. All 3 of these abnormalities were seen in 42% of the patients in the study [24]. Another study by Schmidt et al [17] on patients with PH from chronic thromboembolic disease showed an enlarged main pulmonary artery in 96% of patients. Additionally, CXR may show findings of diffuse lung diseases that can be associated with PH, such as interstitial lung disease, emphysema, etc [10,21].

Echocardiography transthoracic resting

Transthoracic Doppler echocardiography should always be performed in the initial evaluation whenever PH is suspected. Doppler echocardiography has a sensitivity of 79% to 100% and specificity of 68% to 98% in detecting moderate PH. However, detection of mild PH is more limited [14,25]. Continuous-wave Doppler measurement of the peak tricuspid regurgitation velocity is used in combination with additional echocardiographic signs suggestive of PH to assign an echocardiographic probability of PH (low, intermediate, or high) [2,26]. Additional echocardiographic variables that may be suggestive of PH include diameter of the pulmonary artery, estimate of right atrial pressure, increased right atrial size, abnormal bowing and motion of the interventricular septum, blood flow velocity and pattern out of the right ventricle, right ventricular hypertrophy, and increased right ventricle size compared with the left ventricle [10,27-30].

Transthoracic Doppler echocardiography is also useful for evaluating cardiac anatomy, 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 [11]. Echocardiographic evaluation of the right ventricular myocardial performance index and tricuspid annular plane systolic excursion index should be measured concomitantly with mean pulmonary artery pressure (PAP), as PAP in patients with advanced PH may decrease with deterioration of right ventricular function [31]. Studies have shown that real-time 3-D echocardiography is superior in evaluation of right ventricular volumes and ejection fraction compared with conventional 2-D echocardiography [29,32].

RHC is necessary for confirmation of PH in patients with intermediate or high echocardiographic probability of PH prior to treatment initiation. As mentioned above, transthoracic Doppler echocardiography is not reliable to screen for mild, asymptomatic PH. Additional limits of transthoracic Doppler echocardiography include acoustic window restrictions (particularly in patients with underlying lung disease), limitations due to body habitus, and operator dependence. Further evaluation with additional noninvasive examinations including CT and MRI may be obtained. RHC is available for further evaluation if noninvasive examinations fail to yield a diagnosis [2,10].

Patients at high risk for development of PAH may benefit from screening using transthoracic Doppler echocardiography [11]. At-risk patients include 1) individuals with a known genetic mutation associated with PAH or a first-degree relative with idiopathic PAH (IPAH), 2) patients with scleroderma spectrum of disease, 3) patients with congenital heart disease and systemic-to-pulmonary shunts, and 4) patients with portal hypertension prior to liver transplant [33]. However, as noted above, mild, asymptomatic PH is not reliably detected by Doppler echocardiography, and if there is high clinical suspicion, RHC may be pursued [2,28].

Transesophageal echocardiography

Transesophageal echocardiography (TEE) is a more invasive technique than resting transthoracic echocardiography (TTE). It requires conscious sedation. Although complications of TEE are rare when performed by an experienced echocardiographer, pharyngeal and esophageal trauma have been reported. In general, PH is well assessed by resting TTE. TEE may be considered to further evaluate for the presence of congenital shunts such as sinus venosus defect and anomalous pulmonary venous return. However, noninvasive techniques such as cardiac MRI and CT can also easily assess these entities and are recommended over TEE [2].

Ventilation/perfusion scans

The algorithm recommended by the American College of Cardiology Working Group recommends V/Q scanning in all patients with unexplained PH, primarily to assess for CTEPH [2,34]. V/Q scans are the examination of choice in evaluating for CTEPH and differentiating CTEPH from other causes of PH [35,36]. V/Q scanning demonstrated a sensitivity of 90% to 100% and specificity of 94% to 100% for differentiation between IPAH and CTEPH [37]. 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 [2,10].

A study by Tunariu et al [36] 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 with 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 [38]. MR can also be used to also assess ventilation and perfusion in centers with experience [39].

Computed tomography and computed tomography angiography

Both CT and CTPA (CT angiography [CTA] chest/CTPA chest) are very useful studies in the evaluation of PH and characterization of associated changes in the pulmonary parenchyma as well as the cardiovascular system. A main pulmonary artery diameter of ≥29 mm has been shown to be 87% sensitive and 89% specific, with a positive predictive value (PPV) of 97%, for PH [3,40]. However, additional studies have shown that the sensitivity and specificity of main pulmonary artery diameter can vary depending on the presence of lung disease, and a main pulmonary artery diameter <29 mm does not exclude PH [41-43]. PH is almost always present when the main pulmonary artery is larger than the adjacent ascending aorta (PPV of 96%) [14,22]. Other findings suggesting PH on CT/CTPA include a ratio of segmental pulmonary artery to accompanying bronchus >1:1, mosaic attenuation of the lungs, pericardial thickening or effusion, enlargement of the right ventricle, and straightening of the interventricular septum. Enlargement of the bronchial arteries to a diameter of >1.5 mm can also be seen in patients with PH [3,22,40,44-46]. Extrinsic compression of the left main coronary artery by a dilated main pulmonary artery, an uncommon finding in PH, has also been reported on CT [47]. In end-stage PH, linear calcifications along central pulmonary artery walls compatible with atheromatous plaques may be present [14,22]. A small retrospective study by Chan et al [48] has shown that additional CT predictors of PH include right ventricular free wall thickness ≥6 mm, right ventricular lumen/left ventricular lumen ≥1.28, and true right and left descending pulmonary artery diameters of 16 mm and 21 mm, respectively. Subsequent evaluation with RHC is necessary for confirmation of PH.

There are many etiologies of PH, including chronic pulmonary embolism (PE), IPAH, PCH, PVOD, left-to-right shunts, and congenital heart disease, as well as many diffuse lung diseases. Many of these diseases are best characterized by cross-sectional imaging, particularly CT/CTPA, and many have additional specific imaging findings on CT/CTPA in addition to the findings compatible with PH that have been discussed above.

CTPA is regularly used to evaluate for thromboembolic disease and is the standard of care at most institutions [22,49]. When compared with V/Q scanning, it is also an accurate diagnostic modality for CTEPH, with a sensitivity of 83% to 100% and a specificity of 89% to 97% [35,50]. CT findings of CTEPH include CT findings of PH (detailed above), as well as findings of chronic PE, which include eccentric thrombus within the pulmonary arteries, recanalized thrombus with or without calcification, abrupt cut-off and narrowing of an affected pulmonary artery, and thin linear webs within the affected arteries. Mosaic attenuation of the lung parenchyma is often present as well, with decreased vessel size in the areas of low attenuation [22,51-58]. Enlarged bronchial arteries and systemic arteries are commonly present [35]. Bronchiectasis has also been reported [59]. Small studies have shown that dual-energy CT perfusion and angiography is also both highly sensitive and specific in diagnosis of CTEPH and can assess both anatomy and perfusion in CTEPH [60-62].

IPAH is an uncommon disease characterized by a plexiform lesion, which is a network of capillary-like channels in the wall of a dilated muscular pulmonary artery. CT findings of IPAH include enlarged pulmonary arteries, small pericardial effusion, and centrilobular ground-glass attenuation surrounding the torturous and enlarged centrilobular arterioles. Subtle mosaic attenuation may be present. Sometimes small centrilobular nodules can also be seen on CT [22,63-65].

There are 2 rare diseases (PCH and PVOD) associated with PH that can be suggested by findings on CT/CTPA. PCH is a diffuse proliferation of capillaries throughout the pulmonary interstitium that is associated with obstruction of venules. CT will show enlarged pulmonary arteries, centrilobular ground-glass nodules, and interlobular septal thickening [14,66-68]. PVOD is a disease that results from pulmonary vein intimal fibrosis and subsequent pulmonary vein thrombosis. This causes pulmonary venous hypertension and edema and ultimately

PH as the increased pulmonary vascular pressure backs up into the main pulmonary artery. CT will show enlarged pulmonary arteries, pleural effusion, and interlobular septal thickening. The left atrium is normal in size, a key feature distinguishing PVOD from left heart mitral valve disease. Mediastinal lymph node enlargement has also been reported. PCH and PVOD both often require surgical biopsy for definitive diagnosis [14,66,69-71].

Congenital heart disease as well as left-to-right shunts including atrial septal defect, ventricular septal defect, patent ductus arteriosus, and anomalous pulmonary venous return can be diagnosed with CTPA [14], although MR is the preferred modality for these diseases.

Diffuse lung diseases associated with PH include interstitial lung disease, emphysema, sarcoidosis, connective tissue disease, and pulmonary Langerhans cell histiocytosis. High-resolution CT (HRCT) is indicated for evaluating diffuse lung disease and for the screening of patients with chronic unexplained dyspnea (see the ACR Appropriateness Criteria® "Chronic Dyspnea—Suspected Pulmonary Origin" [72] and "Dyspnea—Suspected Cardiac Origin" [73]). In many cases, HRCT may obviate the need for open lung biopsy.

For the purposes of distinguishing between CT and CT angiography (CTA), American College of Radiology (ACR) Appropriateness Criteria topics use the definition in the Practice Parameter for the Performance and Interpretation of Body Computed Tomography Angiography (CTA) [74]:

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

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

Magnetic resonance imaging and magnetic resonance angiography

MRI is another excellent noninvasive imaging study for the evaluation of PH and is the best noninvasive imaging examination for the evaluation of right ventricular morphology and function. MRI should be performed at centers with experience. Both cardiac MRI and pulmonary MR angiography (MRA) show morphologic findings of PH similar to those seen on CT/CTPA: pulmonary artery enlargement and pruning of peripheral vasculature on MRA and right ventricular hypertrophy and dilation as well as straightening of the interventricular septum on cardiac MRI [1,4,38,75-78]. The combination of MRA and MR perfusion imaging of the lung can also reliably diagnose CTEPH [1,4,35,38,79]. MRI and MRA are often used to follow patients with known PH to assess for early changes in right ventricular function.

MR cine imaging is the gold standard to evaluate right ventricular function and size and can evaluate for right ventricular wall motion changes seen in PH. Right and left ventricular mass can also be accurately determined [80], which can then be used to calculate a ventricular mass index (right ventricular mass/left ventricular mass). A ventricular mass index that is >0.6 is compatible with PH [3,81]. Functional abnormalities seen in cardiac remodeling secondary to PH include right ventricular hypokinesis, leftward bowing and/or paradoxical movement of the interventricular septum, right ventricular dysfunction (increased end-diastolic volume, reduced ejection fraction, reduced cardiac index, reduced stroke volume), and pulmonary and tricuspid insufficiency [1,4,77,81-85].

Phase-contrast imaging techniques can measure average blood flow velocity of the main pulmonary artery, which correlates with mean pulmonary arterial pressure. Decreased pulmonary artery blood flow velocity correlates to increased vascular resistance [10,81]. A small study by Kreitner et al [86] has suggested that estimation of mean pulmonary arterial pressure from high-temporal-resolution phase-contrast MRI is possible, but further investigation in a larger population is still needed. Heterogeneous flow in the main pulmonary artery as well as decreased pulmonary artery distensibility can be seen in PH. Pulmonary artery distensibility is the change in the pulmonary artery area throughout the cardiac cycle. A decrease in pulmonary artery distensibility is an early sign of increased pulmonary vascular resistance and poor outcomes in PH patients [10,80,81,83,87,88]. Additionally, it can measure right ventricular stroke volume and cardiac output and can quantify intracardiac and extracardiac shunts by measuring the pulmonary to systemic flow ratio [1,3,4,22,89]. MRI is an excellent tool (sensitivity of 93% to 100% and specificity of 87% to 100%) to detect and quantify cardiovascular shunts that are difficult to identify on echocardiography, including sinus venosus atrial septal defects, patent ductus arteriosus, and anomalous pulmonary venous return [14,90-92].

Delayed-contrast MRI typically shows enhancement of the right ventricle at its insertion points into the interventricular septum in PH [1,3,80,81,83]. Recently, 4-D flow MRI techniques have been used to noninvasively measure hemodynamic alterations that occur with PH: decreased wall shear stress, increased tricuspid regurgitation velocity, and abnormal vortex blood flow pattern within the main pulmonary artery that is associated with early-onset systolic retrograde flow [93-96]. Small studies have shown that MRI can also be used as a noninvasive method for obtaining functional information to monitor treatment response and the long-term effects of vasodilator therapy [22,97,98].

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 requires. 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 [10,99]. Contraindications to intravenous (IV) gadolinium chelate contrast, which is required for certain sequences, include allergy to gadolinium and renal dysfunction [100].

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 with experience. 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 RHCs.

Right heart catheterization

RHC is the gold standard for the diagnosis of PAH and is typically performed after all other noninvasive examinations have been completed to confirm the diagnosis of PH as well as assess its severity. At experienced institutions, it has morbidity and mortality rates of 1.1% and 0.055%, respectively [2,11,101].

RHC directly measures PAP and cardiac function. A pulmonary vascular resistance of >3 Wood units is necessary to establish a diagnosis of PAH. Vasoreactivity testing of the pulmonary circulation may also be performed at the time of RHC in selected patients with IPAH, heritable PAH, and drug-induced PAH to determine candidacy for calcium channel blocker treatment [2,102].

Pulmonary angiography

Prior to multidetector CT, catheter pulmonary angiography was considered the reference standard for assessing PE. Studies have demonstrated that CTPA is as reliable as digital subtraction angiography in the evaluation of CTEPH [55]. Findings of CTEPH on CTPA and catheter pulmonary angiography include webs or bands with or without stenotic dilatation, intimal irregularities, and abrupt narrowing or occlusion of segmental or larger vessels. Catheter pulmonary angiography is now used almost exclusively for thrombolysis.

Fluorine-18-2-fluoro-2-deoxy-D-glucose positron emission tomography/computed tomography

FDG-PET/CT allows *in vivo* imaging of metabolic processes and is complementary to the structural/anatomic information provided by cross-sectional imaging modalities such as CT and MRI. FDG-PET/CT is well established for the diagnosis and follow-up of malignancy but is also becoming a valuable imaging modality for the characterization and diagnosis of various inflammatory conditions [103,104]. The use of FDG-PET/CT in the evaluation of PH is extremely limited. There is increased FDG uptake in the lungs of patients with IPAH as well as within the right ventricular myocardium in patients with right ventricular dysfunction secondary to PH [83,105-107]. A study by Tatebe et al [108] shows poorer prognosis in PH patients with a standardized uptake value of ≥8.3 in the region of the right ventricular free wall. Gated FDG-PET/CT is an available method that evaluates both right ventricular function and myocardial glucose metabolism [109]. The main potential utility of FDG-PET/CT in patients with PH is in distinguishing rare mimics of CTEPH, including pulmonary artery sarcoma and medium- to large-vessel vasculitis such as Takayasu arteritis, both of which will demonstrate increased FDG uptake [3,110-112]. Differentiating CTEPH from these rare mimics is critical because of important treatment implications [113,114].

Summary of Recommendations

- CXR is an appropriate study in the initial diagnostic evaluation of PH, based on multiple studies. It has an overall high sensitivity and specificity for detecting the presence of PH. However, CXR is known to be insensitive in the detection of mild PH, and therefore a normal CXR cannot exclude it. Further imaging evaluation is recommended if the CXR is normal and there are persistent unexplained symptoms such as dyspnea or other risk factors for PH.
- Resting TTE is the screening test of choice in the initial evaluation of suspected PH and should always be performed in the evaluation of suspected PH. Similar to CXR, it can be limited in the evaluation of mild, asymptomatic PH.
- TEE is more invasive than TTE. Although it can also evaluate for congenital shunts in addition to PH, noninvasive studies such as CT and MRI can do this as well and are recommended over TEE.
- V/Q scans should be obtained in all patients with PH to assess for CTEPH. A V/Q scan is the examination of choice to evaluate for CTEPH. CTA (CTPA) and MRA are additional tools that are also available to evaluate for CTEPH.
- CT chest with IV contrast and CTA (CTPA) chest with IV contrast are excellent noninvasive imaging examinations that can characterize findings of PH as well as often suggest an underlying cause. In some situations (for example, if there is suspicion for occult interstitial lung disease), an HRCT without IV contrast may also be useful.
- MRI is an excellent noninvasive imaging examination that readily characterizes findings associated with PH. It is the best available noninvasive examination to evaluate right ventricular morphology and function, which are closely associated with PH prognosis.
- RHC is the gold standard for the diagnosis of PH and should be performed in all cases of suspected PH after noninvasive examinations are completed to confirm the diagnosis prior to treatment initiation.
- Catheter pulmonary angiography is now primarily reserved for thrombolysis. It can be performed when considering surgical or percutaneous embolectomy.
- The use of FDG-PET/CT in evaluating PH is extremely limited. Its main potential utility is in distinguishing rare mimics of CTEPH, such as pulmonary artery sarcoma.

Summary of Evidence

Of the 114 references cited in the *ACR Appropriateness Criteria® Suspected Pulmonary Hypertension* document, all are categorized as diagnostic references, including 12 good-quality studies and 22 quality studies that may have design limitations. There are 78 references that may not be useful as primary evidence. There are 2 references that are meta-analysis studies.

The 114 references cited in the *ACR Appropriateness Criteria® Suspected Pulmonary Hypertension* document were published from 1962 through 2016.

Although there are references that report on studies with design limitations, 12 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**
O	0 mSv	0 mSv
☢	<0.1 mSv	<0.03 mSv
☢☢	0.1-1 mSv	0.03-0.3 mSv
☢☢☢	1-10 mSv	0.3-3 mSv
☢☢☢☢	10-30 mSv	3-10 mSv
☢☢☢☢☢	30-100 mSv	10-30 mSv

*RRL assignments for some of the examinations cannot be made, because the actual patient doses in these procedures vary as a function of a number of factors (eg, region of the body exposed to ionizing radiation, the imaging guidance that is used). The RRLs for these examinations are designated as "Varies".

Supporting Documents

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

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Appendix 1. Updated Clinical Classification of Pulmonary Hypertension [2,9]

Group 1. Pulmonary arterial hypertension (PAH)
1.1 Idiopathic PAH
1.2 Heritable PAH
1.2.1 *BMPR2* mutation
1.2.2 *ALK1*, *ENG*, *SMAD9*, *CAV1*, *KCNK3*
1.2.3 Unknown
1.3 Drug and toxin induced
1.4 Associated with:
1.4.1 Connective tissue disease
1.4.2 Human immunodeficiency virus (HIV) infection
1.4.3 Portal hypertension
1.4.4 Congenital heart disease
1.4.5 Schistosomiasis
Group 1′. Pulmonary venoocclusive disease and/or pulmonary capillary hemangiomatosis
1′.1 Idiopathic
1′.2 Heritable
1′.2.1 *EIF2AK4* mutation
1′.2.2 Other mutations
1′.3. Drug, toxin, and radiation induced
1′.4 Associated with:
1′.4.1 Connective tissue disease
1′.4.2 HIV infection
Group 1″. Persistent pulmonary hypertension of the newborn
Group 2. Pulmonary hypertension due to left heart disease
2.1 Left ventricular systolic dysfunction
2.2 Left ventricular diastolic dysfunction
2.3 Valvular disease
2.4 Congenital/acquired left heart inflow/outflow tract obstruction and congenital cardiomyopathies
2.5 Congenital/acquired pulmonary vein stenosis
Group 3. Pulmonary hypertension due to lung diseases and/or hypoxia
3.1 Chronic obstructive pulmonary disease
3.2 Interstitial lung disease
3.3 Other pulmonary diseases with mixed restrictive and obstructive pattern
3.4 Sleep-disordered breathing
3.5 Alveolar hypoventilation disorders
3.6 Chronic exposure to high altitude
3.7 Developmental lung diseases
Group 4. Chronic thromboembolic pulmonary hypertension and other pulmonary artery obstructions
4.1 Chronic thromboembolic pulmonary hypertension
4.2 Other pulmonary artery obstructions
4.2.1 Angiosarcoma
4.2.2 Other intravascular tumors
4.2.3 Arteritis
4.2.4 Congenital pulmonary artery stenoses
4.2.5 Parasites (hydatidosis)
Group 5. Pulmonary hypertension with unclear and/or multifactorial mechanisms
5.1 Hematological disorders: chronic hemolytic anemia, myeloproliferative disorders, splenectomy
5.2 Systemic disorders: sarcoidosis, pulmonary histiocytosis, lymphangioleiomyomatosis, neurofibromatosis
5.3 Metabolic disorders: glycogen storage disease, Gaucher disease, thyroid disorders
5.4 Others: pulmonary tumoral thrombotic microangiopathy, fibrosing mediastinitis, chronic renal failure (with/without dialysis), segmental pulmonary hypertension