American College of Radiology
ACR Appropriateness Criteria®
Suspected Pulmonary Hypertension

Variant: 1 Suspected pulmonary hypertension. Initial imaging.

Procedure	Appropriateness Category	Relative Radiation Level
US echocardiography transthoracic resting	Usually Appropriate	O
Radiography chest	Usually Appropriate	☢
CT chest with IV contrast	Usually Appropriate	☢☢☢
CTA chest with IV contrast	Usually Appropriate	☢☢☢
US echocardiography transesophageal	May Be Appropriate	O
MRI heart function and morphology without and with IV contrast	May Be Appropriate	O
MRI heart function and morphology without IV contrast	May Be Appropriate	O
V/Q scan lung	May Be Appropriate (Disagreement)	☢☢☢
Catheterization right heart	Usually Not Appropriate	☢☢
Arteriography pulmonary with right heart catheterization	Usually Not Appropriate	☢☢☢☢
MRA chest without and with IV contrast	Usually Not Appropriate	O
MRA chest without IV contrast	Usually Not Appropriate	O
CT chest without and with IV contrast	Usually Not Appropriate	☢☢☢
CT chest without IV contrast	Usually Not Appropriate	☢☢☢

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Summary of Literature Review

Introduction/Background

Pulmonary hypertension (PH), recently redefined in 2018 as a mean pulmonary arterial pressure (mPAP) >20 mm Hg at rest (measured at right heart catherization [RHC]), may be idiopathic or related to a large variety of diseases [1,2]. The term pulmonary arterial hypertension (PAH) is used to describe a population of patients with PH who have precapillary PH (pulmonary artery [PA] 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 PH (CTEPH), PA tumors, and various obstructive or restrictive lung diseases [2,3]. Left untreated, PH leads to right heart failure and death [4].

A series of global meetings has been critical in the evolution of understanding PH, as well as in developing a clinical classification for 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; Nice, France in 2013 and 2018), resulting in various updates to the clinical classification [2,6-9]. The 2018 updated clinical classification has been simplified and now includes group 1, PAH; group 2, PH due to left heart disease; group 3, PH due to lung diseases and/or hypoxia; group 4, PH due to PA obstructions; and group 5, PH with unclear and/or multifactorial mechanisms (see Appendix 1) [2]. The sixth World Symposium on Pulmonary Hypertension also defined specific criteria for precapillary PH, isolated postcapillary PH, and combined pre- and postcapillary PH using an mPAP >20 mm Hg in combination with pulmonary arterial wedge pressure (PAWP) and peripheral vascular resistance (PVR) measurements. Precapillary PH is characterized by mPAP >20 mm Hg, PAWP ≤15 mm Hg, and PVR ≥3 Wood units (groups 1, 3, 4, and 5). Isolated postcapillary PH is characterized by mPAP >20 mm Hg, PAWP >15 mm Hg, and PVR <3 Wood units (groups 2 and 5). Combined pre- and postcapillary PH is characterized by mPAP >20 mm Hg, PAWP >15 mm Hg, and PVR ≥3 Wood units (groups 2 and 5) [2].

Diagnosis of PH remains challenging because of the diverse group of diseases that can cause PH as well as its nonspecific symptoms. Signs and symptoms of PH include dyspnea, fatigue, palpitations, angina, peripheral edema, hepatomegaly, ascites, syncope, and, rarely, unilateral vocal cord paralysis [4,10]. A careful history evaluation 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, arterial blood gases, routine biochemistry, hematology, thyroid function, and serological testing to evaluate for lung disease, liver disease, connective tissue disorders, and HIV, as well as cardiothoracic imaging [10,11].

Special Imaging Considerations

For the purposes of distinguishing between CT and CT angiography (CTA), ACR Appropriateness Criteria topics use the definition in the ACR–NASCI–SIR–SPR Practice Parameter for the Performance and Interpretation of Body Computed Tomography Angiography (CTA) [12]:

"CTA 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 3-D renderings."

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

Initial Imaging Definition

Initial imaging is defined as imaging at the beginning of the care episode for the medical condition defined by the variant. More than one procedure can be considered usually appropriate in the initial imaging evaluation when:

- There are procedures that are equivalent alternatives (i.e., only one procedure will be ordered to provide the clinical information to effectively manage the patient's care)

OR

- There are complementary procedures (i.e., more than one procedure is ordered as a set or simultaneously wherein each procedure provides unique clinical information to effectively manage the patient's care).

Discussion of Procedures by Variant

Variant 1: Suspected pulmonary hypertension. Initial imaging.

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A. Radiography Chest

PH tends to present with nonspecific symptoms; thus, chest radiography is often the first imaging test performed [11]. Multiple historic studies have shown that chest radiography is a useful study in the initial evaluation of PH [13-19]. Miniati et al [20] found that chest radiography has a high sensitivity (96.9%) and specificity (99.1%) for detection of moderate to severe PH. Additionally, chest radiography can show findings of diffuse lung diseases that can be associated with PH, such as interstitial fibrosis and emphysema [10,13]. Although chest radiography does well in detecting the presence/absence of PH in moderate and severe cases of PH, it performs poorly in estimating the PH severity [14]. Additionally, it is known to be insensitive in the detection of mild PH. Thus, a normal chest radiograph 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 [13,15,17,20,21].

Findings of PH on chest radiography include enlargement of the central pulmonary arteries, with or without rapid tapering (pruning), and right heart chamber enlargement [10,17,19,21]. A study by Schmidt et al [17] found that the main PA (MPA) was enlarged (>35 mm from midline to left lateral border of the PA on posterior anterior radiograph) 96% of the time in PH. A measurement of the right descending PA >15 mm in women (>16 mm in men) at the hilum on a posterior anterior view has good sensitivity, specificity, and accuracy for PH (93%, 88%, and 92%, respectively) and is considered to be a very useful finding [16-18]. A diameter of the left descending PA >18 mm on the lateral view is also suggestive of PH, but although it has good sensitivity (93%), the specificity (67%) and accuracy (87%) are poorer [16]. Other studies, such as that of Schmidt et al [17], have found poorer sensitivity for the left descending PA measurement. Miniati et al [20] found the most prevalent radiographic findings in patients with PH were enlarged MPA (97%), enlarged right ventricle (95%), and enlarged right descending PA (93%).

Variant 1: Suspected pulmonary hypertension. Initial imaging.
B. US Echocardiography Transthoracic Resting

Transthoracic Doppler echocardiography is a noninvasive test that is a useful part of the initial evaluation of suspected PH [22]. A recent meta-analysis by Ni et al [23] showed that, overall, it has good sensitivity and fair specificity (85% and 74%, respectively) for detecting moderate to severe PH. However, it does not perform as well in detecting mild PH, particularly cases of PH secondary to lung diseases. Transthoracic Doppler echocardiography uses continuous wave Doppler to measure the peak tricuspid regurgitation velocity, which is used in combination with various echocardiographic signs suggestive of PH to assign a low, intermediate, or high echocardiographic probability of PH [24-26].

Echocardiographic signs suggestive of PH fall into 3 categories: right ventricular (RV) findings, PA findings, and inferior vena cava (IVC)/right atrial (RA) findings. RV findings include RV/left ventricular (LV) basal diameter ratio >1 and flattening of the interventricular septum. PA findings include PA diameter >25 mm, early diastolic pulmonary regurgitation velocity >2.2 m/s, and RV outflow Doppler acceleration time <105 ms and/or mid-systolic notching. IVC/RA findings include an IVC diameter >21 mm with <50% collapse with inspiration and end-systolic RA area >18 cm2 [1]. At least 2 categories (RV, PA, or IVC/RA) need to be positive to count toward the transthoracic Doppler echocardiography assigned PH probability. Low probability for PH is a peak tricuspid regurgitation velocity ≤2.8 m/s and no suggestive echocardiographic signs. Intermediate probability for PH is a peak tricuspid regurgitation velocity ≤2.8 m/s and presence of 2 categories of echocardiographic signs suggestive of PH or peak tricuspid regurgitation velocity 2.9 to 3.4 m/s without additional echocardiographic signs of PH. High probability for PH is a peak tricuspid regurgitation velocity of 2.9 to 3.4 m/s with 2 categories of echocardiographic signs of PH or >3.4 m/s without additional echocardiographic signs of PH [22,24,26,27]. Individuals who fall into intermediate or high probability for PH should have further evaluation with RHC to confirm PH before initiation of therapy per current guidelines [24,27].

Transthoracic Doppler echocardiography is also useful in the assessment of multiple right heart parameters that are influenced by PH, including RA and RV size, RV systolic function, RV strain, tricuspid annular plane systolic excursion, and biventricular index. A biventricular index (RV end-diastolic area to LV end-diastolic area) >0.93 is associated with an increased risk of death in patients with PH [22,27]. The presence of pericardial effusion as well as valvular morphology and function are also easily assessed by echocardiography. An echocardiographic bubble study using agitated saline can be performed during the examination to assess for intracardiac shunts [11]. Studies have shown that real-time 3-D echocardiography evaluates RV volumes and the ejection fraction more accurately than conventional 2-D echocardiography [28]. In addition, pressure gradient-volume diagrams derived from 3-D echocardiography data can be used to reliably estimate RV stroke work in patients with PH [29].

Populations including those with a known genetic mutation associated with PAH, first degree relative with PAH, scleroderma spectrum, congenital heart disease, and portal hypertension before liver transplant are at high risk of developing PAH and may benefit from screening with echocardiography [30]. Patients with a high echocardiographic probability for PH have poorer postsurgical outcomes; thus, PH echocardiographic probability can also be used to risk stratify patients with risk factors for PH before a procedure/surgery [27,31]. As mentioned above, transthoracic Doppler echocardiography will not reliably detect mild, asymptomatic PH. Further evaluation with additional noninvasive examinations including CT and MRI may be obtained if there is persistent, high clinical suspicion for PH. RHC is also useful for further evaluation [10,24].

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C. US Echocardiography Transesophageal

Transesophageal echocardiography (TEE) is more useful in assessing PH than transthoracic echocardiography; however, it is a more invasive technique and requires conscious sedation. An MPA to ascending aorta ratio ≥1 on TEE has a sensitivity and specificity for PH of 84% and 83%, respectively [32]. TEE can also further evaluate intracardiac 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 still recommended over TEE [24].

Variant 1: Suspected pulmonary hypertension. Initial imaging.
D. V/Q Scan Lung

There are no data to suggest ventilation-perfusion (V/Q) scintigraphy as an initial test in the workup of suspected PH. However, current guidelines recommend V/Q scintigraphy of the lungs in all patients with unexplained PH to assess for CTEPH [24]. Identification of CTEPH is important because it allows for potentially curative surgical therapy [33]. V/Q scintigraphy typically shows mismatched wedge-shaped, segmental defects in the setting of CTEPH that can normalize after surgical treatment [34].

The high sensitivity and specificity of V/Q scintigraphy is useful for CTEPH detection [34]. A recent study by Mehari et al [35] comparing V/Q scintigraphy and multidetector CT pulmonary angiography (CTPA) for CTEPH detection found that V/Q scintigraphy had excellent sensitivity (90%) and good specificity (75%), whereas CTPA suffered from poor sensitivity (37%), although it had excellent specificity (100%). This is comparable to a prior study by Tunariu et al [36], which found that V/Q scintigraphy was more sensitive than multidetector CTPA in detecting chronic thromboembolic pulmonary disease amenable to surgery, with V/Q scans demonstrating a sensitivity of 96% to 97% and a specificity of 90% to 95% compared with a sensitivity of 51% and specificity of 99% for multidetector CTPA. However, a study that compared V/Q scanning versus CTPA by He et al [37] showed both techniques had good sensitivity, specificity, and accuracy at a center with significant CTEPH experience (100% versus 92%, 94% versus 95%, and 97% versus 94%, respectively). Compared with planar V/Q scanning, V/Q single-photon emission CT improves sensitivity and specificity, resulting in an accuracy of 94% [38]. The slightly lower specificity of V/Q scintigraphy relative to multidetector CTPA is secondary in patients with idiopathic PAH (IPAH) in whom abnormal V/Q scans can also be demonstrated whom can also demonstrate abnormal V/Q scans [33,38].

Although V/Q scintigraphy has excellent sensitivity for CTEPH, its greatest power lies with its negative predictive value. A normal or low-probability scan essentially excludes the diagnosis of CTEPH, although the V/Q scan may be normal in other causes of PH [10,24,33].

Variant 1: Suspected pulmonary hypertension. Initial imaging.
E. CT Chest

CT chest is another noninvasive technique that can assess for PH. CT chest can be performed with or without intravenous (IV) contrast but is ideally performed with IV contrast to adequately visualize the pulmonary vasculature. CT chest without IV contrast is not a first-line test for suspected PH but may be useful to help look for etiologies of PH in patients who already hold a diagnosis of PH. There are no data support obtaining a CT chest with and without IV contrast in the setting of suspected PH.

An MPA diameter of ≥29 mm on CT is 87% sensitive and 89% specific, with a positive predictive value (PPV) of 97% for PH [39]. However, although an MPA diameter of ≥29 mm has a good PPV for PH, not all who meet this criteria will have PH. In addition, it is well known that the presence of parenchymal lung disease (eg, interstitial lung disease) decreases the sensitivity and specificity of the MPA diameter, and thus, an MPA diameter <29 mm does not exclude PH when parenchymal lung disease is present [39,40]. The ratio of the MPA to the adjacent ascending aorta is an extremely sensitive CT finding for PH. PH is nearly always present when the MPA is larger than the adjacent ascending aorta (PPV of 96%) [39,41-43]. Additional findings of PH on CT include true

right and left descending PA diameters of 16 mm and 21 mm, respectively, a ratio of segmental PA to accompanying bronchus >1:1, enlargement of the right ventricle, RV lumen/LV lumen ≥1, straightening of the interventricular septum, RV free wall thickness ≥6 mm, pericardial thickening or effusion, and enlargement of the bronchial arteries to a diameter of >1.5 mm [42,44,45]. Linear calcification within the PA walls may be present in end-stage PH [21]. Evaluation with RHC remains necessary to confirm PH detected on CT before initiating therapy [24].

In addition to demonstrating findings suggestive of PH, chest CT can also characterize various pulmonary etiologies that cause PH, including IPAH, pulmonary capillary hemangiomatosis (PCH), pulmonary veno-occlusive disease (PVOD), and many diffuse lung diseases. IPAH is characterized by plexiform lesions, which are networks of capillary-like channels in the wall of a dilated muscular PA that appear as ground glass centrilobular nodules. IPAH findings on CT include dilated central pulmonary arteries, centrilobular ground glass nodules associated with enlarged tortuous centrilobular arterioles, and pericardial effusion [21,46]. PCH, a diffuse proliferation of capillaries in the pulmonary interstitium, and PVOD, a disease of pulmonary venous hypertension secondary to pulmonary vein intimal fibrosis and subsequent venous occlusion, are rare diseases whose diagnosis can be suggested by findings on CT. On CT, PCH will have enlarged pulmonary arteries, centrilobular ground glass nodules, and interlobular septal thickening. PVOD presents on CT with enlarged pulmonary arteries, lymphadenopathy, pleural effusion, and interlobular septal thickening in the setting of a normal size left atrium. Surgical biopsy is necessary to confirm the diagnosis of PCH and PVOD [46-48].

Finally, there are many diffuse lung diseases including interstitial lung disease, emphysema, sarcoidosis, connective tissue diseases, and pulmonary Langerhans cell histiocytosis, which can cause PH. These diffuse lung diseases are best characterized by high-resolution CT, which is indicated for evaluating chronic dyspnea (see the ACR Appropriateness Criteria® topics on "Chronic Dyspnea-Noncardiovascular Origin" [49] and "Dyspnea-Suspected Cardiac Origin" [50]).

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F. CTA Chest

CTPA also noninvasively assesses for PH. IV contrast is timed for optimum evaluation of the pulmonary arteries in a CTPA examination. Chest CT findings suggestive of PH (MPA diameter ≥29 mm, MPA to ascending aorta ratio >1, true right and left descending PA diameters of 16 mm and 21 mm, respectively, ratio of segmental PA to accompanying bronchus >1:1, enlargement of the right ventricle, RV lumen/LV lumen ≥1, straightening of the interventricular septum, RV free wall thickness ≥6 mm, pericardial thickening or effusion, and enlargement of the bronchial arteries to a diameter of >1.5 mm) will also be present on CTPA [39,41,44,45]. MPA diameter on CTPA has a sensitivity of 87%, specificity of 89%, and PPV of 97% for PH [39]. Recently, investigators have looked into the prognostic value of the 3-D volume of the MPA segmented from the CTPA versus the axial MPA diameter of ≥29 mm for predicting PH and have found that the 3-D volumetric data outperformed the axial diameter measurement of the MPA [51]. Extrinsic compression of the left main coronary artery by a dilated MPA, an uncommon finding in PH, has also been reported at CTA [52,53]. As with chest CT, evaluation with RHC remains necessary to confirm PH suspected on CTPA before initiating therapy [24].

Like chest CT, CTPA also characterizes pulmonary etiologies that result in PH as described above. In addition, CTPA characterizes cardiovascular conditions that cause PH including CTEPH, intravascular tumor, and left-to-right shunts/congenital heart disease. CTPA findings of CTEPH

include findings of PH as well as findings of chronic pulmonary embolism: eccentric thrombus within pulmonary arteries, abrupt cutoff/narrowing of a PA, linear webs within affected arteries, bronchial artery dilation, and mosaic attenuation of the lungs from mosaic perfusion [21,46]. As discussed above, CTPA has the excellent specificity for CTEPH but suffers from lower sensitivity when compared with V/Q scintigraphy [35-37]. Several studies have analyzed the prognostic value of dual-energy CT derived lung perfused blood volume maps, which depict lung perfusion similar to V/Q perfusion scintigraphy, and have shown moderately good agreement with perfusion scintigraphy and good sensitivity, specificity, and PPV (96%–97%, 76%–86%, and 85%–94%, respectively) for detecting perfusion defects present in CTEPH patients with moderate to severe PH [54-58]. Congenital heart disease (atrial septal defect, ventricular septal defect) as well as other left to right shunts, such as patent ductus arteriosus and anomalous pulmonary venous return, result in PH and can be diagnosed with CTPA; however, MRI remains the preferred modality [46,47]. Intravascular tumor emboli (commonly breast, liver, choriocarcinoma) as well as primary pulmonary sarcomas can also result in PH and appear as lobulated intravascular defects with some degree of enhancement (albeit minimal for some intravascular tumors) on CTPA [46].

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G. MRI Heart Function and Morphology

MRI can also noninvasively assess the MPA and right ventricle for PH with good sensitivity and specificity (92% and 80%, respectively) and is useful for RV morphology and function assessment. RV functional abnormalities secondary to PH-related cardiac remodeling include RV hypokinesis, leftward bowing and/or paradoxical movement of the interventricular septum, RV dysfunction (increased end-diastolic volume, reduced ejection fraction, reduced cardiac index, reduced stroke volume), and pulmonary and tricuspid insufficiency [59-62]. RV mass and LV mass can also be accurately determined, which can then be used to calculate a ventricular mass index (RV mass/LV mass), with a ratio >0.6 being abnormal [61,63,64]. Cardiac MRI shows many of the morphologic changes of PH that are also depicted by chest CT and CTPA: PA enlargement, MPA to adjacent ascending aorta ratio > 1, RV enlargement and hypertrophy, straightening of the interventricular septum, and pericardial thickening and effusion [61,65,66]. Measured parameters of RV ejection fraction, RV end diastolic volume, and RV-PA coupling metrics measured on cardiac MR have been shown to be important prognostic indicators of PH [59,65].

Additional characteristic findings of PH on cardiac MRI include findings on late gadolinium enhancement, T1 mapping, and phase-contrast sequences. Enhancement of the RV insertion points on late gadolinium enhancement imaging is commonly present in individuals with PH and is compatible with fibrosis related to RV stress [60,62,63,67]. T1 mapping will also show abnormally prolonged values at the RV insertion points in PH [68,69]. Cardiac MRI phase-contrast imaging techniques can measure average blood flow velocity of the MPA, which correlates with mPAP. Patients with PH have sluggish PA flow as demonstrated by prolonged acceleration times on phase-contrast imaging. Severity of tricuspid regurgitation can be accurately quantified with 2-D phase-contrast imaging and used to estimate MPA pressure [65,70,71]. More recently, 4-D flow has been used to assess the MPA and detect the hemodynamic alterations that occur with PH: decreased wall shear stress, increased tricuspid regurgitation velocity, and abnormal vortex blood flow pattern within the MPA that is associated with early-onset systolic retrograde flow [65,72-74].

MRI can quantify cardiovascular shunts that are difficult to identify on echocardiography, including sinus venosus atrial septal defects, patent ductus arteriosus, and anomalous pulmonary venous return [65,75]. Cardiac MRI can also monitor progression of PH as well as response to treatment

[21]. Cardiac MRI can accurately diagnose mild PH, a finding missed by other noninvasive imaging modalities, via PA wall stiffness assessment. MPA pulsatility ≤40% diagnoses mild PH with excellent sensitivity and specificity (95% and 94%, respectively) [62,76].

Variant 1: Suspected pulmonary hypertension. Initial imaging.
H. MRA Chest

Pulmonary MR angiography (MRA) shows morphologic changes of PH that are also depicted by MRI heart and CT/CTPA: PA enlargement, MPA to adjacent ascending aorta ratio >1, RV enlargement and hypertrophy, straightening of the interventricular septum, and pericardial thickening and effusion [61,65,66]. MRA has lower sensitivity for the detection of acute and chronic pulmonary embolism compared with CTPA [61]. There are no data to support the use of MRA chest alone as a first-line test for suspected PH; however, the combination of MRA and MR perfusion imaging of the lung can diagnose CTEPH with good sensitivity and specificity (83%–100% and 98%–99%, respectively) in patients who have already been diagnosed with PH [61,63,66,75,77].

Variant 1: Suspected pulmonary hypertension. Initial imaging.
I. Catheterization Right Heart

RHC is an invasive procedure that defines cardiopulmonary hemodynamics and is performed after all noninvasive examinations have been completed to confirm the diagnosis of PH before initiating treatment per current guidelines [24]. RHC directly measures the PAP to confirm the diagnosis of PH as well as the PAWP and cardiac function (thermodilution or Fick method), which are both necessary to determine PVR. The mean PAP, PAWP, and PVR values obtained from RHC are used to classify PH into precapillary PH, isolated postcapillary pH, or combined pre- and postcapillary PH [78-80]. 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 [79]. RHC has morbidity and mortality rates of 1.1% and 0.055%, respectively [81].

Variant 1: Suspected pulmonary hypertension. Initial imaging.
J. Arteriography Pulmonary with Right Heart Catheterization

There are no available data to support the use of catheter pulmonary angiography as an initial test in the workup of suspected PH. Catheter pulmonary angiography was useful for assessing pulmonary embolism before multidetector CT, but multiple studies have now shown that CTPA is as reliable as angiography in the evaluation of CTEPH [24,82]. 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 [83]. Catheter pulmonary angiography is now used almost exclusively for thrombolysis or for presurgical planning in cases of CTEPH [24,82].

Summary of Recommendations

- **Variant 1**: Radiography chest or US echocardiography transthoracic resting or CT chest with IV contrast or CTA chest with IV contrast is usually appropriate for the initial imaging of patients with suspected PH. These procedures are complementary (ie, more than one procedure can be performed), with CTA chest yielding the most information of these 4 tests. The panel did not agree on recommending V/Q scan lung for this clinical scenario. There is insufficient medical literature to conclude whether or not these patients would benefit from V/Q scan lung. Imaging in this patient

population is controversial but may be appropriate.

Supporting Documents

The evidence table, literature search, and appendix for this topic are available at https://acsearch.acr.org/list. The appendix includes the strength of evidence assessment and the final rating round tabulations for each recommendation.

For additional information on the Appropriateness Criteria methodology and other supporting documents, please go to the ACR website at https://www.acr.org/Clinical-Resources/Clinical-Tools-and-Reference/Appropriateness-Criteria.

Appropriateness Category Names and Definitions

Appropriateness Category Name	Appropriateness Rating	Appropriateness Category Definition
Usually Appropriate	7, 8, or 9	The imaging procedure or treatment is indicated in the specified clinical scenarios at a favorable risk-benefit ratio for patients.
May Be Appropriate	4, 5, or 6	The imaging procedure or treatment may be indicated in the specified clinical scenarios as an alternative to imaging procedures or treatments with a more favorable risk-benefit ratio, or the risk-benefit ratio for patients is equivocal.
May Be Appropriate (Disagreement)	5	The individual ratings are too dispersed from the panel median. The different label provides transparency regarding the panel's recommendation. "May be appropriate" is the rating category and a rating of 5 is assigned.
Usually Not Appropriate	1, 2, or 3	The imaging procedure or treatment is unlikely to be indicated in the specified clinical scenarios, or the risk-benefit ratio for patients is likely to be unfavorable.

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, because of both 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 with 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 (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."

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Disclaimer

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

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

Group 1. Pulmonary arterial hypertension (PAH)
1.1 Idiopathic PAH
1.2 Heritable PAH
1.3 Drug- and toxin-induced PAH
–Definite: Aminorex, Fenfluramine, Dexfenfluramine, Benfluorex, Methamphetamines, Desatinib, Toxic rapeseed oil
–Possible: Cocaine, Phenylpropanolamine, L-tryptophan, St. John's wort, Amphetamines, Interferon-α &-β, Alkylating agents, Bosutinib, Direct-acting antiviral agents against hepatitis C virus, Leflunomide, Indirubin (Chinese herb Qing-Dai)
1.4 PAH 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
1.5 PAH long-term responders to calcium channel blockers
1.6 PAH with overt features of venous/capillaries (PVOD/PCH) involvement
-PFTs (decreased DLCO, severe hypoxia), Chest CT (septal lines, centrilobular ground glass

opacities/nodules, mediastinal lymph node enlargement), Possible edema in response to PAH therapy, Biallelic EIF2AK4 mutations, Organic solvent (trichloroethylene) exposure
1.7 Persistent PH of the newborn syndrome
Group 2. PH due to left heart disease
2.1 PH due to left heart failure with preserved LVEF
2.2 PH due to left heart failure with reduced LVEF
2.3 Valvular heart disease
2.4 Congenital/acquired cardiovascular conditions leading to postcapillary PH
Group 3. PH due to lung disease and/or hypoxia
3.1 Obstructive lung disease
3.2 Restrictive lung disease
3.3 Other lung diseases with mixed restrictive/obstructive pattern
3.4 Hypoxia without lung disease
3.5 Developmental lung disorders
Group 4. PH due to pulmonary artery obstructions
4.1 Chronic thromboembolic PH
4.2 Other pulmonary artery obstructions
4.2.1 Sarcoma (high or intermediate grade) or angiosarcoma
4.2.2 Other malignant tumors: renal carcinoma, uterine carcinoma, germ cell tumor of testis, other tumors
4.2.3. Non-malignant tumors (uterine leiomyoma)
4.2.4 Arteritis without connective tissue disease
4.2.5 Congenital pulmonary artery stenoses
4.2.6 Parasites (Hydatidosis)
Group 5. PH with unclear and/or multifactorial mechanisms
5.1 Hematologic disorders: chronic hemolytic anemia, myeloproliferative disorders
5.2 Systemic and metabolic disorders: pulmonary Langerhans cell histiocytosis, Gaucher disease, glycogen storage disease, neurofibromatosis, sarcoidosis
5.3 Others: chronic renal failure with or without hemodialysis, fibrosing mediastinitis
5.4 Complex congenital heart disease
PAH = pulmonary arterial hypertension, PVOD = pulmonary veno-occlusive disease, PCH = pulmonary capillary hemangiomatosis, PH = pulmonary hypertension, LVEF = left ventricular ejection fraction

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