American College of Radiology ACR Appropriateness Criteria® Imaging for Pulmonary Embolism, Known Clot

<u>Variant: 1</u> Adult. Known history of acute pulmonary embolism. Suspected recurrent or residual embolic disease. Initial imaging.

Procedure	Appropriateness Category	Relative Radiation Level
MRA chest with IV contrast	Usually Appropriate	0
CTA pulmonary arteries with IV contrast	Usually Appropriate	∵
V/Q scan lung	Usually Appropriate	૽ ૽
V/Q scan with SPECT or SPECT/CT lung	Usually Appropriate	૽ ૽
US echocardiography transthoracic resting	May Be Appropriate	0
MRA chest without and with IV contrast	May Be Appropriate	0
MRA chest without IV contrast	May Be Appropriate	0
US echocardiography transesophageal	Usually Not Appropriate	0
Radiography chest	Usually Not Appropriate	€
Arteriography pulmonary	Usually Not Appropriate	⊗⊗⊗
Arteriography pulmonary with right heart catheterization	Usually Not Appropriate	⊗⊗⊗
MRI heart function and morphology without and with IV contrast	Usually Not Appropriate	0
MRI heart function and morphology without IV contrast	Usually Not Appropriate	0
CT chest with IV contrast	Usually Not Appropriate	∵
CT chest without and with IV contrast	Usually Not Appropriate	∵
CT chest without IV contrast	Usually Not Appropriate	∵
CT heart function and morphology with IV contrast	Usually Not Appropriate	♦♦

Variant: 2 Adult. Known chronic thromboembolic disease. Surveillance.

Procedure	Appropriateness Category	Relative Radiation Level
CTA pulmonary arteries with IV contrast	monary arteries with IV contrast Usually Appropriate	
US echocardiography transthoracic resting	May Be Appropriate (Disagreement)	0
Arteriography pulmonary	May Be Appropriate	※※※
Arteriography pulmonary with right heart catheterization	neart catheterization May Be Appropriate (Disagreement)	
MRA chest with IV contrast	IV contrast May Be Appropriate	
MRA chest without and with IV contrast	RA chest without and with IV contrast May Be Appropriate (Disagreement)	
MRI heart function and morphology without and with IV contrast May Be Appropriate (Disagreement)		0
MRI heart function and morphology without IV contrast	May Be Appropriate	0
V/Q scan lung	May Be Appropriate (Disagreement)	⊗ ⊗
V/Q scan with SPECT or SPECT/CT lung	May Be Appropriate (Disagreement)	⊗ ⊗
US echocardiography transesophageal	Usually Not Appropriate	0
Radiography chest	Usually Not Appropriate	•
MRA chest without IV contrast Usually Not Appropriate		0
CT chest with IV contrast	Usually Not Appropriate	∵ ∵
CT chest without and with IV contrast	Usually Not Appropriate	∵ ∵
CT chest without IV contrast	Usually Not Appropriate	※ ※



Panel Members

Rahul D. Renapurkar, MD, MBBS^a, Prabhakar Shantha Rajiah, MD^b, Twyla B. Bartel, DO, MBA^c, Ahmed H. El-Sherief, MD^d, Christopher J. Francois, MD^e, Kate Hanneman, MD, MPH^f, Joe Y. Hsu, MD^g, Christopher D. Jackson, MD^h, Veronica Lenge de Rosen, MDⁱ, Lucy M. Safi, DO^j, Lilia M. Sierra-Galan, MD^k, Phillip M. Young, MD^l, Michael A. Bolen, MD^m

Summary of Literature Review

Introduction/Background

Acute pulmonary embolism (PE) is one of the most common cardiovascular diseases and is the third most common cause of mortality [1]. Diagnosing PE can often be delayed due to variable clinical presentation and nonspecific symptoms. Although mortality rates have declined over the years due to improved and timely therapeutic intervention, survivors of acute PE can have recurrent thromboembolic events as well as long-term functional sequelae. Risk factors for recurrent events include unprovoked and symptomatic first PE events, inadequate anticoagulation, and risk factors such as cancer and inherited thrombotic disorders [2]. Long-term sequela of PE can be varied, including symptoms such as dyspnea, chest pain, and decreased exercise capacity, all with significant potential to adversely impact quality of life. Imaging abnormalities such as residual pulmonary clots and right ventricular dysfunction and hemodynamic sequelae such as elevated mean pulmonary artery pressure (mPAP) and pulmonary vascular resistance (PVR) can also be present. See the ACR

Appropriateness Criteria [®] topic on "<u>Suspected Pulmonary Hypertension</u>" [3] for further details. In a patient with a history of acute PE, these have been loosely labeled as post-PE syndrome, although a precise definition of this term is lacking [4].

Common scenarios included in the post-PE syndrome include patients who, following at least 3 months of therapeutic anticoagulation, demonstrate post-PE functional limitation, post-PE cardiac impairment, chronic thromboembolic pulmonary disease (CTEPD), or chronic thromboembolic pulmonary hypertension (CTEPH). CTEPH is the severest form of presentation characterized by at least one mismatched segmental perfusion defect along with elevated mPAP (≥20 mm Hg) and normal pulmonary capillary wedge pressure (≤15 mm Hg) according to the current 2022 European Society of Cardiology/European Respiratory Society Guidelines for the diagnosis and treatment of pulmonary hypertension (PH) [5]. CTEPH is a relatively rare disease; varying incidence rates ranging from 0.6% to 8.2% have been described in the literature, depending on the population studied [4]. In the International CTEPH registry, 74.8% patients had a prior history of PE [6]. CTEPD is a diagnosis reserved for patients with presentation similar to CTEPH but with resting normal mPAP. Incidence rates of CTEPD are not truly known due to difficulty in diagnosis and lack of established data.

Guidelines for surveillance of patients following an acute PE event continue to evolve. Routine imaging surveillance of patients with prior PE and who are asymptomatic is not recommended [7].

Imaging plays a multifaceted role including confirming clinical suspicion of recurrent or residual

thromboemboli, ruling out alternative diagnoses and assessing disease burden, and characterization for further therapeutic intervention. In this document, we discuss the role of different imaging modalities that can play a pivotal role in evaluation. In addition, imaging in suspected PE is covered in the ACR Appropriateness Criteria [®] topic on "Suspected Pulmonary Embolism" [8].

Special Imaging Considerations

For the purposes of distinguishing between CT and CT angiography (CTA), ACR Appropriateness Criteria topics use the definition in the <u>ACR-NASCI-SIR-SPR Practice Parameter for the Performance and Interpretation of Body Computed Tomography Angiography (CTA)</u> [9]: "CTA uses a thin-section CT acquisition that is timed to coincide with peak arterial and/or venous enhancement, depending on the vascular structures to be analyzed. The resultant volumetric data set 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.

As imaging techniques continue to evolve, newer techniques have been increasingly used in assessment of PE. Dual-energy CT (DECT) allows the simultaneous acquisition of data at 2 different x-ray energies; these data can be used to quantify the fraction of individual materials such as iodine within a voxel of lung tissue. Depending on the vendors, different approaches are available for dual-energy analysis, broadly classified as raw data-based analysis that uses projection postprocessing data directly acquired from the CT acquisition and image-based analysis that involves postprocessing of the acquired data. One vendor uses postprocessing technique to generate images that are termed perfused blood volume (PBV) or sometimes pulmonary blood volume images [10]. PBV maps are often color-coded and superimposed as a color-fused overlay over the anatomic data. The biggest advantage of these maps is that they assess regional enhancement of the lung at a single time point, which can provide indirect assessment of lung perfusion. DECT can offer incremental benefits in the detection of PE, risk stratification and diagnosis, and preoperative planning of patients with CTEPH [10]. Similarly, single-photon emission computed tomography (SPECT) and SPECT/CT ventilation-perfusion (V/Q) scanning allows to overcome limitations of 2-D planar V/Q scanning, such as shine-through and better localization of perfusion defects, hence improving the sensitivity, specificity, and accuracy of V/Q imaging [11]. Also, newer sequences such as 4-D flow MRI and MR perfusion can provide noninvasive assessment of pulmonary flow patterns and wall shear stress, which can be used to monitor patients with CTEPH and to assess response to treatment [12,13].

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 (ie, only one procedure will be ordered

to provide the clinical information to effectively manage the patient's care)

OR

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

Discussion of Procedures by Variant

Variant 1: Adult. Known history of acute pulmonary embolism. Suspected recurrent or residual embolic disease. Initial imaging.

In patients with a known history of PE with clinical suspicion of recurrent/residual PE, imaging can be used to confirm the clinical suspicion, characterize the disease burden, and triage the patient for further diagnostic workup or clinical decision making.

Variant 1: Adult. Known history of acute pulmonary embolism. Suspected recurrent or residual embolic disease. Initial imaging.

A. Arteriography pulmonary

With the evolution of CT imaging, pulmonary angiography is not typically useful as a first-line test for suspected recurrent/residual PE. Catheter-based angiography has a major role to play in the confirmation of residual disease if CT is negative and a high index of clinical suspicion exists.

Variant 1: Adult. Known history of acute pulmonary embolism. Suspected recurrent or residual embolic disease. Initial imaging.

B. Arteriography pulmonary with right heart catheterization

With the evolution of CT imaging, pulmonary angiography with right heart catheterization (RHC) is not useful as a first-line test for suspected recurrent/residual PE. Catheter-based angiography has a major role in the confirmation of residual disease if CT is negative and a high index of clinical suspicion exists.

Variant 1: Adult. Known history of acute pulmonary embolism. Suspected recurrent or residual embolic disease. Initial imaging.

C. CT chest with IV contrast

There is no relevant literature to support the use of CT chest with intravenous (IV) contrast to assess suspected recurrent/residual PE.

Variant 1: Adult. Known history of acute pulmonary embolism. Suspected recurrent or residual embolic disease. Initial imaging.

D. CT chest without and with IV contrast

There is no relevant literature to support the use of CT chest without and with IV contrast to assess suspected recurrent/residual PE.

Variant 1: Adult. Known history of acute pulmonary embolism. Suspected recurrent or residual embolic disease. Initial imaging.

E. CT chest without IV contrast

There is no relevant literature to support the use of CT chest without IV contrast to assess suspected recurrent/residual PE.

Variant 1: Adult. Known history of acute pulmonary embolism. Suspected recurrent or residual embolic disease. Initial imaging.

F. CT heart function and morphology with IV contrast

There is no relevant literature to support the use of CT heart function and morphology with IV contrast to assess suspected recurrent/residual PE.

Variant 1: Adult. Known history of acute pulmonary embolism. Suspected recurrent or residual embolic disease. Initial imaging.

G. CTA pulmonary arteries with IV contrast

CTA pulmonary arteries with IV contrast is a useful imaging modality for suspected recurrent or residual PE. Excellent spatial resolution and contrast-to-noise ratio, detailed evaluation of vasculature and lung parenchyma, and an ability to rule out alternative differential diagnoses are some advantages offered by this imaging test. Recurrent clots are often seen as filling defects on the pulmonary arteries, which can be occlusive. The imaging features are usually similar to acute PE, although underlying changes related to chronic thromboembolic disease might be identified [14]. In patients with residual PE, imaging features vary and can be seen as webs, or linear filling defects, areas of stenosis, poorly opacified pulmonary arteries, and subtotal to total occlusive lesions [15]. In one study of 55 patients, central disease was better identified with CT when compared with pulmonary angiography (accuracy of 0.79 for each of the 2 readers compared with accuracy of 0.74 with pulmonary angiogram) [16]. It should be noted that CT technology has evolved considerably since some of these older studies, and the accuracy of CTA has most likely substantially improved since then. Newer CT techniques, such as high-pitch acquisition in a dualsource scanner with potential for electrocardiogram (ECG) triggering, have the advantages of decreased motion artifacts with resultant improved PE detection as well as improved evaluation of cardiovascular structures [17,18]. Similarly, DECT imaging can improve detection and interobserver agreement in detecting acute PE due to the ability to use perfusion maps [19]. Also, monoenergetic images can be used to reduce contrast and potentially rescue a study with suboptimal bolus [20]. DECT-based perfusion maps have a proven role in diagnosing chronic PE with good agreement with V/Q scanning, often used as the initial test of choice. In a study on 80 patients, DECT showed excellent agreement with scintigraphy in diagnosing CTEPH (k = 0.80); combined information from DECT perfusion and CTA images improved diagnostic accuracy [21].

Variant 1: Adult. Known history of acute pulmonary embolism. Suspected recurrent or residual embolic disease. Initial imaging.

H. MRA chest with IV contrast

MR angiography (MRA) is generally considered as an alternative modality in diagnosing acute PE and is useful for diagnosing central or lobar PE [22-24]. Some of the limitations of MRA include decreased sensitivity in the detection of peripheral PE. These limitations were highlighted in the Prospective Investigation of Pulmonary Embolism Diagnosis III (PIOPED III) multicenter trial, which evaluated the sensitivity and specificity of gadolinium-enhanced MRA to a composite reference standard (D-dimer, V/Q scan, CTA) [23,25]. In the study, MRA was technically inadequate in 25% of patients across all centers. In technically adequate studies, MRA identified 57% (59 of 104) of those with PE and had a sensitivity of 78% and a specificity of 99% [23]. Similarly, MRA can help detect chronic thromboembolic disease especially involving the central vessels, although data are conflicting and suffer from limitations such as smaller sample sizes. For example, in one study on 53 patients, the overall sensitivity and specificity of MRA in diagnosing proximal and distal chronic PE were 98% and 94%, respectively. MRA identified more stenoses (29/18), poststenosis dilatation (23/7), and occlusions (37/29) compared with CTA, and MRA perfusion images showed a sensitivity

of 92% for diagnosing chronic PE [26]. In another study on 24 patients with CTEPH, the sensitivity and specificity regarding CTEPH-related changes at the main/lobar and at the segmental levels were 100%/100% and 100%/99% for CTA, 83.1%/98.6% and 87.7%/98.1% for MRA, and 65.7%/100% and 75.8%/100% for pulmonary angiogram, respectively [27].

Variant 1: Adult. Known history of acute pulmonary embolism. Suspected recurrent or residual embolic disease. Initial imaging.

I. MRA chest without and with IV contrast

MRA is generally considered as an alternative modality in diagnosing acute PE and is useful for diagnosing central or lobar PE [22,23]. Some of the limitations of MRA include technically challenging study, longer duration, and decreased sensitivity in the detection of peripheral PE. These limitations were highlighted in the PIOPED III multicenter trial, which evaluated the sensitivity and specificity of gadolinium-enhanced MRA to a composite reference standard (Ddimer, V/Q scan, CTA) [23,25]. In the study, MRA was technically inadequate in 25% of patients across all centers. In technically adequate studies, MRA identified 57% (59 of 104) of patients with PE and had a sensitivity of 78% and a specificity of 99% [23]. Similarly, MRA can help detect chronic thromboembolic disease especially involving the central vessels, although data are conflicting and suffer from limitations such as smaller sample sizes of the studies. For example, in one study on 53 patients, the overall sensitivity and specificity of MRA in diagnosing proximal and distal chronic PE were 98% and 94%, respectively. MRA identified more stenoses (29/18), poststenosis dilatation (23/7), and occlusions (37/29) compared with CTA, and MRA perfusion images showed a sensitivity of 92% for diagnosing chronic PE [26]. In another study on 24 patients with CTEPH, the sensitivity and specificity regarding CTEPH-related changes at the main/lobar and at the segmental levels were 100%/100% and 100%/99% for CTA, 83.1%/98.6% and 87.7%/98.1% for MRA, and 65.7%/100% and 75.8%/100% for pulmonary angiogram, respectively [27].

Limited data exist on the role of noncontrast MRI in evaluation of acute and chronic PE. Commonly used noncontrast MRA sequences are 3-D balanced steady-state free precession MRA that can be respiratory-gated and phase-contrast MRA. In a study of 93 patients with suspected PE, noncontrast MRA was found to have good sensitivity (85%) and specificity (98.6%), with an accuracy comparable to CTA [28].

Variant 1: Adult. Known history of acute pulmonary embolism. Suspected recurrent or residual embolic disease. Initial imaging.

J. MRA chest without IV contrast

Limited data exist on the role of noncontrast MRI in evaluation of acute and chronic PE. In a study of 93 patients with suspected PE, noncontrast MRA was found to have good sensitivity (85%) and specificity (98.6%), with an accuracy comparable to CTA [28].

Variant 1: Adult. Known history of acute pulmonary embolism. Suspected recurrent or residual embolic disease. Initial imaging.

K. MRI heart function and morphology without and with IV contrast

There is no relevant literature to support the use of MRI heart function and morphology without and with IV contrast to assess suspected recurrent/residual PE. MRI heart function can play a crucial role in the noninvasive assessment of hemodynamics and risk stratification, although it offers little value in the initial diagnosis [29].

Variant 1: Adult. Known history of acute pulmonary embolism. Suspected recurrent or residual embolic disease. Initial imaging.

L. MRI heart function and morphology without IV contrast

There is no relevant literature to support the use of MRI heart function and morphology without IV contrast to assess suspected recurrent/residual PE. MRI heart function can play a crucial role in the noninvasive assessment of hemodynamics and risk stratification, although it offers little value in the initial diagnosis.

Variant 1: Adult. Known history of acute pulmonary embolism. Suspected recurrent or residual embolic disease. Initial imaging.

M. Radiography chest

Chest radiography is neither sensitive nor specific for diagnosing recurrent or residual PE and, as such, is not useful. It is often performed as a baseline investigation in patients presenting with symptoms such as shortness of breath or chest pain [30].

Variant 1: Adult. Known history of acute pulmonary embolism. Suspected recurrent or residual embolic disease. Initial imaging.

N. US echocardiography transesophageal

Acute PE can be suspected during transesophageal echocardiography (TEE) when there is a hypokinetic or akinetic mid and basal right ventricular free wall with preserved/hyperkinetic right ventricular apical wall motion, labeled as the McConnell sign [31]. Subsequent studies have shown that this sign is neither sensitive nor specific for diagnosing acute PE, and often these patients undergo subsequent CTA for confirmation of diagnosis [32]. TEE is not routinely indicated. Evaluation of right ventricular systolic pressure by Doppler echocardiography is the most common method for evaluation of PH, although it has to be confirmed with RHC.

Variant 1: Adult. Known history of acute pulmonary embolism. Suspected recurrent or residual embolic disease. Initial imaging.

O. US echocardiography transthoracic resting

Acute PE can be suspected during transthoracic echocardiography (TTE) when there is a hypokinetic or akinetic mid and basal right ventricular free wall with preserved/hyperkinetic right ventricular apical wall motion, labeled as the McConnell sign [31]. Subsequent studies have shown that this sign is neither sensitive nor specific for diagnosing acute PE, and often these patients undergo subsequent CTA for confirmation of diagnosis [32]. TTE remains the initial test of choice in screening patients for CTEPH after acute PE in patients with persistent symptoms [33].

Variant 1: Adult. Known history of acute pulmonary embolism. Suspected recurrent or residual embolic disease. Initial imaging.

P. V/Q scan lung

V/Q scanning has largely been replaced by CTA in the evaluation of patients with suspected acute PE [34,35]. In some situations, perfusion imaging alone can also be used in diagnosing PE. One randomized controlled trial on 1,417 patients showed that more patients (19.2%) were diagnosed with PE with CTA versus 14.2% with V/Q scanning. Some of the limitations of V/Q scanning include high proportion of nondiagnostic studies and inability to rule out alternative diagnoses [34]. The PIOPED II investigators used a modified criteria for evaluation of V/Q scans and demonstrated that, in most of the patients, V/Q scan is definitively diagnostic in most cases [36]. In contrast to the evaluation of acute PE, V/Q scanning remains the initial diagnostic test of choice in the evaluation of residual/chronic PE. In a retrospective study on 227 patients, V/Q scintigraphy was found to have a sensitivity of 96% to 97.4% and a specificity of 90% to 95%; CT pulmonary angiography showed a sensitivity of 51% and a specificity of 99% [37]. CTA findings can be subtle in

residual/chronic PE and can be overlooked.

Variant 1: Adult. Known history of acute pulmonary embolism. Suspected recurrent or residual embolic disease. Initial imaging.

Q. V/Q scan with SPECT or SPECT/CT lung

To improve the diagnostic performance of V/Q scans and decrease the number of nondiagnostic studies, V/Q scan with SPECT has been proposed with studies demonstrating high negative predictive value and improved accuracy [38,39]. The addition of a low-dose CT to SPECT imaging allows the detection of conditions other than PE that can cause V/Q mismatch such as extrinsic vascular compression from neoplasm or lymphadenopathy, vasculitis, and postradiation therapy changes; this improves the diagnostic accuracy. In a recent meta-analysis comparing V/Q-SPECT, V/Q-SPECT/CT Q-SPECT/CT, and Q-SPECT, V/Q SPECT/CT examinations had the highest specificity in diagnosing acute PE [40]. Head-to-head comparison of V/Q-SPECT/CT imaging with CTA has been encouraging, with several studies demonstrating comparable accuracy and suggesting that SPECT and SPECT/CT imaging is a viable alternative to CTA in diagnosing acute PE [41]. Despite all these strengths, gaps in data still exist, and the outcomes of patients investigated with SPECT imaging remain unknown; further studies focusing on outcomes are necessary for successful implementation in clinical practice [42]. Data in the evaluation of chronic PE are similar with studies demonstrating improved sensitivity of SPECT imaging over planar imaging in detection of CTEPH [43].

Variant 2: Adult. Known chronic thromboembolic disease. Surveillance.

In patients with known chronic thromboembolic disease, imaging can be used for active surveillance of the disease burden, evaluation of disease progression, and to provide guidance on the next step of management, whether medical therapy, surgery, or endovascular intervention. Surgical treatment with pulmonary thromboendartectomy (PTE) is the treatment of choice with excellent short- and long-term outcomes, although careful patient selection is crucial [44]. Often a multidisciplinary approach is needed in patient selection, with imaging providing crucial insights in preoperative planning.

Variant 2: Adult. Known chronic thromboembolic disease. Surveillance. A. Arteriography pulmonary

Historically considered as the reference standard for diagnosing CTEPH, pulmonary angiography has fallen out of favor due to the emergence of noninvasive modalities such as CTA. However, it still plays a pivotal role in mapping the disease especially in patients considered for balloon pulmonary angioplasty (BPA). Digital subtracting angiography allows the classification of the lesion in terms of morphology and distribution, which can influence the outcomes and complication rate following BPA [44]. Total occlusions have the lowest success rate, whereas tortuous lesions are associated with increased complications [44]. The addition of cone beam CT allows 3-D cross-sectional imaging down to the subsegmental level and can improve the diagnostic accuracy of lesions over digital subtracting angiography [45]. Routine endovascular imaging with intravascular ultrasound or optical coherence tomography can add value but is not the standard of care [46].

Variant 2: Adult. Known chronic thromboembolic disease. Surveillance. B. Arteriography pulmonary with right heart catheterization

RHC allows definitive assessment of precapillary PH defined by a mPAP >20 mm Hg, a pulmonary artery wedge pressure of \leq 15 mm Hg, and a PVR of \geq 240 dynes × sec × cm⁻⁵ or 3 Wood units [47]. Chronic thromboembolic disease is a diagnosis assigned to symptomatic patients with lung

perfusion defects and evidence of chronic clots without resting PH [48]; exercise-induced RHC or cardiopulmonary exercise testing can aid in diagnosis [49]. Beyond diagnosis, RHC allows comprehensive assessment of the hemodynamic significance of the disease and its severity, which can help in follow-up and deciding further treatment options.

Variant 2: Adult. Known chronic thromboembolic disease. Surveillance. C. CT chest with IV contrast

There is no relevant literature to support the use of CT chest with IV contrast for surveillance of patients with chronic thromboembolic disease. When IV contrast is administered for CT, the study should be performed as a CTA pulmonary arteries.

Variant 2: Adult. Known chronic thromboembolic disease. Surveillance.

D. CT chest without and with IV contrast

There is no relevant literature to support the use of CT chest without and with IV contrast for surveillance of patients with chronic thromboembolic disease.

Variant 2: Adult. Known chronic thromboembolic disease. Surveillance.

E. CT chest without IV contrast

There is no relevant literature to support the use of CT chest without IV contrast for surveillance of patients with chronic thromboembolic disease.

Variant 2: Adult. Known chronic thromboembolic disease. Surveillance.

F. CT heart function and morphology with IV contrast

Some studies have evaluated the role of ECG-gated CT in the noninvasive assessment of pulmonary hemodynamics [50,51], although its routine use is not useful. In one study on 45 patients, the ventricular diameter ratio on the 4-chamber view adjusted by the pulmonary trunk diameter/aortic diameter ratio yielded the best correlation to mPAP [51].

Variant 2: Adult. Known chronic thromboembolic disease. Surveillance. G. CTA pulmonary arteries with IV contrast

CTA pulmonary arteries with IV contrast is useful in the assessment of known chronic thromboembolic disease, allowing detailed evaluation of pulmonary vasculature and lung parenchyma. Often, CTA findings can be divided into those related to chronic PE and those related to PH (if present). Signs of chronic PE such as webs, bands, areas of stenoses, and occlusions are clearly depicted with CTA down to the level of subsegmental branches [30]. Depending on the morphology of clot and associated vessel wall changes, CTA can be used to differentiate the age of the clot and assess for superimposed acute thromboembolic events (acute on chronic disease) [30]. Detailed mapping of the disease allows risk stratification and is one of the key factors in selecting patients for PTE. However, it should be noted that CTEPH findings can be subtle and can be overlooked or missed, especially in inexperienced centers [52]. CTA also plays a modest role in hemodynamic assessment and noninvasive monitoring. Established vascular and lung signs related to PH such as enlargement of pulmonary arteries and mosaic attenuation are well evaluated with CTA, and these can provide indirect assessment of hemodynamic significance. ECG-gated CTA can help in improved evaluation of functional parameters such pulmonary artery distensibility and right ventricular dilation, allowing better assessment of hemodynamics, but is not routinely used [49,51]. Bronchial artery collaterals are nicely depicted with CTA, the presence of which has prognostic value and is associated with good postoperative outcomes [53].

DECT offers several advantages in the surveillance of patients with chronic thromboembolic

disease by its ability to assess morphologic and perfusion information in a single scan. Beyond its ability to improve diagnostic accuracy [19], the pulmonary perfusion maps can be used for disease monitoring and assess disease severity [54]. Also, scoring models using anatomic and perfusion data have been developed that can offer insights into pulmonary hemodynamics, thereby allowing disease monitoring [55]. DECT also offers prognostic information and DECT-based scoring models can be used to predict response to surgery [56]. One of the critical factors in poor response to PTE is underlying microvasculopathy, and PBV maps have shown promise in the noninvasive assessment of microvascular disease; this can help in better patient selection [57]. Studies have shown that quantitative information obtained from perfusion maps can offer insights into differentiating chronic thromboembolic disease from CTEPH, which can have prognostic significance [58]. Perfusion maps can provide incremental information in selecting patients for BPA and allows better targeting of lesions, as well as predicting response to therapy [59,60].

Variant 2: Adult. Known chronic thromboembolic disease. Surveillance. H. MRA chest with IV contrast

The role of MRA and MRI in surveillance of CTEPH continues to evolve. Time-resolved 3-D MRA with IV contrast has been used to assess quantitative metrics such as pulmonary blood flow and pulmonary transit time (PTT). PTT is defined as the time taken for a contrast bolus to reach the left ventricle from right ventricle (RV) and has been proven to be a robust biomarker to identify the presence and severity of PH [61]. Qualitative and quantitative assessment of pulmonary perfusion can be a valuable aid in the monitoring of these patients and predicting response to surgery or BPA [62-64]. In a study of 30 patients with CTEPH, median PTT was significantly lower postsurgery (139 msec) compared with presurgery (193 msec); P = .0002. Median PTT correlated significantly with the mPAP post-PTE (r = 0.52; P < .008) [62]. It should be noted that widespread implementation and acceptance of MRA is still limited due to longer scan times and lack of data in larger patient populations.

Variant 2: Adult. Known chronic thromboembolic disease. Surveillance. I. MRA chest without and with IV contrast

Limited data exist on the role of noncontrast MRA in the surveillance of chronic PE. The role of MRA and MRI in the surveillance of CTEPH continues to evolve. Time-resolved 3-D MRA with IV contrast has been used to assess quantitative metrics such as pulmonary blood flow and PTT. PTT is defined as the time taken for a contrast bolus to reach the left ventricle from RV and has been proven to be a robust biomarker to identify the presence and severity of PH [61]. Qualitative and quantitative assessment of pulmonary perfusion can be a valuable aid in monitoring of these patients and predicting response to surgery or BPA [62-64]. In a study of 30 patients with CTEPH, median PTT was significantly lower postsurgery (139 msec) compared with presurgery (193 msec); P = .0002. Median PTT correlated significantly with the mPAP post-PTE (r = 0.52; P < .008) [62]. It should be noted that widespread implementation and acceptance of MRA is still limited due to longer scan times and lack of data in larger patient populations.

Variant 2: Adult. Known chronic thromboembolic disease. Surveillance. J. MRA chest without IV contrast

Limited data exist on the role of noncontrast MRA in surveillance of chronic PE. Functional imaging with highly polarized gases such as helium can provide insights into the regional ventilation and can help estimate local oxygen uptake from selected regions within the lung in patients with CTEPH [65].

Variant 2: Adult. Known chronic thromboembolic disease. Surveillance.

K. MRI heart function and morphology without and with IV contrast

MRI heart function and morphology offers tremendous potential in the noninvasive evaluation of pulmonary hemodynamics and predicting response to therapy. Several established and newer tools are available that can provide comprehensive information on right ventricular morphology and function, pulmonary macro- and microcirculation, and left ventricle and RV remodeling. MRI is the reference standard for assessment of RV function with excellent reproducibility and is routinely used for longitudinal follow-up of RV function [66]. RV functional metrics such as the eccentricity index and remodeling index can be easily calculated using cine images and correlate well with hemodynamics in patients with CTEPH; these can be used to assess favorable response to therapy [67]. Also, the calculation of RV strain using 2-D and 3-D techniques can be helpful for the assessment of treatment effects [68]. Similarly, T1 mapping has been evaluated for noninvasive monitoring and predicting response to therapy, although data are fairly sparse [69]. A 2-D phasecontrast MRI is routinely used for the assessment of pulmonary blood flow and can show abnormal velocity flow waveforms due to increased vascular resistance [70]. Newer tools such as 4-D flow MRI allow comprehensive evaluation of right heart and pulmonary flow; vortical flow has been demonstrated in patients with PH [71]. Several noninvasive biomarkers of pulmonary hemodynamics such as wall shear stress, vorticity, and stiffness can be calculated using 4-D flow MRI that can be used for longitudinal follow-up and predicting response to therapy [12]. In a small study of 20 patients who underwent 4-D flow imaging before and 6 months after pulmonary endarterectomy, 4-D flow-derived PA volumes decreased (P < .001) and stiffness, velocity, and vorticity increased (P < .01) post–pulmonary endarterectomy. Longitudinal improvements from pre- to post-pulmonary endarterectomy in mPAP were associated with longitudinal decreases in the main pulmonary artery area (r = 0.68; P = .002) [12].

Variant 2: Adult. Known chronic thromboembolic disease. Surveillance. L. MRI heart function and morphology without IV contrast

MRI heart function and morphology offers tremendous potential in the noninvasive evaluation of pulmonary hemodynamics and predicting response to therapy. Several established and newer tools are available that can provide comprehensive information on right ventricular morphology and function, pulmonary macro- and microcirculation, and left ventricle and RV remodeling. MRI is the reference standard for assessment of RV function with excellent reproducibility and is routinely used for longitudinal follow-up of RV function [66]. RV functional metrics such as the eccentricity index and remodeling index can be easily calculated using cine images and correlate well with hemodynamics in patients with CTEPH; these can be used to assess favorable response to therapy [67]. Also, calculation of RV strain using 2-D and 3-D techniques can be helpful for the assessment of treatment effects [68]. Similarly, T1 mapping has been evaluated for noninvasive monitoring and predicting response to therapy, although data are fairly sparse [69]. A 2-D phase-contrast MRI is routinely used for the assessment of pulmonary blood flow and can show abnormal velocity flow waveforms due to increased vascular resistance [70]. Newer tools such as 4-D flow MRI allow comprehensive evaluation of right heart and pulmonary flow; vortical flow has been demonstrated in patients with PH [71]. Several noninvasive biomarkers of pulmonary hemodynamics such as wall shear stress, vorticity, and stiffness can be calculated using 4-D flow MRI that can be used for longitudinal follow-up and predicting response to therapy [12]. In a small study of 20 patients who underwent 4-D flow imaging before and 6 months after pulmonary endarterectomy, 4-D flowderived PA volumes decreased (P < .001) and stiffness, velocity, and vorticity increased (P < .01) post–pulmonary endarterectomy. Longitudinal improvements from pre– to post–pulmonary endarterectomy in mPAP were associated with longitudinal decreases in the main pulmonary artery

Variant 2: Adult. Known chronic thromboembolic disease. Surveillance. M. Radiography chest

Chest radiography can demonstrate evidence of chronic thromboembolic disease and associated PH, such as dilated central pulmonary arteries, enlarged cardiac silhouette, and areas of scarring secondary to pulmonary infarcts; these findings are often nonspecific, and there is no defined role of radiography for active surveillance of these patients.

Variant 2: Adult. Known chronic thromboembolic disease. Surveillance. N. US echocardiography transesophageal

Noninvasive assessment of RV function is important in routine monitoring of patients with chronic thromboembolic disease and CTEPH and is routinely done with TEE. Markers of RV function and remodeling such as fractional area change, tricuspid annular plane systolic excursion, strain, and RV end-diastolic diameter index can be easily assessed with echocardiography and can be used to assess response to medical and surgical/interventional therapy [72,73]. TEE is often not routinely used except in a perioperative setting.

Variant 2: Adult. Known chronic thromboembolic disease. Surveillance. O. US echocardiography transthoracic resting

Noninvasive assessment of RV function is important in routine monitoring of patients with chronic thromboembolic disease and CTEPH and is routinely done with TTE. Markers of RV function and remodeling such as fractional area change, tricuspid annular plane systolic excursion, tricuspid annular velocity (S'), strain, and RV end-diastolic diameter index can be easily assessed with echocardiography and can be used to assess response to medical and surgical/interventional therapy [72,73]. TTE is often not routinely used except in a perioperative setting.

Variant 2: Adult. Known chronic thromboembolic disease. Surveillance. P. V/Q scan lung

Because V/Q scans provide little information on anatomy and RV function, they have a limited role in disease monitoring and assessment of hemodynamics. V/Q scan is, however, routinely used in a postoperative or post-BPA setting to assess response to therapy. Successful therapeutic response is often highlighted by improved perfusion and decreased mismatched defects [74]. Often areas with both ventilation and perfusion abnormalities can show normalization postsurgery [75]. New perfusion defects in unoperated segments can be seen postsurgery, a finding that has been described as steal phenomenon likely caused by redistribution of PVR [76].

Variant 2: Adult. Known chronic thromboembolic disease. Surveillance. Q. V/Q scan with SPECT or SPECT/CT lung

Because of the ability to quantitatively analyze the perfusion defects using SPECT and SPECT/CT V/Q scanning, these can be used for risk assessment and noninvasive assessment of hemodynamics [77,78]. Limited available data have shown a modest to good correlation of the quantitative perfusion defect analysis with hemodynamic parameters such as PVR. In a study on 83 patients with CTEPH, the percentage of perfusion defects correlated positively with mPAP, PVR, and RV pressure (r = 0.316, 0.318,and 0.432,respectively; P < .05) and correlated negatively with the 6-minute walk distance (r = -0.309; P < .05). Larger studies and more clinical validation are, however, needed for integration in clinical practice.

Summary of Highlights

This is a summary of the key recommendations from the variant tables. Refer to the complete narrative document for more information.

- **Variant 1**: CTA pulmonary arteries with IV contrast, V/Q scan (planar or SPECT), and MRA with IV contrast are considered appropriate tests for initial imaging of patients with suspected recurrent or residual pulmonary embolic disease. TTE may be appropriate especially for the initial evaluation of suspected CTEPH. MRA without IV contrast may be appropriate when contrast cannot be administered.
- Variant 2: In patients with known chronic thromboembolic disease, CTA pulmonary arteries with contrast is usually appropriate for surveillance of disease burden, evaluation of disease progression, and to provide guidance on next step of management. Pulmonary arteriography, MRA with IV contrast, and MRI heart function without IV contrast may be appropriate as equivalent alternatives. V/Q scan, MRA without IV contrast, pulmonary arteriography with RHC, and MRI heart function with IV contrast may also be appropriate, although there is no definite consensus on these tests.

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.

Gender Equality and Inclusivity Clause

The ACR acknowledges the limitations in applying inclusive language when citing research studies that predates the use of the current understanding of language inclusive of diversity in sex, intersex, gender, and gender-diverse people. The data variables regarding sex and gender used in the cited literature will not be changed. However, this guideline will use the terminology and definitions as proposed by the National Institutes of Health.

Appropriateness Category Names and Definitions

Appropriateness Category Name	Appropriateness Rating	Appropriateness Category Definition
Usually Appropriate		The imaging procedure or treatment is indicated in the specified clinical scenarios at a favorable riskbenefit 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)		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	
0	0 mSv	0 mSv	
③	<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.

^aCleveland Clinic, Cleveland, Ohio. ^bPanel Chair, Mayo Clinic, Rochester, Minnesota. ^cGlobal Advanced Imaging, PLLC, Little Rock, Arkansas; Commission on Nuclear Medicine and Molecular Imaging. ^dVA Greater Los Angeles Healthcare System, Los Angeles, California. ^eMayo Clinic, Rochester, Minnesota. ^fUniversity Medical Imaging Toronto, Department of Medical Imaging, University of Toronto, Toronto, Ontario, Canada. ^gKaiser Permanente, Los Angeles, California. ^hThe University of Tennessee Health Science Center, Memphis, Tennessee; Society of General Internal Medicine. ⁱBaylor College of Medicine, Houston, Texas. ^jMount Sinai Hospital, New York, New York; American Society of Echocardiography. ^kAmerican British Cowdray Medical Center, Mexico City, Mexico; Society for Cardiovascular Magnetic Resonance. ^lMayo Clinic, Rochester, Minnesota. ^mSpecialty Chair, Cleveland Clinic, Cleveland, Ohio.