American College of Radiology
ACR Appropriateness Criteria®
Pulmonary Arteriovenous Malformation (PAVM)

<u>**Variant 1:**</u>   **Presenting with a transient ischemic attack, or seizures, or brain abscess, or altered sensorium. Chest radiography reveals a lung nodule. Suspected pulmonary arteriovenous malformation (PAVM). Next imaging study.**

Procedure	Appropriateness Category	Relative Radiation Level
US echocardiography transthoracic with IV contrast	Usually Appropriate	O
MRA pulmonary arteries without and with IV contrast	Usually Appropriate	O
CT chest without IV contrast	Usually Appropriate	☢☢☢
CTA pulmonary arteries with IV contrast	Usually Appropriate	☢☢☢
US echocardiography transesophageal with IV contrast	May Be Appropriate	O
Arteriography pulmonary	May Be Appropriate	☢☢☢☢
MRA chest without and with IV contrast	May Be Appropriate (Disagreement)	O
CT chest with IV contrast	May Be Appropriate	☢☢☢
CT chest without and with IV contrast	May Be Appropriate (Disagreement)	☢☢☢
CTA chest with IV contrast	May Be Appropriate (Disagreement)	☢☢☢
US echocardiography transesophageal	Usually Not Appropriate	O
US echocardiography transthoracic resting	Usually Not Appropriate	O
Radiography chest	Usually Not Appropriate	☢
MRA chest without IV contrast	Usually Not Appropriate	O
MRA pulmonary arteries without IV contrast	Usually Not Appropriate	O
Pertechnetate albumin pulmonary scan	Usually Not Appropriate	☢☢☢

Variant 2:   Presenting with shortness of breath, or hemothorax, or hemoptysis. Patient has history of epistaxis and family history of hereditary hemorrhagic telangiectasia (HHT). Suspected PAVM. Initial imaging.

Procedure	Appropriateness Category	Relative Radiation Level
US echocardiography transthoracic with IV contrast	Usually Appropriate	O
MRA pulmonary arteries without and with IV contrast	Usually Appropriate	O
CT chest with IV contrast	Usually Appropriate	☢☢☢
CT chest without IV contrast	Usually Appropriate	☢☢☢
CTA chest with IV contrast	Usually Appropriate	☢☢☢
CTA pulmonary arteries with IV contrast	Usually Appropriate	☢☢☢
Arteriography pulmonary	May Be Appropriate	☢☢☢☢
MRA chest without and with IV contrast	May Be Appropriate	O
CT chest without and with IV contrast	May Be Appropriate	☢☢☢
US echocardiography transesophageal	Usually Not Appropriate	O
US echocardiography transesophageal with IV contrast	Usually Not Appropriate	O
US echocardiography transthoracic resting	Usually Not Appropriate	O
Radiography chest	Usually Not Appropriate	☢
MRA chest without IV contrast	Usually Not Appropriate	O
MRA pulmonary arteries without IV contrast	Usually Not Appropriate	O
Pertechnetate albumin pulmonary scan	Usually Not Appropriate	☢☢☢

Variant 3:   **Asymptomatic with a family history of HHT and suspected PAVM. Initial imaging.**

Procedure	Appropriateness Category	Relative Radiation Level
US echocardiography transthoracic with IV contrast	Usually Appropriate	O
MRA pulmonary arteries without and with IV contrast	Usually Appropriate	O
CT chest without IV contrast	Usually Appropriate	☢☢☢
CTA chest with IV contrast	Usually Appropriate	☢☢☢
CTA pulmonary arteries with IV contrast	Usually Appropriate	☢☢☢
MRA chest without and with IV contrast	May Be Appropriate	O
CT chest with IV contrast	May Be Appropriate	☢☢☢
CT chest without and with IV contrast	May Be Appropriate	☢☢☢
US echocardiography transesophageal	Usually Not Appropriate	O
US echocardiography transesophageal with IV contrast	Usually Not Appropriate	O
US echocardiography transthoracic resting	Usually Not Appropriate	O
Radiography chest	Usually Not Appropriate	☢
Arteriography pulmonary	Usually Not Appropriate	☢☢☢☢
MRA chest without IV contrast	Usually Not Appropriate	O
MRA pulmonary arteries without IV contrast	Usually Not Appropriate	O
Pertechnetate albumin pulmonary scan	Usually Not Appropriate	☢☢☢

Variant 4: **Presenting to establish care with a past history of a treated PAVM. Follow-up (surveillance) imaging following embolization of PAVM.**

Procedure	Appropriateness Category	Relative Radiation Level
US echocardiography transthoracic with IV contrast	Usually Appropriate	O
MRA pulmonary arteries without and with IV contrast	Usually Appropriate	O
CT chest with IV contrast	Usually Appropriate	☢☢☢
CT chest without and with IV contrast	Usually Appropriate	☢☢☢
CT chest without IV contrast	Usually Appropriate	☢☢☢
CTA chest with IV contrast	Usually Appropriate	☢☢☢
CTA pulmonary arteries with IV contrast	Usually Appropriate	☢☢☢
Arteriography pulmonary	May Be Appropriate	☢☢☢☢
MRA chest without and with IV contrast	May Be Appropriate	O
US echocardiography transesophageal	Usually Not Appropriate	O
US echocardiography transesophageal with IV contrast	Usually Not Appropriate	O
US echocardiography transthoracic resting	Usually Not Appropriate	O
Radiography chest	Usually Not Appropriate	☢
MRA chest without IV contrast	Usually Not Appropriate	O
MRA pulmonary arteries without IV contrast	Usually Not Appropriate	O

Variant 5:   **Asymptomatic with abnormal imaging on CT or chest radiography suggestive of PAVM. Next imaging study.**

Procedure	Appropriateness Category	Relative Radiation Level
US echocardiography transthoracic with IV contrast	Usually Appropriate	O
MRA pulmonary arteries without and with IV contrast	Usually Appropriate	O
CTA pulmonary arteries with IV contrast	Usually Appropriate	☢☢☢
Arteriography pulmonary	May Be Appropriate	☢☢☢☢
MRA chest without and with IV contrast	May Be Appropriate	O
MRA chest without IV contrast	May Be Appropriate	O
CT chest with IV contrast	May Be Appropriate (Disagreement)	☢☢☢
CT chest without and with IV contrast	May Be Appropriate (Disagreement)	☢☢☢
CT chest without IV contrast	May Be Appropriate	☢☢☢
CTA chest with IV contrast	May Be Appropriate (Disagreement)	☢☢☢
US echocardiography transesophageal	Usually Not Appropriate	O
US echocardiography transesophageal with IV contrast	Usually Not Appropriate	O
US echocardiography transthoracic resting	Usually Not Appropriate	O
MRA pulmonary arteries without IV contrast	Usually Not Appropriate	O

PULMONARY ARTERIOVENOUS MALFORMATION (PAVM)

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Summary of Literature Review

Introduction/Background

Pulmonary arteriovenous malformations (PAVMs) are structurally abnormal pulmonary to systemic vascular shunts that result from direct communications between the pulmonary artery and pulmonary vein bypassing the intervening pulmonary capillary bed. The pulmonary capillary bed acts as a sieve measuring 8 to 10 µm in diameter that filters the blood coming from the pulmonary arteries. The physiological consequences of the right to left shunt are impairment of gas exchange and filtration of the systemic venous return [1].

About 70% to 90% of the patients with PAVMs have hereditary hemorrhagic telangiectasia (HHT) [2]. HHT is an autosomal dominant disorder associated with mutations in genes coding for endoglin and activin receptor-like kinase 1ALK1. The former leads to a phenotypical presentation with cerebral and PAVMs described as HHT type 1 [3]. The later presents with pulmonary hypertension and hepatic AVMs and is described as HHT type 2 [4]. Of patients with HHT, 1% to 2% have a SMAD4 mutation and a clinical phenotype associated with juvenile polyposis syndrome [5]. The clinical diagnosis of HHT is based on the Curacao criteria [6]. The prevalence of PAVM occurs in 1 in 5,000 individuals in the general population but can vary depending on the geographical distribution of HHT. In areas where HHT is more prevalent, PAVM prevalence can reach between 26 and 56 per 100,000 individuals. In general, there is a 1.5 to 2 times higher incidence of PAVMs in women compared to men but a male-predominance is noted in newborns [7]. Other causes of PAVMs are rare and include trauma, chest surgery, schistosomiasis, actinomycosis, mitral valve stenosis, Fanconi syndrome, cirrhosis with hepatopulmonary syndrome, and metastatic cancer [7].

Clinical manifestations of PAVMs depend on the size, number, type (complex versus simple), and flow through the malformations. Most patients are asymptomatic (25%-58%). Hypoxemia (27%-71%) at rest or exercise, especially orthodeoxia (worsening hypoxemia when upright) and platypnea (worsening dyspnea when upright), are classical presentations as 65% to 83% of PAVMs are in the lower lobes of the lungs [8]. Transient ischemic attacks and cerebral strokes (3.2%-55%), systemic infections, and abscesses including cerebral abscesses (0%-25%) and rarely massive hemoptysis and hemothorax (0%-2%) are other manifestations [1]. Pregnancy is associated with the rapid growth of PAVMs due to hormonal and hemodynamic consequences with a higher risk of complications from lack of filtration and rupture [9].

Treatment of PAVMs involves endovascular occlusion of the feeding artery and in rare instances surgical resection. Percutaneous transcatheter embolization is typically performed for the treatment of PAVMs. The feeding artery is occluded by an embolic device obliterating the arteriovenous shunt. Regardless of the size of the feeding artery, any PAVM detected by CT or catheter angiography should be considered for treatment due to the risk of paradoxical embolism [10,11]. Embolization is performed by deploying coils or plugs in the feeding artery as close to the

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arteriovenous communication as possible. Follow-up CT angiography (CTA) to detect persistence/new lesions within 6 to 12 months followed by every 3 to 5 years is recommended by the international guidelines for the diagnosis and management of HHT [10]. Persistent perfusion of PAVMs following embolization carries the continued risk of paradoxical embolism and is a vexing problem to re-treat with success rates up to 44% to 85% [12]. The persistence rates vary with the embolic material and are primarily due to arterial recanalization. Other causes include pulmonary to pulmonary and systemic to pulmonary perfusion of the venous sac. Persistent sac perfusion following embolization with different embolic materials vary from 5% to 21% for coils alone, 4% to 6% for nitinol vascular plugs, and 0% to 2% for microvascular plugs [13,14].

Special Imaging Considerations

Systematic screening and treatment of HHT-related organ involvement have shown to improve the survival [15]. Clinical signs and symptoms of PAVMs are often absent before the onset of complications. Transthoracic contrast echocardiography (TTCE) with intravenous (IV) contrast material and CT may be helpful for diagnosis and posttreatment evaluation with good sensitivity for detection of treatment-naïve disease. However, the sensitivity is low to detect recanalization rates after treatment [16,17].

Use of maximum intensity projection postprocessing has shown to increase the detection rates and reduce reporting times for small PAVMs both in children and adults. Maximum intensity projection also detects the anatomy and size of the feeding artery with higher accuracy compared to thin-section conventional CT images [18-20].

Postembolization scans for PAVMs are prone to artifacts that interfere with the evaluation of persistent flow. Spectral and dual-energy CT have been studied in other anatomical locations in reducing the coil and other metallic artifacts [21,22]. There are no studies evaluating their role in PAVM, but these techniques may improve posttreatment image interpretation by reducing artifacts.

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) [23]:

> *"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 (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: Presenting with a transient ischemic attack, or seizures, or brain abscess, or altered sensorium. Chest radiography reveals a lung nodule. Suspected pulmonary arteriovenous malformation (PAVM). Next imaging study.

Arteriography Pulmonary

The real-time nature of pulmonary arteriography allows for high accuracy in delineating the angioarchitecture and detection of flow characteristics such as the early draining vein. In a study comparing the specificity of pulmonary arteriography and CTA, pulmonary arteriography was noted to have a higher specificity for detecting the angioarchitecture compared to CTA (100% versus 78%) [24]. Pulmonary angiography is performed as a part of the

treatment procedure and does not have a standalone diagnostic role in detecting PAVM. An exception where pulmonary angiogram would be helpful as an initial diagnostic imaging tool is in a patient who is hemodynamically unstable with clinical suspicion of pulmonary hemorrhage from a PAVM [25].

CT Chest With IV Contrast
Chest CT with IV contrast offers high spatial resolution and can detect the number, size, and distribution of PAVMs accurately. Contrast-enhanced CT provides similar diagnostic accuracy as noncontrast CT due to the high natural contrast inherent to pulmonary anatomy. IV contrast material administration adds a small risk of air embolism in patients with PAVM. Adequate precaution should be taken to prevent air embolism [10,26]. Cross-sectional anatomy displayed on CT chest with IV contrast is useful in treatment planning [10,27].

CT Chest Without and With IV Contrast
There is limited data to support obtaining a CT chest with and without IV contrast in the setting of suspected PAVM. A study by Nawaz et al [24] compared CT with and without IV contrast to digital subtraction angiography (DSA) for assessment of PAVMs. Their study showed superior sensitivity of CT compared to DSA for detection of PAVM; however, the specificity for CT was inferior to that of DSA. Cross-sectional anatomy displayed on CT chest with IV contrast is useful in treatment planning [10,27]. The benefit of using CT chest without and with IV contrast for assessing PAVM compared to stand-alone CT without IV contrast or CT with IV contrast is unclear [24].

CT Chest Without IV Contrast
Noncontrast chest CT scan is helpful in confirming the diagnosis of PAVM. Like CT chest with IV contrast, Noncontrast CT offers high spatial resolution and can detect the number, size, and distribution of PAVMs accurately. Remy et al [28] were able to predict angioarchitecture of the PAVMs in 95% of cases using noncontrast CT and 3-D reconstruction. Cross-sectional anatomy displayed on CT chest without IV contrast is useful in treatment planning [10,27].

CTA Chest With IV Contrast
CTA chest provides similar diagnostic accuracy as noncontrast CT due to the high natural contrast inherent to pulmonary anatomy. IV contrast material administration adds a small risk of air embolism in patients with PAVM. Adequate precaution should be taken to prevent air embolism. Unlike CTA pulmonary arteries (CTPA), the vascular enhancement during CTA is timed for the aorta and its branches and, thus, it may help identify systemic supply to PAVMs via the systemic arteries [10,29,30]. Cross-sectional anatomy displayed on CT chest with IV contrast is useful in treatment planning [10,27].

CTA Pulmonary Arteries With IV Contrast
CTPA specifically assesses the pulmonary vasculature. IV contrast material is timed for optimum evaluation of the pulmonary arteries. Use of CTPA for evaluation of PAVM is used in clinical practice when considering a contrast-enhanced CT scan for evaluating PAVM. Like other CT techniques, CTPA offers high special resolution and can detect the number, size, and distribution of PAVMs accurately. CTPA provides similar diagnostic accuracy as noncontrast CT due to the high natural contrast inherent to pulmonary anatomy. IV contrast material administration adds a small risk of air embolism in patients with PAVM. Adequate precaution should be taken to prevent air [27]. Correlating the PAVM grade with contrast-enhanced echo has been shown to be more sensitive with CTPA compared to noncontrast CT [31]. Cross-sectional anatomy displayed on CT chest with IV contrast is useful in treatment planning [10,27].

MRA Chest Without and With IV Contrast
There is no role for the routine use of MR angiography (MRA) chest without and with IV contrast in the workup of a patient suspected of a PAVM. Similar to chest CTA, MRA chest may identify systemic arterial supply to PAVM.

MRA Chest Without IV Contrast
There is no role for the routine use of MRA chest without IV contrast in the workup of a patient suspected of a PAVM.

MRA Pulmonary Arteries Without and With IV Contrast
Contrast-enhanced MRA pulmonary arteries provides anatomical information about the presence, number, size, and location of PAVMs. Schneider et al [32] evaluated 203 patients with HHT or first-degree relatives with HHT using contrast-enhanced MRA. Patients with definite and uncertain diagnosis of PAVM on MRA underwent pulmonary angiogram. Pulmonary angiogram detected only 77% to 80% of the PAVMs that were seen on MRA. In their study, the majority of the PAVMs not detected by pulmonary angiogram were <5 mm in size. The size criterion was

defined as the size of the PAVM itself and not of the feeding artery. A more recent study by Van den Heuvel et al [33] investigated the sensitivity of contrast-enhanced MRA for detection of PAVMs with a feeding artery >2 cm in children and young adults. They enrolled 53 patients who had a TTCE grade 2 or 3 who underwent chest CT and were found to have the PAVM with a feeding artery >2 cm to receive a contrast-enhanced MRA. The sensitivity of contrast-enhanced MRA to detect PAVMs with a feeding artery size of >2cm was 92%, and the specificity ranged from 67% to 96%.

MRA Pulmonary Arteries Without IV Contrast
There is no role for the routine use of MRA pulmonary angiography without IV contrast in the workup of a patient suspected of a PAVM.

Pertechnetate Albumin Pulmonary Scan
There is no role for pertechnetate albumin pulmonary scan in a modern day practice. Historically, this technique was used to detect and quantify right to left shunting before and after treatment of a PAVM [34,35].

Radiography Chest
The radiographic appearance of a lower lobe pulmonary nodule with a branching afferent artery and dilated efferent vein defines the classical appearance of PAVM on chest radiography. The sensitivity of chest radiography is 60% to 70% with a 98% specificity when the classical findings are present [36]. The afferent and efferent vasculature and smaller PAVMs may be difficult to see on a single-view chest radiograph. Best diagnostic results are obtained when a 2-view chest radiograph, posteroanterior view, and lateral view, is performed [37]. The role of chest radiography in the diagnosis of a suspected PAVM is limited due to its poor sensitivity.

US Echocardiography Transesophageal
There is no role for ultrasound (US) transesophageal echocardiography (TEE) as a standalone diagnostic tool in the evaluation of PAVMs. The usefulness of TEE in the context of PAVM is to rule out intracardiac shunts [38]. Its ability to demonstrate the interatrial septum and the insertion of the pulmonary veins into the left atrium is useful to evaluate the anatomical variations [39].

US Echocardiography Transesophageal With IV Contrast
TEE with IV agitated saline contrast material is not routinely used to diagnose PAVM. Contrast-enhanced TEE may be helpful to locate a PAVM based on the excellent visualization of the 4 pulmonary venous ostia as veins drain into the left atrium. Based on the visualization of contrast material emanating from a particular pulmonary vein, the location of the PAVM in that venous territory can be confirmed [38,40]. In the presence of multiple PAVMs, the usefulness of this imaging modality in identifying the location of the PAVMs is limited.

US Echocardiography Transthoracic Resting
There is no role for transthoracic echocardiography (TTE) in the resting phase for the evaluation of PAVM. It does allow evaluation of intracardiac shunts and assessment of cardiac function [38].

US Echocardiography Transthoracic With IV Contrast
TTCE is an essential diagnostic test for patients suspected of having a PAVM. TTCE with agitated saline has a 98% to 99% sensitivity and a 67% to 91% specificity for detecting PAVMs [41]. The microbubbles are visualized after 3 to 8 cardiac cycles in the left atrium after initial opacification of the right chambers in patients with an intrapulmonary shunt [1]. TTCE does not provide any information regarding the size and location of the PAVM. Based on the appearance of the bubbles in the left atrium a semiquantitative grading system has been developed [42,43]. The grades are defined as 0 with no opacification, grade 1 with <30 bubbles, grade 2 with moderate filling, and grade 3 with complete opacification of the left atrium. The grading system correlates well with the diagnosis of PAVM, with higher grades associated with larger shunts and cerebral complications [41,44,45]. Usefulness of the grading system to predict treatment of PAVM demonstrates that grades 2 and 3 have a positive predictive value of 0.21 (95% confidence interval [CI], 0.05-0.36) and 0.87 (95% CI, 0.79-0.99), respectively [44]. Adverse events including air embolism are rare with TTCE occurring in <1% [46].

Variant 2: Presenting with shortness of breath, or hemothorax, or hemoptysis. Patient has history of epistaxis and family history of hereditary hemorrhagic telangiectasia (HHT). Suspected PAVM. Initial imaging.
Arteriography Pulmonary
The real-time nature of pulmonary arteriography allows for high accuracy in delineating the angioarchitecture and detection of flow characteristics such as the early draining vein. In a study comparing the specificity of pulmonary arteriography and CTA, pulmonary arteriography was noted to have a higher specificity for detecting the

angioarchitecture compared to CTA (100% versus 78%) [24]. Pulmonary angiography is performed as a part of the treatment procedure and does not have a standalone diagnostic role in detecting PAVM. An exception in which pulmonary angiogram would be helpful as an initial diagnostic imaging tool is in a patient who is hemodynamically unstable with clinical suspicion of pulmonary hemorrhage from a PAVM [25].

CT Chest With IV Contrast
Contrast-enhanced CT chest offers high spatial resolution and can detect the number, size, and distribution of PAVMs accurately. Contrast-enhanced CT provides similar diagnostic accuracy as noncontrast CT due to the high natural contrast inherent to pulmonary anatomy. IV contrast material administration adds a small risk of air embolism in patients with PAVM. Adequate precaution should be taken to prevent air embolism [10,26]. Cross-sectional anatomy displayed on CT chest with IV contrast is useful in treatment planning [10,27].

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CTA Chest With IV Contrast
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CTA Pulmonary Arteries With IV Contrast
CTPA specifically assesses the pulmonary vasculature. IV contrast material is timed for optimum evaluation of the pulmonary arteries. Use of CTPA for evaluation of PAVM is used in clinical practice when considering a contrast-enhanced CT scan for evaluating PAVM. Like other CT techniques, CTPA offers high special resolution and can detect the number, size, and distribution of PAVMs accurately. CTPA provides similar diagnostic accuracy as noncontrast CT due to the high natural contrast inherent to pulmonary anatomy. IV contrast material administration adds a small risk of air embolism in patients with PAVM. Adequate precaution should be taken to prevent air [27]. Correlating the PAVM grade with contrast-enhanced echo has been shown to be more sensitive with CTPA compared to noncontrast CT [31]. Cross-sectional anatomy displayed on CT chest with IV contrast is useful in treatment planning [10,27].

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the majority of the PAVMs not detected by pulmonary angiogram were <5 mm in size. The size criterion was defined as the size of the PAVM itself and not of the feeding artery. A more recent study by Van den Heuvel et al [33] investigated the sensitivity of contrast-enhanced MRA for detection of PAVMs with a feeding artery >2 cm in children and young adults. They enrolled 53 patients who had a TTCE grade 2 or 3 who underwent chest CT and were found to have the PAVM with a feeding artery >2 cm to receive a contrast-enhanced MRA. The sensitivity of contrast-enhanced MRA to detect PAVMs with a feeding artery size of >2cm was 92%, and the specificity ranged from 67% to 96%.

MRA Pulmonary Arteries Without IV Contrast
There is no role for the routine use of MRA pulmonary angiography without IV contrast in the workup of a patient suspected of a PAVM.

Pertechnetate Albumin Pulmonary Scan
There is no role for pertechnetate albumin pulmonary scan in a modern day practice. Historically, this technique was used to detect and quantify right to left shunting before and after treatment of a PAVM [34,35].

Radiography Chest
The radiographic appearance of a lower lobe pulmonary nodule with a branching afferent artery and dilated efferent vein defines the classical appearance of PAVM on chest radiography. The sensitivity of chest radiography is 60% to 70% with a 98% specificity when the classical findings are present [36]. The afferent and efferent vasculature and smaller PAVMs may be difficult to see on a single-view chest radiograph. Best diagnostic results are obtained when a 2-view chest radiograph, posteroanterior view, and lateral view is performed [37]. The role of chest radiography in the diagnosis of a suspected PAVM is limited due to its poor sensitivity.

US Echocardiography Transesophageal
There is no role for TEE as a standalone diagnostic tool in the evaluation of PAVMs. The usefulness of TEE in the context of PAVM is to rule out intracardiac shunts [38]. Its ability to demonstrate the interatrial septum and the insertion of the pulmonary veins into the left atrium is useful to evaluate the anatomical variations [39].

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There is no role for TTE in the resting phase for evaluation of PAVM. It does allow evaluation of intracardiac shunts and assessment of cardiac function [38].

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TTCE is an essential diagnostic test for patients suspected of having a PAVM. TTCE with agitated saline has a 98% to 99% sensitivity and a 67% to 91% specificity for detecting PAVMs [41]. The microbubbles are visualized after 3 to 8 cardiac cycles in the left atrium after initial opacification of the right chambers in patients with an intrapulmonary shunt [1]. TTCE does not provide any information regarding the size and location of the PAVM. Based on the appearance of the bubbles in the left atrium a semiquantitative grading system has been developed [42,43]. The grades are defined as 0, with no opacification, grade 1 with <30 bubbles, grade 2 with moderate filling, and grade 3 with complete opacification of the left atrium. The grading system correlates well with the diagnosis of PAVM, with higher grades associated with larger shunts and cerebral complications [41,44,45]. Usefulness of the grading system to predict treatment of PAVM demonstrates that grades 2 and 3 have a positive predictive value of 0.21 (95% CI, 0.05-0.36) and 0.87 (95% CI, 0.79-0.99), respectively [44]. Adverse events including air embolism are rare with TTCE occurring in <1% [46].

Variant 3: Asymptomatic with a family history of HHT and suspected PAVM. Initial imaging.

Arteriography Pulmonary
The real-time nature of pulmonary arteriography allows for high accuracy in delineating the angioarchitecture and detection of flow characteristics such as the early draining vein. In a study comparing the specificity of pulmonary arteriography and CTA, pulmonary arteriography was noted to have a higher specificity for detecting the

angioarchitecture compared to CTA (100% versus 78%) [24]. Pulmonary angiography is performed as a part of the treatment procedure and does not have a standalone diagnostic role in detecting PAVM.

CT Chest With IV Contrast
Contrast-enhanced CT chest offers high spatial resolution and can detect the number, size, and distribution of PAVMs accurately. Contrast-enhanced CT provides similar diagnostic accuracy as noncontrast CT due to the high natural contrast inherent to pulmonary anatomy. IV contrast material administration adds a small risk of air embolism in patients with PAVM. Adequate precaution should be taken to prevent air embolism [10,26]. Cross-sectional anatomy displayed on CT chest with IV contrast is useful in treatment planning [10,27].

CT Chest Without and With IV Contrast
There is limited data to support obtaining a CT chest with and without IV contrast in the setting of suspected PAVM. A study by Nawaz et al [24] compared CT with and without IV contrast to DSA for assessment of PAVMs. Their study showed superior sensitivity of CT compared to DSA for detection of PAVM; however, the specificity for CT was inferior to that of DSA. Cross-sectional anatomy displayed on CT chest with IV contrast is useful in treatment planning [10,27]. The benefit of using CT chest without and with IV contrast for assessing PAVM compared to stand-alone CT without IV contrast or CT with IV contrast is unclear [24].

CT Chest Without IV Contrast
Noncontrast chest CT scan is helpful in confirming the diagnosis of PAVM. Like CT chest with IV contrast, noncontrast CT offers high spatial resolution and can detect the number, size, and distribution of PAVMs accurately. Remy et al [28] were able to predict angioarchitecture of the PAVMs in 95% of cases using noncontrast CT and 3-D reconstruction. Cross-sectional anatomy displayed on CT chest without IV contrast is useful in treatment planning [10,27].

CTA Chest With IV Contrast
CTA chest provides similar diagnostic accuracy as noncontrast CT due to the high natural contrast inherent to pulmonary anatomy. IV contrast material administration adds a small risk of air embolism in patients with PAVM. Adequate precaution should be taken to prevent air embolism. Unlike CTPA, the vascular enhancement during CTA is timed for the aorta and its branches and, thus, it may help identify systemic supply to PAVMs via the systemic arteries [10,29,30]. Cross-sectional anatomy displayed on CT chest with IV contrast is useful in treatment planning [10,27].

CTA Pulmonary Arteries With IV Contrast
CTPA specifically assesses the pulmonary vasculature. IV contrast material is timed for optimum evaluation of the pulmonary arteries. Use of CTPA for evaluation of PAVM is used in clinical practice when considering a contrast-enhanced CT scan for evaluating PAVM. Like other CT techniques, CTPA offers high special resolution and can detect the number, size, and distribution of PAVMs accurately. CTPA provides similar diagnostic accuracy as noncontrast CT due to the high natural contrast inherent to pulmonary anatomy. IV contrast material administration adds a small risk of air embolism in patients with PAVM. Adequate precaution should be taken to prevent air [27]. Correlating the PAVM grade with contrast-enhanced echo has been shown to be more sensitive with CTPA compared to noncontrast CT [31]. Cross-sectional anatomy displayed on CT chest with IV contrast is useful in treatment planning [10,27].

MRA Chest Without and With IV Contrast
There is no role for the routine use of MRA chest without and with IV contrast in the workup of a patient suspected of a PAVM.

MRA Chest Without IV Contrast
There is no role for the routine use of MRA chest without IV contrast in the workup of a patient suspected of a PAVM.

MRA Pulmonary Arteries Without and With IV Contrast
Contrast-enhanced MRA pulmonary arteries provides anatomical information about the presence, number, size, and location of PAVMs. Schneider et al [32] evaluated 203 patients with HHT or first-degree relatives with HHT using contrast-enhanced MRA. Patients with definite and uncertain diagnosis of PAVM on MRA underwent pulmonary angiogram. Pulmonary angiogram detected only 77% to 80% of the PAVMs that were seen on MRA. In their study, the majority of the PAVMs not detected by pulmonary angiogram were <5 mm in size. The size criterion was defined as the size of the PAVM itself and not of the feeding artery. A more recent study by Van den Heuvel et al

[33] investigated the sensitivity of contrast-enhanced MRA for detection of PAVMs with a feeding artery >2 cm in children and young adults. They enrolled 53 patients who had a TTCE grade 2 or 3 who underwent chest CT and were found to have the PAVM with a feeding artery >2 cm to receive a contrast-enhanced MRA. The sensitivity of contrast-enhanced MRA to detect PAVMs with a feeding artery size of >2cm was 92%, and the specificity ranged from 67% to 96%.

MRA Pulmonary Arteries Without IV Contrast
There is no role for the routine use of MRA pulmonary angiography without IV contrast in the workup of a patient suspected of a PAVM.

Pertechnetate Albumin Pulmonary Scan
There is no role for pertechnetate albumin pulmonary scan in a modern day practice. Historically, this technique was used to detect and quantify right to left shunting before and after treatment of a PAVM [34,35].

Radiography Chest
The radiographic appearance of a lower lobe pulmonary nodule with a branching afferent artery and dilated efferent vein defines the classical appearance of PAVM on chest radiography. The sensitivity of chest radiography is 60% to 70% with a 98% specificity when the classical findings are present [36]. The afferent and efferent vasculature and smaller PAVMs may be difficult to see on a single-view chest radiograph. Best diagnostic results are obtained when a 2-view chest radiograph, posteroanterior view, and lateral view is performed [37].

US Echocardiography Transesophageal
There is no role for TEE as a standalone diagnostic tool in the evaluation of PAVMs. The usefulness of TEE in the context of PAVM is to rule out intracardiac shunts [38]. Its ability to demonstrate the interatrial septum and the insertion of the pulmonary veins into the left atrium is useful to evaluate the anatomical variations [39].

US Echocardiography Transesophageal With IV Contrast
TEE with IV agitated saline contrast material is not routinely used to diagnose PAVM. Contrast-enhanced TEE may be helpful to locate a PAVM based on the excellent visualization of the 4 pulmonary venous ostia as veins drain into the left atrium. Based on the visualization of contrast material emanating from a particular pulmonary vein, the location of the PAVM in that venous territory can be confirmed [38,40]. In the presence of multiple PAVMs, the usefulness of this imaging modality in identifying the location of the PAVMs is limited.

US Echocardiography Transthoracic Resting
There is no role for TTE in the resting phase for the evaluation of PAVM. It does allow evaluation of intracardiac shunts and assessment of cardiac function [38].

US Echocardiography Transthoracic With IV Contrast
TTCE is an essential diagnostic test for patients suspected of having a PAVM. TTCE with agitated saline has a 98% to 99% sensitivity and a 67% to 91% specificity for detecting PAVMs [41]. The microbubbles are visualized after 3 to 8 cardiac cycles in the left atrium after initial opacification of the right chambers in patients with an intrapulmonary shunt [1]. TTCE does not provide any information regarding the size and location of the PAVM. Based on the appearance of the bubbles in the left atrium a semiquantitative grading system has been developed [42,43]. The grades are defined as 0 with no opacification, grade 1 with <30 bubbles, grade 2 with moderate filling, and grade 3 with complete opacification of the left atrium. The grading system correlates well with the diagnosis of PAVM, with higher grades associated with larger shunts and cerebral complications [41,44,45]. Usefulness of the grading system to predict treatment of PAVM demonstrates that grades 2 and 3 have a positive predictive value of 0.21 (95% CI, 0.05-0.36) and 0.87 (95% CI, 0.79-0.99), respectively [44]. Adverse events including air embolism are rare with TTCE occurring in <1% [46].

Variant 4: Presenting to establish care with a past history of a treated PAVM. Follow-up (surveillance) imaging following embolization of PAVM.

Arteriography Pulmonary
The real-time nature of pulmonary arteriography allows for high accuracy for delineating the angioarchitecture and detection of flow characteristics such as the early draining vein. Pulmonary angiogram is the reference standard to detect reperfusion of a treated AVM, but the angiogram is performed as a part of the treatment protocol and not done as a diagnostic test [10]. In patients who have inconclusive findings on CT, pulmonary angiography can assess recanalized PAVMs.

CT Chest With IV Contrast

The high natural contrast inherent to pulmonary anatomy on noncontrast CT and the small risk of air embolism while administering IV medications renders CT chest with IV contrast unnecessary in the diagnosis of PAVM [10]. Artifacts from the embolic material limits the usefulness of contrast enhancement as a tool to evaluate reperfusion [47]. The role of contrast-enhanced CT has been studied after embolotherapy for PAVM in the context of detecting systemic collaterals by Brillet et al [29] in 32 patients. They found systemic collaterals in 13 patients that would otherwise have been missed on noncontrast CT. The role of contrast-enhanced CT scan after PAVM embolization is unclear. Cross-sectional anatomy displayed on CT chest with IV contrast is useful in further treatment planning if new or recanalized PAVMs are detected [10,27].

CT Chest Without and With IV Contrast

The high natural contrast inherent to pulmonary anatomy on noncontrast CT and the small risk of air embolism while administering IV medications renders CT chest with IV contrast unnecessary in the diagnosis of PAVM [10]. Artifacts from the embolic material limits the usefulness of contrast enhancement as a tool to evaluate reperfusion [47]. The role of contrast-enhanced CT has been studied after embolotherapy for PAVM in the context of detecting systemic collaterals by Brillet et al [29] in 32 patients. They found systemic collaterals in 13 patients that would otherwise have been missed on noncontrast CT. The role of contrast-enhanced CT scan after PAVM embolization is unclear. Cross-sectional anatomy displayed on CT chest with IV contrast is useful in further treatment planning if new or recanalized PAVMs are detected [10,27].

CT Chest Without IV Contrast

Noncontrast chest CT scan uses the reduction in the size of the sac/draining vein as a predictor for successful embolization. Two different size criteria have been proposed, >70%reduction in the size of the sac [48-50] and >30% reduction in the size of the draining vein [12,30,51]. Both criteria are arbitrary and not validated. Belanger et al [47] compared the sensitivity and specificity of both these size criteria in 108 treated PAVMs with pulmonary angiography as the reference standard. When 70% criterion was used, the sensitivity was between 98% to 100%, and the specificity was 20% to 47%. When the 30% reduction criterion was used, the sensitivity was between 51% to 56%, and the specificity was 86% to 98%. Similar conclusions have been reported by other studies [52]. Another study suggested a draining vein diameter of ≥2.5 mm as a marker for reperfusion of a PAVM postembolization [53]. Cross-sectional anatomy displayed on CT chest with IV contrast is useful in further treatment planning if new or recanalized PAVMs are detected [10,27].

CTA Chest With IV Contrast

CTA also provides similar diagnostic accuracy as noncontrast CT due to the high natural contrast inherent to pulmonary anatomy. Contrast administration adds a small risk of air embolism in patients with PAVM. Adequate precaution should be taken to prevent air embolism. CTA may help identify systemic supply to large PAVMs via the systemic arteries [10,29,30]. Artifacts from the embolic material limit the usefulness of contrast enhancement as a tool to evaluate the reperfusion [47]. The role of contrast-enhanced CT has been studied after embolotherapy for PAVM in the context of detecting systemic collaterals by Brillet et al [29] in 32 patients. They found systemic collaterals in 13 patients that would otherwise have been missed on noncontrast CT. Another study by Remy-Jardin et at [30] showed that long-term CTA follow-up of initially successfully treated PAVMs revealed successful embolotherapy of 75% and partially or completely failed embolotherapy of 25% of PAVMs. Cross-sectional anatomy displayed on CT chest with IV contrast is useful in further treatment planning if new or recanalized PAVMs are detected [10,27].

CTA Pulmonary Arteries With IV Contrast

CTPA specifically assesses the pulmonary vasculature. IV contrast is timed for optimum evaluation of the pulmonary arteries. Although, there is limited literature specific to CTA of pulmonary arteries for evaluation of PAVM. This protocol is used in clinical practice when considering contrast-enhanced CT scan for evaluating PAVM. Like other CT techniques, CTPA offers high special resolution and can detect the number, size, and distribution of PAVMs accurately. CTPA provides similar diagnostic accuracy as noncontrast CT due to the high natural contrast inherent to pulmonary anatomy. IV contrast material administration adds a small risk of air embolism in patients with PAVM. Adequate precaution should be taken to prevent air [27]. Correlating the PAVM grade with contrast-enhanced echo has been shown to be more sensitive with CTPA compared to noncontrast CT [31].

Strategies to mitigate the artifacts from embolic material by adjusting scan parameters and using dual-energy and spectral CT acquisition have shown promise in other anatomical locations. For instance, Zheng et al [54] showed

that spectral CT using metal artifact reduction software improved vascular evaluation of adjacent metallic coils in the brain. Visualization of recanalized flow remains a challenge in posttreatment CT evaluation for reperfusion.

MRA Chest Without and With IV Contrast
There is no role for the routine use of MRA chest without and with IV contrast in the workup of a patient with treated PAVM.

MRA Chest Without IV Contrast
There is no role for the routine use of MRA chest without IV contrast in the workup of a patient with treated PAVM.

MRA Pulmonary Arteries Without and With IV Contrast
Dynamic MRA using time-resolved sequences has the advantage over CT, and it is not affected by the artifact from the embolic material. Time-resolved MRA can detect flow in the pulmonary vein similar to pulmonary arteriography. Shimohira et al [55] compared the sensitivity and specificity of CT and time-resolved MRA with digital subtraction angiography as a reference standard in a multi-institutional retrospective cohort of 205 patients. Using 55% as the threshold reduction of the sac diameter on CT, they found a sensitivity and specificity for CT at 82% and 81%, respectively. Time-resolved MRA on the other hand has a sensitivity and specificity of 89% and 95%, respectively. Similar findings have been reported in 28 PAVMs in 10 patients by Kawai et al [56]. In a recent study, Hong et al [57] studied the feasibility of time-resolved MRA in evaluating the feeding arteries and reperfusion after embolization of PAVMs in 9 patients with 62 PAVMs. They found a sensitivity and specificity of 100% for detection of reperfusion compared to catheter angiography as the reference standard. The visibility of the feeding arteries was compared to CT, and they found good correlation between 0.76 and 0.79 with 2 readers when using 3-D maximum intensity projection images. Time-resolved MRA also has the advantage of detecting reperfusion after repeat embolization for persistence as the burden of coiling will be greater leading to more metallic artifacts on CT [58].

MRA Pulmonary Arteries Without IV Contrast
There is no role for the routine use of MRA pulmonary angiography without IV contrast in the workup of a patient with treated PAVM.

Radiography Chest
The radiographic appearance of a lower lobe pulmonary nodule with a branching afferent artery and dilated efferent vein defines the classical appearance of PAVM on chest radiography. The sensitivity of chest radiography is 60% to 70% with a 98% specificity when the classical findings are present [36]. The afferent and efferent vasculature and smaller PAVMs may be difficult to see on a single-view chest radiograph. Best diagnostic results are obtained when a 2-view chest radiograph, posteroanterior view, and lateral view is performed [37]. Chest radiography has limited usefulness to detect recurrent disease in the setting of prior treated PAVM.

US Echocardiography Transesophageal
There is no role for TEE as a standalone diagnostic tool in the evaluation of PAVMs. The usefulness of TEE in the context of PAVM is to rule out intracardiac shunts [38]. Its ability to demonstrate the interatrial septum and the insertion of the pulmonary veins into the left atrium is useful to evaluate the anatomical variations [39].

US Echocardiography Transesophageal With IV Contrast
TEE with IV agitated saline contrast material is not routinely used to diagnose PAVM. Contrast-enhanced TEE maybe helpful to locate a PAVM based on the excellent visualization of the 4 pulmonary venous ostia as veins drain into the left atrium. Based on the visualization of contrast material emanating from a particular pulmonary vein, the location of the PAVM in that venous territory can be confirmed [38,40]. In the presence of multiple PAVMs, the usefulness of this imaging modality in identifying the location of the PAVMs may be limited.

US Echocardiography Transthoracic Resting
There is no role for TTE in the resting phase for evaluation of PAVM. It does allow evaluation of intracardiac shunts and assessment of cardiac function [38].

US Echocardiography Transthoracic With IV Contrast
TTCE is an essential diagnostic test for patients suspected of having a PAVM. TTCE with agitated saline has a 98% to 99% sensitivity and a 67% to 91% specificity for detecting PAVMs [41]. The microbubbles are visualized after 3 to 8 cardiac cycles in the left atrium after initial opacification of the right chambers in patients with an intrapulmonary shunt [1]. TTCE does not provide any information regarding the size and location of the PAVM. Based on the appearance of the bubbles in the left atrium a semiquantitative grading system has been developed

[42,43]. The grades are defined as 0 with no opacification, grade 1 with <30 bubbles, grade 2 with moderate filling, and grade 3 with complete opacification of the left atrium. The grading system correlates well with the diagnosis of PAVM, with higher grades associated with larger shunts and cerebral complications [41,44,45]. Usefulness of the grading system to predict treatment of PAVM demonstrates that grades 2 and 3 have a positive predictive value of 0.21 (95% CI, 0.05-0.36) and 0.87 (95% CI, 0.79-0.99), respectively [44]. Adverse events including air embolism are rare with TTCE occurring in <1% [46]. However, as a tool to measure the effectiveness of prior treatment, TTCE has low specificity and sensitivity.

Variant 5: Asymptomatic with abnormal imaging on CT or chest radiography suggestive of PAVM. Next imaging study.

Arteriography Pulmonary
The real-time nature of pulmonary arteriography allows for high accuracy in delineating the angioarchitecture and detection of flow characteristics such as the early draining vein. In a study comparing the specificity of pulmonary arteriography and CTA, pulmonary arteriography was noted to have a higher specificity for detecting the angioarchitecture compared to CTA (100% versus 78%) [24]. Pulmonary angiography is performed as a part of the treatment procedure and does not have a standalone diagnostic role in detecting PAVM. An exception where pulmonary angiogram would be helpful as an initial diagnostic imaging tool is in a patient who is hemodynamically unstable with clinical suspicion of pulmonary hemorrhage from a PAVM [25].

CT Chest With IV Contrast
Contrast-enhanced CT chest offers high spatial resolution and can detect the number, size, and distribution of PAVMs accurately. Contrast-enhanced CT provides similar diagnostic accuracy as noncontrast CT due to the high natural contrast inherent to pulmonary anatomy. IV contrast material administration adds a small risk of air embolism in patients with PAVM. Adequate precaution should be taken to prevent air embolism [10,26]. Cross-sectional anatomy displayed on CT chest with IV contrast is useful in treatment planning [10,27].

CT Chest Without and With IV Contrast
There is limited data to support obtaining a CT chest with and without IV contrast in the setting of suspected PAVM. A study by Nawaz et al [24] compared CT with and without IV contrast to DSA for assessment of PAVMs. Their study showed superior sensitivity of CT compared to DSA for detection of PAVM; however, the specificity for CT was inferior to that of DSA. Cross-sectional anatomy displayed on CT chest with IV contrast is useful in treatment planning [10,27]. The benefit of using CT chest without and with IV contrast for assessing PAVM compared to stand-alone CT without IV contrast or CT with IV contrast is unclear [24].

CT Chest Without IV Contrast
Noncontrast chest CT scan is helpful in confirming the diagnosis of PAVM. Like CT chest with IV contrast, Noncontrast CT offers high spatial resolution and can detect the number, size, and distribution of PAVMs accurately. Remy et al [28] were able to predict angioarchitecture of the PAVMs in 95% of cases using noncontrast CT and 3-D reconstruction. Cross-sectional anatomy displayed on CT chest without IV contrast is useful in treatment planning [10,27].

CTA Chest With IV Contrast
CTA chest also provides similar diagnostic accuracy as noncontrast CT due to the high natural contrast inherent to pulmonary anatomy. IV contrast material administration adds a small risk of air embolism in patients with PAVM. Adequate precaution should be taken to prevent air embolism. Unlike CTPA, the vascular enhancement during CTA is timed for the aorta and its branches, and, thus, it may help identify systemic supply to PAVMs via the systemic arteries [10,29,30]. Cross-sectional anatomy displayed on CT chest with IV contrast is useful in treatment planning [10,27].

CTA Pulmonary Arteries With IV Contrast
CTPA specifically assesses the pulmonary vasculature. IV contrast material is timed for optimum evaluation of the pulmonary arteries. Use of CTPA for evaluation of PAVM is used in clinical practice when considering a contrast-enhanced CT scan for evaluating PAVM. Like other CT techniques, CTPA offers high special resolution and can detect the number, size, and distribution of PAVMs accurately. CTPA provides similar diagnostic accuracy as noncontrast CT due to the high natural contrast inherent to pulmonary anatomy. IV contrast material administration adds a small risk of air embolism in patients with PAVM. Adequate precaution should be taken to prevent air [27]. Correlating the PAVM grade with contrast-enhanced echo has been shown to be more sensitive with CTPA

compared to noncontrast CT [31]. Cross-sectional anatomy displayed on CT chest with IV contrast is useful in treatment planning [10,27].

MRA Chest Without and With IV Contrast
There is no role for the routine use of MRA chest without and with IV contrast in the workup of a patient suspected of a PAVM.

MRA Chest Without IV Contrast
There is no role for the routine use of MRA chest without IV contrast in the workup of a patient suspected of a PAVM.

MRA Pulmonary Arteries Without and With IV Contrast
Contrast-enhanced MRA pulmonary arteries provides anatomical information about the presence, number, size, and location of PAVMs. Schneider et al [32] evaluated 203 patients with HHT or first-degree relatives with HHT using contrast-enhanced MRA. Patients with definite and uncertain diagnosis of PAVM on MRA underwent pulmonary angiogram. Pulmonary angiogram detected only 77% to 80% of the PAVMs that were seen on MRA. In their study, the majority of the PAVMs not detected by pulmonary angiogram were <5 mm in size. The size criterion was defined as the size of the PAVM itself and not of the feeding artery. A more recent study by Van den Heuvel et al [33] investigated the sensitivity of contrast-enhanced MRA for detection of PAVMs with a feeding artery >2 cm in children and young adults. They enrolled 53 patients who had a TTCE grade 2 or 3 who underwent chest CT and were found to have the PAVM with a feeding artery >2 cm to receive a contrast-enhanced MRA. The sensitivity of contrast-enhanced MRA to detect PAVMs with a feeding artery size of >2cm was 92%, and the specificity ranged from 67% to 96%.

MRA Pulmonary Arteries Without IV Contrast
There is no role for the routine use of MRA pulmonary angiography without IV contrast in the workup of a patient suspected of a PAVM.

US Echocardiography Transesophageal
There is no role for TEE as a standalone diagnostic tool in the evaluation of PAVMs. The usefulness of TEE in the context of PAVM is to rule out intracardiac shunts [38]. Its ability to demonstrate the interatrial septum and the insertion of the pulmonary veins into the left atrium is useful to evaluate the anatomical variations [39].

US Echocardiography Transesophageal With IV Contrast
TEE with IV agitated saline contrast material is not routinely used to diagnose PAVM. Contrast-enhanced TEE maybe helpful to locate a PAVM based on the excellent visualization of the 4 pulmonary venous ostia as veins drain into the left atrium. Based on the visualization of contrast material emanating from a particular pulmonary vein, the location of the PAVM in that venous territory can be confirmed [38,40]. In the presence of multiple PAVMs, the usefulness of this imaging modality in identifying the location of the PAVMs may be limited.

US Echocardiography Transthoracic Resting
There is no role for TTE in the resting phase for the evaluation of PAVM. It does allow evaluation of intracardiac shunts and assessment of cardiac function [38].

US Echocardiography Transthoracic With IV Contrast
TTCE is an essential diagnostic test for patients suspected of having a PAVM. TTCE with agitated saline has a 98% to 99% sensitivity and a 67% to 91% specificity for detecting PAVMs [41]. The microbubbles are visualized after 3 to 8 cardiac cycles in the left atrium after initial opacification of the right chambers in patients with an intrapulmonary shunt [1]. TTCE does not provide any information regarding the size and location of the PAVM. Based on the appearance of the bubbles in the left atrium a semiquantitative grading system has been developed [42,43]. The grades are defined as 0 with no opacification, grade 1 with <30 bubbles, grade 2 with moderate filling, and grade 3 with complete opacification of the left atrium. The grading system correlates well with the diagnosis of PAVM, with higher grades associated with larger shunts and cerebral complications [41,44,45]. Usefulness of the grading system to predict treatment of PAVM demonstrates that grades 2 and 3 have a positive predictive value of 0.21 (95% CI, 0.05-0.36) and 0.87 (95% CI, 0.79-0.99), respectively [44]. Adverse events including air embolism are rare with TTCE occurring in <1% [46].

Summary of Recommendations

- **Variant 1**: US TTCE is usually appropriate for a next imaging study in a patient presenting with a transient ischemic attack, or seizures, or brain abscess, or altered sensorium. The chest radiograph reveals a lung nodule

and the patient has suspected PAVM. MRA pulmonary arteries without and with IV contrast, CT chest without IV contrast, or CTA pulmonary arteries with IV contrast are complementary procedures each of which are equivalent alternatives (ie, only one procedure will be ordered to provide the clinical information to effectively manage the patient's care). The panel did not agree on recommending MRA chest without and with IV contrast, CT chest without and with IV contrast, or CTA chest with IV contrast for this clinical scenario. There is insufficient medical literature to conclude whether or not these patients would benefit from these procedures in this scenario. Imaging with these procedures in this patient population is controversial but may be appropriate.

- **Variant 2**: US TTCE is usually appropriate for the initial imaging in a patient presenting with shortness of breath, or hemothorax, or hemoptysis. The patient has a history of epistaxis and family history of HHT and, therefore, has suspected PAVM. MRA pulmonary arteries without and with IV contrast, CT chest with IV contrast, CT chest without IV contrast, CTA chest with IV contrast, or CTA pulmonary arteries with IV contrast are complementary procedures each of which are equivalent alternatives (ie, only one procedure will be ordered to provide the clinical information to effectively manage the patient's care).

- **Variant 3**: US TTCE is usually appropriate for the initial imaging in a patient that is asymptomatic with a family history of HHT and has suspected PAVM. MRA pulmonary arteries without and with IV contrast, CT chest without IV contrast, CTA chest with IV contrast, or CTA pulmonary arteries with IV contrast are complementary procedures each of which are equivalent alternatives (ie, only one procedure will be ordered to provide the clinical information to effectively manage the patient's care).

- **Variant 4**: US TTCE, MRA pulmonary arteries without and with IV contrast, CT chest with IV contrast, CT chest without and with IV contrast, CT chest without IV contrast, CTA chest with IV contrast, or CTA pulmonary arteries with IV contrast is usually appropriate for the follow-up (surveillance) imaging in a patient following embolization of PAVM. The patient is presenting to establish care with a past history of a treated PAVM. These procedures are complementary alternatives (ie, one or more procedure will be ordered to provide the clinical information to effectively manage the patient's care).

- **Variant 5**: US TTCE, MRA pulmonary arteries without and with IV contrast, or CTA pulmonary arteries with IV contrast is usually appropriate in a next imaging study in a patient asymptomatic with abnormal imaging on CT or chest radiography suggestive of PAVM. These procedures are complementary alternatives (ie, one or more procedure will be ordered to provide the clinical information to effectively manage the patient's care). The panel did not agree on recommending CT chest with IV contrast, CT chest without and with IV contrast, or CTA chest with IV contrast for this clinical scenario. There is insufficient medical literature to conclude whether or not these patients would benefit from these procedures in this scenario. Imaging with these procedures 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 go to www.acr.org/ac.

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 [59].

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."		

References

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