

**Suspected Pulmonary Hypertension
EVIDENCE TABLE**

Reference	Study Type	Patients/ Events	Study Objective (Purpose of Study)	Study Results	Study Quality
1. Alassas K, Mergo P, Ibrahim el S, et al. Cardiac MRI as a diagnostic tool in pulmonary hypertension. <i>Future Cardiol.</i> 2014;10(1):117-130.	Review/Other-Dx	N/A	To discuss important information provided by CMR that can aid the clinician, particularly relating to morphologic RV alterations and quantification of stiffness, as well as providing a novel prognostic framework.	No results stated in abstract.	4
2. Galie N, Humbert M, Vachiery JL, et al. 2015 ESC/ERS Guidelines for the diagnosis and treatment of pulmonary hypertension: The Joint Task Force for the Diagnosis and Treatment of Pulmonary Hypertension of the European Society of Cardiology (ESC) and the European Respiratory Society (ERS): Endorsed by: Association for European Paediatric and Congenital Cardiology (AEPC), International Society for Heart and Lung Transplantation (ISHLT). <i>Eur Heart J.</i> 2016;37(1):67-119.	Review/Other-Dx	N/A	To provide guidelines for the diagnosis and treatment of PH.	No results stated in abstract.	4
3. Pena E, Dennie C, Veinot J, Muniz SH. Pulmonary hypertension: how the radiologist can help. <i>Radiographics.</i> 2012;32(1):9-32.	Review/Other-Dx	N/A	To examine the radiologist's role in evaluating patients with this PH.	Radiologists play an important role in evaluating patients with this disease.	4
4. Schiebler ML, Bhalla S, Runo J, et al. Magnetic resonance and computed tomography imaging of the structural and functional changes of pulmonary arterial hypertension. <i>J Thorac Imaging.</i> 2013;28(3):178-193.	Review/Other-Dx	N/A	To describe how the known structural and functional changes associated with elevated PAH and PH can be studied with MRI and CT.	By understanding the dynamic relationship that exists between the heart and lungs in this heterogeneous group of diseases, the severity of this disease process can often be inferred. MRI is a promising noninvasive and nonionizing modality that can be used to study the many diseases that cause PAH and PH in a longitudinal fashion.	4
5. Hatano S, Strasser T. <i>Primary pulmonary hypertension: report on a WHO meeting, Geneva, 15-17 October 1973.</i> Geneva; Albany, N.Y.: World Health Organization; distributed by Q Corporation; 1975.	Review/Other-Dx	N/A	Report on primary PH.	N/A	4

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6. Rich S, Rubin LJ, Abenhail L, et al. Executive summary from the World Symposium on Primary Pulmonary Hypertension (Evian, France, September 6-10, 1998). The World Health Organization publication via the Internet. Available at: http://www.who.int/ncd/cvd/pph.html .	Review/Other-Dx	N/A	Executive summary from the World Symposium on Primary Pulmonary Hypertension.	N/A	4
7. Simonneau G, Galie N, Rubin LJ, et al. Clinical classification of pulmonary hypertension. <i>J Am Coll Cardiol</i> . 2004;43(12 Suppl S):5S-12S.	Review/Other-Dx	N/A	Review clinical classification of PH.	Evian classification is well accepted and widely used in clinical practice, especially in specialized centers. In 2003, during the Third World Symposium on Pulmonary Arterial Hypertension held in Venice, Italy, it was decided to maintain the general architecture and philosophy of the Evian classification. However, some modifications have been proposed, mainly to abandon the term “primary PH” and to replace it with “idiopathic PH”; to reclassify pulmonary venoocclusive disease and pulmonary capillary hemangiomatosis; to update risk factors and associated conditions for PAH and to propose guidelines in order to improve the classification of congenital systemic-to-pulmonary shunts.	4

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8. Simonneau G, Robbins IM, Beghetti M, et al. Updated clinical classification of pulmonary hypertension. <i>J Am Coll Cardiol.</i> 2009;54(1 Suppl):S43-54.	Review/Other-Dx	N/A	A review on the updated clinical classification of PH.	In 2003, the clinical classification of PH initially adopted was slightly modified. During the 4th World Symposium held in 2008, it was decided to maintain the general architecture and philosophy of the previous clinical classifications. The modifications adopted during this meeting principally concern Group 1, PAH. This subgroup includes patients with PAH with a family history or patients with IPAH with germline mutations. In the new classification, schistosomiasis and chronic hemolytic anemia appear as separate entities in the subgroup of PAH associated with identified diseases. Finally, it was decided to place pulmonary venoocclusive disease and pulmonary capillary hemangiomatosis in a separate group, distinct from but very close to Group 1 (now called Group 1'). Thus, Group 1 of PAH is now more homogeneous.	4
9. Simonneau G, Gatzoulis MA, Adatia I, et al. Updated clinical classification of pulmonary hypertension. <i>J Am Coll Cardiol.</i> 2013;62(25 Suppl):D34-41.	Review/Other-Dx	N/A	To update clinical classification of PH by adding some specific items related to pediatric PH in order to have a comprehensive classification common for adults and children.	No results stated in abstract.	
10. McCann C, Gopalan D, Sheares K, Screaton N. Imaging in pulmonary hypertension, part 1: clinical perspectives, classification, imaging techniques and imaging algorithm. <i>Postgrad Med J.</i> 2012;88(1039):271-279.	Review/Other-Dx	N/A	To describe the clinical signs and symptoms of PH, explain the current PH classification and illustrate various imaging techniques that are available to investigate this condition. The strengths and weaknesses of each of these techniques will also be highlighted.	No results stated in abstract.	4
11. McGoon M, Gutterman D, Steen V, et al. Screening, early detection, and diagnosis of pulmonary arterial hypertension: ACCP evidence-based clinical practice guidelines. <i>Chest.</i> 2004;126(1 Suppl):14S-34S.	Review/Other-Dx	N/A	Review evidence for screening in susceptible patient groups and the approach to diagnosing PAH when it is suspected, and provide specific recommendations for applying this evidence to clinical practice.	A high level of suspicion is of paramount importance for the diagnosis of PAH, regardless of the underlying cause. Once suspected, a methodical workup using commonly employed diagnostic interventions allows both confirmation of the presence of PAH and elucidation of its etiology. Clarification of etiology is necessary to ensure that the proper therapeutic interventions are implemented. A diagnostic algorithm that is accepted among experienced centers can guide the evaluation of PAH.	4

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12. Montani D, O'Callaghan DS, Jais X, et al. Implementing the ESC/ERS pulmonary hypertension guidelines: real-life cases from a national referral centre. <i>Eur Respir Rev.</i> 2009;18(114):272-290.	Review/Other-Dx	N/A	To describe a series of clinical cases of PH due to various etiologies that were referred to a large national PH expert referral center. In each case, the assessment and therapeutic approach undertaken is described in the context of the new European Society of Cardiology and the European Respiratory Society guidelines.	European Society of Cardiology and the European Respiratory Society guidelines are clinically relevant.	4
13. Chetty KG, Brown SE, Light RW. Identification of pulmonary hypertension in chronic obstructive pulmonary disease from routine chest radiographs. <i>Am Rev Respir Dis.</i> 1982;126(2):338-341.	Observational-Dx	34 patients	To determine the value of routine chest radiographs in the identification of PH in COPD. MPAP was measured in patients with moderate to severe COPD (FEV1, 1,010 +/- 460 ml) and was correlated with the following 3 indexes derived from the chest roentgenogram: (1) the hilar thoracic index, (2) the diameter of the descending branch of the right pulmonary artery, (3) the hilar width, and (4) the cardiothoracic ratio.	The PAP best correlated with the hilar thoracic index ($r = 0.74, P < 0.01$) and was significantly correlated with the other 3 indexes. However, the accuracy with which the PAP could be predicted was only +/- 21 mm Hg. Authors conclude that the chest radiograph is useful in screening patients with COPD for elevated PAP, but that it cannot be used to predict the PAP accurately.	4
14. Frazier AA, Burke AP. The imaging of pulmonary hypertension. <i>Semin Ultrasound CT MR.</i> 2012;33(6):535-551.	Review/Other-Dx	N/A	To discuss the value of different imaging modalities in the diagnosis PH.	Radiologic imaging provides an essential tool for the comprehensive analysis of the pulmonary vasculature, lung parenchyma, and cardiac morphology and function in PH. Chest radiography, V/Q imaging, and CT help to localize the primary site of disease as either precapillary or postcapillary, and further may provide insight into the underlying etiology of PH.	4
15. Lupi E, Dumont C, Tejada VM, Horwitz S, Galland F. A radiologic index of pulmonary arterial hypertension. <i>Chest.</i> 1975;68(1):28-31.	Observational-Dx	250 patients	A new index derived from the plain chest radiograph is provided for assessing normal and elevated PAP.	The index was significantly different in groups with and without PH and was abnormal (above 38% in 111/150) patients with cardiovascular disease and PAH. None of the cases with increased pulmonary flow from cardiac shunts but normal PAP had an abnormal index. Thus, an abnormal index suggested PAH but correlated poorly with the extent of hypertension.	4

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16. Matthay RA, Schwarz MI, Ellis JH, Jr., et al. Pulmonary artery hypertension in chronic obstructive pulmonary disease: determination by chest radiography. <i>Invest Radiol.</i> 1981;16(2):95-100.	Observational-Dx	61 men and 42 normal control subjects	To determine whether analysis of the chest radiograph can reveal the presence of pulmonary artery hypertension in COPD.	The right descending pulmonary artery was enlarged (>16 mm) in 43/46 patients (93%) with an elevated MPAP, and the left descending pulmonary artery diameter also was enlarged (>18 mm) in 43/46. Combined increased right descending pulmonary artery and increased left descending pulmonary artery diameter measurements permitted correct diagnosis in 45/46 patients (98%) with pulmonary artery hypertension, including all 26 a mild elevation of MPAP (21–30 mmHg). There was a significant correlation between PAP and both right descending pulmonary artery and left descending pulmonary artery measurements. Analysis of right descending pulmonary artery and left descending pulmonary artery diameters on the plain chest radiograph is a sensitive and accurate method of detecting the presence and severity of pulmonary artery hypertension in COPD.	4
17. Schmidt HC, Kauczor HU, Schild HH, et al. Pulmonary hypertension in patients with chronic pulmonary thromboembolism: chest radiograph and CT evaluation before and after surgery. <i>Eur Radiol.</i> 1996;6(6):817-825.	Observational-Dx	50 patients	To assess the value of morphometric data on conventional radiography and CT predicting the presence and degree of PH and to assess the reversibility after surgery.	In 14 X-ray patients and 18 CT patients, with follow-up after surgical thromboendarterectomy the reversibility of these changes was assessed. A dilated pulmonary trunk was the most common abnormality (96% each on X-ray and CT). Pulmonary arteries were dilated on X-ray in 40% (right) and 14% (left), and on CT in 92% (right) and 96% (left). The best correlation with mean arterial pressure was found measuring the pulmonary trunk on CT ($r = 0.43$, $P < 0.01$). After surgery, reversibility was most significant for the pulmonary trunk on CT ($P < 0.0001$). In patients with chronic pulmonary embolism, PH can best be predicted by assessing the diameter of the pulmonary trunk both on X-ray and CT. No close correlation is present between the extent of any parameter and the level of pulmonary pressure.	4

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18. Teichmann V, Jezek V, Herles F. Relevance of width of right descending branch of pulmonary artery as a radiological sign of pulmonary hypertension. <i>Thorax</i> . 1970;25(1):91-96.	Observational-Dx	112 healthy subjects	To elucidate the diagnostic importance of the right descending branch, the authors investigated the relation between its diameter and any PH. Catheterization data was compared with the width of the right descending branch and the authors tried to find out the difference, if any, between the radiographic appearance of PH in chronic bronchitis and in other diseases.	Considering (in men over 40 years of age) a diameter of the right descending branch of 18 mm or more as pathological, the reliability of the diagnosis of PH in chronic bronchitis was 72-2% with readable films, or 64-2% when not reliably readable chest films were included. Nevertheless, a similar comparison in patients with mitral stenosis indicates that the radiographic picture of PH differs according to the initial disease.	4
19. Woodruff WW, 3rd, Hoeck BE, Chitwood WR, Jr., Lyerly HK, Sabiston DC, Jr., Chen JT. Radiographic findings in pulmonary hypertension from unresolved embolism. <i>AJR Am J Roentgenol</i> . 1985;144(4):681-686.	Observational-Dx	22 patients	To evaluate plain chest radiographs in a series of patients with pulmonary artery hypertension and concomitant chronic pulmonary embolism seen at Duke University Medical Center between 1 968 and 1984.	In 22 patients, no normal radiographs were seen. Findings included cardiomegaly (86.4%) with right-sided enlargement (68.4%), right descending pulmonary artery enlargement (54.5%), azygos vein enlargement (27.3%), mosaic oligemia (68.2%), chronic volume loss (27.3%), atelectasis and/or effusion (22.7%), and pleural thickening (13.6%). Good correlation with specific areas of diminished vascularity was seen on chest radiographs compared with pulmonary angiograms.	3
20. Miniati M, Monti S, Airo E, et al. Accuracy of chest radiography in predicting pulmonary hypertension: a case-control study. <i>Thromb Res</i> . 2014;133(3):345-351.	Observational-Dx	108 patients	To assess the accuracy of chest radiography in predicting PH.	82 patients had PH confirmed at RHC. Weighted sensitivity of chest radiography was 96.9% (95% CI, 94.9% to 98.2%), and weighted specificity 99.8% (95% CI, 99.6% to 99.9%). By considering the 165 patients who underwent RHC, weighted sensitivity of chest radiography was unchanged, and weighted specificity decreased to 99.1%. None of the patients with PH were misclassified as having left heart failure, and vice versa.	2
21. Algeo S, Morrison D, Ovitt T, Goldman S. Noninvasive detection of pulmonary hypertension. <i>Clin Cardiol</i> . 1984;7(3):148-156.	Observational-Dx	79 consecutive patients and 50 consecutive control patients	Plain chest x-rays, twelve-lead electrocardiograms, and M-mode echocardiograms were analyzed in control patients and patients with PH documented at cardiac catheterization in order to determine the relative values of these noninvasive techniques as screening tests in detecting PH.	The sensitivity and specificity of selected findings previously described as being associated with PH were calculated for each test. All test results were found to have sensitivities too low to function as satisfactory noninvasive screening techniques. Most findings were highly specific for PH when present. Patients with pulmonary vascular disease were detected more frequently than those with PH due to pulmonary venous congestion.	3

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22. Barbosa EJ, Jr., Gupta NK, Torigian DA, Gefter WB. Current role of imaging in the diagnosis and management of pulmonary hypertension. <i>AJR Am J Roentgenol.</i> 2012;198(6):1320-1331.	Review/Other-Dx	N/A	To describe classification schemes and imaging findings in the diagnosis and management of PH.	PH is a complex pathophysiologic condition in which several clinical entities increase pressure in the pulmonary circulation, progressively impairing cardiopulmonary function and, if untreated, causing RV failure. Current classification schemes emphasize the necessity of an early, accurate etiologic diagnosis for a tailored therapeutic approach. Imaging plays an increasingly important role in the diagnosis and management of suspected PH.	4
23. Chang CH. The normal roentgenographic measurement of the right descending pulmonary artery in 1,085 cases. <i>Am J Roentgenol Radium Ther Nucl Med.</i> 1962;87:929-935.	Review/Other-Dx	1,085 patients	To report normal roentgenographic measurement of the right descending pulmonary artery.	No results stated in abstract.	4
24. Rich S, Dantzker DR, Ayres SM, et al. Primary pulmonary hypertension. A national prospective study. <i>Ann Intern Med.</i> 1987;107(2):216-223.	Observational-Dx	187 patients	To evaluate diagnosis of patients with primary PH. A national registry was used to collect data from 32 centers on patients diagnosed by uniform criteria as having primary PH.	The most frequent presenting symptoms included dyspnea (60%), fatigue (19%), and syncope (or near syncope) (13%). Raynaud phenomenon was present in 10% (95% of whom were female) and a positive antinuclear antibody test, in 29% (69% female). Pulmonary function studies showed mild restriction (forced vital capacity, 82% of predicted) with a reduced diffusing capacity for carbon monoxide, and hypoxemia with hypocapnia. The mean (+/- SD) right atrial pressure was 9.7 +/- 6 mm Hg; MPAP, 60 +/- 18 mm Hg; cardiac index, 2.3 +/- 0.9 L/min X m2; and pulmonary vascular resistance index, 26 +/- 14 mm Hg/L/min X m2 for the group. Although no deaths or sustained morbid events occurred during the diagnostic evaluation of the patients, the typically long interval from initial symptoms to diagnosis emphasizes the need to develop strategies to make the diagnosis earlier.	3

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25. Taleb M, Khuder S, Tinkel J, Khouri SJ. The diagnostic accuracy of Doppler echocardiography in assessment of pulmonary artery systolic pressure: a meta-analysis. <i>Echocardiography</i> . 2013;30(3):258-265.	Meta-analysis	9 articles	To estimate the accuracy, sensitivity, and specificity of DE in the assessment of pulmonary artery systolic pressure.	The correlation between pulmonary artery systolic pressure estimated by Doppler echocardiography and RHC ranged from ($r = 0.65, P < 0.001$) to ($r = 0.97, P < 0.001$). The pooled sensitivity, specificity, and accuracy of Doppler echocardiography for the diagnosis of PH were 88% (95% CI, 84%–92%), 56% (95% CI, 46%–66%), and 63% (95% CI, 53%–73%), respectively.	M
26. Janda S, Shahidi N, Gin K, Swiston J. Diagnostic accuracy of echocardiography for pulmonary hypertension: a systematic review and meta-analysis. <i>Heart</i> . 2011;97(8):612-622.	Review/Other-Dx	29 studies	To perform a systematic review and quantitative meta-analysis to determine the correlation of pulmonary pressures obtained by echocardiography vs RHC and to determine the diagnostic accuracy of echocardiography for PH.	The summary correlation coefficient between systolic pulmonary arterial pressure estimated from echocardiography vs measured by RHC was 0.70 (95% CI, 0.67 to 0.73; $n=27$). The summary sensitivity and specificity for echocardiography for diagnosing PH was 83% (95% CI, 73 to 90) and 72% (95% CI, 53 to 85; $n=12$), respectively. The summary diagnostic OR was 13 (95% CI, 5 to 31). Echocardiography is a useful and noninvasive modality for initial measurement of pulmonary pressures but due to limitations, RHC should be used for diagnosing and monitoring PH.	4
27. Lang RM, Badano LP, Mor-Avi V, et al. Recommendations for cardiac chamber quantification by echocardiography in adults: an update from the American Society of Echocardiography and the European Association of Cardiovascular Imaging. <i>Eur Heart J Cardiovasc Imaging</i> . 2015;16(3):233-270.	Review/Other-Dx	N/A	To update recommendations to the previously published guidelines for cardiac chamber quantification.	This document provides updated normal values for all 4 cardiac chambers, including 3D echocardiography and myocardial deformation, when possible, on the basis of considerably larger numbers of normal subjects, compiled from multiple databases.	4
28. Lau EM, Manes A, Celermajer DS, Galie N. Early detection of pulmonary vascular disease in pulmonary arterial hypertension: time to move forward. <i>Eur Heart J</i> . 2011;32(20):2489-2498.	Review/Other-Dx	N/A	To discuss the possible strategies devoted to the early detection of pulmonary vascular disease in PAH patients.	No results stated in abstract.	4

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29. Mocerri P, Baudouy D, Chiche O, et al. Imaging in pulmonary hypertension: Focus on the role of echocardiography. <i>Arch Cardiovasc Dis.</i> 2014;107(4):261-271.	Review/Other-Dx	N/A	To describe the cardiac alterations related to PH, and to review and discuss the extent to which echocardiography plays a key role in early diagnosis and prognosis in PH.	Echocardiography is a widely available, cost-effective, safe and reliable examination, which provides us with major diagnostic and prognostic information. Comparison of serial assessments allows monitoring of the efficacy of advanced therapies. More recent ultrasound techniques, such as 3D echocardiography and speckle-tracking, are promising; they may provide additional data regarding RV and pulmonary artery mechanics in PH and may allow the preclinical detection of high-risk patients.	4
30. Rudski LG, Lai WW, Afilalo J, et al. Guidelines for the echocardiographic assessment of the right heart in adults: a report from the American Society of Echocardiography endorsed by the European Association of Echocardiography, a registered branch of the European Society of Cardiology, and the Canadian Society of Echocardiography. <i>J Am Soc Echocardiogr.</i> 2010;23(7):685-713; quiz 786-688.	Review/Other-Dx	N/A	To establish a standard uniform method for obtaining right heart images for assessing RV size and function and as an impetus for the development of databases to refine the normal values.	This guidelines document provides clinicians basic views to assess the right ventricle and right atrium, the various parameters to assess RV systolic and diastolic function, and the normal reference values from pooled data. This will enable echocardiographers to distinguish an abnormal right ventricle from a normal one. It is hoped that this document will lead to further work in establishing normal ranges in larger populations and that the application of the values included will enhance the value of echocardiography in recognizing RV dysfunction in clinical practice, in improving disease detection and in patient follow-up.	4

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31. Aduen JF, Castello R, Daniels JT, et al. Accuracy and precision of three echocardiographic methods for estimating mean pulmonary artery pressure. <i>Chest</i> . 2011;139(2):347-352.	Observational-Dx	117 patients	To compare the accuracy and precision of 3 echocardiographic methods for estimating MPAP.	The mean +/- SD of the differences between invasive MPAP and the 3 echocardiographic methods were -1.6 +/- 7.7 mm Hg for the mean gradient method, -3.7 +/- 7.4 mm Hg for the Chemla formula, and -3.2 +/- 7.6 mm Hg for the Syyed formula. Median absolute differences were 5.5 mm Hg (mean gradient), 5.7 mm Hg (Chemla; $P=.45$ vs mean gradient), and 6.0 mm Hg (Syyed; $P=.23$ vs mean gradient). Accuracy (calculated MPAP within 10 mm Hg of RHC-measured MPAP) was 81% (mean gradient), 77% (Chemla), and 76% (Syyed). Echocardiographic estimation of MPAP by the mean gradient method had similar accuracy and precision compared with the Chemla and Syyed methods. The acceptable accuracy of these methods suggests that they are equally suitable for clinical use.	2
32. Di Bello V, Conte L, Delle Donne MG, et al. Advantages of real time three-dimensional echocardiography in the assessment of right ventricular volumes and function in patients with pulmonary hypertension compared with conventional two-dimensional echocardiography. <i>Echocardiography</i> . 2013;30(7):820-828.	Observational-Dx	54 patients	To point out the value of real time 3D echocardiography and tissue Doppler imaging in the evaluation of patients affected by PH, compared with conventional 2D echocardiography.	In this study, RV fractional area change, and tricuspid annular plane systolic excursion showed marked alterations in patients with PH compared to the control group (C): (RV fractional area change: [PH] 0.29 +/- 0.07 vs [C] 0.49 +/- 0.05%, $P<0.0001$; tricuspid annular plane systolic excursion: [PH] 15.3 +/- 3.2 vs [C] 21.1 +/- 2.6 mm, $P>0.0001$). The 3D RV EDV was significantly higher in PH than in C (PH) (138.7 +/- 25.3 vs [C] 82.8 +/- 12.5 mL, $P<0.0001$) as well as 3D RV ESV (PH) (97.6 +/- 21.5 vs [C] 39.3 +/- 9.5 mL, $P<0.0001$). The 3D RV ejection fraction was significantly lower in the PH group than in healthy subjects (31.8 +/- 6.8 vs [C] 52.5 +/- 4.7%, $P<0.0001$).	2

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33. Lancellotti P, Budts W, De Wolf D, et al. Practical recommendations on the use of echocardiography to assess pulmonary arterial hypertension--a Belgian expert consensus endorsed by the Working Group on Non-Invasive Cardiac Imaging. <i>Acta Cardiol.</i> 2013;68(1):59-69.	Review/Other-Dx	N/A	To provide practical recommendations for the use of echocardiography in the evaluation of PH and its consequences on the RV.	Due to its widespread availability, echocardiography is used as the first-line imaging modality to detect pulmonary PH and assess RV function in daily routine. As such, echocardiography is the key examination to detect the presence of PH, to provide valuable prognostic information and to give an orientation to therapeutic strategies. In addition to detection and screening, echocardiography also provides clues for the differential diagnosis of PH.	4
34. Hoepfer MM, Barbera JA, Channick RN, et al. Diagnosis, assessment, and treatment of non-pulmonary arterial hypertension pulmonary hypertension. <i>J Am Coll Cardiol.</i> 2009;54(1 Suppl):S85-96.	Review/Other-Dx	N/A	Medical literature regarding the role of PH in chronic obstructive lung disease, interstitial lung disease, chronic thromboembolic PH, and left heart disease was reviewed, and recommendations regarding diagnosis and treatment of PH in these conditions are provided.	Given the lack of robust clinical trials addressing PH in any of these conditions, it is important to conduct further studies to establish the role of medical therapy in non-PAH PH.	4
35. Giannouli E, Maycher B. Imaging techniques in chronic thromboembolic pulmonary hypertension. <i>Curr Opin Pulm Med.</i> 2013;19(5):562-574.	Review/Other-Dx	N/A	To examine the findings of various imaging techniques in CTEPH and their contribution in the diagnostic and therapeutic evaluation of the disease.	CTEPH is a potentially curable cause of PH via pulmonary thromboendarterectomy. It is probably more common than previously thought, and can be misdiagnosed as patients present with nonspecific symptoms. Radiologic imaging plays a central role in early detection and accurate diagnosis of CTEPH. Furthermore, cross-sectional imaging can correctly assess the technical feasibility of pulmonary thromboendarterectomy and assist in postoperative follow-up. V/Q scan can be useful in screening, as if it is normal, CTEPH can be ruled out. CT and MRA represent the future for diagnosis and management of CTEPH. Newer noninvasive technologies (MR, dual energy CTA) for assessment of lung perfusion and pulmonary hemodynamics may help optimizing selection of operative candidates.	4

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36. Tunariu N, Gibbs SJ, Win Z, et al. Ventilation-perfusion scintigraphy is more sensitive than multidetector CTPA in detecting chronic thromboembolic pulmonary disease as a treatable cause of pulmonary hypertension. <i>J Nucl Med.</i> 2007;48(5):680-684.	Observational-Dx	227 patients	Retrospective study to compare the value of V/Q scintigraphy with CTPA in detecting chronic thromboembolic pulmonary disease.	78 patients (group A) had a final diagnosis of CTEPH and 149 (group B) had non-CTEPH etiology. Among group A, V/Q scintigraphy was reported as high probability in 75 patients, intermediate probability in 1 patient, and low probability in 2 patients. CTPA was positive in 40 patients and negative in 38 patients. Among group B, V/Q scintigraphy was reported as low probability in 134, intermediate probability in 7, and high probability in 8 patients. CTPA was negative in 148 patients and false-positive in 1 patient. Statistical analysis showed V/Q scintigraphy to have a sensitivity of 96%–97.4% and a specificity of 90%–95%. CTPA showed a sensitivity of 51% and a specificity of 99%. Results demonstrate that V/Q scintigraphy has a higher sensitivity than CTPA in detecting CTEPH as a potential curable cause of PH.	4
37. Fedullo PF, Auger WR, Kerr KM, Rubin LJ. Chronic thromboembolic pulmonary hypertension. <i>N Engl J Med.</i> 2001;345(20):1465-1472.	Review/Other-Dx	N/A	A review on CTEPH.	No results stated.	4
38. Ley S, Ley-Zaporozhan J, Pitton MB, et al. Diagnostic performance of state-of-the-art imaging techniques for morphological assessment of vascular abnormalities in patients with chronic thromboembolic pulmonary hypertension (CTEPH). <i>Eur Radiol.</i> 2012;22(3):607-616.	Observational-Dx	24 patients	To determine the most comprehensive imaging technique for the assessment of pulmonary arteries in patients with CTEPH.	Based on image quality, there was no non-diagnostic examination by either imaging technique. DSA did not sufficiently display 1 main, 3 lobar and 4 segmental arteries. The pulmonary trunk was not assessable by DSA. 1 patient showed thrombotic material at this level only by multidetector-CTA and MRA. Sensitivity and specificity of multidetector-CTA regarding CTEPH-related changes at the main/lobar and at the segmental levels were 100%/100% and 100%/99%, of contrast-enhanced MRA 83.1%/98.6% and 87.7%/98.1%, and of DSA 65.7%/100% and 75.8%/100%, respectively. Echocardiography-gated multidetector-CTA proved the most adequate technique for assessment of the pulmonary arteries in the diagnostic workup of CTEPH patients.	3

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39. Rajaram S, Swift AJ, Telfer A, et al. 3D contrast-enhanced lung perfusion MRI is an effective screening tool for chronic thromboembolic pulmonary hypertension: results from the ASPIRE Registry. <i>Thorax</i> . 2013;68(7):677-678.	Observational-Dx	132 patients	To evaluate the diagnostic utility of lung perfusion MRI.	Of 132 patients, 78 were diagnosed as having CTEPH. Lung perfusion MRI correctly identified 76 patients as having CTEPH with an overall sensitivity of 97%, specificity 92%, PPV 95% and NPV 96% compared with perfusion scintigraphy (sensitivity 96%, specificity 90%) and CTPA (sensitivity 94%, specificity 98%). No cases of surgically accessible CTEPH were missed with either modality.	2
40. Castaner E, Gallardo X, Rimola J, et al. Congenital and acquired pulmonary artery anomalies in the adult: radiologic overview. <i>Radiographics</i> . 2006;26(2):349-371.	Review/Other-Dx	N/A	To review the features of various congenital and acquired anomalies of the pulmonary arteries, with an emphasis on their CT appearance and possible effects on the heart.	In the appropriate clinical setting (eg, infection or vasculitis), an awareness of the radiologic manifestations of possible pulmonary arterial complications may enable an early diagnosis.	4
41. Alhamad EH, Al-Boukai AA, Al-Kassimi FA, et al. Prediction of pulmonary hypertension in patients with or without interstitial lung disease: reliability of CT findings. <i>Radiology</i> . 2011;260(3):875-883.	Observational-Dx	134 patients	To study the reliability of pulmonary vascular measurements based on CT in the prediction of PH in patients with advanced interstitial lung disease compared with those without interstitial lung disease.	Main pulmonary artery diameter was significantly greater in patients with PH than in those without PH in both groups (group A, $P=.008$; group B, $P=.02$). A pulmonary artery diameter >25 mm in patients with interstitial lung disease was predictive of PH, with a sensitivity of 86.4% (32/37), a specificity of 41.2% (26/63), a PPV of 46.3% (32/69), and a NPV of 83.8% (26/31). In patients without interstitial lung disease, a pulmonary artery diameter >31.6 mm and a left pulmonary artery diameter >21.4 mm were predictive of PH (sensitivity, 47.3% [9/19]; specificity, 93.3% [14/15]; PPV, 90.0% [9/10]; and NPV, 58.3% [14/24]).	2

**Suspected Pulmonary Hypertension
EVIDENCE TABLE**

Reference	Study Type	Patients/ Events	Study Objective (Purpose of Study)	Study Results	Study Quality
42. Tan RT, Kuzo R, Goodman LR, Siegel R, Haasler GB, Presberg KW. Utility of CT scan evaluation for predicting pulmonary hypertension in patients with parenchymal lung disease. Medical College of Wisconsin Lung Transplant Group. <i>Chest</i> . 1998;113(5):1250-1256.	Observational-Dx	45 patients	To determine the utility of CT-determined main pulmonary artery diameter for predicting PH in patients with parenchymal lung disease.	CT-determined main pulmonary artery diameter was 35+/-6 mm in patients with PH and 27+/-2 mm in control subjects. main pulmonary artery diameter \geq 29 mm had a sensitivity of 87%, specificity of 89%, PPV of 0.97, and positive likelihood ratio of 7.91 for predicting PH; in the subgroup of patients with parenchymal lung disease (n=28, PH and control subjects), main pulmonary artery diameter \geq 29 mm had a sensitivity of 84%, specificity of 75%, PPV of 0.95, and positive likelihood ratio of 3.36 for predicting PH. The most specific findings for the presence of PH were both main pulmonary artery diameter \geq 29 mm and segmental artery-to-bronchus ratio >1:1 in 3 or 4 lobes (specificity, 100%). There was no linear correlation between the degree of PH and main pulmonary artery diameter ($r=0.124$). CT-determined main pulmonary artery diameter has excellent diagnostic value for detection of PH in patients with advanced lung disease. Therefore, standard chest CT scans can be used to screen for PH as a cause of exertional limitation in patients with parenchymal lung disease. Because CT is commonly used to evaluate parenchymal lung disease, this information is readily available.	4
43. Zisman DA, Karlamangla AS, Ross DJ, et al. High-resolution chest CT findings do not predict the presence of pulmonary hypertension in advanced idiopathic pulmonary fibrosis. <i>Chest</i> . 2007;132(3):773-779.	Observational-Dx	65 patients	To determine whether the CT-determined extent and severity of pulmonary fibrosis and diameter of the main pulmonary artery could be used to diagnose PH in advanced idiopathic pulmonary fibrosis patients.	Chest CT-determined fibrosis score, ground-glass opacity score, honeycombing score, total profusion score, diameter of the main pulmonary artery, and the ratio of the pulmonary artery to aorta diameter did not differ between those with and without PH. There was no significant correlation between MPAP and any of the chest CT-determined measures.	4

**Suspected Pulmonary Hypertension
EVIDENCE TABLE**

Reference	Study Type	Patients/ Events	Study Objective (Purpose of Study)	Study Results	Study Quality
44. Baque-Juston MC, Wells AU, Hansell DM. Pericardial thickening or effusion in patients with pulmonary artery hypertension: a CT study. <i>AJR Am J Roentgenol.</i> 1999;172(2):361-364.	Review/Other-Dx	45 patients	To determine the prevalence of pericardial thickening or effusion revealed by CT in patients with pulmonary artery hypertension.	The prevalence of an increased pericardial score and increased maximum pericardial thickening was higher in group 3 than in group 1 or group 2 ($P=.02$ and $<.001$, respectively). Anterior pericardial recess thickening was markedly increased in group 3 ($P<.0001$). For all patients, significant correlations (Spearman's rank correlation coefficient = $.44-.56$, $P<.005-.0001$) were found between MPAP and all pericardial measures. On CT, pericardial thickening or effusion is a frequent finding in patients with severe PH.	4
45. Coulden R. State-of-the-art imaging techniques in chronic thromboembolic pulmonary hypertension. <i>Proc Am Thorac Soc.</i> 2006;3(7):577-583.	Review/Other-Dx	N/A	To describe a rational approach to imaging in PH, how it is used in the diagnosis of CTEPH, and how it is used to assess operability when CTEPH is present.	No results stated in abstract.	4
46. Fischer A, Misumi S, Curran-Everett D, et al. Pericardial abnormalities predict the presence of echocardiographically defined pulmonary arterial hypertension in systemic sclerosis-related interstitial lung disease. <i>Chest.</i> 2007;131(4):988-992.	Observational-Dx	41 patients	To determine the prevalence and significance of pericardial abnormalities in systemic sclerosis-related interstitial lung disease.	59% had an abnormal pericardium, 49% had a pericardial effusion, 56% had an abnormal anterior pericardial recess, and 49% had an abnormal total pericardial score. An abnormal pericardium was more common in men than women. Subjects with and without pericardial abnormalities were otherwise similar with respect to age, systemic sclerosis classification, autoantibodies, interstitial lung disease radiographic pattern, and presence of esophageal dilation. Both groups had similar median percentage of predicted total lung capacity, percentage of predicted forced vital capacity, percentage of predicted FEV(1), and percentage of predicted diffusion capacity of the lung for carbon monoxide. Subjects with pericardial abnormalities were more likely to have coexistent PAH (35% vs 75%; $P=0.02$) and a higher median RV systolic pressure (31 mm Hg vs 44 mm Hg; $P=0.03$). Multiple logistic regression revealed that total pericardial score was the best individual predictor of the presence of transthoracic echocardiography-defined PAH.	3

**Suspected Pulmonary Hypertension
EVIDENCE TABLE**

Reference	Study Type	Patients/ Events	Study Objective (Purpose of Study)	Study Results	Study Quality
47. Eksinar S, Gedevanishvili A, Koroglu M, et al. Extrinsic compression of the left main coronary artery in pulmonary hypertension. <i>JBR-BTR</i> . 2005;88(4):190-192.	Review/Other-Dx	1 patient	Case report to demonstrate significant compression of the left main coronary artery on coronary angiography and thorax CT examinations.	Coronary angiography is the modality of choice for diagnosis.	4
48. Chan AL, Juarez MM, Shelton DK, et al. Novel computed tomographic chest metrics to detect pulmonary hypertension. <i>BMC Med Imaging</i> . 2011;11:7.	Observational-Dx	101 patients	To determine whether PH can be assessed by measuring a set of CT chest-based metrics that has a relationship to RHC.	Multiple regression analyses controlling for age, sex, ascending aortic diameter, body surface area, thoracic diameter and pulmonary wedge pressure showed that a main pulmonary artery diameter ≥ 29 mm (OR=4.8), right descending pulmonary artery diameter ≥ 19 mm (OR=7.0), true right descending pulmonary artery diameter ≥ 16 mm (OR=4.1), true left descending pulmonary artery diameter ≥ 21 mm (OR=15.5), RV free wall ≥ 6 mm (OR=30.5), RV wall/LV wall ratio ≥ 0.32 (OR=8.8), RV/LV lumen ratio ≥ 1.28 (OR=28.8), main pulmonary artery/ascending aorta ratio ≥ 0.84 (OR=6.0) and main pulmonary artery/descending aorta ratio ≥ 1.29 (OR=5.7) were significant predictors of PH in this population of hospitalized patients.	3
49. Piazza G, Goldhaber SZ. Chronic thromboembolic pulmonary hypertension. <i>N Engl J Med</i> . 2011;364(4):351-360.	Review/Other-Dx	N/A	A review on thromboembolic PH.	No results stated.	4
50. He J, Fang W, Lv B, et al. Diagnosis of chronic thromboembolic pulmonary hypertension: comparison of ventilation/perfusion scanning and multidetector computed tomography pulmonary angiography with pulmonary angiography. <i>Nucl Med Commun</i> . 2012;33(5):459-463.	Observational-Dx	51 patients	To compare the diagnostic efficacy of pulmonary V/Q scanning and CTPA using pulmonary angiography as the golden standard.	51 patients (44.7%) had a final diagnosis of CTEPH. V/Q scan showed high probability, intermediate probability, and low probability/normal scan in 52, 3, and 59 patients, respectively. CTPA revealed 50 patients with CTEPH and 64 patients without CTEPH. The sensitivity, specificity, and accuracy of the V/Q scan were 100%, 93.7%, and 96.5%, respectively, with threshold 1, and 96.1%, 95.2%, and 95.6%, respectively, with threshold 2; similarly, the sensitivity, specificity, and accuracy of CTPA were 92.2%, 95.2%, and 93.9%, respectively.	2

**Suspected Pulmonary Hypertension
EVIDENCE TABLE**

Reference	Study Type	Patients/ Events	Study Objective (Purpose of Study)	Study Results	Study Quality
51. Arakawa H, Stern EJ, Nakamoto T, Fujioka M, Kaneko N, Harasawa H. Chronic pulmonary thromboembolism. Air trapping on computed tomography and correlation with pulmonary function tests. <i>J Comput Assist Tomogr.</i> 2003;27(5):735-742.	Observational-Dx	9 patients and 9 controls	To evaluate lung attenuation on inspiratory/expiratory CT and spiral CTA from patients with chronic pulmonary embolism and correlate the CT findings with pulmonary function test results.	Lower attenuation with mosaic perfusion and air trapping were identified in 6 and 9 patients, respectively (mean scores, 8.1 and 11.3, respectively). Air trapping was identified in 19 (42.2%) of 45 segments with lower attenuation on inspiratory images, but was also noted in 31 segments with normal inspiratory attenuation. Air trapping was associated with the presence of proximal arterial stenosis ($P<0.01$), and the area showed less contrast enhancement than the adjacent lung ($P<0.05$). Extent of air trapping correlated inversely with pulmonary function test parameters of peripheral airway obstruction such as maximum mid-expiratory flow rate ($r=-0.86$, $P=0.003$). On the other hand, extent of mosaic perfusion did not correlate with pulmonary function test. Air trapping is commonly seen in chronic embolism and is found in areas of relative hypoperfusion. The extent of air trapping correlates with parameters of peripheral airway obstruction.	3
52. Castaner E, Gallardo X, Ballesteros E, et al. CT diagnosis of chronic pulmonary thromboembolism. <i>Radiographics.</i> 2009;29(1):31-50; discussion 50-33.	Review/Other-Dx	N/A	To review the risk factors, clinical characteristics, and pathogenesis of chronic pulmonary embolism; and describe the optimal technique for CTA and the CT diagnostic criteria for chronic pulmonary thromboembolism. Finally, to briefly discuss the differential diagnoses, diagnosis, and treatment of this entity.	No results stated in abstract.	4

**Suspected Pulmonary Hypertension
EVIDENCE TABLE**

Reference	Study Type	Patients/ Events	Study Objective (Purpose of Study)	Study Results	Study Quality
53. Han D, Lee KS, Franquet T, et al. Thrombotic and nonthrombotic pulmonary arterial embolism: spectrum of imaging findings. <i>Radiographics</i> . 2003;23(6):1521-1539.	Review/Other-Dx	N/A	To discuss and illustrate the imaging findings in acute and chronic pulmonary thromboembolism and in nonthrombotic pulmonary embolism and correlate these findings with clinical and pathologic findings.	Along with clinical diagnosis and laboratory examinations such as D-dimer tests, imaging plays a key role in the diagnosis of diverse forms of pulmonary embolism. Multi-detector row helical CT is a highly comprehensive and noninvasive method for evaluating patients with suspected pulmonary thromboembolism. Various biologic and nonbiologic agents can cause nonthrombotic pulmonary embolism. Familiarity with the specific imaging features of pulmonary embolism should facilitate prompt identification of the underlying abnormalities.	4
54. King MA, Ysrael M, Bergin CJ. Chronic thromboembolic pulmonary hypertension: CT findings. <i>AJR Am J Roentgenol</i> . 1998;170(4):955-960.	Review/Other-Dx	N/A	To show the CT findings in patients with CTEPH.	Although the clinical diagnosis of CTEPH can be difficult because of the nonspecificity of signs amid symptoms, helical CT provides the radiologist with an opportunity to diagnose this rare but treatable cause of pulmonary artery hypertension.	4
55. Reichelt A, Hoeper MM, Galanski M, Keberle M. Chronic thromboembolic pulmonary hypertension: evaluation with 64-detector row CT versus digital subtraction angiography. <i>Eur J Radiol</i> . 2009;71(1):49-54.	Observational-Dx	27 patients	To evaluate the role of 64-row CT in the diagnostic workup of patients with CTEPH using DSA as the method of diagnostic reference.	Sensitivity and specificity of CT regarding CTEPH-related pathological changes in general were 98.3% and 94.8% at main/lobar level and 94.1% and 92.9% at segmental level, respectively. Sensitivity and specificity of CT regarding the different pathological criteria of CTEPH (complete obstruction, intimal irregularities, bands and webs, indirect signs) were 88.9-100% and 96.1-100% at main/lobar level and 84.3-90.5% and 92-98.7% at segmental level, respectively. Results show that CT is an accurate and reliable noninvasive alternative to conventional DSA in the diagnostic workup in patients with CTEPH.	3

**Suspected Pulmonary Hypertension
EVIDENCE TABLE**

Reference	Study Type	Patients/ Events	Study Objective (Purpose of Study)	Study Results	Study Quality
56. Remy-Jardin M, Duhamel A, Deken V, Bouaziz N, Dumont P, Remy J. Systemic collateral supply in patients with chronic thromboembolic and primary pulmonary hypertension: assessment with multi-detector row helical CT angiography. <i>Radiology</i> . 2005;235(1):274-281.	Observational-Dx	36 patients	To compare retrospectively the frequency of systemic collateral supply in patients who have CTEPH with the frequency of systemic collateral supply in patients who have primary PH by using multi-detector row helical CTA.	The degree of PH was comparable in groups 1 and 2. Abnormally enlarged systemic arteries were identified in 16 (73%) of 22 patients from group 1 and in 2 (14%) of 14 patients from group 2 ($P=.002$). The systemic collateral supply in group 1 comprised enlargement of both bronchial and nonbronchial systemic arteries in 9 (56%) of the 16 patients; the remaining 7 patients had an exclusive enlargement of bronchial systemic arteries ($n = 6$, 38%) or nonbronchial ($n = 1$, 6%) systemic arteries. A total of 31 enlarged nonbronchial systemic arteries were depicted, including 13 inferior phrenic arteries, 10 intercostal arteries, 7 internal mammary arteries, and 1 lateral thoracic artery. The mean \pm SD of abnormal nonbronchial systemic arteries per patient was 1.4 ± 1.9 . No relationship was found between the mean number of abnormally enlarged nonbronchial systemic arteries and the CT angiographic features of chronic pulmonary embolism. The results demonstrate the higher frequency of abnormally enlarged bronchial and nonbronchial systemic arteries in patients who have CTEPH compared with patients who have primary PH; this finding could help distinguish these 2 entities on CT angiograms.	4
57. Roberts HC, Kauczor HU, Schweden F, Thelen M. Spiral CT of pulmonary hypertension and chronic thromboembolism. <i>J Thorac Imaging</i> . 1997;12(2):118-127.	Review/Other-Dx	N/A	To describe the utility of spiral CT in PH and chronic thromboembolism.	No results stated in abstract.	4

**Suspected Pulmonary Hypertension
EVIDENCE TABLE**

Reference	Study Type	Patients/ Events	Study Objective (Purpose of Study)	Study Results	Study Quality
58. Sherrick AD, Swensen SJ, Hartman TE. Mosaic pattern of lung attenuation on CT scans: frequency among patients with pulmonary artery hypertension of different causes. <i>AJR Am J Roentgenol.</i> 1997;169(1):79-82.	Review/Other-Dx	64 patients	To determine the frequency with which a mosaic pattern of lung attenuation is seen on chest CT scans in patients with various causes of PAH.	Peak PAP of the patients in the study averaged 74 mm Hg (range, 36-194 mm Hg). 21 patients had PAH due to lung disease: 17 patients, due to cardiac disease; and 23 patients, due to vascular disease. 3 other patients had PAH due to miscellaneous causes. Of the 23 patients with PAH due to vascular disease, 17 patients (74%) had a mosaic pattern of lung attenuation. Of the 21 patients with PAH due to lung disease, 1 patient (5%) had a mosaic pattern of lung attenuation. Among the 17 patients with PAH due to cardiac disease, 2 patients (12%) had a mosaic pattern of lung attenuation. A mosaic pattern of lung attenuation was seen significantly more often in patients with PAH due to vascular disease than in patients with PAH due to cardiac or lung disease. A mosaic pattern of lung attenuation can be seen on CT scans in patients with PAH due to vascular disease, cardiac disease, or lung disease. However, the mosaic pattern is seen significantly more often in patients with PAH due to vascular disease than in patients with PAH due to cardiac or lung disease.	4
59. Remy-Jardin M, Remy J, Louvegny S, Artaud D, Deschildre F, Duhamel A. Airway changes in chronic pulmonary embolism: CT findings in 33 patients. <i>Radiology.</i> 1997;203(2):355-360.	Observational-Dx	33 patients (group 1) and 19 controls (group 2)	To evaluate airway changes in chronic pulmonary embolism with CT.	In group 1, cylindrical bronchial dilatation was found in 21 (64%) patients vs 2 (11%) patients in group 2 ($P < .001$) at the level of segmental and/or subsegmental bronchi and in the absence of obstructive syndrome. Bronchial wall thickening was identified in 4 (12%) patients in group 1 and in 2 (11%) patients in group 2 ($P = .6$). In group 1, concordance was found between the location of bronchial dilatation and that of completely obstructed and retracted pulmonary arteries ($\kappa = 0.70$), with a lower lobe predominant for bronchial dilatation. Follow-up CT scans demonstrated no changes in airway caliber over time. Chronic pulmonary embolism may lead to ipsilateral proximal bronchial dilatation	4

**Suspected Pulmonary Hypertension
EVIDENCE TABLE**

Reference	Study Type	Patients/ Events	Study Objective (Purpose of Study)	Study Results	Study Quality
60. Dournes G, Verdier D, Montaudon M, et al. Dual-energy CT perfusion and angiography in chronic thromboembolic pulmonary hypertension: diagnostic accuracy and concordance with radionuclide scintigraphy. <i>Eur Radiol.</i> 2014;24(1):42-51.	Observational-Dx	40 patients	To evaluate the diagnostic accuracy of dual-energy CT perfusion and CTA vs V/Q scintigraphy in CTEPH, and to assess the per-segment concordance rate of dual-energy CT and scintigraphy.	14 patients were diagnosed with CTEPH and 26 with other etiologies. Dual-energy CT perfusion and CTA correctly identified all CTEPH patients with sensitivity/specificity values of 1/0.92 and 1/0.93, respectively. At a segmental level, dual-energy CT perfusion showed moderate agreement (kappa = 0.44) with scintigraphy. Agreement between CTA and scintigraphy ranged from fair (kappa = 0.31) to slight (kappa = 0.09) depending on whether completely or partially occlusive patterns were considered, respectively.	2
61. Hoey ET, Mirsadraee S, Pepke-Zaba J, Jenkins DP, Gopalan D, Screaton NJ. Dual-energy CT angiography for assessment of regional pulmonary perfusion in patients with chronic thromboembolic pulmonary hypertension: initial experience. <i>AJR Am J Roentgenol.</i> 2011;196(3):524-532.	Observational-Dx	20 patients	To assess the utility of dual-energy pulmonary CTA for noninvasive assessment of regional pulmonary perfusion in patients with CTEPH.	A strong correlation existed between dual-energy CT-derived perfusion and mosaic attenuation pattern when both lobar ($r > 0.6$; $n = 20$; $P < 0.006$) and whole-lung scores were assessed ($r = 0.77$; $n = 20$; $P < 0.001$). There was no statistically significant correlation between dual-energy CT perfusion and vascular obstructive index, MPAP, or pulmonary vascular resistance ($P > 0.08$). Of 42 completely occluded lobes, 27 (64%) had demonstrable residual perfusion (mismatching), suggesting that blood supply was maintained via systemic collaterals.	2
62. Nakazawa T, Watanabe Y, Hori Y, et al. Lung perfused blood volume images with dual-energy computed tomography for chronic thromboembolic pulmonary hypertension: correlation to scintigraphy with single-photon emission computed tomography. <i>J Comput Assist Tomogr.</i> 2011;35(5):590-595.	Observational-Dx	51 patients	To evaluate the feasibility and diagnostic utility of lung perfused blood volume images generated by dual-energy CT, as compared with pulmonary perfusion scintigraphy, for patients with CTEPH.	All examinations were acquired without complications, and the contrast enhancement of the pulmonary artery was sufficient for diagnosis of vascular thromboses. In the lung perfused blood volume images, in 76 (8.3%) of 918 segments, it was difficult to assess perfusion because of artifacts. The agreement between the 2 modalities was good (kappa = 0.70). The sensitivity of lung perfused blood volume in detecting perfusion defects was 96%; the specificity was 76%; the PPV was 94%, and the NPV was 29%.	2
63. Horton MR, Tuder RM. Primary pulmonary arterial hypertension presenting as diffuse micronodules on CT. <i>Crit Rev Comput Tomogr.</i> 2004;45(5-6):335-341.	Review/Other-Dx	1 patient	To present a case of PAH in a woman with a history of fenfluramine and phentermine use who presented with diffuse micronodules on CT scan.	No results stated in abstract.	4

**Suspected Pulmonary Hypertension
EVIDENCE TABLE**

Reference	Study Type	Patients/ Events	Study Objective (Purpose of Study)	Study Results	Study Quality
64. Nolan RL, McAdams HP, Sporn TA, Roggli VL, Tapson VF, Goodman PC. Pulmonary cholesterol granulomas in patients with pulmonary artery hypertension: chest radiographic and CT findings. <i>AJR Am J Roentgenol.</i> 1999;172(5):1317-1319.	Review/Other-Dx	20 patients	To describe the chest radiographic and CT findings of pulmonary cholesterol granulomas in patients with PAH.	Histopathologic evidence of cholesterol granulomas was found in 5 (25%) of 20 patients with severe PH. In 3 of these 5 patients, the granulomas manifested on chest radiographs and CT as small centrilobular nodules mimicking the appearance of sarcoidosis, bronchiolitis, hypersensitivity pneumonitis, or aspiration.	4
65. Sztrymf B, Yaici A, Girerd B, Humbert M. Genes and pulmonary arterial hypertension. <i>Respiration.</i> 2007;74(2):123-132.	Review/Other-Dx	N/A	To provide an overview of familial PAH.	Familial PAH was first described more than 50 years ago. Before the availability of modern genetic tools, studies of the genealogies demonstrated that these cases segregated as an autosomic dominant trait, with an incomplete penetrance and a genetic anticipation phenomenon by which age at onset of the disease is decreasing in the subsequent generations. Germline mutations in the gene coding for the bone morphogenetic protein receptor II are present in more than 70% of familial PAH and up to 26% of idiopathic, apparently sporadic cases (IPAH). Incomplete penetrance (around 20%) is a major pitfall because familial PAH becomes ignored when the disease skips 1 or several generations. Genetic counseling is complex, with a significant number of bone morphogenetic protein receptor II mutation healthy carriers screened in some families. Incomplete penetrance puts them in the anxious situation of being potentially affected in the future by this devastating condition or to transmit this risk to their offspring. Nevertheless, genetic testing and counseling is about to become a standard in the management of PAH. Recent international guidelines on PAH state that genetic testing is recommended in familial PAH and that IPAH patients have to be informed about the availability of such testing.	4

**Suspected Pulmonary Hypertension
EVIDENCE TABLE**

Reference	Study Type	Patients/ Events	Study Objective (Purpose of Study)	Study Results	Study Quality
66. Frazier AA, Franks TJ, Mohammed TL, Ozbudak IH, Galvin JR. From the Archives of the AFIP: pulmonary veno-occlusive disease and pulmonary capillary hemangiomas. <i>Radiographics</i> . 2007;27(3):867-882.	Review/Other-Dx	N/A	To discuss the radiologic manifestations that help to distinguish pulmonary venoocclusive disease and pulmonary capillary hemangiomas from PAH.	Pulmonary venoocclusive disease and pulmonary capillary hemangiomas are clinically indistinguishable from a primary PAH disorder such as primary PH or CTEPH. This distinction, however, is essential for appropriate pharmacologic intervention as well as for timely evaluation for lung transplantation.	4
67. Hansell DM. Small-vessel diseases of the lung: CT-pathologic correlates. <i>Radiology</i> . 2002;225(3):639-653.	Review/Other-Dx	N/A	Review highlights some of the less obvious imaging manifestations of occlusive and inflammatory diseases of the small pulmonary vessels.	In this review, emphasis was placed on the basic distinction between occlusive and inflammatory diseases of the small vessels of the lungs. An advantage of this simple categorization is the corresponding distinct differences in the CT manifestations of occlusive vs inflammatory small-vessel diseases.	4
68. Lippert JL, White CS, Cameron EW, Sun CC, Liang X, Rubin LJ. Pulmonary capillary hemangiomas: radiographic appearance. <i>J Thorac Imaging</i> . 1998;13(1):49-51.	Review/Other-Dx	N/A	Case report on pulmonary capillary hemangiomas.	The typical radiographic appearance of pulmonary capillary hemangiomas is a diffuse bilateral reticulonodular pattern associated with enlarged central pulmonary arteries.	4
69. Maltby JD, Gouverne ML. CT findings in pulmonary venoocclusive disease. <i>J Comput Assist Tomogr</i> . 1984;8(4):758-761.	Review/Other-Dx	1 patient	A case report of pulmonary venoocclusive disease in which chest radiography, when supplemented with CT after contrast medium administration, was specific enough to make the diagnosis without resorting to more invasive, potentially dangerous procedures.	A brief review of the literature, with emphasis on etiology, pathogenesis, and possible therapeutic approaches, emphasizes the importance of early diagnosis of this rare and usually fatal disease.	4
70. Mandel J, Mark EJ, Hales CA. Pulmonary veno-occlusive disease. <i>Am J Respir Crit Care Med</i> . 2000;162(5):1964-1973.	Review/Other-Dx	N/A	A summary on the current state of knowledge regarding pulmonary venoocclusive disease.	Pulmonary venoocclusive disease remains a rare and poorly understood syndrome that likely represents a final common pathway of disease caused by a variety of insults. The importance of diagnosing the condition is due largely to its poorer prognosis than either primary PH or CTEPH; this necessitates more rapid evaluation and listing for lung transplantation. In addition, the frequent deterioration that has been reported in response to vasodilators necessitates that these agents be used more judiciously than when primary PH is present.	4

**Suspected Pulmonary Hypertension
EVIDENCE TABLE**

Reference	Study Type	Patients/ Events	Study Objective (Purpose of Study)	Study Results	Study Quality
71. Swensen SJ, Tashjian JH, Myers JL, et al. Pulmonary venoocclusive disease: CT findings in eight patients. <i>AJR Am J Roentgenol.</i> 1996;167(4):937-940.	Review/Other-Dx	8 patients	To describe the CT findings of pulmonary venoocclusive disease.	7 of the 8 patients had interlobular septal thickening. All 8 patients had regions of ground-glass opacity. 4 of the 8 patients had a mosaic pattern of lung attenuation. No enlarged hilar or mediastinal nodes were revealed. 5 patients had bilateral pleural effusions. The most common CT findings in these 8 patients with pulmonary venoocclusive disease were smooth interlobular septal thickening, diffuse multifocal regions of ground-glass opacity, pleural effusions, enlarged central pulmonary arteries, and pulmonary veins of normal caliber. 4 patients had a mosaic pattern of lung attenuation on the CT scans. These findings are highly suggestive of pulmonary venoocclusive disease and may be helpful in difficult cases. Definitive diagnosis requires lung biopsy.	4
72. American College of Radiology. ACR Appropriateness Criteria®: Chronic Dyspnea — Suspected Pulmonary Origin. Available at: https://acsearch.acr.org/docs/69448/Narrative/ .	Review/Other-Dx	N/A	Evidence-based guidelines to assist referring physicians and other providers in making the most appropriate imaging or treatment decision for a specific clinical condition.	No results stated in abstract.	4
73. American College of Radiology. ACR Appropriateness Criteria®: Dyspnea — Suspected Cardiac Origin. Available at: https://acsearch.acr.org/docs/69407/Narrative/ .	Review/Other-Dx	N/A	Evidence-based guidelines to assist referring physicians and other providers in making the most appropriate imaging or treatment decision for a specific clinical condition.	No results stated in abstract.	4
74. American College of Radiology. ACR–NASCI–SIR–SPR Practice Parameter for the Performance and Interpretation of Body Computed Tomography Angiography (CTA). Available at: http://www.acr.org/~media/ACR/Documents/PGTS/guidelines/Body_CTA.pdf .	Review/Other-Dx	N/A	Guidance document to promote the safe and effective use of diagnostic and therapeutic radiology by describing specific training, skills and techniques.	N/A	4

**Suspected Pulmonary Hypertension
EVIDENCE TABLE**

Reference	Study Type	Patients/ Events	Study Objective (Purpose of Study)	Study Results	Study Quality
75. Marcus JT, Vonk Noordegraaf A, Roeleveld RJ, et al. Impaired left ventricular filling due to right ventricular pressure overload in primary pulmonary hypertension: noninvasive monitoring using MRI. <i>Chest</i> . 2001;119(6):1761-1765.	Observational-Dx	12 patients and 14 controls	To analyze the effect of primary PH on cardiac function using MRI.	In the patients, leftward ventricular septal bowing was quantified in early diastole by the septal curvature of -0.14 ± 0.07 cm(-1), and the septal to free-wall curvature ratio of -0.42 ± 0.21 . LV EDV and LV SV correlated negatively with diastolic PAP ($P=0.004$ and $P=0.04$, respectively). In patients vs control subjects, RV SV was reduced (52 ± 12 mL vs 82 ± 11 mL, $P<0.0001$); LV peak filling rate was smaller (2.2 ± 0.7 EDV/s vs 3.3 ± 0.5 EDV/s, $P<0.001$); LV EDV was smaller (81 ± 23 mL vs 117 ± 19 mL, $P=0.001$); and LV SV was smaller (49 ± 18 mL vs 83 ± 13 mL, $P<0.0001$). In primary PH, RV pressure overload leads to leftward ventricular septal bowing and reduced RV output. By decreased blood delivery, LV filling is reduced, which results in decreased LV SV by the Frank-Starling mechanism.	4
76. McCann GP, Gan CT, Beek AM, Niessen HW, Vonk Noordegraaf A, van Rossum AC. Extent of MRI delayed enhancement of myocardial mass is related to right ventricular dysfunction in pulmonary artery hypertension. <i>AJR Am J Roentgenol</i> . 2007;188(2):349-355.	Observational-Dx	15 patients	To assess the presence and extent of delayed contrast enhancement of ventricular myocardium in pulmonary artery hypertension.	All patients showed delayed contrast enhancement at the insertion points of the RV free wall to the interventricular septum (15 inferior, 13 anterior). The mean weight of the delayed contrast-enhanced myocardial mass was 3.1 ± 1.9 g (size range, 0.3-3.9% of the total myocardial mass). The extent of the delayed contrast-enhancing myocardium was inversely related to the RVEF ($r = -0.63$, $P=0.001$), RV SV ($r = -0.67$, $P=0.006$), and RV ESV index ($r = -0.51$, $P=0.05$) but not to any invasively measured hemodynamic index or N-terminal pro brain natriuretic peptide. Myocardial delayed contrast enhancement occurs frequently in patients with severe symptomatic pulmonary artery hypertension and is inversely related to measures of RV systolic function.	3

**Suspected Pulmonary Hypertension
EVIDENCE TABLE**

Reference	Study Type	Patients/ Events	Study Objective (Purpose of Study)	Study Results	Study Quality
77. McLure LE, Peacock AJ. Imaging of the heart in pulmonary hypertension. <i>Int J Clin Pract Suppl.</i> 2007(156):15-26.	Review/Other-Dx	N/A	Review techniques currently used to image the heart in patients with PH. Imaging modalities discussed include echocardiography, radionuclide ventriculography, cardiac CT and CMR focusing on the rapidly evolving technique of CMR.	No results stated in abstract.	4
78. van Wolferen SA, Marcus JT, Boonstra A, et al. Prognostic value of right ventricular mass, volume, and function in idiopathic pulmonary arterial hypertension. <i>Eur Heart J.</i> 2007;28(10):1250-1257.	Observational-Dx	64 patients	To examine the relationship between RV structure and function and survival in IPAH.	During a mean follow-up of 32 months, 19 patients died. A low SV, RV dilatation, and impaired LV filling independently predicted mortality. In addition, a further decrease in SV, progressive RV dilatation, and further decrease in LV EDV at 1-year follow-up were the strongest predictors of mortality. According to Kaplan–Meier survival curves, survival was lower in patients with an inframedian SV index 25 mL/m2, a supramedian RV EDV index 84 mL/m2, and an inframedian LV EDV 40 mL/m2. The RV contains prognostic information in IPAH. A large RV volume, low SV, and a reduced LV volume are strong independent predictors of mortality and treatment failure.	3
79. Nikolaou K, Schoenberg SO, Attenberger U, et al. Pulmonary arterial hypertension: diagnosis with fast perfusion MR imaging and high-spatial-resolution MR angiography--preliminary experience. <i>Radiology.</i> 2005;236(2):694-703.	Observational-Dx	29 patients	To determine prospectively the accuracy of a MR perfusion imaging and MRA protocol for differentiation of CTEPH and primary PH by using parallel acquisition techniques.	A correct diagnosis was made in 26 (90%) of 29 patients by using this comprehensive MRI protocol. Results of MR perfusion imaging demonstrated 79% agreement (ie, identical diagnosis on a per-patient basis) with those of perfusion scintigraphy, and results of MRA demonstrated 86% agreement with those of DSA and/or CTA. Interobserver agreement was good for both MR perfusion imaging and MRA (0.63 and 0.70, respectively). The combination of fast MR perfusion imaging and high-spatial resolution MRA with parallel acquisition techniques enables the differentiation of primary PH from CTEPH with high accuracy.	3

**Suspected Pulmonary Hypertension
EVIDENCE TABLE**

Reference	Study Type	Patients/ Events	Study Objective (Purpose of Study)	Study Results	Study Quality
80. Swift AJ, Rajaram S, Condliffe R, et al. Diagnostic accuracy of cardiovascular magnetic resonance imaging of right ventricular morphology and function in the assessment of suspected pulmonary hypertension results from the ASPIRE registry. <i>J Cardiovasc Magn Reson.</i> 2012;14:40.	Observational-Dx	233 patients	To compare the diagnostic accuracy of a variety of CMR parameters to identify PH confirmed at cardiac catheterization in unselected patients with suspected PH attending a referral center.	Ventricular mass index was the CMR measurement with the strongest correlation with MPAP ($r = 0.78$) and the highest diagnostic accuracy for the detection of PH (area under the receiver operating characteristic curve of 0.91) compared to an receiver operating characteristic curve of 0.88 for echocardiography calculated MPAP. Late gadolinium enhancement, ventricular mass index ≥ 0.4 , retrograde flow ≥ 0.3 L/min/m ² and PA relative area change $\leq 15\%$ predicted the presence of PH with a high degree of diagnostic certainty with a PPV of 98%, 97%, 95% and 94% respectively. No single CMR parameter could confidently exclude the presence of PH.	3
81. Iwasawa T. Diagnosis and management of pulmonary arterial hypertension using MR imaging. <i>Magn Reson Med Sci.</i> 2013;12(1):1-9.	Review/Other-Dx	N/A	To review the current status of MRI of the right side of the heart and pulmonary circulation in patients with PAH and other associated pulmonary diseases.	No results stated in abstract.	4
82. Boxt LM. MR imaging of pulmonary hypertension and right ventricular dysfunction. <i>Magn Reson Imaging Clin N Am.</i> 1996;4(2):307-325.	Review/Other-Dx	N/A	Review MRI of PH and RV dysfunction.	MRI provides direct, noninvasive visualization of the RV chamber as well as the myocardium itself, allowing reliable demonstration of morphologic changes in the size and shape of the ventricle, thickness of the myocardium, and presence of abnormal infiltration by fat or edema. Furthermore, because MRI techniques do not depend upon geometric assumptions about the complex shape of the right ventricle, they may be used for accurate and reproducible quantitation of RV volume and myocardial mass.	4
83. Lopez-Costa I, Bhalla S, Raptis C. Magnetic resonance imaging for pulmonary hypertension: methods, applications, and outcomes. <i>Top Magn Reson Imaging.</i> 2014;23(1):43-50.	Review/Other-Dx	N/A	To describe the utility of MRI in PH.	No results stated in abstract.	4

**Suspected Pulmonary Hypertension
EVIDENCE TABLE**

Reference	Study Type	Patients/ Events	Study Objective (Purpose of Study)	Study Results	Study Quality
84. Marcu CB, Beek AM, Van Rossum AC. Cardiovascular magnetic resonance imaging for the assessment of right heart involvement in cardiac and pulmonary disease. <i>Heart Lung Circ.</i> 2006;15(6):362-370.	Review/Other-Dx	N/A	Review of the contemporary use of CMR for the evaluation of right heart involvement in various cardiopulmonary diseases.	CMR is a virtually 3D tomographic technique which has entered the mainstream of clinical cardiovascular imaging over the last decade. Compared to other imaging methods CMR allows the accurate quantification of RV volumes, myocardial mass, and transvalvular flow with the added benefit of tissue characterization and without the use of ionizing radiation.	4
85. Roeleveld RJ, Marcus JT, Faes TJ, et al. Interventricular septal configuration at mr imaging and pulmonary arterial pressure in pulmonary hypertension. <i>Radiology.</i> 2005;234(3):710-717.	Observational-Dx	39 subjects	To investigate whether a relationship exists between septum shape and systolic PAP in patients with PH.	Of 39 subjects, 37 had PH. Maximal distortion of normal septal shape was found during RV relaxation phase. Systolic PAP was proportional to septal curvature: $r=0.77$ ($P<.001$), slope=-114.7, and intercept=67.2. In the 2 vasodilator responsive subjects, a significant reduction of leftward ventricular septal bowing was observed in response to reduction of RV pressure. In 37 patients with PH, systolic PAP higher than 67 mm Hg may be expected when leftward curvature is observed.	4

Suspected Pulmonary Hypertension
EVIDENCE TABLE

Reference	Study Type	Patients/ Events	Study Objective (Purpose of Study)	Study Results	Study Quality
86. Kreitner KF, Wirth GM, Krummenauer F, et al. Noninvasive assessment of pulmonary hemodynamics in patients with chronic thromboembolic pulmonary hypertension by high temporal resolution phase-contrast MRI: correlation with simultaneous invasive pressure recordings. <i>Circ Cardiovasc Imaging</i> . 2013;6(5):722-729.	Observational-Dx	19 patients	To create a model for estimating MPAP and pulmonary vascular resistance in patients with CTEPH by high temporal resolution phase-contrast MRI and to correlate the results with simultaneously acquired, invasive catheter-based measurements (simultaneously measured MPAP) and with RHC measurements.	Velocity- and flow-time curves of phase-contrast MRI were used to calculate absolute acceleration time, maximum of mean velocities, volume of acceleration, and maximum flow acceleration (dQ/dt). On the basis of these parameters, multiple linear regression analysis revealed maximum achievable model fit (B=0.902) for the following linear combination equation to calculate MPAP (MPAP_cal): MPAP_cal=69.446-(0.521 x absolute acceleration time)-(0.570 x mean velocities)+(1.507 x volume of acceleration)+(0.002 x dQ/dt). There was a statistically significant equivalence of MPAP_cal and simultaneously measured MPAP with a goodness of fit of 0.892. Pulmonary vascular resistance was overestimated by calculated pulmonary vascular resistance on the basis of phase-contrast MRI in comparison with RHC-based measurements by a median of -112 dyn.s.cm(-5), the pairwise regression formula revealed a goodness of fit of 0.792.	3
87. Gan CT, Lankhaar JW, Westerhof N, et al. Noninvasively assessed pulmonary artery stiffness predicts mortality in pulmonary arterial hypertension. <i>Chest</i> . 2007;132(6):1906-1912.	Observational-Dx	70 patients	To investigate whether proximal pulmonary artery stiffness, in terms of area distensibility and noninvasively assessed relative area change, calculated as relative cross-sectional area change, predicts mortality in patients with PAH.	In 70 patients, the diagnosis PAH was confirmed, and 16 subjects served as control subjects. In comparison with control subjects, proximal pulmonary arteries of patients were distended (685 +/- 214 mm ² vs 411 +/- 153 mm ² , <i>P</i> <0.001), less distensible (area distensibility = 0.46 +/- 0.38.10(-2) mm Hg(-1) vs 3.69 +/- 1.96.10(-2) mm Hg(-1), <i>P</i> <0.0001), and relative area change was smaller (20 +/- 10% vs 58 +/- 21%, <i>P</i> <0.0001) [mean +/- SD]. Relative area change showed an inverse curvilinear relation with MPAP (R ² = 0.47). 18 patients (26%) died because of cardiopulmonary causes. Patients with a pulmonary artery relative area change ≤16% had a worse prognosis than those with a value >16% (log-rank <i>P</i> <0.001). Relative area change predicted mortality better than area distensibility.	3

* See Last Page for Key

**Suspected Pulmonary Hypertension
EVIDENCE TABLE**

Reference	Study Type	Patients/ Events	Study Objective (Purpose of Study)	Study Results	Study Quality
88. Swift AJ, Rajaram S, Condliffe R, et al. Pulmonary artery relative area change detects mild elevations in pulmonary vascular resistance and predicts adverse outcome in pulmonary hypertension. <i>Invest Radiol.</i> 2012;47(10):571-577.	Observational-Dx	134 patients	To evaluate the clinical use of MRI measurements related to pulmonary artery stiffness in the evaluation of PH.	The relationship between pulmonary vascular resistance and pulmonary artery relative area change was best reflected by an inverse linear model. Patients with mild elevation in pulmonary vascular resistance (<4 Woods units) demonstrated reduced relative area change ($P=0.02$) and increased RV mass index ($P<0.0001$) without significant loss of RV function ($P=0.17$). At follow-up of 0 to 40 months, 18 patients with PH had died (16%). Analysis of Kaplan-Meier plots showed that both area change and relative area change predicted mortality (log-rank test, $P=0.046$ and $P=0.012$, respectively). Area change and relative area change were also predictors of mortality using univariate Cox proportional hazards regression analysis ($P=0.046$ and $P=0.03$, respectively).	3
89. Ley S, Mereles D, Puderbach M, et al. Value of MR phase-contrast flow measurements for functional assessment of pulmonary arterial hypertension. <i>Eur Radiol.</i> 2007;17(7):1892-1897.	Observational-Dx	25 patients	To compare the pulmonary hemodynamics between healthy volunteers and patients with PAH and correlate MR flow measurements with echocardiography.	In PAH patient's transthoracic echocardiography and RHC served as the gold standard. In comparison to volunteers, the PAH patients showed significantly reduced pulmonary velocities ($P=0.002$), blood flow ($P=0.002$) and pulmonary distensibility ($P=0.008$). In patients, the time to peak velocity was shorter ($P<0.001$), and the velocity rise gradient was steeper ($P=0.002$) than in volunteers. While in volunteers the peak velocity in the aorta was reached earlier, it was the reverse in patients. Patients showed a significant bronchoscopic shunt ($P=0.01$).	3
90. Bremerich J, Reddy GP, Higgins CB. MRI of supracristal ventricular septal defects. <i>J Comput Assist Tomogr.</i> 1999;23(1):13-15.	Review/Other-Dx	2 patients	MR features of 2 cases of supracristal ventricular septal defect are described.	In both patients, axial spin-echo T1-weighted images demonstrated a defect between the base of the aorta and the upper posterior aspect of the RV infundibulum. Cine MRI in the 2 cases showed left-to-right shunting with a flow jet in the distal RV outflow tract that propagated into the main pulmonary artery. Both patients had prolapse of the right sinus of Valsalva, and 1 had aortic insufficiency.	4

**Suspected Pulmonary Hypertension
EVIDENCE TABLE**

Reference	Study Type	Patients/ Events	Study Objective (Purpose of Study)	Study Results	Study Quality
91. Ferrari VA, Scott CH, Holland GA, Axel L, Sutton MS. Ultrafast three-dimensional contrast-enhanced magnetic resonance angiography and imaging in the diagnosis of partial anomalous pulmonary venous drainage. <i>J Am Coll Cardiol.</i> 2001;37(4):1120-1128.	Observational-Dx	20 patients	To evaluate patients with suspected anomalous pulmonary veins and atrial septal defects using fast cine MRI and ultrafast 3D MRA.	Partial anomalous pulmonary venous drainage was demonstrated in 16/20 patients and was excluded in 4 patients. MRI correctly diagnosed anomalous pulmonary veins and atrial septal defects in all patients (100%) who underwent surgery. For the diagnosis of anomalous pulmonary veins, the MRI and catheterization results agreed in 74% of patients and the MRI and transesophageal echocardiography agreed in 75% of patients. For atrial septal defects, MRI agreed with catheterization and transesophageal echocardiography in 53% and 83% of patients, respectively. Fast cine MRI with 3D contrast-enhanced MRA provides rapid and comprehensive anatomic definition of anomalous pulmonary veins and atrial septal defects in patients with adult congenital heart disease in a single examination.	3
92. Wang ZJ, Reddy GP, Gotway MB, Yeh BM, Higgins CB. Cardiovascular shunts: MR imaging evaluation. <i>Radiographics.</i> 2003;23 Spec No:S181-194.	Review/Other-Dx	N/A	Review MRI techniques that are most useful for detecting, localizing, and quantifying shunts in atrial, ventricular, and atrioventricular septal defects; patent ductus arteriosus; aortopulmonary window; and partial anomalous pulmonary venous return.	MRI is a valuable tool for depicting cardiac anatomy and quantifying function. It is particularly useful for the evaluation of cardiac shunts in supracristal ventricular septal defect, atrioventricular septal defect, and partial anomalous pulmonary venous return, in which echocardiography and conventional angiography have limited use. MRI is also well suited for the noninvasive quantification of shunt volume and functional evaluation of shunt severity.	4
93. Barker AJ, Roldan-Alzate A, Entezari P, et al. Four-dimensional flow assessment of pulmonary artery flow and wall shear stress in adult pulmonary arterial hypertension: results from two institutions. <i>Magn Reson Med.</i> 2015;73(5):1904-1913.	Observational-Dx	19 healthy subjects and 17 PAH subjects	To compare pulmonary artery flow using Cartesian and radially sampled 4D flow MRI at 2 institutions.	Vmax, Qmax, SV, and wall shear stress at all locations were significantly lower ($P<0.05$) in PAH compared with healthy subjects. The limits of agreement were 0.16 m/s, 2.4 L/min, 10 mL, and 0.31 N/m(2) for Vmax, Qmax, SV, and wall shear stress, respectively. Differences between Qmax and SV using Cartesian and radial sequences were not significant. Plane placement and acquisition exhibited isolated, site-based differences between Vmax and wall shear stress.	3

**Suspected Pulmonary Hypertension
EVIDENCE TABLE**

Reference	Study Type	Patients/ Events	Study Objective (Purpose of Study)	Study Results	Study Quality
94. Odagiri K, Inui N, Miyakawa S, et al. Abnormal hemodynamics in the pulmonary artery seen on time-resolved 3-dimensional phase-contrast magnetic resonance imaging (4D-flow) in a young patient with idiopathic pulmonary arterial hypertension. <i>Circ J</i> . 2014;78(7):1770-1772.	Review/Other-Dx	1	To present a patient's pulmonary arterial hemodynamics, gadolinium-enhanced MRA and phase-resolved 3-D phase-contrast MRI (MRI; 4D-flow) of the pulmonary artery.	The present report shows that 4D-flow is a useful noninvasive method for the qualitative and quantitative characterization of not only blood flow but also wall shear stress in PAH patients.	4
95. Reiter G, Reiter U, Kovacs G, et al. Magnetic resonance-derived 3-dimensional blood flow patterns in the main pulmonary artery as a marker of pulmonary hypertension and a measure of elevated mean pulmonary arterial pressure. <i>Circ Cardiovasc Imaging</i> . 2008;1(1):23-30.	Observational-Dx	102 patients	To investigate characteristic differences in 3D blood flow patterns in the main pulmonary artery of patients with manifest PH, patients with latent PH, and subjects without PH to determine the blood flow pattern-related measures for mPAP and PH.	Main findings were as follows: (1) Manifest PH coincides with the appearance of a vortex of blood flow in the main pulmonary artery (sensitivity and specificity of 1.00, 95% CIs of 0.84 to 1.00 and 0.87 to 1.00, respectively), and (2) the relative period of existence of the vortex correlates significantly with mean pulmonary arterial pressure at rest (correlation coefficient of 0.94). To test the diagnostic performance of the vortex criterion, we furthermore investigated 55 patients in a blinded prospective study (22 with manifest PH, 32 with latent PH, and 1 healthy subject), which resulted in a sensitivity of 1.00 and specificity of 0.91 (95% CIs of 0.84 to 1.00 and 0.76 to 0.98, respectively). Comparison of catheter-derived MPAP measurements and calculated MPAP values resulted in a SD of differences of 3.6 mm Hg.	2
96. Roldan-Alzate A, Frydrychowicz A, Johnson KM, et al. Non-invasive assessment of cardiac function and pulmonary vascular resistance in an canine model of acute thromboembolic pulmonary hypertension using 4D flow cardiovascular magnetic resonance. <i>J Cardiovasc Magn Reson</i> . 2014;16:23.	Observational-Dx	6 adult female beagles	To quantify RV and LV function, pulmonary artery flow, tricuspid valve regurgitation velocity, and aorta flow from a single 4D flow CMR sequence in a canine model of acute thromboembolic PH.	Biases between 4D flow CMR and balanced steady-state free precession were 0.8 mL and 1.6 mL for RV EDV and RV ESV, respectively, and 0.8 mL and 4 mL for LV EDV and LV ESV, respectively. Flow in the MPA, right pulmonary artery, and left pulmonary artery did not change after induction of acute PAH ($P=0.42-0.81$). MPA, right pulmonary artery, and left pulmonary artery flow determined with 4D flow CMR was significantly lower than with 2D flow ($P<0.05$). The correlation between pulmonary artery flow/tricuspid valve regurgitation velocity and pulmonary vascular resistance was 0.95. The average pulmonary artery flow/aorta flow was 0.96 +/- 0.11.	3

**Suspected Pulmonary Hypertension
EVIDENCE TABLE**

Reference	Study Type	Patients/ Events	Study Objective (Purpose of Study)	Study Results	Study Quality
97. Alunni JP, Degano B, Arnaud C, et al. Cardiac MRI in pulmonary artery hypertension: correlations between morphological and functional parameters and invasive measurements. <i>Eur Radiol.</i> 2010;20(5):1149-1159.	Observational-Dx	40 patients	To compare CMR with RHC in patients with PH and to evaluate its ability to assess PH severity.	Interventricular septum position was correlated with PAPs and PVR (pulmonary vascular resistance). Median PAPs and resistance were significantly higher in patients with an abnormal septal position compared with those with a normal position. Correlations were good between the RVEF and PVR, RV ESV and PAP, percentage of RV area change and PVR, and diastolic and systolic ventricular area ratio and PVR. These parameters were significantly associated with PH severity. CMR can help to assess the severity of PH.	3
98. Gan CT, Holverda S, Marcus JT, et al. Right ventricular diastolic dysfunction and the acute effects of sildenafil in pulmonary hypertension patients. <i>Chest.</i> 2007;132(1):11-17.	Observational-Dx	25 PH patients and 11 control subjects	To determine whether RV diastolic function is impaired in PH patients, and whether it is related to RV mass and afterload.	Compared to control subjects, patients had prolonged mean (+/- SD) Isovolumic relaxation time (133.5 +/- 53.2 vs 29.3 +/- 20.8 ms, respectively; $P < 0.001$), decreased E (3.0 +/- 1.6 vs 6.4 +/- 2.5 s(-1), respectively; $P < 0.001$) and E/A ratio (1.1 +/- 0.7 vs 5.3 +/- 4.9, respectively; $P < 0.001$), and increased A (3.0 +/- 1.4 vs 1.5 +/- 0.9 s(-1), respectively; $P = 0.001$). Isovolumic relaxation time was related to RV mass ($r(25) = 0.56$; $P = 0.005$) and pulmonary vascular resistance ($r(25) = 0.74$; $P < 0.0001$). Sildenafil therapy reduced RV afterload and improved RV diastolic and systolic function. Isovolumic relaxation time was correlated with N-terminal pro-brain natriuretic peptide (NT-proBNP) level ($r = 0.70$; $P < 0.001$), and was inversely related to cardiac index ($r = -0.70$; $P < 0.001$) and RV ejection fraction ($r = -0.69$; $P < 0.001$). In PH patients, RV diastolic dysfunction is related to RV mass and afterload. RV diastolic function improves by reducing afterload. The correlations between diastolic function and prognostic parameters showed that diastolic function is most impaired in patients with severe disease.	4

**Suspected Pulmonary Hypertension
EVIDENCE TABLE**

Reference	Study Type	Patients/ Events	Study Objective (Purpose of Study)	Study Results	Study Quality
99. Nazarian S, Hansford R, Roguin A, et al. A prospective evaluation of a protocol for magnetic resonance imaging of patients with implanted cardiac devices. <i>Ann Intern Med.</i> 2011;155(7):415-424.	Observational-Dx	438 patients with devices who underwent 555 MRI studies	To define the safety of a protocol for MRI at the commonly used magnetic strength of 1.5 T in patients with implanted cardiac devices.	In 3 patients (0.7% [95% CI, 0% to 1.5%]), the device reverted to a transient back-up programming mode without long-term effects. RV sensing (median change, 0 mV [IQR, -0.7 to 0 V]) and atrial and right and LV lead impedances (median change, -2 O [IQR, -13 to 0 O], -4 O [IQR, -16 to 0 O], and -11 O [IQR, -40 to 0 O], respectively) were reduced immediately after MRI. At long-term follow-up (61% of patients), decreased RV sensing (median, 0 mV, [IQR, -1.1 to 0.3 mV]), decreased RV lead impedance (median, -3 O, [IQR, -29 to 15 O]), increased RV capture threshold (median, 0 V, IQR, [0 to 0.2 O]), and decreased battery voltage (median, -0.01 V, IQR, -0.04 to 0 V) were noted. The observed changes did not require device revision or reprogramming. With appropriate precautions, MRI can be done safely in patients with selected cardiac devices. Because changes in device variables and programming may occur, electrophysiologic monitoring during MRI is essential.	4
100. American College of Radiology. <i>Manual on Contrast Media</i> . Available at: http://www.acr.org/Quality-Safety/Resources/Contrast-Manual .	Review/Other-Dx	N/A	Guidance document on contrast media to assist radiologists in recognizing and managing risks associated with the use of contrast media.	N/A	4

**Suspected Pulmonary Hypertension
EVIDENCE TABLE**

Reference	Study Type	Patients/ Events	Study Objective (Purpose of Study)	Study Results	Study Quality
101. Hoepfer MM, Lee SH, Voswinckel R, et al. Complications of right heart catheterization procedures in patients with pulmonary hypertension in experienced centers. <i>J Am Coll Cardiol.</i> 2006;48(12):2546-2552.	Observational-Dx	7,218 total RHC procedures: Retrospective period - 5,727 Prospective period - 1,491	To assess the risks associated with RHC procedures in patients with PH.	The overall number of serious adverse events was 76 (1.1%, 95% CI, 0.8% to 1.3%). The most frequent complications were related to venous access, followed by arrhythmias and hypotensive episodes related to vagal reactions or pulmonary vasoreactivity testing. The vast majority of these complications were mild to moderate in intensity and resolved either spontaneously or after appropriate intervention. 4 fatal events were recorded in association with any of the catheter procedures, resulting in an overall procedure-related mortality of 0.055% (95% CI 0.01% to 0.099%). When performed in experienced centers, RHC procedures in patients with PH are associated with low morbidity and mortality rates.	4
102. Rosenkranz S. Pulmonary hypertension: current diagnosis and treatment. <i>Clin Res Cardiol.</i> 2007;96(8):527-541.	Review/Other-Dx	N/A	The article provides an overview of the definition, classification, pathophysiology, and clinical presentation of various forms of PH. Furthermore, it summarizes the recommended diagnostic workup and the current treatment options particularly in PAH, with special emphasis on prostanoids, endothelin receptor antagonists, and phosphodiesterase type 5 inhibitors such as sildenafil. Finally, novel developments are being discussed which currently represent an exciting field of research.	No results stated in abstract.	4
103. Love C, Tomas MB, Tronco GG, Palestro CJ. FDG PET of infection and inflammation. <i>Radiographics.</i> 2005;25(5):1357-1368.	Review/Other-Dx	N/A	Review role of FDG PET in the evaluation of infection and inflammation.	FDG PET promises to be helpful in the diagnosis of infection and inflammation. FDG PET will likely assume increasing importance in assessing fever of undetermined origin, spinal osteomyelitis, vasculitis, and sarcoidosis and may even become the radionuclide imaging procedure of choice in the evaluation of some or all of these pathologic conditions.	4

**Suspected Pulmonary Hypertension
EVIDENCE TABLE**

Reference	Study Type	Patients/ Events	Study Objective (Purpose of Study)	Study Results	Study Quality
104. Zhuang H, Alavi A. 18-fluorodeoxyglucose positron emission tomographic imaging in the detection and monitoring of infection and inflammation. <i>Semin Nucl Med.</i> 2002;32(1):47-59.	Review/Other-Dx	N/A	Review role of FDG-PET in the detection and monitoring of infection and inflammation.	It is becoming evident that PET imaging will play a major role in the treatment of patients with suspected infection and inflammation. PET has been shown to be particularly valuable in the evaluation of chronic osteomyelitis, infected prostheses, sarcoidosis, fever of unknown origin, and acquired immunodeficiency syndrome.	4
105. Hagan G, Southwood M, Treacy C, et al. (18)FDG PET imaging can quantify increased cellular metabolism in pulmonary arterial hypertension: A proof-of-principle study. <i>Pulm Circ.</i> 2011;1(4):448-455.	Observational-Dx	14 patients and 6 controls	The hypothesis of this study is that increased FDG uptake may be present in the lungs, large pulmonary arteries and right ventricle of patients with PH, and that this uptake would be related to markers of immune activation.	There were no correlations between FDG uptake and high-sensitivity C-reactive protein or inflammatory cytokine levels. N-Terminal Probrain natriuretic peptide correlated with RV uptake in those with PH ($r=0.55$, $P=0.04$). Authors found increased FDG uptake in the lung parenchyma and RV of subjects with IPAH. The lung uptake might be useful as a surrogate marker of increased cellular metabolism and immune activation as underlying mechanisms in this disease. Further evaluation of the impact of targeted therapies in treatment-naive patients and the significance of RV uptake is suggested.	4

**Suspected Pulmonary Hypertension
EVIDENCE TABLE**

Reference	Study Type	Patients/ Events	Study Objective (Purpose of Study)	Study Results	Study Quality
106. Oikawa M, Kagaya Y, Otani H, et al. Increased [18F]fluorodeoxyglucose accumulation in right ventricular free wall in patients with pulmonary hypertension and the effect of epoprostenol. <i>J Am Coll Cardiol.</i> 2005;45(11):1849-1855.	Observational-Dx	24 patients	To examine whether RV FDG accumulation is increased in patients with PH using gated PET and whether RV FDG accumulation changes after therapy with epoprostenol.	The corrected RV SUV of FDG was significantly correlated with the pulmonary vascular resistance, MPAP, right atrial pressure, RV wall stress, and plasma brain natriuretic peptide levels, but not with the RV wall thickness and mass. After pulmonary vasodilator therapy with epoprostenol for 3 months, the corrected RV SUV of FDG significantly decreased in the responders, but not in the nonresponders, and the percentage change of the corrected RV SUV of FDG was significantly correlated with the percentage change of the pulmonary vascular resistance ($r = 0.78$; $P < 0.01$) and RV systolic wall stress ($r = 0.76$; $P < 0.05$). The RV FDG accumulation corrected for the partial volume effect was significantly increased in accordance with the severity of the RV pressure overload in patients with PH. Furthermore, the corrected RV FDG accumulation was decreased after the treatment with epoprostenol in accordance with the degree of reduction in the pulmonary vascular resistance and RV peak-systolic wall stress.	3
107. Yang T, Wang L, Xiong CM, et al. The ratio of (18)F-FDG activity uptake between the right and left ventricle in patients with pulmonary hypertension correlates with the right ventricular function. <i>Clin Nucl Med.</i> 2014;39(5):426-430.	Observational-Dx	38 patients	To assess possible relationship between FDG uptake of ventricles and the function/hemodynamics of the RV in patients with PH.	The SUV of RV and SUV of LV were significantly higher in glucose-loading condition than in fasting condition. In both fasting and glucose-loading conditions, SUV of RV and SUV of RV/SUV of LV showed reverse correlation with RVEF derived from CMR. In addition, in both fasting and glucose-loading conditions, SUVR and SUVR/L showed positive correlations with pulmonary vascular resistance. However, only SUVR/L in glucose-loading condition could independently predict RVEF after adjusted for age, body mass index, sex, mean right atrial pressure, mean pulmonary arterial pressure, and pulmonary vascular resistance ($P = 0.048$).	3

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EVIDENCE TABLE**

Reference	Study Type	Patients/ Events	Study Objective (Purpose of Study)	Study Results	Study Quality
108. Tatebe S, Fukumoto Y, Oikawa-Wakayama M, et al. Enhanced [18F]fluorodeoxyglucose accumulation in the right ventricular free wall predicts long-term prognosis of patients with pulmonary hypertension: a preliminary observational study. <i>Eur Heart J Cardiovasc Imaging</i> . 2014;15(6):666-672.	Observational-Dx	27 patients	To examine whether enhanced RV FDG accumulation by gated PET has a prognostic impact in patients with PH.	FDG-PET examination showed that cRV-SUV was significantly higher in the CW group compared with the non-CW group (10.1 vs 7.6, $P=0.02$). Univariate Cox hazard analysis showed that cRV-SUV was significantly correlated with the time to CW (hazard ratio 1.25, 95% CI, 1.04-1.51, $P=0.02$), which remained significant even after adjustment of World Health Organization functional class. Kaplan-Meier analysis showed that the patients with cRV-SUV ≥ 8.3 had poor prognosis compared with those with cRV-SUV < 8.3 (log-rank $P=0.005$ for time to CW and $P=0.07$ for mortality).	2
109. Wang L, Zhang Y, Yan C, et al. Evaluation of right ventricular volume and ejection fraction by gated (18)F-FDG PET in patients with pulmonary hypertension: comparison with cardiac MRI and CT. <i>J Nucl Cardiol</i> . 2013;20(2):242-252.	Observational-Dx	23 patients	To prospectively compare gated FDG-PET myocardial imaging (gated PET), CMR, and cardiac CT for the assessment of RV volume and ejection fraction in patients with PH.	Gated PET showed a moderate correlation ($r = 0.680$, $P < .001$) for RV EDV, good correlation for RV ESV ($r = 0.757$, $P < .001$) and RVEF ($r = 0.788$, $P < .001$) with CMR, and good correlation for RV EDV ($r = 0.767$, $P < .001$), RV ESV ($r = 0.837$, $P < .001$), and RVEF ($r = 0.730$, $P < .001$) with cardiac CT. Bland-Altman analysis revealed systematic underestimation of RV EDV and RV ESV and overestimation of RVEF with gated PET compared with CMR and cardiac CT. The correlation between RV ESV ($r = 0.863$, $P < .001$), RV ESV ($r = 0.903$, $P < .001$), and RVEF ($r = 0.853$, $P < .001$) of CMR and those of cardiac CT was excellent; Bland-Altman analysis showed only a slight systematic variation between CMR and cardiac CT. There were statistically significant negative correlations between RV-corrected SUV and RVEF-CMR ($r = -0.543$, $P < .01$), Corrected SUV R/L and RVEF-CMR ($r = -0.521$, $P < .05$), RV-corrected SUV and RVEF-cardiac CT ($r = -0.429$, $P < .05$), Corrected SUV R/L and RVEF-cardiac CT ($r = -0.580$, $P < .01$), respectively.	2

**Suspected Pulmonary Hypertension
EVIDENCE TABLE**

Reference	Study Type	Patients/ Events	Study Objective (Purpose of Study)	Study Results	Study Quality
110. Chong S, Kim TS, Kim BT, Cho EY, Kim J. Pulmonary artery sarcoma mimicking pulmonary thromboembolism: integrated FDG PET/CT. <i>AJR Am J Roentgenol</i> . 2007;188(6):1691-1693.	Review/Other-Dx	N/A	A case is presented on pulmonary artery sarcoma that showed high FDG uptake on integrated FDG-PET/CT.	FDG-PET/CT finding can be helpful in differentiating a pulmonary artery sarcoma from pulmonary thromboembolism.	4
111. James OG, Christensen JD, Wong TZ, Borges-Neto S, Koweek LM. Utility of FDG PET/CT in inflammatory cardiovascular disease. <i>Radiographics</i> . 2011;31(5):1271-1286.	Review/Other-Dx	N/A	Review role of FDG-PET/CT in inflammatory cardiovascular disease.	CT and MRI play important roles in the assessment of structural abnormalities of the cardiovascular system, and combined PET and CT may depict inflammatory processes before structural changes occur. Familiarity with the PET/CT appearances of inflammatory processes in the myocardium, pericardium, and vessels is important for accurate and prompt diagnosis.	4
112. Matsunaga N, Hayashi K, Sakamoto I, Ogawa Y, Matsumoto T. Takayasu arteritis: protean radiologic manifestations and diagnosis. <i>Radiographics</i> . 1997;17(3):579-594.	Review/Other-Dx	100 patients	Article presents the recently described radiologic features of the early (systemic or prepulseless) phase and the common and uncommon appearances of the late (occlusive) phase of Takayasu arteritis.	Familiarity with the varied chest radiographic, angiographic, CT, and MRI features of Takayasu arteritis will permit earlier diagnosis and treatment.	4
113. Hu XP, Xu JP, Liu NN. Primary pulmonary artery sarcoma: surgical management and differential diagnosis with pulmonary embolism and pulmonary valve stenosis. <i>J Card Surg</i> . 2009;24(6):613-616.	Review/Other-Dx	6 patients	To review 6 cases of primary pulmonary artery sarcomas and discuss clinical features, differential diagnosis, surgical treatment, and outcome of the tumors.	Histological examinations showed 5 malignant mesenchymomas and 1 fibrosarcoma. 1 patient died of refractory PH during operation. 2 patients died 4 months postoperatively because of brain metastases. 2 patients were alive for 3 and 9 months, respectively after the operation with recurrent tumor. 1 patient is alive even 2 years after resection with no signs of recurrence or metastasis. Because of similar clinical features, pulmonary artery sarcomas are often confused with other pulmonary vascular obstructive diseases. CT scanning and gadolinium-enhanced MRI could be useful methods for differential diagnosis. The prognosis is very poor. The survival time after resection varies from several months to several years depending on the presence of recurrence or metastasis. Early diagnosis and radical surgical resection presents the only opportunity for a potential cure.	4

**Suspected Pulmonary Hypertension
EVIDENCE TABLE**

Reference	Study Type	Patients/ Events	Study Objective (Purpose of Study)	Study Results	Study Quality
114. Mukhtyar C, Guillevin L, Cid MC, et al. EULAR recommendations for the management of large vessel vasculitis. <i>Ann Rheum Dis.</i> 2009;68(3):318-323.	Review/Other-Dx	Expert group (10 rheumatologists, 3 nephrologists, 2 immunologists, 2 internists, 1 clinical epidemiologist and 1 rep from a drug regulatory agency	To develop European League Against Rheumatism (EULAR) recommendations for the management of large vessel vasculitis.	7 recommendations were made relating to the assessment, investigation and treatment of patients with large vessel vasculitis. The strength of recommendations was restricted by the low level of evidence and EULAR standardized operating procedures. On the basis of evidence and expert consensus, management recommendations for large vessel vasculitis have been formulated and are commended for use in everyday clinical practice.	4

Evidence Table Key

Study Quality Category Definitions

- *Category 1* The study is well-designed and accounts for common biases.
- *Category 2* The study is moderately well-designed and accounts for most common biases.
- *Category 3* There are important study design limitations.
- *Category 4* The study is not useful as primary evidence. The article may not be a clinical study or the study design is invalid, or conclusions are based on expert consensus. For example:
 - a) the study does not meet the criteria for or is not a hypothesis-based clinical study (e.g., a book chapter or case report or case series description);
 - b) the study may synthesize and draw conclusions about several studies such as a literature review article or book chapter but is not primary evidence;
 - c) the study is an expert opinion or consensus document.
- M = Meta-analysis

Dx = Diagnostic

Tx = Treatment

Abbreviations Key

CI = Confidence interval

CMR = Cardiovascular magnetic resonance imaging

COPD = Chronic obstructive pulmonary disease

CT = Computed tomography

CTA = Computed tomography angiography

CTEPH = Chronic thromboembolic pulmonary hypertension

CTPA = Computed tomography pulmonary angiography

DSA = Digital subtraction angiography

EDV = End-diastolic volume

ESV = End-systolic volume

FDG-PET = Fluorine-18-2-fluoro-2-deoxy-D-glucose-positron emission tomography

IPAH = Idiopathic pulmonary arterial hypertension

IQR = Interquartile range

LV = Left ventricular

MPAP=Mean pulmonary artery pressure

MRA = Magnetic resonance angiography

MRI = Magnetic resonance imaging

NPV = Negative predictive value

OR = Odds ratio

PAH = Pulmonary arterial hypertension

PAP= Pulmonary artery pressure

PH = Pulmonary hypertension

PPV = Positive predictive value

RHC = Right heart catheterization

RV = Right ventricular

RVEF = Right ventricular ejection fraction

SD = Standard deviation

SUV = Standardized uptake value

SV = Stroke volume

V/Q = Ventilation-perfusion