Imaging of Pulmonary Hypertension

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KEYWORDS

• Pulmonary hypertension • Pulmonary arterial hypertension
• Chronic thromboembolic pulmonary hypertension • Systemic sclerosis • Cor pulmonale
• Magnetic resonance imaging • Computed tomography

KEY POINTS

- Pulmonary hypertension from worsening of left ventricular function is a common disease and is becoming more of a public health issue as the world's population ages.
- The critical issue with all noninvasive imaging modalities is that there is no ability to measure pulmonary artery pressure.
- Noninvasive imaging has a critical role in the initial diagnosis and follow-up of patients with pulmonary hypertension.
- Computed tomography and magnetic resonance (MR) imaging can help identify the specific cause of pulmonary hypertension.
- Cardiac MR imaging is increasingly used to assess the impact of pulmonary hypertension treatment on right ventricular function.

INTRODUCTION

Pulmonary hypertension (PH) is a diverse group of entities that affect the pulmonary vasculature. These diseases can secondarily affect the right heart by causing a chronic increase in right heart pressure, or primary heart disease of the left ventricle (LV) can be a secondary cause of PH.[1,2] PH is defined as a mean pulmonary artery pressure (mPAP) of 25 mm Hg or greater, determined from right heart catheterization. Further classification of PH is based on additional hemodynamic parameters, including cardiac output and pulmonary capillary wedge pressure. The most recent update to the classification of the different categories of PH is based on conditions that have similar hemodynamic and pathologic findings and management.[3] Five groups are included in the updated classification from 2013 (**Table 1**): group 1 (pulmonary arterial hypertension [PAH]), group 2 (PH secondary to left heart disease), group 3 (PH secondary to diffuse lung disease or chronic hypoxia), group 4 (chronic thromboembolic PH [CTEPH]), and group 5 (PH caused by unclear or multifactorial mechanisms).

A complete analysis of the clinical presentation, treatment, and all the subtypes of PH and their respective imaging findings is beyond the scope of this article. Imaging often provides the initial evidence that PH may be present and can help to identify the specific cause of PH in many cases. It is important to recognize that PH secondary to left heart disease (group 2) is the most common cause of PH[4] and to be aware of specific radiological findings that can have a critical impact on management, such as CTEPH, pulmonary veno-occlusive disease (PVOD), and pulmonary capillary hemangiomatosis (PCH). Furthermore, cross-sectional imaging is

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Table 1
2013 Classification of PH

Group	Type	Examples
1	PAH	Idiopathic PAH; heritable PAH; drug-induced PAH; PAH associated with CTD, HIV, portal hypertension, CHD, schistosomiasis
1′	PVOD PCH	—
1″	PPHN	—
2	PH 2° to left heart disease	LV systolic dysfunction; LV diastolic dysfunction; valvular disease; cardiomyopathies
3	PH 2° to lung disease and/or hypoxia	COPD, ILD, sleep apnea, altitude, and so forth
4	CTEPH	—
5	Unclear cause or multifactorial	Hematological diseases, systemic diseases, and so forth

Abbreviations: CHD, congenital heart disease; COPD, chronic obstructive pulmonary disease; CTD, connective tissue disease; CTEPH, chronic thromboembolic PH; HIV, human immunodeficiency virus; ILD, interstitial lung disease; PAH, pulmonary arterial hypertension; PCH, pulmonary capillary hemangiomatosis; PPHN, persistent PH of newborn; PVOD, pulmonary veno-occlusive disease.

Adapted from Simonneau G, Gatzoulis MA, Adatia I, et al. Updated clinical classification of pulmonary hypertension. J Am Coll Cardiol 2013;62:D34–41.

used to assess the right ventricle (RV) and measure the effects of PH on RV function. The underlying pathophysiology and the roles of right heart catheterization, pulmonary angiography, and PET in PH are not discussed here. This article highlights the use of computed tomography (CT) and magnetic resonance (MR) imaging in the diagnosis and management of PH.

CHEST RADIOGRAPHY

Chest radiographs can detect the characteristic changes in the pulmonary arteries (PAs) and RV in patients with advanced PH.[5,6] The central PAs are typically dilated (**Figs. 1** and **2**) and the right heart (atrium and ventricle) may be enlarged (see **Fig. 2**). Abnormalities in the lungs may be detected with chest radiography in patients with PAH

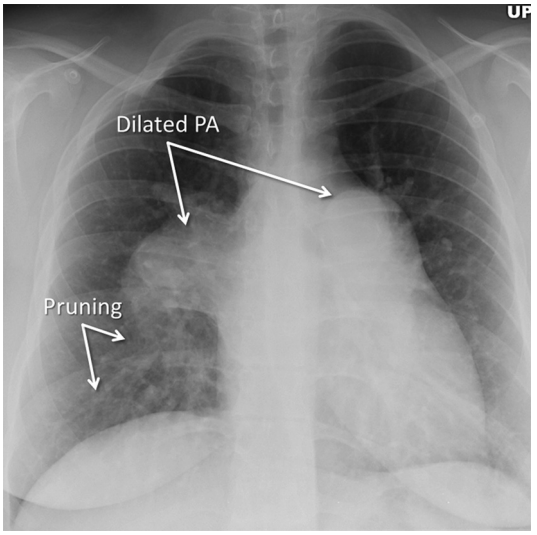

Fig. 1. Frontal chest radiograph of a patient with PAH (group 1) secondary to uncorrected atrial septal defect. The central PAs are severely dilated and there is abrupt tapering of the branch pulmonary arteries resulting in a pruning appearance.

secondary to systemic sclerosis (SSc) or PH secondary to diffuse lung disease, such as emphysema or diffuse lung disease. Chest radiography is appropriate for the initial evaluation of patients with unexplained dyspnea or symptoms that could be attributable to PH. However, chest radiography is insensitive for the identification of patients with mild to moderate PH.[7]

ECHOCARDIOGRAPHY

Transthoracic two-dimensional (2D) Doppler echocardiography (TTE) is the most commonly used noninvasive imaging modality for estimating PA pressures and assessing cardiac function in PH. TTE can be used to screen for PH and may be appropriate in patients with a family history of PAH, congenital heart disease (CHD) and systemic to pulmonary shunts, portal hypertension, or systemic diseases associated with PH.[8,9] Furthermore, serial TTE is routinely performed to monitor the effects of therapy on estimated PA pressures and cardiac function. However, TTE is limited in its ability to comprehensively and accurately evaluate the entire pulmonary vasculature and RV. TTE is particularly useful in determining whether left heart disease or CHD is the cause of PH, but is more restricted in its ability to completely characterize the extent of disease in other causes of PH.

The systolic PA pressure can be estimated with 2D Doppler (**Fig. 3**) using the simplified Bernoulli equation, $\Delta P = 4v^2$,[10] in which v is the peak velocity of the tricuspid regurgitation (TR) jet. The systolic