



Noninvasive assessment of pulmonary hypertension: key insights to maximize chest computed tomography

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Abstract

According to the 2022 European Society of Cardiology (ESC)/European Respiratory Society (ERS) guidelines, a right heart catheterization-derived mean pulmonary arterial pressure (mPAP) > 20 mmHg at rest defines pulmonary hypertension (PH). As PH can ultimately lead to death, multidisciplinary early diagnosis, proper framework, and prompt management are crucial. ESC/ERS PH classification encompasses five groups, each sharing pathogenesis, hemodynamics, and management, i.e., group I, pulmonary arterial hypertension; II, left heart disease; III, lung disease and/or hypoxia; IV, chronic thromboembolic PH; and V, with unclear and/or multifactorial mechanisms. Chest CT is pivotal in PH grouping, comprehensively evaluating the lung, heart, pulmonary vessels, and mediastinum. In this review, we synoptically illustrate the chest CT findings of PH and provide a CT-based logical framework suggesting a PH group hypothesis. Improving knowledge of PH CT features and their combination empowers radiologists to contribute valuably to the PH multidisciplinary clinical discussion.

Keywords Pulmonary hypertension · CT findings · DECT · Noninvasive assessment

Abbreviations

CGGN	Centrilobular ground-glass nodules	HIV	Human immunodeficiency virus
CHD	Congenital heart disease	ILD	Interstitial lung disease
CLD-PH	Chronic lung disease-related PH	IpcPH	Isolated postcapillary PH
CMRI	Cardio magnetic resonance imaging	IPF	Idiopathic pulmonary fibrosis
COPD	Chronic obstructive pulmonary disease	LA	Left atrial
CpcPH	Combined pre- and postcapillary PH	LAM	Lymphangioleiomyomatosis
CT	Computed tomography	LHD	Left heart disease
CTD	Connective tissue disease	LV	Left ventricle
CTEPD	Chronic thromboembolic pulmonary disease	MDP	Methyl diphosphonate
CTEPH	Chronic thrombo-embolic pulmonary hypertension	mPAP	Mean pulmonary arterial pressure
CPFE	Combined pulmonary fibrosis and emphysema	MRI	Magnetic resonance imaging
CPTA	CT pulmonary angiography	NCT	Non-contrast CT
DECTA	Dual-energy CT angiography	PA	Pulmonary artery
DTPA	Diethylenetriamine pentaacetate	PAH	Pulmonary arterial hypertension
ES	Eisenmenger Syndrome	PAP	Pulmonary arterial pressure
ESC	European Society of Cardiology	PAWP	Pulmonary Artery Wedge Pressure
ESR	European Respiratory Society	PH	Pulmonary hypertension
		PH-LHD	PH associated with left heart disease
		PVOD	Pulmonary veno-occlusive disease
		PCH	Pulmonary capillary hemangiomatosis
		PVR	Pulmonary vascular resistance
		RV	Right ventricle
		SAD	Small airway disease
		SSc	Systemic sclerosis
		V/Q	Ventilation/perfusion

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TEE	Transesophageal echocardiography
TTE	Transthoracic echocardiography
WU	Wood units

Introduction

Pulmonary hypertension (PH) refers to a broad group of conditions sharing a pathologic elevation of the mean pulmonary artery pressure (mPAP) [1, 2]. In this review, we aimed to give non-dedicated radiologists basic knowledge on chest computed tomography (CT) role in PH, highlighting its inherent capability of comprehensively evaluating all the chest compartments to provide clinicians with critical diagnostic and prognostic information. Following a brief overview of key PH clinical issues and imaging modalities, we present a per-chest compartment analysis of typical CT findings in PH. We provide reference figures and synoptic tables to help the radiologist valuably contribute to the PH multidisciplinary team clinical discussion.

Definition and classifications

Current guidelines from the European Society of Cardiology (ESC)/European Respiratory Society (ERS) define PH as a right heart catheterization-defined mean mPAP value > 20 mmHg at rest [1]. PH can further be classified into pre-capillary PH, isolated postcapillary PH (IpcPH), and combined pre- and postcapillary PH (CpcPH) according to different combinations of pulmonary arterial wedge pressure (PAWP) and pulmonary vascular resistance (PVR) values (Table 1) [3, 4].

According to the leading presumed cause, physicians clinically classify PH into five groups (Table 2, Fig. 1), sharing pathogenesis, hemodynamics, and management

strategies. Basically, the group I (also named pulmonary arterial hypertension, PAH) includes cases that are idiopathic, heritable, or associated with various conditions, e.g., drugs, connective tissue disease, and congenital heart disease; group II is associated with left heart disease; group III is associated with lung disease and/or hypoxia; group IV mainly includes chronic thromboembolic PH (CTEPH); and group V gather cases with unclear and/or multifactorial mechanisms, e.g., sarcoidosis. Notably, all the groups may include pre- and postcapillary components contributing to mPAP elevation. The ESC/ERS guidelines recommend a patient's multiparametric risk assessment to steer management and stratify the mortality risk [1].

Group I. PAH is a progressive disease characterized by pulmonary artery cell proliferation, ultimately leading to luminal narrowing [5]. It can affect all age groups, with a higher prevalence among older patients with comorbidities. The current incidence approximates six cases per million adults [6]. The major causes are idiopathic, heritable, drug-induced, connective tissue disease (CTD), and congenital heart disease-associated [5]. Among CTDs, systemic sclerosis (SSc) is the leading cause of PAH in European countries [7]. Other possible causes include HIV, schistosomiasis, and portal hypertension. PAH also includes pulmonary veno-occlusive disease/pulmonary capillary hemangiomatosis (PVOD/PCH), which is a rare, potentially life-threatening, and rapidly evolving disease with overt features of venous/capillary involvement [8].

Group II. PH associated with left heart disease (LHD-PH) represents the most common cause of PH [1]. A retrograde transmission of increased left-sided pressures into the pulmonary circulation results in PH [5], pointing out endothelial dysfunction and pulmonary vascular remodeling. Hemodynamically, LHD-PH can present either an IpcPH or CpcPH component. Patients are typically elderly,

Table 1 Hemodynamic definition of pulmonary hypertension according to the 2022 European Society of Cardiology/European Respiratory Society guidelines

Definition	Characteristics	Clinical groups
Pulmonary hypertension (PH)	mPAP > 20 mmHg	All
Pre-capillary PH	mPAP > 20 mmHg PAWP ≤ 15 mmHg PVR ≤ 2 WU	PAH (I); Lung disease (III); CTEPH (IV); Unclear and/or multifactorial mechanism (V)
Isolated postcapillary PH (IpcPH)	mPAP > 20 mmHg PAWP > 15 mmHg PVR ≤ 2WU	Left Heart disease (II); Left heart + lung disease overlap (II + III); Unclear and/or multifactorial mechanism (V)
Combined pre- and postcapillary PH (CpcPH)	mPAP > 20 mmHg PAWP > 15 mmHg PVR > 2WU	Left Heart disease (II); Left heart + lung disease overlap (II + III); Unclear and/or multifactorial mechanism (V)

mPAP mean pulmonary arterial pressure, PAWP pulmonary arterial wedge pressure, PVR pulmonary vascular resistance, WU wood units

Table 2 Hemodynamic definition of pulmonary hypertension according to the 2022 European Society of Cardiology/European Respiratory Society guidelines

Group 1: Pulmonary arterial hypertension (PAH)
1.1 Idiopathic
1.1.1 Non-responders at vasoreactivity testing
1.1.2 Acute responders at vasoreactivity testing
1.2 Heritable
1.3 Associated with drugs and toxins
1.4 Associated with:
1.4.1 Connective tissue disease
1.4.2 HIV infection
1.4.3 Portal hypertension
1.4.4 Congenital heart disease
1.4.5 Schistosomiasis
1.5 PAH with features of venous/capillary (PVOD/PCH/PCH) involvement
1.6 Persistent PH of the newborn
Group 2: PH associated with left heart disease
2.1 Heart failure:
2.1.1 with preserved ejection fraction
2.1.2 with reduced or mildly reduced ejection fraction
2.2 Valvular heart disease
2.3 Congenital/acquired cardiovascular conditions leading to postcapillary PH
Group 3: PH associated with lung disease and/or hypoxia
3.1 Obstructive lung disease or emphysema
3.2 Restrictive lung disease
3.3 Lung disease with mixed restrictive/obstructive pattern
3.4 Hypoventilation syndromes
3.5 Hypoxia without lung disease (e.g., high altitude)
3.6 Developmental lung disorders
Group 4: PH associated with pulmonary artery obstructions
4.1 Chronic Thromboembolic Pulmonary Hypertension (CTEPH)
4.2 Other pulmonary artery obstruction
Group 5: PH with unclear and/or multifactorial mechanism
5.1 Hematological disorder
5.2 Systemic disorder
5.3 Metabolic disorder
5.4 Chronic renal failure with or without hemodialysis
5.5 Pulmonary tumor thrombotic microangiopathy
5.6 Fibrosing mediastinitis

PH pulmonary hypertension, *HIV* human immunodeficiency virus

often presenting with metabolic syndrome, structural left heart disease, or left atrial (LA) dilatation [6].

Group III. Chronic lung disease-related PH (CLD-PH) represents the second most common cause of PH. The primary contributors to CLD-PH include emphysema, restrictive lung diseases, hypoventilation syndromes, and lymphangioleiomyomatosis (LAM). In patients with chronic lung disease, structural modification of lung parenchyma may lead to vascular changes and, ultimately, to PH. Continuous or intermittent hypoxia is common in chronic lung disease and may result in pulmonary vessel constriction,

release of growth factors, and subsequent intimal hyperplasia conditioning an increased vascular tone [9].

Group IV. This group includes PH conditions caused by obstructive pulmonary artery (PA) lesions. CTEPH is the most common cause. Non-resolved thrombi result in organization and fibrosis, thus narrowing vessels and impairing blood flow [6]. Group IV PH may rarely be due to PA sarcoma or large-vessel pulmonary vasculitis.

Group V. This group encompasses a wide range of disorders. The etiology is multifactorial, involving increased pre- and postcapillary pressure and direct effects on pulmonary

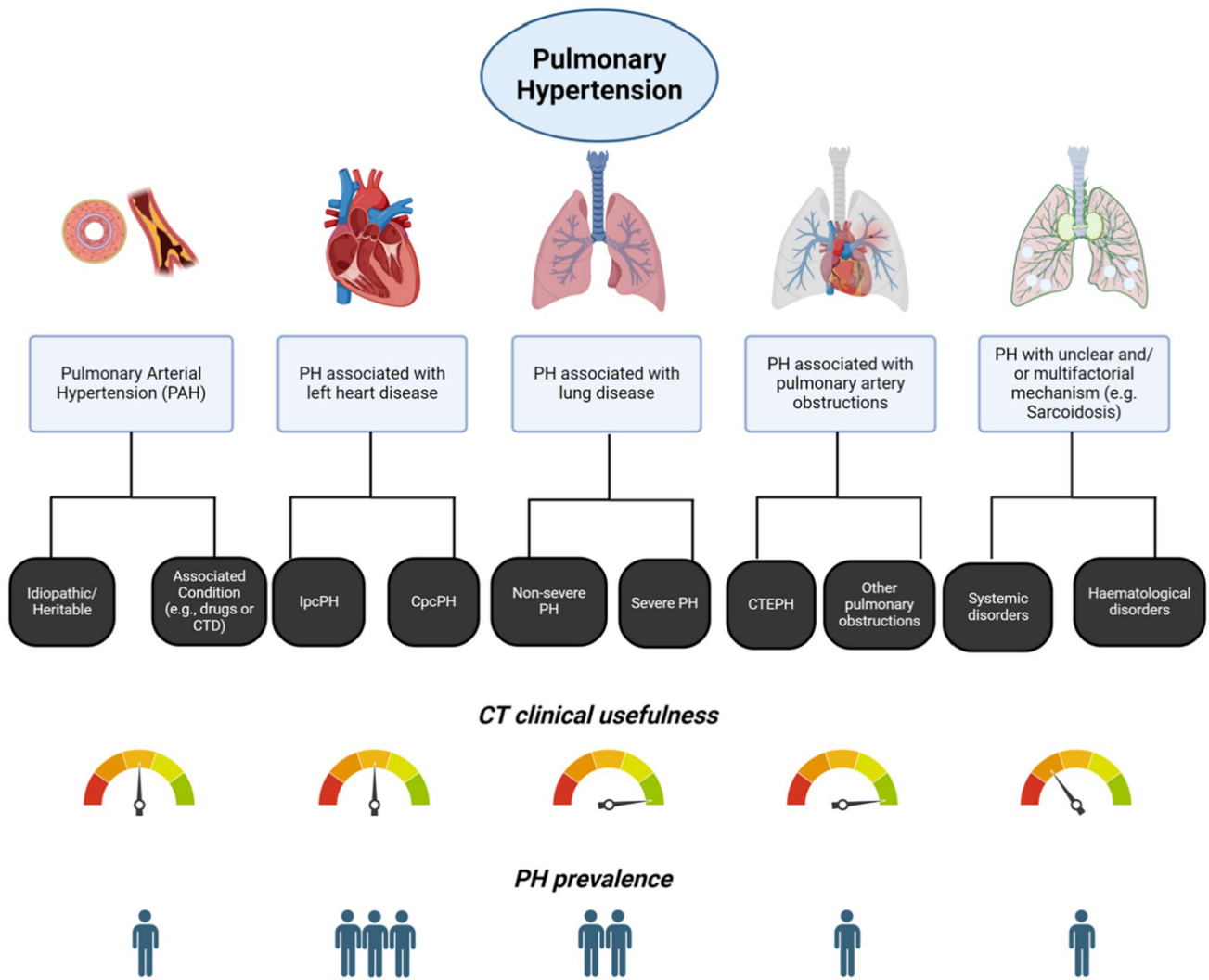


Fig. 1 Clinical pulmonary hypertension (PH) classification according to the 2022 European Society of Cardiology/European Respiratory Society guidelines. For each PH group, a visual speedometer indicates the potential CT clinical utility level according to evidence from

the literature, with green being the most useful. The figure was created with BioRender.com. *CTD* connective tissue disease; *CT* computed tomography; *PH* pulmonary hypertension; *IpcPH* isolated post-capillary PH; *CpcPH* combined pre- and postcapillary PH

vasculature [1]. Example conditions include myeloproliferative disorders, chronic renal failure, sarcoidosis, and thyroid disease [6].

Overview of imaging modalities

Even though the diagnosis of PH relies on right heart catheterization, noninvasive imaging plays a pivotal role in the PH assessment, aiming to evaluate the heart, lung parenchyma, airways, pulmonary vasculature, and mediastinum, as well as suggest critical diagnostic and prognostic information. Table 3 provides a synoptic overview of the strengths and limitations of the main imaging modalities.

Current guidelines recommend transthoracic echocardiography (TTE) as the first-line, noninvasive imaging investigation when suspecting PH [1]. Indeed, regardless of the underlying cause, PH leads to echocardiography-detectable right ventricle (RV) pressure overload and dysfunction. Notably, echocardiography allows defining the probability of PH (low, intermediate, or high), thus prompting further investigation, including cardiac catheterization. Furthermore, TTE provides a reproducible assessment of potential left heart causes of PH, including valvular disease, left ventricle (LV) systolic or diastolic dysfunction, congenital heart disease (CHD), and intra-cardiac shunts [10]. Besides conventional TTE, speckle-tracking strain imaging and 3D echocardiography are gaining ground in monitoring right heart structure and function [11]. The assessment of RV

Table 3 Overview of imaging modalities in pulmonary hypertension

	Chest Radiography	Echocardiography	Chest NCT	Chest CTPA	Chest DECTA	CMRI	V/Q	Pulmonary angiography
PH detection	+	+++	+	++	++	+/-	-	-
Lung	+	-	+++	+++	+++	-	-	-
Heart	+/-	+++	+	++	++	++	-	-
Pulmonary vessels	+/-	+	+	++	++	++	+++	+++
Mediastinum	-	-	+	+++	+++	+++	-	-
PH etiology	+	++	+	++	++	++	++	+
Strengths	Availability; 1st line imaging	1st line imaging; define the probability of PH (low, intermediate, or high), thus prompting further investigation	Lung parenchyma evaluation	Comprehensive evaluation of all the chest compartments; PH etiology	Lung “perfusion” map	No radiation exposure; evaluation of right ventricle size and function	Pivotal in ruling out or detecting CTEPH	Endovascular treatment planning
Limitations	Unhelpful in mild PH and etiology assessment	Limited role in the assessment of etiology	Limited role in the assessment of heart and pulmonary vessels	Limited hemodynamics assessment; limited evaluation of distal pulmonary arteries	Needs more robust validation	Limited role in the evaluation of lung and vessels	Limited in case of patients with comorbidities; no comprehensive evaluation	Invasiveness

PH pulmonary hypertension, NCT non-contrast computed tomography, CTPA CT pulmonary angiography, DECTA dual-energy CT angiography, CMRI cardiac magnetic resonance imaging, V/Q ventilation/perfusion scan, CTEPH chronic thromboembolic pulmonary hypertension

systolic pressure is obtained with the tricuspid regurgitation jet velocity and size/collapsibility of the inferior vena cava [12]. In selected patients, transesophageal echocardiography (TEE) may further ambiguous findings, e.g., mitral valve disease, ultimately refining the patient treatment [10].

Chest radiography (CXR) is the first-line imaging modality in patients with PH, stating that presenting symptoms, such as dyspnea, are usually nonspecific. Albeit unhelpful in mild PH, CXR demonstrates high sensitivity and specificity in detecting moderate to severe PH [13]. Indeed, CXR findings can suggest PH, e.g., central PAs with or without peripheral pruning, and right heart chambers enlargement [14], while certain abnormalities can elicit the suspicion of specific PH groups, e.g., left heart chambers' enlargement, gross valve calcification, and pulmonary venous congestion suggest group II; interstitial fibrosis, bronchiectasis changes or extensive emphysema suggest group III; the Hampton's hump, i.e., wedge-shaped, pleural-based opacification due to pulmonary infarction, and the Westermark sign, i.e., focal peripheral hyperlucency due to oligemia resulting in a collapsed appearance of vessels distal to the occlusion, suggest group IV [15].

Cardiac Magnetic Resonance Imaging (CMRI) is the gold standard in evaluating RV size and function, allowing highly reproducible tissue characterization and distinguishing ischemic perfusion from fibrotic processes [16]. Furthermore, it provides high-resolution 3D volumetric visualization of the RH, allowing identification of congenital cardiac abnormalities and quantification of blood flow, and helps recognize regurgitant valvular flow and the relationship between RV and LV [17].

Radionuclide ventilation–perfusion scanning (V/Q) is pivotal in ruling out or detecting CTEPH in patients with suspected or newly diagnosed PH, stating > 90% sensitivity and specificity [1, 18]. V/Q is performed with gas (Xe133), aerosolized droplets (Technetium 99 M, DTPA/MDP/Sulphur colloid) or carbon particles (Technegas) for the ventilation phase, and Technetium labeled macro-aggregated albumin for the perfusion phase [19]. A segmental mismatched perfusion defect with preserved ventilation to the corresponding regions at V/Q in PH patients defines CTEPH. Nevertheless, V/Q can display nonmatched perfusion defects in up to 10% of patients with PAH (including PVOD/PCH) [20]. Therefore, a high probability V/Q in patients with PH does not always correspond to CTEPH.

The role of chest CT in the noninvasive work-up

Non-contrast CT (NCT)

Non-contrast CT (NCT) can provide valuable information in patients with unexplained dyspnea and suspected PH, as well as in patients with confirmed PH to help diagnose group I, III, or V PH. Concerning group I PH, NCT can detect indirect signs of PH resulting from pulmonary microvasculopathy, such as centrilobular micronodules, peripheral neo-vascularization, and areas of ground-glass attenuation [21]. NCT may also suggest PVOD/PCH when distinguishing imaging features are present, i.e., centrilobular ground-glass opacities, interlobular septa thickening, and mediastinal lymphadenopathy, particularly when these findings coexist [22, 23]. Moreover, NCT can detect lung fibrosis or emphysema, thus pointing toward group III PH, as well as signs of sarcoidosis, pulmonary Langerhans cell histiocytosis, and neurofibromatosis, raising the suspicion of group V PH.

Pulmonary CT angiography (CTPA)

The key benefit of pulmonary CT angiography (CTPA) is helping diagnose CTEPH via several direct and indirect signs, including thrombus within the vascular walls, bands in the PAs, filling defects, mosaic perfusion, and hypertrophic bronchial arteries. CTPA can also depict cardiovascular abnormalities such as intra-cardiac shunts, abnormal pulmonary venous return, patent ductus arteriosus, and pulmonary arteriovenous malformation [1]. Additionally, CTPA can suggest PH when revealing PA dilatation (> 30 mm), a PA-to-aorta ratio higher than 0.9, enlarged right heart chambers, RV wall thickness ≥ 6 mm, RV/LV ratio higher than 1, and septal deviation $\geq 140^\circ$ [24].

Dual-energy CT angiography (DECTA)

Dual-energy CT angiography (DECTA) enables displaying pulmonary "perfusion" maps derived from the iodine content within the pulmonary vasculature, enabling noninvasive analysis of lung microcirculation [25]. DECTA demonstrated excellent agreement with V/Q in diagnosing CTEPH [26], with higher accuracy, reduced radiation exposure, and concurrent lung and heart morphology assessment [27]. DECTA proved high sensitivity in predicting the disease level of CTEPH, distinguishing between proximal and distal disease, thereby providing valuable road-mapping for pulmonary endarterectomy aimed at improving hemodynamics [28]. Notably, DECTA-derived pulmonary perfusion patterns enable differentiating acute pulmonary embolism and

CTEPH [20, 29], thus helping in CTEPH early diagnosis. Moreover, DECTA can detect perfusion changes in a small proportion of group I PH, including idiopathic PAH and PVOD/PCH [30]. The perfusion patterns of PAH considerably differ from peripheral CTEPH, being almost exclusively patchy and homogeneous in the former and patchy or pulmonary embolism type in the latter [31, 32].

Per-chest compartment analysis of CT findings in PH

We hereafter resume typical chest CT findings in PH according to the most involved chest compartment, i.e., lung, heart, pulmonary vessels, and mediastinum. Table 4 resumes some of the most frequent clinical conditions associated with PH and the corresponding CT imaging findings.

Lung

Several CLDs can cause PH, including those of group III PH (interstitial lung diseases, chronic obstructive pulmonary disease [COPD]), and combined pulmonary fibrosis and

emphysema [CPFE]) and group V PH (sarcoidosis, pulmonary Langerhans' cell histiocytosis, and neurofibromatosis type 1) [33]. Chest CT is paramount in detecting PH-related lung findings, including signs of emphysema and/or airway disease, interstitial lung disease and fibrosis, centrilobular nodules, interlobular septal thickening, subpleural peripheral opacities, and mosaic attenuation pattern [24, 34].

Emphysema and airway disease

Permanent enlargement of airspaces beyond the terminal bronchioles and destruction of alveolar walls define lung emphysema (Fig. 2a) [22, 35]. Along with emphysema, chest CT can display signs of airway disease, including bronchial wall thickening of the central airways and distal airway changes such as tree-in-bud opacities, centrilobular micronodules, air trapping, bronchiolectasis, and oligemia. Different combinations of such findings mirror conditions that lead to functional irreversible airflow limitation in the framework of COPD [36]. PH frequently complicates COPD, with a prevalence approximating 40% of patients and much higher in end-stage disease, with the majority presenting with mild-to-moderate PH [37]. Hence, assessing

Table 4 Some of the most frequent clinical PH-associated conditions, with corresponding group and typical CT imaging findings

Clinical condition	PH group	CT findings
Idiopathic PAH	I	Lung nodules, RV dilatation, deviated interventricular septum, lung mosaic attenuation pattern
CTD-related PAH	I	Lung ill-defined centrilobular ground-glass micronodules; esophageal dilatation; possible mild interstitial abnormalities
Pulmonary arteriovenous malformation	I	Rounded pulmonary opacities, feeding artery, and draining vein
Porto-pulmonary hypertension	I	Esophageal varices, nodular and shrunken liver, abdominal ascites, and splenomegaly
Pulmonary veno-occlusive disease (PVOD/PCH)	I	Centrilobular ground-glass nodules, interlobular septal thickening, and mediastinal lymph-node enlargement
Atrial septal defect/ventricular septal defect	II	Left-to-right intra-cardiac contrast shunting
Left heart disease	II	Left atrium enlargement; lung signs of pulmonary edema (e.g., areas of increased lung attenuation, smooth septal thickening, bronchovascular bundle thickening, pleural effusion)
COPD	III	Centrilobular/paraseptal/panlobular emphysema with upper prevalence, lung overinflation, signs of airway disease
ILD with fibrosis	III	Volume loss, traction bronchiectasis, reticulation, honeycombing
CTEPH	IV	Endoluminal filling defects, webs, mural thrombi, narrowing of peripheral arteries, mosaic attenuation pattern, hypertrophic bronchial artery
Pulmonary artery sarcoma	IV	Endoluminal filling defects and expansion into the vessel lumen with an acute angle
Sarcoidosis	V	Bilateral hilar/mediastinal lymph-node enlargement, with/without eggshell or icing sugar calcification, perilymphatic lung abnormalities
Pulmonary Langerhans cell histiocytosis	V	Diffuse cystic lung disease with a predominant upper and middle distribution, nodules ranging in number from a few to innumerable, and possibly cavitating
Fibrosing mediastinitis	V	Mediastinal infiltrative mass encasing/invading adjacent structures

CTD connective tissue disease, COPD chronic obstructive pulmonary disease, CTEPH chronic thromboembolic pulmonary hypertension, ILD interstitial lung disease, PAH pulmonary arterial hypertension

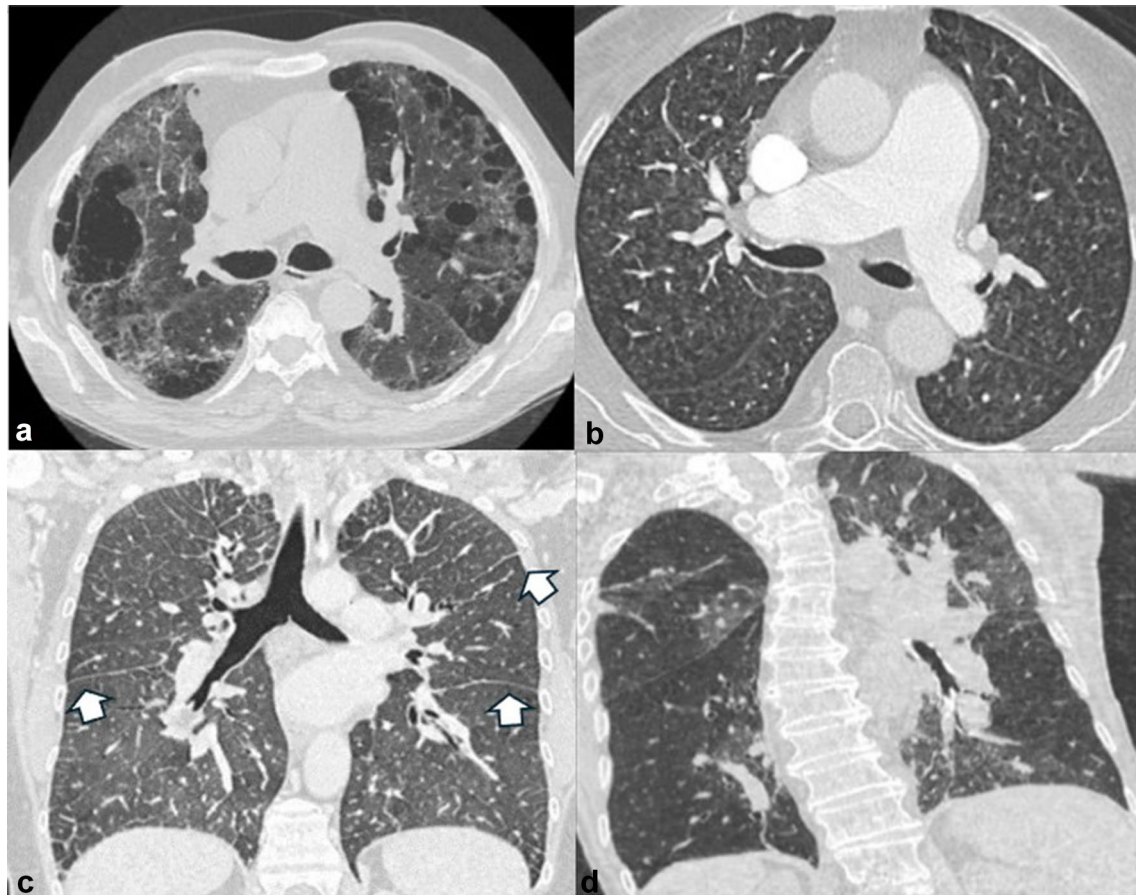


Fig. 2 Lung CT findings in pulmonary hypertension. **a** Emphysema and fibrosis. **b** Centrilobular ground-glass nodules. **c** Smooth interlobular septal thickening (arrow). **d** Subpleural peripheral scars

CT signs of emphysema and airway disease in patients with suspected/established PH is crucial. It is essential to emphasize that, since COPD is a common condition in the general population, in patients with coexisting PH, a comprehensive clinical, imaging, and functional assessment is mandatory to ascertain if PH results from COPD or it is due to a coincident non-group III etiology [38].

Interstitial lung disease and fibrosis

Interstitial lung disease (ILD) is an umbrella term indicating a broad, heterogeneous group of lung conditions sharing restrictive physiology and impaired gas exchange, albeit with highly variable causes and pathological, clinical, and imaging features. Connective tissue disease-related interstitial lung disease (CTD-ILD), idiopathic pulmonary fibrosis (IPF), chronic hypersensitivity pneumonitis, pulmonary Langerhans cell histiocytosis, and sarcoidosis are the ILDs that most commonly complicate with PH [39]. Other conditions include combined pulmonary

fibrosis and emphysema (CPFE) and LAM [1]. Notably, all the above-mentioned ILD-related PH conditions belong to group III except sarcoidosis and pulmonary Langerhans cell histiocytosis, which fall under group V due to their multifactorial nature. They all share general pathophysiology mechanisms, including alveolar hypoxia, reactive vasoconstriction, and various parenchymal and vascular remodeling processes, ultimately leading to PH (Fig. 2a) [39].

From a practical viewpoint, CT detection and quantification of ILD extent is essential, irrespective of the PH group. Indeed, mild parenchymal lung changes (i.e., fibrosis and/or emphysema) in patients with idiopathic PAH are associated with worse survival [40], probably identifying a distinct PH phenotype of idiopathic PAH closer to group II PH [41]. Moreover, among patients with SSc-related PH, those with extensive ILD (i.e., CT extent > 30%) are associated with worse survival than those with PAH and low DLCO but no or limited ILD [42, 43].

Centrilobular ground-glass nodules

Centrilobular ground-glass nodules (CGGN) represent an easily overlooked but typical pulmonary finding in patients with PH (Fig. 2b). Ill-defined CGGN are common in PAH, presumably reflecting cholesterol granulomas, hemorrhage, or plexogenic arterial lesions. CGGN can also be present in patients with long-standing left-to-right heart shunt and, most importantly, in various ILD, including subacute hypersensitivity pneumonitis and smoking-related respiratory bronchiolitis [44]. Notably, a correct interpretation of CGGN is crucial. Indeed, when CGGN presents as isolated in the framework of idiopathic PAH, it is not associated with lower DLCO nor worse survival [45]. On the contrary, the situation is much different when CGGN is part of an ILD or reflects PVOD/PCH, carrying the risk of aggravated prognosis.

Interlobular septal thickening

The morphology of interlobular septal thickening can help group PH. Indeed, smooth, irregular, and nodular thickening points toward group II PH (i.e., LHD, Fig. 2c), group III PH (lung fibrosis), or group V PH (sarcoidosis, fibrosing mediastinitis), respectively. In the absence of signs of LHD or ILD, interlobular septal thickening may mirror the post-capillary congestion typical of patients with PVOD/PCH, prompting the search for accoupling CGGN and mediastinal lymph-node enlargement.

Subpleural peripheral opacities

Nonspecific, subpleural peripheral bands or consolidation areas resembling parenchymal scars can frequently be encountered in CTEPH (Fig. 2d) [46], thus prompting careful evaluation of PAs (see further in the text).

Mosaic attenuation pattern

The mosaic attenuation pattern is present in nearly 20% of patients with PH [47]. While possible in all the PH clinical groups, its distribution tends to be heterogeneous or patchy, perivascular in idiopathic PAH, and segmental and well defined in CTEPH [48].

Heart

Technological advances in chest CT acquisition, mainly related to high temporal resolution, make first-hand heart and coronary artery imaging feasible. Indeed, various cardiac findings are detectable on non-ECG-gated CT scans, including cardiac shunts, coronary and valvular calcifications, and chamber enlargement [49, 50].

Congenital heart diseases

PH can develop in patients with CHDs due to pulmonary over-circulation (left-to-right shunt, from systemic to pulmonary circulation), falling under group I PH. Several CHD characterized by chronic and untreated pulmonary over-circulation can ultimately lead to Eisenmenger Syndrome (ES), i.e., the progressive development of pulmonary vasculopathy (increased PVR) with reversal of shunt flow (from right-to-left or bidirectional) [51]. Examples include atrial septal defects, partial anomalous pulmonary venous return, ventricular septal defects, and patent ductus arteriosus. Notably, patients with large post-tricuspid defects and complex anatomical lesions and/or those associated with trisomy 21 are at higher risk of early PH development [52]. Less frequently, PH may complicate obstructive left heart lesions, thus falling under group II PH [1]. Patients who undergo radical correction at a later age, or partially corrective intervention only, may complicate with PH even when the CHD is associated with small defects/shunts [1].

CT is crucial in investigating PAH associated with congenital heart disease, providing detailed information on cardiac chambers, great arteries, pulmonary vasculature, parenchyma, and mediastinal structures in a single acquisition with high spatial resolution (Fig. 3a). This is particularly beneficial when acoustic windows are limited (constraining echocardiography), in case of lung disease, or when patients cannot undergo MRI. Communication between chambers can be visualized, and shunt direction can be inferred from the contrast flow. CTPA is the preferred noninvasive method for evaluating proximal and distal PAs, identifying PA dilation and in-situ thrombus formation due to sluggish blood flow. CT excels in assessing extracardiac features, notably native or surgically created systemic-to-pulmonary shunts [53].

Left atrium enlargement

The close correlation between LA enlargement and group II PH is well established. Previous studies found an excellent diagnostic accuracy of CTPA-derived LA diameter and area, thus distinguishing patients with group II PH from those with non-group II PH [17, 54–56]. The LA diameter should be the largest anterior–posterior diameter on the slice in the middle 50% of the LA craniocaudal extension (Fig. 3b) [57]. LA area can be calculated through manual or automatic segmentation [57].

Coronary artery and valvular calcification

LHD causes backpressure and increases the hydrostatic pressure in the pulmonary veins, eventually resulting in increased pressure in PAs [58].

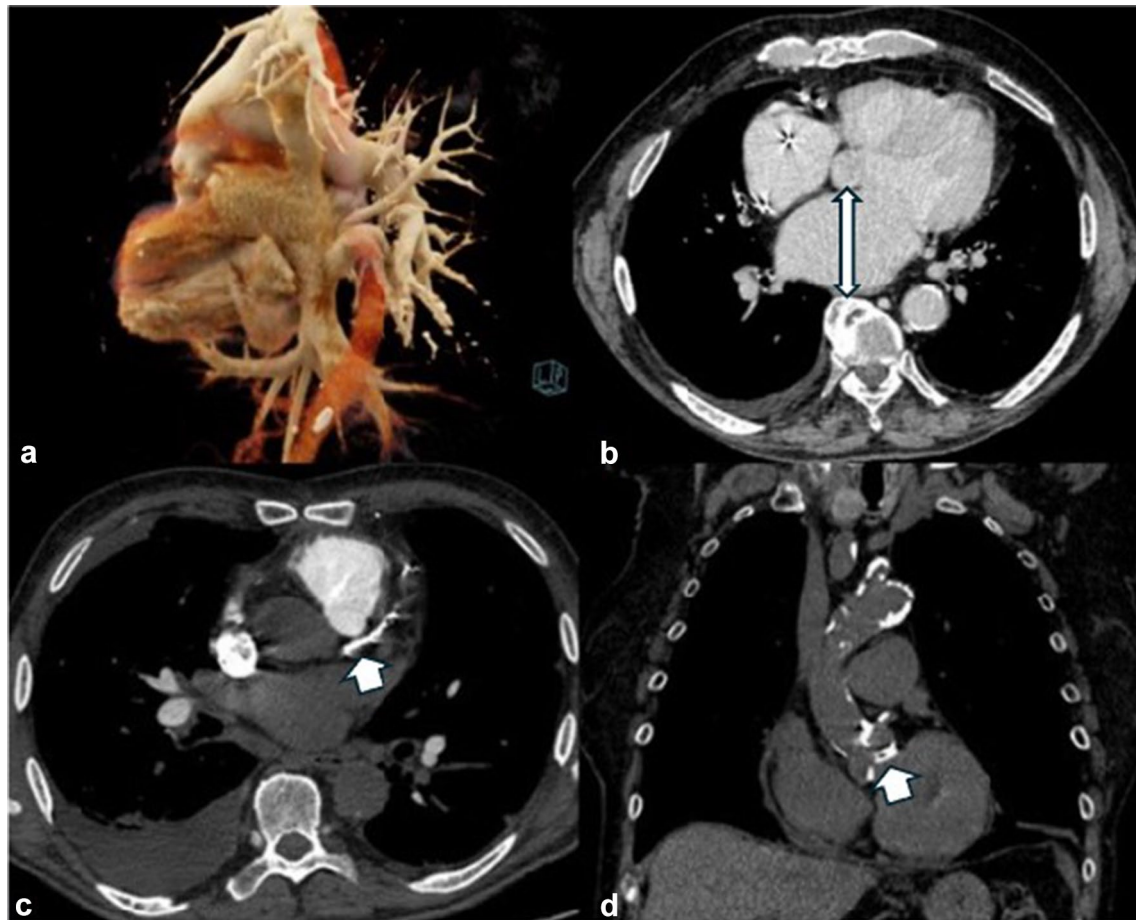


Fig. 3 Heart CT findings in pulmonary hypertension. **a** 3D volume rendering image of the heart and pulmonary vessels showing adulthood-discovered congenital malformation with inferior vena cava draining into the left atrium (LA). **b** LA enlargement. How to meas-

ure the LA anteroposterior diameter is reported (double-sided arrow). **c** Left coronary artery calcification (arrow). **d.** Aortic valve calcification (arrow)

Aortic or mitral valvular thickening and calcification can indicate valvular stenosis, the calcification degree correlating with the valvular stenosis severity [59]. Valvular calcification can be identified as high-attenuation material in the aortic valve and mitral region. Concerning the aortic valve, care should be taken to differentiate valve calcification from calcification in the aortic root, mitral annulus, or coronary (Fig. 3c) [60].

Coronary artery calcification can be identified as high-attenuation material in the path of a coronary artery (Fig. 3d). For non-gated thoracic CT in routine clinical practice, a simple visual ordinal score performed on a whole-patient basis has been recommended [60].

Pulmonary vessels

Identifying CT signs about pulmonary vessels helps recognize group IV PH, mostly CTEPH (Fig. 4). Other causes of pulmonary vascular obstruction, such as malignant tumors

(sarcomas, renal carcinoma, uterine carcinoma, germ-cell tumors of the testis), arteritis without connective tissue disease, and congenital pulmonary arterial stenosis, are far less common and described elsewhere [1].

Signs of chronic thromboembolic pulmonary disease

Chronic thromboembolic pulmonary disease (CTEPD) refers to symptomatic patients due to post-thromboembolic fibrotic obstructions within the PA, with or without PH [1]. CTEPD patients typically exhibit mismatched perfusion defects on V/Q and signs of chronic, organized, fibrotic clots on CTPA or angiography after at least three months of therapeutic anticoagulation, including ring-like stenoses, webs/slits, and chronic total occlusions (pouch lesions or tapered lesions) [1]. CTEPH is the preferred term for patients with CTEPD complicating with PH. In 0.6–4.4% of acute PE, abnormal, persistent residual thromboembolic occlusion, combined with a variable degree of microscopic vasculopathy, leads to

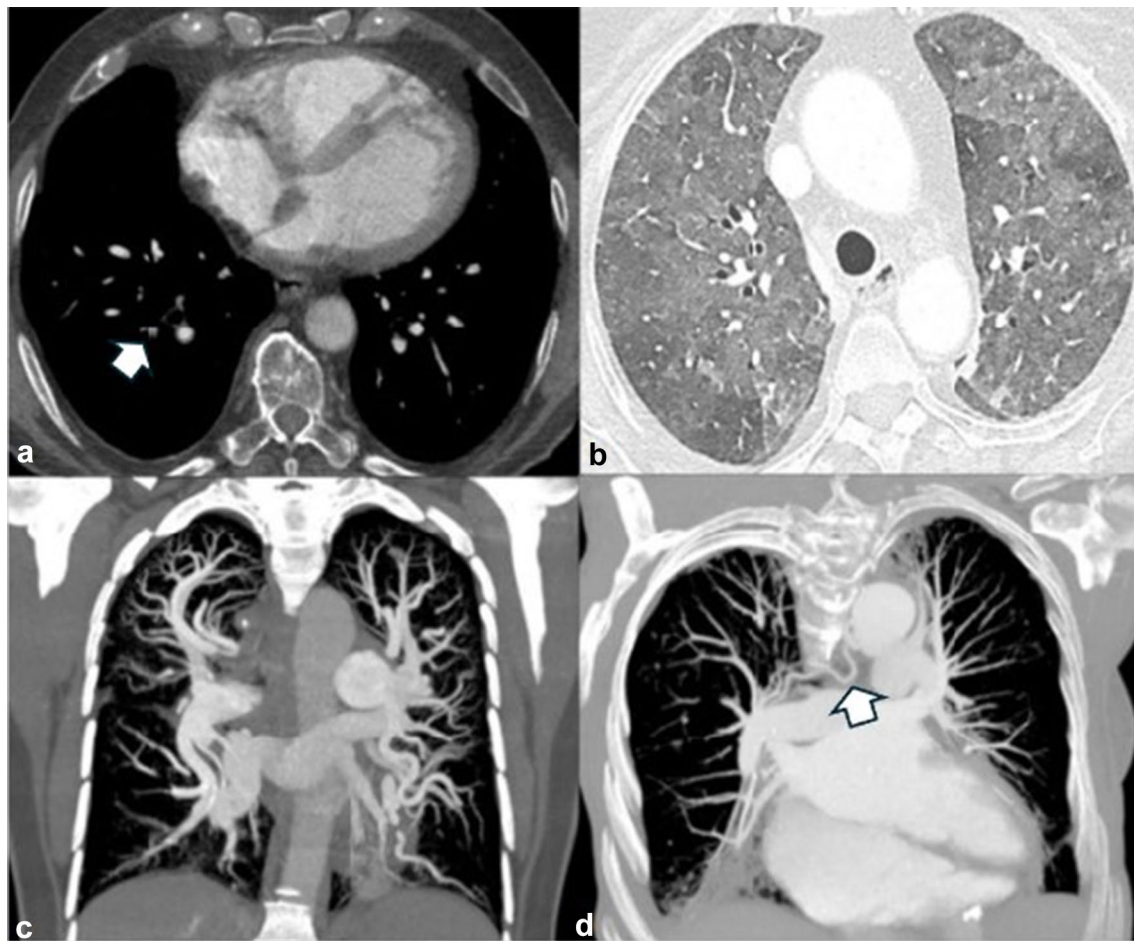


Fig. 4 Pulmonary vessel CT findings in pulmonary hypertension. **a** Chronic, non-occlusive thrombi in pulmonary arteries (arrow). **b** Mosaic attenuation pattern reflecting oligemia. **c**. Pulmonary artery branch abruption, abnormal tapering, and corkscrewing. **d** Right

hypertrophic bronchial artery (red arrow). **c, d** Thin-slab MIP coronal reformatted images show pulmonary artery branch abruption, abnormal tapering, and corkscrewing (c), and right hypertrophic bronchial artery (arrow in d)

CTEPH [61, 62]. If untreated, CTEPH can ultimately evolve into right heart failure [21].

Imaging modalities to assess CTEPH include echocardiography, V/Q, CTPA, and right heart catheterization with pulmonary angiography [63]. Typical CTPA findings include signs of non-occlusive and occlusive chronic thrombi [64]. Non-occlusive thrombi present as linear filling defects, forming bands across the PAs and creating webs; alternatively, they may appear as eccentric thrombi with an obtuse vessel wall margin (Fig. 4a). Occlusive chronic thrombi occlude the vessel lumen, causing vascular constriction (unlike acute thrombi, which enlarge the vessel). PA branch abruption, abnormal tapering, and corkscrewing are other frequent CTEPH findings (Fig. 4b) [64].

Mosaic attenuation CT pattern is a crucial, indirect sign of CTEPH [65]. Vessel narrowing results in hypoperfused lung areas, appearing as well-defined abnormally hypodense regions alternating with abnormally hyperdense areas, with patchy distribution, reflecting vascular perfusion differences

(Fig. 4c). Indeed, patent vessels resulting in increased blood flow supply the hyperdense areas, while the hypodense areas are located distal to vascular narrowing/occlusion. Small airway disease (SAD) is another cause of mosaic attenuation CT pattern due to hypoxic vasoconstriction. While large airway wall thickening in the hypodense areas is a frequent SAD-associated finding, patients with CTEPH typically exhibit peripheral scarring resulting from previous infarctions, thus helping differentiate the two conditions [64].

Collateral circulation develops in CTEPH, likely due to increased vascular resistance and blood flow blockages. Consequently, enlarged, tortuous bronchial artery collaterals running through the mediastinum into the hila are frequent findings, particularly in areas affected by obstruction (Fig. 4d). Although these collateral vessels assist in sustaining perfusion to lung tissue beyond thrombotic blockages, the exposure of the pulmonary circulation to the elevated pressure values of the systemic circulation can induce vascular remodeling and, in turn, arteriopathy [66].

Right-ventricular hypertrophy is frequently encountered in long-standing PH. Notably, right atrial dilatation coexists and is detectable at CT in many patients with CTEPH due to a combination of elevated right heart pressures and right ventricular dilatation conditioning functional tricuspid regurgitation [64].

Mediastinum

Along with pulmonary, cardiac, and vascular signs, several mediastinal findings can help diagnose and group PH. We focus on three abnormalities we frequently encounter when dealing with PH, which are lymph-node enlargement, esophageal dilatation, and signs of portal hypertension.

Lymph-node enlargement

Conventionally, a short-axis cut-off of 10 mm defines an enlarged lymph node (LN) [67]. Enlarged mediastinal LNs are a frequent CT finding, lacking specificity. It can be encountered in many different PH-related pathologies

pertaining to all five clinical groups. Indeed, mediastinal LN enlargement is common in group II PH and has been reported in 45% of patients with CTEPH, possibly due to right-sided failure and increased lymphatic flow [68]. Nevertheless, a couple of conditions worth mentioning, including sarcoidosis and PVOD/PCH.

Symmetric LN enlargement at stations 4R, 7, 11L, and 11R in sarcoidosis defines the classical Garland's triad (Fig. 5a, b). LNs typically present focal, *eggshell*, or *icing sugar* calcifications [69]. It has been supposed that enlarged LNs can determine pulmonary vascular compression or extrinsic stenoses, thus restricting blood flow and, ultimately, causing PH [70]. It must be pointed out that the pathophysiology of sarcoidosis-associated PH is multifactorial, which is why it pertains to group V PH. Indeed, several other mechanisms have been hypothesized, including advanced lung parenchyma involvement, cardiac sarcoidosis-inducing systolic/diastolic LV dysfunction, PVOD/PCH, higher risk of thromboembolic disease, and sarcoidosis-triggered vascular granulomatous inflammation [71].

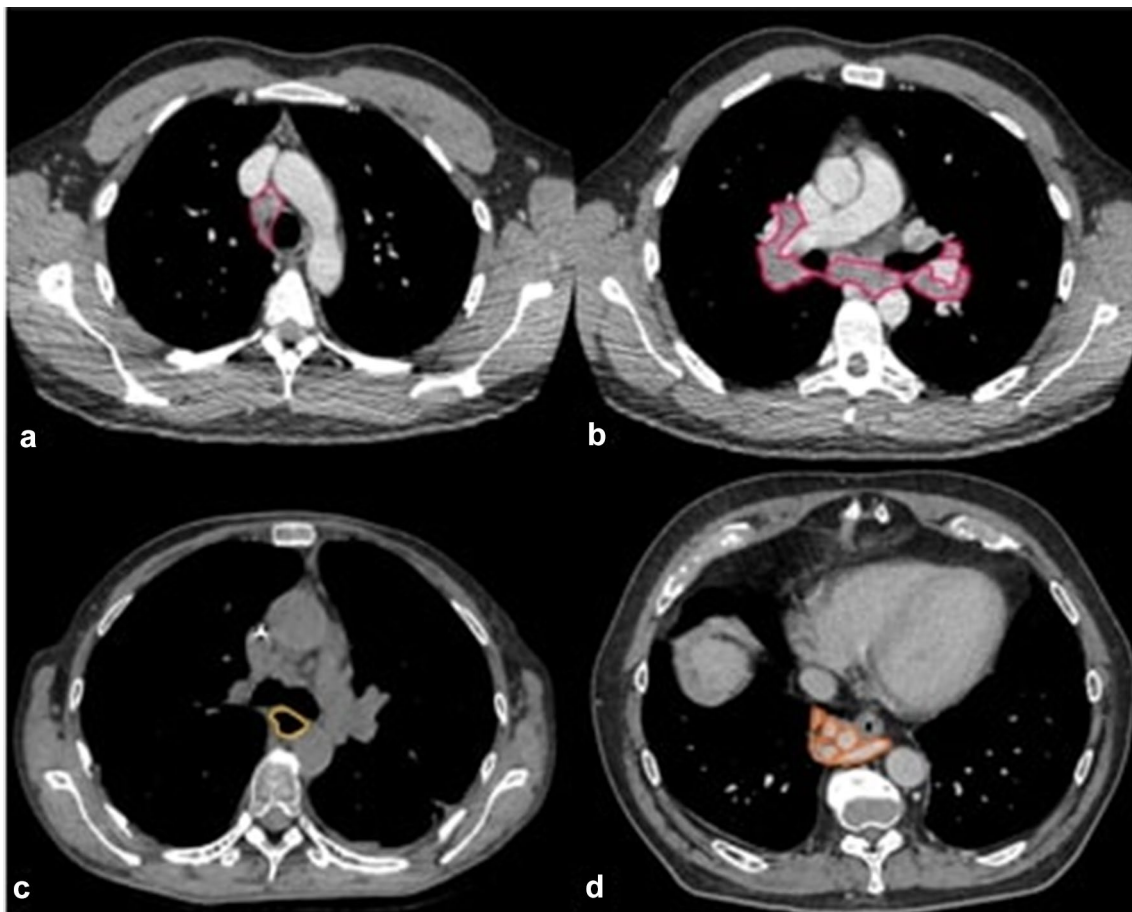


Fig. 5 Mediastinal CT findings in pulmonary hypertension. **a, b** Garland's sarcoidosis triad includes right paratracheal (**a**) and symmetric, bilateral hilar lymph-node enlargement (**b**). **c** Esophageal dilatation in systemic sclerosis. **d** Dilated paraesophageal veins in portal hypertension

In PVOD/PCH, mediastinal LN enlargement is present in up to 84% of cases [72]. It probably derives from venous congestion, veno-lymphatic shunts, and angiogenetic factors typically encountered in this condition [73, 74]. Albeit such enlarged LNs have no specific station location, when associated with centrilobular ground-glass opacities and interlobular septa smooth thickening, they are highly suggestive of PVOD/PCH [75].

Esophageal dilatation

An axial diameter cut-off value > 10 mm at the carina level or a maximum diameter > 15 mm defines esophageal dilatation [76]. The dilatation of the lower two-thirds of the esophagus, particularly below the level of the aortic arch, suggests systemic sclerosis (SSc) (Fig. 5c) [77]. In patients with SSc, esophageal smooth muscle becomes atrophic, with replacing fibrous tissue leading to severe dysmotility, dilatation, and absent peristalsis [78]. Esophageal abnormalities have been implicated in SSc-related ILD development. In patients with known/suspected PH, the association of esophageal dilatation and ILD should raise the suspicion of SSc-related PH, thus prompting further tests [79].

Signs of portal hypertension

Persistent portal hypertension leads to the development of portosystemic collaterals. CT displays varices as distinct, round, tubular, or winding structures exhibiting smooth, homogeneous attenuation and enhancing to a similar degree as portal and mesenteric veins after contrast injection [80, 81]. Mediastinal varices include (i) esophageal veins, presenting with nodular thickening of the esophageal wall and enhancing nodular intraluminal protrusions with scalloped borders near the esophageal hiatus (Fig. 5d); (ii) paraesophageal veins, manifesting as dilated collateral vessels encircling the esophagus and descending thoracic aorta; (iii) pericardiophrenic veins, which are dilated veins in the cardiophrenic angle, often associated with cirrhosis due to inferior vena cava membranous obstruction [81]. The presence of these mediastinal signs, along with other indicators such as other dilated collateral vessels, nodular and shrunken liver, ascites, and splenomegaly, supports the suspicion of portal hypertension-associated PAH, also known as portopulmonary hypertension [82].

Conclusion

PH is a complex condition encompassing a broad range of different clinical scenarios, thus requiring multidisciplinary interaction to prompt adequate diagnosis, risk stratification, and management. The crucial and constantly evolving role of

chest CT in the noninvasive work-up of patients with known or suspected PH mandates adequate knowledge of typical and less frequent CT findings to adequately interpret them and valuably contribute to the PH multidisciplinary clinical discussion.

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Declarations

Conflict of interest All authors declare no conflict of interest.

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