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### CHEST IMAGING





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# Imaging in chronic thromboembolic pulmonary hypertension: review of the current literature

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#### **ABSTRACT**

Chronic thromboembolic pulmonary hypertension (CTEPH) is a severe, life-threatening complication of pulmonary embolism with pulmonary hypertension (PH). The combination of insufficient resolution of thrombi following pulmonary emboli and accompanying microvascular disease results in PH. Advances in imaging can offer better insight into CTEPH diagnosis and management, but lack of disease awareness among radiologists has been shown to be a cause of CTEPH misdiagnosis or delayed diagnosis. This review highlights features pertinent to CTEPH diagnosis. The primary focus is on different modalities with their distinctive signs and newly developed technologies employing artificial intelligence systems.

#### **KEYWORDS**

Chronic thromboembolic pulmonary diseases, chronic thromboembolic pulmonary hypertension, pulmonary embolism, pulmonary hypertension, pulmonary angiography, computed tomography, dual-energy computed tomography, magnetic resonance imaging, four-dimensional magnetic resonance imaging, artificial intelligence

hronic thromboembolic pulmonary hypertension (CTEPH) is characterized by incomplete resolution of thrombus in the pulmonary arteries. It originates from deep veins in the lower limbs and is associated with pulmonary hypertension (PH). The accumulation of residual chronic thrombus in the pulmonary arteries triggers fibrotic proliferation and secondary microvasculopathy, leading to an elevation in pulmonary arterial pressure and pulmonary vascular resistance (PVR). The majority of thrombosis resolves following acute pulmonary thromboembolism (PE), but almost 5% of cases persist as chronic PE. Known and unknown factors are responsible for incomplete clot resolution. Abnormal fibrinogen structure and function, inhibition of thrombus angiogenesis, and inflammatory thrombosis are the main factors triggering microvasculopathy. There are also certain risk factors for CTEPH, including antiphospholipid syndrome and elevated factor VIII, splenectomy, chronic inflammatory processes, vascular shunts, infections, cancer, and non-O blood groups.

As per the 2022 European Society of Cardiology/European Respiratory Society (ESC/ERS) PH guidelines, CTEPH, placed in Group 4 PH, is a form of precapillary PH that is hemodynamically characterized by mean pulmonary arterial pressure (mPAP) measured at rest >20 mmHg, PVR >2 Wood units, and pulmonary artery wedge pressure <15 mmHg.<sup>3</sup> In the presence of comorbidities, mPAP can exceed 15 mmHg. These parameters should be supported with imaging modalities to establish an accurate diagnostic approach. Right heart catheterization (RHC) is therefore an integral component in CTEPH evaluation (Graphic 1).

CTEPH is defined by the presence of perfusion defects at least 3 months after anticoagulation following acute PE. Abnormal vascular organization and disorganized vascular remodeling lead to characteristic abnormalities in pulmonary arteries, including eccentric clots, stenosis, webs, bands, pruning, and pouch defects. Increased arterial tortuosity is also a feature of CTEPH.<sup>5</sup> A cohort of patients with chronic PE without PH is in the "Chronic Thromboembolic Pulmonary Disease" category.<sup>3,6</sup>

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The diagnosis of CTEPH can be challenging even with contemporary diagnostic modalities.

Although it may be a relatively uncommon disease, the diagnosis is often underestimated due to non-specific signs and symptoms as well as a lack of awareness among the radiological community.<sup>7</sup>

The initial period of CTEPH, commencing approximately 3 months post-acute PE and extending into the first year, is characterized as the "honeymoon period." Right ventricular hypertrophy and interventricular septal bowing may initially compensate for rising pulmonary pressures. During this phase, the pulmonary arteries may not attain critical values for mPAP and PVR. Prolonged elevation of mPAP measured at rest >30 mmHg leads to cor pulmonale and right heart failure, with low survival.<sup>8</sup>

A distinctive feature of CTEPH that sets it apart from other PH groups is the potential for cure with surgical pulmonary endarterectomy (PEA). There is generally a dramatic response to PEA surgery, with a rapid and substantial decrease in mPAP and permanent improvement of pulmonary hemodynamics.

## Methods

This review was conducted to synthesize and critically evaluate current imaging techniques and emerging technologies in the diagnosis and management of CTEPH. A systematic search was conducted across the PubMed, Scopus, and Google Scholar Databases, focusing on the literature published between January 2000 and April 2025. The search terms included "CTEPH," "dual-energy computed tomography," "photon-counting CT," "4D MRI,". "cardiac MRI in PH," "Artificial Intelligence in pulmonary hypertension," and

### Main points

- Chronic thromboembolic pulmonary hypertension (CTEPH) should be excluded in all patients presenting with pulmonary hypertension.
- CTEPH can be easily overlooked on imaging; hence, a high degree of suspicion is needed during imaging review.
- In challenging cases, novel diagnostic methodologies such as dual-energy computed tomography can be instrumental in achieving an accurate diagnosis.
- Consulting with CTEPH expert centers is highly beneficial for making informed decisions.

"pulmonary endarterectomy." Studies were chosen based on their clinical significance, methodological quality, and relevance to current imaging practices. Inclusion criteria encompassed peer-reviewed English-language articles, clinical trials, reviews, and quideline documents. Exclusion criteria included case reports, non-peer-reviewed abstracts, and non-English texts. Major sections of the manuscript are structured thematically according to modality, with integration of the recent ESC/ERS guidelines (2022) and expert consensus recommendations. All authors independently performed the literature screening, and any possible disagreement was resolved by consensus.

# Diagnostic approach to chronic thromboembolic pulmonary hypertension

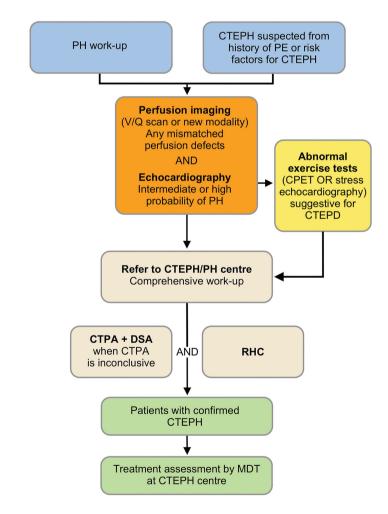
### Ventilation/perfusion scan

Ventilation/perfusion (V/Q) scintigraphy is the widely accepted screening modality

for excluding CTEPH. Guidelines still affirm a V/Q scan as a first step in the diagnostic process in CTEPH. Abnormal perfusion with preserved V/Q mismatch is suspicious for CTEPH, but as perfusion defects can also be seen in acute PE, large vessel vasculitis, pulmonary arterial malignancy, veno-occlusive disease, and vascular stenosis,9 it is necessary to perform anatomical imaging to confirm CTEPH. Single-photon emission computed tomography (SPECT) imaging is preferred over planar scintigraphy due to its multiplanar imaging capability and better diagnostic capacity (Figures 1 and 2). Low radiation dose exposure is a major advantage of V/Q scintigraphy. It has been shown that approximately 27–136-times less radiation exposure is required than with other diagnostic modalities using X-ray sources.10

## **Computed tomography**

Computed tomography (CT) pulmonary angiography (CTPA) plays a central role in



**Graphic 1.** Chronic thromboembolic pulmonary hypertension diagnosis algorithm adapted from the 2022 European Society of Cardiology/European Respiratory Society guidelines.<sup>12</sup> PH, pulmonary hypertension; CTEPH, chronic thromboembolic pulmonary hypertension; V/Q, ventilation/perfusion; CPET, chronic pulmonary hypertension; DSA, digital subtraction angiography; RHC, right heart catheterization; MDT, CTEPH Multidisciplinary Team.

the diagnosis of both acute thromboembolic pulmonary disease and CTEPH. In the setting of acute pulmonary embolism, CTPA is the first-line imaging modality recommended by the 2019 ESC/ERS guidelines, providing rapid and accurate identification of thrombi in the pulmonary arteries. In acute PE, CTPA not only allows evaluation of pulmonary arterial patency but also provides prognostic information through detection of right ventricular strain [e.g., right ventricle (RV)/left ventricle (LV) ratio, septal bowing] and extra-vascular findings. In the chronic setting, according to the 2022 ESC/ERS guidelines, echocardiography is the recommended first-line diagnostic modality (recommendation IB), whereas CTPA is included in the diagnostic algorithm for CTEPH (recommendation IC). The CTPA technique is a valuable anatomical tool in the assessment of CTEPH, particularly for visualizing eccentric wall-adherent thrombi, vessel stenosis, webs, and bands. Right ventricular remodeling is a key feature that develops as a consequence of long-standing pressure overload. Remodeling manifests as right ventricular hypertrophy due to increased afterload, followed by progressive dilatation in advanced stages, which ultimately leads to impaired contractility and right heart failure. The CTPA method can indirectly demonstrate these changes through findings such as increased RV free wall thickness, RV dilation, interventricular septal flattening or bowing toward the LV, and enlargement of the right atrium and inferior vena cava (IVC). These features, when combined with vascular abnormalities, strongly support the diagnosis of CTEPH and provide valuable prognostic information. 11,12

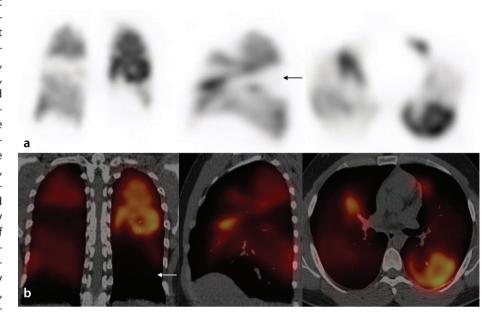
To provide optimal evaluation of the pulmonary and cardiovascular system, CTPA should be performed in the pulmonary arterial phase. Structured evaluation of CT includes assessment of vasculature, cardiac chambers, and lung parenchyma. Although high-quality CTPA can effectively demonstrate proximal CTEPH, it is still possible to miss distal CTEPH with conventional CTPA. One of the major limitations of CTPA in the context of CTEPH is its reduced sensitivity in evaluating subsegmental pulmonary arteries, primarily due to vessel tapering and suboptimal opacification in the distal pulmonary vasculature. These technical limitations can lead to underestimation of the distal disease burden, particularly in chronic thromboembolic disease affecting small-caliber vessels.

In addition to assessing the extent of the clot burden, CTPA offers key advantages in evaluating associated underlying lung and/

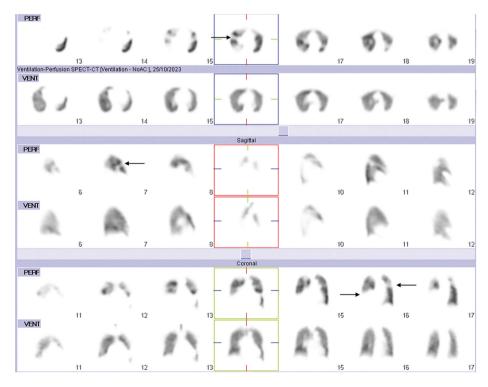
or cardiac diseases, as well as other potential causes of acute chest pain.<sup>13</sup>

Besides diagnosis, CTPA also has the potential to predict patient outcomes. Pulmonary arterial occlusion exceeding 30% considerably elevates vascular resistance, leading to PH and subsequent elevated right

ventricular afterload as well as an increased RV/LV ratio.<sup>14</sup> It is crucial to recognize that a substantial clot burden in the pulmonary bed poses a severe risk for right ventricular and cardiac dysfunction. During every CTPA evaluation process, the pulmonary trunk and main branch diameters and the RV/LV diameter ratio should be measured to determine



**Figure 1.** Selected images of coronal, sagittal, and axial single-photon emission computed tomography (SPECT) perfusion (**a**, top panel) and corresponding fusion images of SPECT perfusion and CT (**b**, bottom panel). Multiple bilateral lobar perfusion defects (white arrow) and segmental (black arrow) distribution in a 45-year-old man with chronic thromboembolic pulmonary hypertension.



**Figure 2.** Selected images of coronal, sagittal, and axial perfusion (top) and ventilation (bottom) from a ventilation/perfusion single-photon emission computed tomography series in a 40-year-old woman with chronic thromboembolic pulmonary hypertension. Multiple bilateral mismatched perfusion defects (black arrows) are observed.

whether PH is present. A main pulmonary trunk diameter >29 mm, an RV/LV ratio >0.9 on a four-chamber view, and leftward septal bowing are findings that indicate a high sensitivity for predicting PH on CTPA. These measurements should be interpreted in the context of CTEPH (Figures 3 and 4).<sup>2,6,15</sup> In addition, CTEPH is often associated with secondary changes in right heart morphology, including right ventricular hypertrophy, right atrial enlargement, and IVC dilation features, that reflect chronic pressure overload.

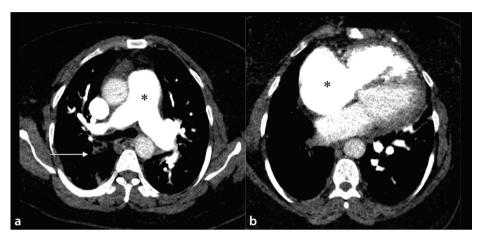
Besides these PH findings, obtuse angle-shaped, eccentrically positioned thrombus; asymmetric pruning or cut-off pulmonary artery branches; irregular linear filling defects, webs, or bands; and compensatory bronchial artery hypertrophy are all signs of CTEPH (Table 1, Figure 5). Generally, bronchial arteries originate from the systemic arterial circulation, most commonly from the descending thoracic aorta. Hypertrophy of the bronchial arteries, which often arise from the aorta or systemic vessels, represents com-

pensatory collateral circulation in CTEPH. These enlarged bronchial arteries may contribute to retrograde systemic-to-pulmonary flow. During PEA, this retrograde flow poses a substantial risk of intraoperative bleeding, making recognition of bronchial artery hypertrophy on imaging crucial for surgical planning. Pulmonary artery calcification, although relatively uncommon, may be observed in patients with chronic, long-standing CTEPH and is considered a marker of disease chronicity.<sup>1</sup>

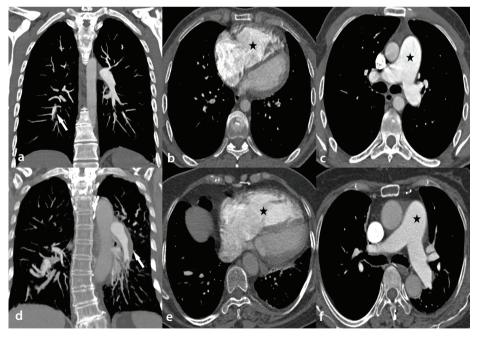
It is crucial to acknowledge that a portion of patients with CTEPH may also have concurrent acute PE. Consequently, CTPA can effectively reveal the presence of acute–subacute and chronic thrombi combinations and associated complications.<sup>16</sup>

Although CTPA is a foundational imaging modality in diagnosing acute pulmonary embolism and is also pivotal in evaluating CTEPH, its reliance on ionizing radiation necessitates strict adherence to the "as low as reasonably achievable" principle to balance diagnostic benefits with patient safety. Practical strategies for dose reduction include justification of each CT scan, automatic tube current modulation, lowering kilovolt (kV) peak, adjusting pitch, delaying scan initiation to limit unnecessary exposure, and optimizing scan range.17 Additionally, the use of iodinated contrast media entails a risk of contrast-induced acute kidney injury, particularly in patients with pre-existing renal dysfunction. Pre-hydration protocols and contrast dose minimization are advised in atrisk groups.18

In the imaging evaluation of CTEPH, standardized CTPA protocols are crucial for ensuring optimal diagnostic accuracy and reproducibility. Acquisition should be conducted using low-kV settings (typically 80-140 kV), selected based on body habitus or through automatic tube voltage selection systems to improve vascular contrast and reduce radiation dose. The tube current should be automatically modulated to maintain consistent image quality across varying anatomical thicknesses. Short gantry rotation times are critical to reduce motion artifacts, particularly in patients who are unstable or dyspneic. The use of thin collimation and 1-mm slice thickness reconstructions is recommended to ensure high spatial resolution. Scanning may be performed in either craniocaudal or caudocranial directions; the latter is preferable in patients with respiratory compromise to reduce basal motion artifacts. Although deep inspiratory breath-hold remains the



**Figure 3.** Axial computed tomography pulmonary angiography images. **(a)** Marked dilatation of the main pulmonary artery (star), exceeding the diameter of the adjacent ascending aorta due to pulmonary hypertension. Absence of visible vascular structures in the right lower lobe, suggestive of segmental occlusion due to chronic thromboembolic disease (white arrow). **(b)** Cardiomegaly with prominent right atrial and ventricular enlargement in pulmonary hypertension (star).



**Figure 4.** Computed tomography pulmonary angiography (CTPA) findings in patients with chronic thromboembolic pulmonary hypertension. Top row (**a-c**): patient 1. (**a**) Coronal reformatted CTPA image demonstrating chronic intraluminal thrombi within a segmental pulmonary artery (arrow); (**b**) axial image showing right ventricular dilatation secondary to pressure overload (star); (**c**) axial image illustrating main pulmonary artery enlargement consistent with pulmonary hypertension (star). Bottom row (**d-f**): patient 2. (**d**) Coronal reformatted CTPA image demonstrating chronic intraluminal thrombi (arrow); (**e**) axial image showing right ventricular dilatation (star); (**f**) axial image illustrating main pulmonary artery enlargement (star).

**Table 1.** Imaging findings in acute versus chronic thromboembolic disease and imaging modalities

Imaging finding	Imaging modality
Acute	
Centrally located thrombus	CTPA, CPA
Acute angle with vessel wall	CTPA, CPA
Polo-mint sign, floating	CTPA, CPA
Convex margins	CTPA, CPA
Preserved or increased caliber	CTPA, CPA
Parenchymal infarcts, reverse halo sign	Chest CT
Chronic	
Total occlusion— "cut-off" sign	CTPA, CPA
Obtuse angle with vessel wall	CTPA, CPA
Linear filling defects, bands, webs	CTPA, CPA
Concave margins	CTPA, CPA
Eccentric thrombus abutting the vessel wall	CTPA, CPA
Asymmetric pruning/decreased vessel caliber	CTPA, CPA
Bronchial artery hypertrophy	CTPA, CPA
Mosaic attenuation pattern	Dual-energy CTPA, V/Q scan

Note: Both CTPA and Chest CT refer to standard protocols as well as their dual-energy CT implementations. CTPA, computed tomography pulmonary angiography; Chest CT, thorax computed tomography; CPA, conventional pulmonary angiography; V/Q, ventilation/perfusion.



**Figure 5.** Coronal reformatted computed tomography pulmonary angiography image in chronic thromboembolic pulmonary hypertension. Segmental occlusion of the left lower lobe pulmonary artery is evident. Segmental occlusions are also present in the right lung. Marked bronchial artery hypertrophy (white arrows) is best observed on the coronal maximum intensity projection image.

preferred respiratory maneuver, shallow breathing or breath-holding at vital capacity may be necessary for symptomatic patients.

Intravenous contrast administration should be tailored to patient size, with a preferred flow rate of 3–5 mL/s and contrast volumes ranging from 80 to 100 mL, using agents with iodine concentrations of 300–370 mg/mL. Bolus triggering, ideally with region-of-interest placement in the pulmonary artery or ascending aorta, ensures optimal

timing of image acquisition. For comprehensive assessment in suspected CTEPH, the use of a generalized chest CT angiography protocol that includes simultaneous opacification of both pulmonary and systemic arteries is recommended. This approach is not a conventional triple-rule-out protocol aimed at coronary artery evaluation; rather, the acquisition is optimized in patients with CTEPH to achieve simultaneous opacification of the pulmonary arteries and systemic

arterial supply, allowing for evaluation of bronchial artery hypertrophy and systemic-to-pulmonary collaterals. Recognizing these systemic collaterals is crucial for both diagnosis and surgical planning in CTEPH. Advanced techniques such as dual-energy (DE) CT or photon-counting (PC) CT offer additional perfusion mapping capabilities, highlighting perfusion defects not easily seen on standard morphological imaging. Subtraction imaging may also be employed when pre-contrast data is available, particularly for assessing iodine distribution in the lung parenchyma. In special populations such as pregnant patients, modifications include the use of low tube voltage, caudocranial scanning, and enhanced contrast injection protocols to counteract increased blood volume and cardiac output while minimizing radiation exposure, particularly to the fetus. The application of model-based iterative or deep-learning reconstruction techniques is strongly encouraged to reduce image noise without increasing the dose.7

A potential pitfall in CTPA interpretation is transient interruption of contrast (TIC). This occurs when deep inspiration or abrupt intrathoracic pressure changes during injection lead to an admixture of unopacified IVC blood with contrast-opacified superior vena cava blood, which produces heterogeneous opacification of the pulmonary arteries and may mimic true filling defects. In addition to TIC, several other artifacts may complicate image interpretation. Respiratory motion can generate pseudo-filling defects, and beam hardening from dense contrast in the superior vena cava or subclavian veins may obscure adjacent pulmonary arteries. Inadequate contrast timing may result in insufficient pulmonary arterial enhancement, rendering the study non-diagnostic, and partial volume averaging—particularly adjacent to calcified vessel walls—may falsely appear as intraluminal thrombus. Furthermore, in patients with right heart dysfunction, delayed or heterogeneous contrast distribution can create flow-related artifacts that resemble vascular obstruction. Awareness of these pitfalls and the application of optimized scanning protocols—including proper bolus tracking, patient instruction, and thin-slice reconstructions—are crucial to avoid misinterpretation and ensure accurate diagnosis.19

# **Dual-energy computed tomography**

Dual-energy computed tomography (DECT) lung perfusion has gained wide-spread recognition as a valuable tool for identifying perfusion abnormalities. Iodine

maps can increase diagnostic confidence for CTEPH identification, and DECT perfusion imaging has been shown to reach close to 100% sensitivity and specificity for the diagnosis of CTEPH when combined with CTPA. The perfusion defects are typically wedgeshaped and located peripherally, distinguishing them from the more diffuse or central perfusion abnormalities often observed in conditions such as emphysema;20 DECT perfusion defects provide pixel-wise quantification of the spatial area of hypoperfusion.<sup>21</sup> The presence of perfusion defects in cases where there is no morphological evidence of chronic thromboembolism should raise suspicion of distal CTEPH (Figures 6 and 7).<sup>22</sup>

Furthermore, DECT scanners possess an additional capability for contrast media reduction through virtual monoenergetic imaging (VMI). The reconstruction of images at low energies (40-50 keV) with VMI application enhances vascular opacity, enabling the utilization of low-contrast media on DECT. Mosaic perfusion of the lungs is an integral feature of CTEPH, characterized by geographical areas of hypoperfused and normal or hyperperfused areas (Figure 8). Mosaic attenuation is not specific to CTEPH and may also be seen in small airway diseases, such as chronic obstructive pulmonary disease (COPD), hypersensitivity pneumonitis, and constrictive bronchiolitis. In CTEPH, the vascular etiology is suggested by a paucity of vessels in the hypoperfused zones, whereas in airway diseases, the affected areas often demonstrate air trapping on expiratory CT, which may be subtle in COPD but is usually more conspicuous in hypersensitivity pneumonitis or constrictive bronchiolitis. Recognizing these differences is essential in the differential diagnosis of mosaic attenuation patterns. It should also be noted that similar imaging findings—though typically less pronounced—may be encountered in bronchial obstructive pathologies, particularly in advanced stages of emphysema. Other lung abnormalities in CTEPH include parenchymal infarcts and related sequelae.

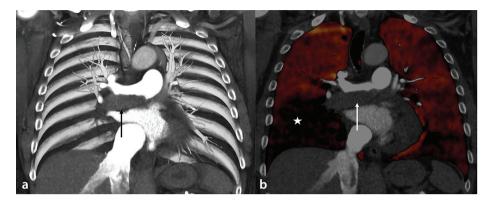
### Photon-counting computed tomography

Photon-counting computed tomography (PCCT) with novel detector technology permits ultra-high resolution (UHR) imaging of the lungs and pulmonary vasculature. This enables the visualization of subtle abnormalities at the microcirculatory level, providing detailed morphological insights and differentiating chronic thromboembolic lesions from artifacts. The PCCT method offers key radiation dose reductions while maintaining or improving image quality.<sup>23</sup> In CTEPH, the

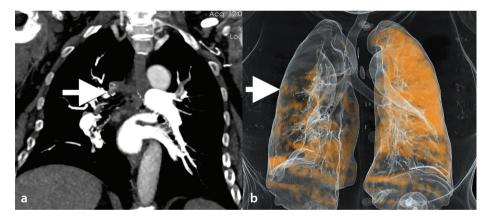
subpleural region of the lungs is the most affected area, characterized by major perfusion defects due to the presence of small-vessel disease. Poor subpleural perfusion can be precisely assessed using the UHR capability of PCCT, whereas CTEPH microvasculopathy, characterized by thin arterioles, venules, and ill-defined micronodules in the subpleural region of the lungs, which is not detectable

with other imaging modalities, could be identified using PCCT.<sup>24</sup>

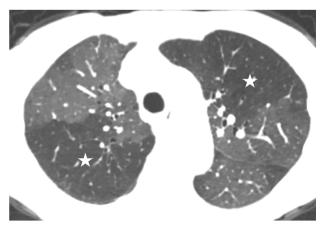
Additionally, four-dimensional (4D)-CT perfusion imaging with PCCT allows dynamic visualization of blood flow, enabling quantification of time-dependent perfusion parameters. This method provides complementary functional data alongside anatomical CTPA



**Figure 6. (a)** Coronal computed tomography pulmonary angiography shows eccentric thrombus in the right main pulmonary artery (black arrow) and occlusion of the right lower lobe vessels. **(b)** Dual-energy iodine map demonstrates a corresponding lobar perfusion defect in the right lung (star). Perfusion is also abnormal in the left lung due to subsegmental thrombi.



**Figure 7.** Coronal reformatted dual-energy computed tomography pulmonary angiography and volume-rendered perfusion images. **(a)** Lobar occlusion of the right upper lobe pulmonary artery due to chronic thromboembolic pulmonary hypertension (arrow); **(b)** corresponding three-dimensional volume-rendered perfusion image reveals marked perfusion defects in the right lung (arrow).



**Figure 8.** Geographical areas of normal (gray areas) and reduced (black areas, star) perfusion, known as mosaic attenuation in chronic thromboembolic pulmonary hypertension.

findings, particularly useful in patients with suspected distal or microvascular disease. Although 4D perfusion imaging with PCCT offers valuable functional data, it is not yet part of routine clinical practice. Its current use remains largely investigational, and further multicenter validation studies are required before clinical implementation.

### Catheter pulmonary angiography

Catheter pulmonary angiography (CPA) remains a cornerstone in the diagnostic workup of CTEPH, particularly when planning interventions such as PEA or balloon pulmonary angioplasty (BPA) (Figure 9). The CPA technique enables dynamic, high-resolution visualization of pulmonary arterial anatomy and is especially valuable for detecting chronic changes such as pouch defects, webs, bands, and abrupt vascular cutoffs. These findings help differentiate CTEPH from acute thromboembolic disease. However, CPA is highly operator dependent and subject to projectional limitations, which can lead to underestimation of distal disease burden. Overlapping vascular structures or suboptimal contrast timing may also obscure subtle lesions. Thus, high-quality biplane imaging and expert interpretation are essential. Furthermore, CPA offers the advantage of being combined with RHC in a single session, allowing for both anatomical and hemodynamic assessment. However, it is an invasive modality associated with procedural risks, including contrast nephropathy, vascular injury, arrhythmias, and, rarely, perforation. Its use should be balanced against clinical necessity and patient comorbidities.

Despite advances in non-invasive imaging modalities, RHC is still mandatory for hemodynamic evaluation. It is possible to combine RHC with CPA for detailed assessment of pulmonary circulation. 11,25

# Optical coherence tomography and intravascular ultrasound

Optical coherence tomography (OCT) and intravascular ultrasound (IVUS) are real-time intravascular high-resolution imaging modalities for detecting or characterizing PE and simultaneously measuring the luminal diameter.<sup>26</sup> The resolution capacity of the OCT system is almost 10-fold higher than that of IVUS for the evaluation of subsegmental branches of pulmonary arteries. Although not used routinely, both systems have been shown to be useful for the identification of BPA targets;<sup>27</sup> however, caution must be exercised, as it is possible to induce

pulmonary injury during OCT by forceful injection of a contrast jet. Forceful injection of contrast during OCT may result in pulmonary vascular injury, including barotrauma, vessel dissection, or even rupture, particularly in fragile or chronically diseased arterial segments.<sup>28</sup>

### Magnetic resonance imaging

Cardiac magnetic resonance imaging (MRI) is a comprehensive modality that combines basic morphologic imaging with advanced functional and tissue characterization techniques. Its value is particularly evident in CTEPH, where chronic pressure overload leads to progressive right ventricular remodeling. Cine steady-state free precession sequences are the cornerstone of cardiac MRI and provide dynamic visualization of cardiac motion, allowing accurate measurement of right ventricular and LV volumes, ejection fraction, and wall thickness. In CTEPH, these sequences typically reveal right ventricular hypertrophy and dilatation, interventricular septal flattening or leftward bowing, and right atrial enlargement, which together reflect the chronic hemodynamic stress imposed by elevated pulmonary pres-

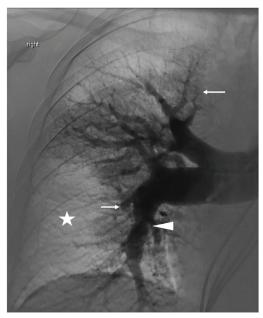
Beyond morphology and function, contrast-enhanced sequences add critical tissue information. Late gadolinium enhancement is a robust marker of focal fibrosis, most commonly observed at the right ventricular insertion points in patients with long-standing PH, including CTEPH. This localized fibrosis is

thought to result from repetitive mechanical stress at the interventricular junction and has been associated with adverse outcomes.

More advanced characterization is possible with mapping techniques. Specifically, T1 mapping and extracellular volume guantification provide a quantitative measure of diffuse interstitial expansion and fibrosis, changes that cannot be captured by late gadolinium enhancement alone. By contrast, T2 mapping is primarily sensitive to myocardial edema and can highlight reversible injury or acute strain superimposed on chronic remodeling. In the context of CTEPH, T1 mapping values and extracellular volume fractions are often elevated, reflecting diffuse myocardial changes secondary to pressure overload, whereas T2 mapping may help identify additional acute components of myocardial stress.

These MRI-derived parameters have particular importance in CTEPH because they correlate strongly with invasive hemodynamic measurements. Increased right ventricular volumes, reduced ejection fraction, abnormal strain, and elevated T1 or extracellular volume values are associated with relatively high PVR and mPAP.

Although mPAP itself is determined only by RHC, cardiac MRI provides reliable non-invasive surrogates that mirror hemodynamic burden and help in risk stratification. Moreover, cardiac MRI is valuable for follow-up after interventions such as PEA or BPA. Cine imaging can demonstrate reverse remodeling, with a reduction in right ventricular size



**Figure 9.** Digital subtraction angiography imaging of the right lung in a patient with chronic thromboembolic pulmonary hypertension with segmental occlusions and abrupt truncations (arrows), stenosis in the right lower lobe artery (arrowhead), and reduced parenchymal perfusion (star) in the capillary phase.

and improved function, whereas mapping techniques may show partial normalization of previously elevated T1 or extracellular volume values. These changes parallel clinical and hemodynamic improvement and underscore the role of MRI not only in diagnosis but also in monitoring therapeutic response in CTEPH.<sup>29</sup>

Recent studies have shown that newly improved MRI scanners with faster image acquisition and perfusion capabilities have a 97% sensitivity and 92% specificity for the diagnosis of CTEPH.30 To evaluate slow-flowing blood in pulmonary arteries, it is useful to combine the spin-echo double inversion recovery (black-blood) sequence with the half-Fourier single-shot turbo spin-echo sequence.31 Dynamic contrast-enhanced (DCE) MRI with three-dimensional (3D) T1-weighted gradient-echo sequences with more speedy acquisitions (Figure 10) is used for the delineation of vascular changes associated with CTEPH and can help discriminate CTEPH from other causes of PH.32 Quantitative evaluation, such as blood flow, volume, and mean transit time values, are additional benefits of DCE-MRI for better evaluation of microcirculation in patients with CTEPH in terms of progress or response to treatment.33

Compared with pulmonary angiography and CTPA, DCE-MRI had a slightly lower sensitivity than CTPA between 83% and 86% for depicting total occlusion and a sensitivity of almost 70% for the diagnosis of chronic thrombi, webs, and linear bands.<sup>34</sup>

Ferumoxytol (FE) and oxygen-enhanced MRI (oeMRI) studies have recently been used as alternative methods for lung perfusion. FE exhibits exceptional superparamagnetic properties and is gaining popularity as a potential alternative to gadolinium-based contrast agents due to safety concerns. Consequently, pulmonary MRI with FE could serve as a viable diagnostic approach, particularly in pregnant women and individuals with chronic kidney disease. FE-enhanced MRI offers a wide range of vascular indications, including CTEPH, and can be further enhanced with 4D imaging techniques. The oeMRI method facilitates the visualization of primary lung function, gas exchange, and perfusion mechanisms. In comparison to other imaging techniques, oeMRI exhibits moderate diagnostic compatibility with V/Q scans. Notably, oeMRI demonstrates superior sensitivity to ventilation scintigraphy.35,36 Although FE-enhanced MRI may serve as an alternative in patients with renal dysfunction, the American College of Radiology recommends avoiding its use during pregnancy unless absolutely necessary, following a thorough risk-benefit analysis.<sup>37</sup>

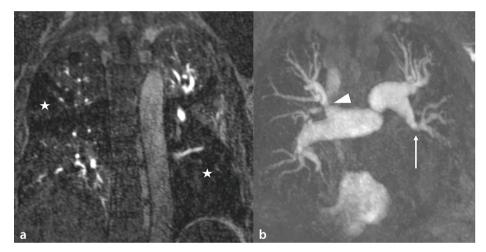
# Four-dimensional magnetic resonance imaging

4D flow MRI can provide a comprehensive and real-time objective evaluation of different blood-flow parameters, such as wall shear stress, vorticity, kinetic energy, and pressure gradients.<sup>38</sup> This application holds strong potential for evaluation of the pulmonary vascular system (Figure 11). Early onset of retrograde flow in the main pulmonary artery is an important abnormality in PH. A linear correlation between mPAP and vortex value in PH has been shown; a relative vortex flow time over 14.3% of the cardiac cycle has 97% sensitivity and 96% specificity for PH.<sup>39</sup> Vorticity in the main pulmonary artery also

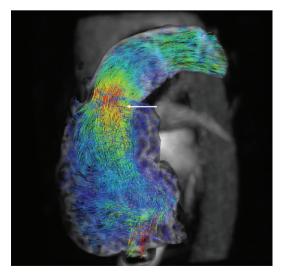
correlates with PVR, and helicity reveals main pulmonary artery stiffness with right ventricular outflow function.<sup>40,41</sup> Despite these promising findings, multicenter validation is required to establish the utility of 4D flow in PH diagnosis.

# Phase-resolved functional lung magnetic resonance imaging

Currently, there is a focus on lung perfusion imaging without using intravenous contrast media. The phase-resolved functional lung (PREFUL) technique is a novel, non-contrast MRI application that utilizes artificial intelligence (AI)-driven subtraction images of lungs. It possesses the potential to serve as a diagnostic aid in a spectrum of lung diseases, particularly in patients with PH and CTEPH. The primary objective of the PREFUL technique is to discern variations in perfusion



**Figure 10. (a)** Magnetic resonance perfusion and **(b)** angiography imaging in chronic thromboembolic pulmonary hypertension (CTEPH). Occlusion of the left lower lobe (arrow) and origin stenosis of the right upper lobe (arrowhead) are associated with corresponding perfusion defects on the left side in CTEPH (star).



**Figure 11.** Magnetic resonance four-dimensional flow image of the right ventricle outflow tract and pulmonary artery, with abnormal flow vortex (arrow) in the main pulmonary artery due to pulmonary hypertension in chronic thromboembolic pulmonary hypertension.

defects between inspiration and expiration image groups.<sup>42</sup> Prospective multicenter validation is necessary prior to the adoption of this technique in routine clinical practice.

### Positron emission tomography

Fluorine-18 fluorodeoxyglucose (18F-FDG) is a radiopharmaceutical analog that is predominantly utilized in positron emission tomography (PET)-CT to evaluate glucose metabolism. The PET-CT approach is a valuable imaging modality employed in tumor diagnostics, primarily based on the elevated glucose metabolism observed within tumor cells.

Uptake of <sup>18</sup>F-FDG in PH has been reported in the RV, central pulmonary artery, and lung parenchyma. Increased <sup>18</sup>F-FDG uptake in these areas may reflect underlying inflammation, vascular remodeling, or increased metabolic demand due to pressure overload. These findings may help differentiate CTEPH from other pulmonary vascular diseases and provide insight into disease activity or prognosis.43 The most critical role of 18F-FDG in Group 4 PH is in the differential diagnosis between CTEPH and pulmonary artery tumors; pulmonary artery malignancy has a poor prognosis and is occasionally misdiagnosed as PE, leading to incorrect treatment or surgery.

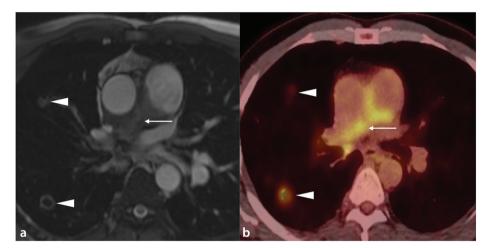
In certain cases, distinguishing CTEPH from tumors (particularly angiosarcomas) and vasculitis can be challenging due to the location and shape of the thrombus (Figures 12 and 13). Consequently, the high cellularity of tumors is generally associated with elevated uptake of <sup>18</sup>F-FDG, which serves as a valuable marker for correct diagnosis and may allow surgery or chemotherapy to be performed promptly.<sup>44</sup>

Another important role of PET-CT is its capability to diagnose large-vessel vasculitis, such as Behçet's, Takayasu, and giant cell arteritis. Early diagnosis of vasculitis plays a valuable role in reducing complications such as aneurysm, PE, and CTEPH.<sup>45</sup>

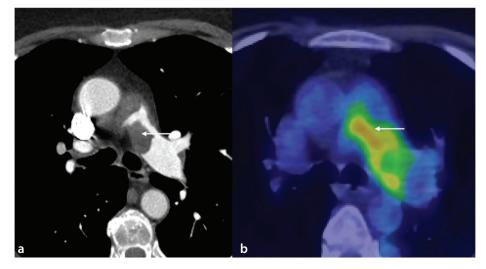
# Evidence-based evaluation of imaging techniques for chronic thromboembolic pulmonary hypertension

The assessment of imaging modalities in pulmonary vascular disease, particularly in CTEPH, reveals important variability in both evidence strength and methodological quality.

A critical review of imaging methods used for CTEPH shows that the strength of evidence supporting each technique varies between studies and that many studies have methodological limitations. This emphasizes the need to clearly understand the diagnostic accuracy, potential sources of bias, and clinical applicability of each imaging modality. Although V/Q scintigraphy remains the gold-standard first-line test for CTEPH screening, with reported sensitivity of 90%-100% and specificity of 94%-100%,46 a systematic review using the Quality Assessment of Diagnostic Accuracy Studies-2 tool revealed that most V/Q SPECT studies have significant methodological bias, with 11 of 13 studies rated high risk in ≥2 domains with important applicability concerns.<sup>47</sup> The diagnostic performance of CTPA in detecting CTEPH is limited, with reported sensitivity and specificity of 76% and 96%, respectively, at the patient level. However, its accuracy improves when high-resolution, multi-detector CT scanners are employed and images are interpreted by experienced radiologists. The CTPA approach may miss distal disease (potential risk of underdiagnosis), and evaluation of image quality and reader expertise remains critical. Although DECT also offers perfusion insights, formal evidence for diagnostic accuracy is still emerging.<sup>12</sup> In a study involving 74 patients with suspected CTEPH, DCE-MRI demonstrated a sensitivity of 100% for screening, compared with 97% for V/Q SPECT. The level of evidence supporting this finding was classified as level 3, Stage 3 technical efficacy. Given its radiation-free nature, DCE-MRI represents a promising imaging modality; however, its clinical utility requires



**Figure 12.** Large vessel vasculitis with pulmonary arterial involvement. (a) Axial magnetic resonance image with soft tissue thickening involving the right pulmonary artery (thin arrow) and small cavitating and non-cavitating lesions in the right lung (arrowheads); (b) corresponding axial positron emission tomography/computed tomography shows both pulmonary vascular and lung parenchymal lesions to be fluorodeoxyglucose avid.



**Figure 13.** Pulmonary artery sarcoma. **(a)** Computed tomography pulmonary angiography image shows expansile soft tissue lesion in the main and left pulmonary arteries (thin arrow); **(b)** corresponding positron emission tomography/computed tomography imaging shows intense fluorodeoxyglucose avidity in the left pulmonary artery.

confirmation through larger, prospective studies.<sup>48</sup> Digital subtraction angiography (DSA) is currently regarded as the reference standard when CTPA findings are inconclusive. However, no formal assessment of risk of bias or grading of the level of evidence is provided in the current guidelines. Its use remains standard but is mostly supported by expert consensus and procedural experience rather than comparative trials.

The 2022 ESC/ERS guidelines for PH, which also encompass CTEPH as a distinct subtype, provide structured diagnostic recommendations. In these guidelines, CTPA is explicitly recommended in the diagnostic work-up of patients with suspected PH to evaluate for CTEPH (class I, level C). This grading reflects a strong consensus regarding the central role of CTPA. Beyond CTPA, the guidelines emphasize a multimodality and multidisciplinary approach. Perfusion lung or V/Q scintigraphy remains a cornerstone for screening unexplained PH and is also listed as a class I, level C recommendation to exclude or confirm the probability of CTEPH. Additional imaging modalities, such as DSA and chest CT, are considered useful in selected contexts (class IIa), particularly for anatomical delineation and interventional planning. According to the 2022 ESC/ERS guidelines, the diagnostic approach to CTEPH in symptomatic patients should follow a structured and multidisciplinary pathway. The entire diagnostic process should be coordinated within experienced, high-volume CTEPH centers, where Multidisciplinary Team (MDT) comprising PH specialists, radiologists, surgeons, and interventionalists can guide appropriate management decisions.

### **Reporting recommendations**

Reports should clearly state the extent of pulmonary artery analyzability; in cases where only central arteries are evaluable or image quality is compromised, the study should be labeled as indeterminate or non-diagnostic. Quantitative assessment of contrast enhancement is advisable, and attenuation values should be reported when suboptimal opacification is suspected. Simultaneous evaluation of mediastinal and lung window images is essential to differentiate true emboli from artifacts. The interpretation and communication of CTPA findings in suspected CTEPH should follow a standardized reporting structure that encompasses both diagnostic and prognostic information, facilitating clinical decision-making and longitudinal patient care. Central to this approach is the inclusion of a core set of

imaging findings—derived through expert consensus—that have demonstrated prognostic significance. These include the RV/LV diameter ratio measured on axial images. central embolus location, the presence of an isolated subsegmental PE, septal bowing, right ventricular hypertrophy, bronchial artery dilatation, intravascular webs or bands, and pulmonary artery retraction. Supplementary findings such as main pulmonary artery diameter, complete arterial occlusion, and organized mural thrombi are considered clinically useful and should be reported. For perfusion studies (e.g., DECT or V/Q), quantification of the perfusion defect extent (e.g., as % of total lung volume) should be reported. Further evaluation with MRI for verification of CTEPH diagnosis should primarily focus on right ventricular functional and morphological parameters, including the ventricular mass index, RV ejection fraction, and RV longitudinal strain. Additionally, MRI can provide indirect estimations of hemodynamic parameters, such as mPAP, ventriculoarterial coupling (RV-PA coupling), end-systolic elastance, and arterial elastance; however, these values are not directly measured but rather estimated through derived indices and are ideally confirmed by invasive RHC.7,49

# Multidisciplinary team and expert centers for chronic thromboembolic pulmonary hypertension

The idea of a "CTEPH MDT" was first postulated at the fifth World Symposium on PH in 2013 and has subsequently been permanently embedded into the CTEPH treatment algorithm. Recent studies increasingly support the concept that CTEPH-MDTs should be based in high-volume centers with established experience in pulmonary PEA and BPA. Concentrating expertise in such centers has been associated with improved diagnostic accuracy, optimized patient selection, and superior long-term outcomes. A "CTEPH expert center" should include an experienced CTEPH cardiovascular surgeon, PH pulmonologist, and cardiologist (preferentially a BPA specialist), as well as a cardiothoracic radiologist with specialist expertise in CTEPH.50 Given the diagnostic complexities, expert radiological opinion has assumed a pivotal role in the implementation of a successful CTEPH program. The specialist MDT is essential to ensure accurate diagnosis, appropriate timing, and precise surgical and BPA planning. In addition, a considerable number of patients with CTEPH require long-term follow-up with CTEPH-experienced medical professionals to

ensure optimal and prolonged survival.

In addition to surgical and interventional options, medical therapies such as Riociguat treatment, an approved soluble guanylate cyclase stimulator, play a crucial role in patients who are inoperable, have persistent PH following PEA, or are undergoing BPA.<sup>51</sup> A combined approach integrating pharmacologic treatment with procedural interventions is increasingly considered to improve hemodynamics and clinical status in selected patients.<sup>12,50</sup>

### **Pulmonary endarterectomy**

PEA is one of the treatment options for CTEPH in the absence of life-threatening serious comorbidities. This open-heart surgery necessitates a true bilateral endarterectomy through the pulmonary arteries. The proximal extension of the cast defines the level of disease, and the classification is made according to the material removed surgically. Level I defines a disease in the right or left main pulmonary artery; level II is a disease in the lobar branches; level III is in the proximal segmental arteries; and level IV is in the distal segmental and subsegmental pulmonary branches (Figure 14).<sup>52</sup>

Residual thrombi, particularly in the distal branches of the pulmonary artery, should be reported following PEA surgery.

Cardiopulmonary bypass is followed by circulatory arrest in deep hypothermia, which is necessary to keep the surgical field free of blood via the bronchopulmonary collaterals and protect brain function. Identification of the correct plane of dissection in the segmental and sub-segmental branches and removing the material en bloc with its tail is pivotal.

PEA remains a challenging surgery and is determined by multiple factors, such as technical operability (e.g., accessibility of the obstructions and surgical expertise) and the risk-benefit ratio, as determined by preoperative hemodynamics and comorbidities.

Following PEA surgery, the literature reported a survival rate exceeding 90% within 3 years. However, if patients with CTEPH had not undergone surgery, the survival rate during this period was only 70%.<sup>8,53</sup>

### **Balloon pulmonary angioplasty**

BPA has emerged as a viable alternative treatment option for patients with CTEPH who experience thrombi formation within the distal–proximal segmental locations of level III–IV PAs. An alternative option for BPA is in-



**Figure 14.** Pulmonary endarterectomy specimen of chronic thrombus demonstrates typical yellowish-white chronic thrombus with tails.

operable CTEPH or in patients with residual lesions following PEA.<sup>12</sup> The BPA method requires extensive training and has been shown to have a learning curve to reduce complications. Before and after the BPA procedure, pressure gradient analysis should be performed to evaluate the procedural success.<sup>54</sup> Vascular injury due to wire perforation is the most feared complication of BPA, with a rate of approximately 7.7%.<sup>55</sup> An evolving trend is the combined use of PEA and BPA as a "hybrid approach."

## Artificial intelligence in chronic thromboembolic pulmonary hypertension imaging

Applying AI in diagnostic imaging involves utilizing machine learning (ML) algorithms and methodologies to analyze images, thereby assisting in the interpretation and optimal diagnosis. It is important to note that ML is a subset of AI, which broadly encompasses various computational techniques designed to simulate human decision-making. Given the rarity and diagnostic complexity of CTEPH, Al-driven tools—particularly ML-based algorithms—could provide vital support to radiologists by enhancing the detection of subtle vascular abnormalities and improving diagnostic consistency across centers. ML can assist in quantifying vascular morphology by automatically segmenting pulmonary vessels on CT imaging. Additionally, vascular density, volume fraction, volume ratio of the vessels, and pulmonary artery tortuosity can be measured in patients with CTEPH using ML.56

Promising diagnostic methods for Albased CT techniques include CT morphometry (pulmonary vasculature analysis) and computational fluid dynamics (CFD). Al-based CT morphometry is a specialized technique employed to detect vascular remodeling. It utilizes Al-driven vasculature analysis derived from CT scans to quantify the varying sizes of vessels. Vascular pruning in CTEPH has been associated with the loss of vascular volume and density.<sup>57</sup>

CFDs focuses on detecting the flow velocity and wall shear stress in pulmonary artery branches with the assistance of Al. There is a reduction in wall shear stress in patients with PH compared with healthy individuals. This technique is potentially beneficial in determining the therapeutic response to PEA and BPA.<sup>58</sup>

The application of ML in CTEPH research presents several issues, primarily due to the condition's rarity, resulting in limited patient cohorts. Additionally, assessing the substantial heterogeneity of vascular pathologies associated with CTEPH poses considerable difficulties. Leveraging the major advancements in ML technology, further research on CTEPH imaging utilizing Al has the potential to be of immense value in CTEPH diagnosis, risk stratification, and therapy evaluation but requires validation in large cohorts.

In conclusion, this review highlighted the known and newly developed imaging modalities, including Al-based techniques, for the diagnosis of CTEPH. Despite advances in diagnostic modalities over the past two decades, CTEPH remains challenging to diagnose and manage. Radiologists play a crucial role in improving the outcomes of patients with CTEPH. The decision-making process in CTEPH diagnosis and management requires an expert MDT. Although new modalities and techniques have many advantages, their utilization is limited in many centers. In the

future, CTEPH expert centers should connect with each other to provide second opinions for complex cases as well as to expand radiologist training. These improvements could be beneficial for pooling all resources and gaining optimal expertise.

Future research on CTEPH imaging should focus on the integration of advanced modalities, such as PCCT, DECT, 4D-flow MRI, and Al-driven image analysis. Prospective multicenter trials are required to validate their diagnostic and prognostic utility in diverse patient populations. Standardized endpoints, including the 6-minute walk distance, mPAP, PVR, and overall survival, should be adopted.

In addition, Al-based techniques such as CT morphometry and CFD have shown potential in quantifying pulmonary vascular pruning, tortuosity, and altered hemodynamics. These technologies could support personalized therapy planning and early identification of disease progression.

International collaboration between expert centers is essential for establishing imaging-based biomarkers, creating open-access datasets, and developing consensus protocols to advance the field.

### **Footnotes**

### **Conflict of interest disclosure**

Mehmet Ruhi Onur, MD, is Editor in Chief Diagnostic and Interventional Radiology. He had no involvement in the peer-review of this article and had no access to information regarding its peer-review. Other authors have nothing to disclose.

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