

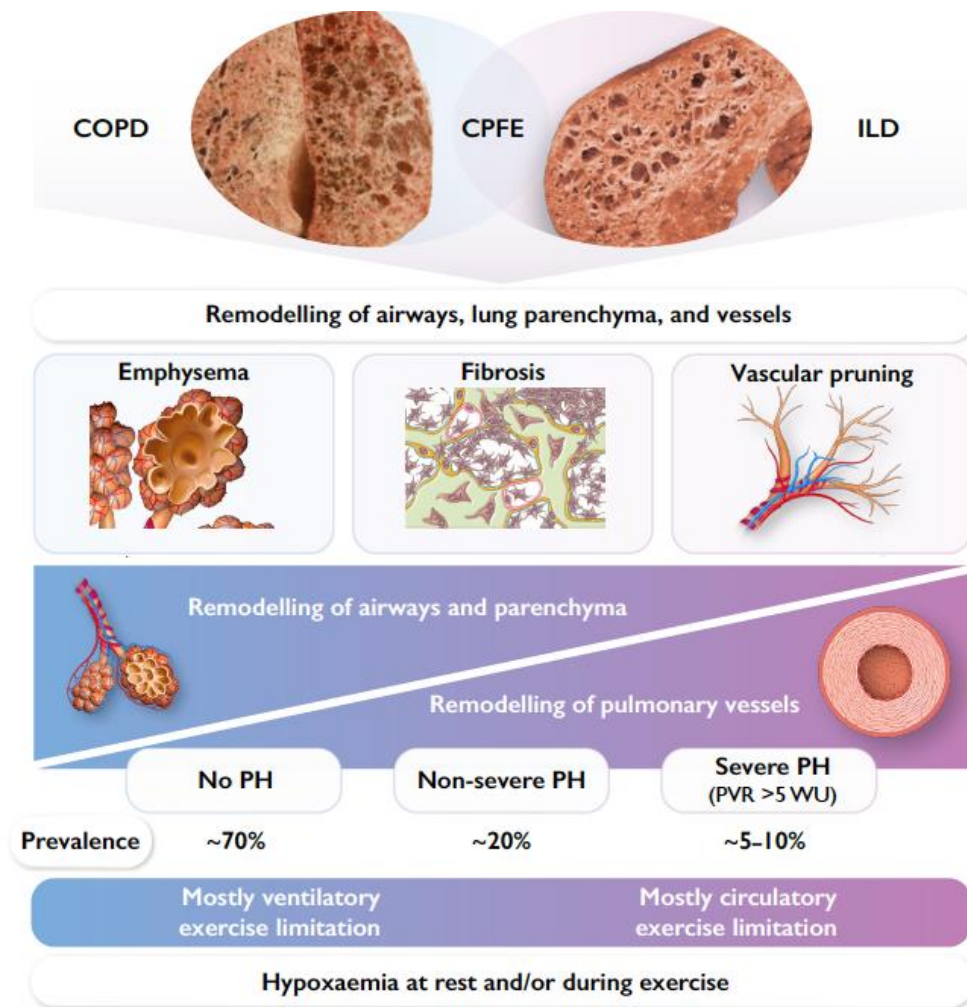


Figure 1. ESC 2022 GL on PH. Pathophysiology of pulmonary hypertension associated with lung disease (group 3).

COPD, chronic obstructive pulmonary disease; CPFE, combined pulmonary fibrosis and emphysema; ILD, interstitial lung disease; PH, pulmonary hypertension; PVR, pulmonary vascular resistance; WU, Wood units. Shown are underlying lung diseases (upper panel); contributing pathogenic pulmonary alterations of airways, parenchyma, and vessels (middle panel); and the relation of airway/parenchymal remodelling and vascular remodelling to the degree of PH and its consequences for exercise limitation (ventilatory vs. circulatory, lower panel).

Table 1. ESC 2022 GL on Pulmonary Hypertension. Recommendations for pulmonary hypertension associated with lung disease and/or hypoxia

If PH is suspected in patients with lung disease, echocardiography ^a to be performed and the results interpreted in conjunction with ABG, PFTs including DLCO, and CT imaging	I C
In patients with lung disease and suspected PH, optimize treatment of the underlying lung disease and, where indicated, hypoxaemia, sleep-disordered breathing, and/or alveolar hypoventilation	I C
In patients with lung disease and suspected severe PH, or where there is uncertainty regarding the treatment of PH, referral to a PH centre ^b	I C
In patients with lung disease and severe PH, an individualized approach to treatment	I C
Refer eligible patients with lung disease and PH for LTx evaluation	I C
In patients with lung disease and suspected PH, RHC if the results are expected to aid management decisions	I C
Inhaled treprostinil in patients with PH associated with ILD	IIb B
The use of ambrisentan is not recommended in patients with PH associated with IPF	III B
The use of riociguat is not recommended in patients with PH associated with IIP	III B
The use of PAH medication is not recommended in patients with lung disease and non-severe PH ^c	III C

	GRADE		
	Quality of evidence	Strength of recommendation	
PDE5is may be considered in patients with severe PH associated with ILD	Very low	Conditional	IIb C
(individual decision-making in PH centres)	Very low	Conditional	III C
The use of PDE5is in patients with ILD and non-severe PH is not recommended			

ABG, arterial blood gas analysis; CT, computed tomography; DLCO, Lung diffusion capacity for carbon monoxide; IIP, idiopathic interstitial pneumonia; IPF, idiopathic pulmonary fibrosis; ILD, interstitial lung disease; LTx, lung transplantation; PAH, pulmonary arterial hypertension; PDE5i, phosphodiesterase 5 inhibitor; PFT, pulmonary function test; PH, pulmonary hypertension; RHC, right heart catheterization.

^a Assessments should ideally be made when the patient is clinically stable, as exacerbations can significantly raise pulmonary artery pressure.

^b This recommendation does not apply to patients with end-stage lung disease who are not considered candidates for LTx.

^c This does not include inhaled treprostinil, which may be considered in patients with PH associated with ILD, irrespective of PH severity.