

Table 1. ESC 2022 GI on Pulmonary Hypertension. Haemodynamic definitions of pulmonary hypertension

Definition	Haemodynamic characteristics
PH	mPAP >20 mmHg
Pre-capillary PH	mPAP >20 mmHg PAWP $\leq$ 15 mmHg PVR >2 WU
lpcPH	mPAP >20 mmHg PAWP >15 mmHg PVR $\leq$ 2 WU
CpcPH	mPAP >20 mmHg PAWP >15 mmHg PVR >2 WU
Exercise PH	mPAP/CO slope between rest and exercise >3 mmHg/L/min

CO, cardiac output; CpcPH, combined post- and pre-capillary pulmonary hypertension; lpcPH, isolated post-capillary pulmonary hypertension; mPAP, mean pulmonary arterial pressure; PAWP, pulmonary arterial wedge pressure; PH, pulmonary hypertension; PVR, pulmonary vascular resistance; WU, Wood units.

Some patients present with elevated mPAP (>20 mmHg) but low PVR ($\leq$ 2 WU) and low PAWP ($\leq$ 15 mmHg); this haemodynamic condition may be described by the term 'undclassified PH' (see text for further details).

Table 2. ESC 2022 GI on Pulmonary Hypertension. Clinical classification of pulmonary hypertension

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 ^a
1.3 Associated with drugs and toxins ^a
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) 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 ^b
2.2 Valvular heart disease
2.3 Congenital/acquired cardiovascular conditions leading to post-capillary PH
GROUP 3 PH associated with lung diseases 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 thrombo-embolic PH
4.2 Other pulmonary artery obstructions ^c
GROUP 5 PH with unclear and/or multifactorial mechanisms
5.1 Haematological disorders ^d
5.2 Systemic disorders ^e
5.3 Metabolic disorders ^f
5.4 Chronic renal failure with or without haemodialysis
5.5 Pulmonary tumour thrombotic microangiopathy
5.6 Fibrosing mediastinitis

HF, heart failure; HIV, human immunodeficiency virus; PAH, pulmonary arterial hypertension; PCH, pulmonary capillary haemangiomatosis; PH, pulmonary hypertension; PVOD, pulmonary veno-occlusive disease.

^aPatients with heritable PAH or PAH associated with drugs and toxins might be acute responders.

^bLeft ventricular ejection fraction for HF with reduced ejection fraction: $\leq 40\%$; for HF with mildly reduced ejection fraction: 41–49%.

^cOther causes of pulmonary artery obstructions include: sarcomas (high or intermediate grade or angiosarcoma), other malignant tumours (e.g. renal carcinoma, uterine carcinoma, germ-cell tumours of the testis), non-malignant tumours (e.g. uterine leiomyoma), arteritis without connective tissue disease, congenital pulmonary arterial stenoses, and hydatidosis.

^dIncluding inherited and acquired chronic haemolytic anaemia and chronic myeloproliferative disorders.

^eIncluding sarcoidosis, pulmonary Langerhans's cell histiocytosis, and neurofibromatosis type 1.

^fIncluding glycogen storage diseases and Gaucher disease.

Table 3. ESC 2022 GI on Pulmonary Hypertension. World Health Organization classification of functional status of patients with pulmonary hypertension

Class	Description^a
WHO-FC I	Patients with PH but without resulting limitation of physical activity. Ordinary physical activity does not cause undue dyspnoea or fatigue, chest pain, or near syncope
WHO-FC II	Patients with PH resulting in slight limitation of physical activity. They are comfortable at rest. Ordinary physical activity causes undue dyspnoea or fatigue, chest pain, or near syncope
WHO-FC III	Patients with PH resulting in marked limitation of physical activity. They are comfortable at rest. Less than ordinary activity causes undue dyspnoea or fatigue, chest pain, or near syncope
WHO-FC IV	Patients with PH with an inability to carry out any physical activity without symptoms. These patients manifest signs of right HF. Dyspnoea and/or fatigue may even be present at rest. Discomfort is increased by any physical activity

PH, pulmonary hypertension; WHO-FC, World Health Organization functional class.

^a Functional classification of PH modified after the New York Heart Association functional classification according to the World Health Organization 1998.

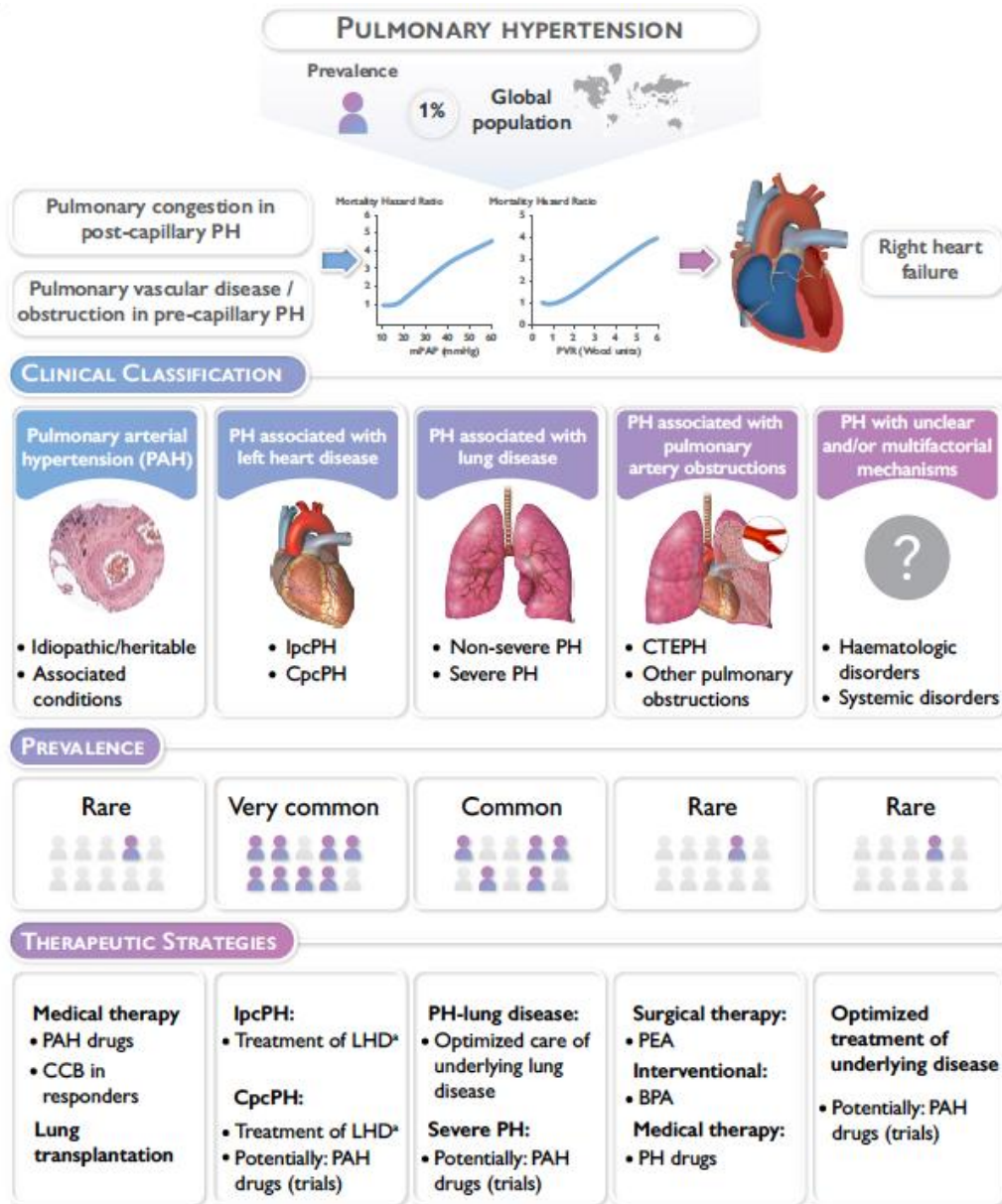


Figure 1. ESC 2022 GI on Pulmonary Hypertension. Central illustration.

BPA, balloon pulmonary angioplasty; CCB, calcium channel blocker; CTEPH, chronic thrombo-embolic pulmonary hypertension; CpCPH, combined post- and pre-capillary pulmonary hypertension; IpcPH, isolated post-capillary pulmonary hypertension; LHD, left heart disease; PAH, pulmonary arterial hypertension; PEA, pulmonary endarterectomy; PH, pulmonary hypertension.

a Treatment of heart failure according to the ESC Guidelines for the diagnosis and treatment of acute and chronic heart failure. Treatment of left-sided valvular heart disease according to the 2021 ESC/EACTS Guidelines for the management of valvular heart disease.

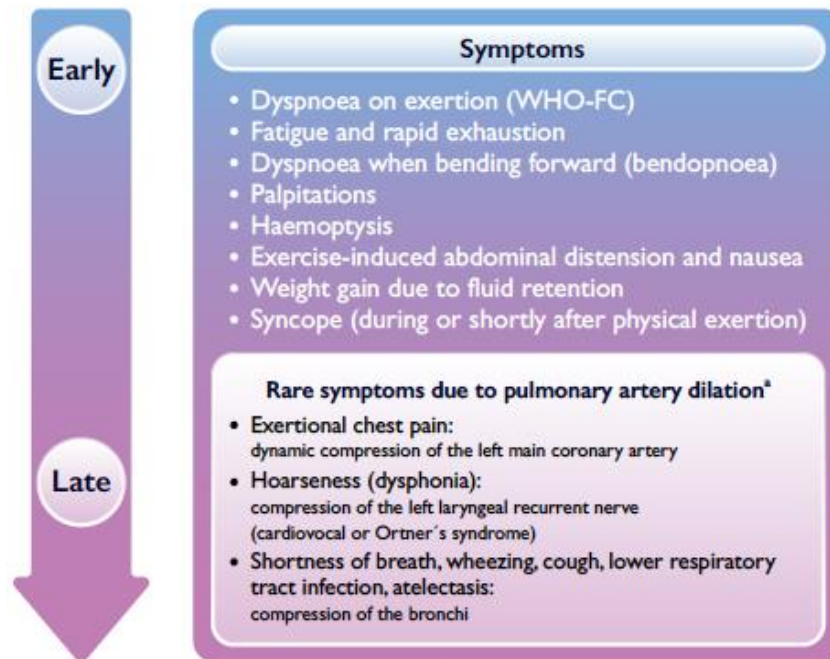


Figure 2. ESC 2022 GI on Pulmonary Hypertension. Symptoms in patients with pulmonary hypertension.

WHO-FC, World Health Organization functional class.

^a Thoracic compression syndromes are found in a minority of patients with PAH with pronounced dilation of the pulmonary artery, and may occur at any disease stage and even in patients with otherwise mild functional impairment.

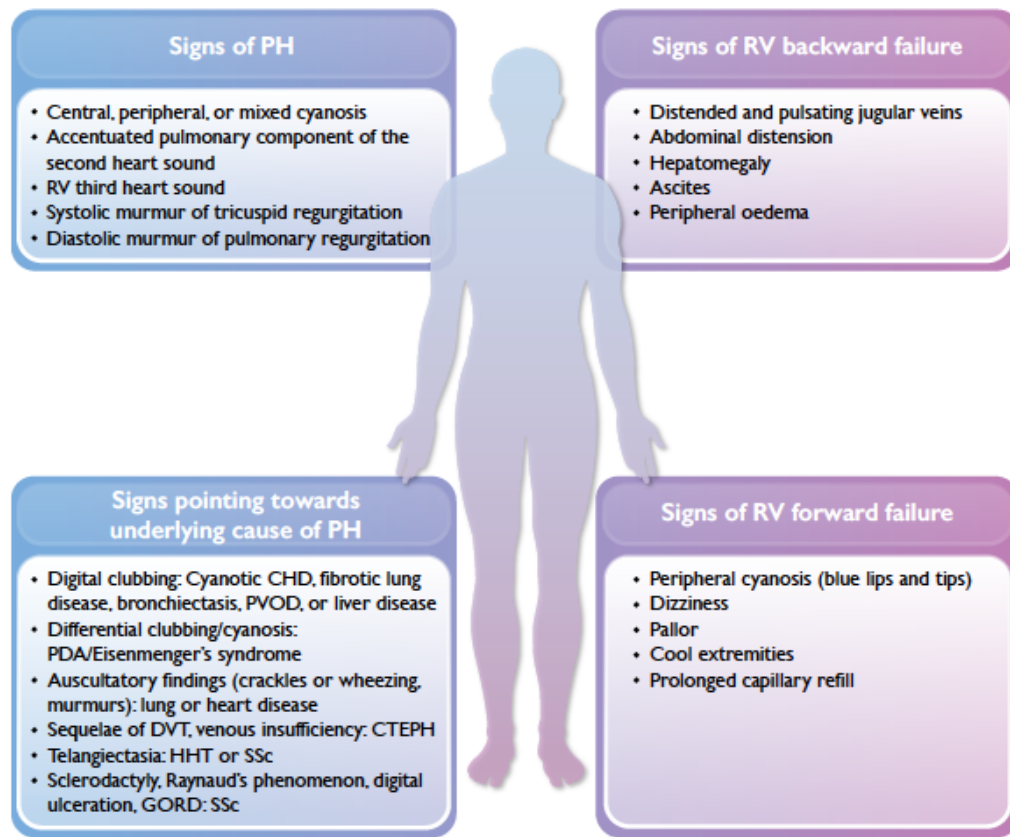


Figure 3. ESC 2022 GI on Pulmonary Hypertension. Clinical signs in patients with pulmonary hypertension.

CHD, congenital heart disease; CTEPH, chronic thrombo-embolic pulmonary hypertension; DVT, deep venous thrombosis; GORD, gastro-oesophageal reflux disease; HHT, hereditary haemorrhagic telangiectasia; PDA, patent ductus arteriosus; PH, pulmonary hypertension; PVOD, pulmonary veno-occlusive disease; RV, right ventricle; SSc, systemic sclerosis.

Table 4. ESC 2022 GI on Pulmonary Hypertension. Radiographic signs of pulmonary hypertension and concomitant abnormalities

Signs of PH and concomitant abnormalities	Signs of left heart disease/ pulmonary congestion	Signs of lung disease
Right heart enlargement	Central air space opacification	Flattening of diaphragm (COPD/emphysema)
PA enlargement (including aneurysmal dilatation)	Interlobular septal thickening 'Kerley B' lines	Hyperlucency (COPD/emphysema)
Pruning of the peripheral vessels	Pleural effusions	Lung volume loss (fibrotic lung disease)
'Water-bottle' shape of cardiac silhouette ^a	Left atrial enlargement (including splayed carina) Left ventricular dilation	Reticular opacification (fibrotic lung disease)

COPD, chronic obstructive pulmonary disease; PA, pulmonary artery; PH, pulmonary hypertension.

^aMay be present in patients with PH with advanced right ventricular failure and moderate pericardial effusion.

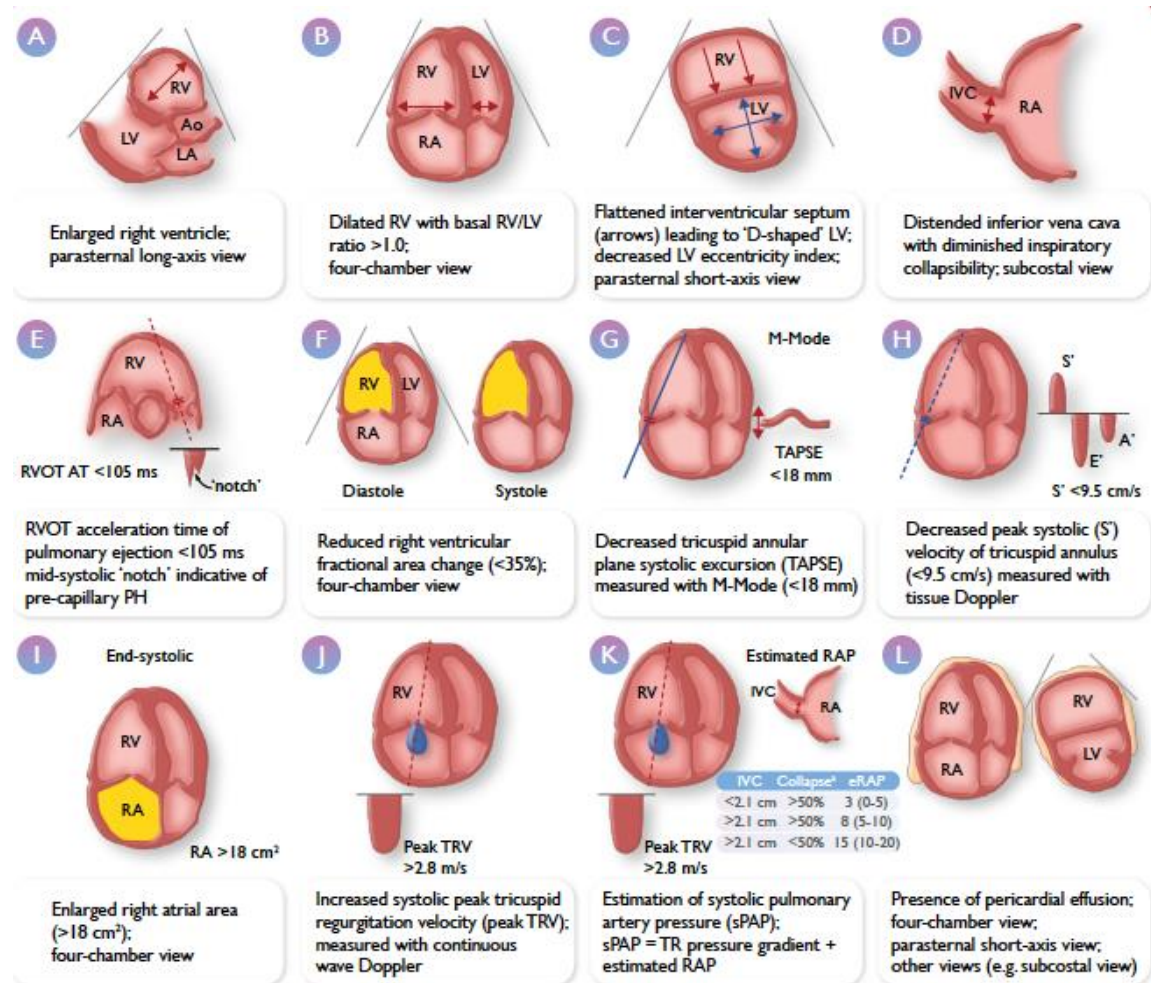


Figure 4. ESC 2022 GI on Pulmonary Hypertension. Transthoracic echocardiographic parameters in the assessment of pulmonary hypertension.

Ao, aorta; IVC, inferior vena cava; LA, left atrium; LV, left ventricle; PH, pulmonary hypertension; RA, right atrium; RAP, right atrial pressure; RV, right ventricle; RVOT AT, right ventricular outflow tract acceleration time; sPAP, systolic pulmonary artery pressure; TAPSE, tricuspid annular plane systolic excursion; TR, tricuspid regurgitation; TRV, tricuspid regurgitation velocity. ^aRefers to collapse on inspiration.

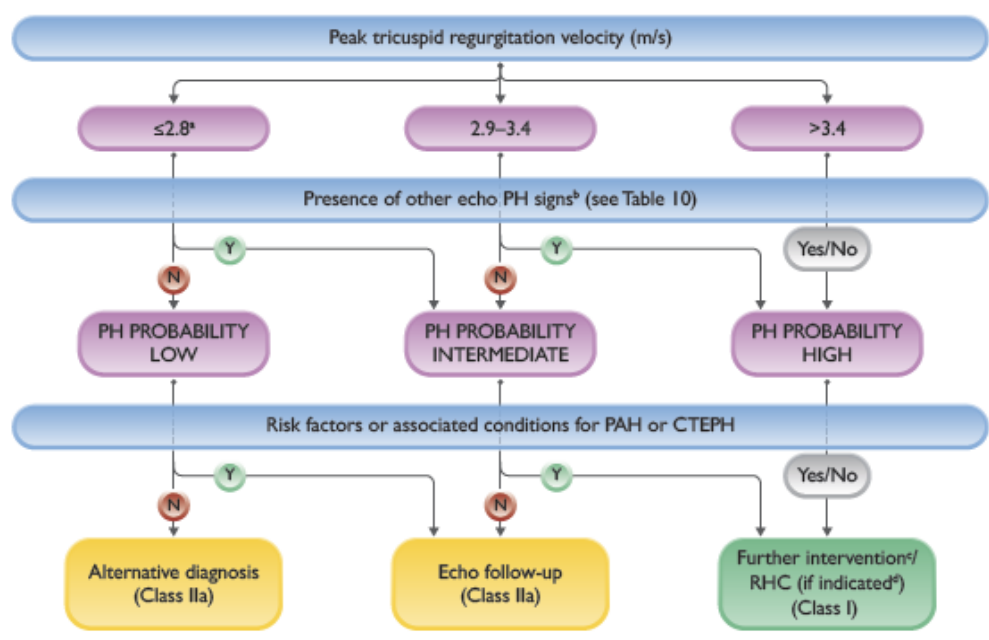


Figure 5. ESC 2022 GI on Pulmonary Hypertension. Echocardiographic probability of pulmonary hypertension and recommendations for further assessment.

CPET, cardiopulmonary exercise testing; CTEPH, chronic thrombo-embolic pulmonary hypertension; echo, echocardiography; LHD, left heart disease; N, no; PAH, pulmonary arterial hypertension; PH, pulmonary hypertension; RHC, right heart catheterization; TRV, tricuspid regurgitation velocity; Y, yes.

a Or unmeasurable. The TRV threshold of 2.8 m/s was not changed according to the updated haemodynamic definition of PH.

b Signs from at least two categories in the Table describing additional echo signs. (A/B/C) must be present to alter the level of echocardiographic probability of PH.

c Further testing may be necessary (e.g. imaging, CPET).

d RHC should be performed if useful information/a therapeutic consequence is anticipated (e.g. suspected PAH or CTEPH), and may not be indicated in patients without risk factors or associated conditions for PAH or CTEPH (e.g. when mild PH and predominant LHD or lung disease are present).

Table 5. ESC 2022 GI on Pulmonary Hypertension. Electrocardiogram abnormalities in patients with pulmonary hypertension

Typical ECG abnormalities in PH

- P pulmonale (P >0.25 mV in lead II)
- Right or sagittal axis deviation (QRS axis >90° or indeterminable)
- RV hypertrophy (R/S >1, with R >0.5 mV in V1; R in V1 + S in lead V5 > 1 mV)
- Right bundle branch block—complete or incomplete (qR or rSR patterns in V1)
- RV strain pattern^a (ST depression/T-wave inversion in the right pre-cordial V1–4 and inferior II, III, aVF leads)
- Prolonged QTc interval (unspecific)^b

ECG, electrocardiogram; PH, pulmonary hypertension; QTc, corrected QT interval; RV, right ventricular.

^aPresent in advanced PH.

^bPatients with pulmonary arterial hypertension can present with a prolonged QTc interval (although non-specific), which may reflect RV dysfunction and delayed myocardial repolarization, and is an independent predictor of mortality.

Table 6. ESC 2022 GI on Pulmonary Hypertension. Additional echocardiographic signs suggestive of pulmonary hypertension

A: The ventricles	B: Pulmonary artery	C: Inferior vena cava and RA
RV/LV basal diameter/area ratio >1.0	RVOT AT <105 ms and/or mid-systolic notching	IVC diameter >21 mm with decreased inspiratory collapse (<50% with a sniff or <20% with quiet inspiration)
Flattening of the interventricular septum (LVEI >1.1 in systole and/or diastole)	Early diastolic pulmonary regurgitation velocity >2.2 m/s	RA area (end-systole) >18 cm ²
TAPSE/sPAP ratio <0.55 mm/mmHg	PA diameter >AR diameter PA diameter >25 mm	

AR, aortic root; IVC, inferior vena cava; LV, left ventricle; LVEI, left ventricle eccentricity index; PA, pulmonary artery; RA, right atrium; RV, right ventricle; RVOT AT, right ventricular outflow tract acceleration time; sPAP, systolic pulmonary arterial pressure; TAPSE, tricuspid annular plane systolic excursion; TRV, tricuspid regurgitation velocity.

a Signs contributing to assessing the probability of PH in addition to TRV. Signs from at least two categories (A/B/C) must be present to alter the level of echocardiographic probability of PH.

Table 7. ESC 2022 GI on Pulmonary Hypertension. Haemodynamic measures obtained during right heart catheterization

Measured variables	Normal value
Right atrial pressure, mean (RAP)	2–6 mmHg
Pulmonary artery pressure, systolic (sPAP)	15–30 mmHg
Pulmonary artery pressure, diastolic (dPAP)	4–12 mmHg
Pulmonary artery pressure, mean (mPAP)	8–20 mmHg
Pulmonary arterial wedge pressure, mean (PAWP)	≤ 15 mmHg
Cardiac output (CO)	4–8 L/min
Mixed venous oxygen saturation (SvO ₂) ^a	65–80%
Arterial oxygen saturation (SaO ₂)	95–100%
Systemic blood pressure	120/80 mmHg
Calculated parameters	
Pulmonary vascular resistance (PVR) ^b	0.3–2.0 WU
Pulmonary vascular resistance index (PVRI)	3–3.5 WU·m ²
Total pulmonary resistance (TPR) ^c	< 3 WU
Cardiac index (CI)	2.5–4.0 L/min·m ²
Stroke volume (SV)	60–100 mL
Stroke volume index (SVI)	33–47 mL/m ²
Pulmonary arterial compliance (PAC) ^d	> 2.3 mL/mmHg

WU, Wood units.

^aDerived from blood sample taken from the pulmonary artery; compartmental oximetry to exclude an intracardiac shunt is recommended when SvO₂ > 75%.

^bPVR, (mPAP–PAWP)/CO.

^cTPR, mPAP/CO.

^dPAC, SV/(sPAP–dPAP).

Table 8. ESC 2022 GI on Pulmonary Hypertension. Recommendations for right heart catheterization and vasoreactivity testing

Right heart catheterization

RHC is performed to confirm the diagnosis of PH (especially PAH or CTEPH) and to support treatment decisions. I B

In patients with suspected or known PH, RHC is performed in experienced centres. I C

RHC comprises a complete set of haemodynamics and is performed following standardized protocols. I C

Vasoreactivity testing

Vasoreactivity testing in patients with I/H/DPAH to detect those who can be treated with high doses of a CCB I B

Vasoreactivity testing is performed at PH centres I C

Consider a positive response to vasoreactivity testing by a reduction in mPAP ≥ 10 mmHg to reach an absolute value of mPAP ≤ 40 mmHg with an increased or unchanged CO^a I C

Inhaled nitric oxide, inhaled iloprost, or i.v. epoprostenol for performing vasoreactivity testing I C

Vasoreactivity testing, for identifying candidates for CCB therapy, is not recommended in patients with PAH other than I/H/DPAH, and in PH groups 2, 3, 4, and 5 III C

CCB, calcium channel blocker; CO, cardiac output; CTEPH, chronic thrombo-embolic pulmonary hypertension; I/H/DPAH, idiopathic, heritable, drug-associated pulmonary arterial hypertension; i.v., intravenous; mPAP, mean pulmonary arterial pressure; PAH, pulmonary arterial hypertension; PH, pulmonary hypertension; RHC, right heart catheterization.

^a Testing should also be performed in patients with a baseline mPAP ≤ 40 mmHg, in whom the same responder criteria apply.

Table 9. ESC 2022 GI on Pulmonary Hypertension. Route of administration, half-life, dosages, and duration of administration of the recommended test compounds for vasoreactivity testing in pulmonary arterial hypertension

Compound	Route	Half-life	Dosage	Duration
Nitric oxide ¹²⁹	inh	15–30 s	10–20 p.p.m.	5–10 min ^a
Iloprost ^{130,131}	inh	30 min	5–10 μg ^b	10–15 min ^c
Epoprostenol ¹²⁹	iv.	3 min	2–12 ng/kg/min	10 min ^d

Inh, inhaled; iv., intravenous.

^aMeasurement as a single step within the dose range.

^bAt mouth piece.

^cMeasurement as a single step, temporize full effect.

^dIncremental increase in 2 ng/kg/min intervals, duration of 10 min at each step.

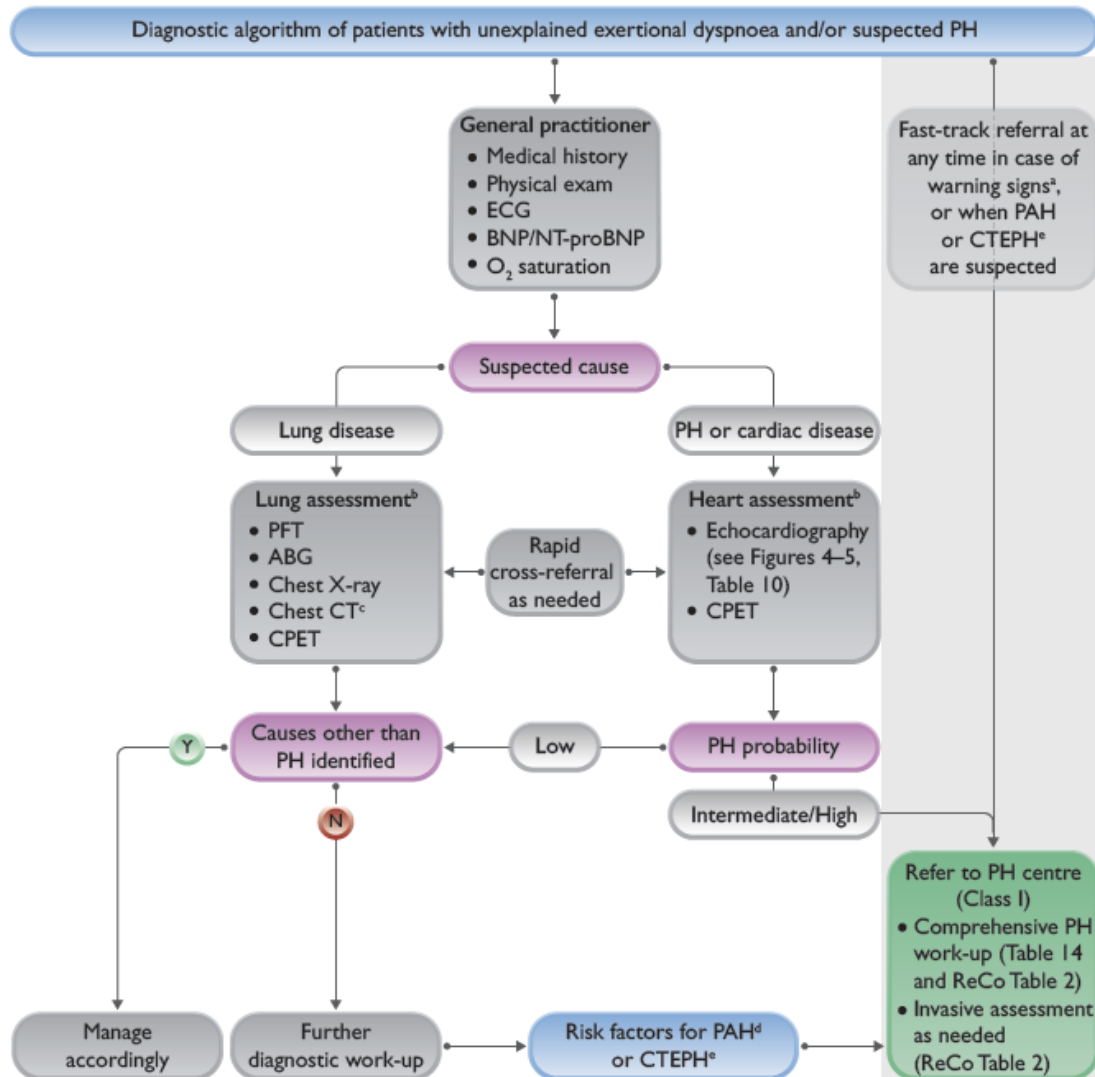


Figure 6. ESC 2022 GI on Pulmonary Hypertension. Diagnostic algorithm of patients with unexplained dyspnoea and/or suspected pulmonary hypertension.

ABG, arterial blood gas analysis; BNP, brain natriuretic peptide; CPET, cardiopulmonary exercise testing; CT, computed tomography; CTEPH, chronic thrombo-embolic pulmonary hypertension; ECG, electrocardiogram; HIV, human immunodeficiency virus; N, no; NT-proBNP, N-terminal pro-brain natriuretic peptide; PAH, pulmonary arterial hypertension; PE, pulmonary embolism; PFT, pulmonary function tests; PH, pulmonary hypertension; ReCo, recommendation; Y, yes.

a Warning signs include rapid progression of symptoms, severely reduced exercise capacity, pre-syncope or syncope on mild exertion, signs of right heart failure.

b Lung and heart assessment by specialist as per local practice.

c As indicated; CT pulmonary angiography recommended if PH suspected.

d Includes connective tissue disease (especially systemic sclerosis), portal hypertension, HIV infection, and family history of PAH.

e History of PE, permanent intravascular devices, inflammatory bowel diseases, essential thrombocythaemia, splenectomy, high-dose thyroid hormone replacement, and malignancy.

Table 10. ESC 2022 GI on Pulmonary Hypertension. Recommendations for diagnostic strategy***Echocardiography***

Echocardiography as the first-line, non-invasive, diagnostic investigation in suspected PH	I B
Assign an echocardiographic probability of PH, based on an abnormal TRV and the presence of other echocardiographic signs suggestive of PH	I B
Maintain the current threshold for TRV (>2.8 m/s) for echocardiographic probability of PH according to the updated haemodynamic definition	I C
Based on the probability of PH by echocardiography, further testing should be considered in the clinical context (i.e. symptoms and risk factors or associated conditions for PAH/ CTEPH)	IIa B
In symptomatic patients with intermediate echocardiographic probability of PH, CPET may be considered to further determine the likelihood of PH	IIb C

Imaging

Ventilation/perfusion or perfusion lung scan in patients with unexplained PH to assess for CTEPH	I C
CT pulmonary angiography in the work-up of patients with suspected CTEPH.	I C
Routine biochemistry, haematology, immunology, HIV testing, and thyroid function tests in all patients with PAH, to identify associated conditions	I C
Abdominal ultrasound for the screening of portal hypertension	I C
Chest CT in all patients with PH	IIa C
Digital subtraction angiography in the work-up of patients with CTEPH	IIa C

Other diagnostic tests

Pulmonary function tests with DLCO in the initial evaluation of patients with PH	I C
Open or thoracoscopic lung biopsy is not recommended in patients with PAH	III C

CPET, cardiopulmonary exercise testing; CT, computed tomography; CTEPH, chronic thrombo-embolic pulmonary hypertension; DLCO, Lung diffusion capacity for carbon monoxide; HIV, human immunodeficiency virus; PAH, pulmonary arterial hypertension; PH, pulmonary hypertension; TRV, tricuspid regurgitation velocity.

Table 11. ESC 2022 GI on Pulmonary Hypertension. Recommendations for screening and improved detection of pulmonary arterial hypertension and chronic thrombo-embolic pulmonary hypertension

Systemic sclerosis

Annual evaluation of the risk of having PAH in patients with SSc	I B
In adult patients with SSc with >3 years' disease duration, an FVC $\geq$ 40%, and a DLCO <60%, the DETECT algorithm to identify asymptomatic patients with PAH	I B
In patients with SSc, where breathlessness remains unexplained following non-invasive assessment, RHC to exclude PAH	I C
Assessing the risk of having PAH based on an evaluation of breathlessness, in combination with echocardiogram or PFTs and BNP/NT-proBNP, in patients with SSc	IIa B
Policies to evaluate the risk of having PAH in hospitals managing patients with SSc	IIa C
In symptomatic patients with SSc, exercise echocardiography or CPET, or CMR to aid decisions to perform RHC	IIb C
In patients with CTD with overlap features of SSc, an annual evaluation of the risk of PAH	IIb C

CTEPH/CTEPD

In patients with persistent or new-onset dyspnoea or exercise limitation following PE, Further diagnostic evaluation to assess for CTEPH/CTEPD	I C
For symptomatic patients with mismatched perfusion lung defects beyond 3 months of anticoagulation for acute PE, referral to a PH/ CTEPH centre after considering the results of echocardiography, BNP/ NT-proBNP, and/or CPET	I C

Other

Counselling regarding the risk of PAH and annual screening in individuals who test positive for PAH-causing mutations and in first-degree relatives of patients with HPAH	I B
In patients referred for liver transplantation, echocardiography as a screening test for PH	I C
Further tests (echocardiography, BNP/ NT-proBNP, PFTs, and/or CPET) in symptomatic patients with CTD, portal hypertension, or HIV to screen for PAH	IIa B

BNP, brain natriuretic peptide; CMR, cardiac magnetic resonance; CPET, cardiopulmonary exercise testing; CTD, connective tissue disease; CTEPD, chronic thrombo-embolic pulmonary disease; CTEPH, chronic thrombo-embolic pulmonary hypertension; DLCO, Lung diffusion capacity for carbon monoxide; FVC, forced vital capacity; HIV, human immunodeficiency virus; HPAH, heritable pulmonary arterial hypertension; NT-proBNP, N-terminal pro-brain natriuretic peptide; PAH, pulmonary arterial hypertension; PE, pulmonary embolism; PFT, pulmonary function test; PH, pulmonary hypertension; RHC, right heart catheterization; SSc, systemic sclerosis.

Table 12. ESC 2022 GI on Pulmonary Hypertension. Characteristic diagnostic features of patients with different forms of pulmonary hypertension

Diagnostic tool	Characteristic findings/features	Group 1 (PAH)	Group 2 (PH associated with left heart disease)	Group 3 (PH associated with lung disease)	Group 4 (PH associated with pulmonary artery obstructions)
5.1.1 Clinical presentation	Clinical features	Variable age, but young, female patients may be predominantly affected. ^{a161} Clinical presentation depends on associated conditions and phenotype See Section 5.1.1	Mostly elderly patients, female predominance in case of HFpEF. ¹⁶¹ History and clinical findings suggestive of LHD	Mostly elderly patients, male predominance. ¹⁶¹ History and clinical findings suggestive of lung disease. Smoking history common	Variable age, but elderly male and female equally affected. History of VTE (CTEPH may occur in the absence of a VTE history). Risk factors for CTEPH See Section 10.1
	Oxygen requirement for hypoxaemia	Uncommon, except for conditions with low DLCO or right-to-left shunting	Uncommon	Common, often profound hypoxaemia in severe PH	Uncommon; common in severe cases with predominantly distal pulmonary artery occlusions
5.1.3 Chest radiography		RA/RV/PA size ↑ Pruning of peripheral vessels	LA/LV size ↑ Cardiomegaly Occasional signs of congestion (interstitial oedema/Kerley lines, alveolar oedema, pleural effusion)	Signs of parenchymal lung disease	RA/RV/PA size ↑ Number and size of peripheral vessels ↓ Occasional signs of pulmonary infarction
5.1.4 Pulmonary function tests and ABG	Spirometry/PFT impairment	Normal or mildly impaired	Normal or mildly impaired	Abnormal as determined by the underlying lung disease	Normal or mildly impaired
	DLCO	Normal or mild-to-moderately reduced (low DLCO in SSC-PAH, PVOD, and some IPAH phenotypes)	Normal or mild-to-moderately reduced, especially in HFpEF	Often very low (<45% predicted)	Normal or mild-to-moderately reduced
	Arterial blood gas PaO ₂ PaCO ₂	Normal or reduced Reduced	Normal or reduced Usually normal	Reduced Reduced, normal, or increased	Normal or reduced Normal or reduced
5.1.5 Echocardiography		Signs of PH (increased sPAP, enlarged RA/RV) Congenital heart defects may be present See Section 5.1.5	Signs of LHD (HFpEF, HFpEF, valvular) and PH (increased sPAP, enlarged RA/RV) See Section 8	Signs of PH (increased sPAP, enlarged RA/RV) See Section 5.1.5	Signs of PH (increased sPAP, enlarged RA/RV) See Section 5.1.5
5.1.6 Lung scintigraphy	Planar – SPECT V/Q	Normal or matched	Normal or matched	Normal or matched	Mismatched perfusion defect
5.1.7 Chest CT		Signs of PH or PVOD See Section 5.1.7	Signs of LHD Pulmonary oedema Signs of PH	Signs of parenchymal lung disease Signs of PH	Intravascular filling defects, mosaic perfusion, enlarged bronchial arteries Signs of PH
5.1.11 Cardiopulmonary exercise testing		High VE/VCO ₂ slope Low P _{ET} CO ₂ , decreasing during exercise No EOv	Mildly elevated VE/VCO ₂ slope Normal P _{ET} CO ₂ increasing during	Mildly elevated VE/VCO ₂ slope Normal P _{ET} CO ₂	High VE/VCO ₂ slope Low P _{ET} CO ₂ , decreasing during
			exercise EOv	increasing during exercise	exercise No EOv
5.1.12 Right heart catheterization		Pre-capillary PH	Post-capillary PH	Pre-capillary PH	Pre- (or post-) capillary PH

ABG, arterial blood gas analysis; CT, computed tomography; CTEPH, chronic thrombo-embolic pulmonary hypertension; DLCO, Lung diffusion capacity for carbon monoxide; EOv, exercise oscillatory ventilation; HFpEF, heart failure with preserved ejection fraction; HFREF, heart failure with reduced ejection fraction; IPAH, idiopathic pulmonary arterial hypertension; LA, left atrium; LHD, left heart disease; LV, left ventricle; PA, pulmonary artery; PaCO₂, partial pressure of arterial carbon dioxide; PAH, pulmonary arterial hypertension; PaO₂, partial pressure of arterial oxygen; P_{ET}CO₂, end-tidal partial pressure of carbon dioxide; PFT, pulmonary function test; PH, pulmonary hypertension; PVOD, pulmonary veno-occlusive disease; RA, right atrium; RV, right ventricle; sPAP, systolic pulmonary arterial pressure; SPECT, single-photon emission computed tomography; SSC-PAH, systemic sclerosis-associated pulmonary arterial hypertension; VE/VCO₂, ventilatory equivalent for carbon dioxide; V/Q, ventilation perfusion scintigraphy; VTE, venous thrombo-embolism.

↓, reduced; ↑, increased.

^aHowever, it may affect individuals of all ages and sexes; diagnosis in males should not be delayed.

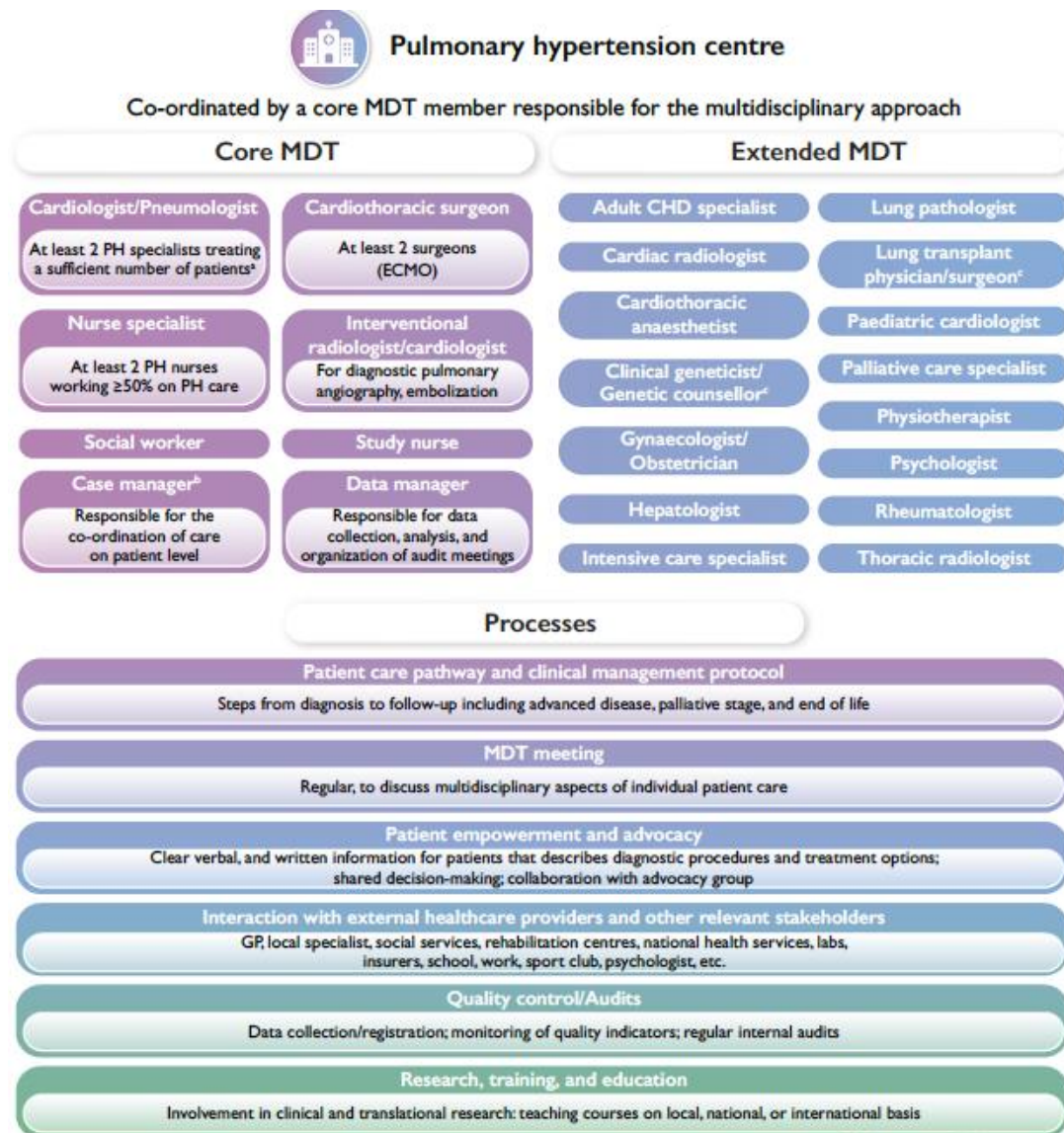


Figure 7. ESC 2022 GI on Pulmonary Hypertension. Pulmonary hypertension centre schematic.

CHD, congenital heart disease; ECMO, extracorporeal membrane oxygenation; GP, general practitioner; MDT, multidisciplinary team; PH, pulmonary hypertension.

a Number adapted according to specific country characteristics.

b Case manager can be a nurse specialist, social worker, physiotherapist, or administrative assistant in function of the centre organization.

c Can be located in partner centres.

Table 13. ESC 2022 GI on Pulmonary Hypertension. Recommendations for pulmonary hypertension centres

PH centres should:

Provide care by a multidisciplinary team (cardiologist, pneumologist, rheumatologist, nurse specialist, radiologist, psychological and social work support, and appropriate on-call expertise)	I C
Have direct links and quick referral patterns to other services (such as genetic counselling, PEA/BPA, LTx, and adult congenital heart disease service)	I C
Maintain a patient registry	I C
Collaborate with patient associations	I C
Accreditation of the PH centres should be considered (e.g. https://ec.europa.eu/health/ern/assessment_en)	IIa C
PH centres' participation in collaborative clinical research	IIa C
PH centres should follow-up a sufficient number of patients to maintain expertise (at least 50 patients with PAH or CTEPH and at least two new referrals per month with documented PAH or CTEPH), and consider establishing collaborations with high-volume centres	IIa C
BPA, balloon pulmonary angioplasty; CTEPH, chronic thrombo-embolic pulmonary hypertension; LTx, lung transplantation; PAH, pulmonary arterial hypertension; PEA, pulmonary endarterectomy; PH, pulmonary hypertension.	