Pulmonary Hypertension

2022 ESC/ERS · The threshold moved — and the group decides the treatment

mPAP > 20 mmHg
lowered from 25

THE DEFINITION CHANGED *numbers that most textbooks still get wrong*

PULMONARY HYPERTENSION

Mean pulmonary arterial pressure > 20 mmHg at rest, measured by right heart catheterization. **Lowered from the long-standing ≥ 25 mmHg**, based on catheterization data in healthy individuals showing the true upper limit of normal is around 20.

PRE-CAPILLARY / PAH

Requires **PVR > 2 Wood units** — **lowered from > 3** — together with **PAWP ≤ 15 mmHg**. Two Wood units is the upper limit of normal and the lowest prognostically relevant threshold.

UNCLASSIFIED PH

A new category: **mPAP > 20 mmHg with PVR ≤ 2 WU and PAWP ≤ 15 mmHg**. Elevated pressure without elevated resistance or left-sided loading — typically a high-flow state. It is described, not treated with PAH drugs.

GROUP IT — THIS IS THE WHOLE POINT *the five groups have opposite treatments*

Group	Cause	Treatment principle
1 · PAH	Idiopathic, heritable, drug-induced, connective tissue disease, congenital heart disease, portopulmonary, HIV	**The only group for PAH-targeted therapy.** Pre-capillary hemodynamics required.
2 · Left heart disease the commonest group	Heart failure with reduced or preserved EF, valve disease	**Treat the left heart. PAH drugs are harmful here** — they can precipitate pulmonary edema by increasing flow into a congested circuit.
3 · Lung disease / hypoxia	COPD, ILD, sleep-disordered breathing, hypoventilation	**Treat the lung disease and correct hypoxemia** with oxygen. PAH drugs are generally not indicated and may worsen V/Q matching.
4 · CTEPH	Chronic thromboembolic disease	**Potentially curable** — see below. Must not be missed.
5 · Multifactorial	Hematologic, systemic, metabolic, sarcoidosis, chronic kidney disease	Treat the underlying condition; targeted therapy is case-by-case.

DIAGNOSE IT PROPERLY *echo screens, catheter confirms, and one test must not be skipped*

1. **Echocardiography is the screening test** — estimated tricuspid regurgitant velocity, right ventricular size and function, and signs of pressure overload. **It cannot make the diagnosis**, only raise the probability.
2. **Right heart catheterization is mandatory** to confirm PH, establish the hemodynamic phenotype, and before committing anyone to targeted therapy. Treating on echo alone is a serious error.
3. **A V/Q scan is required in every patient** to exclude CTEPH. **V/Q is the screening test, not CT pulmonary angiography** — CTPA can miss chronic organized thrombus and gives false reassurance. Missing CTEPH means missing the one potentially curable form.
4. **Then complete the workup:** PFTs with DLCO, HRCT chest, overnight oximetry or sleep study, connective tissue serology, HIV, liver function and abdominal imaging for portal hypertension.
5. **Refer group 1 and group 4 to a specialist PH center.** These are complex, expensive therapies with narrow indications, and outcomes are demonstrably better in expert hands.

TREATING GROUP 1 AND GROUP 4 *risk stratification drives intensity*

PAH (GROUP 1)

Vasoreactivity testing at catheterization in idiopathic, heritable and drug-induced disease — a small minority are responders and do well on **high-dose calcium channel blockers**. Everyone else receives targeted therapy: **endothelin receptor antagonists**, **PDE5 inhibitors or riociguat**, and **prostacyclin pathway agents**. **Initial combination therapy is standard**, with **parenteral prostacyclin for high-risk patients**. Reassess risk regularly and escalate to reach a low-risk profile.

CTEPH (GROUP 4)

Lifelong anticoagulation in everyone. Refer for assessment of **pulmonary endarterectomy**, which is **potentially curative** and the treatment of choice for operable disease. For inoperable or residual disease, **balloon pulmonary angioplasty** and **riociguat**. Every CTEPH patient deserves an operability opinion from an expert center before being declared inoperable.

DECOMPENSATED RIGHT HEART FAILURE *the ward emergency — and it behaves nothing like left heart failure*

Priority	Approach
Find the trigger	**Infection, arrhythmia, non-adherence, pulmonary embolism, anemia, surgery, or pregnancy.** Decompensation is nearly always precipitated.
Optimize volume carefully	Most are **congested and need diuresis**. But the failing right ventricle is **preload-sensitive in both directions** — too much fluid worsens septal shift, too little drops output. **Fluid challenges are hazardous.**
Maintain systemic pressure	**Norepinephrine** is generally the vasopressor of choice — right coronary perfusion depends on systemic pressure exceeding right ventricular pressure. Consider inotropic support with **dobutamine or milrinone**. **Hypotension here is a spiral**, not a number to tolerate.
Avoid the aggravators	**Hypoxia, hypercapnia and acidosis all raise pulmonary vascular resistance** — correct them promptly. Restore **sinus rhythm** where possible; these patients depend heavily on atrial contribution and tolerate tachyarrhythmia poorly.
Intubation is high risk	**Induction and positive pressure can precipitate arrest.** Sedatives drop preload and systemic pressure while positive pressure raises right ventricular afterload. **Avoid intubation where possible**; if unavoidable, have vasopressors running and the most experienced operator present.

PEARLS & PITFALLS

- **The thresholds moved.** mPAP > 20 and PVR > 2 WU, not 25 and 3 — older references and many local protocols are out of date.
- **Do not give PAH drugs for group 2 or 3.** In left heart disease they can precipitate pulmonary edema, and in lung disease they worsen gas exchange.
- **V/Q scan, not CTPA, screens for CTEPH.** A normal CTPA does not exclude it.
- **Never diagnose or treat on echo alone.** Right heart catheterization defines the phenotype and the phenotype defines the treatment.
- **Syncope or chest pain in PH is ominous** — it signals a failing right ventricle and warrants urgent specialist input.
- **Be very careful with anesthesia and fluids.** These patients tolerate hypotension, hypoxia and hypercapnia poorly, and induction can precipitate right ventricular failure and arrest.

References: 2022 ESC/ERS Guidelines for the diagnosis and treatment of pulmonary hypertension (Eur Respir J / Eur Heart J, 2022) · ACC key points summary of the 2022 guideline.

Educational aid; verify against local specialist protocol and patient factors.

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