REBELLIOUS MEDIC · RESPIROLOGY ALGORITHM SERIES

Pulmonary Hypertension

Diagnostic workup, risk stratification, and group-specific management (PAH, left heart disease, lung disease, CTEPH) for adults with suspected or confirmed pulmonary hypertension.

2022 ESC/ERS Guidelines
Diagnosis and Treatment of Pulmonary Hypertension
ESC/ERS Task Force · Eur Heart J 2022;43:3618–3731

COR — I · Recommended | IIa · Reasonable | IIb · May Consider | III · Not Recommended

1 Diagnosis — RHC & Vasoreactivity

COR / LOE	
I B/C	**RHC** confirms diagnosis and supports treatment decisions; perform at experienced centres with a standardized hemodynamic protocol.
I B/C	**Vasoreactivity testing** in I/H/DPAH to identify CCB candidates, at PH centres, using inhaled NO, iloprost, or IV epoprostenol.
I C	**Positive response** = mPAP fall ≥10 mmHg to ≤40 mmHg with increased/unchanged CO.
III C	Vasoreactivity testing **not indicated** outside I/H/DPAH or in groups 2–5.

2 Diagnostic Strategy & Screening

COR / LOE	
I B	**Echocardiography** first-line non-invasive test; assign PH probability from TRV plus supportive echo signs.
I C	V/Q or perfusion scan + CTPA for CTEPH workup; biochemistry/hematology/HIV/thyroid + abdominal US in all PAH; PFTs with DLCO.
I B/C	**Systemic sclerosis:** annual PAH risk screening; DETECT algorithm if >3 yr disease, FVC ≥40%, DLCO <60%; RHC if unexplained dyspnea.
I C	**CTEPH screening:** evaluate persistent dyspnea after PE; refer to PH/CTEPH centre for mismatched perfusion defects beyond 3 mo anticoagulation.

3 Risk Stratification

COR / LOE	
I B	**Multiparametric** assessment from clinical, exercise, biochemical, echo, and hemodynamic data; low-risk profile is the treatment goal.
I B	**At diagnosis:** three-strata model (low/intermediate/high), incorporating hemodynamics. **Follow-up:** four-strata model using WHO-FC, 6MWD, BNP/NT-proBNP.

4 General Measures & Special Circumstances

COR / LOE	
I A/C	**Supportive care:** supervised exercise training; psychosocial support; immunization; diuretics for RV failure; LTOT if PaO2 <8 kPa; correct iron deficiency.
III C	**Avoid** ACEi/ARB/ARNI/SGLT2i/beta-blockers/ivabradine unless required by comorbidity.
I C	**Pregnancy:** counsel against pregnancy, provide contraceptive advice, refer to an experienced PH centre.
III Harm · B	**ERAs and riociguat are teratogenic** — avoid in pregnancy.

5 Vasoreactive PAH — CCB Therapy

COR / LOE	
I C	**Responders:** high-dose CCB for I/H/DPAH vasoreactivity responders; reassess with RHC at 3–4 months.
I C	**Continue vs. switch:** continue CCB if WHO-FC I/II with marked hemodynamic improvement (mPAP <30, PVR <4 WU); switch to PAH therapy if FC III/IV or no improvement.
III C	**Non-responders:** CCBs not recommended without a vasoreactivity study or in non-responders.

6 Initial & Sequential Combination Therapy

COR / LOE	
I B	**Initial combo:** ambrisentan + tadalafil, or macitentan + tadalafil.
III B	Upfront **triple therapy** (macitentan+tadalafil+selexipag) not recommended.
I C	**Escalation:** base treatment escalation on risk assessment per the treatment algorithm.
I B	**Add-on (mortality benefit):** macitentan to PDE5i/prostacyclin analogue; selexipag to ERA/PDE5i; oral treprostinil to ERA or PDE5i/riociguat.
I B	**Add-on (exercise):** add sildenafil to epoprostenol to improve exercise capacity.
III B	**Avoid combos:** bosentan + sildenafil, and riociguat + PDE5i, not recommended.

7 Associated PH Groups — LHD, Lung Disease, CTEPH

GROUP 2 — LEFT HEART DISEASE

COR / LOE	
I A	Optimize underlying cardiac disease before PH workup.
III A	PAH-approved drugs not recommended.

GROUP 3 — LUNG DISEASE

COR / LOE	
I C	Optimize lung disease/hypoxemia/sleep-disordered breathing; refer to PH centre if severe PH.
III B/C	Avoid ambrisentan in IPF, riociguat in IIP, and PAH meds in non-severe PH.

GROUP 4 — CTEPH WORKUP

COR / LOE	
I C	Lifelong anticoagulation for all; test for antiphospholipid syndrome; multidisciplinary CTEPH team review.

GROUP 4 — CTEPH TREATMENT

COR / LOE	
I B	**PEA** is treatment of choice if surgically accessible; **BPA** if inoperable or residual PH; **riociguat** if inoperable or persistent/recurrent PH after PEA.

8 Referral, Transplant & PH Centres

COR / LOE	
I C	**LTx referral/listing:** refer if inadequate response to oral combo therapy with intermediate-high/high risk or REVEAL >7; list if high risk or REVEAL ≥10 despite optimized therapy incl. parenteral prostacyclin.
I C	**ICU right HF:** involve PH-experienced physicians; treat causative factors; use inotropes/vasopressors, fluid management, and PAH drugs as appropriate.

PH centres: multidisciplinary team, patient registry, quick-referral links to genetics/PEA-BPA/LTx/ACHD services, collaboration with patient associations (I, C).

Humbert M, Kovacs G, Hoeper MM, et al. 2022 ESC/ERS Guidelines for the Diagnosis and Treatment of Pulmonary Hypertension. *Eur Heart J.* 2022;43(38):3618–3731.