



PAH Clinical Scenarios: Advances in Diagnostic Assessment, Guideline Recommended Care, and Emerging Treatments

Clinical Brief #2 – Treating PAH

References

Tweet 2 References:

Humbert M, Kovacs G, Hoeper MM, et al. 2022 ESC/ERS guidelines for the diagnosis and treatment of pulmonary hypertension. *Eur Heart J*. 2022;43:3618-3731.

Humbert M, Kovacs G, Hoeper MM, et al. 2022 ESC/ERS guidelines for the diagnosis and treatment of pulmonary hypertension. *Eur Respir J*. 2023;61:2200879.

Additional References:

Chin KM, Sitbon O, Doelberg M, et al. Three- versus two-drug therapy for patients with newly diagnosed arterial hypertension. *J Am Coll Cardiol*. 2021;78:1393-1403.

Galiè N, Humbert M, Vachiery J-L, et al. 2015 ESC/ERS Guidelines for the diagnosis and treatment of pulmonary hypertension. *Eur Heart J*. 2016;37:67-119.

Humbert M, Kovacs G, Hoeper MM, et al. 2022 ESC/ERS guidelines for the diagnosis and treatment of pulmonary hypertension. *Eur Heart J*. 2022;43:3618-3731.

Humbert M, Kovacs G, Hoeper MM, et al. 2022 ESC/ERS guidelines for the diagnosis and treatment of pulmonary hypertension. *Eur Respir J*. 2023;61:2200879.

Leeper B, Powell B. Pulmonary arterial hypertension. *Nurs Crit Care*. 2019;14:14-22.

Vasquez ZG, Klinger JR. Guidelines for the treatment of pulmonary arterial hypertension. *Lung*. 2020;198:581-596.



Glossary

6MWD, 6-minute walk distance
AE, adverse event
BNP, brain natriuretic peptide
cAMP, cyclic adenosine monophosphate
cGMP, cyclic guanosine monophosphate
CHD, coronary heart disease
cMRI, cardiac magnetic resonance imaging
CPET, cardiopulmonary exercise testing
CTD, connective tissue disease
CV, cardiovascular
DLCO, diffusing capacity of the lungs for carbon monoxide
ERA, endothelin receptor antagonist
ERS, European Respiratory Society
ESC, European Society of Cardiology
FC, functional class
GMP, guanosine monophosphate
GTP, guanosine-5'-triphosphate
I/H/D-PAH, idiopathic, hereditary, or drug-induced pulmonary arterial hypertension
IP, prostacyclin receptor
INH, inhaled
IV, intravenous
NO, nitric oxide
NT-proBNP, N-terminal pro-brain natriuretic peptide
PAH, pulmonary arterial hypertension
PCA, prostacyclin analogue
PDE-5i phosphodiesterase-5 inhibitor
PGI₂, prostaglandin 12
PH, pulmonary hypertension
PRA, prostacyclin receptor agonist
PVR, pulmonary vascular resistance
RCT, randomized, controlled trial
REMS, Risk Evaluation and Mitigation Strategies
SC, subcutaneous
sGC, soluble guanylate cyclase
T2DM, type 2 diabetes mellitus
w/, with
WHO, World Health Organization