PAH and PAH-LHD: New Treatment Insights

New 2022 PAH Treatment Algorithm

The European Society of Cardiology (ESC) and the European Respiratory Society (ERS) jointly released updated guidelines (ESC/ERS Guidelines) for the diagnosis and treatment of pulmonary hypertension (PH), with an emphasis on diagnosing and treating pulmonary arterial hypertension (PAH) and chronic thromboembolic pulmonary hypertension (CTEPH) (Humbert, Kovacs et al. 2022). These guidelines include several changes (Figure 1):

- An updated treatment algorithm for PAH that highlights the importance of cardiopulmonary comorbidities and risk assessment at diagnosis and follow-up
- A focus on the role of combination therapies
    - Combination therapy with a phosphodiesterase 5 inhibitor (PDE-5i), an endothelin receptor antagonist (ERA), and/or prostacyclin analogues should be considered in patients with idiopathic, heritable, or drug-associated PAH, depending on the severity of presentation
    - In those patients presenting with intermediate-low risk of death while receiving ERA/PDE-5i therapy, switching from PDE-5i to riociguat may be considered

New 2022 PH-LHD Recommendations

In addition to the new PAH treatment algorithm, the ESC/ERS Guidelines include 84 new recommendations across every category of recommendation. Among the most significant changes are the following:

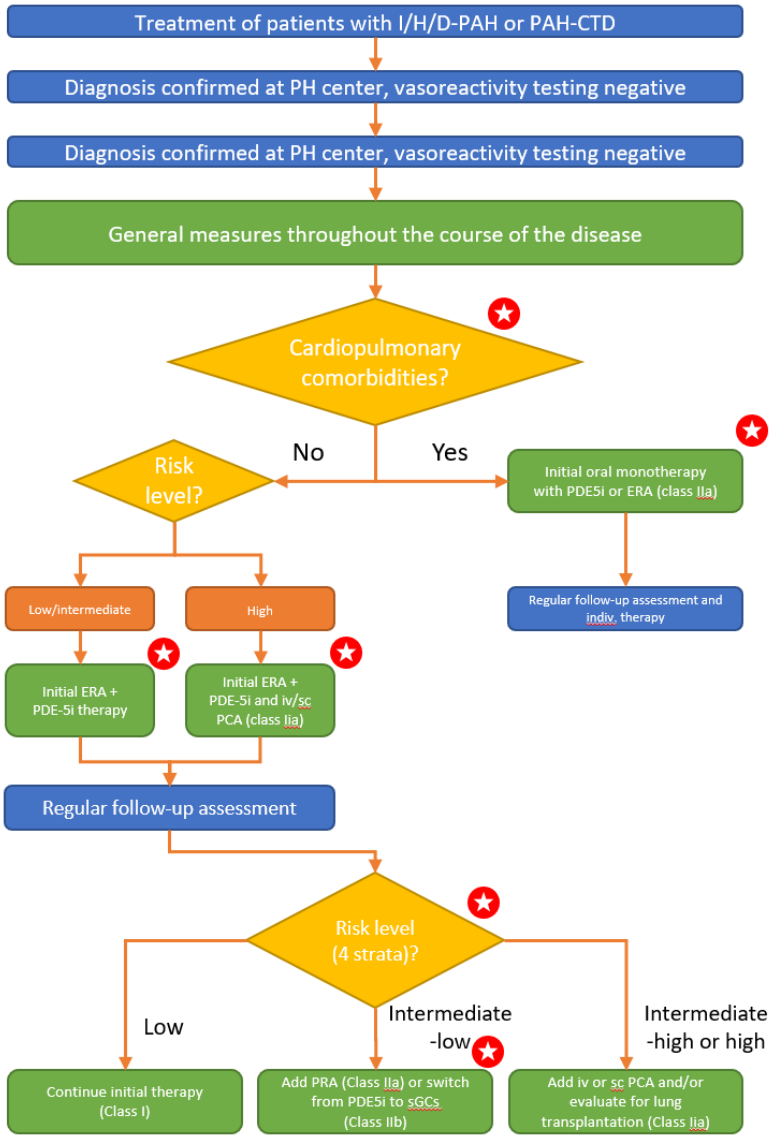
- Four (4) recommendations related to diagnostic strategy
- Five (5) new and one (1) revised recommendation for women of childbearing potential
- Five (5) new recommendations for the treatment of non-vasoreactive patients with idiopathic, heritable, or drug-associated PAH who present with and without cardiopulmonary comorbidities
- New recommendations for patients who present with PAH associated with drugs or toxins (4 recommendations), connective tissue disease (1 recommendation), or human immunodeficiency virus (HIV, 2 recommendations)
- Five (5) new recommendations regarding shunt closure in patients
- Five (5) new recommendations for PH associated with left heart disease

The ESC/ERS Guidelines also describe other new content in this version of the guidelines:

- Reconsideration of the hemodynamic definition of PH
- An updated classification of PH
- Expansion of the risk-stratification table

Finally, throughout the document the authors address questions with direct consequences for clinical practitioners for specific subgroups of PH, including initial treatment, use of an oral PDE-5i, and the use of PH drugs prior to balloon pulmonary angioplasty.

Figure 1: Evidence-based PAH treatment algorithm for patients with idiopathic, heritable, drug-associated, and connective tissue disease-associated PAH[1]



Adapted from Humbert M, Kovacs G, Hoeper MM, et al. 2022 ESC/ERS Guidelines for the diagnosis and treatment of pulmonary hypertension: Developed by the task force for the diagnosis and treatment of pulmonary hypertension of the European Society of Cardiology (ESC) and the European Respiratory Society (ERS). Endorsed by the International Society for Heart and Lung Transplantation (ISHLT) and the European Reference Network on rare respiratory diseases (ERN-LUNG). European Heart Journal. 2022;doi:10.1093/eurheartj/ehac237.

[1] ★Denotes changes to guideline. ERA, endothelin receptor antagonist; I/H/D-PAH, idiopathic, heritable, or drug-associated pulmonary arterial hypertension; iv, intravenous; PAH-CTD, PAH associated with connective tissue disease; PCA, prostacyclin analogue; PDE-5i, phosphodiesterase 5 inhibitor; PH, pulmonary hypertension; PRA, prostacyclin receptor agonist; sc, subcutaneous; sGCs, soluble guanylate cyclase stimulator.

References

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