

Pulmonary Hypertension Diagnosis and Management

Several tools and investigations are available to help diagnose pulmonary hypertension (PH). Echocardiogram is usually the initial test while right heart catheterization confirms PH when pulmonary artery pressures are above 20 mmHg at rest. Chest X-ray can also clue the clinician. Management for PH includes treating the underlying disorder. Several pharmacotherapies are available such as calcium channel blockers, endothelin receptor antagonists, prostacyclin analogs, and PDE-5 inhibitors.



PLAY PICMONIC

Diagnosis

Echocardiogram

[Echoing Cardiogram](#)

An echocardiogram is the best initial diagnostic test for pulmonary hypertension. It can help detect right ventricular hypertrophy or dilation and can estimate pulmonary artery pressures.

Right Heart Catheterization

[Right Heart Cathether-cat](#)

Right heart catheterization (RHC) is the best confirmatory test for pulmonary hypertension. This involves a wire introduced from a peripheral vein and guided to the pulmonary artery after which a balloon is inflated at the tip that helps measure the pressure.

≥ 20 mmHg at Rest

[Greater-than \(20\) Dollar-bill on Bed-rest-bed](#)

A mean pulmonary artery pressure (mPAP) above 20 mmHg at rest is diagnostic of pulmonary hypertension as mentioned in the proceedings of the 6th World Symposium on Pulmonary Hypertension. Normal mean pulmonary artery pressure is 10–14 mm Hg.

Chest X-Ray

[Chest X-Ray](#)

90% of patients with pulmonary arterial hypertension have an abnormal chest X-ray. Findings include enlarged right atrium, elevated cardiac apex (right heart enlargement sign), enlarged pulmonary arteries, "pruning" of peripheral pulmonary vessels, and an enlarged pulmonary trunk.

MANAGEMENT

Treat Underlying Disorder

[Treating Disorders Under the Table](#)

Treating the underlying cause of pulmonary hypertension will help alleviate symptoms and might bring down pulmonary artery pressures. The classification of PH is discussed in a separate Picmonic.

Calcium Channel Blockers

Calcium-channel Blockers

Patients who respond well to vasodilators after an initial vasoreactivity test are often administered calcium channel blockers first.

Endothelin Receptor Antagonists

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Endothelin receptor antagonists work by blocking the endothelin-1 receptor. This action inhibits the vasoconstricting effects of endothelin-1. Bosentan, macitentan, and ambrisentan are examples of these drugs.

Prostacyclin Analogs

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Prostacyclin (PGI_2) acts as a potent vasodilator on pulmonary and systemic arterial vascular beds, inhibits proliferation of vascular smooth muscle cells, and inhibits platelet aggregation. Epoprostenol and iloprost are prostacyclin analogs that are used for pulmonary arterial hypertension.

PDE-5 inhibitors

Phosphodiesterase-5 Inhibitors

Phosphodiesterase type 5 (PDE-5) inhibitors work by inhibiting PDE-5 which normally degrades cGMP. This results in an increasing amount of cGMP thus prolonging the vasodilatory effects of nitric oxide (NO). Sildenafil and tadalafil are well known PDE-5 inhibitors.