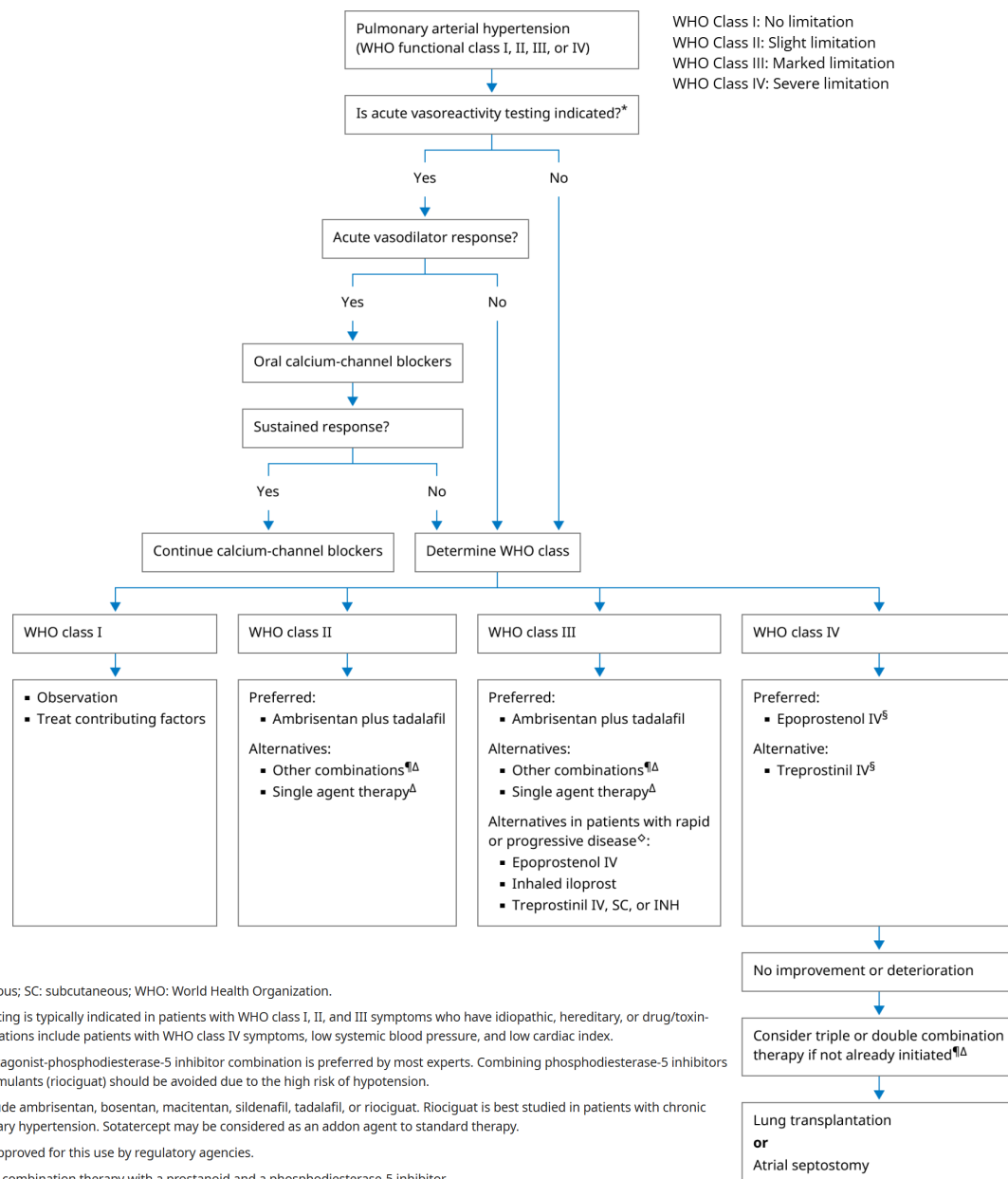


Pulmonary Arterial Hypertension

Pulmonary Arterial Hypertension (PAH) is a rare disorder characterized by **elevated blood pressure in the lungs** caused by narrowing of small pulmonary arteries. This increased vascular resistance forces the heart to work harder to pump blood, leading to reduced cardiac effectiveness over time. PAH affects approximately 15 to 50 people per million in the United States and Europe. The condition places a **significant economic burden** on healthcare systems due to the high cost of medications and frequent hospitalizations. Mortality and morbidity in PAH are influenced by factors such as medication adherence, patient monitoring, and disease severity.

PAH Treatment Algorithm



PAH Medications

Drug Class	Drug	Route	Adverse Effects	Cost of 90D
Prostacyclin Pathway Agonists	Epoprostenol	IV	Hypotension, hypoxemia, severe nausea/vomiting	\$1,129.41
	Treprostinil	IV, SQ, Inhaled, Oral	Hypotension, jaw pain, flushing	*Inhaled: \$31,070.04 Oral: \$16,862.85
	Iloprost	Inhaled		\$4,200.00
	Selexipag	IV, Oral	Flushing, diarrhea, headache, jaw pain	\$43,101.30
Endothelin Receptor Antagonists	Bosentan	Oral	Hepatotoxicity, flushing, edema, anemia, pregnancy category X	\$5,052.03
	Ambrisentan			\$4,818.26
	Macitentan			\$26,455.23
Nitric Oxide-cGMP Enhancers	Riociguat	Oral	Dizziness, headache, syncope, pregnancy category X	\$12,475.24
Phosphodiesterase Type 5 Inhibitors	Sildenafil	IV, Oral	Headache, flushing, epistaxis, vision changes	\$17.04
	Tadalafil	Oral		\$370.83
TGF-beta Receptor Modulator / Activin Trap	Sotatercept	SQ	Erythrocytosis, thrombocytopenia	~ \$14,000
Calcium Channel Blockers	Nifedipine ER	Oral	Systemic hypotension & peripheral edema	\$10.05
	Diltiazem ER			\$132.55
	Amlodipine			\$2.66

*Consists of Nebulizer and DPI formulations

In 2023-2024, Arkansas Health Network (AHN) treated 103 patients with PAH, incurring an **annual cost of \$2,066,668** for drug therapies. Our analysis of the patients responsible for this expenditure revealed **> 80% appropriate medication use**, confirmed by PAH diagnoses, medication adherence, and therapy aligned with WHO classifications. These patients are under close supervision by their PCPs, cardiologists, and pulmonologists. Therefore, we do not identify an immediate need or intervention to more cost-effective therapies for this patient group.

Disclaimer: This information is not intended to dictate or substitute for the professional judgment of a health care practitioner in a particular case. Please verify with insurance availability on formulary

Sources

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