American Lung Association.

PAH Medication and Treatment Guide

Pathways	Type of Medication	Medication Names	What it does	Common Side Effects	Delivery Method	Additional Information
Nitric Oxide	Phosphodiesterase Inhibitors (PDE5 Inhibitors)	Tadalafil, Sildenafil	Helps relax and widen blood vessels in the lungs, making it easier for blood to flow through them	Headache, flushing (warmth and redness in the face, neck and upper chest), dizziness, upset stomach, runny or stuffy nose, muscle pain, back pain	Oral	May improve ability to do physical activity and help slow disease from getting worse
	Soluble Guanylate Cyclase Stimulators (SGCS)	Adempas®, Riociguat	Helps relax and widen blood vessels in the lungs, allowing blood to flow more easily	Headache, indigestion, nausea, dizziness, low blood pressure, anemia, constipation *Note: Riociguat must not be used at the same time as PDE5 inhibitors (e.g. sildenafil or tadalafil) due to the risk of significant hypotension.	Oral	May improve ability to do physical activity and help slow the disease from getting worse

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Endothelin	Endothelin Receptor Antagonists (ERAs)	Bosentan, Ambrisentan, Macitentan	Helps relax and widen blood vessels in the lungs, making it easier for blood to flow and reducing strain on the heart	Headache, swelling in ankles, feet, legs and abdomen, flushing, cold-like symptoms, unusual or rapid weight gain or weight loss, tingling of hands or feet *Note: Bosentan, ambrisentan and macitentan can cause harm to an unborn baby. Talk to your provider about potential risks and discuss family planning, including the need for effective birth control	Oral	May improve ability to do physical activity and helps slow disease progression
Prostacyclin	Prostacyclin Analogue	Epoprostenol (Flolan®, Veletri®), Treprostinil (Remodulin®, Orenitram®, Tyvaso®), Iloprost (Ventavis®), Tyvaso DPI (inhaled)	Helps relax and widen blood vessels in the lungs, making it easier for blood to flow through them	Flushing, headache, muscle pain, cough, diarrhea, nausea, vomiting, indigestion, throat irritation, flu-like symptoms, rash	Oral, inhaled, IV or subcutaneous	May improve ability to do physical activity, reduce symptoms and help slow disease progression

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Prostacyclin	Selective IP Receptor Agonist	Selexipag (Uptravi®)	Helps relax and widen blood vessels in the lungs	Headache, jaw pain, muscle pain, joint pain, flushing, rash, anemia, nausea	Oral, Delivery method is only Oral	May slow disease from getting worse
Biologics	Sotatercept-(csrk) (Winrevair)	Sotatercept-csrk (Winrevair)	Helps reduce the risk of PAH worsening events, including hospitalization and the need for transplantation, or death	Headache, nosebleed, rash, tiny blood vessels that look pink or red on the skin, diarrhea, dizziness, redness *Note: While taking this medication, you will need regular blood tests to monitor your blood cell and platelet levels and check for an increase in red blood cells. Your healthcare provider will want to monitor you closely to ensure this treatment remains safe for you.	Subcutaneous	Improves ability to do physical activity, reduces the risk of physical condition and symptoms getting worse
Combination Medication (ERA/PDE5i)	Macitentan and Tadalafil	Opsynvi®	Helps prevent narrowing and remodeling of blood vessels in the lungs	Edema/fluid retention, anemia, and headache migraine	Oral	May help slow disease progression, reduce the risk of hospitalizations for PAH and improve ability to do physical activity