

Pathways	Type of Medication	Medication Names	What it does	Common Side Effects	Delivery Method	Additional Information
Nitric Oxide	Phosphodiesterase Inhibitors (PDE5 Inhibitors)	Tadalafil, Sildenafil	Allows the lungs to relax and the blood vessels to expand, making it easier for your heart to pump blood through your body	Headache, flushing, lightheaded, nausea, runny or stuffy nose, muscle pain, back pain	Oral, subcutaneous (short-term only for subcutaneous)	Slows disease from getting worse, improves ability to do physical activity
	Soluble Guanylate Cyclase Stimulators (SGCS)	Adempas, Riociguat	Allows the blood vessels in the lungs to relax helping more blood to flow through	Headache, indigestion, gastritis, dizziness, low blood pressure, anemia GERD, constipation, bleeding	Oral	Slows disease from getting worse, improves ability to do physical activity
Endothelin Receptor Antagonists (ERAs)	Endothelin Receptor Antagonists (ERAs)	Bosentan, Ambrisentan, Macitentan	Lowers blood pressure in the lungs, helping heart to pump blood more efficiently	Headache, joint pain, low blood pressure, fainting, flushing, irregular heartbeat, cold symptoms	Oral	Improves ability to do physical activity, slows disease from getting worse, relieves symptoms
Prostaglandin	Prostacyclin Analogue	Epoprostenol, Flolan, Iloprost, Orenitram, Treprostinil, Treprostinil Inhaled, Remodulin, Tyvaso, Veletri, Ventavis	Allows the blood vessels in the lungs to relax; inhaled treatments relieve shortness of breath	Flushing, headache, muscle pain, cough, diarrhea, nausea, vomiting, indigestion, throat irritation, flu-like symptoms, rash	Oral, inhaled, IV or subcutaneous	Improves ability to do physical activity

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Nitric Oxide	Selective IP Receptor Agonist	Selexipag	Allows the blood vessels in the lungs to relax	Headache, jaw pain, muscle pain, joint pain, flushing, rash, anemia, nausea, pain/redness at injection site if injected	Oral, subcutaneous	Slows disease from getting worse
	Prostacyclin Mimetics	Tyvaso DPI (Inhaled)	Relaxes blood vessels, helping to widen the narrowed blood vessels in the lungs for better blood flow	Cough, headache, throat irritation, nausea, flushing, dizziness	Inhaled	Improves ability to do physical activity, slows worsening of symptoms
Biologics	Sotatercept-(csrk) (Winrevair)	Sotatercept-csrk (Winrevair)	Works to correct imbalances that impact the structure of your blood vessels	Headache, nosebleed, rash, tiny blood vessels that look pink or red on the skin, diarrhea, dizziness, redness	Subcutaneous	Improves ability to do physical activity, reduces the risk of physical condition and symptoms getting worsening
Combination Medication (ERA/PDE5i)	Macitentan and Tadalafil	Opsynvi	Counteracts the thickening of the arterial wall and organ damage; promotes muscle relaxation and widening of blood vessels	Edema/fluid retention, anemia, and headache migraine	Oral	Slows disease from getting worse and decreases risk of hospitalizations, improves ability to do physical activity