Management of Pulmonary Arterial Hypertension (PAH)

Your provider may prescribe one or multiple medications depending on the cause of pulmonary hypertension, how you tolerate the medications, and the stage of the disease.

**Oxygen Therapy**: Replaces the low oxygen in your blood. Oxygen may be prescribed for use when sleeping and throughout the day.

**Anticoagulants or "blood thinners"**: To prevent blood clots from forming or getting larger. These medications require regular labs for monitoring, and taking them increases the risk of bleeding. Drug names: Coumadin (Warfarin).

**Diuretics or "water pills"**: To help remove extra fluid from legs, lungs, and heart and make breathing easier. Drug names: Furosemide (Lasix), Bumetadine (Bumex), Torsemide (Demadex).

**Potassium**: To replace low potassium levels in the blood. Potassium is important for heart function. Drug names: K-Dur, Potassium Chloride.

**Magnesium**: To replace low magnesium levels in the blood. Magnesium is important for heart function. Drug names: Magnesium Oxide, Magnesium Complex.

**Digoxin**: To help improve heart strength and pumping function. It can also be used to regulate heart rate and some abnormal heart rhythms. Drug names: Digoxin, Lanoxin.

**Aldosterone Antagonists**: In addition to removing fluid from the body, this medication helps to keep potassium in the blood and helps the heart to work better. Drug names: Spironolactone (Aldactone), Eplerenone (Inspra).

**Additional Measures**:

**Immunizations**: Get an annual influenza vaccine (flu shot). Stay up to date on pneumococcal vaccine (pneumonia shot). It is critical to prevent infections, because infections such as the flu or pneumonia can cause serious complications for a patient with lung disease.

**Lifestyle**: Diet, exercise, and healthy habits such as avoiding alcohol, drugs, and not smoking can also help to improve your symptoms and health outcomes. Please discuss safe exercise and healthy diet options with your provider.
Specialty Medications to treat Pulmonary Arterial Hypertension (PAH)

- The following medications may be prescribed by your provider after a thorough review of PAH-specific test results.
- Your provider may prescribe one or multiple medications depending on the cause of pulmonary hypertension, how you tolerate the medications, and stage of disease.
- These medications help improve breathing and symptoms of PAH.
- The doses may need to be adjusted over time.
- These medications should be taken regularly and as directed. Some require lab tests before starting and while you take the medication.

**Calcium Channel Blockers:** only a very small percentage of patients will benefit from this kind of medication. It can lower pulmonary pressure and can help to improve the pumping function of the right side of the heart.

**Type of medication:** tablet, taken by mouth.

**Possible side effects:** swelling in your legs, can lower your blood pressure

**Drug names:** Nifedipine (Procardia), Diltiazem (Cardizem), Amlodipine (Norvasc) and others.

**PDE-5 Inhibitors:** Dilates (opens) blood vessels throughout the body, including the heart and lungs.

**Type of medication:** Tablet, taken by mouth.

**Possible side effects:** headache, flushing, heartburn, muscle pain, and dizziness.

**Drug names:** Sildenafil (Viagra, Revatio), Tadalafil (Cialis, Adcirca).

**Note:** PDE-5 Inhibitor medications have a dangerous interaction with other medications containing nitrates (such as nitroglycerin, isosorbide dinitrate). Please notify care providers if you are taking any of these medications.

**Endothelin Receptor Antagonists:** Endothelin makes blood vessels narrow. These medications block endothelin to help keep blood vessels open.

**Type of medication:** Tablet, taken by mouth.

**Possible side effects:** nasal congestion, headache, swelling in legs or weight gain. Other possible side effects that your provider will watch for: anemia, liver function abnormalities, and fetal birth defects.

**Drug names:** Bosentan (Tracleer), Ambrisentan (Letairis), Macitentan (Opsumit).

**Note:** These medications require periodic blood tests to check liver function and for pregnancy. Women of childbearing age are required by the FDA to be on a reliable method of birth control while on this medication. Your provider will discuss these options before starting this medication.
Prostacyclins: Dilate (open) pulmonary arteries.

Type of medication: liquid--inhaled with nebulizer or given by continuous intravenous (IV) or subcutaneous (SQ) infusion, tablet (this formulation has some restrictions, that your provider can discuss with you).

Possible side effects: Headache, jaw pain, flushing, nausea, diarrhea, cough (when inhaled).

Drug names: Epoprostenol (Flolan, Veletri), Treprostinil sodium (Remodulin, Tyvaso), Iloprost (Ventavis),

Note: These medications may be started in the clinic or in the hospital to ensure safety and monitoring when the medication is started.

Soluable Guanylate cyclase (sGC) Stimulant: Stimulates cells to open the blood vessels that connect the heart and lungs and also can help prevent narrowing of the blood vessels and may aid in preventing blood clot formation.

Type of medication: Tablet, taken by mouth.

Possible side effects: Headache, swelling of legs or feet, weight gain, low blood pressure, anemia, and fetal birth defects.

Drug names: Riociguat (Adempas).

Note: SGC medications have a dangerous interaction with other medications containing nitrates (such as nitroglycerin, isosorbide dinitrate). These medications should not be combined with PDE-5 Inhibitors (sildenafil or tadalafil). Women of childbearing age are required by the FDA to be on a reliable method of birth control while on this medication. Your provider will discuss these options before starting this medication.