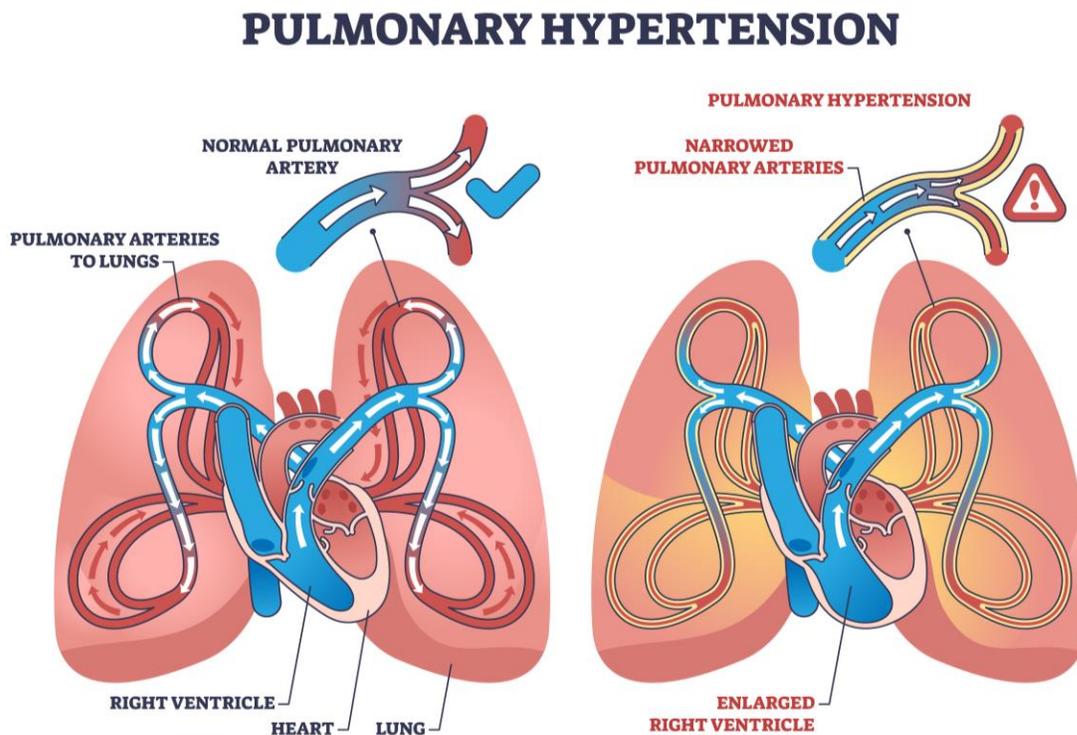


Pulmonary Arterial Hypertension

Pulmonary arterial hypertension (PAH) is when blood pressure in the pulmonary arteries is high. The pulmonary arteries carry blood from the right side of the heart to the lungs, so they have oxygen (Picture 1).

Pulmonary hypertension happens when the pulmonary arteries become stiff, tight, or narrowed. Over time, scar tissue forms. The narrowing makes it hard for blood to flow through the arteries, causing PAH. This puts strain on the heart, making it large and weak. Over time, the heart may not keep up with the body's needs. This is called **heart failure**.



Picture 1 Normal blood flow (left) and blood flow with pulmonary hypertension (right).

Signs and Symptoms

Older children, teens, and adults may have 1 or more of these symptoms:

- Tired
- Short of breath
- Trouble walking up stairs
- Chest pain
- Swollen ankles or legs
- Blue colored lips or nail beds
- Dizzy, faints
- Enlarged nail beds (clubbing)

Types of PAH

Type	Cause
Idiopathic PAH	No cause can be found.
Heritable PAH	Passed down from a parent (inherited).
PAH with types of heart disease that you are born with	Examples include: <ul style="list-style-type: none">• Repaired and unrepaired atrial septal, and ventricle septal defects (VSD)• Patent ductus arterious (PDA) that is still open
PAH connected with other conditions	Examples include: <ul style="list-style-type: none">• Connective tissue diseases• Portal hypertension
Drug- and toxin-induced PAH	Certain drugs and poisons (toxins) have been linked to PAH. Examples include: <ul style="list-style-type: none">• Some weight loss drugs• Toxic rapeseed oil

Tests

You may need to have the following tests done to diagnose PAH:

- **Chest X-ray** – used to see if the pulmonary arteries are large.
- **Blood work** – rules out other conditions.
- **Electrocardiogram (EKG)** – checks for heart rhythm changes and heart damage.
- **Pulmonary function test** – looks for other lung diseases.
- **Sleep study** – looks for oxygen levels and sleep problems (sleep apnea).

- **Echocardiogram (Echo)** – checks for increased pressure in the lungs and blood flow in the heart. It looks at the size and strength of the heart.
- **Lung scan (V/Q scan)** – looks for blood clots and how much blood and air is going to each lung.
- **Right heart catheterization** – procedure to measure pressures in heart. **This is the best way to diagnose PAH.**

Treatment

There is no cure for most forms of PAH. However, there are medicines that may help you:

- **Phosphodiesterase Inhibitors (PDE-5)** – these are medicines taken by mouth or through a feeding tube.
 - PDE-5 relaxes blood vessels. This reduces pressure in the lungs.
 - Examples of medicines that help the body to relax blood vessels in the lungs are: Sildenafil (Revatio[®]) and Tadalafil (Adcirca[®]).
- **Endothelin receptor antagonists (ERAs)** – these are medicines taken by mouth or through a feeding tube.
 - **Too much** endothelin will cause blood vessels to get narrow and tight. This can lead to high blood pressure in the lungs.
 - Examples of medicines that **lessen the endothelin** in the body: Tracleer[®], Opsumit[®], Letaris[®].
- **Prostacyclins** – these medicines are breathed in (inhaled), put into the body through a tube (catheter), taken by mouth or through a feeding tube.
 - These helps keep all blood vessels open so blood can flow easily. **Too little** of this in your body may increase blood pressure in the lungs.
 - Examples of medicines that help **make more prostacyclins** in your body:
 - Tyvaso[®] (inhaled)
 - Ventavis[®] (inhaled)
 - Uptravi[®] (by mouth)
 - Orenitram[®] (by mouth)
 - Remodulin[®] and Flolan[®] (through an IV [intravenous] tube or through skin)
- **Oxygen** – helps relax blood vessels in the lungs so you can breathe easier.
- **Blood thinners (anticoagulants)** – these may be used to lower the chance of blood clots in the lungs.
- **Water pills (diuretics)** – these are used to decrease swelling in the belly, legs, or feet.

Daily Living

With treatment, you can go to school or work and have an active life. You must:

- Go to your cardiologist at least 1 time each year.
- Know the signs and symptoms of PAH and when you feel different than usual.
- Tell your doctor or health care provider if your symptoms get worse.
- Take your medicines as prescribed each day.
- Check with your cardiologist before:
 - Taking part in sports or exercise.
 - Any type of surgery.
 - Taking over-the-counter (OTC) decongestant medicine.
 - Having dental work. You may need antibiotics before your dentist appointment.
 - Getting on an airplane.
- Get plenty of rest.
- Keep up on all vaccines and get your flu shot each year.
- Get a pneumonia vaccine every 5 years.
- Eat a heart-healthy diet that includes chicken, fish, fruit, whole grains, and vegetables.
- Limit the amount of salt (sodium) you eat. It can increase swelling. Buy items that say low sodium. Avoid fast food.

When to Call the Doctor

Call your cardiologist for any of the following:

- Irregular heart rate
- Fainting
- Chest pain
- Fast heart beat
- Poor appetite

When to Call 911

Call 911 for emergency help if you:

- Are coughing up blood.
- Have a seizure.
- Have trouble breathing, like can't catch your breath, gasping, wheezing, or grunting.
- Have changes to your skin color, like grayish blue or very pale.
- Are hard to wake up (lethargic), act confused, or don't know what you're doing.