



## Pulmonary Arterial Hypertension (PAH)

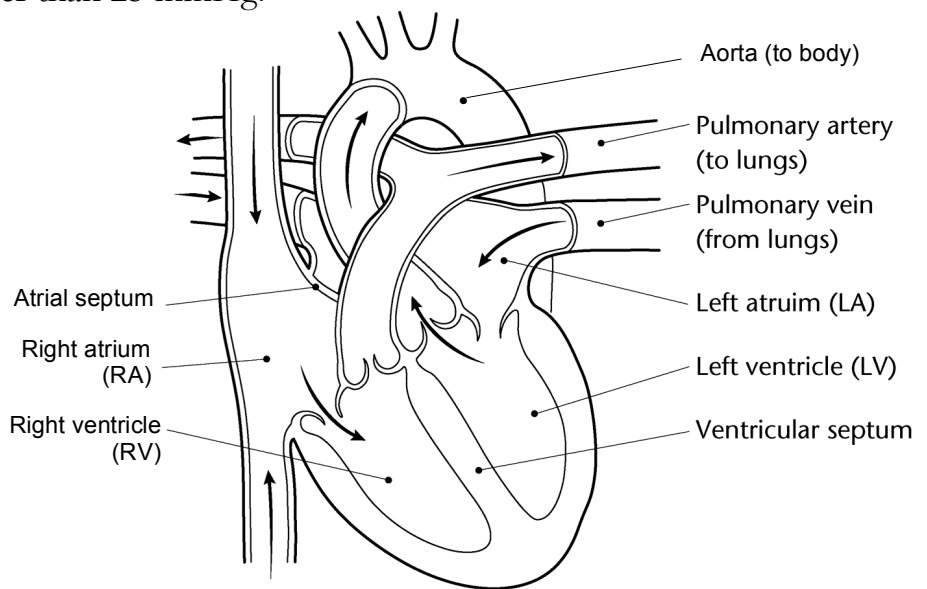
Pulmonary hypertension is a rare lung disease in which the blood pressure in the pulmonary arteries are high. The pulmonary arteries are blood vessels that carry blood from your right side of the heart to the lungs to receive oxygen (Picture 1).

Pulmonary hypertension develops when the pulmonary arteries become stiff and thick. This causes a tightening or narrowing of the pulmonary arteries. Over time scar tissue develops. These changes (narrowing) make blood flow difficult to pass through the pulmonary arteries resulting in PAH. Normal pressure in this artery is 15 mmHg. Patients with PAH have pulmonary artery pressures greater than 25 mmHg.

High pressure in the pulmonary artery makes it difficult for the right ventricle to pump blood into the lungs.

The strain on the heart causes the heart to become enlarged and weak.

Eventually the heart fails to keep up with the body's demands. This is known as **heart failure**.



**Picture 1** Inside the heart.

### Signs and Symptoms

#### Infants and young children

Poor growth	Rapid breathing	Lethargy (drowsiness)
Poor appetite	Respiratory Distress	Blue colored lips or skin

#### Older children, adolescents, and adults

Difficulty lying flat	Shortness of Breath	Dizziness, fainting spells
Swelling of ankles or legs	Chest Pain	Blue colored lips or nail beds
Difficulty walking up stairs	Tiredness	Enlargement of nail beds (clubbing)

## Types of Pulmonary Hypertension

Types of PAH	Cause
Idiopathic PAH	No underlying cause can be found.
Heritable PAH	It is inherited from a parent. The child is born with the tendency to have PAH.
PAH associated with certain types of heart disease which have been present since birth	Includes those born with a hole in the heart between the left and right sides Examples include: <ul style="list-style-type: none"> <li>▪ Unrepaired Atrial Septal Defect</li> <li>▪ Unrepaired Ventricle Septal Defect</li> </ul>
PAH associated with other conditions	Connective tissue diseases, HIV infection, and portal hypertension
Drug-and toxin-induced PAH	Certain drugs and toxins have been linked to the development of PAH. Examples include: <ul style="list-style-type: none"> <li>▪ Aminorex</li> <li>▪ Certain weight loss drugs</li> <li>▪ Toxic rapeseed oil</li> </ul>

## Testing

There are many tests the doctor may want to use. For an accurate diagnosis, certain tests may rule out other conditions or confirm PAH.

Diagnostic Test	Purpose of Test
Chest x-ray	May show enlarged pulmonary arteries
Blood work	Rules out other conditions
Electrocardiogram	May show changes in heart rhythm May indicate damage to the heart
Echocardiogram	May detect abnormal pressure and flow rates in the vessels of heart and lungs. Looks at the size and strength of the heart.
Lung scan (V/Q Scan)	Rules out blood clots in the lungs or lung disease
Pulmonary Function Test	Rules out other lung disease
Sleep study	Looks for sleeping disorder (sleep apnea)
Right Heart Catheterization	Best way to make the diagnosis of PAH

## Treatment Options

Although there is no known cure for most forms of PH, there are medicines available to help treat PAH. These medicines are based on natural chemicals in your body that may not be balanced.

Type of Treatment	Action
<p><b>Endothelin Receptor Antagonists (ERAs)</b></p> <p>These are oral (taken by mouth) medicines.</p>	<p><b>Too much</b> endothelin will cause blood vessels to narrow and tighten. This can lead to high blood pressure in the lungs.</p> <p>Examples of medications that <b>lessen the endothelin</b> in the body:</p> <ul style="list-style-type: none"> <li>▪ Tracleer<sup>®</sup></li> <li>▪ Opsumit<sup>®</sup></li> <li>▪ Letaris<sup>®</sup></li> </ul>
<p><b>Phosphodiesterase Inhibitors (PDE-5)</b></p> <p>These are oral (taken by mouth) medicines.</p> <p>PDE-5 works like nitric oxide.</p>	<p>Nitric oxide helps relax blood vessels. <b>Low levels</b> of this can lead to high blood pressure in the lungs.</p> <p>Examples of medications that <b>make more nitric oxide</b> in the body to help relax blood vessels:</p> <ul style="list-style-type: none"> <li>▪ Sildenafil (Revatio<sup>®</sup>)</li> <li>▪ Tadalafil (Adcirca<sup>®</sup>)</li> </ul>
<p><b>Prostacyclins</b></p> <p>These medicines are either:</p> <ul style="list-style-type: none"> <li>▪ inhaled (breathed in)</li> <li>▪ infused into the body through a catheter (tube)</li> <li>▪ taken orally (by mouth)</li> </ul> <p>Oral prostacyclins are newer drugs for PAH.</p>	<p>Helps to keep arteries open so blood can flow easily. <b>Too little</b> of this in your body may increase blood pressure in your lungs.</p> <p>Examples of medications that help <b>make more prostacyclins</b> in your body:</p> <ul style="list-style-type: none"> <li>▪ Tyvaso<sup>®</sup> (inhaled)</li> <li>▪ Ventavis<sup>®</sup> (inhaled)</li> <li>▪ Remodulin<sup>®</sup> and Flolan<sup>®</sup> (through an IV tube or through skin)</li> <li>▪ Uptravi<sup>®</sup> (oral) was FDA approved December 2015</li> <li>▪ Orenitram<sup>®</sup> (oral)</li> </ul>
<p><b>Oxygen</b></p>	<p>This can help relax blood vessels in your lungs.</p>
<p><b>Anticoagulants</b></p>	<p>Known as “blood thinners.” These are sometimes used to decrease blood clots in your lungs.</p>
<p><b>Diuretics</b></p>	<p>Known as “water pills.” These are used to decrease swelling in the abdomen, legs or feet.</p>

## Living with Pulmonary Hypertension

With the best possible therapy, going to school or work is possible and expected. Patients with PAH can have active lives. All PAH patients must:

- Be seen at least yearly by a cardiologist who specializes in pulmonary arterial hypertension.
- Know the signs and symptoms of PAH.
- Let the doctor know if symptoms get worse.
- If the doctor has prescribed medicines for PAH, make sure these are taken every day.
- Check with the cardiologist before participating in sports or exercise.
- Get plenty of rest.
- Check with the cardiologist before any type of surgery.
- Check with the cardiologist before taking over-the-counter decongestants.
- Ask the cardiologist if antibiotics are needed before dental work.
- Keep up all immunizations. All PAH patients should get a flu shot every year.
- A child should receive a pneumonia vaccine every five years.
- Always eat a “heart healthy diet.” Include chicken, fish, fresh fruit, whole grains and vegetables.
- Restrict salt in the diet. Salt may increase swelling. Buy items that say “low sodium” and avoid “fast food.”

## When to Call the Doctor

Call the cardiologist if any of the following things occur:

Difficulty breathing	Fainting
Chest pain	Coughing up blood
Rapid heart beat	Poor appetite
Irregular heart rate	Sweating during feedings (infants)