

## Pulmonary Arterial Hypertension



### What Is Pulmonary Hypertension?

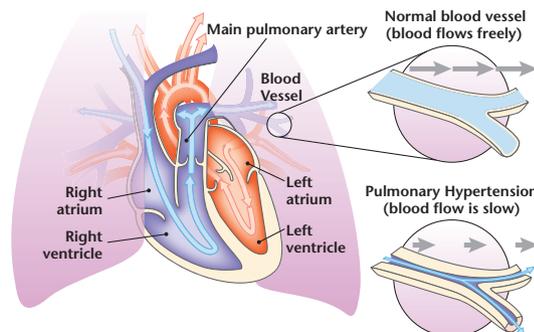
To understand pulmonary hypertension (PH) it helps to understand how blood flows throughout your body. While the heart is one organ, it works like two pumps that are connected to one another. There is a left side and a right side of the heart, each with two different jobs.

The left side of the heart (left atrium) takes oxygen-rich blood coming from your lungs and the left ventricle pumps this blood throughout your body. Since the left side of your heart has to pump blood such a great distance, the left side of your heart is designed to pump against a fairly high pressure. This pressure is easily measured with a blood pressure cuff and is called your blood pressure. When your **blood pressure** is too high, it is called **systemic hypertension** or simply, **hypertension**.

After your blood has delivered oxygen to the tissues of your body, the blood needs to come back to the lungs to get more oxygen. It does this by returning the blood to the right side of the heart (right atrium) and then the right ventricle pumps the blood into your lungs, so the process can start over again. The blood does not need to travel very far to get from the right side of your heart to your lungs. Therefore, the right side of your heart pumps against less pressure than the left side of your heart. The right side of your heart is therefore normally a *low-pressure* system. The pressure that the right side of your heart is pumping against is called your **pulmonary pressure**. When this pressure is too high, it is called **pulmonary hypertension** (PH). How the pressure in the right side of your heart is measured will be discussed in a later section.

### Is There More Than One Type of Pulmonary Hypertension?

Yes, you may hear the terms pulmonary hypertension (PH) or pulmonary arterial hypertension (PAH). Both terms mean that the pressure on the right side of your heart is higher than normal. Pulmonary hypertension (PH) is a general term that means that the blood pressure on the right side of your heart is too high, but does not explain why it is high. The high pressure could be caused by a medical condition such as; chronic lung disease, blood clots in the blood vessels in your lungs, or the left side of your heart is weakened (causing a back-up of pressure on the right side of the heart). This type of pulmonary hypertension was called “secondary pulmonary hypertension” but is now referred to as PH, because the cause is known to be from lung disease, heart disease, or



chronic thromboemboli (blood clots).

Pulmonary Arterial Hypertension (PAH) used to be called “primary pulmonary hypertension”. PAH occurs when the blood vessels in the lung are directly diseased (unlike the other forms of PH where the increased pressure is due to another reason like chronic lung or heart disease) and become thick and narrow. The pressure on the right side of your heart increases as it tries to pump blood through these narrow blood vessels. In PAH the pressure that the right side of your heart is pumping against is usually a much higher pressure than in patients who have PH from other causes.

### What Happens to the Body with Pulmonary Arterial Hypertension?

With PAH, the arteries become too narrow to handle the amount of blood that must be pumped through the lungs. This causes several things to happen: a backup of blood in the veins returning blood to the heart; an increase in the pressure that the right side of your heart has to pump against to push blood through your lungs; and a strain on the right side of your heart due to the increased work that it has to do. If this increased pressure is not treated, the right side of your heart can become overworked, become very weak and may possibly fail. Because the blood has difficulty getting through the lungs to pick up oxygen, your blood oxygen level may be lower than normal. This can put a strain not only on your heart, but also decrease the amount of oxygen getting to your brain.

### What are the Symptoms of Pulmonary Arterial Hypertension?

There may be no signs or symptoms of PAH in its early stages. You might first notice that you become short of breath more easily. You might also notice you are more tired (fatigued) than usual. Some patients also may feel “light headed” or even pass out. Swelling (edema) of your feet and ankles is common with PAH and may progress to swelling of your belly (ascites). Chest pain may also occur and can be mistaken for a heart attack. You may feel your heart racing or pounding (palpitations). The oxygen level in your blood may become very low.

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## How is Pulmonary Arterial Hypertension Diagnosed?

Because the different kinds of pulmonary hypertension are treated differently, it is important that your health care provider takes the time and orders the necessary tests to find out what kind of pulmonary hypertension you have. You can help your health care provider in diagnosing your condition by telling them what kind of symptoms you are having and if there is any worsening of these symptoms. For example, let your health care provider know if you notice any of the following:

- Increased shortness of breath
- Dizziness
- Feeling like you might faint
- Fainting
- Chest pain
- Heart palpitations (feeling like your heart is racing or pounding)
- Increased swelling of your feet, legs or belly
- Your lips and/or fingers turn blue

If your health care provider thinks you may have PAH, they will order tests to see if there is a strain on the right side of your heart. Usually the first test they order will be an ultrasound of your heart (echocardiogram). If the echocardiogram shows the pressure on the right side of your heart may be high, they may order a cardiac catheterization. During a cardiac catheterization, a rubber tube (catheter) is placed through a blood vessel into the chambers of your heart to measure the pressure in the right side of your heart (See ATS Patient Series at <http://patients.thoracic.org/information-series/en/resources/arterial-catheterization.pdf>). A cardiac catheterization is the best way to measure the blood pressure in the right side of your heart.

## What Causes Pulmonary Arterial Hypertension?

The cause of PAH is often difficult to determine. Your health care provider will likely order several tests to see if they can find a cause. PAH can be from some known causes, such as inherited (called *familial*) PAH, or be caused by reasons that are never known (called *idiopathic* PAH). Other known causes of PAH (called *associated* PAH) are:

- Connective tissue diseases such as scleroderma or lupus
- Use of prescription amphetamines or diet pills
- Use of illicit drugs such as cocaine and methamphetamines
- Congenital heart defects
- Liver disease/cirrhosis
- HIV

## Is There a Cure for Pulmonary Arterial Hypertension?

PAH is a serious disease that at this time has no cure but there are treatments available and new medications are in clinical trials. Every patient's disease is different and it is important that you discuss your care with your health care

provider. Early diagnosis and treatment are important to try to limit the progression of PAH.

## How is Pulmonary Arterial Hypertension Treated?

If you have PAH, there are several types of treatment available. These include medicines you swallow (oral medications), medicines that you breathe directly into your lungs (inhaled) and medicines that are given continuously, directly into a vein or under your skin with a pump (infused). These PAH medications are given to open the blood vessels in your lungs, improve the blood flow through your lungs and reduce the strain on your heart. Most patients with PAH will need to take medicines (diuretics, also called "water pills") that remove excess fluid that tends to buildup in the body. You may also need to use oxygen if your oxygen level is low. Additionally, patients with PAH may be referred to a pulmonary rehabilitation program to help them become more active and breathe better. There are many new medications being studied and your provider may suggest or recommend these for the treatment of your PAH as part of clinical research trials.

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### Resources:

**National Heart Lung and Blood Institute (NHLBI) at**  
<http://www.nhlbi.nih.gov/health/health-topics/topics/pah/>  
**Pulmonary Hypertension Association at**  
<http://www.phassociation.org>

## Rx Action Steps

If you have been diagnosed with PAH, it is important to find a health care provider who has experience in treating patients with PAH. Centers specializing in PAH ([www.phassociation.org/Patients/FindADoctor](http://www.phassociation.org/Patients/FindADoctor)) may be the best option as they have dedicated doctors, nurses, and other staff to assist in your care. You should see your health care provider regularly and contact them if you notice any change in your symptoms including the following:

- ✓ Increasing shortness of breath
- ✓ Swelling
- ✓ Feeling light headed
- ✓ If you have passed out.

**Doctor's Office Telephone:**

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