

### What is pulmonary hypertension?

If you have pulmonary hypertension (PH), it means that the blood pressure in your lungs is higher than normal. The heart has two sides that send blood to two places—the lungs and the body. When your caregiver uses a blood pressure cuff on your arm, s/he is measuring the pressure of arterial blood in your body. Blood pressure is always measured at two points: when the heart beats (systole) and when the heart relaxes (diastole). Normal blood pressure in the body should be less than 120/80mmHg. In the lungs, the pressure should be less than 40/18mmHg, or under 25mmHg on average. If the average (mean) pressure in the lungs is higher than 25mmHg, you have PH.

### How do you measure pressure in the lung (pulmonary) artery?

The most accurate way to measure lung pressure is by catheterization. A small tube is placed in a vein and directed into your heart and lung arteries. A direct pressure measurement is taken through the tube. A less invasive way to measure lung pressure is through an ultrasound of the heart (also called an echocardiogram or echo). Through echo, your healthcare providers can measure the speed of blood flow throughout the heart. They can then estimate the pressure in different parts of your heart. This allows them to determine if you have any narrowing of an artery, which would increase resistance to flow. This lets them estimate your lung pressure but is not as dependable as a catheterization.

### What causes pulmonary hypertension?

Science tells us that pressure equals flow times resistance. If you have ever played with a hose, you know that there are several ways to increase pressure. You can turn the spigot and increase the amount of water flowing through the hose. You can also bend or block the hose and make the water flow through a smaller opening, increasing resistance to flow. PH can be caused by problems with too much blood flow to the heart. It can also be caused by narrowing or stiffening in the heart and blood vessels, which increases resistance.

### Can congenital heart disease (CHD) cause PH?

There are many problems in the heart that can cause PH. If you have a connection (shunt) in your heart that creates extra blood flow to the lungs, this can cause PH. The most common defects that cause PH are holes in the heart such as atrial septal defects (ASDs), ventricular septal defects (VSDs), and patent ductus arteriosus (PDA). Other defects can also cause PH, including truncus arteriosus, double-outlet right ventricle (DORV) and any defect in which there is only a single working ventricle.

If the filling pressure on the left side of your heart gets too high, it can also cause PH. The pressure can rise due to a thickening of the heart's wall. The heart must allow blood in order to be able to pump it to your body. You might have noticed that a thin, stretched balloon is easier to blow up than a thick, stiff balloon. If your heart wall is thick or stiff, it

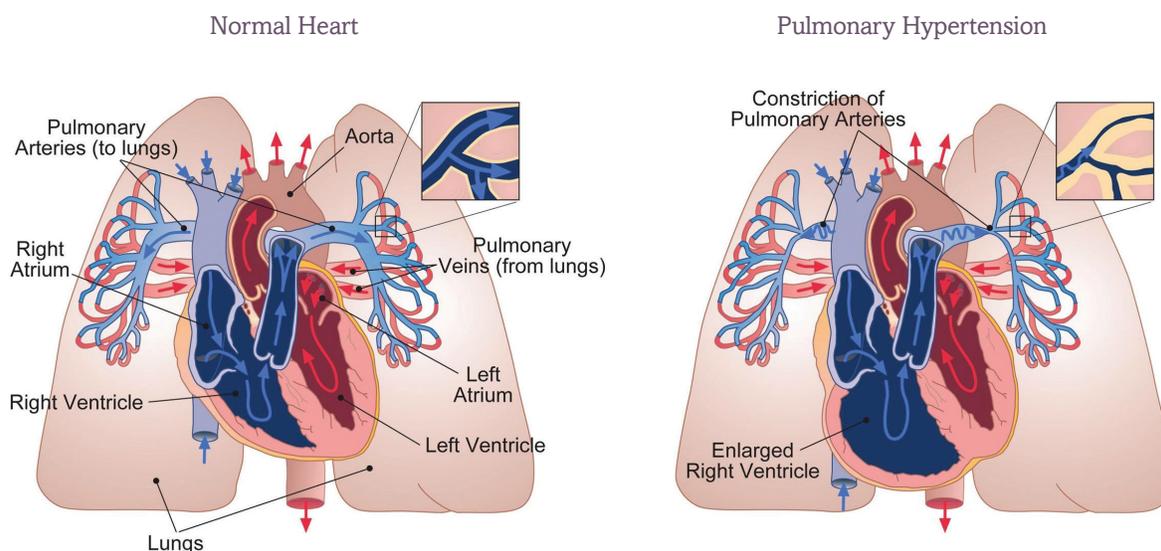


Image courtesy of the Columbus Ohio Adult Congenital Heart Disease Program at Nationwide Children's Hospital Heart Center, Columbus, Ohio

becomes harder to fill; you need higher filling pressure to drive the blood into the left side of the heart. This can cause your blood to get “backed up” in the lungs and cause PH.

### **What are the other causes of PH?**

Beyond the heart, there are many problems that can cause or worsen PH. Some other causes of PH are genetic mutations, rheumatologic disease such as scleroderma, interstitial lung disease, and liver cirrhosis.

**Problems in the lungs:** Heart and lung development are connected, so CHD patients can have inborn problems in their lung structures. Your lungs may not have developed enough oxygen-processing areas. Certain heart defects can cause the lung’s blood vessels to become blocked or compressed. Your lung tissue may also be damaged by scarring from heart surgeries. If your CHD caused spine or ribcage problems, your lungs may not be able to expand normally. If your heart is too big, this can also compress your lungs. CHD patients can sometimes develop blood clots in the lungs. All of these things can decrease your heart’s ability to process oxygen. When this happens, your lung pressure can increase as your heart tries to increase the body’s oxygen supply.

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**Problems outside the heart and lungs:** There are a number of general diseases that can cause PH similar to that seen with CHD. Examples are liver cirrhosis, HIV infection, or scleroderma. Obesity and obstructive sleep apnea may result in milder forms of elevated lung pressures. If you have PH, it is especially important that you maintain a heart-healthy weight and good eating habits.

Other possible causes of PH include thyroid problems, immune system problems, infections, anemia, parasites, and severe vitamin B deficiency. Certain drugs, such as methamphetamine or diet pills, can also make lung pressure rise.

If you have CHD and PH, it is important that your healthcare provider look beyond your heart and check for all possible causes. Many CHD patients find that there are several factors involved in their lung problems.

### **What are the treatments for PH?**

PH is like a fever; it is a sign that there is a problem somewhere that is causing lung pressure to rise. The first step in treatment is making sure that the cause of the rise is found. Different kinds of causes require different kinds of treatment.

**Heart structure problems:** If a heart structure problem is causing your PH, your caregivers may be able to correct it. For example, lung pressure might be lowered by closing a hole and/or opening blockages that increase pressure. These interventions might happen through surgery or through catheter-based interventions. The same procedures that are used to correct blood flow through the heart can often lower lung pressure. If you had previous heart repair, and you now have PH, it is very important that an ACHD specialty center assess your anatomy and determine if additional heart repair is needed.

**Lung damage problems:** When heart defects cause high pressure in the lungs over a long time period, it can result in permanent damage. The lungs can become scarred and inflamed, leaving the cells less able to transport oxygen efficiently. Once this happens, PH will continue even if the heart is repaired.

**Medication therapies:** There are many new medications now that can improve lung function in patients who have permanent PH caused by their CHD. There are five main types of medications that work to increase the lung cell’s ability to provide oxygen to your body:

- Calcium channel blockers (nifedipine, amlodipine)
- Endothelin receptor antagonists (bosentan, ambrisentan)
- Phosphodiesterase V inhibitors (sildenafil, tadalafil)
- Soluble guanylate cyclase stimulators (riociguat)
- Prostanoids (epoprostenol, treprostinil, iloprost)

Different medications affect different cell functions. All medications prescribed for PH require careful monitoring by experts in CHD and PH. While these medications do not cure PH, they can significantly improve health and well-being. Medication therapies are often combined for best results, and many new drugs are being developed to help patients with PH.

The good news about PH and CHD is that in the last 15 years there has been an enormous increase in both knowledge and effective treatments. If you are an ACHD patient and have been told that there are no treatments for your PH, we encourage you to seek care at an ACHD program offering PH expertise. This way you can ensure you learn about any new options available to help you feel better and live longer.

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