Pulmonary Arterial Hypertension (PAH)

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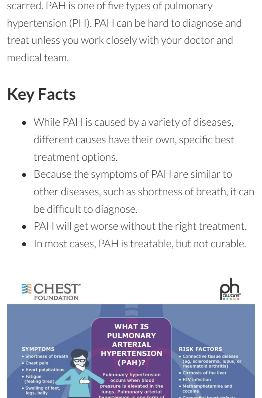
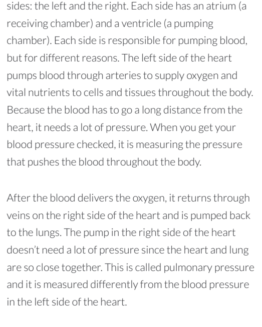
Learn About Pulmonary Arterial Hypertension



Pulmonary arterial hypertension (PAH) is a rare and serious disease that happens when there is high blood pressure in the lungs. PAH is caused when the small blood vessels in your lungs become narrowed and scarred. PAH is one of five types of pulmonary hypertension (PH). PAH can be hard to diagnose and treat unless you work closely with your doctor and medical team.

Key Facts

- While PAH is caused by a variety of diseases, different causes have their own, specific best treatment options.
- Because the symptoms of PAH are similar to other diseases, such as shortness of breath, it can be difficult to diagnose.
- PAH will get worse without the right treatment.
- In most cases, PAH is treatable, but not curable.



How Pulmonary Arterial Hypertension Affects Your Body

To understand how PAH affects the body, you first need to understand the way lungs contribute to the normal flow of blood. The heart is divided into two sides: the left and the right. Each side has an atrium (a receiving chamber) and a ventricle (a pumping chamber). Each side is responsible for pumping blood, but for different reasons. The left side of the heart pumps blood through arteries to supply oxygen and vital nutrients to cells and tissues throughout the body. Because the blood has to go a long distance from the heart, it needs a lot of pressure. When you get your blood pressure checked, it is measuring the pressure that pushes the blood throughout the body.

After the blood delivers the oxygen, it returns through veins on the right side of the heart and is pumped back to the lungs. The pump in the right side of the heart doesn't need a lot of pressure since the heart and lung are so close together. This is called pulmonary pressure and it is measured differently from the blood pressure in the left side of the heart.

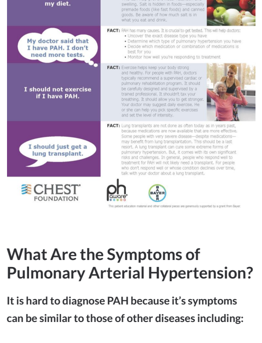
When the pumping pressure is too high in the left side of the heart, it is called hypertension or high blood pressure. Similarly, when the pumping pressure is too high in the right side of the heart, it is called pulmonary hypertension (PH).

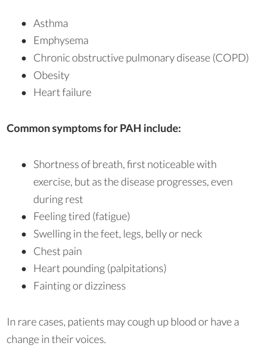
PAH (pulmonary arterial hypertension) is one form of PH. In this case, high blood pressure in the right side of the heart is caused from blood vessels in the lungs that have thickened and narrowed. Because of this narrowing of blood vessels, the right side of the heart has to pump harder – meaning use more pressure – to get blood to pass through. It also leads to a back-up of blood returning from the veins to the heart. As a result, the right side of the heart works too hard to keep up with the blood flow, which can weaken the heart and, over time, make it fail. Additionally, it takes longer for the blood to be cleansed through the lungs and pick up oxygen, which can lead to a lower-than-normal blood oxygen level. Oxygen is vital not only to cells and tissue, it is a critical component for healthy brain function. That's why diagnosing and treating PAH is so critical.

Measuring Blood Pressure

You're probably familiar with getting your blood pressure checked with a blood pressure cuff. This measures the pressure in the left side of the heart and is recorded in millimeters of mercury (mm Hg). There are two parts to this blood pressure measure. The first number reflects the systolic measure, which measures the pressure when your heart beats. The second number is the diastolic measure, which measures the pressure when your heart rests between beats. A normal blood pressure level is 120 (systolic) over 80 (diastolic).

Pulmonary Arterial Hypertension Symptoms, Causes and Risk Factors





What Are the Symptoms of Pulmonary Arterial Hypertension?

It is hard to diagnose PAH because it's symptoms can be similar to those of other diseases including:

- Asthma
- Emphysema
- Chronic obstructive pulmonary disease (COPD)
- Obesity
- Heart failure

Common symptoms for PAH include:

- Shortness of breath, first noticeable with exercise, but as the disease progresses, even during rest
- Feeling tired (fatigue)
- Swelling in the feet, legs, belly or neck
- Chest pain
- Heart pounding (palpitations)
- Fainting or dizziness

In rare cases, patients may cough up blood or have a change in their voices.

What Causes Pulmonary Arterial Hypertension?

Some of the most common diseases or conditions associated with PAH are:

- Connective tissue and/or autoimmune diseases, such as scleroderma, lupus, or rheumatoid arthritis
- Cirrhosis of the liver
- HIV infection
- Methamphetamine and cocaine use
- Congenital heart defects
- Rarely, an inherited disease

If no other disease are associated with PAH, it is referred to as idiopathic pulmonary arterial hypertension (IPAH).

What Are Risk Factors?

Risk factors for PAH include:

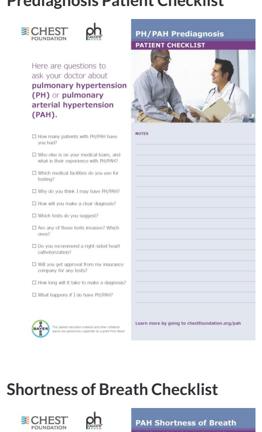
- Obesity
- Illegal drug use
- Family history of pulmonary hypertension
- Taking some appetite-suppressant medications
- Living in high altitudes
- Sleep apnea

Diagnosing and Treating Pulmonary Arterial Hypertension

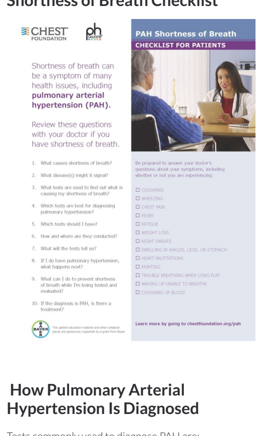
If your doctor suspects you have PAH, you will require several tests to prove first, that you have PH and specifically PAH and second, what type it is so that you can be treated appropriately. These tests help your doctor examine and directly measure the blood pressure in your lungs.

Print out and take these checklists to your appointment to facilitate better conversations with your health-care provider.

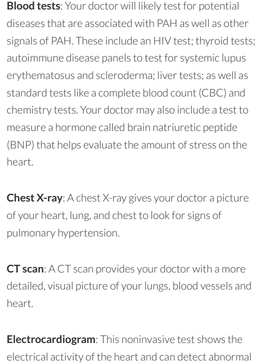
Questions About Your Diagnosis, Treatment, and Lifestyle



Prediagnosis Patient Checklist



Shortness of Breath Checklist



How Pulmonary Arterial Hypertension Is Diagnosed

Tests commonly used to diagnose PAH are:

Blood tests: Your doctor will likely test for potential diseases that are associated with PAH as well as other signals of PAH. These include an HIV test; thyroid tests; autoimmune disease panels to test for systemic lupus erythematosus and scleroderma; liver tests; as well as standard tests like a complete blood count (CBC) and chemistry tests. Your doctor may also include a test to measure a hormone called brain natriuretic peptide (BNP) that helps evaluate the amount of stress on the heart.

Chest X-ray: A chest X-ray gives your doctor a picture of your heart, lung, and chest to look for signs of pulmonary hypertension.

CT scan: A CT scan provides your doctor with a more detailed, visual picture of your lungs, blood vessels and heart.

Electrocardiogram: This noninvasive test shows the electrical activity of the heart and can detect abnormal heart rhythms. It is a useful test to identify different causes for your symptoms associated with PAH.

Pulmonary function tests: These simple breathing tests measure how much air you can hold in your lungs and how much air moves in and out of your lungs. Pulmonary function tests may signal other lung diseases, like asthma or pulmonary fibrosis, which often causes scarring that can narrow lung blood vessels.

Exercise tolerance test (also known as the 6-minute walk test): This test is used to compare your exercise capacity, oxygen levels and symptoms over time and to evaluate how these characteristics change over time and with therapy.

Cardiopulmonary exercise testing (CPET or CPX): This test measures how well your heart and lungs are performing both at rest and during exercise. CPET helps your doctor understand the amount of oxygen your body is using, the amount of carbon dioxide your body is producing, and your breathing pattern. Results can help determine whether or not you may have PAH.

Ventilation-perfusion scan (VQ scan): This test examines air and blood flow to the lungs and creates images doctors use to look for blood clots in the lungs.

Echocardiogram: This noninvasive test – a type of ultrasound of the heart – looks at the chambers and valves of the heart, determining their size and function. An echocardiogram will detect some signs of PH in most patients with the disease, making it very practical. While it does not directly measure pulmonary arterial pressure (PAP), it does give doctors an idea if PH is present and how severe it is. Unfortunately, it cannot guarantee the diagnosis of PH by itself, which means further testing is likely.

Cardiac MRI (Magnetic Resonance Imaging): This is an MRI test that evaluates the heart size and function more accurately than the echocardiogram and gives a better picture of the heart muscles, valves, and blood work. It may also show any congenital heart disease (an abnormality in the heart that developed before birth).

A cardiac MRI also helps doctors identify and quantify any shunting of blood that may be happening between the right and left sides of the heart. Normally, the right and left sides of the heart function completely separately and no blood is exchanged between them. Sometimes PH or PAH can cause the blood to flow in a different pattern, which can impact how well the heart and lungs work.

Right-sided heart catheterization: Your doctor will likely recommend a right-sided heart catheterization because it is the clearest test for showing which of the five forms of PH you have. This test involves placing a small tube, known as a catheter, into a large vein in the neck, arm, or groin. This catheter is then threaded through the different chambers of the heart and lung to measure the pressure in each. How much blood the heart pumps each minute, known as cardiac output, is also measured. The amount of resistance to blood flow in the lungs, known as the pulmonary vascular resistance, can be calculated from these measurements and are important indicators of how severe your PAH is.

During heart catheterization, it is likely that you will undergo a trial to assess if a medication is effective at dilating your blood vessels. There are two types of medications that can be tested:

- Medications delivered by breathing (Nitric oxide), or
- Medications given through an IV (prostacyclin or adenosine).

The results help guide your doctor's choice of medications that can be used to manage your PH. Mostly the trial tells your doctor whether or not regular high blood pressure medications, known as calcium channel blockers, will be helpful for you.

How Pulmonary Arterial Hypertension Is Treated

In most cases, PAH is treatable, but not curable. Treatment for PAH is very individualized and is designed to relieve symptoms and slow the progress of the disease. The exact course of your PAH treatment depends on you, the severity of your symptoms, the test findings, and your support system at home. Fortunately, the treatment of PAH has changed during the last 20 years. Several medications have been approved for PAH that don't work for other forms of PH. In all likelihood, you will need to take more than one medication to get the best results. In addition to medications, some patients also need oxygen to make breathing easier.

Medical treatment is different for every patient and is usually adjusted to enable the patient to meet goals that are important to the patient and the physician. These goals, both from the lab and clinical standpoints, are often adjusted to the individual and often require discussion with the physician.

Stick to Your Treatment Plan. It is important that you follow all the instructions your doctor gives you for taking medications and making lifestyle improvements to keep you in the best health possible. Finding the right medications can take some trial and error, but you need to stay the course. Stopping the medications can be very dangerous and should only be done under your doctor's supervision.

Using Oxygen

Some people with PAH may need to use extra oxygen to help them breathe better and improve their daily life. Oxygen is not addictive, so you don't have to worry about how much you use. Oxygen can help:

- Make breathing easier
- Improve sleep, mood, and mental ability
- Your body work better during the day

There are three types of oxygen:

- **Compressed gas oxygen**, which comes in steel or aluminum containers in different sizes for home use and traveling
- **Liquid oxygen**, which changes liquid into oxygen that you can breathe
- **Oxygen concentrators**, electrical devices that remove nitrogen from the air, which makes oxygen more concentrated

Questions to ask If your doctor prescribes oxygen include:

- How do I make sure I receive the right amount of oxygen?
- When and how often should I use my oxygen container?
- Which type of oxygen equipment will be best for my lifestyle?

Traveling with Oxygen

Airlines have rules about using oxygen. You cannot travel on an airplane with an oxygen canister. You must use either an airline-supplied oxygen canister or an FAA-certified portable oxygen container. There may be other rules, so check with your airline and/or oxygen supplier at least 2 weeks before you plan to travel.

Lung Transplantation and PAH

Lung transplantation is a final treatment option for some patients with PAH when other medical therapy is no longer effective.

Lung transplantation may prolong survival and improve

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infections, kidney failure, rejection. Although long-term outcomes with lung transplantation are improving, they are still far from ideal. Therefore, it is often a very complex decision and needs to be made along with the family, caregivers, your PAH medical team, as well as a transplant team.

Living with Pulmonary Arterial Hypertension

Living with PAH can be difficult to manage. You will need to be active in your care. It is important that you know as much about your disease and medications as possible and work with your doctor to find the best treatment plan for you.

Because PAH is a rare and complex disease, good care for PAH patients involves a close relationship with a PAH specialist as well as support from family and friends. Be sure you work with a clinical team with lots of experience treating patients with PH and PAH. They will understand the variety of diagnostic tests and the most current treatment advances that might improve your situation. They will also have access to medical