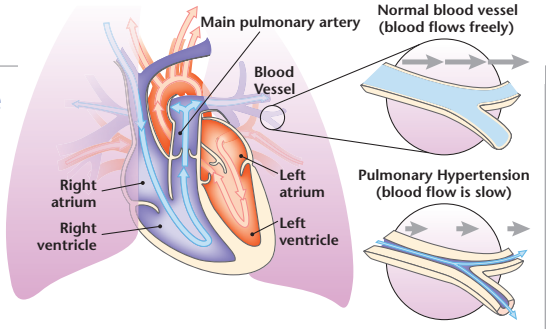
Diagnosis and Treatment of Pulmonary Hypertension



Pulmonary hypertension is a problem in which the blood pressure is high in the lungs from any cause. There are five different types of pulmonary hypertension that are grouped based on the causes. All forms of pulmonary hypertension are serious and can be life-threatening. Pulmonary hypertension can develop at any age. Different types of PH can require different treatment. There is usually no cure, but treatment can often help reduce symptoms and limit progression. When another chronic disease or cause is found, the treatment may need to be directed at the cause as well. This fact sheet will focus on adults and review the diagnosis and treatment of PH. For more information about pulmonary hypertension in children and the diagnosis and the different types of pulmonary hypertension see the Patient Information Series fact sheets at www.thoracic.org/patients.

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How is pulmonary hypertension diagnosed?

Pulmonary Hypertension can be difficult to diagnose and therefore it is important to be evaluated by a team that has expertise in PH. Your healthcare provider will likely order several other tests to see if they can find a cause if it is not clear that it is due to an underlying chronic disease.

Many tests will be ordered to determine the severity of disease and the type of pulmonary hypertension.

- One of the first tests performed is often an ultrasound of the heart called an echocardiogram (echo). This painless procedure is often used to help screen for and make a diagnosis of PH by estimating the pressures in the right heart and assessing how well the heart is functioning. The echo will show if there is strain on the right side of the heart. Echocardiograms are used to follow disease progression. Other heart conditions that produce symptoms similar to PH may be diagnosed with an echocardiogram.
- A chest x-ray may show enlargement of the size of the heart and/or pulmonary arteries and evaluate for lung disease. A CT (CAT Scan) of the chest can show abnormal blood vessels or early lung disease which may not be noticed on x-ray.
- An electrocardiogram (EKG) may be performed to evaluate for enlargement of the right side of the heart or changes in cardiac muscle function. The EKG checks the electrical impulses of the heart. Electrodes are attached to the person's skin, and a recording of these impulses is made. An EKG alone is not enough to make a PH diagnosis.
- A six-minute walk test is used to assess exercise capacity, or endurance. During the test, a person walks a as far as they can while wearing a pulse oximeter to measure heart rate and oxygen levels. The distance walked is measured and compared to previous six-minute walk tests. This test is also used to follow response to therapy and to help determine disease severity.
- Pulmonary function tests (PFTs) are typically done to evaluate air flows, lung volumes and oxygen uptake. The tests can show if you have lung disease. For more information on pulmonary function testing, go to www.thoracic.org/patients.
- A sleep study or polysomnography may be performed if there is concern for sleep disordered breathing as a contributing factor or cause of PH.
- Blood tests may be done to look for the cause of PH such as to see if a person has a collagen vascular disease, signs of infection, or HIV antibodies. Blood tests may be used to check the oxygen level. One test, the brain natriuretic peptide (BNP) can help assess the strain on the heart and may also be used to monitor response to treatment. Blood tests may be done to check liver and kidney function with use of medications.
- Nuclear scan (Ventilation/Perfusion Scan or V/Q Scan)
  A nuclear scan tests for blood clots in the lungs by making a picture of air and blood flow to the lungs. A small dose of radioactive material is breathed in, and another small dose is injected via a blood vessel into the lungs. Multiple images are taken as these materials flow through the airways and blood vessels of the lungs.
- Right heart (Cardiac) catheterization. This is the gold standard in diagnosis of PH. If the echocardiogram shows the pressure on the right side of your heart may be high, your healthcare provide may advise that you get a right heart (cardiac) catheterization. During a catheterization, a special 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 and blood vessels in the lungs.
  Through this procedure, the degree of PH can be measured, response to therapy (oxygen/medications) can be tested and anatomy of the heart and blood vessels can be studied. Cardiac catheterization can help guide therapy as well as help monitor disease progression.

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A cardiac catheterization is the best way to measure the blood pressure in the right side of your heart. If your mean pulmonary artery pressures are more than 25 mm Hg on right heart catheterization you have PH.

How is pulmonary hypertension treated?

Treatment depends in part on how symptomatic you are. The World Health Organization (WHO) has a severity of symptoms scoring system that is used to help make decisions about treatment. You will work with your PH specialist to consider that score and other clinical information about you to decide what treatment you may need and how long you need treatment.

There are many treatments are available which may relieve some symptoms and may slow the progression of the disease. Early treatment is important because advanced disease may be less responsive to therapy.

Treatment usually includes use of specific PH disease modifying medications. Other medications include diuretics (water pills) and possibly blood thinners (anticoagulants). These PH medications are given to relax the blood vessels in the lungs, improve blood flow through the lungs, and reduce strain on the right heart.

There are three different categories of pulmonary hypertension modifying drugs. They can be given in different forms such as pills to swallow, in a vein or injected under the skin by a pump or breathed in through an inhaler. Your PH specialist will decide if you need one or a combination of drugs from three categories based on how sick or severe your symptoms are. The WHO functional class is a tool that helps determine how much PH affects you in daily life. In general, the higher the WHO functional class (more symptomatic) you will get combination therapies. Your treating physician should be specialized in the field of pulmonary hypertension as these drugs could have very dangerous side effects. 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.

Most people with PAH will need to take medicines (diuretics, also called "water pills") that remove excess fluid that tends to buildup in the body. You also may need to be on blood thinners.

It is also recommended that you follow a low salt diet, and weigh yourself daily, especially for those with right heart failure, and ensure your immunizations are up to date.

Your healthcare provider may suggest that you wear oxygen with sleep or all the time if your oxygen level is low. For more information about oxygen therapy, see ATS Patient Information Series fact sheet at www.thoracic.org/patients.

People being treated for PH need to have regular appointments with their specialist. They often need regular testing including blood tests and echocardiograms to determine how well their medications are working. You need to contact your PH specialist or healthcare provider right away with any changes in symptoms. While the current coronavirus pandemic may pose unique challenges, it is important and safe for you to receive ongoing and uninterrupted care for your PH.

People with PH should avoid contact sports and only take part in activities that allow them to rest when tired. You may be referred to a pulmonary rehabilitation or exercise program to help you be more active and breathe better. For more information about Pulmonary rehab see ATS Patient Information Series fact sheet at www.thoracic.org/patients

If medications fail, for some people with PH, treatment options may include surgical measures to decompress the heart or lung transplantation. These two advanced procedures are only done in highly specialized centers after careful consideration.

What is the prognosis for pulmonary hypertension?

The prognosis depends on the type and severity of PH (such as whether it is the only problem or is associated with another condition). For PH that occurs with another condition, the prognosis is often tied to the prognosis of the underlying disease or condition. In contrast, PH that occurs without an obvious cause (i.e. PAH) is a progressive disease, which is life-shortening. At this time, for almost all types of PH, there is no cure. However, with early and aggressive treatment, improved survival has been reported. Advances in research continue to provide hope for the future treatment of PH.

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Rx *Action Steps*

If you have trouble breathing with exercise or fainting spells, that are not improving with treatment or getting worse, ask your healthcare provider about checking for pulmonary hypertension.

- ✔ If you have been diagnosed with pulmonary hypertension, seek care from a PH specialist.
- ✔ Take all medications as prescribed and call right away with any change in symptoms.
- ✔ Talk with your PH specialist about what activities you can safely do.

Healthcare Provider's Contact Number:

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For More Information

American Thoracic Society
- www.thoracic.org/patients/
  - CTEPH
  - Oxygen Therapy
  - Pulmonary Function Testing
  - Pulmonary Rehabilitation
  - Sleep Studies
  - What is PH

PHAware Global
- www.phaware.global

Pulmonary Hypertension Association
- www.phassociation.org

PHA Europe–European Pulmonary Hypertension Association
- http://www.phaeurope.org/

US National Library of Medicine
- https://medlineplus.gov/pulmonaryhypertension.html

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