

**Pulmonary
Hypertension and
Interstitial
Lung Disease**

PH&I

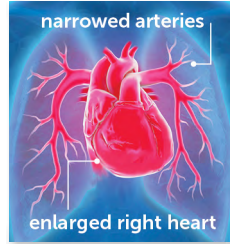


Pulmonary Hypertension Association
Empowered by hope

ABOUT

PULMONARY HYPERTENSION

Pulmonary hypertension is a complex and often misunderstood disease. PH sometimes is confused with systemic hypertension or “high blood pressure” that affects arteries throughout the body.



PH refers to pressure within the blood vessels of the lungs. The blood vessels can become stiff and narrow, which makes it more difficult for the right side of the heart to pump blood through them.

There are five types of PH based on different causes. Each form of PH is different, so it is important for newly diagnosed patients to find PH specialists who can pinpoint the cause of their PH. The specialist then develops a treatment plan specifically for the patient’s type of PH.

Every person with PH is different, and scientists continually conduct new research to improve the outlook for people living with PH. With proper care and treatment, people with PH can live many years.



NORMAL mean pulmonary artery pressure is between 8-20 mmHg at rest.

PULMONARY HYPERTENSION is defined as a resting mean pulmonary artery pressure at or above 20 mmHg.



PH Related to Interstitial Lung Disease

There are many types of interstitial lung disease, which are related to the network of lung tissue that supports air sacs in the lungs. Interstitial lung diseases are characterized by changes in the air sacs, such as scarring, inflammation or both.

Interstitial lung diseases can be caused by autoimmune conditions or exposure to certain medicines, dust, chemicals, mold, smoke or other inhaled substances.

Idiopathic pulmonary fibrosis is the most common type of interstitial lung disease. Idiopathic means without a known cause and fibrosis means scarring. Pulmonary fibrosis is characterized by scarring in the lungs that continues to get worse.

Patients often complain of breathlessness (especially with activity), dry cough and fatigue.

Pulmonary fibrosis is characterized by scarring in the lungs that continues to get worse.

CT scan showing healthy lungs.



CT scans showing lung fibrosis.



When that happens, doctors usually refer patients to a pulmonologist. The pulmonologist might order a breathing (pulmonary function) test and a high-resolution CAT scan of the chest. HRCT is a type of high-resolution CAT scan that shows a detailed view of the lung.

Diagnosis and treatment usually take place at a center specializing in interstitial lung disease. The results of the HRCT, along with blood work findings, determine whether a lung biopsy is necessary.

Some patients with pulmonary fibrosis develop PH, which can worsen shortness of breath and cause dizziness.

Diagnosing PH

Patients with interstitial lung disease can develop PH for many reasons. It's important to determine the root cause of PH for each patient.

Echocardiogram: An ultrasound of the heart that provides a rough estimate of the pressure in the lungs. It assesses the function of how blood flows through the heart by examining the right and left ventricles, valves, etc.

Pulmonary function tests: Non-invasive tests that measure how much air the lungs can hold, how much and how quickly someone can blow air, and how oxygen and carbon dioxide are exchanged between the air entering the lungs and the blood circulating through the lungs (diffusion capacity of carbon monoxide).

Ventilation-perfusion (V/Q) scan: Determines whether there are undissolved clots in the blood vessels of the lungs.

Natriuretic peptide tests: Measure specific proteins in a blood sample and help screen for heart failure. Tests of the proteins NT-proBNP (N-terminal pro b-type natriuretic peptide) and BNP (brain natriuretic peptide) are two examples. Elevated levels of these proteins can signify heart failure.

Right heart catheterization: The most accurate diagnostic test for PH. It measures the pressures and flow directly in the pulmonary artery. The invasive procedure usually takes place on an outpatient basis in the hospital. Doctors guide a small tube (catheter) through the right side of the heart and into the pulmonary artery to directly measure the pulmonary artery pressure.

In addition to directly measuring pressures in the heart and lungs, a right heart catheterization can determine whether patients have problems with a stiff left side of the heart. Left heart disease also can cause PH. In those cases, doctors refer patients to a cardiologist to treat the heart disease.

For people with interstitial lung disease, it's important to rule out obstructive sleep apnea through a sleep study, as sleep apnea can cause PH.

If doctors can't identify other causes of PH, they can diagnose PH due to the patient's interstitial lung disease.

Treatment Options

Anyone with ILD can benefit from simple measures, including attention to fluid balance and vaccinations for influenza and pneumonia. People with PH-related to idiopathic pulmonary fibrosis often have low oxygen levels with exercise and at night, so those people need to use oxygen during sleep and with activity.

In addition, it is important to get treatment for interstitial lung disease. The Food and Drug Administration has approved several medications to treat pulmonary fibrosis and other fibrotic interstitial lung diseases.

Inhaled treprostinil is an FDA-approved drug to improve exercise capacity and treat ILD-associated PH. When PH is severe, doctors at specialized PH centers might try other medications.

PH often, but not always, develops among people with advanced interstitial lung diseases. PH can indicate that their lung disease is getting worse.

Lung transplantation is an option for some people with advanced interstitial lung disease. Patients need to be evaluated at a transplant center to

A monitored exercise program called pulmonary rehabilitation can decrease symptoms and improve endurance for people with PH and interstitial lung diseases.

determine whether they are candidates for lung transplant. For people deemed appropriate for lung transplant, PH can trigger placement on the transplant waiting list.

With these conditions, it is important to stay as active as possible. A monitored exercise program called pulmonary rehabilitation can decrease

symptoms and improve endurance for people with PH and interstitial lung diseases. Pulmonary rehabilitation takes place on an outpatient basis at a hospital and requires referral by a treating pulmonologist.



ADDITIONAL RESOURCES

Pulmonary Fibrosis Foundation | [PulmonaryFibrosis.org](https://www.pulmonaryfibrosis.org)

REFERENCES

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About the Pulmonary Hypertension Association

Headquartered in Washington, D.C., the Pulmonary Hypertension Association is the oldest and largest nonprofit patient association dedicated to the pulmonary hypertension community. PHA's mission is to extend and improve the lives of those affected by PH.

PHA engages people with PH and their families, caregivers, health care providers and researchers, who work together to advocate for the PH community, provide support to patients, caregivers and families, offer up-to-date education and information on PH, improve quality patient care, and fund and promote research. For information, visit PHAssociation.org.

PHA's mission is to extend and improve the lives of those affected by pulmonary hypertension. PHA's vision is a world without PH, empowered by hope.



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