Pulmonary Hypertension & IDIOPATHIC PULMONARY FIBROSIS
Pulmonary hypertension (PH) is a complex and often misunderstood disease. The term PH simply means high blood pressure in the arteries of the lungs. In regular hypertension — also known as high blood pressure or systemic hypertension, which you can check with a blood pressure cuff — the pressure in the arteries of the entire body is higher than it should be. In PH, the blood vessels specifically in the lungs are affected. They can become stiff and narrow, and the right side of the heart must work harder to pump blood through them. There are five groups of PH based on different causes.

Each form of PH is different, so it is important for newly diagnosed patients to find a PH specialist who can accurately pinpoint what is causing their PH. They then can develop a treatment plan for that specific type as soon as possible after a confirmed diagnosis. Every individual with PH is different, and new research with the potential to improve the outlook for people living with this disease is conducted on a continual basis. Once in the care of a PH-treating health care team and on appropriate therapy, individuals with PH can live for 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 **25 mmHg**.

*PH, or high blood pressure in the lungs, frequently results from a narrowing of the small blood vessels in the lungs, which leads to a larger right side of the heart.*
Interstitial lung disease (ILD) is a diagnosis that includes many different conditions. These each involve the support tissue in the lungs that sits between the blood vessels and the air sacs, called the interstitium. ILDs are characterized by different specific types of changes in the interstitium, sometimes by scarring, in other instances by inflammation and often by both. There are many reasons why someone may develop an ILD, such as an underlying autoimmune disease or exposure to various chemicals or other irritants like mold.

Idiopathic pulmonary fibrosis (IPF) is the most common type of interstitial lung disease. Idiopathic means without a known cause and fibrosis means scarring. IPF is characterized by scarring in the lungs that continues to get worse. Patients tend to be older and often complain of breathlessness (especially with exertion), dry cough and fatigue. When that happens, many patients are referred to a pulmonologist, who may order a breathing test (pulmonary function test) and a high-resolution CAT scan (HRCT) of the chest. HRCT is a type of CAT scan that provides the necessary resolution to show a detailed view of the lung. This diagnosis and treatment of ILD is often completed at a center specializing in these diseases. How the HRCT looks, along with blood work findings, determines if a lung biopsy is necessary.

CT scan showing lung fibrosis.¹
CT scan showing healthy lungs.

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Some patients with IPF also develop PH, which may explain why some patients with IPF develop worse breathing problems, lower extremity swelling or dizziness with activity.

**DIAGNOSING PH IN PEOPLE WITH IPF**

It is very important to determine what the root cause of PH is for each patient. The screening test most frequently used in PH is an echocardiogram. An echocardiogram is an ultrasound of the heart that provides a rough estimate of the pressure in the lungs and assesses the function of the right ventricle. A definitive PH diagnosis is made by right heart catheterization, a procedure during which a small tube (catheter) is guided through the right side of the heart and into the pulmonary artery to directly measure the pulmonary artery pressure. If no other cause of PH has been discovered, the diagnosis of PH due to IPF is made.

Patients with IPF can develop PH for many reasons. In addition to directly measuring the pressures in the heart and lungs, a right heart catheterization is useful to determine if patients have problems with a stiff left side of the heart. This also may cause PH, called PH due to heart failure with preserved ejection fraction (HFP EF-PH). In this case, a patient is referred to a cardiologist to help treat that condition. It also is important to rule out obstructive sleep apnea by performing a sleep study, as sleep apnea also can cause PH. Finally, a lung ventilation-perfusion scan (V/Q scan) is important to exclude blood clots in the lung that cause World Health Organization Group 4 PH (chronic thromboembolic pulmonary hypertension, or CTEPH).
TREATMENT OPTIONS FOR PH IN IPF

All patients benefit from simple measures, including attention to fluid balance and vaccinations for influenza and pneumonia. Individuals with PH-IPF often have low oxygen levels with exercise and at night, so it is very important that they use oxygen during sleep and with activity.

Although several medications have been approved for treatment of IPF, there are currently no medications that have been shown to treat the PH component of IPF. Several small studies indicated that medications used to treat other types of PH may be useful in PH-IPF. Sometimes, when a doctor feels that the PH is very severe and not entirely explained by the IPF, they may prescribe a targeted PH therapy. The decision to use PH medications in PH-IPF can be complicated and requires careful follow-up at a center experienced in treating individuals with PH. Treatment options for IPF-related scarring should be reviewed and discussed with a specialist.
Lung transplantation is an option for some patients with advanced IPF. A patient needs to be evaluated at a transplant center to determine if they are a candidate for lung transplantation. PH often develops in patients with advanced IPF, usually indicating that the IPF is getting worse. For patients who have been deemed appropriate candidates for lung transplantation, development of PH is one of the triggers to be placed on the transplant waiting list.

It is very important to stay as active as possible. A monitored exercise program called pulmonary rehabilitation has been shown to be useful to decrease symptoms and improve endurance for patients with PH, IPF and PH-IPF. Pulmonary rehabilitation is administered on an outpatient basis at a hospital and requires a referral by a treating pulmonologist.

*For patients who have been deemed appropriate candidates for lung transplantation, development of PH is one of the triggers to be placed on the transplant waiting list.*
ABOUT THE PULMONARY HYPERTENSION ASSOCIATION

The Pulmonary Hypertension Association (PHA) is a community of individuals with PH, caregivers, families, medical professionals and researchers who work together to empower people battling the disease, while supporting research toward improved treatments. PHA was the first organization in the world dedicated to providing comprehensive PH patient and caregiver support, medical education, research and services that improve patients’ quality of life.

PHA facilitates more than 200 active support groups around the country and serves as a model for more than 85 international PH organizations. In addition to in-person support, PHA offers a telephone support line, an email mentorship program, educational patient programs and a variety of print and online publications. Every two years, PHA hosts its International PH Conference and Scientific Sessions, which brings together nearly 1,500 individuals with PH, caregivers and medical professionals to share information on the latest research, treatments and lifestyle coping tips.

PHA also has provided more than $8 million for PH research and offers continuing education programs for medical professionals through the PHA Online University and an in-person CME program.

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