Pulmonary Fibrosis Foundation™

# Pulmonary Hypertension Related to ILD

## RECOGNIZING SYMPTOMS AND TAKING ACTION

Pulmonary hypertension (PH) is a condition in which high blood pressure develops in the pulmonary arteries (blood vessels in the lungs). PH has many causes, including genetic mutations; illicit drug use; and associated conditions including connective tissue disease, congenital heart disease, and chronic lung conditions such as interstitial lung disease (ILD).

### What happens in the heart and lungs with PH?

To understand PH, it is important to understand the way blood travels through the body. There are two sides to circulation and two sides to the heart. In left-sided, or "systemic," circulation, the left side of the heart pumps oxygen-rich blood through the arteries to supply oxygen to the body, including the brain, muscles, and major organs. When oxygen-depleted blood comes back to the heart from the body, it travels through veins to the right side of the heart, which then pumps blood to the lungs so more oxygen can be absorbed and carbon dioxide eliminated. (See diagram on next page.) PH is a condition in which there is increased pressure and resistance in right-sided circulation, also called pulmonary circulation.

### How can pulmonary hypertension be related to ILD?

The many causes of ILD can be associated with PH. When pulmonary fibrosis (PF) is present, there is a greater chance of developing PH. The ways in which ILD can lead to PH are not completely understood. It is thought that ILD-caused damage and changes to the tissue and structure of the lungs can create some increase in pressure in the pulmonary vessels.

### What symptoms are caused by PH?

In PH related to ILD (PH-ILD), some symptoms are very similar to the most common symptoms of ILD without PH, including shortness of breath and fatigue. PH-ILD may also cause dizziness or chest discomfort. Strain on the heart caused by PH-ILD can lead to a backup of fluid that can cause leg or abdominal swelling. PH-ILD can also cause hypoxemia (low oxygen levels), sometimes lower than seen with ILD alone.

### How common is PH-ILD?

About one in 20 patients, or 5%, seeing a doctor for the first time for PF may also have PH. Some studies have shown that 15-50% of patients with idiopathic pulmonary fibrosis (IPF) may have PH. These numbers are similar for patients with autoimmune disease, hypersensitivity pneumonitis, and some other types of ILD. According to some studies, PH affects up to 65% of patients with both emphysema and PF.

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## How is PH-ILD diagnosed?

Screening tests for PH-ILD may include pulmonary function tests (PFTs), six-minute walk testing, computed tomography (CT) scans, and monitoring of certain blood tests that may indicate high pressure or heart failure possibly caused by PH. If your care team suspects PH after the screening tests, they may order an echocardiogram, or ultrasound test, of your heart. A right heart catheterization is necessary to confirm the diagnosis of PH-ILD.

## How is PH-ILD treated?

While there is no cure for PH, treatments are available to help manage the condition. The treatment plan for PH-ILD may include a type of medication called a pulmonary vasodilator that lowers blood pressure by relaxing the pulmonary blood vessels so the heart can beat more efficiently and pump more blood. Often, treatment also includes supportive care such as supplemental oxygen and pulmonary rehabilitation. For patients with symptoms of fluid retention, dietary changes and diuretics are often prescribed. Appropriate candidates may also be evaluated for lung transplantation. Palliative care, which focuses on symptom relief and quality of life, may be especially helpful for patients with more severe symptoms. Treatment of PH-ILD is best undertaken by an experienced medical center that can closely monitor a patient's response to therapy.

## What medications are FDA-approved to treat PH-ILD?

The inhaled form of the vasodilator trepostinil (brand names Tyvaso or Yutrepia) was approved by the FDA in 2022 for PH-ILD. It is inhaled directly into the lungs, where it helps blood vessels relax and open so the heart can pump blood more easily. There are two ways to take inhaled treprostinil: a nebulizer inhalation system and a dry powder inhaler (DPI), both of which are taken four times a day. Other measures and medications may be recommended to manage side effects, which may include low systemic blood pressure (as measured by a blood pressure cuff), coughing, headaches, shortness of breath, dizziness, nausea, fatigue, diarrhea, and throat irritation.

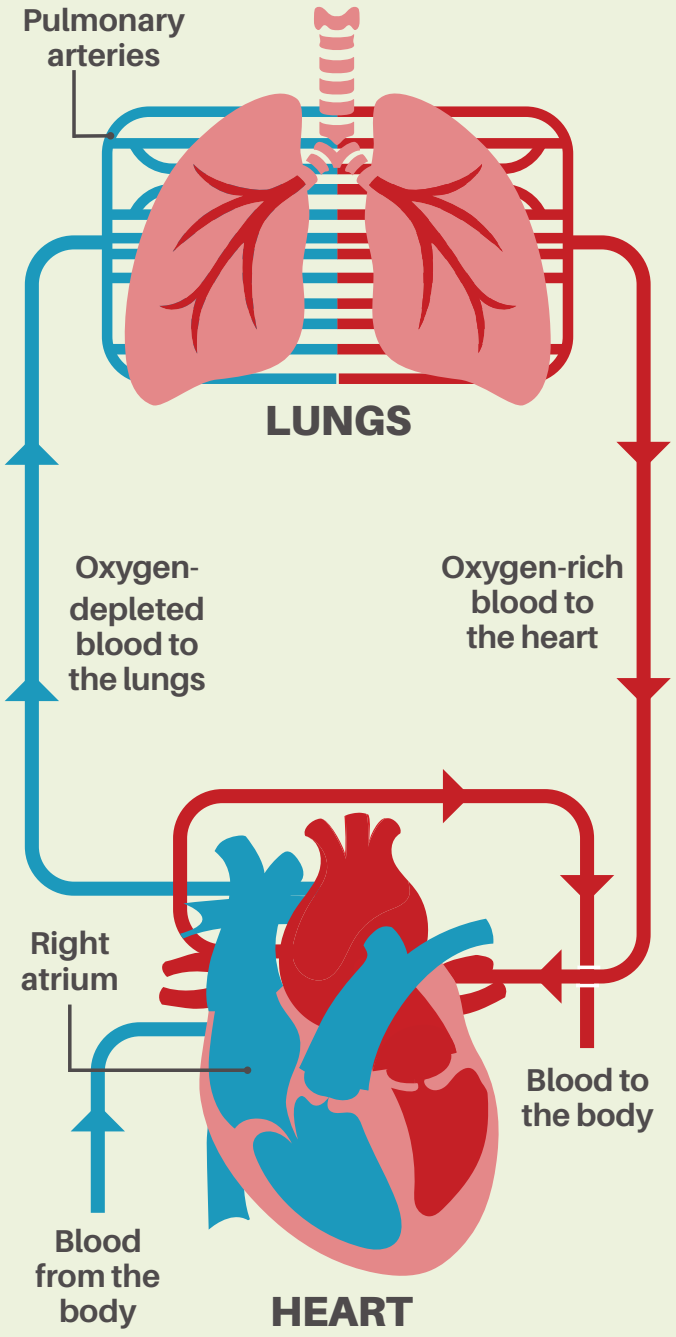
## Are there other medications for PH-ILD?

Study results have been mixed on a class of medications called phosphodiesterase type 5 (PDE5) inhibitors. Future studies should further assess the effectiveness of PDE5 inhibitors for treatment of PH-ILD.

## Where can I learn more?

Pulmonary Fibrosis Foundation educational materials
pulmonaryfibrosis.org/patients-caregivers/education-resources/other-educational-resources

Pulmonary Hypertension Association
phassociation.org

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