



PH-ILD

# GET TO KNOW PULMONARY HYPERTENSION ASSOCIATED WITH INTERSTITIAL LUNG DISEASE (PH-ILD)

Your guide to understanding what PH-ILD is, how it is diagnosed, and how an approved class of treatment may help.

# UNDERSTANDING PH-ILD

Learning about your diagnosis and symptoms is important so you can receive effective treatment that may help manage your PH-ILD.



## What is interstitial lung disease (ILD)?

ILD is the name for a group of serious, progressive conditions, most of which cause scarring of lung tissue, making it harder to breathe and get enough oxygen into your bloodstream.

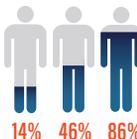
Many different conditions are forms of ILD, including but not limited to:

- Idiopathic pulmonary fibrosis (IPF) or pulmonary fibrosis (PF)
- Combined pulmonary fibrosis and emphysema (CPFE)
- Connective tissue disease (CTD)



## What is pulmonary hypertension (PH)?

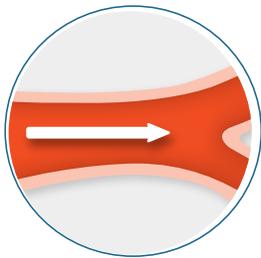
PH is a serious and progressive condition in which there is high blood pressure in the arteries of the lungs and the right side of the heart. People with ILD are often at high risk of developing PH, known as pulmonary hypertension associated with interstitial lung disease (PH-ILD).



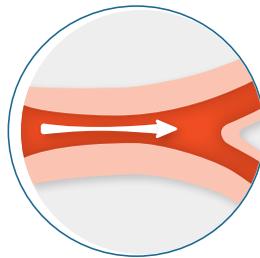
## How common is PH if you have ILD?

The proportion of patients with ILD who develop PH varies depending on the form and severity of ILD. For example, studies have shown that 14% of patients with early-stage IPF have PH, a number that increases to 46% as the disease advances and further rises to 86% with end-stage IPF.

## How does PH-ILD affect my lungs and heart?



**Normal** pulmonary artery

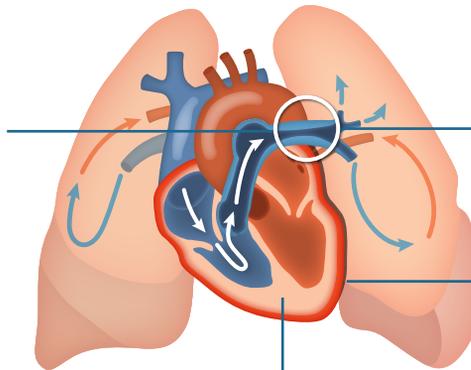


**Narrow** pulmonary artery

1

Damage to the lung due to ILD causes pulmonary arteries to stiffen and thicken, narrowing the arteries so there is less space for blood to flow.

**Pulmonary arteries**  
(blood vessels that carry blood from the right side of your heart to your lungs)



2

As a result, blood pressure in your pulmonary arteries goes up, causing PH.

3

This causes your heart to work harder to pump blood through the lungs.

4

High blood pressure causes the right side of the heart to become enlarged and weakened.

# DIAGNOSING PH-ILD

## How do I know if I have developed PH?

Many symptoms of PH are very similar to that of ILD, including shortness of breath, dizziness, fatigue, and chest discomfort.

- Symptoms of PH may develop slowly over time and can add to the overall burden of the symptoms of ILD
- After you have been diagnosed with ILD, it is important to be screened regularly for PH so you can receive appropriate treatment

### Symptoms of PH include:



**SHORTNESS  
OF BREATH**



**TIREDFNESS**



**CHEST PAIN  
OR PRESSURE**



**FAINING**



**DIZZINESS**



**RAPID  
HEARTBEAT**



**SWOLLEN  
ABDOMEN**



**SWOLLEN LEGS  
AND ANKLES**

**TALK TO YOUR DOCTOR ABOUT THE SYMPTOMS OF PH  
AND HOW THEY MAY DIFFER FROM THOSE OF ILD.**



## How is PH-ILD diagnosed?

Your doctor may perform several tests to determine if you have developed PH. Some of these tests may already be done regularly as a part of the routine monitoring of your ILD.



**Electrocardiogram (EKG or ECG)** to measure the electrical activity of your heart



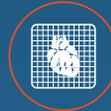
**Pulmonary function test (PFT)** to see how well your lungs are working



**6-minute walk test (6MWT)** to see how far you can walk in 6 minutes



**Echocardiogram (echo)** to check the size and function of your heart



**Chest x-ray and ventilation perfusion scan (VQ scan)** to get images of your heart, lungs, and blood vessels



**Blood tests** to check your oxygen levels and for signs of liver, kidney, or heart problems



**Right heart catheterization (RHC)** to measure the pressure inside your heart and the blood vessels of your lungs

**RHC IS THE MOST CONCLUSIVE WAY TO DIAGNOSE PH.**

# IMPACT OF PH-ILD

## How are the effects of PH-ILD on everyday activities measured?

A functional class assessment can help your doctor understand how much PH affects your everyday activities.

### The functional classes are defined as follows:

NYHA Functional Class I	No limitations on physical activity. Ordinary physical activity doesn't cause shortness of breath, fatigue, chest pain, or near fainting.
NYHA Functional Class II	Some limitations on physical activity. Although comfortable at rest, ordinary activity causes shortness of breath, fatigue, chest pain, or near fainting.
NYHA Functional Class III	Clear limitations on physical activity. Although comfortable at rest, even less than ordinary activity causes shortness of breath, fatigue, chest pain, or near fainting.
NYHA Functional Class IV	Any level of physical activity is uncomfortable. There may be signs of heart failure, and shortness of breath and/or fatigue may be present even when resting.

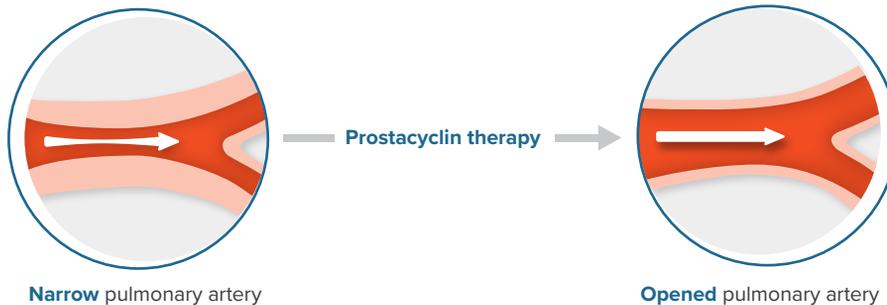
NYHA = New York Heart Association.

# TREATING PH-ILD

## What treatment is available for PH-ILD and how does it work?

Inhaled prostacyclin therapy is the only FDA-approved type of treatment for PH-ILD.

- Prostacyclin is a natural substance that helps keep your pulmonary arteries open
- Currently, the only approved prostacyclin therapy for PH-ILD is called treprostinil, which works by acting like the prostacyclin produced in your body



In a clinical study, patients with PH-ILD who took an inhaled treprostinil solution walked further on average and lowered the chance of their PH-ILD worsening than those who took a placebo solution.

- Prostacyclin therapy has also been proven safe and effective in patients with another type of PH called pulmonary arterial hypertension (PAH)

# GET TO KNOW PH-ILD



## KNOW THE RISKS

If you have ILD, you are at high risk of developing PH, a serious and progressive condition.



## KNOW WHAT TO LOOK FOR

Symptoms of PH are similar to those of ILD, so it is crucial to start regular screening early.



## KNOW WHAT CAN BE DONE

If you develop PH-ILD, there are treatments that may help.

**LEARN MORE ABOUT A  
TREATMENT THAT COULD BE  
RIGHT FOR YOU.**



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