



# Breathing

LIFE

#BreathingLife

#PFMonth

PF Awareness Month  
September 2021

## FACT SHEET: PULMONARY FIBROSIS (PF)

PF is a progressive, irreversible, chronic lung disease causing scarring of the lung tissue with no known cure. In Europe it currently affects more than 300,000 people with over 50,000 losing their lives to PF each year. The most common type of PF is Idiopathic Pulmonary Fibrosis (IPF), accounting for 200,000 patients in Europe. The average life expectancy following diagnosis of PF or IPF is about three to five years, but early diagnosis and early treatment could improve this.

**Disease mechanism:** PF means scarring in the lungs, (pulmonary = lung, fibrosis = scar tissue). The scar tissue blocks the movement of oxygen from inside the tiny air sacs in the lungs into the bloodstream. Low oxygen levels (and the scar tissue) can cause a feeling of shortness of breath, particularly when walking and exercising. PF gets worse over time as the amount of scarring in the lungs build up. As this occurs, a person's breathing becomes more difficult, eventually resulting in shortness of breath, even at rest.

**A family of diseases:** PF is a family of more than 200 different lung disease, which is part of a larger group called interstitial lung diseases (ILD). ILD includes all diseases that have inflammation and/or scarring in the lung. In ILDs the injury and damage occur in the walls of the air sacs of the lung, and the tissue and space around these air sacs. Some ILDs don't include scar tissue. PF is an interstitial lung disease that includes scar tissue in the lung. The most common type of PF is IPF, which has no known cause. IPF is more common in men, but the number of cases of IPF in women is increasing.

**Causes of PF:** Some cases of PF are caused by autoimmune diseases like rheumatoid arthritis, scleroderma or Sjogren's syndrome, certain viral infections, or gastroesophageal reflux disease.

Familial PF is rare, but genes have been linked to PF.

PF can be caused by exposure to hazardous materials as for instance asbestos, breathing in bird or animal droppings and cigarette smoking also increases the risk of developing PF. Radiation treatments and certain types of medications can cause PF.

**PF diagnosis:** Most people with PF develop symptoms between the ages of 50 and 70 years. Because the symptoms of PF may be similar to symptoms of other lung diseases PF is often misdiagnosed and many people wait long to get their diagnosis.

### Symptoms of PF

- Shortness of breath, particularly during exercise
- Dry, hacking cough
- Fast, shallow breathing
- Gradual unintended weight loss
- Tiredness
- Aching joints and muscles
- Clubbing (widening and rounding) of the tips of the fingers or toes

**Treatment of PF:** There is no cure for PF. Current treatments are aimed at slowing the course of the disease, relieving symptoms, and helping to stay active and healthy. Research advancements allow people to live longer and have a better quality of life.