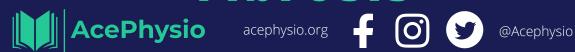
# Idiopathic Pulmonary Fibrosis







Idiopathic Pulmonary Fibrosis (IPF): fibrotic lung disease with unknow aetiology that progresses over the course of several years. It is characterised by scar tissue formation within the lungs, dyspnoea, and a significantly shortened lifespan after diagnosis.

#### **Causes:**

IPF is associated with several factors such as cigarette smoking, exposure to occupational dusts, gastro-oesophageal reflux disease (GORD), diabetes and several viral infections.

## Pathophysiology:

- Healing of primary pulmonary damage  $\rightarrow$  excess deposition of collagen + fibrous tissue  $\rightarrow$  progressive scarring of lung tissue + reduced lung compliance  $\rightarrow$  progressive dyspnoea → hypoxemia
  - ullet Hypertrophy of type II pneumocytes ullet excess collagen production ullet thickening of interstitial layer between alveoli + capillaries  $\rightarrow$  impaired gaseous exchange + sclerosis of lung parenchyma  $\rightarrow$  restricted lung expansion
  - Chronic, irreversible and eventual fatal disease

#### **Presentation**

- Dry, non-productive cough exacerbated by exertion
- Dyspnoea
- Cyanosis
- Finger clubbing
- Dry basal crackles on auscultation
- Significant respiratory failure with increasing pulmonary tissue damage



# **Diagnosis**

- Exclude known causes of interstitial lung disease
- High-resolution chest CT scan honeycombing, thickening of interstitial walls at the lung bases and periphery +/- traction bronchiectasis
- Chest x-ray basilar + peripheral reticular opacities (fine network of lines)
- Biopsy of lung tissue interstitial scarring, honeycombing, scar tissue
- Spirometry FEV1 and FVC are reduced - FEV1/FVC ratio is normal or increased

# Management

## Rehabilitation:

Respiratory muscle training in patients with impaired respiratory

function - induces diaphragm hypertrophy + reduction in perceived breathlessness and exertion

- Education on smoking cessation prevents further oxidative damage and disease progression
- Pulmonary rehabilitation involves aerobic, strength and mobility training, psychosocial support & education on energy conservation techniques - leads to improvement in walking capacity and quality of life
- Supplemental oxygen therapy for patients with severe hypoxaemia (PaO<sub>2</sub> ≤55 mmHg or oxygen saturation ≤89%) at rest or upon exertion - Oxygen therapy may improve exercise tolerance and reduce the risk of developing pulmonary hypertension and cor pulmonale
- Ergonomic assessment of home environment to optimise function
- Teach Active Cycle of Breathing Technique- breathing control may alleviate episodes of dyspnoea - thoracic expansion exercises enhances the recruitment of alveoli

Medical: Antifibrotic medication - slows disease progression; Seasonal influenza vaccine, Proton-pump inhibitor; Lung transplant - definitive treatment in severe cases

### Want to learn more?

With AcePhysio the learning journey doesn't stop here! Take a look at our further reading

recommendations below to become an expert in Idiopathic Pulmonary Fibrosis:

- 1. Vainshelboim B. Exercise training in idiopathic pulmonary fibrosis: is it of benefit? Breathe (Sheff). 2016 Jun;12(2):130-8 2. Raghu G, Rochwerg B, Zhang Y, et al. An official ATS/ERS/JRS/ALAT clinical practice guideline: treatment of idiopathic pulmonary fibrosis - an update of the 2011 clinical practice guideline. Am J Respir Crit Care Med. 2015 Jul 15;192(2):e3-19
- 3. Kozu R, Jenkins S, Senjyu H. Effect of disability level on response to pulmonary rehabilitation in patients with idiopathic pulmonary fibrosis. Respirology. 2011 Nov;16(8):1196-202