Idiopathic Pulmonary Fibrosis

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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 → excess deposition of collagen + fibrous tissue → progressive scarring of lung tissue + reduced lung compliance → progressive dyspnoea → hypoxemia
- Hypertrophy of type II pneumocytes → excess collagen production → thickening of interstitial layer between alveoli + capillaries → impaired gaseous exchange + sclerosis of lung parenchyma → 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_2 ≤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