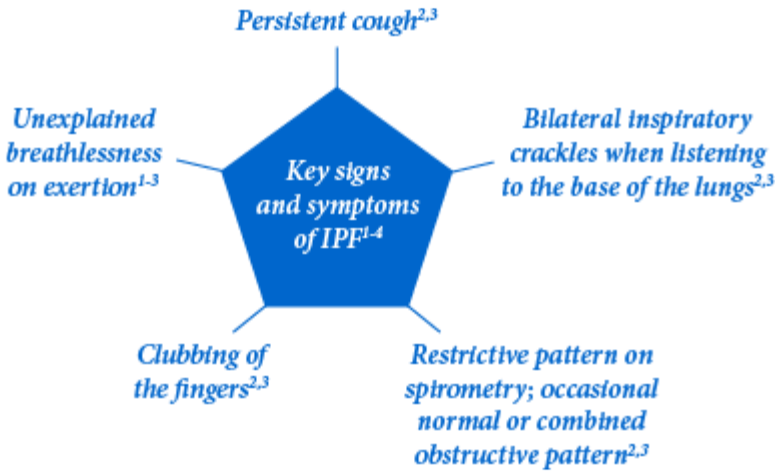
Presentation of IPF

Key clinical features of IPF

Symptoms at presentation

Patients with idiopathic pulmonary fibrosis (IPF) may present with a number of different symptoms.[1-3]



General risk factors of IPF[1]

- Patients who are older than 50 years of age
  - Incidence increases with older age, typically presenting in the 60s and 70s
- Men are at greater risk than women
- The majority of patients have a history of cigarette smoking

Listening to the lungs

Lung auscultation is currently the best way to recognise IPF early.[4] A key clinical feature for recognising IPF is the presence of bilateral Velcro®-like crackles that can be heard when listening to the basal section of the patient's lungs. Crackles are discontinuous, short, explosive sounds that, in IPF, resemble the sound of a strip of Velcro® slowly separating.[4] Identification of crackles should prompt a pulmonary function test.

For further details on what to listen for, including audio examples, see *Listening to IPF*

Pulmonary function test

IPF is a restrictive lung disease limiting lung capacity and impairing lung expansion.[5] All patients with suspected interstitial lung disease (ILD) should undergo spirometry and diffusing capacity pulmonary function tests to provide an assessment of the restrictive impairment.[5] The observed restrictive impairment typically manifests as reduced measures of lung capacity and volume, such as forced vital capacity, total lung capacity and residual volume.[5]

These measures may appear normal in patients with IPF and a concomitant obstructive disease, such as chronic bronchitis or emphysema.[5] It is also possible for a patient with IPF to have normal pulmonary function at rest; however, the diffusing capacity of the lungs for carbon monoxide (DLco) at presentation is a more reliable guide of disease status than other resting lung function variables; research has shown that a DLco <40% is suggestive of advanced fibrotic disease.[5]

References

1. Raghu G, et al. *Am J Respir Crit Care Med*. 2011;183:788–824. **2.** NICE Clinical Guidelines 163: Idiopathic Pulmonary Fibrosis. June 2013. **3.** NICE Clinical Pathway: Idiopathic Pulmonary Fibrosis overview. April 2015. **4.** Cottin V, et al. *Eur Respir J*. 2012;40:519–521. **5.** Wells AU, et al. *Thorax*. 2008;63:v1–v58.