

# HOSPITAL PHYSICIAN®

## PULMONARY DISEASE BOARD REVIEW MANUAL

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## Idiopathic Pulmonary Fibrosis

### Series Editor:

**Robert A. Balk, MD, FACP, FCCP, FCCM**

*Professor of Internal Medicine, Rush Medical College*

*Director of Pulmonary and Critical Care Medicine*

*Rush-Presbyterian-St. Luke's Medical Center*

*Chicago, IL*

### Contributing Author:

**Larry C. Casey, MD, PhD, FACP**

*Associate Professor of Medicine, Rush Medical College*

*Associate Director of Pulmonary and Critical Care Medicine*

*Rush-Presbyterian-St. Luke's Medical Center*

*Chicago, IL*

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#### I. INTRODUCTION

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Idiopathic pulmonary fibrosis (IPF) is an interstitial lung disease of unknown etiology. In the past, the medical literature has been confusing because of the various names that have been used to describe IPF. These include cryptogenic fibrosing alveolitis, Hamman-Rich syndrome, diffuse interstitial fibrosis, idiopathic interstitial pneumonia, honeycomb lung, fibrosing alveolitis, and usual interstitial pneumonia (UIP). The 2 most commonly used terms are IPF and cryptogenic fibrosing alveolitis. The primary distinction between IPF and cryptogenic fibrosing alveolitis is that cryptogenic fibrosing alveolitis includes patients with well-defined connective tissue diseases. By definition, IPF is idiopathic and thus has no clear association with any under-

lying disease, drug, or occupational or environmental factor known to cause pulmonary fibrosis.

The prevalence of IPF in the general population is difficult to determine. Open lung biopsy has been considered the gold standard for the diagnosis of IPF but has not been examined in large population-based studies. Tertiary care centers have reported prevalence rates of 3 to 6 cases per 100,000. An epidemiologic study from a county in New Mexico found a prevalence of approximately 30 cases per 100,000.<sup>1</sup> Most IPF patients present after age 50 years, with a peak incidence of presentation after age 70 years. Up to 70% of patients with IPF are current or former smokers.

There are clear cases of familial IPF, which tends to manifest itself at an earlier age (20–40 years) and to have a more rapidly progressive course. The genetics of familial IPF have yet to be confirmed, but it is thought