

Clinical Cases in Interstitial Lung Disease

<https://annenber.net/ILD-CME>

Key Findings in IPF

Interstitial pulmonary fibrosis is more common in males and with advancing age. Although IPF is a **diagnosis of exclusion**, signs and symptoms such as shortness of breath, nonproductive cough, Velcro crackles, digital clubbing, fatigue, **honeycomb pattern** (particularly on high-resolution computed tomography), and fibroblastic foci are strongly suggestive of IPF. **Changes over time** of the forced vital capacity, diffusing capacity of the lungs for carbon monoxide, and pulmonary artery pressure are useful prognostic indicators.



Personalized Treatment

An understanding of the role of **genetics** in IPF has evolved recently and is beginning to inform clinical trials and the development of **targeted therapies**. As yet, however, the impact on clinical practice has been minimal. **Personalized treatment** of IPF also includes avoidance of inhalational exposures and adoption of a healthy lifestyle.

Acute Exacerbation

Identifying and, if found, treating **causes of acute deterioration** in lung function **other than IPF is critical**. **Supportive care** often consists of high-flow nasal cannula oxygen, antibiotics, and corticosteroids. Other treatments are often used, but are not supported by good evidence. Preliminary evidence suggests that antifibrotic therapy, ie, nintedanib and pirfenidone, may prevent or reduce the risk of an acute exacerbation.

Holistic Care

Pulmonary rehabilitation is an essential component of holistic care for all patients with ILD. Additional care **should be individualized** to manage the many facets of ILD. The presence of comorbidities and disease complications should be assessed at diagnosis and periodically thereafter and managed in **collaboration with other providers**. Oxygen should be initiated as needed, and patients should be referred for evaluation for future lung transplantation.

Treatment for IPF

Current guidelines for IPF strongly recommend the use of **oxygen** and, in select patients, **lung transplantation**. Conditional recommendations for IPF include the **antifibrotic medications** nintedanib and pirfenidone, as well as **antacids** and **pulmonary rehabilitation**. The choice of antifibrotic therapy primarily depends on the side effect profile and other considerations such as drug interactions.

Diagnostic Evaluation

The diagnosis of an ILD is best made through a multidisciplinary discussion in which the clinical, radiologic, and histopathologic evidence is considered.

Palliative Care

Palliative care should be implemented at the time of diagnosis of ILD with the goal of **optimizing daily functioning and quality of life**. Holistic care is an essential component of palliative care and should be provided by a multidisciplinary team to include treatment beyond focusing on slowing disease progression.

