INTERSTITIAL LUNG DISEASE:
ALTERING THE DISEASE COURSE THROUGH EFFECTIVE AND INDIVIDUALIZED MANAGEMENT

D Differential Diagnosis

Interstitial lung diseases (ILDs) represent a broad category of heterogeneous diseases characterized by variable amounts of inflammation and/or **fibrosis of the lung**, primarily of the interstitium. Because the prognosis and treatment vary widely among these diseases, especially for interstitial pulmonary fibrosis (IPF), an accurate differential diagnosis is essential. Critical components of the **diagnostic evaluation** include a thorough history and physical examination, selected laboratory parameters, high-resolution computed tomography, and lung function testing.

T Treatment Principles

Early diagnosis and **early initiation of individualized therapy** are important. Removing the cause of the ILDs, if known, is essential. Hypoxemia should always be treated to reduce complications. A holistic approach to treatment that involves the primary care provider is essential, since comorbidities (eg, chronic obstructive pulmonary disease, gastroesophageal reflux disease, coronary artery disease) are common. **Palliative care** should be initiated at the time of diagnosis with the goal of optimizing quality of life throughout the disease course.

P Pharmacologic Treatment

For many ILDs — other than IPF — reducing inflammation is the goal; this generally involves a corticosteroid, often with immunosuppressive therapy (eg, mycophenolate). However, for IPF, the goal is to reduce fibrosis.

Nintedanib and pirfenidone are **antifibrotic therapy**, indicated for IPF treatment. The choice of antifibrotic therapy is based on patient factors, side effects, and tolerability. Targeted therapies based on increased understanding of the role of genetics in the pathogenesis are being investigated.

N Nonpharmacologic Treatment

Pulmonary rehabilitation is essential for all patients with IPF since functional capacity and quality of life are improved.

Lung transplantation is an option for select patients with advanced IPF, who have progressive respiratory failure, but carries substantial risks, including death. Evaluation should begin before significant disease progression, ideally shortly after diagnosis.

A Acute Exacerbations

Acute exacerbations are a **complication of IPF** and may contribute to early death. Excluding other causes of pulmonary decompensation, such as heart failure or pulmonary embolism, is imperative.

Available treatments for an acute exacerbation, eg, corticosteroids, cyclophosphamide, tacrolimus, or hemofiltration, have limited effectiveness. Consequently, **preventative strategies** (eg, antifibrotic therapy) are important.

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