Interstitial lung disease (ILD) is the presence of fibrosis and/or inflammation of the lung parenchyma not due to infection or malignancy. 1

**STEP 1: RECOGNIZE ILD**

**TYPICAL PRESENTATION**

- Subacute or chronic cough
- Exertional dyspnea (breathlessness)

**While performing an H&P:**

- Listen carefully for crackles
- Look for evidence of autoimmune disease on the skin and joints
- Walk your patient informally in the hallway to assess for desaturation (>3% drop)

Order initial testing based on your H&P. Consider:

- Spirometry (normal or low FVC often with a normal FEV1/FVC ratio in ILD)
- Lung Volumes (normal or low TLC)
- DLCO (frequently reduced in ILD)
- Chest x-ray (unexplained, persistent markings)

If you suspect ILD, perform a high resolution CT scan of the chest. HRCT includes the following:

- Inspiratory and expiratory supine imaging
- High-spatial frequency algorithm reconstruction at 0.625 to 2.5mm thickness
- No IV contrast required

**MANAGEMENT CHECKLIST**

- Recommend smoking cessation
- Advise patients to purchase a pulse oximeter to monitor oxygen levels immediately after activity: stairs, hills, level ground, exercise, showering
- Prescribe different oxygen prescriptions at rest, with routine activities, with exercise, and with sleep to maintain oxygen levels over 90% 24 hours per day
- Consider nocturnal oximetry or polysomnography
- Prescribe pulmonary rehabilitation
- Advise patients to achieve a healthy weight
- Administer influenza and pneumococcal vaccination
- Tell your patients about reliable resources like the Pulmonary Fibrosis Foundation (PFF). They can visit pulmonaryfibrosis.org or call 844.TalkPFF (844.825.5733) for information
- Provide information about your local PFF support group
- Consider lung transplant evaluation
- Consider clinical trial enrollment
- Consider pharmacological treatment after a diagnosis is confirmed

**SUPPORT FOR YOU FROM THE PULMONARY FIBROSIS FOUNDATION**

The Pulmonary Fibrosis Foundation mobilizes people and resources to provide access to high quality care and leads research for a cure so people with pulmonary fibrosis will live longer, healthier lives.

To learn more about how the PFF can help support you, contact the PFF Patient Communication Center at 844. TalkPFF (844.825.5733) or pcc@pulmonaryfibrosis.org, or visit the PFF online at pulmonaryfibrosis.org.

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**Step 2A: Perform a detailed history and physical**

**Disease**

- Drug/Radiation-induced ILD
  - Chemotherapy, amiodarone, nitrofurantoin, other drugs
  - Check PneumoTox.com, Radiation therapy to the chest

- Connective Tissue Disease
  - Joints, skin, Raynaud's, effusions, dry eyes/mouth, muscle weakness, or pain
  - Occupational history
  - Familial history of ILD, sarcoidosis, home oxygen use, autoimmune disease

- Vasculitis
  - Exposure to mold sources (forced air heating, hot tubs, humidifiers, water damage or visible mold)
  - Exposure to birds, down bedding, farming, or agriculture

- Chronic Hypersensitivity Pneumonitis
  - Exposure to mycoplasma, fungal/fungal (forced air heating, hot tubs, humidifiers, water damage or visible mold)
  - Exposure to birds, down bedding, farming, or agriculture

- Familial ILD
  - Family history of ILD, sarcoidosis, home oxygen use, autoimmune disease

**Questions/Findings**

- Disease-related ILD or pain
- Vasculitis
- Drug/Radiation-induced ILD
- Connective Tissue Disease
- Chronic Hypersensitivity Pneumonitis
- Familial ILD

**Step 2B: Order relevant blood tests**

**Disease**

- Eosinophilic pneumonia
  - CBC with differential

- Sarcoidosis
  - Serum calcium

- Scleroderma/MCTD
  - ANA, Scl70, centromere, U1RNP

- Rheumatoid Arthritis
  - RF, C CP

- Sjogren's
  - ANA, Ro/SSA, La/SSB

- Idiopathic inflammatory myositis
  - Anti-PL3 and MPO (ANCA)

- Vasculitis
  - CTD-ILD

- Chronic Hypersensitivity Pneumonitis
  - HP panel (controversial)

- CT-ILD

- ESR, CRP

**Step 2C: Consider surgical lung biopsy**

- **Involve a multidisciplinary team at an ILD center to determine whether a surgical lung biopsy is required.**
- **A biopsy is performed, ask the surgeon to take multiple appropriately sized biopsies from 2-3 lobes (not just the lingula or middle lobe). Biopsy underaffected and mild-to-moderately affected areas. Do not biopsy only the most severely affected area. Have the biopsy reviewed by a pulmonary pathologist with expertise in ILD.**

**ATS/ERS/JRS/ALAT TREATMENT GUIDELINES FOR IPF**

**STRONG RECOMMENDATION:** "Most individuals should receive this intervention."

**CONDITIONAL RECOMMENDATION:** "Recognize that different choices will be appropriate for individual patients and that you must help each patient arrive at a management decision consistent with his or her values and preferences. **The majority of individuals in this situation would want the suggested course of action, but many would not.** Please note that corticosteroid monotherapy, N-acetylcysteine monotherapy, and combination therapy with prednisone + azathioprine are not recommended for IPF."

- Please note that corticosteroid monotherapy, N-acetylcysteine monotherapy, and combination therapy with prednisone + azathioprine are not recommended for IPF.

- Long-term oxygen therapy**
- Lung transplantation**

- Strongly recommended for IPF**
- Conditional recommendation for IPF**
- Pulmonary rehabilitation
- Nintedanib
- Pirfenidone
- Antidepressant therapy
- Corticosteroids during an "acute exacerbation"

- These recommendations apply only to adults diagnosed with idiopathic pulmonary fibrosis (IPF) and do not apply to those with other forms of pulmonary fibrosis or interstitial lung disease.

- *While indicated