## 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, pneumococcal, and COVID-19 vaccinations
- ☐ Provide information about your local PFF support group
- ☐ Consider lung transplant evaluation
- ☐ Consider clinical trial enrollment
- ☐ Consider pharmacological treatment after a diagnosis is confirmed
- ☐ 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

1 Rosas IO, et al. Interstitial lung disease: NHLBI Workshop on the Primary Prevention of Chronic Lung Diseases. Ann Am Thorac Soc. 2014 Apr;11 Suppl 3:S169–77.

## SUPPORT FOR YOU FROM THE PULMONARY FIBROSIS FOUNDATION

The mission of the Pulmonary Fibrosis Foundation is to accelerate the development of new treatments and ultimately a cure for pulmonary fibrosis. Until this goal is achieved, the PFF is committed to advancing improved care of patients with PF and providing unequaled support and education resources for patients, caregivers, family members, and health care providers.

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

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# ILD POCKET GUIDE

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
- Consider prone imaging when mild dependent ground-glass attenuation is present

## STEP 2: ATTEMPT TO IDENTIFY THE CAUSE OF ILD

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

| Disease | Questions/Findings |
|---|---|
| Drug/Radiation-induced ILD | Chemotherapy, amiodarone, nitrofurantoin, other drugs<br>Check PneumoTox.com. Radiation therapy to the chest |
| Connective Tissue | Joints, skin, Raynaud's, reflux, dry eyes/mouth, muscle weakness<br>Disease-related ILD or pain |
| Vasculitis | Sinus disease, hoarseness, hematuria, hemoptysis |
| Chronic Hypersensitivity Pneumonitis | • Exposure to mold sources (forced air heating, hot tubs, humidifiers, water damage or visible mold)<br>• Exposure to birds, down bedding, farming, or agriculture |
| Pneumoconioses | Occupational history |
| Familial ILD | Family history of ILD, sarcoidosis, home oxygen use, autoimmune disease |

### Step 2B: Order relevant blood tests

| Disease | Blood tests |
|---|---|
| Eosinophilic pneumonia | CBC with differential |
| Sarcoidosis | Serum calcium |
| Scleroderma/MCTD | ANA, Scl70, centromere, U1RNP |
| Rheumatoid Arthritis | RF, CCP |
| Sjogren's | ANA, Ro/SSA, La/SSB |
| Idiopathic inflammatory myositis | ANA, Jo-1, CK, myoglobin, aldolase, consider myositis panel |
| Vasculitis | Anti-PR3 and MPO (ANCA), creatinine |
| Chronic Hypersensitivity Pneumonitis | HP panel (controversial) |
| CTD-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.

If 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 unaffected 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

**Strongly recommended for IPF*:**

Long-term oxygen therapy**
Lung transplantation**

Conditionally recommended for IPF*:

Pulmonary rehabilitation
Nintedanib
Pirfenidone
Antacid 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
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." and "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 + N-acetylcysteine are **not** recommended for IPF.

From Raghu G, *et al., Am J Respir Crit Care Med 2015;192(2):238–48,* and Raghu G, *et al., Am J Respir Crit Care Med 2011;183(6):788–824.*

**When indicated