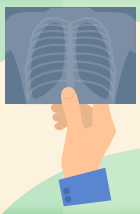
Pathways to a Diagnosis of IPF

NEW OR SUSPECTED ILD

Patients may present with:

- Dyspnea and/or cough
- Crackles on lung auscultation
- Exertional hypoxia
- Restrictive physiology on PFTs
- Abnormal chest imaging

ILD CONFIRMED BY CHEST CT



IS THIS ILD IDIOPATHIC?

1. **Review current and past medical conditions and medications.**
   - Prior cancer treated with chemotherapy or radiation
   - Medication history including amiodarone, nitrofurantoin, chemotherapy regimens
2. **Detdetailed social and environmental history**
   - Former or current tobacco use
   - Illicit inhaled or IV drug use
   - Environmental exposures (birds, down, mold, hot tubs, asbestos, etc)
3. **Detailed occupational history**
   - Agricultural work, ranching, exposure to metal and wood dust, hair dressers, mining, construction, stone cutting, work around asbestos
4. **Family history**
   - Are there family members with lung fibrosis or autoimmune disease?
5. **Are there signs or symptoms of autoimmune disease?**
   - Detailed review of symptoms
   - Physical examination with attention to autoimmune features
   - Family history
   - Serologies for autoimmune disease

YES

NO

EVALUATE CHEST HRCT PATTERN [1]

UIP	Probable UIP	Indeterminate	Alternative diagnosis
• Subpleural and basal predominant • Reticular abnormality • Honeycombing with or without peripheral traction bronchiectasis or bronchiolectasis	• Subpleural and basal predominant; • Reticular abnormality • Peripheral traction bronchiectasis or bronchiolectasis; • May have mild ground glass opacities	• Subpleural and basal predominant; • Subtle reticulation • May have mild ground glass or distortion suggesting an early UIP pattern • CT features do not suggest any specific etiology	• CT features and/or distribution suggest another diagnosis. • Examples include cysts, predominant ground glass, marked mosaic attenuation, consolidation, nodules, upper or mid-lung predominant, peribronchovascular or perilymphatic distribution. • May include extra-parenchymal findings suggesting an alternate diagnosis.

UIP

Probable UIP Indeterminate Alternative Diagnosis

Consider BAL, surgical lung biopsy. Pursue multidisciplinary discussion (MDD) for decision making. If lung biopsy is performed, review pathology and HRCT findings in MDD for consensus diagnosis.

Further evaluation (including HRCT) leading to a diagnosis?

YES

NOT IPF
Alternative Diagnosis

NOT IPF
Alternative Diagnosis

IPF DIAGNOSIS

MANAGEMENT OF IPF

Pharmacotherapy	Disease Course	Oxygen	Manage Comorbidities	Pulmonary Rehab	Transplant Referral
Two FDA approved therapies for IPF. Both slow rate of FVC decline by approximately 50%. Nintedanib – 150mg po BID [2] Pirfenidone – 801mg po TID [3] Both require LFT monitoring Not approved for use in combination Consider IPF clinical trials	Educate patients regarding variability in disease course. Predictive tools such as GAP index can provide staging information.[4] **Acute exacerbation:** acute and clinically significant worsening of respiratory symptoms over 2-4 weeks. Often requires hospitalization. *Patients should notify MD with any change in respiratory symptoms.*	Determine patient oxygen requirement: • At rest • With exertion • During sleep Prescribe oxygen if patients desaturate below 89%. Obstructive Sleep Apnea (OSA) is common and may contribute to sleep-related oxygen requirement.	Pulmonary Hypertension • Common (30-50%) and may contribute to mortality.[5] Obstructive Sleep Apnea • Prevalence up to 88% in IPF and classic clinical indicators may be absent.[6] Gastro esophageal Reflux Disease • Prevalence up to 88% in IPF and greater than 50% may be proximal.[7-9] • May be asymptomatic in a majority.	Improves: - 6 minute walk distance (6MWD) - Quality of life - Dyspnea Effect is not sustained upon stopping rehab participation.[10-12]	Disease course unpredictable; refer early. Indications for referral:[13] • UIP pattern on imaging or lung biopsy • FVC<80% of DLCO <40% • Any dyspnea or functional limitation due to IPF • Any O2 requirement *It is appropriate to refer at the time of diagnosis* 5 year transplant survival in IPF ~50%

REFERENCES

[1] Raghu G, et al. Diagnosis of Idiopathic Pulmonary Fibrosis. An Official ATS/ERS/JRS/ALAT Clinical Practice Guideline. Am J Respir Crit Care Med. 2018; 198(5): e44-68. | [2] Richeldi L, et al. Efficacy and safety of nintedanib in idiopathic pulmonary fibrosis. N Engl J Med. 2014; 370(22): 2017-82. | [3] King TE, et al. A phase 3 trial of pirfenidone in patients with idiopathic pulmonary fibrosis. N Engl J Med. 2014; 370(22):2083-92. | [4] Ley B, et al. A multidimensional index and staging system for idiopathic pulmonary fibrosis. Ann Intern Med. 2012; 156(10): 684-91. | [5] Raghu G, et al. Comorbidities in idiopathic pulmonary fibrosis patients: a systematic literature review. Eur Resir J. 2015; 46(4): 1113-30. | [6] Lancaster LH, et al. Obstructive sleep apnea is common in idiopathic pulmonary fibrosis. Chest. 2009; 136(6): 772-8. | [7] Tobin RW, et al. Increased prevalence of gastroesophageal reflux in patients with idiopathic pulmonary fibrosis. Am J Respir Crit Care Med. 1998; 158(6): 1804-8. | [8] Savarino E, et al. Gastro-oesophageal reflux and gastric aspiration in idiopathic pulmonary fibrosis patients. Eur Respir J. 2013; 42(5): 1322-31. | [9] Raghu G, et al. High prevalence of abnormal acid gastro-oesophageal reflux in idiopathic pulmonary fibrosis. Eur Respir J. 2006; 27(1): 136-42. | [10] Ferreira A, et al. Pulmonary rehabilitation in interstitial lung disease: benefits and predictors of response. Chest. 2009; 135(2): 442-7. | [11] Holland AE, et al. Short term improvement in exercise capacity and symptoms following exercise training in interstitial lung disease. Thorax. 2008; 63(6): 549-54. | [12] Swigris JJ, et al. Benefits of pulmonary rehabilitation in idiopathic pulmonary fibrosis. Respir Care. 2008; 56(6): 783-9. | [13] Weill D, et al. A consensus document for the selection of lung transplant candidates: 2014—An update from the Pulmonary Transplantation Council of the International Society for Heart and Lung Transplantation. The Journal of Heart and Lung Transplantation. 2015; 34:1-15.

This activity is supported by an independent educational grant from Boehringer Ingelheim Pharmaceuticals, Inc.

njhealth.org

1.877.CALLNJH (1.877.225.5654)

©2018 National Jewish Health

National Jewish Health®

Breathing Science is Life.®