Initial Outpatient Evaluation of Interstitial Lung Disease

Case related questions.

Scenario:
Mrs. F is a 65-year-old woman with a history of hypothyroidism who presents for a second opinion regarding interstitial lung disease. She reports that she first noted dyspnea about 2 years ago, which was attributed to issues with her thyroid medications. She never had a chest x-ray. Her shortness of breath was mildly progressive, but a few months ago, she developed a respiratory infection, and her breathing worsened significantly. She went to a local pulmonologist, who ordered a computed tomography (CT) scan, see Figure 1. Before proceeding with care locally, she wanted a second opinion and has come to see you.

Question 1: Given there is a broad differential diagnosis, how can the many etiologies of interstitial lung disease (ILD) be classified to facilitate work-up?

Question 2: What is a systematic approach to evaluation when considering the broad differential diagnosis of interstitial lung disease? What historical facts would help narrow the differential?

Scenario cont.:
Mrs. F notes 2 years of dyspnea with recent dramatic worsening. She notes lower back and knee pain, but no other joint symptoms. She thinks her fingers turn cool and numb in the cold, but she hasn't noted color changes. Other than occasional over-the-counter medicines, she has only ever taken levothyroxine. She has never smoked. She is from Mexico and spends about six months per year there still. There is no known mold or water damage in her homes. She previously owned a pet bird several years ago and does not have a hot tub.

Question 3: You plan to obtain updated pulmonary function tests, a six-minute walk test, review the CT scan with the radiologist and see her again in a month to discuss the results. What, if any, laboratory testing would you order?

Question 4: Are there specific radiologic patterns of ILD that can be helpful in diagnosis?

Scenario cont.:
You review Mrs. F's scan with the radiologist. There are clear peripheral reticular interstitial changes, but there is a significant amount of upper lobe involvement, making this less consistent with a UIP pattern. The primary radiographic differential diagnosis is NSIP versus fibrotic HP. Her labs return with an elevated ANA (1:640) and RF (96), but are otherwise normal, including the serum HP panel. PFTs show moderately severe restriction with a moderate diffusion defect, and her walk test demonstrated the lowest SpO2 at 92% with a distance of 880 feet. You suspect a diagnosis of interstitial pneumonia with autoimmune features (IPAF), but feel that fibrotic HP has not been excluded. She presents for follow-up.

Question 5: What is the most appropriate next diagnostic step?

Question 6: If you were to proceed with bronchoscopy and bronchoalveolar lavage (BAL), what BAL cellular profile would be most consistent with hypersensitivity pneumonitis?

Scenario cont.:
Because the differential includes fibrotic HP and NSIP/IPAF, the case is discussed at a multidisciplinary case conference. Options of treating with steroids for a short period are discussed as is the option of a biopsy. After extensive discussion with the patient about the recommendations, decision is made to pursue a surgical lung biopsy (considerations are patient preference to know exactly what she is dealing with vs the potential harm from a biopsy). You decide that BAL will not increase your ability to distinguish between the leading diagnoses. She undergoes a surgical lung biopsy without complication.

Question 7: When her biopsy results return, what is the best way to determine a final diagnosis?

Scenario conclusion:
Mrs. F's biopsy demonstrated patchy fibrosis with marked lymphoid aggregates and moderate to severe interstitial chronic inflammatory infiltrates, suggesting an underlying collagen vascular disease. A multidisciplinary discussion was held, and given the positive autoimmune serologies, atypical CT pattern for IPF, and pathology findings not consistent with fibrotic hypersensitivity pneumonitis, the consensus diagnosis was connective tissue disease-associated ILD/IPAF. Mrs. F was started on mycophenolate mofetil and her PFTs have remained stable while on therapy.

Initial Outpatient Evaluation of Interstitial Lung Disease

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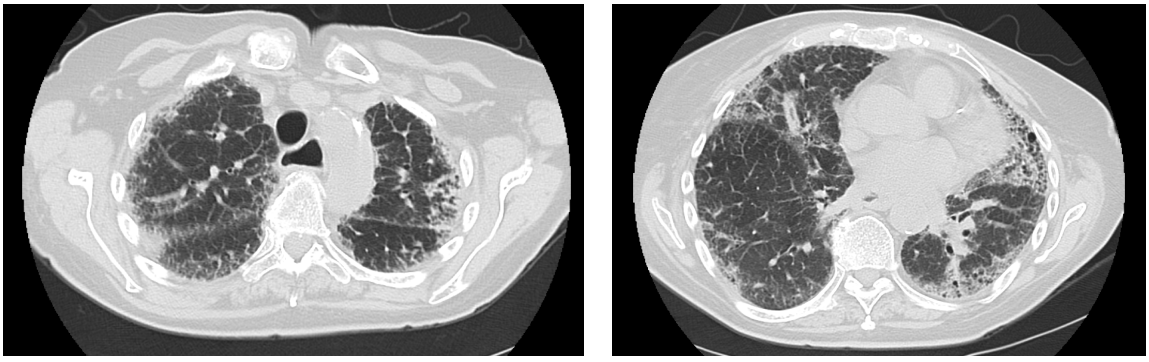
Educational Objectives:

1. Review the differential diagnosis of interstitial lung disease (ILD)
2. Develop a standard approach for an initial outpatient visit for ILD
3. Discuss the role of chest CT, bronchoscopy, and lung biopsy in the multidisciplinary diagnosis of ILD

Scenario:

Mrs. F is a 65-year-old woman with a history of hypothyroidism who presents for a second opinion regarding interstitial lung disease. She reports that she first noted dyspnea about 2 years ago, which was attributed to issues with her thyroid medications. She never had a chest x-ray. Her shortness of breath was mildly progressive, but a few months ago, she developed a respiratory infection, and her breathing worsened significantly. She went to a local pulmonologist, who ordered a computed tomography (CT) scan, see Figure 1. Before proceeding with care locally, she wanted a second opinion and has come to see you.

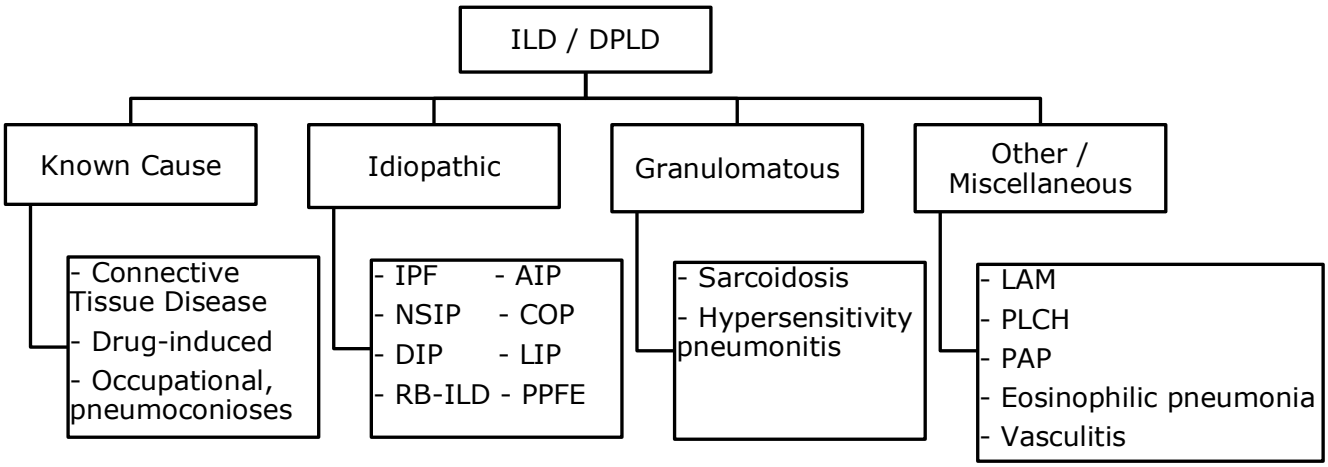
Figure 1. Representative axial CT scan images for the patient Mrs. F.



Question 1: Given there is a broad differential diagnosis, how can the many etiologies of ILD be classified to facilitate work-up?

Multiple ways exist to classify interstitial lung diseases, also known as diffuse parenchymal lung diseases (DPLDs). Figure 2 is a suggested method to organize the main categories of DPLDs.

Figure 2. Suggested schematic to organize the main types of ILD



***IPF= idiopathic pulmonary fibrosis, NSIP= non-specific interstitial pneumonitis, DIP= desquamative interstitial pneumonia, RB-ILD= respiratory bronchiolitis-ILD, AIP= acute interstitial pneumonia, COP= cryptogenic organizing pneumonia, LIP= lymphocytic interstitial pneumonia, PPFE= pleuroparenchymal fibroelastosis, HP= hypersensitivity pneumonitis, LAM= lymphangioleiomyomatosis, PLCH= pulmonary Langerhan's cell histiocytosis, PAP= pulmonary alveolar proteinosis. Major pneumoconioses include asbestosis, silicosis, and coal-worker's lung.*

Figure adapted from King TE. Idiopathic interstitial pneumonias: progress in classification, diagnosis, pathogenesis and management. Trans Am Clin Climatol Assoc 2004; 115: 43-78; and Travis et al. An official American Thoracic Society/European Respiratory Society statement: update of the international multidisciplinary classification of the idiopathic interstitial pneumonias. Am J Respir Crit Care Med. 2013;188:733-748.

Question 2: What is a systematic approach to evaluation when considering the broad differential diagnosis of interstitial lung disease? What historical facts would help narrow the differential?

The BTS guidelines provide the following "memory card" for initial ILD visits that can facilitate a systematic work-up:

Table 1. Memory card for the first clinic visit in suspected ILD

In addition to routine respiratory history and examination: • History of acid reflux, symptoms suggestive of connective tissue disease, including Raynaud's phenomenon • Detailed occupational history • Exposure to birds and other potential antigens • Current and previous drugs
Investigations for most (if not all) patients with suspected ILD: • Full blood count, chemistry, ESR, CRP, autoimmune serologies • Full pulmonary function test

• HRCT • Oxygen saturation at rest and with exertion
Additional tests in selected patients: • Suspected sarcoidosis: serum ACE level (controversial) • Suspected HP: precipitating antibody panel (controversial) • Suspected vasculitis: ANCA, urinalysis

Table adapted from Wells AU, Nirani H, British Thoracic Society Interstitial Lung Disease Group. Interstitial lung disease guideline: the British Thoracic Society in collaboration with Thoracic Society of Australia and New Zealand and the Irish Thoracic Society. Thorax. 2008;63:v1-v58.

The following historical elements can help narrow the differential diagnosis:

a. Symptom duration
   i. Acute: Drug-induced, HP, IPF exacerbation, AIP, COP, acute eosinophilic PNA
   ii. Episodic: HP, COP, chronic eosinophilic pneumonia

b. History of autoimmune diseases or symptoms
   i. Common diseases include rheumatoid arthritis, polymyositis/dermatomyositis, anti-synthetase syndrome, lupus, scleroderma, and Sjogren's
   ii. Symptoms may include arthritis/arthralgias, Raynaud's, dysphagia, characteristic rashes, and sicca symptoms. In some patients, lung disease may precede signs/symptoms of systemic autoimmune disease.
   iii. Some patients have ILD and symptoms or serologic results suggesting rheumatologic disease but do not meet the defined criteria for any specific autoimmune diagnosis. Such patients are considered to have interstitial pneumonia with autoimmune features (IPAF).

c. Medications/Drugs
   i. MANY, some examples in Table 2. The British Thoracic Society guidelines have an online summary and Pneumotox.com provides a searchable database.

Table 2. Medications implicated in pneumonitis/ILD

Antibiotics	Nitrofurantoin, daptomycin
Disease-modifying antirheumatic drugs	sulfasalazine, methotrexate, gold penicillamine, leflunomide, etanercept
Cardiovascular agents	amiodarone
Chemotherapeutic agents	bleomycin, cyclophosphamide, selective EGFR-inhibitors, immune checkpoint inhibitors
Illicit drugs	heroin, methadone, talc
Miscellaneous	radiation, aspirin, interferons

Table adapted from Wells AU, Nirani H, British Thoracic Society Interstitial Lung Disease Group. Interstitial lung disease guideline: the British Thoracic Society in collaboration with Thoracic Society of Australia and New Zealand and the Irish Thoracic Society. Thorax. 2008;63:v1-v58.

d. Occupational and other exposures: There are MANY potential environmental exposures to organic and inorganic antigens and a thorough history is needed including current and prior occupations, housing, pets, hobbies, etc. There are several published HP-specific questionnaires that can assist with history taking. See Table 3 for a summary of the major exposures and antigen types that were derived from this work:

Table 3. Major Exposures Causing Hypersensitivity Pneumonitis

Microbial Particulate Matter	Plant & Animal Proteins	Chemical Agents
• Water damage, flooding • Moldy hay/silage • Visible or smelly mold • Humidifiers/air conditioners • Hot tubs, steam saunas • Moldy wood • Organic matter (manure, compost) • Musical (wind) instruments • Humid internal environment • Dentistry products • CPAP	• Birds, bird droppings • Down or feather products • Farming • Vegetable production • Food production • Wood dust • Esparto grass	• Isocyanates • Metalworking fluids • Vapors, gases, fumes • Cleaning agents • Pesticides • Hair spray • Heated plastic vapors

e. Smoking history: There are a few DPLDs associated with cigarette smoking:
   i. Desquamative Interstitial Pneumonia (DIP)
   ii. Respiratory Bronchiolitis-Interstitial Lung Disease (RB-ILD)
   iii. Pulmonary Langerhans Cell Histiocytosis (PLCH)
   iv. Combined pulmonary fibrosis/emphysema (CPFE)

f. Family history:
   i. Familial pulmonary fibrosis (FPF), in which more than one relative is affected, can present with a variety of ILD phenotypes, though IPF and unclassifiable are the most common. FPF confers a poorer prognosis compared with sporadic ILD, so obtaining a family history can convey prognostic information as well as raise suspicion for a causative genetic variant.
   ii. Genetic variants causing ILD include those affecting surfactant metabolism and telomere maintenance. However, only about 1/4 of FPF can be explained by currently known genetic variants.

Scenario cont.:
Mrs. F notes 2 years of dyspnea with recent dramatic worsening. She notes lower back and knee pain, but no other joint symptoms. She thinks her fingers turn cool and numb in the cold, but she hasn't noted color changes. Other than occasional over-the-counter medicines, she has only ever taken levothyroxine. She has never smoked. She is from Mexico and spends about six months per year there still. There is no known mold or water damage in her homes. She previously owned a pet bird several years ago and does not have a hot tub.

Question 3: You plan to obtain updated pulmonary function tests, a six-minute walk test, review the CT scan with the radiologist and see her again in a month to discuss the results. What, if any, laboratory testing would you order?
There is no clear consensus on what laboratory tests need to be ordered. Because ILD can be the first manifestation of any of the above connective tissue diseases, consider sending the following tests:

- ANA
- RF, CCP
- Anti-Jo-1
- Myomarker panel
- Anti-Scl-70
- RNA polymerase III
- SS-A/SS-B
- Sm/RNP
- ANCA with reflex MPO/PR3
- CK, aldolase

For hypersensitivity pneumonitis, serum precipitins (a select IgG panel) can be sent. However, this a limited panel that does not cover the wide array of possible antigens, and a negative result does not exclude HP.

Question 4: Are there specific radiologic patterns of ILD that can be helpful in diagnosis?

The ATS clinical practice guideline on IPF defines the radiologic patterns of abnormality as classified into UIP, probable UIP, indeterminate for UIP, and alternative diagnosis, Table 4. Although the UIP pattern is not unique to IPF, radiologic UIP or probable UIP patterns in the absence of a suspected cause (such as autoimmune disease or inhaled antigen) can be sufficient to diagnose IPF per these guidelines.

High-resolution ILD protocol chest CTs are preferred as these typically include prone/supine imaging (to exclude dependent atelectasis) and inspiratory/expiratory images to evaluate for air trapping (such as can be seen in HP). They also provide thinner cuts with post-processing to enhance edges for sharper image quality than conventional CT chest protocols.

Table 4. CT classification of UIP

UIP	Probable UIP	Indeterminate for UIP	Alternative Diagnosis
Subpleural and basal predominant; distribution is often heterogeneous*	Subpleural and basal predominant; distribution is often heterogeneous	Subpleural and basal predominant	Findings suggestive of another diagnosis, including: • CT features: ° Cysts ° Marked mosaic attenuation ° Predominant GGO ° Profuse micronodules ° Centrilobular nodules ° Nodules ° Consolidation • Predominant distribution: ° Peribronchovascular ° Perilymphatic ° Upper or mid-lung • Other: ° Pleural plaques (consider asbestosis) ° Dilated esophagus (consider CTD) ° Distal clavicular erosions (consider RA) ° Extensive lymph node enlargement (consider other etiologies) ° Pleural effusions, pleural thickening (consider CTD/drugs)
Honeycombing with or without peripheral traction bronchiectasis or bronchiolectasis†	Reticular pattern with peripheral traction bronchiectasis or bronchiolectasis May have mild GGO	Subtle reticulation; may have mild GGO or distortion ("early UIP pattern") CT features and/or distribution of lung fibrosis that do not suggest any specific etiology ("truly indeterminate for UIP")	

Definition of abbreviations: CT = computed tomography; CTD = connective tissue disease; GGO = ground-glass opacities; RA = rheumatoid arthritis; UIP = usual interstitial pneumonia.
*Variants of distribution: occasionally diffuse, may be asymmetrical.
†Superimposed CT features: mild GGO, reticular pattern, pulmonary ossification.

Table borrowed from Raghu et al. Diagnosis of Idiopathic Pulmonary Fibrosis. American Journal of Respiratory and Critical Care Medicine. Volume 198 Number 5. 2018. PMID 30168753.

Alternative diagnosis CT patterns suggest a diagnosis other than IPF. One of these patterns is NSIP, which is characterized by diffuse ground glass (classically with subpleural sparing) and basilar-predominant fibrotic changes and is most commonly associated with autoimmune disease, drug reactions, as well as idiopathic disease. The ATS has also published guidelines defining radiologic patterns that are typical for nonfibrotic and fibrotic HP (including mosaicism/air trapping, triple density sign, etc).

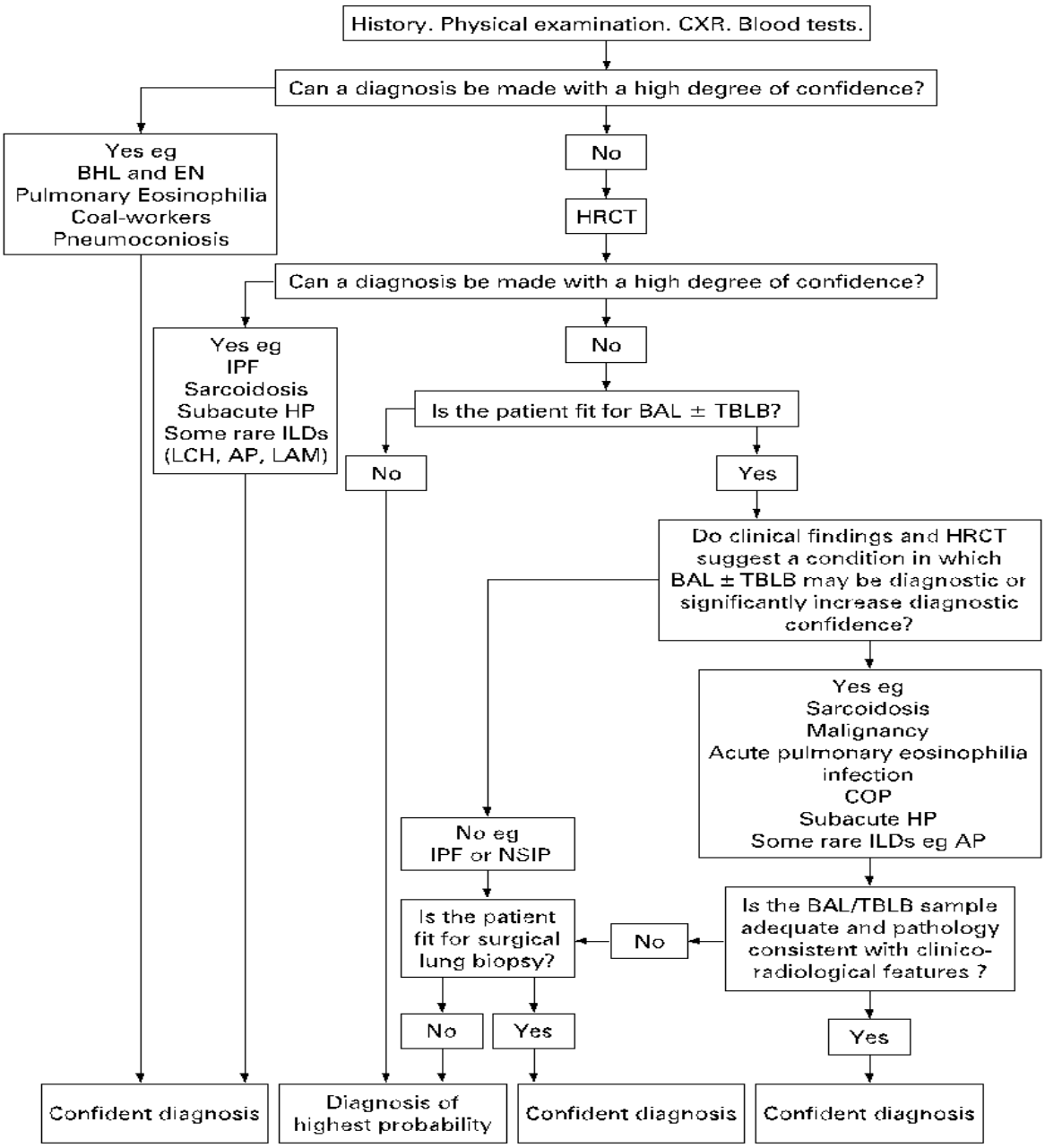
Scenario cont.:
You review Mrs. F's scan with the radiologist. There are clear peripheral reticular interstitial changes, but there is a significant amount of upper lobe involvement, making this less consistent with a UIP pattern. The primary radiographic differential diagnosis is NSIP versus fibrotic HP. Her labs return with an elevated ANA (1:640) and RF (96), but are otherwise normal, including the serum HP panel. PFTs show moderately severe restriction with a moderate diffusion defect, and her walk test demonstrated the lowest SpO2 at 92% with a distance of 880 feet. You suspect a diagnosis of interstitial pneumonia with autoimmune features (IPAF), but feel that fibrotic HP has not been excluded. She presents for follow-up.

Question 5: What is the most appropriate next diagnostic step?

- Empiric treatment (e.g. steroid trial, antifibrotic)?
- Bronchoscopy with BAL +/- transbronchial lung biopsy or cryobiopsy?
- Referral for surgical lung biopsy?

There is no definite answer, but the adjacent algorithm from the 2008 BTS guidelines and the recently updated ATS guidelines (Figure 3) can help us make the decision. Because the HRCT did not provide us with a diagnosis that can be made with a "high degree of confidence," we should consider proceeding with another diagnostic step. Additionally, for this patient, we should review the list of diagnoses for which a BAL/TBBx and cryobiopsy might increase the diagnostic confidence. Review at Multidisciplinary Discussion (Pulmonology, Radiology, Pathology) can also be helpful prior to making a decision about referral for biopsy and whether this will ultimately change management.

Figure 3. Diagnostic approach for ILD



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Question 6: If you were to proceed with bronchoscopy and bronchoalveolar lavage (BAL), what BAL cellular profile would be most consistent with hypersensitivity pneumonitis?

BAL lymphocytosis is most suggestive of hypersensitivity pneumonitis. In general, BAL alone has limited utility in the diagnosis of ILD, see Table 5. It is most helpful in cases of eosinophilic pneumonia, pulmonary hemorrhage, and pulmonary alveolar proteinosis.

Table 5. Bronchoalveolar lavage findings that are potentially useful in ILD diagnosis

BAL finding	Consistent interpretation/suggested diagnosis
Eosinophils ⩾25%	Eosinophilic pneumonia
Lymphocytes ⩾25%	Sarcoidosis, HP, cellular NSIP, drug reaction, CBD, LIP, lymphoproliferative disorder
Neutrophils ⩾50%	AIP, DAD, AEIPF, pulmonary infection
Bloody fluid	Pulmonary haemorrhage, DAH
High haemosiderin score	DAH, DAD
CD1a+ cells >4%	PLCH
Milky BAL fluid with PAS-positive amorphous debris	PAP
Monotypic lymphocytes	Pulmonary lymphomatous malignancy
Malignant cells	Pulmonary malignancy
Squamous epithelial cells >5%	Unsuitable sample due to upper airway secretion contamination
Bronchial epithelial cells >5%	BAL sample may be unsuitable for cell analysis

Table borrowed from Meyer KC, Raghu G. Bronchoalveolar lavage for the evaluation of interstitial lung disease: is it clinically useful? Eur Respir J. 2011;38:761-769.

Scenario cont.:
Because the differential includes fibrotic HP and NSIP/IPAF, the case is discussed at a multidisciplinary case conference. Options of treating with steroids for a short period are discussed as is the option of a biopsy. After extensive discussion with the patient about the recommendations, decision is made to pursue a surgical lung biopsy (considerations are patient preference to know exactly what she is dealing with vs the potential harm from a biopsy). You decide that BAL will not increase your ability to distinguish between the leading diagnoses. She undergoes a surgical lung biopsy without complication.

Question 7: When her biopsy results return, what is the best way to determine a final diagnosis?

To more confidently make a final diagnosis, it is important to have a multidisciplinary discussion involving pulmonologists, radiologists, and pathologists. Flaherty et al. conducted a study to assess the utility of the multidisciplinary discussion. The study consisted of several steps, from pulmonologists and radiologists reviewing the HRCT individually to a consensus discussion including pulmonologists, radiologists, and pathologists.

Although there is no way to know if the final diagnoses were "correct," the researchers measured inter-observer agreement at each stage, outlined in Table 6:

Table 6. Interobserver agreement at each diagnostic step

TABLE 3. INTEROBSERVER AGREEMENT AT EACH DIAGNOSTIC STEP

Step	Clinicians [κ (95% CI)]	Radiologists [κ (95% CI)]	Clinicians–Radiologists [κ (95% CI)]	All Observers [κ (95% CI)]
1	0.41 (0.29, 0.52)	0.72 (0.57, 0.86)	0.39 (0.29, 0.49)	NA
2	0.51 (0.37, 0.64)	0.80 (0.67, 0.93)	0.44 (0.34, 0.54)	NA
3	0.67 (0.54, 0.79)	0.78 (0.65, 0.91)	0.55 (0.44, 0.66)	NA
4	0.75 (0.64, 0.86)	0.84 (0.72, 0.96)	0.78 (0.70, 0.86)	0.79 (0.71, 0.86)
5	0.86 (0.76, 0.95)	0.90 (0.80, 0.99)	0.88 (0.81, 0.96)	0.88 (0.81, 0.94)

Table borrowed from Flaherty KR, King TE, Raghu G, et al. Idiopathic interstitial pneumonia: what is the effect of a multidisciplinary approach to diagnosis? Am J Respir Crit Care Med. 2004;170:904-910.

Importantly, the final diagnosis was more influenced by pathologists in cases of non-IPF idiopathic interstitial pneumonias compared to IPF.

Figure 4. Multidisciplinary discussion to integrate radiologic and histologic pattern can help make confident IPF diagnoses

IPF suspected*		Histopathology pattern†			
		UIP	Probable UIP	Indeterminate for UIP or biopsy not performed	Alternative diagnosis
HRCT pattern	UIP	IPF	IPF	IPF	Non-IPF dx
	Probable UIP	IPF	IPF	IPF (Likely)‡	Non-IPF dx
	Indeterminate	IPF	IPF (Likely)‡	Indeterminate§	Non-IPF dx
	Alternative diagnosis	IPF (Likely)‡	Indeterminate§	Non-IPF dx	Non-IPF dx

Figure borrowed from Raghu et al Am J Respir Crit Care Med 205e18-e47. DOI: 10.1164/rccm.202202-0399ST

Scenario conclusion:
Mrs. F's biopsy demonstrated patchy fibrosis with marked lymphoid aggregates and moderate to severe interstitial chronic inflammatory infiltrates, suggesting an underlying collagen vascular disease. A multidisciplinary discussion was held, and given the positive autoimmune serologies, atypical CT pattern for IPF, and pathology findings not consistent with fibrotic hypersensitivity pneumonitis, the consensus diagnosis was connective tissue disease-associated ILD/IPAF. Mrs. F was started on mycophenolate mofetil and her PFTs have remained stable while on therapy.

References:

1. King TE. Idiopathic interstitial pneumonias: progress in classification, diagnosis, pathogenesis and management. *Trans Am Clin Climatol Assoc* 2004; 115: 43-78.
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Post-Test Case Related Questions

1. Which of the following ILDs is NOT most commonly seen in patients with current smoking?

    a. Pulmonary Langerhan's Cell Histiocytosis
    b. Respiratory Bronchiolitis Interstitial Lung Disease
    c. Lymphangioleiomyomatosis
    d. Desquamative Interstitial Pneumonia

2. A 35-year-old woman presents to the office for new onset dyspnea and possible interstitial changes on her CXR. She has a CT scan and ultimately undergoes a bronchoscopy with bronchoalveolar lavage and transbronchial lung biopsy as part of her evaluation. The BAL fluid reveals the presence of CD1a+ cells. Which of the following is the most likely diagnosis based on the BAL fluid findings alone?

    a. Pulmonary Alveolar Proteinosis
    b. Hypersensitivity Pneumonitis
    c. Lymphocytic Interstitial Pneumonia
    d. Pulmonary Langerhan's Cell Histiocytosis

3. A 65-year-old man with a history only notable for hypertension presents to the clinic for evaluation of ILD. He has no clear systemic symptoms to suggest connective tissue disease, and the remainder of the clinical history is largely unrevealing. Serologic testing is unremarkable. He has a high-resolution CT chest that reveals findings concerning either idiopathic pulmonary fibrosis or non-specific interstitial pneumonitis. However, at this point, you remain uncertain about his underlying diagnosis and decide to consider additional testing with tissue biopsy. He has a history of hypertension treated with lisinopril 5 mg daily and no other significant co-morbidities. His PFTs reveal an FEV1 of 50% predicted (declined from 65% one year prior). He does not desaturate with ambulation. His 6MWT distance is normal for his age. Which of the following would be the best approach to his management moving forward according to established BTS guidelines?

    a. Bronchoscopy with bronchoalveolar lavage
    b. Bronchoscopy with bronchoalveolar lavage and transbronchial lung biopsy
    c. Surgical lung biopsy
    d. Initiate empiric treatment for the most likely cause of his ILD and monitor with serial PFTs and six-minute walk tests

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