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SECOND EDITION

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Principles and Practice of Hospital Medicine

Second Edition

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CHAPTER 233

Interstitial Lung Diseases/Diffuse Parenchymal Lung Diseases

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1. Does the hospitalized patient with unexplained dyspnea have Interstitial Lung Disease (ILD)?
2. How and why should idiopathic pulmonary fibrosis (IPF) be differentiated from other forms of ILD?
3. How do you distinguish an acute exacerbation of IPF from other causes of worsening in ILD?
4. What are the indications for pulmonary consultation? For bronchoscopy in the diagnosis or evaluation of ILD?
5. When do you use newer antifibrotic agents in hospitalized patients with ILD?
6. How do you discharge patients with high oxygen requirements to home?

INTRODUCTION

■ EPIDEMIOLOGY

The interstitial lung diseases (ILDs) are a heterogeneous group of disorders with the common feature of inflammatory or fibrotic injury to the lung parenchyma. These disorders are also described as the diffuse parenchymal lung diseases. Numerous potential etiologies for ILD may be broadly divided into five categories: idiopathic, drug/medication related, environmental/occupational, genetic/hereditary and autoimmune associated (Table 233-1). While considered rare, these diseases affect approximately 500,000 individuals in the United States each year and result in 40,000 deaths, comparable to the number of deaths from breast cancer. The exact epidemiology is difficult to determine due to misidentification of patients as having more common disorders such as congestive heart failure or chronic obstructive pulmonary disease (COPD). The epidemiology varies based on the ILD subtype.

CLASSIFICATION AND COMMON PRESENTATION

The term ILD encompasses a diverse group of diseases. The American Thoracic Society and European Respiratory Society developed a two-level classification system to facilitate the clinical evaluation of ILD (Figure 233-1). This system divides ILDs initially based on specific mechanisms of disease into: disorders of known etiology, idiopathic interstitial pneumonias (IIPs), granulomatous diseases and rare diseases. The idiopathic interstitial pneumonias are further classified into a second level based on histologic appearance. We will focus primarily on the IIPs and mention several other ILDs of particular relevance to the hospitalist. A general approach to the diagnostic evaluation of suspected ILD is presented in Figure 233-2.

■ IDIOPATHIC PULMONARY FIBROSIS

Idiopathic pulmonary fibrosis primarily affects individuals over the age of 60. Recent epidemiologic studies show the prevalence increases with each successive decade from 18.7-23.3/100,000 in 55 to 64 year olds to 29.3-50.0/100,000 in 65 to 74 year olds to 48.4-87.9/100,000 in adults over than 85. IPF affects approximately 200,000 people in the United States and results in 20,000 deaths per year. A notable exception to the older age of onset is familial IPF. In this genetic disorder, patients may present two to three decades earlier with symptomatic disease. The median survival is 3 to 5 years from the time of diagnosis in symptomatic patients. Diagnosis is made based on a consistent history (slowly progressive dyspnea), findings of dry, velcro-like crackles on examination and a radiograph with basilar predominant interstitial changes (Figure 233-3). The hallmarks of IPF on chest computed tomography (CT) include basilar predominant subpleural reticulation and honeycombing with traction bronchiectasis (Figure 233-4). Although ground glass opacities may be seen during acute exacerbations, this should be a minor feature of a baseline CT study. The diagnosis of IPF mandates the exclusion of other etiologies including environmental exposures, medications and autoimmune diseases.

■ NONSPECIFIC INTERSTITIAL PNEUMONIA (NSIP)

Nonspecific interstitial pneumonia (NSIP) is the form of ILD most common in patients with autoimmune disease. This form of ILD is more common in women than men and has an earlier age of onset than IPF. The diagnosis is typically suspected based on the clinical

TABLE 233-1 Etiologies of Interstitial Lung Disease (ILD) by Category

Autoimmune
Scleroderma
Rheumatoid arthritis
Systemic lupus erythematosis
Polymyositis/dermatomyositis
Sjogren syndrome
Occupational/environmental
Asbestosis
Silicosis
Berylliosis
Bird fancier's lung
Idiopathic
Medication-induced
Common/conventional:
Amiodarone
Bleomycin
Methotrexate
Biologics
TNF inhibitors
Genetic/hereditary
Mucin 5b (Muc5b) variant
Hermansky-Pudlak
Surfactant protein A&C deficiency
Telomerase mutations

symptoms and radiograph showing patchy ground glass opacities (GGO) with minimal fibrosis (Figure 233-4). Open lung biopsy is necessary for histologic diagnosis. For the purpose of therapy, a radiographic "diagnosis" of NSIP is often sufficient. The prognosis of NSIP is generally good but there is a subset of patients who develop progressive fibrosis and have a worse outcome.

■ RESPIRATORY BRONCHIOLITIS-ILD AND DESQUAMATIVE INTERSTITIAL PNEUMONIA

Respiratory-bronchiolitis interstitial lung disease (RB-ILD) and desquamative interstitial pneumonia (DIP) represent a poorly appreciated spectrum of smoking-related lung disease. Although they occur most frequently in smokers, they may occasionally occur in nonsmokers. These disorders may develop years after starting smoking. Unlike the more commonly encountered COPD and emphysema, RB-ILD and DIP are largely reversible with avoidance of cigarette smoke, including second hand exposures. The age of onset is variable, but tends to be younger than IPF. Definitive diagnosis requires lung biopsy. Individuals who develop this disorder without apparent tobacco smoke exposure may be treated with corticosteroids, but the response is variable.

■ ORGANIZING PNEUMONIA

Organizing pneumonia is a common and nonspecific lung injury pattern which can be encountered in a number of situations including medication-related, with autoimmune disease and following viral infections. When the cause is known or suspected, it is called organizing pneumonia. When the cause is unknown, the diagnosis is cryptogenic organizing pneumonia. The presentation may be virtually identical to that of pneumonia. Patients frequently receive one or several courses of antibiotics prior to diagnosis. The chest CT may show patchy ground glass opacities or dense infiltrates (Figure 233-4). The diagnosis is made by surgical lung biopsy but is often suspected based on clinical presentation. Prognosis is generally good as patients typically respond to corticosteroids.

■ ACUTE INTERSTITIAL PNEUMONITIS

Acute interstitial pneumonitis (AIP), sometimes referred to as the Hamman-Rich syndrome, is the most dreaded presentation of ILD. This enigmatic disorder may occur in the absence of any apparent trigger, although it may also complicate autoimmune disease. The age at onset is variable. Patients present with rapidly progressive dyspnea over a few weeks to months. Radiographs are indistinguishable from acute respiratory distress syndrome (ARDS), leading some people to refer to AIP as a form of idiopathic ARDS. Infection must be excluded and/or treated empirically. The prognosis is dismal with over 70% of patients dying of respiratory failure within weeks of presentation. High-dose corticosteroids have been used with occasional success in AIP particularly in the setting of underlying autoimmune disease.

■ SARCOIDOSIS

Sarcoidosis, the most common form of ILD, is a systemic disorder which can manifest in the skin, lung, heart, liver, bone marrow or the peripheral or central nervous system. The etiology remains unknown.

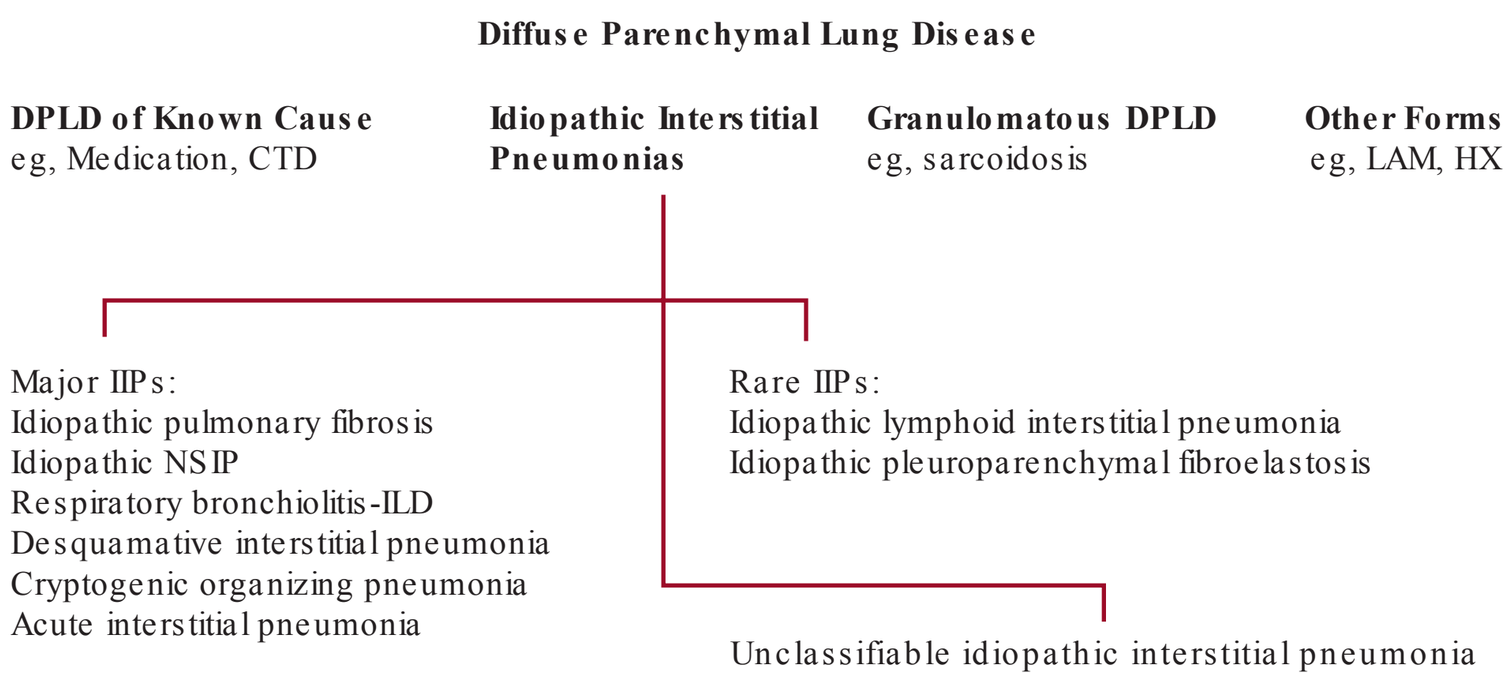

Figure 233-1 ATS/ERS Classification of Interstitial Lung Disease (Diffuse Parenchymal Lung Disease, DPLD). Adapted from An official American Thoracic Society/European Respiratory Society statement: Update of the international multidisciplinary classification of the idiopathic interstitial pneumonias. This updated joint statement of the American Thoracic Society (ATS), and the European Respiratory Society (ERS) was adopted by the ATS board of director the ERS Executive Committee in 2013. Am J Respir Crit Care Med. 2013;188(6):733-748.

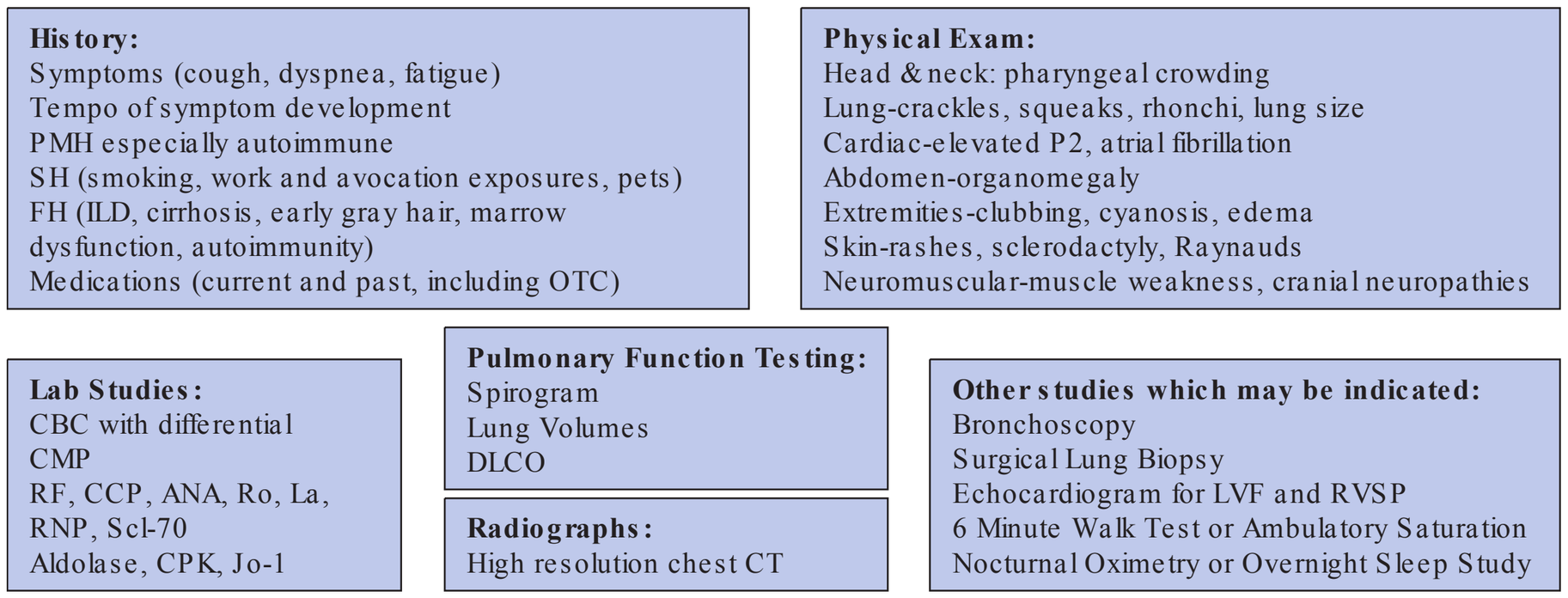

Figure 233-2 Standardized evaluation for Interstitial Lung Disease (ILD). A standardized evaluation for a new diagnosis of ILD includes a comprehensive history and physical examination as well as pulmonary function tests (PFTs), chest computed tomography (CT) and targeted laboratory studies. In appropriate situations, bronchoscopy and/or surgical biopsy may be indicated. Assessment of oxygen requirement and cardiac function are relevant in many patients. ANA, antinuclear antibody; CBC, complete blood count; CCP, anticitrullinated protein antibody; CMP, comprehensive metabolic panel; CPK, creatine phosphokinase; DLCO, diffusing capacity of the lung for carbon monoxide; FH, family history; Jo-1, Jo-1 antibody; La, La antibody; LVF, left ventricular function; OTC, over the counter medication; P2, second heart sound; PMH, past medical history; RF, rheumatoid factor; RNP, ribonucleoprotein antibody; Ro, Ro antibody; RVSP, right ventricular systolic pressure; Scl70-anti, Scl70 antibody; SH, social history.

It is more common in women, African Americans and people of Scandanavian and Irish descent. The diagnosis of pulmonary sarcoidosis is suggested by upper lobe interstitial changes on radiography with or without associated bilateral mediastinal and hilar lymph node enlargement (Figure 233-3). In contrast to many other forms of ILD, crackles on examination are a feature in later stage disease. Diagnosis is made by identification of noncaseating granulomas on biopsy of involved tissue. Pulmonary sarcoidosis is typically diagnosed by endobronchial biopsy, transbronchial lung biopsy or fine needle aspiration of involved lymph nodes. The clinical course is highly variable, with some individuals developing destructive fibrocavitary disease and others having asymptomatic lymph node enlargement.

■ AUTOIMMUNE ASSOCIATED ILD

Autoimmune-associated ILD should be suspected in young patients presenting with ILD in the setting of a constellation of systemic symptoms. In this case, the patient presented with pneumomediastinum in the setting of new onset polymyositis. The recognition of systemic symptoms may be difficult in the context of a dramatic presentation. However, if these symptoms are identified, biochemical and serologic testing can quickly confirm the underlying diagnosis. While the sensitivity and specificity of studies may vary regionally, CPK, aldolase, ANA, RF, RNP, Ro/La, Scl-70 and myositis antibodies (including Jo-1) are useful as an initial screen.

■ DRUG ASSOCIATED ILD

Drug-associated ILD can be difficult to identify since the causative medication may have been present for months or years prior to the onset of lung disease. The number of medications associated with ILD is increasing rapidly. This is especially true of the many biological agents in clinical use, particularly in the treatment of autoimmune disease and malignancy. A useful resource for determining if a medication has been associated with the development of ILD is Pneumotox (www.pneumotox.com). The recognition of drug-associated ILD

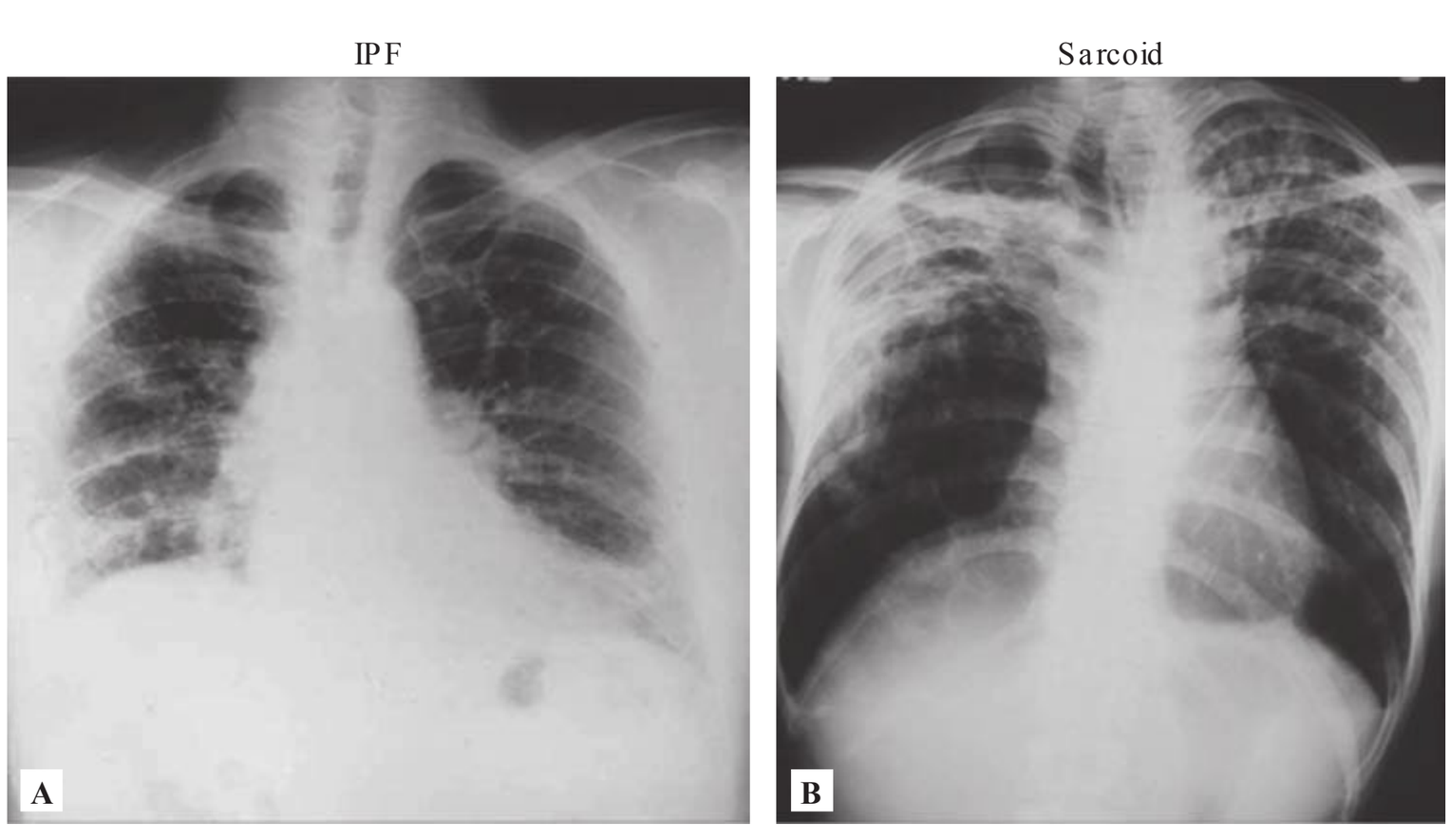

Figure 233-3 Representative chest x-rays that can be seen in patients with idiopathic pulmonary fibrosis (IPF) and sarcoid. IPF is characterized by basilar predominant interstitial markings. By contrast, sarcoid typically shows an upper lobe distribution of fibrosis which may also be accompanied by bilateral hilar adenopathy.

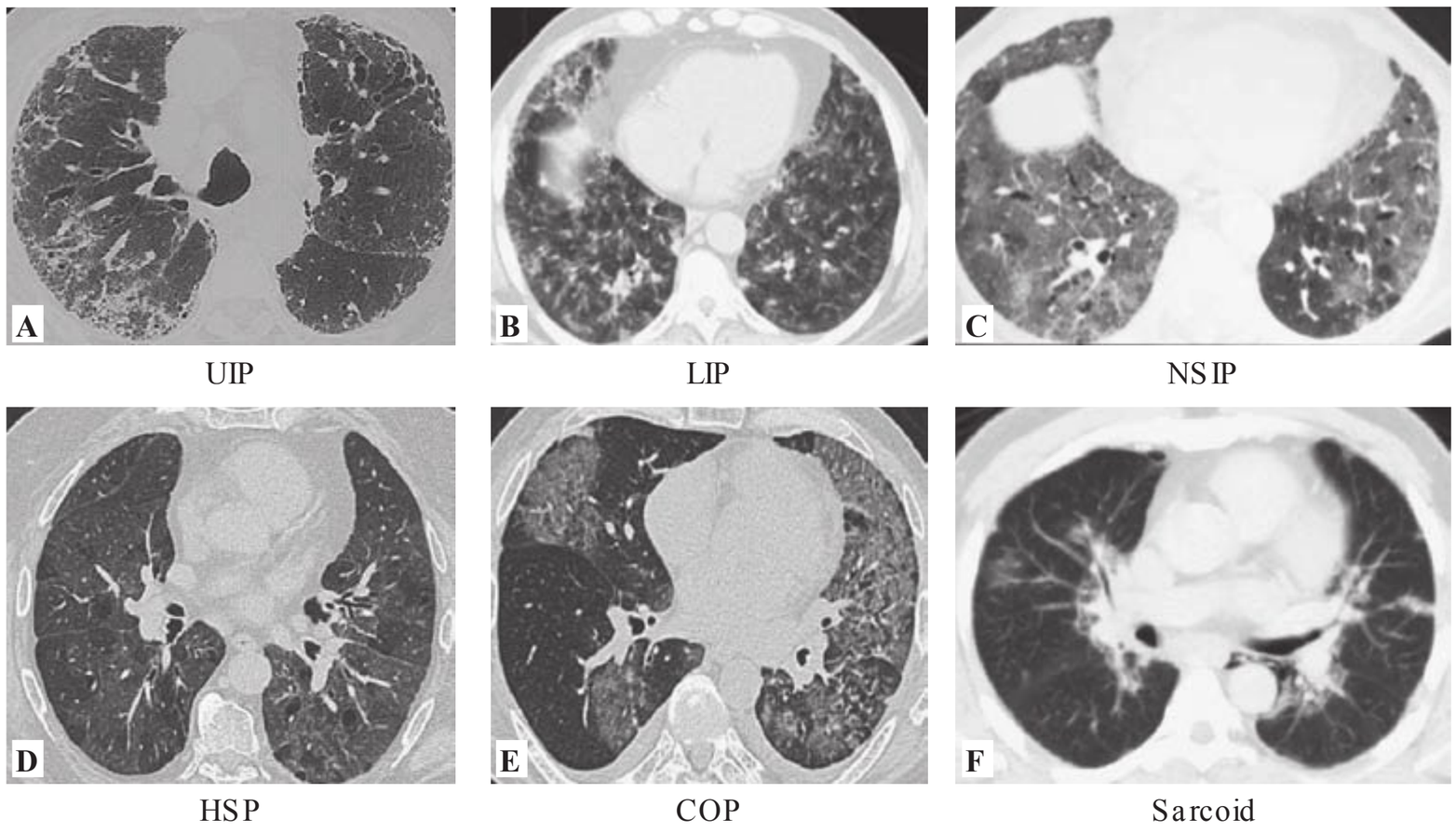

Figure 233-4 Representative computed tomography (CT) patterns that can be seen in patients with interstitial lung disease (ILD). ILD can be characterized by many different radiographic patterns. With the exception of usual interstitial pneumonia (UIP), there is substantial overlap among different forms of ILD. A number of the patterns are shown with the associated pathology noted. (A) Usual interstitial pneumonia (UIP) is characterized honeycombing, traction bronchiectasis and increased reticular markings. (B) Lymphocytic interstitial pneumonia (LIP) may demonstrate a micronodular pattern. (C) Nonspecific interstitial pneumonia (NSIP) often appears as a diffuse GGO. (D) Hypersensitivity pneumonitis (HSP) appears as a nearly homogeneous pattern of GGO. (E) Cryptogenic organizing pneumonia (COP) appears as a patchy infiltrate which may be GGO or dense. (F) Sarcoid can present with bronchovascular infiltrates and hilar adenopathy.

depends on ruling out other potential etiologies such as heart failure or an underlying autoimmune disease. In addition to evaluating prescription medications, patients should be asked about over-the-counter and herbal remedies as well as dietary supplements. Discontinuation of the offending medication results in improvement of symptoms within weeks to months. Some patients may require a several week course of systemic corticosteroids to limit lung injury while the medication is washing out.

■ HYPERSENSITIVITY PNEUMONITIS

Hypersensitivity pneumonitis (HP) is caused by an inappropriate inflammatory response to an environmental exposure. Hypersensitivity pneumonitis may present as an acute febrile, flu-like illness but may progress to a chronic phase that results in lung fibrosis. Characteristic findings include a mixed restrictive/obstructive pattern on pulmonary function testing and a CT with a combination of ground glass opacities and air trapping (Figure 233-4). It may be difficult to distinguish HP from other fibrosing lung diseases such as fibrotic NSIP and IPF. While the history may be suggestive of a potential offending antigen, it is often difficult to identify the cause. Common occupations associated with HP include farming, chemical manufacturing and bird breeding. Household exposures include mold, hot tub exposures and domestic and farm animals (ie, house birds and chickens). The key to treatment is identifying the causative agent and eliminating continued exposure. In situations in which the cause remains unclear, a trial of immunosuppression may help in slowing the progression of disease.

DIAGNOSIS

■ CLINICAL FEATURES

The presenting symptoms of dyspnea, cough and fatigue are common to a number of more prevalent diseases including pneumonia, heart failure, and COPD. A detailed history may bring out features which are helpful in discriminating these potential diagnoses. Have the symptoms truly been acute or has there been a more protracted period of decline? While ILD may have a fulminant acute presentation, many patients when prompted will give a history of a more subacute decline preceding the acute illness. Have the symptoms responded appropriately to a reasonable therapeutic intervention? Patients who fail to respond to multiple courses of antibiotics or who have worsening dyspnea on exertion despite prior efforts at diuresis should raise the concern for ILD.

Ultimately the diagnosis is based on a comprehensive history and physical (Figure 233-2). The history is critical in initially diagnosing ILD and distinguishing IPF with its very poor prognosis from other potentially more treatable forms of ILD. Many a patient with bird fancier's lung has passed through the hands of capable physicians who neglected to ask about environmental exposures. It is not unusual to find that a patient has been seen and hospitalized repeatedly for pneumonia or bronchitis.

Physical findings on pulmonary examination may be nonspecific with crackles or squeaks or normal sounds. Perhaps the most useful finding is the basilar predominant "velcro" crackles often heard in IPF. Clubbing is variably associated with ILD, but may occur with other pulmonary and nonpulmonary disorders. Systemic signs of autoimmune disease (Raynaud's, arthritis, rash) can be helpful in raising the suspicion for a collagen vascular disease-associated ILD.

■ PULMONARY FUNCTION TESTING (PFT)

The role of spirometry in the monitoring of airways disease (COPD, asthma) in the hospitalized patient is well established. Less so is the role of pulmonary function testing in the patient with ILD. The classic features of ILD are restrictive, including forced vital capacity less than 80% predicted for age without evidence of obstruction, and total lung capacity less than 80% predicted. In addition, diffusing capacity (DLCO) is typically reduced with a single breath DLCO less than 80% predicted. In the acute setting PFTs are often deferred; however, establishing posttreatment PFTs are invaluable in subsequent patient care. Not only do PFTs provide an objective measure of disease severity, they can provide an important clue as to coexistent pulmonary hypertension (PH) if the DLCO is reduced out of proportion to the lung volumes. This may also be seen in patients who

develop ILD in the setting of concurrent emphysema, appropriately named combined pulmonary fibrosis with emphysema.

■ IMAGING

Chest radiographs may show increased interstitial markings or reticulo-nodular changes which provide an initial suggestion of ILD. High-resolution chest CT scans are significantly more helpful, with representative CT patterns illustrated in Figure 233-4. A diagnosis of IPF can be made by a characteristic radiograph and exclusion of all other etiologies.

■ BRONCHOSCOPY AND LUNG BIOPSY

Definitive histologic diagnosis of IIPs other than IPF depends on lung biopsy. Bronchoalveolar lavage may be helpful in the diagnosis of infection which may complicate or mimic ILD. It is also useful for assessment of eosinophilia which is characteristic of a number of forms of ILD. Transbronchial biopsies may be diagnostic for sarcoidosis or for excluding malignancy but have poor sensitivity for diagnosing most other ILDs. Recently, a cryoprobe has been developed that can obtain larger bronchoscopic biopsy specimens although its utility in diagnosing ILD has not been established.

Surgical biopsy diagnosis should be reserved for patients with atypical presentations or who fail to respond appropriately to initial therapy. The decision to pursue surgical biopsy in a hospitalized patient with suspected ILD should be made in consultation with a pulmonologist. Patients with ILD frequently have multiple comorbidities that may complicate surgical biopsy, such as underlying cardiac disease. Many anecdotal reports suggest exacerbation of lung disease in the setting of a surgical lung biopsy. These exacerbations may be life threatening and often result in a permanent decrement in lung function. This risk should temper the decision to pursue surgical biopsy in many patients. In addition, many patients do not need biopsies. ILD presenting in the setting of a known trigger (such as medication or autoimmune disease) rarely requires a tissue diagnosis.

- Early involvement of pulmonary consultation in patients with suspected ILD is critical due to the potential complexity of diagnosis and the need for long-term follow-up. A specialist should become involved early in the hospitalization to guide diagnostic and therapeutic decision making. Patients with a new diagnosis of ILD need to establish care with a pulmonologist with experience in this field.

DISTINGUISHING IPF FROM OTHER ILDS

It is critical to distinguish IPF from other causes of interstitial lung disease because the approach to long-term management is fundamentally different. A surgical biopsy with findings of usual interstitial pneumonia (UIP) is not pathognomonic for IPF. Several other causes of ILD, including collagen vascular disease and hypersensitivity pneumonitis, may be associated with a UIP pattern of injury. Many ILDs are inflammatory in nature and respond to long-term immunosuppressive therapy. However, IPF does not respond to immunosuppression or anti-inflammatory medications. Immunosuppressive therapy, particularly the combination of prednisone and azathioprine, leads to worse outcomes in IPF patients. Newer antifibrotic therapies such as pirfenidone and nintedanib slow the rate of lung function decline in some patients with IPF but, to date, no therapy is available that reverses the course of this fatal disease. The role of the newer antifibrotic agents in other ILDs remains unknown.

TRIAGE AND HOSPITAL ADMISSION

■ CRITERIA FOR HOSPITAL ADMISSION

Patients with known ILD who experience significant respiratory decline frequently benefit from hospitalization for treatment of infection, cardiac comorbidity or intensification of immunosuppression. Since the etiology of worsening dyspnea is often uncertain at admission, a thoughtful evaluation is required in all ILD patients. Patients with ILD typically have exercise induced hypoxemia. However, new resting hypoxemia or worsening exercise desaturation requires evaluation. It is critical to assess patients both at rest and with exertion. Frequently, minimal exertion such as talking, standing or walking, will reveal desaturation.

- Any patient with new resting hypoxemia merits hospital admission. Patients with worsening exercise desaturation may require inpatient evaluation. Clinicians should assess patients both at rest and with exertion.

It is challenging to recognize a new diagnosis of ILD in a patient admitted for progressive dyspnea or hypoxemia. This diagnosis requires a high level of suspicion on the part of the treating physician as symptoms and signs may be nonspecific. Any patient with a new diagnosis of ILD should undergo a standardized evaluation for possible etiology (Figure 233-2).

■ CRITERIA FOR ICU ADMISSION

The admission of patients with ILD to the ICU continues to be a controversial decision. For some patients, ICU care including intubation is both appropriate and lifesaving. This group includes patients with pulmonary manifestations of autoimmune disease. The role of intubation is less certain in IPF patients with an acute exacerbation. An acute exacerbation of IPF is defined as a decline in lung function in the absence of an inciting event such as infection (Table 233-2). The survival rate following intubation in this setting is extremely poor. Patients and their families should be appropriately advised regarding the potential for ventilator dependence. Ultimately, the benefit of ICU care is dependent on the precipitating event and the potential to reverse this event without significant long-lasting lung injury.

MANAGEMENT

■ GENERAL APPROACH

The appropriate management of patients admitted to the hospital for pulmonary decompensation from ILD depends on accurate identification of the precipitating event. In general, ILD decompensation may be divided into pulmonary and nonpulmonary categories. Pulmonary etiologies include worsening of underlying ILD (ie, acute exacerbations of IPF), infections, pneumothorax, and thromboembolic disease. Nonpulmonary etiologies of ILD decompensation include cardiac dysfunction (ie, pulmonary hypertension, right heart failure, left heart failure, or ischemia) and neuromuscular dysfunction. All patients with pulmonary decompensation should undergo chest imaging. Chest CT imaging can identify significant infections and is more sensitive to changes in underlying lung disease including the appearance of new ground glass opacities. If there is a suspicion for thromboembolic disease, chest CT with IV contrast provides a simple method for evaluation. However, CT imaging is often not adequate for differentiating infection versus inflammation. Empiric treatment of possible infection should be considered since ILD patients typically have limited pulmonary reserve. Cultures including sputum and blood should be obtained if the patient is febrile

TABLE 233-2 Diagnosis of Acute Exacerbation of Idiopathic Pulmonary Fibrosis (IPF)

Diagnostic criteria:
Previous or concurrent diagnosis of idiopathic pulmonary fibrosis*
Unexplained worsening or development of dyspnea within 30 d
High-resolution computed tomography with new bilateral ground-glass abnormality and/or consolidation superimposed on a background reticular or honeycomb pattern consistent with usual interstitial pneumonia pattern **
No evidence of pulmonary infection by endotracheal aspirate or bronchoalveolar lavage***
Exclusion of alternative causes, including the following:
• Left heart failure
• Pulmonary embolism
• Identifiable cause of acute lung injury§

Patients with idiopathic clinical worsening who fail to meet all five criteria due to missing data should be termed "suspected acute exacerbations."
*If the diagnosis of idiopathic pulmonary fibrosis is not previously established according to American Thoracic Society/European Respiratory Society consensus criteria, this criterion can be met by the presence of radiologic and/or histopathologic changes consistent with usual interstitial pneumonia pattern on the current evaluation.
**If no previous high-resolution computed tomography is available, the qualifier "new" can be dropped.
***Evaluation of samples should include studies for routine bacterial organisms, opportunistic pathogens, and common viral pathogens.
§Causes of acute lung injury include sepsis, aspiration, trauma, reperfusion pulmonary edema, pulmonary contusion, fat embolization, inhalational injury, cardiopulmonary bypass, drug toxicity, acute pancreatitis, transfusion of blood products, and stem cell transplantation.
(Reproduced with permission from Collard HR, Moore BB, Flaherty KR, et al. Acute exacerbations of idiopathic pulmonary fibrosis. Am J Respir Crit Care Med. 2007;176(7):636-643.)

or exhibits cough productive of sputum. Bronchoscopy or induced sputum should be considered for patients with dry cough. Opportunistic infection (eg, Pneumocystis jiroveci [PCP]) should be considered and ruled out if the patient is immunosuppressed.

Cardiac disease is increasingly recognized as a major comorbidity in ILD. Both pulmonary hypertension and acute coronary syndrome (ACS) should be considered in the hospitalized patient with ILD. Echocardiography may show transiently elevated right heart pressures; right heart catheterization is generally advisable prior to starting therapy for PH in patients with ILD. Other common comorbidities in ILD patients include depression and anxiety, sleep disordered breathing, gastroesophageal reflux disease and diabetes mellitus. These disorders impact the presentation of a pulmonary decompensation and often require specific therapy.

- In ILD decompensation, echocardiogram may show transient moderately elevated right heart pressures. However, findings of severe pulmonary hypertension, right ventricular (RV) enlargement, or decreased RV function should prompt further evaluation for alternate etiologies of the PH including exclusion of thromboembolic disease.

TREATMENT AND MONITORING

Inpatient treatment of ILD patients must be tailored to the etiology of decompensation. The treatment of acute exacerbations of IPF involves pulse corticosteroids (1 g IV methylprednisolone daily for three doses). Prednisone is then administered at 1 mg/kg oral daily dose for 2 to 4 weeks. In the setting of high-dose steroids, prophylaxis for PCP should be started. The newer antifibrotic agents (pirfenidone and nintedanib) have not been studied in the setting of IPF exacerbations and should not be initiated in the hospital without the consultation of a pulmonologist familiar with their use.

The most significant complication associated with pulmonary decompensation is a lasting decline in lung function. The occurrence of pulmonary exacerbations is clearly associated with poorer prognosis. Oxygen requirements may require frequent reassessment.

DISCHARGE PLANNING

Prior to discharge, patients should be evaluated for both resting and exercise oxygen requirements to maintain an oxygen saturation greater than 88%. Nocturnal oxygen use is indicated for patients with nocturnal desaturation. A common problem encountered in discharge planning is high supplemental oxygen requirements. In general, patients are able to receive up to 6 L/min by nasal cannula. Beyond this oxygen level, patients may require face-mask oxygen at home. If the goals at discharge are for rehabilitation in a recovering patient, plans to reassess oxygen requirement and decrease oxygen use should be made at discharge. Patients may also use home pulse oximetry monitoring to allow more autonomy in titrating oxygen. Patients and their families should be aware that maintaining oxygen saturation greater than or equal to 88% is the goal. Oxygen may be reduced or taken off at rest if this criterion is met. Conversely, oxygen should be increased with activity to maintain this goal. For secondary prevention, patients should receive the pneumonia vaccine (if not previously vaccinated or boosted) and influenza vaccine if appropriate to the season. Follow-up with a pulmonary specialist is indicated for all patients with ILD.

QUALITY IMPROVEMENT: A MULTIDISCIPLINARY APPROACH TO ILD

Care of patients with ILD is best accomplished by a multidisciplinary approach. While definitive therapies for some forms of ILD are not available, there is increasing evidence that patients with ILD benefit from formalized pulmonary rehabilitation. Like vaccination, this simple intervention may improve patient survival and quality of life.

In addition to active interventions aimed at prolonging life, attention to quality of life, particularly in the end stage of ILD, is critical. Understanding and transmitting prognosis accurately and sensitively is a key component of patient care. Patients newly diagnosed with ILD are frequently unaware of the often poor prognosis, particularly with IPF. Reassuring patients and their families regarding the ability to keep patients comfortable at the end of life is critical. Involve a pulmonary specialist to help patients understand available therapeutic options. While discussions of end-of-life decisions are often more appropriate in the outpatient setting, the frequency of acute exacerbations means that these discussions are also needed in the acute inpatient setting. Involvement of hospice care may be helpful particularly for patients with acute exacerbations of IPF for whom improvement in function is unlikely.

SUGGESTED READINGS

An Official ATS/ERS/JRS/ALAT Statement: Idiopathic Pulmonary Fibrosis: Evidence-based Guidelines for Diagnosis and Management. Am J Respir Crit Care Med. 2011;183:788-824.

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