Approach to the Diagnosis of Interstitial Lung Disease

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KEYWORDS

• Interstitial lung disease • Idiopathic interstitial pneumonia
• Diagnosis

Interstitial lung diseases (ILDs) are a heterogeneous group of more than 150 disease entities that differ significantly with respect to prevention, therapy, and prognosis. The current classification scheme of ILDs is shown in **Fig. 1**.[1]

The diagnostic strategy in a patient with ILD is based on considerations regarding the dynamic time course (acute, subacute, chronic), the cause (known or unknown), and the context of the disease at presentation (presence of extrapulmonary/systemic disease manifestations). **Fig. 2** summarizes the main disease categories that have to be differentiated during the diagnostic process.[1–3]

Once an interstitial disease process has been recognized in a patient, there are 3 crucial questions that have to be addressed in the diagnostic workup:

1. Is there a discernible cause for the disease?
2. If no cause is identifiable, is it idiopathic pulmonary fibrosis (IPF)?
3. If there is no cause of the disease and if it is not IPF, should surgical lung biopsy be recommended?

After a diagnosis has been established, the severity and dynamics of the disease have to be assessed and monitored, with or without therapy. Diagnosis and disease severity/dynamics are fundamental for treatment decisions and to predict prognosis. The diagnostic approach to ILD may have to be adapted to different clinical scenarios that eventually lead to presentation of a patient:

1. Patient presents with clinical symptoms (eg, dry cough, dyspnea).
2. Patient is at risk of ILD due to known exposures (eg, amiodarone, asbestos).
3. Patient is at risk of ILD due to family history.
4. Patient is asymptomatic but presents with chance finding on chest radiography or computed tomography.
5. Patient is asymptomatic but presents with chance finding on pulmonary functioning test (eg, restrictive pattern, reduced gas transfer).

This article deals with diagnostic approaches suitable for patients presenting with clinical symptoms of ILD in the first place.

CLINICAL EVALUATION

History Taking

A comprehensive patient history taking is of crucial importance for the diagnosis of ILD. There are 4 main questions to be answered: (1) when did respiratory symptoms start, (2) how did the disease develop over time to the present, (3) are there or have there been any exposures to

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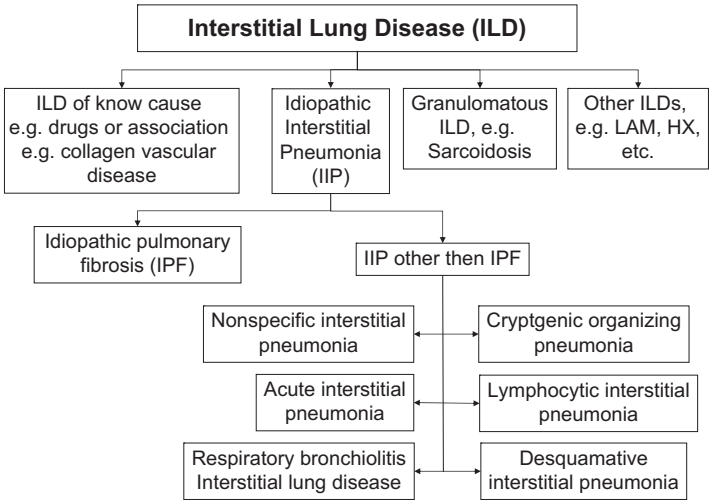

Fig. 1. Classification of ILDs. HX, histiocytosis X; LAM, lymphangiomyomatosis. (*Adopted from* American Thoracic Society/European Respiratory Society international multidisciplinary consensus classification of the idiopathic interstitial pneumonias. Am J Respir Crit Care Med 2002;165:279; with permission.)

etiologic agents known to cause ILD, and (4) what is the severity of symptoms at presentation.[1]

The disease chronology can be subdivided into 4 categories: (1) acute, days up to a few weeks; (2) subacute, 4 to 12 weeks; (3) chronic, longer than 12 weeks; and (4) episodic, ie, symptomatic phases that are followed by asymptomatic phases. In addition, all available radiographs of the lung should be reviewed to characterize the nature and development of the radiologic pattern. Flitting opacities on chest imaging studies may drive the differential diagnosis to focus on eosinophilic pneumonia,

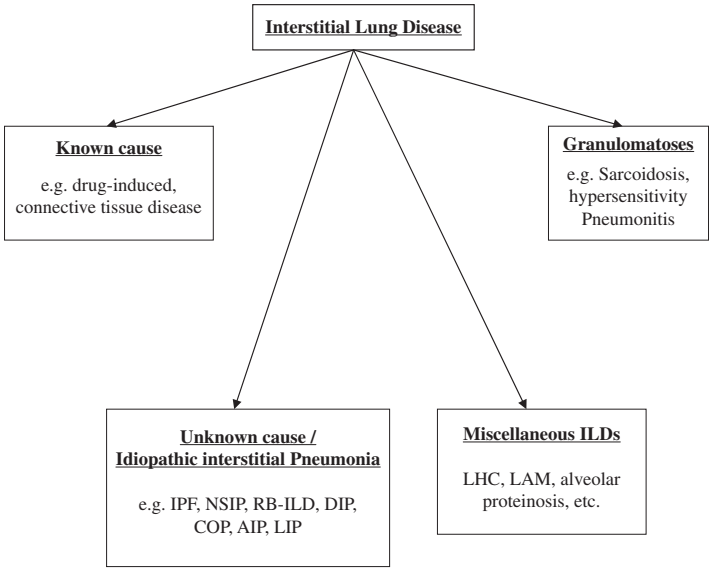

Fig. 2. Overview of different ILD disease categories. AIP, acute interstitial pneumonia; COP, cryptogenic organizing pneumonia; DIP, desquamative interstitial pneumonia; IPF, idiopathic pulmonary fibrosis; LAM, lymphangioleiomyomatosis; LCH, Langerhans cell histiocytosis; LIP, lymphoid interstitial pneumonia; NSIP, nonspecific interstitial pneumonia; RB-ILD, respiratory bronchiolitis and interstitial lung disease. (*Data from* American Thoracic Society/European Respiratory Society international multidisciplinary consensus classification of the idiopathic interstitial pneumonias. Am J Respir Crit Care Med 2002;165:279; with permission.)