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REVIEW ARTICLE

ADULT INTERSTITIAL LUNG DISEASES: AN APPROACH TO DIAGNOSIS AND MANAGEMENT

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I. INTRODUCTION:

Adult interstitial lung diseases (ILD), also called diffuse parenchymal (infiltrative) lung diseases (DPLDs), are a heterogeneous group of disorders that affects not only the interstitial compartment, but also alveolar airspaces, blood vessels and distal airway. ILD covers an immense diversity of lung pathology which includes diseases caused by occupational and environmental exposure, drugs, poisons, physical agents, collagen vascular disorders, infectious agents, diffuse malignancy, idiopathic interstitial pneumonias and various other diseases. Although more than 200 diseases can result in interstitial involvement, the resulting clinical, physiologic and radiographic manifestations are often similar.

The diversity of this disease group makes it difficult to provide representative estimates of the size of disease burden it presents, but the approximate incidence of ILD has been estimated at 31.5 per 100,000 men and 26.1 per 100,000 women. Thus, ILD are less common than the major lung disorders, but collectively they amount to a significant disease burden for society and an appreciable work load for the pulmonary physicians.

Children's interstitial lung disease (chILD) are rare than ILDs that affect adults. Idiopathic pulmonary fibrosis (IPF) is an idiopathic interstitial pneumonia (IIP) and it is the most prominent adult ILD, mostly occurs after the fifth decade of life; this entity is not found in children. Unlike in adults, most ILDs in children are found to have an underlying cause. In addition, the clinical significance of the histologic classification differs significantly between children and adults. For example, usual interstitial pneumonitis (UIP), the histological pattern associated with IPF in adults, is rarely described in children. Desquamative interstitial pneumonitis (DIP) another known type of IIP, which is associated with steroid responsiveness and a better prognosis in adults, has a very poor prognosis in children, particularly in infants. Thus management of ILD in children differs from that in adults.

The initial evaluation of patients with ILD is aimed at identifying the etiology of the ILD and its severity. The results of laboratory, radiographic and pulmonary function tests guide the decisions about whether to pursue bronchoalveolar lavage and/or transbronchoscopic, thoracoscopic or open lung biopsy.

This article reviews the latest data and proposes an approach for the initial evaluation of adult ILD.

II. ETIOLOGY:

The causes of ILD are extremely extensive. In general the pathogenesis is thought to center around injury to the lung followed by attempts to heal the injury. Whether, this injury represents an ongoing event, a series of multiple events, or an abnormal response to an event that is no longer present remains unclear. Unfortunately, the etiology often remains elusive, with the most common causes of ILD are related to occupational and environmental exposure, sarcoidosis, idiopathic pulmonary fibrosis (IPF) and pulmonary fibrosis associated collagen vascular disease. The Potential Causes /Categories of Interstitial Lung Disease are presented in Table 1.

III. CLASSIFICATION:

ILD, also termed diffuse parenchymal lung diseases (DPLDs), are a group of disorders that involve infiltration of alveolar airspaces or thickening of pulmonary interstitial structures. More precise definition is difficult, partly because of the disparity of individual disorders involved and party because alveolar and interstitial pathology is a recognized component of many lung diseases which are appropriately classified elsewhere.

A new classification released in 2002 by Thoracic Society (ATS) and European Respiratory Society (ERS) algorithm for DPLD or ILD has now been accepted internationally (Fig. 1).

Table 1. Potential Causes /Categories of Interstitial Lung Disease.

Cause	Categories
Occupational or other inhaled organic agents	Bird fancier's lung Farmer's lung Bagassosis Byssinosis Malt worker's lung Coffee worker's lung
Occupational or other inhaled inorganic agents	Silicosis Asbestosis Coal worker's pneumoconiosis Talc pneumoconiosis Berylliosis Hard metal fibrosis
Collagen vascular disease related	Systemic lupus erythematosus Rheumatoid arthritis Scleroderma Ankylosing spondylitis Sjogrens syndrome Bechet`s disease Dermatopolymyositis
	Mixed connective tissue disease
Drug related	Chemotherapeutics (Bleomycin, Methotrexate, Busulfan) Drug induced Lupus (Phyenytoin, procainamide) Antiarrhythmics (Amiodarone) Antibiotics (Nitrofurantoin, sulfasalazine) Gold
Physical agents & toxins	Radiation / Radiotherapy Paraquat toxicity Oxygen
Primary disease diagnosis	Sarcoidosis Amyloidosis Tuberous sclerosis Neurofibromatosis Niemann-Pick disease
	Lymphangioleiomyomatosis Pulmonary Langerhans cell histiocytosis
Neoplastic diseases	Bronchoalveolar carcinoma Lymphangitis carcinomatosis
Vasculitides	Churg -Strauss syndrome Wegener`s granulomatosis
Alveolar filling diseases	Alveolar protienosis Lipoid pneumonia Pulmonary lymphoma Chronic aspiration
	Eosinophilic pneumonia
Disorders of circulation	Pulmonary edema Pulmonary veno-occlusive disease
Infection	Tuberculosis Residue of active infection of any type
Idiopathic interstitial pneumonias	IPF (UIP) NSIP COP AIP LIP RB-ILD DIP

AIP = acute interstitial pneumonia; COP= cryptogenic organizing pneumonia; DIP = desquamative interstitial pneumonia: IIP = idiopathic interstitial pneumonia; IPF = idiopathic pulmonary fibrosis; LIP = lymphocytic interstitial pneumonia; NSIP = nonspecific interstitial pneumonia; RBILD = respiratory bronchiolitis interstitial lung disease: UIP = usual interstitial pneumonia.

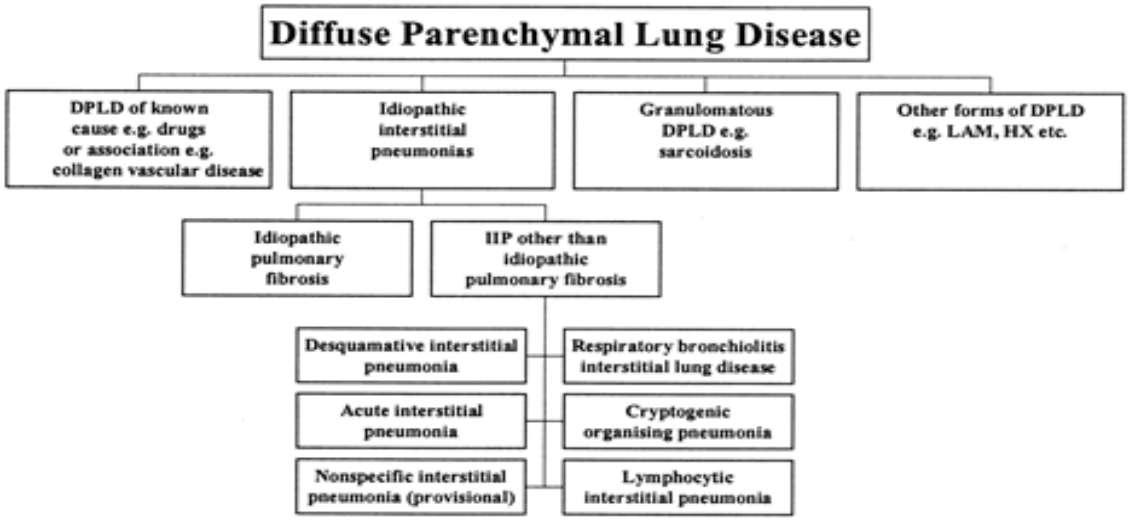

Fig. 1. American Thoracic Society (ATS) and European Respiratory Society (ERS) algorithm for DPLD.

Diffuse parenchymal lung diseases (DPLDs) consist of disorders of known causes (collagen vascular disease, environmental, or drug-related) as well as disorders of unknown cause. The latter include idiopathic interstitial pneumonias (IIPs), granulomatous lung disorders (e.g., sarcoidosis), and other forms of interstitial lung disease (ILD) including lymphangioleiomyomatosis (LAM), pulmonary Langerhans cell histiocytosis/histiocytosis X (HX), and eosinophilic pneumonia. The most important distinction among the IIPs is that between idiopathic pulmonary fibrosis and the other interstitial pneumonias, which include nonspecific interstitial pneumonia (NSIP) (a provisional term), desquamative interstitial pneumonia (DIP), respiratory bronchiolitis-associated interstitial lung disease (RB-ILD), acute interstitial pneumonia (AIP), cryptogenic organizing pneumonia (COP) and lymphocytic interstitial pneumonia (LIP).

IV. SUSPICIOUS OF ILD:

Patients with adult ILD typically present with progressive exertional dyspnea and/or persistent nonproductive cough. Unfortunately, as symptoms are often subtle, nonspecific, and slowly progressive, it is common for patients to realize the true duration of symptoms only in retrospect. Furthermore, some patients are without symptoms and are identified through abnormal findings on radiographs or pulmonary function studies. As such, the physician needs to maintain a suspicion of ILD to facilitate appropriate and early diagnostic testing.

Maintaining a suspicion for ILD is perhaps even more critical for patients at risk of developing ILD from environmental exposures (inhaled allergens, dusts, drugs) or concomitant systemic illnesses (such as collagen vascular disease) Table 2.

Table 2. How to suspect ILD.

Symptoms:
• Progressive exertional dyspnea
• Persistent nonproductive cough
• History of environmental exposures or concomitant systemic illnesses
Abnormal investigations:
• Chest x ray
• Lung functions abnormality

V. DIAGNOSTIC APPROACH:

The ATS and ERS recommend an integrated clinical, radiologic, and pathologic approach to the diagnosis of DPLDs. This suggested integrated approach includes a comprehensive assessment of clinical history, physical examination findings, selected laboratory studies, imaging studies and in selected patients transbronchial or surgical lung biopsy (SLB) (Fig. 2).

1. CLINICAL HISTORY:

- ***General Characteristics***

A careful history is required in the initial evaluation of patients with suspected ILD. The presence of a collagen vascular illness should heighten the suspicion of ILD, as many collagen vascular diseases are associated with pulmonary parenchymal involvement.

Important features include onset, sex, age, smoking history, drug exposures, and assessment of occupational & environmental conditions.

Onset: The onset of symptom development can provide important clues. In vast majority of ILDs the symptoms and signs are chronic i.e. months or years as in IIPs, sarcoidosis and pulmonary histiocytosis X. In some however, symptoms may be acute (days to weeks) or subacute (weeks to months) as in extrinsic allergic alveolitis (EAA), drug induced, AIP and acute eosinophilic pneumonia.

Sex: Sex may affect predisposition to certain types of ILDs or influence the clinical course of others. For example, lymphangioleiomyomatosis and pulmonary involvement in tuberous sclerosis are predominantly seen in premenopausal female patients and women with IPF have an improved prognosis compared with men.

Age: Age is helpful as some ILDs are more common in younger individuals (sarcoidosis, familial idiopathic pulmonary fibrosis and Gaucher's disease), while IPF are more common in older patients.

Occupational history: A strict chronological listing of the patient's lifelong employment must be sought, including specific duties and known exposure to avian, animal, and fish proteins, as well as fungal spores, asbestos, silica, cobalt, beryllium, aluminum, isocyanates, and copper sulfate. The degree of exposure, duration, latency of exposure and use of protective devices need to be inquired about.

Environmental exposure: Review of the environment (home & work) is valuable. It is important to determine if the patient has had exposure to pets (especially birds), air conditioners, humidifiers & evaporating cooling systems. In EAA respiratory symptoms and abnormal chest x-ray are often temporarily related to workplace or hobby. Symptoms may diminish or disappear after the patient leaves the exposure and reappear on returning back to the exposure.

Smoking History: Smoking can alter both the development of ILD and the course of disease. In fact, several ILDs are seen almost exclusively in smokers, including RB-ILD, DIP, and eosinophilic granuloma. Interestingly, cigarette smoking has also been implicated in the development of IPF and can influence disease course. In contrast, EAA is less common in smokers.

Current/Previous medications: A detailed account of current and previous medication use should be obtained because numerous prescription medications have been associated with the development of ILD. In addition to over-the-counter medications, the history should also probe for recreational drug use such as cocaine.

Family History: The family history may be occasionally helpful since familial association have been identified with some cases of IPF, sarcoidosis, tuberous sclerosis, neurofibromatosis and niemann-Pick disease. As such, a careful family history may be helpful in narrowing the differential diagnosis and may identify other family members with an earlier stage of disease.

- ***Symptoms:***

Although patients with ILD typically present with progressive exertional dyspnoea and/or persistent nonproductive cough, other unusual chest symptoms may provide clues to the etiology of an ILD. For example, haemoptysis may be noted in patients with alveolar hemorrhage syndromes, pulmonary vascular diseases, lymphangioleiomyomatosis, tuberous sclerosis, and chronic mitral valve disease. Pleuritic chest pain may be seen in patients with collagen vascular illness, or a pneumothorax in patients with lymphangioleiomyomatosis, tuberous sclerosis, or eosinophilic granuloma. Wheezing is an uncommon symptom in ILD. It has been described in cases of Lymphangitis carcinomatosis, Churg -Strauss syndrome, chronic eosinophilic pneumonia and EAA.

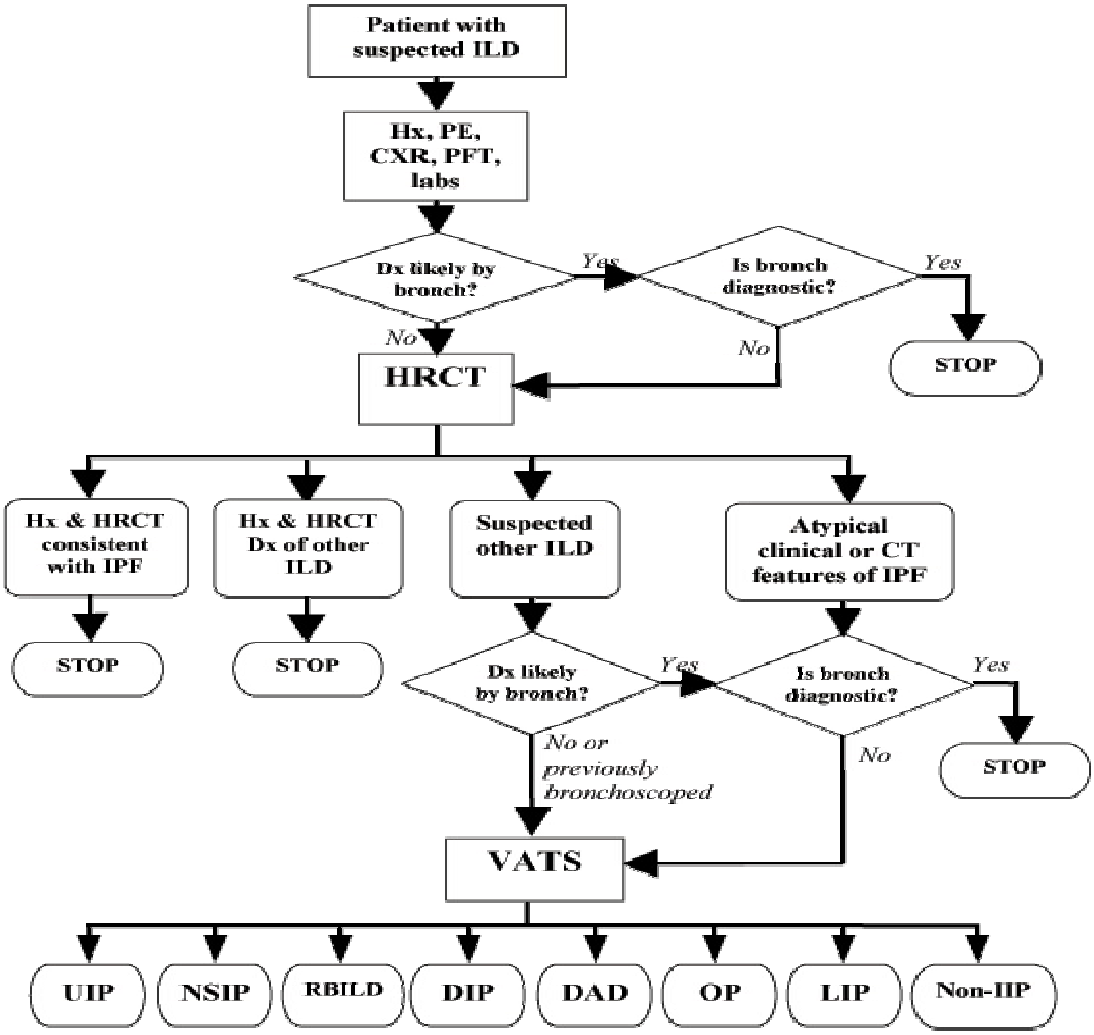

Fig 2. Algorithm outlining an approach for evaluating a patient with suspected interstitial lung disease.

Hx = history; PE = physical examination; PFT = pulmonary function tests; HRCT= high resolution CT scan chest; bronch = bronchoscopy; Dx = diagnosis; VATS = video-assisted thoracoscopy;; ILD= interstitial lung diseases; UIP = usual interstitial pneumonia; NSIP = nonspecific interstitial pneumonia; RBILD = respiratory bronchiolitis interstitial lung disease; DIP = desquamative interstitial pneumonia; DAD = diffuse alveolar damage; OP = organizing pneumonia; LIP = lymphocytic interstitial pneumonia; IIP = idiopathic interstitial pneumonia.

2. CLINICAL EXAMINATION:

The physical examination findings for ILD are generally nonspecific.

- ***Pulmonary findings:***

The characteristic finding is dry bibasilar crackles, although inspiratory high-pitched rhonchi ("squeaks") can be seen with bronchiolar disorders. Clubbing (most common in IPF) and signs of right heart failure can also be seen with advanced disease. Cyanosis is uncommon and is usually a late manifestation indicative of advanced disease.

- ***Extrapulmonary findings:***

Careful attention should be given to extrapulmonary manifestations of disease. e.g. Skin lesions: Erythema nodosum ⇒ Sarcoidosis, tuberculosis, Butterfly rash ⇒ Systemic lupus erythematosus, Telangiectasia ⇒

Scleroderma, Eye changes: Uveitis⇒ Sarcoidosis, Bechet`s syndrome & Keratoconjunctivitis sicca ⇒ LIP, Salivary glands: ⇒ Sarcoidosis, LIP & Musculoskeletal: ⇒ Dermatopolymyositis.

3. *LABORATORY TESTS:*

A minimum panel of initial laboratory investigations includes a complete blood count with differential, erythrocytes sedimentation rate, electrolytes, renal function studies, liver function studies, antinuclear antibodies, antineutrophil cytoplasmic antibody, rheumatoid factor, and a urinalysis.

Routine laboratory tests are often not helpful because findings are nonspecific. Anemia or elevated erythrocytes sedimentation rate are commonly observed. Antinuclear antibodies & rheumatoid factor are clear markers of collagen vascular disease, but are also often positive in IPF. Antineutrophil cytoplasmic antibody helps to confirm Wegener's granulomatosis, but is also found in other vasculitides. Eosinophilia is a requirement for the diagnosis of eosinophilic pneumonia, although it may be encountered in other causes of DPLD. Presence of precipitating antibodies to an offending antigen may be helpful in confirming exposure when EAA is suspected, although they too are non-diagnostic.

4. *RADIOLOGY:*

Radiographic studies are usually abnormal in patients with ILD, although chest radiographs (CXRs) can be normal in approximately 10% of patients.

- ***CXR:*** To identify the radiological pattern of disease, its anatomical distribution (upper or lower zone) & presence of honeycombing, hilar adenoapthy or pleural affection is helpful in narrowing the differential diagnosis. The most common abnormality is reticular or nodular pattern e.g. IPF, sarcoidosis, asbestosis & miliary tuberculosis. However, alveolar filling pattern is not unusual e.g. alveolar cell carcinoma, alveolar proteinosis & pulmonary edema. Upper zones of lung are affected in silicosis, EAA & sarcoidosis. While IPF, asbestosis, pulmonary fibrosis associating collagen vascular disorders tend to involve the lower lung zones. Hilar adenoapthy is a characteristic of sarcoidosis, lymphoma & other malignant disease. Pleural effusions are seen collagen vascular disease, malignancy, infection & asbestosis, in which pleural thickening or plaques may also occur.

- ***High resolution CT scan (HRCT) of Chest:*** HRCT has dramatically altered the diagnostic evaluation of patients with ILD. HRCT is now the imaging modality of choice in anyone with suspected ILD. The technique allows a detailed evaluation of the lung parenchyma by using 1- to 2-mm-thick slices reconstructed with an algorithm that maximizes spatial resolution. Several studies have confirmed that abnormalities can be identified when they are not visible on CXR. HRCT is particularly likely to be diagnostic in patients with IPF (figure 3), lymphangitis carcinomatosis, sarcoidosis, silicosis, subacute HP, and pulmonary alveolar proteinosis. Common characteristic of HRCT appearances are presented in Table 3.

Table 3. Common appearances on HRCT in relation to individual diseases.

Appearance	Diseases
Nodules	Sarciodosis Silicosis Coal worker's pneumoconiosis Miliary Tuberculosis Eosinophilic granuloma
Interlobar septal thickening	Usual interstitial pneumonia Collagen vascular disease Sarcoidosis Asbestosis Lymphangitis carcinomatosis
Alveolar opacification / ground-glass shadowing	Desquamative interstitial pneumonia Cryptogenic organizing pneumonia Extrinsic Allergic Alveolitis Alveolar cell carcinoma Alveolar proteinosis Eosinophilic pneumonia
Cystic change (Honeycomb)	Usual interstitial pneumonia Collagen vascular disease Silicosis Asbestosis Coal worker's pneumoconiosis Lymphangioleiomyomatosis
Pleural involvement	Asbestosis Collagen vascular disease Drug induced (e.g. Amiodarone)

The ATS/ERS statement on the classification of IIPs suggests a confident HRCT diagnosis of UIP (IPF) in the setting of a bilateral basilar/subpleural distribution, more reticular infiltrates, and less ground glass. While a more diffuse distribution, less reticular infiltrates, and more ground glass were thought to have NSIP.

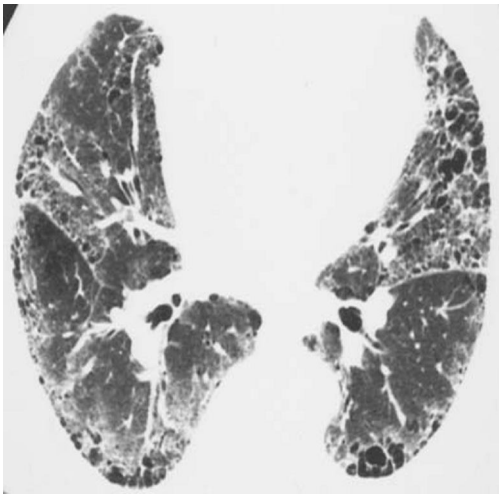

Fig 3. Computed tomography image through the lower lungs in a patient with IPF showing bilateral basilar/subpleural reticular infiltrates with predominantly subpleural honeycombing.

5. PULMONARY FUNCTION TESTING:

All patients with suspected ILD should undergo full lung function evaluation including spirometry, lung volumes, diffusing lung capacity for carbon monoxide (DLCO) and arterial blood gases. Most ILDs share a common pattern of physiologic abnormalities characterized by a restrictive ventilatory defect with reduced lung volumes, DLCO and arterial hypoxemia at rest or on exercise. Some ILDs such as Lymphangioleiomyomatosis, sarcoidosis, EAA can produce an obstructive ventilatory defect. In alveolar hemorrhage, the gas transfer factor is markedly increased. In practice, however, aside from proving objective evidence of the presence of abnormal physiology, lung functions tests do not usually contribute appreciably to specific diagnosis of individual DPLD, but the results of such testing may have important prognostic value.

6. BIOPSY:

A clear diagnosis of many of the ILDs relies on histological evidence. Samples of the lung tissue can be obtained by various methods.

- ***Bronchoalveolar lavage (BAL):*** Bronchoscopic BAL is straightforward to perform, confers little risk, narrow the differential diagnosis and can be diagnostic in cases of infectious causes of ILD, certain occupational exposures to inorganic dusts, malignancy and pulmonary alveolar proteinosis. The value of differential cell count of BAL in the diagnosis of ILD has been extensively explored and found to be lees helpful.

- ***Transbronchial lung biopsy (TBLx):*** Bronchoscopic TBLx can be performed at the same time as BAL and adds only a slight additional risk of bleeding and pneumothorax. It can be very useful in the diagnosis of some ILDs e.g. sarcoidosis, lymphangitis carcinomatosis, eosinophilic pneumonia or infection. The combination of BAL, TBLx and transbronchial needle aspiration from mediastinal lymph node has proven very sensitive for the diagnosis of sarcoidosis. Unfortunately, TBLx is of limited value in the diagnosis of IPF and other subgroups of IIPs owing to the small amount of tissue that is obtained.

- ***Percutaneous needle biopsy:*** is a relatively simple technique done under local anesthesia with fluoroscopic, ultrasound or CT guidance allowing precise sampling from localized areas of abnormality. Again, small amount of tissue is obtained with risk of bleeding & pneumothorax.

- ***Surgical lung biopsy (SLB):*** SLB is required in the diagnosis of most types of ILD. In some cases, either the above procedures have failed or more substantial samples of lung tissue are required or even direct inspection of the lung is necessary to identify areas for sampling, surgical open lung biopsy either through thoracotomy or more recently through video assisted thoracoscopy (VATS) may be carried out. Mortality and morbidity is relatively low with VATS. In some cases, notably IPF, with classic HRCT chest picture and after exclusion of other mimicking diagnosis, SLB is not needed for a diagnosis. Some patients who have severe end stage ILD or significant concomitant illness may be wise not to proceed to SLB.

  The diagnostic findings of SLB in IIPs have become increasingly standardized with recent consensus statements of ATS and ERS. Nevertheless, significant interobserver disagreement remains among expert pathologists.

Summary of Diagnostic Approach

The diagnosis of ILD remains a challenging diagnostic dilemma. (Fig. 2) demonstrates a potential diagnostic algorithm for patients with suspected ILD, reflecting an adaptation of published recommendations. In the initial evaluation, a careful history and physical examination are performed followed by selective laboratory studies, a

chest radiograph, and physiologic studies. If a diagnosis can be reliably achieved by bronchoscopy with BAL and transbronchial biopsy, this may be diagnostic at this stage. If not, or if bronchoscopy is nondiagnostic, HRCT assumes a pivotal role in further diagnostic efforts. A typical history, physical examination, and HRCT picture can assure a diagnosis of UIP/IPF with a high degree of certainty, assuming no alternate etiology. If the clinical features are inconsistent with this diagnosis, or in the setting of atypical HRCT features, SLB should be considered. A histologic diagnosis of UIP in any sample should be considered diagnostic. If no alternate explanation is readily available as an etiology for this diagnosis (e.g. collagen vascular illness or exposure), then a diagnosis of IPF should be considered. A histologic picture of NSIP should lead the clinician to intensify a search for an underlying process, including collagen vascular disease, a drug exposure, or EAA.

VI. TREATMENT:

- ***General measures***

Treatment of the individual ILDs outlined above varies according to disease and although therapies (such as corticosteroids) are used for several diagnoses, others, (such as lung lavage for alveolar proteinosis) are very individual. The specific therapy for individual ILDs is beyond the scope of this review, although a brief summary of typical therapies for the more common ILDs is outlined in Table 4.

The approach to therapy in relation to a specific clinical diagnosis also varies considerably between countries for various reasons that relate to accessibility of diagnostic procedures, differences in philosophy & expectation regarding the need for precise diagnosis or finical influence.

General measures that apply across most forms of ILD includes: early referral to Pulmonology specialist, smoking cessation, pulmonary rehabilitation, supplemental oxygen therapy if needed and treatment of other commonly associated diseases that commonly exists in ILD patients and that are sometimes believed to contribute in worsening of symptoms e.g. gastroesophageal reflux disease, pulmonary hypertension and depression and anxiety.

Patients who have intractable dyspnea may benefit from supplemental oxygen and low dose opioids that reduce the perception of dyspnea. For end-stage disease, lung transplantation remains a restricted, but often effective option.

Table 4. Outline guide to the treatment of ILDs.

Diagnosis	Typical therapy
Pneumoconiosis	Removal from exposure
Extrinsic Allergic Alveolitis	Removal from exposure Corticosteroids
Collagen vascular disease	Specific therapy of disease
Drug reactions	Removal from exposure Corticosteroids
Sarcoidosis	Corticosteroids
Lymphangioleiomyomatosis	Progesterone
Vasculitic disease	Corticosteroids/immunosuppressant
Alveolar proteinosis	Lung lavage
Idiopathic pulmonary fibrosis	Corticosteroids/immunosuppressant
Eosinophilic pneumonia	Corticosteroids

- Specific measures

There is ongoing controversy over the role of specific treatments in subgroups of ILD due to a lack of good quality evidence for many of the specific treatments proposed. At times, no specific treatment may be an appropriate option after discussion between the patient and the specialist.

Idiopathic pulmonary fibrosis:

No therapy has been proven to improve the survival or otherwise significantly alter the clinical course of IPF. There is no evidence to support the widespread use of high dose prednisolone alone, and its use is associated with significant morbidity. A trial of prednisolone (tapering over 3 months from 0.5 mg/kg/day to 10

mg/day maintenance) with azathioprine (2 mg/kg/day) and n-acetyl cysteine (600 mg three times daily) may be warranted, but strong evidence for benefit is lacking. However, enrolment of patients into trials of new therapies is considered to be "best current practice" as this offers sufferers with IPF the chance to receive new agents that may be more effective than current treatments. If the patient is a suitable candidate a referral for consideration of lung transplantation is essential.

In pulmonary fibrotic disorders including IIPs other than IPF:

Anti-inflammatory therapy is broadly appropriate and benefits most patients, but a clear treatment strategy is essential. The art of management is to distinguish accurately between inherently stable fibrotic disease (with treatment not required), progressive predominantly fibrotic disease (with low-dose long-term treatment warranted to retard progression) and the presence of major associated inflammation (justifying initial high-dose treatment).

Sarcoidosis:

Treatment for pulmonary sarcoidosis is only indicated if there is progressive disease on radiology and lung function, or if there are significant symptoms or extrapulmonary disease. Prednisolone (0.5mg/kg/day) is usually first line treatment for 1 month, weaning to a maintenance dose that controls symptoms and then continuing for up to 6–24 months.

Hypersensitivity pneumonitis:

Avoidance of the antigen is essential and prednisolone therapy may be required if there is severe or progressive disease.

In conclusion, ILDs are a group of complex disorders usually presenting with progressive dyspnea, restrictive pulmonary physiology and an abnormal chest radiograph. Because of overlapping signs and symptoms, the evaluation of these patients is often frustrating; the key to understanding and correctly diagnosing ILD is the development and utilization of a disciplined evaluation process (Fig. 2). The use of such a standardized, logical evaluation will yield a diagnosis in the majority of patients with ILD. Referral to a pulmonology specialist and involvement of multidisciplinary teams are the mainstays of management. General management strategies include: pulmonary rehabilitation, smoking cessation, oxygen therapy as required, and treatment of commonly associated diseases. Specific management varies according to the underlying diagnosis.

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