The diffuse parenchymal lung diseases (DPLD), often collectively referred to as the “interstitial lung diseases” (ILD) encompass approximately 150 entities in which the lung is altered by combination of interstitial and/or granulomatous inflammation and usually — fibrosis. Most of these diseases affect not only the interstitium of the lung (structure situated in between the alveoli) but also alveolar space, walls of bronchioles, blood and lymphatic vessels and pleura.

This heterogenous group of disorders is classified together because of similar clinical, radiologic, physiologic and pathologic manifestations. The common features of ILD include relatively subacute to chronic course (usually longer than 6 months), progressive dyspnea on exertion, bilateral interstitial-alveolar infiltrates, restrictive pulmonary dysfunction, diminished diffusing capacity for carbon monoxide, exercise-induced hypoxemia, hypocarbia in the initial stages, and nonspecific histological findings (varying degrees of inflammation, fibrosis and remodeling).

Infections, cardiac failure and malignant diseases are usually not included in this group, although they should be included in the differential diagnosis.

Few data are available on the epidemiology of ILD in general population. In USA task force study in 1972 revealed that diffuse lung diseases constituted 15% of all problems encountered by practicing pulmonary physicians.

In a population-based study in New Mexico in the period 1988–1990 prevalence of ILD was estimated as 81/100 000 males and 67/100 000 females and incidence of ILD was estimated as 32/100 000 per year in males and of 26/100 000 per year in females (table 1).

ILD include disorders of known cause (e.g. drug, radiation therapy, environmental exposures and connective tissue diseases), but in majority — the cause is unknown. Among them sarcoidosis represents granulomatous diseases but other e.g. lymphangioleiomyomatosis or pulmonary Langerhans cell granulomatosis — represent specific forms of interstitial diseases. The most common and important group of ILD is idiopathic interstitial pneumonia (IIP).

Recently ILD are classified broadly into 7 main groups (table 2). It seems likely that these diseases are underdiagnosed in life and misclassified at death.

There are number reasons why the management of diffuse lung disease is inconsistent: 1) however the clinical symptoms, radiologic lesions and functional disturbances are very similar, but individual disease has specific pathogenesis, natural history and likely responsiveness to treatment, 2) many physicians equate all diffuse lung disease as having the same prognosis as most lethal individual disease has specific pathogenesis, natural history and usually — fibrosis. Most of these diseases affect not only the interstitium of the lung (structure situated in between the alveoli) but also alveolar space, walls of bronchioles, blood and lymphatic vessels and pleura.

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Interstitial Lung Disease: Clinical Evaluation and Ways to Diagnosis

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of progression, or stability of the disease. The use of high resolution computed tomography (HRCT) over the past 10 years has revolutionized the approach to ILD. Recently it should be considered as a standard procedure.

The abnormalities seen on chest X-ray and CT-scan in ILD range from a “ground glass” appearance to end-stage honeycombing and include changes described as acinar, reticular, micronodular and reticulonodular. Unfortunately none of these patterns is specific for any particular type of ILD. The distribution of chest-X-ray infiltrates may be of some value in predicting disease etiology: bilateral hilar and paratracheal lymphadenopathy may suggest sarcoidosis or berylliosis, while coexistent pleural plaques or calcifications may indicate asbestos-related pleuropulmonary disease. The pattern of abnormality may be characteristic in several diseases and is virtually pathognomonic in most cases of idiopathic pulmonary fibrosis (table 4).

Table 1 Prevalence and incidence of interstitial lung diseases, (Demetsd)

<table>
<thead>
<tr>
<th>Diagnosis</th>
<th>Prevalence</th>
<th>Incidence Per 105 per year</th>
</tr>
</thead>
<tbody>
<tr>
<td>Male</td>
<td>Female</td>
<td>Male</td>
</tr>
<tr>
<td>Total interstitial lung disease</td>
<td>80,9</td>
<td>67,2</td>
</tr>
<tr>
<td>Idiopathic pulmonary fibrosis</td>
<td>20,2</td>
<td>13,2</td>
</tr>
<tr>
<td>Post inflammatory pulmonary fibrosis</td>
<td>10,1</td>
<td>14,3</td>
</tr>
<tr>
<td>Sarcoidosis</td>
<td>8,3</td>
<td>8,8</td>
</tr>
<tr>
<td>Connective tissue diseases</td>
<td>7,1</td>
<td>11,6</td>
</tr>
<tr>
<td>Drugs and radiation</td>
<td>1,2</td>
<td>2,2</td>
</tr>
<tr>
<td>Occupational/ environmental</td>
<td>20,8</td>
<td>0,6</td>
</tr>
</tbody>
</table>

Table 2 Clinical classification of the interstitial lung diseases

1. Drug-induced lung diseases
2. Environmental and occupational exposures
   a) Pneumoconiosis
   b) Hypersensitivity pneumonitis (HP)
3. Connective tissue diseases (CTD)
4. Primary diseases — unique entities
   a) Sarcoidosis
   b) Langerhans cell granulomatosis (LCG)-histiocytosis X
   c) Lymphangioleiomyomatosis — LAM
   d) Diffuse alveolar hemorrhage (DAH)
   e) Eosinophilic pneumonia — EP
   f) Pulmonary alveolar proteinosis
   g) Alveolar microlithiasis
   h) Amyloidosis
5. Inherited diseases
6. Idiopathic interstitial pneumonias
   a) Idiopathic pulmonary fibrosis-IPF
   b) Nonspecific interstitial pneumoni — NSIP
   c) Acute interstitial pneumonia — AIP
   d) Respiratory bronchiolitis- interstitial lung disease-RB-ILD
   e) Desquamative interstitial pneumonia — DIP
   f) Lymphomatoid interstitial pneumonia — LIP
   g) Cryptogenic organizing pneumonia — COP

Table 3 Systemic signs accompanying ILD

<table>
<thead>
<tr>
<th>Sign/symptom</th>
<th>Disease</th>
</tr>
</thead>
<tbody>
<tr>
<td>Systemic hypertension</td>
<td>CTD, Goodpasture S., Wegener G., MPA</td>
</tr>
<tr>
<td>Erythema nodosum</td>
<td>Sarcoïdosis, Behcet S. (BS), CTD</td>
</tr>
<tr>
<td>Maculopapular rash/cutaneous vasculitis</td>
<td>CTD, WG, drug induced (DI), MPA.</td>
</tr>
<tr>
<td>Teleangiectasia</td>
<td>Scleroderma</td>
</tr>
<tr>
<td>Raynaud’s phenomena</td>
<td>CTD, idiopathic pulmonary fibrosis (IPF)</td>
</tr>
<tr>
<td>Subcutaneous nodules</td>
<td>rheumatoid arthritis, neurofibromatosis, sarcoïd.</td>
</tr>
<tr>
<td>Keratoconjunctivitis sicca</td>
<td>Sjoegren syndrom, CTD.</td>
</tr>
<tr>
<td>Uveitis</td>
<td>Sarc., BS.</td>
</tr>
<tr>
<td>Lymphadenopathy</td>
<td>Sarc., HP, Churg-Strauss S (Ch-SS).</td>
</tr>
<tr>
<td>Pericarditis</td>
<td>CTD, Radiotherapy, Ch-SS</td>
</tr>
<tr>
<td>Hepatosplenomegaly</td>
<td>Sarc., CTD, LIP</td>
</tr>
<tr>
<td>Diabetes insipidus</td>
<td>Sarc., Langerhans cell granulomatosis</td>
</tr>
<tr>
<td>Neurologic abnormalities</td>
<td>Sarc., NF, CTD, WG, Ch-SS</td>
</tr>
<tr>
<td>Arthritis</td>
<td>Sarc., CTD, WG, IPF</td>
</tr>
</tbody>
</table>

Table 4 Diseases, in which HRCT shows characteristic patterns

1. Idiopathic pulmonary fibrosis
2. Langerhans cell granulomatosis
3. Lymphangioleiomyomatosis
4. Sarcoidosis
5. Hypersensitivity pneumonitis
6. Pulmonary proteinosis
7. Microlithiasis
8. Asbestosis

Table 5 Diagnostic value of BAL in ILD

1. Opportunistic infection: Pneumocystis carini, fungi, and tuberculosis
2. Alveolar haemorrhage: hemosiderin-laden macrophages
3. Pulmonary alveolar proteinosis: milky effluent, PAS-positive noncellular substance, foamy macrophages
4. Eosinophilic pneumonias-eosinophilia > 25%
5. Asbestosis: asbestos bodies
6. Lipid aspiration: cytoplasmic lipid vacuoles within alveolar macrophages
7. Microlithiasis: calcic bodies
useful particularly to rule out infections as a cause of diffuse infiltrates. Although it is infrequently diagnostic by itself (table 5), when combined with clinical data and radiographic imaging, BAL cell patterns can be useful in reaching a likely diagnosis without performing lung biopsy.

Generally diagnosis of interstitial lung disease should be confirmed by biopsy if HRCT is not diagnostic and unless the patient’s general condition precludes the procedure — e.g. severe respiratory failure or uncontrolled heart failure. In patients in whom tomographic appearances may be pathognomonic decision to perform lung biopsy depends on assessment of individual cases. A patient over 65 or with poor lung function, especially with hypoxia at rest is at high risk from any operation. It is often argued that treatment will be the same with or without confirmation of the diagnosis by biopsy. It is true if empirical treatment always produce an improvement, but during recurrence of disease the next step in management is more difficult. It is important to perform a biopsy before treatment started. Transbronchial lung biopsy will often confirm diagnosis in several diseases and in other ILD open lung biopsy /or videothoracoscopy is necessary (table 6).

The physician who evaluates ILD has to amass specific knowledge that relates to a large number of potential diagnoses. A clear diagnosis confirmed by biopsy allows clinicians and patients to discuss fully the implications of the disease, to develop a clear plan of treatment and weigh up the advantages and disadvantages of treatment. Morbidity and mortality from open biopsy are low.

**REFERENCES**