'INTERSTITIAL LUNG DISEASE' ASSIGNMENT

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INTRODUCTION

Interstitial lung Disease (ILD) or diffuse parenchymal lung disease (DPLD) is a group of respiratory diseases affecting the interstitium (the tissue) and space around the alveoli (air sacs) of the lungs. It concerns alveolar epithelium, pulmonary capillary endothelium, basement membrane and perivascular and perilymphatic tissues. It may occur when an injury to the lungs triggers an abnormal healing response. Ordinarily, the body generates just the right amount of tissue to repair damage, but in interstitial lung disease the repair process is disrupted, and the tissue around the air sacs (alveoli) becomes scarred and thickened. This makes it more difficult for oxygen to pass into the bloodstream. The disease presents itself with the following symptoms- shortness of breath, non-productive coughing, fatigue, and weight loss, which tend to develop slowly, over several months. The average rate of survival for someone with this disease is between three and five years. The term ILD is used to distinguish these diseases from obstructive airways diseases.

There are specific types in children, known as children's interstitial lung diseases. The acronym chILD is sometimes used for this gp of diseases.

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Prolonged ILD may result in pulmonary fibrosis, but this is not always the case. Idiopathic pulmonary fibrosis is interstitial lung disease for which no obvious cause can be identified (idiopathic) and is associated with typical findings both radiographic (basal and pleural-based fibrosis with honeycombing) and pathologic (temporally and spatially heterogeneous fibrosis, histopathologic honeycombg and fibroblastic foci)

An ILD may be classified as to whether its cause is <u>not known</u> (Idiopathic) or <u>known</u> (secondary)

<u>IDIOPATHIC</u> - ILDs with an unknown cause

Idiopathic interstitial pneumonia is the term given to ILDs with an unknown cause. they represent the majority of cases of interstitial lung diseases. They were subclassified by the American Thoracic Society in 2002 into 7 subgroups -

- Idiopathic pulmonary fibrosis (IPF)
  - most common subgroup
- Desquamative interstitial pneumonia (DIP)
- Acute interstitial pneumonia (AIP)
  - also k/a Hamman-Rich syndrome

- Nonspecific interstitial pneumonia (NSIP)
- Respiratory bronchiolitis - associated interstitial lung disease (RB-ILD)
- Cryptogenic organizing pneumonia (COP)
  - also known by other name bronchiolitis obliterans organizg pneumonia (BOOP)
- Lymphoid interstitial pneumonia (LIP)

SECONDARY - ILDs with a known etiology

A Connective tissue and auto immune diseases
- Sarcoidosis
- Rheumatoid arthritis
- Systemic lupus erythematosus
- Systemic sclerosis
- Polymyositis
- Dermatomyositis
- Antisynthetase syndrome

B Inhaled substances (Pneumoconiosis)
- Inorganic
  - Silicosis
  - Asbestosis
  - Berylliosis
  - Industrial printg chemicals (eg. Carbon black)

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- Organic
- Hypersensitivity pneumonitis (extrinsic allergic alveolitis)

C Drug - Induced
- Antibiotics (eg. Nitrofurantoin)
- Chemotherapeutic drugs
- Anti-arrythmic agents
- Cigarette smoking
  - Smoking - related interstitial fibrosis (SRIF)

D Infection
- Coronavirus disease 2019 (Covid-19)
- Atypical pneumonia
- Pneumocystis pneumonia (PCP)
- Tuberculosis
- Chlamydia trachomatis
- Respiratory syncytial virus

E Malignancy
- lymphangitic carcinomatosis

F Childhood interstitial lung disease and ILD predominately in children
- Diffuse development disorders
- Growth abnormalities & deficient alveolarisation
- Infant conditions of undefined cause
- ILD related to alveolar surfactant region.

DIAGNOSIS

Investigation is tailored towards the symptoms and signs. A proper and detailed history looking for the occupational exposures and for signs of conditions listed above is the first and probably the most imp part of the workup in patients with ILD. Pulmonary function tests usually show a restrictive defect with ↓ed diffusion capacity.

A lung Biopsy is req if the clinical history and imagig are not clearly suggestive of a specific diagnosis or malignancy cannot otherwise be ruled out. In cases where a lung biopsy is indicated, a trans-bronchial biopsy is usually unhelpful, and a surgical lung biopsy is often req.

PULMONARY FUNCTION TESTING

Most patients with suspected ILD are likely to undergo complete pulmonary fxn testg. These tests are useful in diagnosis and determing severity of disease.

Although, there is a large diversity in interstitial lung disease, most follow a restrictive pattern. Restrictive defects are defined by ↓ed TLC (total lung capacity), RV (residual

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volume), FVC (forced vital capacity) and FEV1 (forced expiratory volume in one sec). As both FVC and FEV1 are reduced, the FVC to FEV1 ratio remains normal or is ↑sed.

As disease progression ↑ses and the lungs become stiffer lung volumes will continue to ↓se. Lower TLC, RV, FVC and FEV1 scores are associated with a more severe disease progression and poorer prognosis.

X-ray and CT (radiography and Computed tomography)

Chest radiography is usually the first test to detect interstitial lung diseases, but the chest radiography can be normal in up to 10% of patients, especially early in disease process.

High resolution CT of the chest is the preferred modality and differs from routine CT of chest.

Conventional (regular) CT chest examines 7-10 mm slices obt at 10mm intervals; high resolution CT examines 1-1.5 mm slices at 10 mm intervals usg a high spatial frequency reconstructn algorithm. The HRCT therefore provides approx 10 times more resoluth than regular CT scans, allowg HRCT to elicit details that can't otherwise be visualized.

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Genetic Testing

For some types of paediatric ILDs and few forms adult ILDs, genetic causes have been identified. These may be identified by blood tests. For a limited no. of cases, this is a definite advantage, as a precise mol diagnosis can be done; frequently then there is no need for a lung biopsy.

Testing is available for -

ILDs related to alveolar surfactant region

- → surfactant protein B deficiency (mutations in SFTPB)
- → surfactant protein C deficiency (mutation in SFTPC)
- → ABCA3 deficiency (mutations in ABCA3)
- → Brain lung thyroid syndrome (Mutations in TTF1)
- → Congenital pulmonary alveolar proteinosis (mutations in CSFR2A and/or CSFR2B)

Diffuse development disorder

- → Alveolar capillary dysplasia (mutations in FoxF1)

Idiopathic pulmonary fibrosis

- → Mutations in telomerase reverse transcriptase (TERT)
- → Mutations in telomerase RNA component (TERC)
- → Mutations in the regulator of telomere elongation helicase 1 (RTEL1)
- → Mutations in poly (A)- specific ribonuclease (PARN)

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TREATMENT

ILDs is not a single disease but encompasses many different pathological processes; hence, treatment is different for each disease. If a specific occupational exposure cause is found, the person should avoid that environment. If a drug cause is suspected, the drug should be discontinued.

Many cases due to unknown or connective tissue-based causes are treated with corticosteroids, such as prednisolone. Some people respond to immunosuppresant treatment.

Oxygen therapy
Oxygen therapy at home is recommended in those with significantly low oxygen levels.

Pulmonary rehabilitation
Pulmonary rehabilitation appears to be useful with the benefits being sustainable longer term with improvement in exercise capacity, dyspnoea, and quality of life. Lung transplantation is an option if the ILD progresses despite therapy in appropriately selected patients with no other contraindications

Antibodies And Inhibitors
Food and Drug Administration approved a new drug for treatment of idiopathic

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pulmonary fibrosis (IPF). This drug, Ofev (nintedanib), is marketed by Boehringer Ingelheim Pharmaceuticals. This drug has been shown to slow the decline of lung fxn, although the drug has not been shown to reduce mortality or improve lung fxn. The estimated cost of drug per year is approx $94,000

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References

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