Interstitial Lung Disease

by Dr Liam Chawke. Dr Kate O'Reilly, Mater Misericordiae University Hospital..

Irish Thoracic Society

Common signs and symptoms:

- Cough, exertional dyspnoea (progressive)
- CTD features
  - Digital clubbing /Digital ulcers
  - Gottrens papules / Ulcers sclerodactyly/calcinosis / evidence of joint inflammation /Mechanics hands
  - Heliotrope rash/ shawl rash /
  - Esophageal dysmotility / Sicca symptoms / Malar flush

Investigation checklist - Bloods

Connective tissue disease screen (ENA etc.) / Rheumatoid factor /anti-CCP / Anti U1 RNP / Extended myositis panel / ANCA (MPO and PR3 also if positive)
ESR / CRP / CK
Specific IgG antibodies in Hypersensitivity pneumonitis (if indicated)
VEGF-D (in selected cases)
Antibodies for suspected acute/chronic hypersensitivity pneumonitis
Genetic screen – familial / rare causes of ILD (in selected cases)

Investigation checklist continued

Lung imaging:
- CXR, High resolution CT Thorax

PFTs
- Spirometry & DLCO
- Total lung capacity / Lung volumes
- Sniff testing – if indicated (myositis ILD)

Bronchoalveolar lavage
- Differential WCC (useful for HP / eosinophilic lung disease

Lung biopsy
- VATS lung biopsy (gold standard indicated)
- Cryoprobe assisted transbronchial lung biopsy (no recommendation for/against)
- Transbronchial lung biopsy (useful in stage 2 sarcoidosis / Chronic hypersensitivity pneumonitis)

- BNP / ECG / Echocardiogram – to assess for PH
- Right heart catherisation - for confirmation of PH

Other imaging
- Whole body MRI – in selected cases (? Myositis)

Occupational / environmental exposure checklist

Classification of Interstitial Lung Disease (ATS guidelines 2013)

Idiopathic Interstitial Pneumonia	Rheumatologic	Primary disease related	Iatrogenic / Miscellaneous	Environmental / Inherited
• IPF (UIP) • NSIP • Desquamative Interstitial pneumonia • RB-ILD • Acute interstitial pneumonia • Lymphocytic interstitial pneumonia • Cryptogenic organising pneumonia • **Rare** • Unclassifiable IIP • Idiopathic pleuroparenchymal fibroelastosis	• Rheumatoid Arthritis • Systemic sclerosis • Anti-synthetase syndrome (PM/DM) • Mixed CTD ILD • Sjogren Syndrome • SLE • Ankylosing spondylitis • Vasculitis (i.e. Goodpastures syndrome)	• Sarcoidosis • Pulmonary Langerhans cell histiocytosis • LAM / BHD • Eosinophilic lung disease • Pulmonary Alveolar Proteinosis • Amyloidosis • IBD associated	• Drug induced ILD(www.pneumotox.com) • Radiation pneumonitis / fibrosis • Interstitial pneumonia with autoimmune features • Acute fibrinous and organizing pneumonia	• acute / chronic hypersensitivity pneumonitis • Inorganic dust/fibre-fume related (pneumoconiosis, asbestosis, silicosis • Familial interstitial pneumonia • Hermansky-Pudlak syndrome • Metabolic storage disease

Radiology pathology in ILD	Distribution on CT	Typical radiographic features	Typical pathologic feature
Usual interstitial pneumonia	Peripheral / subpleural . Basiliar / bilateral	Reticular markings / traction bronchiectasis / honeycombing	Fibrosis with honeycombing Fibroblastic foci Subpleural distribution Heterogenous lung involvement Absence of features of suggesting alternative diagnosis
NSIP	Peripheral / subpleural / basiliar / bilateral	GGO / reticular markings / NSIP line / Minimal or no honeycombing	Homogenous interstitial fibrosis and/or inflammation Rare honeycombing
OP	Diffuse / often peripheral and patchy	Patchy GGO and consolidation Sometime nodular	Plugs of connective tissue in small airways Patchy distribution Little of no fibrosis Preservation of lung architecture
DAD	Diffuse	GGO / Airspace consolidation	Hyaline membranes / oedema / diffuse

Management of ILD

Pharmacological	• Pirfenidone or Nintedanib for IPF • Immunosuppressive agents for CTD-ILD (nintedanib in Scleroderma ILD) • Emerging evidence of Pirfenidone and Nintenanib for progressive fibrotic ILD
Pulmonary rehab	Any ILD
Oxygen	Long term and ambulatory therapy as indicated
Lung transplantation	Single / Double lung transplantation as indicated
Palliative Care	For management of refractory dyspnoea / progressive ILD despite treatment / those unsuitable for lung transplantation