Recognizing ILD with Progressive Lung Disease Phenotype

### Step 1: Correctly Diagnose ILD

- **ROS looking for symptoms of connective tissue disease**
  - Raynaud’s phenomena, dry mouth/eyes, joint erythema/edema
- **Medicines**
  - Bleomycin, amiodarone, methotrexate, nitrofurantoin
- **Occupational history**
  - Asbestos exposure, silica exposure
- **Environmental history**
  - Birds, mold, dust

### Testing

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### Make the Diagnosis

- **Connective Tissue-Related ILD**
  - More commonly younger and female
  - May or may not have other signs/symptoms of CTD
  - Raynaud’s skin tightening, telangiectasias in scleroderma
  - Gottron’s papules in myositis
  - Morning stiffness, joint pain in RA
  - HRCT pattern
    - Typically NSIP (scleroderma and myositis) but can be UIP (RA)
    - LIP in Sjogren’s

- **Hypersensitivity Pneumonitis (HP)**
  - Exposure can be overt or covert
  - Has acute, subacute and chronic forms
  - Chronic fibrotic HP can look like IPF
  - HRCT Pattern
    - Reticulation and ground glass
    - Can be upper lung predominate
    - Air trapping on expiratory imaging

- **Idiopathic Pulmonary Fibrosis (IPF)**
  - Age > 50
  - No identifiable causes of ILD (meds, CTD, exposures)
  - UIP on HRCT or lung biopsy
  - UIP on HRCT
    - Basilar and peripheral changes
    - Reticulation
    - Honeycombing with or without traction bronchiectasis
    - Absence of other feature not consistent with UIP

### HRCT

- Thin cut - <= 2 mm
- No contrast
- Prone images
- Expiratory images

### PULMONARY FUNCTION TESTING

- Lung volumes
- Airflow
- Diffusion capacity for CO

### INDICES OF OXYGENATION

- Activity
- Rest
- Sleep

### AUTOIMMUNE SEROLOGIES

- ANA with reflex including SSA, SSB Abs
- Rheumatoid factor and anti-centromere antibody
- Myositis panel, aldolase, CPK.
- Scl-70, centromere

### Step 2: Determine severity and risk for progression/mortality

- Disease risk is determined largely by type of ILD; poorest outcomes are seen with IPF
- There are significant limitations to predicting disease course for individual patients
- Predictive tools for ILD are extrapolated from data in IPF

### Baseline Features

- Older age
- Male sex
- Tobacco use
- Low BMI
- Pulmonary hypertension
- Emphysema
- Honeycombing
- Extent of fibrosis
- Baseline 6MWD
- Baseline FVC
- Baseline DLCO

### Longitudinal Features

- Acute exacerbations
- Worsening symptoms requiring hospitalization
- Changes in FVC
- Changes in DLCO
- Changes in 6MWD

### Treatment

- Specific ILD diagnosis
- Mortality risk
- Co-morbidities
- Disease behavior and prior response to therapy

- IPF: pirfenidone and nintedanib
- Non-IPF: Limited data for non-IPF ILDs. Commonly used therapies include mycophenolate mofetil, cyclophosphamide, rituximab, azathioprine, corticosteroids

- Monitor closely for response to therapy and adjust if needed
- Manage co-morbidities
- Consider transplant referral early
- Consider drug trials
- Maintain normoxia and enroll in pulmonary rehab
- Palliative/hospice care if needed

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