# Recognizing ILD with Progressive Lung Disease Phenotype

## STEP 1 Correctly Diagnose ILD

### DETAILED HISTORY

- **ROS looking for symptoms of connective tissue disease** (e.g. Raynaud's phenomena, dry mouth/eyes, joint erythema/edema)
- **Medicines** (e.g. bleomycin, amiodarone, methotrexate, nitrofurantoin)
- **Occupational history** (e.g. asbestos exposure, silica exposure)
- **Environmental history** (e.g. birds, mold, down)

### DETAILED EXAM

- **Signs of connective tissue disease** (e.g. skin tightening of the fingers, telangiectasias, "mechanics hands", gottron's papules)
- **Looking for evidence of severity** (e.g. right ventricular heave, JVD, extent of inspiratory crackles)

### Testing

**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

### 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 Sjogrens

**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

## 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

## STEP 3 Treatment

### INFLUENCED BY

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

### TREATMENTS

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

### MANAGEMENT

- 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

*This activity is supported by an independent educational grant from Boehringer Ingelheim Pharmaceuticals, Inc.*

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