

DIAGNOSIS AND LONGITUDINAL MANAGEMENT OF SYSTEMIC SCLEROSIS INTERSTITIAL LUNG DISEASE (SSc-ILD)



Pre-existing ILD with suspected CTD

Refer to Rheumatologist

Pre-existing SSc
No ILD or ILD not yet diagnosed



Interdisciplinary Evaluation

Dyspnea with suspected ILD

Refer to Pulmonologist

DIAGNOSTIC EVALUATION

- HRCT
- PFT
- ECHO
- Serologies
- Assessment of Oxygen Needs
- Modified Rodnan Skin Score
- Swallowing/Aspiration Evaluation
- Esophageal Motility/GERD
- Age-appropriate Cancer Screening

ASSESS RISK FOR PROGRESSIVE ILLD PHENOTYPE

Antibody status and clinical factors are most informative related to the risk of progressive ILLD

- Early disease (within 5 years of scleroderma diagnosis)
- Patients with anti Scl 70 or a nucleolar pattern ANA (anti Th/To, U3 RNP or PmScl)
- Any patient with scleroderma is at risk of developing ILLD but it is most common in diffuse cutaneous scleroderma
- Patients with anti-centromere antibodies are at lower risk for progressive ILLD and greater risk for pulmonary hypertension

CONSENSUS CLINICAL SUMMARY DIAGNOSIS = SSc-ILD

Initial Therapeutic Strategies

- ILLD Drug Therapy**
- Cyclophosphamide (CYC)
 - Mycophenolate Mofetil (MMF)
 - Nintedanib
 - Prednisone (low dose) - short term. Use caution in patients at risk for scleroderma renal crisis.
 - Azathioprine and Rituximab are reasonable alternatives to CYC and MMF

- Non-Pharmacologic Therapy**
- Oxygen to maintain normoxia
 - Pulmonary Rehab
 - Vaccination
 - Sleep with HOB elevated
 - Avoid eating within 3 hours of lying down
 - Aggressive reflux management

- Drug Therapy for Non-ILLD manifestations of SSc**
- Consult with appropriate specialist

Surveillance
Q 3-6 month assessment for therapeutic response and longitudinal management

- Spirometry
- DLCO
- 6MWT
- Annual Echo
- Skin Exam
- Vascular Assessment
- HRCT if concerned about progression and/or worsening symptoms
- Consider alternative therapies and organ transplantation for progressive unresponsive disease
- Consider age-appropriate cancer screening
- CT every 1-2 years to monitor malignancy