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

Interdisciplinary Evaluation

Pre-existing ILD with suspected CTD
- HRCT
- PFT
- ECHO

Pre-existing SSc ILD not yet diagnosed
- Obtain PFT’s and HRCT with inspiratory, expiratory and prone imaging (Routine chest CT is inadequate).

Dyspnea with suspected ILD
- Refer to Pulmonologist

Refer to Rheumatologist

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 ILD PHENOTYPE

RISK

Patients with nucleolar pattern ANA, anti-Th/To antibodies or anti-Scl-70 antibodies

Patients with SSc Phenotypes: Limited Cutaneous SSc | Diffuse Cutaneous SSc | Scleroderma Sine Scleroderma | Overlap Syndrome

Patients with anti-centromere or anti-polymerase III antibodies are at lower risk for progressive ILD and greater risk for PHN (ACA) and renal crisis (RNAP)

CONSENSUS CLINICAL SUMMARY

DIAGNOSIS = SSc-ILD

ILD 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

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

surveillance
- Spirometry
- Dlco
- 6MWT
- Annual Echo
- Annual HRCT
- Skin Exam
- Vascular Assessment
- Consider alternative therapies and organ transplantation for progressive unresponsive disease
- Consider age-appropriate cancer screening

Initial Therapeutic Strategies

Respiratory Institute

This activity is supported by an independent educational grant from Boehringer Ingelheim Pharmaceuticals, Inc. © National Jewish Health, 2009.