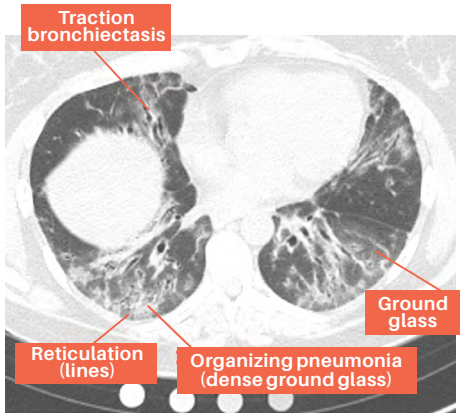
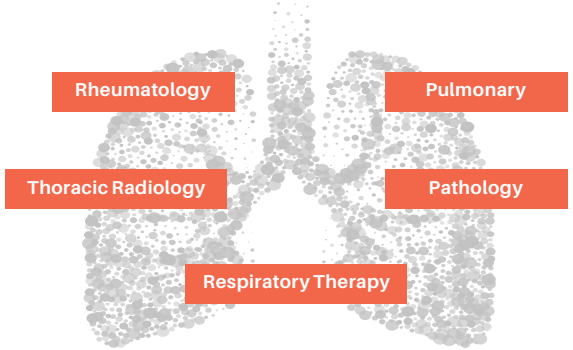



*HRCT images courtesy of Gregory P. Cosgrove, MD

A multidisciplinary discussion (MDD) for patients suspected to have CTD-ILD has many benefits and is crucial to the diagnostic process. MDD can increase diagnostic confidence by refining a provisional diagnosis, enhance interobserver agreement on the diagnosis, and increase diagnostic precision. It is important that CTD patients with evidence of respiratory disease are referred to a pulmonologist. In addition to rheumatology and pulmonary, other areas of specialty that are often involved in MDD for CTD-ILD include thoracic radiology, and pathology. Patients benefit from improved care coordination when MDD is utilized.




Mitto S, Fell CD. Semin Respir Crit Care Med 2014;35:249.

## SUPPORT FOR YOU FROM THE PULMONARY FIBROSIS FOUNDATION

The Pulmonary Fibrosis Foundation mobilizes people and resources to provide access to high quality care and leads research for a cure so people with pulmonary fibrosis will live longer, healthier lives.

To learn more about how the PFF can help support you, contact the PFF Patient Communication Center at **844.TalkPFF** (844.825.5733) or **pcc@pulmonaryfibrosis.org**, or visit the PFF online at **pulmonaryfibrosis.org**.

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# RHEUMATOLOGY POCKET GUIDE

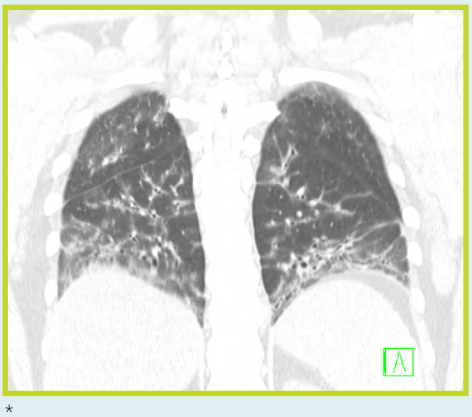
Understanding Connective Tissue Disease-Related Interstitial Lung Disease

## TABLE 1:
## DIAGNOSTIC CLUES THAT SUGGEST ILD IN THE SETTING OF A PREVIOUSLY DIAGNOSED CTD

| Clues | Symptoms | Signs | Physiologic | Radiographic |
|---|---|---|---|---|
| | Early-<br>Fatigue/Dyspnea on exertion<br><br>Late-<br>Dyspnea at rest | Crackles on auscultation<br><br>Digital clubbing<br><br>Oxygen desaturation with exercise or at rest | Restrictive pattern on PFTs<br>- Low FVC<br>- Low TLC<br><br>Diffusion Impairment<br>- Low DLCO | High-resolution chest CT findings consistent with ILD<br><br>Reticulation (peripheral "lines" in the subpleural space)<br><br>Ground glass (hazy) opacities and/or Traction bronchiectasis (dilation of the airways larger than the adjacent blood vessel)<br><br>Honeycomb change (peripheral, subpleural "cysts" in rows) |
| **Tips** | Many patients are asymptomatic in early ILD<br><br>Symptoms may be nonspecific | Certain CTDs have specific risk factors for ILD as described in Table 2. | Early in course of ILD, PFTs may be normal, or there may be an isolated decrease in the DLCO | NSIP is the most common radiographic pattern of ILD in CTD |

## TABLE 2: PREVALENCE AND RISK FACTORS FOR ILD BASED ON THE UNDERLYING CTD

| | Prevalence of ILD | Risk factors on history and physical | Risk factors based on laboratory studies |
|---|---|---|---|
| **Systemic sclerosis** | >65% | Male sex<br>African American race<br>Diffuse cutaneous sclerosis | Scl-70 antibody<br>PM/Scl antibody |
| **Rheumatoid arthritis** | 30-76% | Male sex<br>Older age<br>Cigarette smoking | RF antibody<br>CCP antibody |
| **Sjogren's syndrome** | 25% | Male sex<br>Older age<br>Cigarette smoking | ANA antibody<br>RF antibody |
| **Myositis** | 23-65% | Amyopathic dermatomyositis<br>Older age<br>Arthritis | Jo-1 (PL1) antibody<br>PL7 antibody<br>PL12 antibody<br>MDA5 antibody |
| **Systemic lupus erythematosus** | 3-13% | Not well defined | dsDNA antibody |
| **Mixed connective tissue disease** | 18-66% | Older age<br>Raynaud phenomenon<br>Dysphagia | Anti-Smith antibody |



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Non-specific interstitial pneumonia (NSIP) is the most common HRCT pattern seen in lung disease associated with CTD, although other interstitial patterns, such as usual interstitial pneumonia, are possible. The NSIP pattern is more commonly related to CTD than idiopathic NSIP. Distinctive characteristics seen in NSIP are symmetric, bibasilar, ground glass, reticulation, traction bronchiectasis (dilation and distortion of the bronchi or bronchioles in areas of fibrosis), and subpleural sparing.[1]


[1]References:

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Ahuja J, et al. Radiol Clin N Am 2016; 54: 1015-1031.

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