AMERICAN THORACIC SOCIETY DOCUMENTS

Treatment of Systemic Sclerosis–associated Interstitial Lung Disease: Evidence-based Recommendations

An Official American Thoracic Society Clinical Practice Guideline

Ganesh Raghu, Sydney B. Montesi, Richard M. Silver, Tanzib Hossain, Madalina Macrea, Derrick Herman, Hayley Barnes, Ayodeji Adegunsoye, Arata Azuma, Lorinda Chung, Gregory C. Gardner, Kristin B. Highland, Marie Hudson, Robert J. Kaner, Martin Kolb, Mary Beth Scholand, Virginia Steen, Carey C. Thomson, Elizabeth R. Volkmann, Fredrick M. Wigley, Dee Burlile, Karen A. Kemper, Shandra L. Knight, and Marya Ghazipura; on behalf of the American Thoracic Society Assembly on Clinical Problems

THIS OFFICIAL CLINICAL PRACTICE GUIDELINE OF THE AMERICAN THORACIC SOCIETY WAS APPROVED MAY 2023

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SSc-ILD Definition

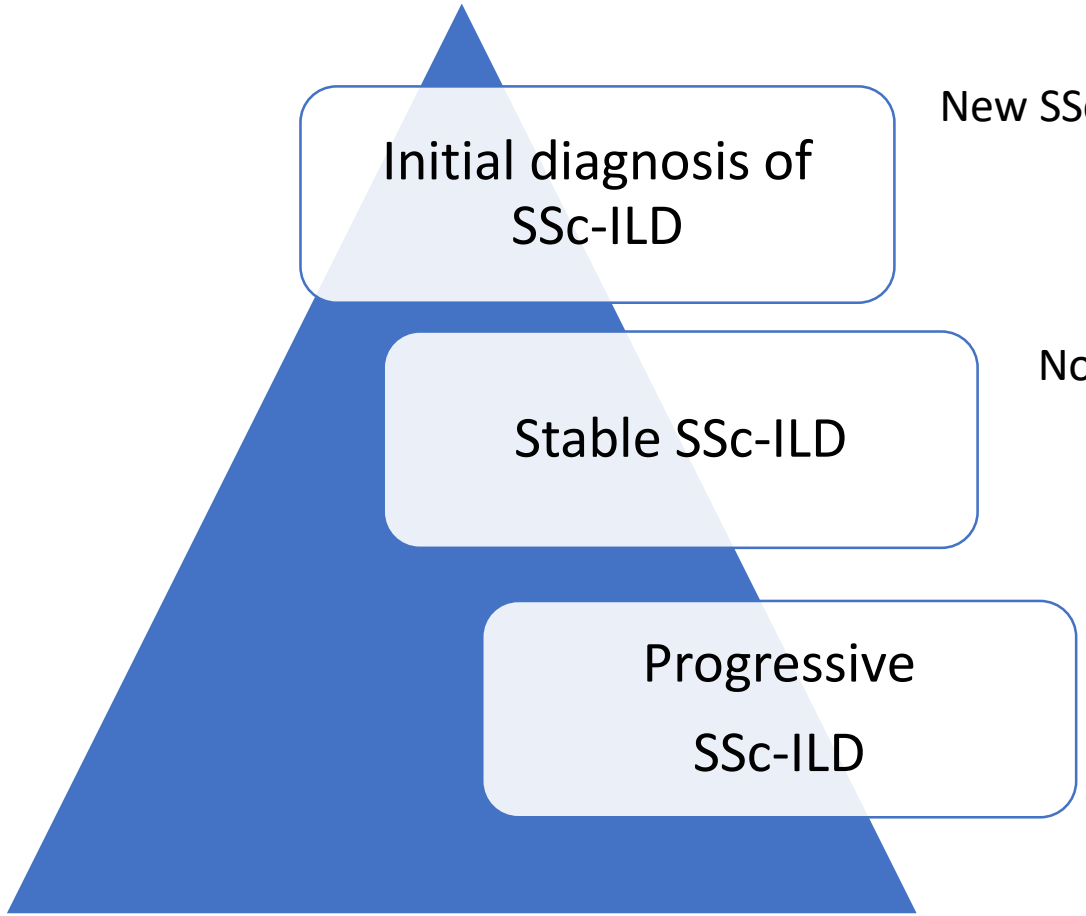
SSc:

- American Rheumatology Association 1980 criteria or
- 2013 ACR-EULAR classification criteria

ILD:

- Radiologic presence of
  - Reticulation, Traction bronchiectasis, traction bronchiolectasis, honeycomb cysts,
  - Ground-glass opacities or air space consolidation,
  - any of the **recognized patterns** of interstitial pneumonias
  - **UIP, probable UIP, indeterminate for UIP, NSIP, OP, LIP**, pleuro-parenchymal fibroelastosis, or unclassifiable interstitial pneumonias reported in the context of SSc.

SSC- 3 subgroups



New SSc-ILD diagnosis before treatment initiation

Not meeting criteria for progressive SSc-ILD

2 of 3 criteria:

1) **Worsening dyspnea or cough**
2) **Physiological evidence of disease progression (>5% absolute decline in FVC or >10% absolute decline in DLCO (adjusted for hemoglobin)**
3) **Radiological evidence of disease progression/ Increase in the extent or severity of ILD features on CT assessed visually**

6 Therapies assessed

- Cyclophosphamide
- Mycophenolate
- Tocilizumab
- Rituximab
- Nintedanib
- Pirfenidone
- Nintedanib plus mycophenolate
- Pirfenidone and mycophenolate

Recommendations