

# 2023 American College of Rheumatology (ACR) Guideline for the Screening, Monitoring, and Treatment of Interstitial Lung Disease in People with Systemic Autoimmune Rheumatic Disease

## Public Comments

*The 2023 American College of Rheumatology (ACR) Guideline for the Screening, Monitoring, and Treatment of Interstitial Lung Disease in People with Systemic Autoimmune Rheumatic Disease call for public comment was posted on the ACR website August 29, 2022. The announcement was emailed to the Practice Guidelines Subcommittee, Quality of Care Committee, and ACR Board of Directors, and was included in multiple ACR publications and on ACR social media platforms. Six (6) responses were received via the online form. The public comment period closed on September 29, 2022.*

### **RESPONSES RECEIVED ONLINE:**

- Name: Richard Zamore
- Institution: Lahey Hospital and Medical Center
- Position: Faculty
- Disclosure (optional): Nothing to disclose

#### Comment:

A few comments on the project plan:

1) While I understand the goal to keep the scope narrow for clinical practice guidelines, the inclusion criteria will leave out individuals who have ILD with autoimmune features, such as a patient with ILD, raynauds, and a positive SCL-70 antibody, but without features of systemic sclerosis. Guidance on these patients would be useful if possible.

2) Some definitions will be helpful. For example, "patients with rheumatic disease at increased risk for developing ILD" (459). Who is included in this? Is this all patients with RA, for example?

3) My understanding is a lot of treatment is based on the pattern of ILD, either NSIP or UIP, and I do not see mention of this in the project plan. In fact, the inclusion criteria mentions "Progressive fibrosing ILD" (1464), whereas NSIP is not necessarily a fibrosing type of ILD, but still falls within the domain of ARD-ILD.

- Name: Vivek Nagaraja
- Institution: Mayo Clinic Arizona (Division of Rheumatology)
- Position: Chair & Senior Associate Consultant
- Disclosure (optional): Nothing to disclose

#### Comment:

1. In the PICO section, do history / physical include information from PRO data? It will be useful to make this distinction and include comparison with PRO data where possible (PRO measures of impact on general health and ILD specific symptomatology).

2. Page 43: With the increasing evidence of data on ANCA vasculitis associated ILD, I hope the committee considers including this disease sub-group for the guideline. It may have a direct impact on more awareness to look for ILD in this patient population.

3. Page 44: I am surprised to see Pulmonary Rehabilitation excluded from interventions. PR has shown to have a meaningful impact on quality of life in patients with ILD and is an important supplemental intervention to pharmacologic interventions. I hope the guideline committee re-considers to include this intervention.

- Name: Grant Schulert
- Institution: Cincinnati Childrens Hospital
- Position: Associate Professor
- Disclosure (optional): Novartis, SOBI - consulting fees

Comment:

I don't see any pediatric trained panel members - while less common this population should be considered in both cSLE/JDM but also systemic JIA and monogenic diseases.

- Name: Belinda Birnbaum
- Institution: Bryn Mawr Medical Specialists Association
- Position: Rheumatologist
- Disclosure (optional): Nothing to disclose

Comment:

This project seems truly ambitious and wonderful. Thanks to the PI's. I am wondering if:

- 1) Any recommendations/definitions will be given to response to treatment, disease stabilization vs improvement
- 2) Any timeline will be given to how often to do testing, and when to assess response and need to change therapy
- 3) Many of the PI's involved appear to be at tertiary care centers. I would love to see involvement of community rheumatologists. Management, access to pulmonary testing and resources can differ.
- 4) Given the lack of access to rheumatologists in rural areas and a workforce shortage, any thoughts of this group to establishing a national network of ILD experts that could be accessed remotely for second opinions?
- 5) I didn't see any mention of use of PJP prophylaxis If any of these issues are addressed in the proposal, my apologies.

- Name: Aleksander Feoktistov
- Institution: NYC HHC/ Kings County
- Position: Chief of Rheumatology
- Disclosure (optional): Nothing to disclose

Comment:

As you aware, ACP ILD classification created group called IPAF - Interstitial pneumonitis with autoimmune features. I do not think that was reasonable. One of the reasons to that was the approach to CTD definition. ACP used classification criteria for clinical trials which can help in the field, but it is not diagnostic criteria, therefore, patients who might have been diagnosed with CTD by the specialist was misclassified. What criteria - clinical, classification will be used in the guidelines?

Another big part of the interstitial lung diseases does not get enough attention. Those are CTD antibody-associated interstitial lung disease without overt clinical presentation.

In our Rheumatology clinic we have about 50 patients with ILD, significant percentage are antibody - associated without clinical picture as CCP-ILD, U1-RNP-ILD, RNase P-ILD, some myositis associated antibodies as well as combination of those.

Will guidelines for RA-ILD will be applicable to CCP-ILD?

- Name: Ann Chauffe
- Institution: University of Florida
- Position: Assistant Professor of Medicine
- Disclosure (optional): Boehringer Ingelheim – speaker / consultant Novartis Pharmaceutical – investigator Priovant Therapeutics – investigator UCB – investigator Argenx – investigator Vera Therapeutics - investigator

Comment:

I am a rheumatologist at the University of Florida with a specific interest in ILD. In August 2022, I gave a talk at the Louisiana state rheumatology meeting on the treatment of rheumatic disease-ILD. Likewise, I spent ~50 hours reviewing the autoimmune ILD literature. Overall, there is a paucity of prospective data to drive treatment decisions. That being said, there did seem to be differences in response to treatment in the various underlying autoimmune disease (RA vs myositis, vs SLE). I would consider adjusting the project plan to the underlying autoimmune disease state driving the ILD. What works in RA-ILD doesn't always appear to be as effective in systemic sclerosis-ILD. It would also be important to breakout treatment by underlying ILD phenotype, as treatment for more inflammatory patterns, such as NSIP, differs from more fibrotic patterns, such as UIP.

Autoimmune ILD is an area I am passionately interested. If there is space, I and would be honored to join the ACR SARD-ILD project. Ann Chauffe, DO, MPH