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

RESPONSE TO PUBLIC COMMENTS

Thank you to those who took the time to review the project proposal and provide comments.

Comments related to scope of the guideline.

Many commenters suggested consideration of important affected patient groups, interventions, and outcomes. Due to the many possible interventions, diseases, and outcomes, we initially had thousands of PICO questions. The scope of the guidelines needed to be focused to address what we considered to be the most critical and clinically important of these.

Several patient groups will not be directly addressed by these guidelines. Patients with interstitial pneumonia and autoimmune features (IPAF) represent a heterogenous population and this classification was created for research purposes rather than as a clinical diagnosis. We suggest clinicians use judgment in classifying these early or incomplete presentations. For instance, a patient with interstitial lung disease, Raynaud's, and anti-Scl-70 antibodies may reasonably be considered to have an early presentation of systemic sclerosis. For the ANCA-associated vasculitides (AAV), we defer to the recent 2021 ACR/VF AAV treatment guidelines. We gave special consideration to pediatric patients, however, upon consulting the pediatrician who led the JIA ACR guidelines, we deferred inclusion given the predominance of adult clinicians in our core group and the existing Childhood Interstitial Lung Disease (CHILD) treatment guidance. Systemic lupus erythematosus (SLE) was not included in the ILD Guidelines as there are protean manifestations of lung disease in patients with SLE, and the treatment of the lung involvement most often aligns with treatment of the underlying SLE. While ILD can occur in patients with SLE, these patients often have multi system involvement and treatment guidance relates to their SLE.

Non-pharmacologic interventions had to be removed from the scope of this guideline to allow a comprehensive assessment of pharmacologic interventions, although we will comment on non-pharmacologic interventions such as pulmonary rehabilitation that are generally of benefit to patients with lung disease. To further limit the scope of the guidelines, we had to exclude radiographic patterns on CT thorax (e.g., usual interstitial pneumonia and nonspecific interstitial pneumonia) and pneumocystis jiroveci pneumonia (PJP) prophylaxis. Patient Reported Outcomes for ILD are not routinely integrated within clinical practice and therefore felt to be outside the scope of these clinical practice guidelines.

Diagnosis/classification of ILD and definitions of response to treatment

Diagnoses of ILD for the purposes of this guideline are based on clinician judgement. It is beyond the scope of this guideline to develop classification/diagnostic criteria. When evaluating published evidence, we will use established response definitions.

Timeline for screening and monitoring

Evidence related to frequency of screening and monitoring will be evaluated and will be addressed, as feasible.

Involvement of community rheumatologists and access of testing and resources

Access to testing and resources will be addressed in this guideline. We will ensure there is community representation.

Creation of a network of ILD experts to help provide second opinions remotely

This type of network is a good idea for the future but is outside the scope of these guidelines.

Different recommendations for different autoimmune conditions

The voting panel will consider whether different recommendations should be made for patients with different autoimmune disease diagnoses. Guidelines will also explicitly state that the treatment recommendations will be focused on the ILD rather than on the other clinical manifestations of the rheumatic disease. Clinical judgement will be needed to apply these guidelines to the treatment of individual patients with consideration of other factors, including disease manifestations and comorbidities.

Specific details about diagnostic tests

Based on radiologist input, non-contrast CT scans are currently typically performed with thin slices and are appropriate for ILD screening. Chest CT angiography (CTA), however, may not provide adequate resolution to provide a screening tool for ILD, if done for other reasons. Our guideline will focus on high resolution chest CT and will include mention of appropriate modalities for screening.

Full pulmonary function tests will be assessed including spirometry, lung volumes, and diffusion capacity for carbon monoxide (DLCO).