

# Interstitial Lung Disease Autoantibody Panel

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Interstitial lung diseases (ILDs) are characterized by impairment of lung function due to the accumulation of extracellular matrix proteins.<sup>1</sup> In some cases, ILD may be the first manifestation of a [connective tissue disease](#).<sup>2</sup> The detection of antibodies may help to establish a diagnosis of connective tissue disease-associated ILD, aid in prognosis, and support treatment decisions.<sup>2,3</sup>

## Disease Overview

ILDs are a group of disorders that may arise from connective tissue disease, systemic autoimmune rheumatic disease, genetic abnormalities, pneumotoxicity, infections, or unknown causes.<sup>2,3</sup> Distinguishing between ILD types is important for disease management, prognosis, and treatment decision-making.<sup>2,3</sup>

Antibody testing for ILD may be useful to identify antibodies associated with connective tissue disease and is recommended in conjunction with a complete clinical examination, patient history, imaging, nonspecific laboratory testing, and other testing as appropriate depending on results.<sup>2,3</sup> Refer to the ARUP Consult [Interstitial Lung Diseases](#) topic for more information about the typical testing strategy for ILD.

## Test Description

This antibody panel test may be useful for the evaluation of patients with ILD. Panel tests that detect a variety of autoantibodies are the most effective approach to identify connective tissue disease-associated ILD.

Patients with signs and symptoms of an [inflammatory myopathy](#) (eg, progressive proximal muscle weakness and/or other clinical findings suggestive of polymyositis/antisynthetase syndrome, dermatomyositis, and/or necrotizing autoimmune myopathy) in the absence of lung disease may benefit from evaluation with a targeted panel.

**Featured ARUP Testing**

[Interstitial Lung Disease Autoantibody Panel 3018869](#)

**Method:** Qualitative Immunoprecipitation / Semi-Quantitative Multiplex Bead Assay / Qualitative Immunoblot / Semi-Quantitative Enzyme-Linked Immunosorbent Assay (ELISA) / Quantitative Immunoturbidimetry / Semi-Quantitative Indirect Fluorescent Antibody (IFA)

Additional ARUP Myositis Panels		
ARUP Panel to Consider	Clinical Utility	Additional Test Information
<p><a href="#">Dermatomyositis and Polymyositis Panel 3018866</a></p> <p>Includes a subset of the antibodies on this panel that are specific to dermatomyositis and polymyositis</p>	<p>May be useful for the evaluation of patients with progressive proximal muscle weakness and/or with cutaneous manifestations suggestive of dermatomyositis</p>	<p><a href="#">Dermatomyositis and Polymyositis Panel Test Fact Sheet</a></p>
<p><a href="#">Polymyositis Panel 3018868</a></p> <p>Includes a subset of the antibodies on this panel that are specific to polymyositis</p>	<p>May be useful for the evaluation of patients with progressive proximal muscle weakness and antisynthetase syndrome</p>	<p><a href="#">Polymyositis Panel Test Fact Sheet</a></p>
<p><a href="#">Dermatomyositis Autoantibody Panel 3018870</a></p> <p>Includes a subset of the antibodies on this panel that are specific to dermatomyositis</p>	<p>May be useful for the evaluation of patients with characteristic cutaneous manifestations of dermatomyositis with or without muscle weakness</p>	<p><a href="#">Dermatomyositis Autoantibody Panel Test Fact Sheet</a></p>
<p><a href="#">Extended Myositis Panel 3018867</a></p> <p>Antibodies overlap with the antibodies on this panel</p>	<p>May be useful for the evaluation of patients with progressive proximal muscle weakness and/or other clinical findings suggestive of polymyositis/antisynthetase syndrome, dermatomyositis, necrotizing autoimmune myopathy, or overlap syndromes associated with connective tissue disease</p>	<p><a href="#">Extended Myositis Panel Test Fact Sheet</a></p>

## Antibodies Tested

This panel detects a selection of antibodies associated with ILD. For more information about the clinical associations with each of these antibodies, visit the ARUP Consult [Interstitial Lung Disease](#) topic.

Interstitial Lung Disease Autoantibody Panel: Antibodies Detected and Methodology	
Interstitial Lung Disease-Associated Antibodies	
Antibody	Method
Cyclic citrullinated peptide Ab, IgG and IgA	Semiquantitative ELISA
Rheumatoid factor	Quantitative immunoturbidimetry
RNA polymerase III Ab, IgG	Semiquantitative ELISA
Scleroderma (Scl-70) (ENA) Ab, IgG	Semiquantitative multiplex bead assay
Myositis-Specific Antibodies <sup>a</sup>	
Antibody	Method
EJ (glycyl-tRNA synthetase) Ab	Qualitative immunoprecipitation
Ha (tyrosyl-tRNA synthetase) Ab	Qualitative immunoblot and qualitative immunoprecipitation
Ks (asparaginyl-tRNA synthetase) Ab	Qualitative immunoblot and qualitative immunoprecipitation
Jo-1 (histidyl-tRNA synthetase) Ab, IgG	Semiquantitative multiplex bead assay
MDA5 (CADM-140) Ab	Qualitative immunoblot
NXP2 (nuclear matrix protein-2) Ab	Qualitative immunoblot
OJ (isoleucyl-tRNA synthetase) Ab	Qualitative immunoprecipitation
PL-7 (threonyl-tRNA synthetase) Ab	Qualitative immunoprecipitation
PL-12 (alanyl-tRNA synthetase) Ab	Qualitative immunoprecipitation
SRP (signal recognition particle) Ab	Qualitative immunoprecipitation
Zo (phenylalanyl-tRNA synthetase) Ab	Qualitative immunoblot and qualitative immunoprecipitation
Myositis-Associated Antibodies <sup>b</sup>	
Antibody	Method
Antinuclear Ab (ANA), Hep-2, IgG <sup>c</sup>	Semiquantitative indirect fluorescent antibody
Ku Ab	Qualitative immunoprecipitation
PM/Scl-100 Ab, IgG	Qualitative immunoblot
SSA-52 (Ro52) (ENA) Ab, IgG	Semiquantitative multiplex bead assay
SSA-60 (Ro60) (ENA) Ab, IgG	Semiquantitative multiplex bead assay

<sup>a</sup>Myositis-specific antibodies are generally regarded as mutually exclusive with rare exceptions. The occurrence of two or more myositis-specific antibodies should be carefully evaluated in the context of the patient's clinical presentation. Refer to the ARUP Consult [Inflammatory Myopathies – Myositis](#) topic for more information about myositis.

<sup>b</sup>Myositis-associated antibodies may be found in patients with overlap syndromes and other conditions and are generally not specific for myositis.

<sup>c</sup>The presence of ANA is a feature of systemic autoimmune rheumatic diseases, however, ANA lacks diagnostic specificity and may occur in the general population. Positive ANA must be confirmed by more specific serologic tests. For more information, refer to the [Antinuclear Antibody \(ANA\) With Hep-2 Substrate](#) Test Fact Sheet.

Ab, antibody; ELISA, enzyme-linked immunosorbent assay; ENA, extractable nuclear antigen; IgA, immunoglobulin A; IgG, immunoglobulin G

Some antibodies may be orderable separately; refer to the [ARUP Laboratory Test Directory](#).

## Test Interpretation

### Results

- **Positive:** Antibody detected.
  - May support a clinical diagnosis of connective tissue disease-associated ILD.
  - Results for specific antibodies may be reported as low/weak positive, positive, or high/strong positive.
  - Antinuclear antibody (ANA) results are reported as a pattern and titer. For more information on the interpretation of ANA results, refer to the [Antinuclear Antibody \(ANA\) With Hep-2 Substrate](#) Test Fact Sheet.
  - Additional interpretive information for positive antibodies may be provided on the Patient Report.
- **Negative:** Antibody not detected.

### Limitations

Results are not diagnostic in the absence of other findings; the complete clinical context should be considered.

Negative results do not rule out a diagnosis of connective tissue disease-associated ILD.

## References

1. Myall KJ, West A, Kent BD. [Sleep and interstitial lung disease](#). *Curr Opin Pulm Med*. 2019;25(6):623-628.
2. Antin-Ozerkis D, Hinchcliff M. [Connective tissue disease-associated interstitial lung disease: evaluation and management](#). *Clin Chest Med*. 2019;40(3):617-636.
3. Kalchiem-Dekel O, Galvin JR, Burke AP, et al. [Interstitial lung disease and pulmonary fibrosis: a practical approach for general medicine physicians with focus on the medical history](#). *J Clin Med*. 2018;7(12):476.

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