

### LABORATORY TEST DIRECTORY

# Extended Myositis Panel

Last Literature Review: March 2024 Last Update: January 2025

Myositis is characterized by inflammation of the skeletal muscles involved in movement. <sup>1,2</sup> The detection of antibodies may help to establish a diagnosis, aid in prognosis, and support treatment decisions.

### Disease Overview

Myositis may occur in a number of inflammatory myopathies, including polymyositis/antisynthetase syndrome, dermatomyositis, necrotizing

autoimmune myopathy, and sporadic inclusion body myositis, as well as overlap syndromes with connective tissue diseases.<sup>1,2</sup> The primary symptom of all forms of myositis is progressive muscle weakness that may develop over a period of weeks, months, or years.<sup>1,2</sup> Other symptoms may include joint pain and fatigue.<sup>1,2</sup>

Antibody testing for myositis should be considered after a standard workup for inflammatory myopathies because it may aid in distinguishing between myopathies, 1.2 which can have important implications for therapy and prognosis.

Refer to the ARUP Consult Inflammatory Myopathies – Myositis topic for more information about myositis and the typical testing strategy for inflammatory myopathies.

## **Test Description**

This antibody panel test 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. Clinical phenotypes for specific antibody-associated inflammatory myopathies often overlap, and targeted panels allow for rapid identification of associated antibodies. Use of the most targeted panel, ie, the panel that most closely matches the patient's complete clinical phenotype, is recommended.

Additional ARUP Myositis Panels		
ARUP Panel to Consider	Clinical Utility	Additional Test Information
Dermatomyositis and Polymyositis Panel 3018866  Includes a subset of the antibodies on this panel that are specific to dermatomyositis and polymyositis	May be useful for the evaluation of patients with progressive proximal muscle weakness and/or with cutaneous manifestations suggestive of dermatomyositis	Dermatomyositis and Polymyositis Panel Test Fact Sheet
Polymyositis Panel 3018868 Includes a subset of the antibodies on this panel that are specific to polymyositis	May be useful for the evaluation of patients with progressive proximal muscle weakness and antisynthetase syndrome	Polymyositis Panel Test Fact Sheet
Dermatomyositis Autoantibody Panel 3018870 Includes a subset of the antibodies on this panel that are specific to dermatomyositis	May be useful for the evaluation of patients with characteristic cutaneous manifestations of dermatomyositis with or without muscle weakness	Dermatomyositis Autoantibody Panel Test Fact Sheet
Interstitial Lung Disease Autoantibody Panel 3018869 Antibodies overlap with the antibodies on this panel	May be useful for the evaluation of patients with interstitial lung disease with or without other signs and symptoms of myositis	Interstitial Lung Disease Autoantibody Panel Test Fact Sheet

### Featured ARUP Testing

#### Extended Myositis Panel 3018867

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

# **Antibodies Tested**

This panel detects a selection of antibodies specific to or associated with myositis. For more information about the clinical associations with each of these antibodies, visit the ARUP Consult Inflammatory Myopathies – Myositis topic.

Extended Myositis	Panel: Antibodies Detected and Methodology	
Myositis-Specific Antibodies <sup>a,b</sup>		
Dermatomyositis Antibodies <sup>c</sup>		
Antibody	Method	
MDA5 (CADM-140) Ab	Qualitative immunoblot	
Mi-2 (nuclear helicase protein) Ab	Qualitative immunoprecipitation	
NXP2 (nuclear matrix protein-2) Ab	Qualitative immunoblot	
P155/140 Ab	Qualitative immunoprecipitation	
SAE1 (SUMO activating enzyme) Ab	Qualitative immunoblot	
TIF-1 gamma (155 kDa) Ab	Qualitative immunoblot	
Polymyositis Antibodies <sup>d</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	
OJ (isoleucyl-tRNA synthetase) Ab	Qualitative immunoprecipitation	
PL-7 (threonyl-tRNA synthetase) Ab	Qualitative immunoprecipitation	
PL-12 (alanyl-tRNA synthetase) Ab	Qualitative immunoprecipitation	
Smith/RNP (ENA) Ab, IgG	Qualitative immunoprecipitation	
Zo (phenylalanyl-tRNA synthetase) Ab	Qualitative immunoblot and qualitative immunoprecipitation	
Myositis-Associated Antibodies <sup>e</sup>		
Antibody	Method	
Antinuclear Ab (ANA), Hep-2, IgG <sup>f</sup>	Semiquantitative indirect fluorescent antibody	
Fibrillarin (U3 RNP) Ab, IgG	Qualitative immunoblot	
Ku Ab	Qualitative immunoprecipitation	
PM/ScI-100 Ab, IgG	Qualitative immunoblot	
Smith/RNP (ENA) Ab, IgG	Semiquantitative multiplex bead assay	
SSA-52 (Ro52) (ENA) Ab, IgG	Semiquantitative multiplex bead assay	
SSA-60 (Ro60) (ENA) Ab, IgG	Semiquantitative multiplex bead assay	

<sup>&</sup>lt;sup>a</sup>This subset of antibodies is also available as a combined dermatomyositis and myositis panel.

<sup>&</sup>lt;sup>b</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>c</sup>This subset of antibodies is also available as a dermatomyositis panel.
- <sup>d</sup>This subset of antibodies is also available as a polymyositis panel.
- <sup>e</sup>Myositis-associated antibodies may be found in patients with overlap syndromes and other conditions and are generally not specific for myositis.

<sup>f</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; ENA, extractable nuclear antigen; IgG, immunoglobulin G; RNP, ribonucleoprotein

Some antibodies may be orderable separately; refer to the ARUP Laboratory Test Directory.

## **Test Interpretation**

### Results

- · Positive: Antibody detected.
  - · Supports a clinical diagnosis of dermatomyositis, polymyositis, necrotizing autoimmune myopathy, and/or an overlap syndrome.
  - 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.
  - 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.
  - Myositis-associated antibodies may be found in patients with overlap syndromes and other conditions, and are generally not specific for myositis.
- Negative: Antibody not detected.

#### Limitations

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

Negative results do not rule out a diagnosis of inflammatory myopathy or overlap syndrome.

### References

- 1. Selva-O'Callaghan A, Pinal-Fernandez I, Trallero-Araguás E, et al. Classification and management of adult inflammatory myopathies. *Lancet Neurol.* 2018;17(9):816-828
- 2. Schmidt J. Current classification and management of inflammatory myopathies. J Neuromuscul Dis. 2018;5(2):109-129.

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