

## Overview

### Useful For

Evaluating patients with clinical features of idiopathic inflammatory myositis, especially those with clinical features suggestive of anti-synthetase syndrome or interstitial lung disease

### Testing Algorithm

For more information see [Connective Tissue Disease Cascade](#).

### Special Instructions

- [Connective Tissue Disease Cascade](#)

### Method Name

Multiplex Flow Immunoassay

### NY State Available

Yes

## Specimen

### Specimen Type

Serum

### Specimen Required

#### Collection Container/Tube:

**Preferred:** Serum gel

**Acceptable:** Red top

**Submission Container/Tube:** Plastic vial

**Specimen Volume:** 0.5 mL

**Collection Instructions:** Centrifuge and aliquot serum into a plastic vial.

### Forms

If not ordering electronically, complete, print, and send a [Kidney Transplant Test Request](#) with the specimen.

### Specimen Minimum Volume

0.35 mL

### Reject Due To

Gross hemolysis	Reject
Gross lipemia	Reject

Gross icterus	OK
Heat-Treated	Reject

### Specimen Stability Information

Specimen Type	Temperature	Time	Special Container
Serum	Refrigerated (preferred)	21 days	
	Frozen	21 days	

### Clinical & Interpretive

#### Clinical Information

Based on their specificity, autoantibodies in idiopathic inflammatory myopathies (IIM) are grouped into myositis specific (MSA) and myositis associated autoantibodies (MAA).<sup>(1-3)</sup> Among the MSA, autoantibodies against aminoacyl-tRNA synthetases (aaRSs) represent the most common antibodies and can be detected in 25% to 35% of patients with mainly anti-synthetase syndrome (ASSD).<sup>(1-4)</sup> ASSD is an autoimmune disease characterized by the presence of autoantibodies targeting one of several aaRSs along with clinical features including interstitial lung disease, myositis, Raynaud's phenomenon, arthritis, mechanic's hands, and fever.<sup>(3,4)</sup> The family of aaRSs consists of highly conserved cytoplasmic and mitochondrial enzymes, one for each amino acid, which are essential for the RNA translation machinery and protein synthesis. Along with their main functions, aaRSs are involved in the development of immune responses, regulation of transcription, and gene-specific silencing of translation.<sup>(4)</sup>

Anti-Jo-1 antibody is the most frequently detected anti-aaRS antibody in ASSD and targets the histidyl tRNA synthetase which catalyses the binding of the histidine to its cognate tRNA during protein synthesis.<sup>(4,5)</sup> Other described anti-aaRSs reported in ASSD include PL-7 (threonyl), PL-12 (alanyl), OJ (isoleucyl), EJ (glycyl), KS (asparaginyl), Zo (phenylalanyl) and Ha (tyrosyl).<sup>(4)</sup> The presence of these autoantibodies has become a key feature for classification and diagnosis of IIM and is increasingly used to define clinically distinguishable IIM subsets. Each anti-ARS antibody seems to define a distinctive clinical phenotype.<sup>(3)</sup>

In addition to the characteristic features associated with the presence of anti-Jo-1 antibodies in patients with ASSD, testing for anti-aaRS autoantibodies including anti-Jo-1 antibody may be indicated with cytoplasmic speckled pattern using HEp-2 substrate by indirect immunofluorescence assay.<sup>(6,7)</sup> In the context of ASSD, their presence may be associated with positivity for anti-Ro52 antibodies which is an MAA.<sup>(8)</sup> In routine clinical testing, anti-Jo-1 antibody testing may be performed using a variety of solid-phase immunoassays such as the enzyme-linked immunosorbent assay, line immunoassay, chemiluminescence immunoassay, fluorescent enzyme immunoassay, and multiplex immunoassay such as the BioPlex.<sup>(6,9,10)</sup> The performance characteristics of these assays for the detection of anti-Jo-1 antibody have not been extensively investigated to establish comparability.<sup>(9,10)</sup>

For more information see [Connective Tissue Disease Cascade](#).

#### Reference Values

<1.0 U (negative)

> or =1.0 U (positive)

Reference values apply to all ages.

## Interpretation

A positive result for anti-Jo 1 antibody is suggestive of anti- synthetase syndrome or may indicate a risk for myositis with or without interstitial lung disease.

## Cautions

A negative test for Jo 1 antibodies does not exclude the diagnosis of idiopathic inflammatory myositis.

## Clinical Reference

1. Satoh M, Tanaka S, Ceribelli A, Calise SJ, Chan EK. A comprehensive overview on myositis-specific antibodies: New and old biomarkers in idiopathic inflammatory myopathy. *Clin Rev Allergy Immunol.* 2017;52(1):1-19
2. Mariampillai K, Granger B, Amelin D, et al. Development of a new classification system for idiopathic inflammatory myopathies based on clinical manifestations and myositis-specific autoantibodies. *JAMA Neurol.* 2018;75(12):1528-1537
3. Cavagna L, Nuno L, Scire CA, et al. Clinical spectrum time course in anti Jo-1 positive antisynthetase syndrome: Results from an international retrospective multicenter study. *Medicine (Baltimore).* 2015;94(32):e1144
4. Galindo-Feria AS, Notarnicola A, Lundberg IE, Horuluoglu B. Aminoacyl-tRNA synthetases: On anti-synthetase syndrome and beyond. *Front Immunol.* 2022;13:866087
5. Freist W, Verhey JF, Ruhlmann A, Gauss DH, Arnez JG. Histidyl-tRNA synthetase. *Biol Chem.* 1999;380(6):623-646
6. Damoiseaux J, Andrade LEC, Carballo OG, et al. Clinical relevance of HEp-2 indirect immunofluorescent patterns: the International Consensus on ANA patterns (ICAP) perspective. *Ann Rheum Dis.* 2019;78(7):879-889
7. Tebo AE. Autoantibody testing in idiopathic inflammatory myopathies. *J Appl Lab Med.* 2022;7(1):387-390
8. Rutjes SA, Vree Egberts WT, Jongen P, et al. Anti-Ro52 antibodies frequently co-occur with anti-Jo-1 antibodies in sera from patients with idiopathic inflammatory myopathy. *Clin Exp Immunol.* 1997;109(1):32-40
9. Cavazzana I, Fredi M, Ceribelli A, et al. Testing for myositis specific autoantibodies: Comparison between line blot and immunoprecipitation assays in 57 myositis sera. *J Immunol Methods.* 2016;433:1-5
10. Espinosa-Ortega F, Holmqvist M, Alexanderson H, et al. Comparison of autoantibody specificities tested by a line blot assay and immunoprecipitation-based algorithm in patients with idiopathic inflammatory myopathies. *Ann Rheum Dis.* 2019;78(6):858-860

## Performance

### Method Description

Recombinant Jo 1 antigen is coupled covalently to polystyrene microspheres, which are impregnated with fluorescent dyes to create a unique fluorescent signature. Jo 1 antibodies, if present in diluted serum, bind to the Jo 1 antigen on the microspheres. The microspheres are washed to remove extraneous serum proteins. Phycoerythrin (PE)-conjugated antihuman IgG antibody is then added to detect IgG anti-Jo 1 bound to the microspheres. The microspheres are washed to remove unbound conjugate, and bound conjugate is detected by laser photometry. A primary laser reveals the fluorescent signature of each microsphere to distinguish it from microspheres that are labeled with other antigens, and a secondary laser reveals the level of PE fluorescence associated with each microsphere. Results are calculated by comparing the median fluorescence response for Jo 1 microspheres to a 4-point calibration curve. (Package insert: Bioplex 2200 ANA Screen. Bio-Rad Laboratories, 02/2019)

### PDF Report

No

**Day(s) Performed**

Monday through Saturday

**Report Available**

1 to 3 days

**Specimen Retention Time**

14 days

**Performing Laboratory Location**

Mayo Clinic Laboratories - Rochester Superior Drive

**Fees & Codes****Fees**

- Authorized users can sign in to [Test Prices](#) for detailed fee information.
- Clients without access to Test Prices can contact [Customer Service](#) 24 hours a day, seven days a week.
- Prospective clients should contact their account representative. For assistance, contact [Customer Service](#).

**Test Classification**

This test has been cleared, approved, or is exempt by the US Food and Drug Administration and is used per manufacturer's instructions. Performance characteristics were verified by Mayo Clinic in a manner consistent with CLIA requirements.

**CPT Code Information**

86235

**LOINC® Information**

Test ID	Test Order Name	Order LOINC® Value
JO1	Jo 1 Ab, IgG, S	33571-1

Result ID	Test Result Name	Result LOINC® Value
JO1	Jo 1 Ab, IgG, S	33571-1