Comprehensive, Innovative Profile and Tests to Help Identify Inflammatory Myopathies
Inflammatory Myopathies

Idiopathic inflammatory myopathies (IIM), commonly known as myositis, are rare conditions that can affect multiple organs apart from muscle and often lead to a severe impairment of the quality of life.

Diagnosis and treatment are often challenging and usually require testing of auto-antibodies.¹

Labcorp offers a myositis profile performed using RIPA and EIA methodologies. MyoMarker® 3 Plus profile includes both myositis-specific antibodies (MSA) and myositis-associated antibodies (MAA), as well as anti-SAE-1 antibody.

Clinical Utility

IIMs are a heterogeneous group of disorders characterized by muscle weakness, resulting from chronic muscle inflammation of unknown cause. Patients with IIM have a variety of autoantibodies with various clinical associations that fall into two main groups:

- **Myositis specific autoantibodies (MSA):** MSA are highly specific for patients with polymyositis (PM), dermatomyositis (DM), anti-synthetase syndrome, and necrotizing myositis
- **Myositis associated autoantibodies (MAA):** MAA appears in myositis overlap syndromes and in other connective tissue diseases, which correlate with certain clinical and/or pathophysiological conditions
Diagnosis and treatment are often challenging and usually require testing of auto-antibodies.¹
Myositis Profile and Related Testing

MyoMarker® 3 Plus Profile includes both myositis-specific antibodies (MSA) and myositis-associated antibodies (MAA), as well as anti-SAE-1 antibody.

MSA are found only in patients with myositis and have been shown to be highly specific for patients with polymyositis (PM), dermatomyositis (DM), anti-synthetase syndrome, necrotizing myositis and overlap syndromes. MAA can be found in patients with overlap syndromes such as polymyositis/scleroderma and may also be found in non-overlap syndromes.

Labcorp now offers MyoMarker® 3 Plus Profile performed using RIPA and EIA methodologies.

RIPA gel radiography utilizes immunoprecipitation, gel electrophoresis and autoradiography to identify radioisotope-labelled proteins from human erythroleukemic cell extracts that are targeted by autoantibodies in patient serum. RIPA gel radiography is a powerful, reliable technology that has been used and perfected for more than 30 years to identify many PM/DM autoantibodies with high sensitivity, specificity and reproducibility. It is the original methodology of studies that defined various myositis-specific and myositis-associated antibodies.²

<table>
<thead>
<tr>
<th>Test Name</th>
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<tbody>
<tr>
<td>MyoMarker® 3 Plus Profile (RDL)</td>
<td>520085</td>
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</table>

**Myositis-Specific Antibodies**

- Anti-Jo-1
- Anti-PL-7
- Anti-PL-12
- Anti-EJ
- Anti-OJ
- Anti-SRP
- Anti-Mi-2
- Anti-TIF-1gamma (part of P155/140 Kd)
- Anti-MDA-5-Ab (CADM-140)
- Anti-NXP-2 (P140)
- Anti-SAE-1

**Myositis-Associated Antibodies**

- Anti-PM/Scl-100
- Anti-SS-A 52kD
- Anti-Ku
- Anti-U1 RNP
- Anti-U2 RNP
- Anti-U3 RNP (Fibrillarin)
# Myositis-Specific Antibodies (MSA)

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<tr>
<td>Anti-Synthetase Profile (RDL)</td>
<td>520193</td>
<td>RIPA Gel Radiography, ELISA</td>
<td>Anti-synthetase syndrome: myositis, non-erosive arthritis, ILD, Raynaud’s phenomenon, unexplained fever, mechanic’s hands. Anti-Jo-1 is the most common MSA, in about 30% of adult IIM, 1-3% of JM. Anti-PL-7, Anti-PL-12 in 3-4%. Anti-E,J, Anti-OJ &lt;2%.1-3</td>
</tr>
<tr>
<td>Anti-SRP Ab (RDL)</td>
<td>520014</td>
<td>RIPA Gel Radiography</td>
<td>Closely associated with necrotizing myositis, acute onset, rapidly progressive, severe weakness, high CK, frequent cardiac and lung involvement, poor response or refractory to treatment. 5-13% of adult IIM (higher frequency in Asians and African Americans) and &lt;2% of JM.4-6</td>
</tr>
<tr>
<td>Anti-Mi-2 Ab (RDL)</td>
<td>520000</td>
<td>RIPA Gel Radiography (detects autoantibodies to both alpha and beta Mi-2 subunits)</td>
<td>Associated with classic DM with mild to moderate weakness and hallmark cutaneous features (shawl rash, heliotrope rash, V-sign, Gottron’s papules), good response to treatment and lower incidence of cancer compared to other DM. 9-24% of adult DM, 4-10% of JDM.4-6</td>
</tr>
<tr>
<td>Anti-TIF-1gamma Ab (RDL) (synonymous with Anti-P155/140)</td>
<td>520017</td>
<td>ELISA</td>
<td>Highly associated with malignancy which is found in 50-100% of positive adults. 89% specificity and 78% sensitivity for cancer-associated DM. No cancer association in children. 13-31% of adult DM, 22-29% of JDM.4-7</td>
</tr>
<tr>
<td>Anti-MDA-5-Ab (CADM-140) (RDL)</td>
<td>520002</td>
<td>ELISA</td>
<td>Associated with CADM, i.e. skin involvement and absent or mild muscle symptoms; rapidly progressive ILD: skin ulcerations and papules, oral ulcerations, arthritis. 10-48% Asian and 0-13% Caucasian adult DM, 7-38% of JDM.4-7</td>
</tr>
<tr>
<td>Anti-NXP-2 (P140) Ab (RDL)</td>
<td>520004</td>
<td>ELISA</td>
<td>In adult DM, associated with malignancy and ILD, significant muscle weakness, elevated CK. In JDM, associated with cutaneous calcinosis cutis. 1-17% of adult DM, 23-25% of JDM.4-6</td>
</tr>
<tr>
<td>Anti-SAE1 Ab, IgG (RDL)</td>
<td>520011</td>
<td>ELISA</td>
<td>Very specific for DM. Cutaneous DM with typical skin lesions at initial disease onset progressing to myositis. 6-8% of Caucasian, 2% Asian adult PM/DM.4-6</td>
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**ANA:** Antinuclear Antibody  
**AIM:** Autoimmune Myositis  
**CADM:** Clinically Amyopathic Dermatomyositis  
**CK:** Creatine Kinase  
**DM:** Dermatomyositis  
**dSSc:** Diffuse Systemic Sclerosis  
**ELISA:** Enzyme-linked Immunosorbent Assay  
**IIM:** Idiopathic Inflammatory Myopathy  
**ILD:** Interstitial Lung Disease  
**JDM:** Juvenile Dermatomyositis  
**JM:** Juvenile Myositis  
**MDA5:** Melanoma Differentiation-Associated Gene 5  
**NXP-2:** Nuclear Matrix Protein 2  
**PM:** Polymyositis  
**RA:** Rheumatoid Arthritis  
**RIPA:** Radioimmunoprecipitation Assay  
**RNP:** Ribonucleoprotein  
**SAE:** Small Ubiquitin-like Modifier Activating Enzyme  
**SCL:** Scleroderma  
**SDS:** Sodium Dodecyl Sulfate Polyacrylamide Gel Electrophoresis  
**sIBM:** Sporadic Inclusion Body Myositis  
**SLE:** Systemic Lupus Erythematosus  
**SRP:** Signal Recognition Particle  
**SS:** Sjögren’s Syndrome  
**SSc:** Systemic Sclerosis  
**Anti-TIF-1gamma:** Transcriptional Intermediary Factor 1  
**UCTD:** Undifferentiated Connective Tissue Disease
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<td>Anti-PM/Scl-100 Ab (RDL)</td>
<td>520007</td>
<td>ELISA</td>
<td>3-9% worldwide frequency. Approximately 20% of dSSc, 18% of overlap syndrome and 2-4% of SSc; associated with digital ulcers and lung fibrosis.8,9</td>
</tr>
<tr>
<td>Anti-Ku Ab (RDL)</td>
<td>520030</td>
<td>ELISA</td>
<td>Most commonly associated with SSc overlap disorders (with myositis or lupus). Common clinical features include diffuse skin changes, ILD, myositis, Raynaud’s, sicca.8,10</td>
</tr>
<tr>
<td>Anti-SS-A 52kD Ab, IgG (RDL)</td>
<td>520015</td>
<td>ELISA</td>
<td>Reported in a variety of autoimmune diseases such as SLE, SS, RA, SSc, DM, malignancies and fibromyalgia. 5.9% of ANA-positive individuals.11</td>
</tr>
<tr>
<td>Anti-U1 RNP Ab (RDL)</td>
<td>520034</td>
<td>ELISA</td>
<td>Positive in 95-100% of MCTD. May also occur in SLE, IIM.12</td>
</tr>
<tr>
<td>Anti-U2 RNP Ab (RDL)</td>
<td>520024</td>
<td>RIPA Gel Radiography</td>
<td>Commonly associated with scleroderma/myositis overlap syndrome.13</td>
</tr>
<tr>
<td>Anti-U3 RNP Antibodies (Fibrillarin) (RDL)</td>
<td>520019</td>
<td>RIPA Gel Radiography</td>
<td>Mostly found in SSc/myositis overlap. 4-10% of diffuse SSc, &lt;2% of limited SSc. More prevalent in African Americans. Associated with pulmonary arterial hypertension (PAH), myositis, cardiac and renal involvement.8,14</td>
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<td>Anti-HMGCR Ab (RDL)</td>
<td>520057</td>
<td>ELISA</td>
<td>Specific for necrotizing myositis, statin-induced. Highly elevated DK. 5% of adult PM/DM. 63% of patients had exposure to statin prior to developing muscle weakness. Antibody levels can be used for monitoring in statin-exposed patients.4,6,15</td>
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<tr>
<td>Anti-cN-1A Ab (NT5c1A) IBM (RDL)</td>
<td>520061</td>
<td>ELISA</td>
<td>Highly associated with sIBM (33-76% sensitivity and 87-100% specificity. Most common IIM in patients older than 50 y/o with poor response to conventional immunotherapies.36</td>
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References