



MYOMARKER® PROFILE

Comprehensive, Innovative Profile and Tests to Help Identify Inflammatory Myopathies



MYOMARKER® PROFILE

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.



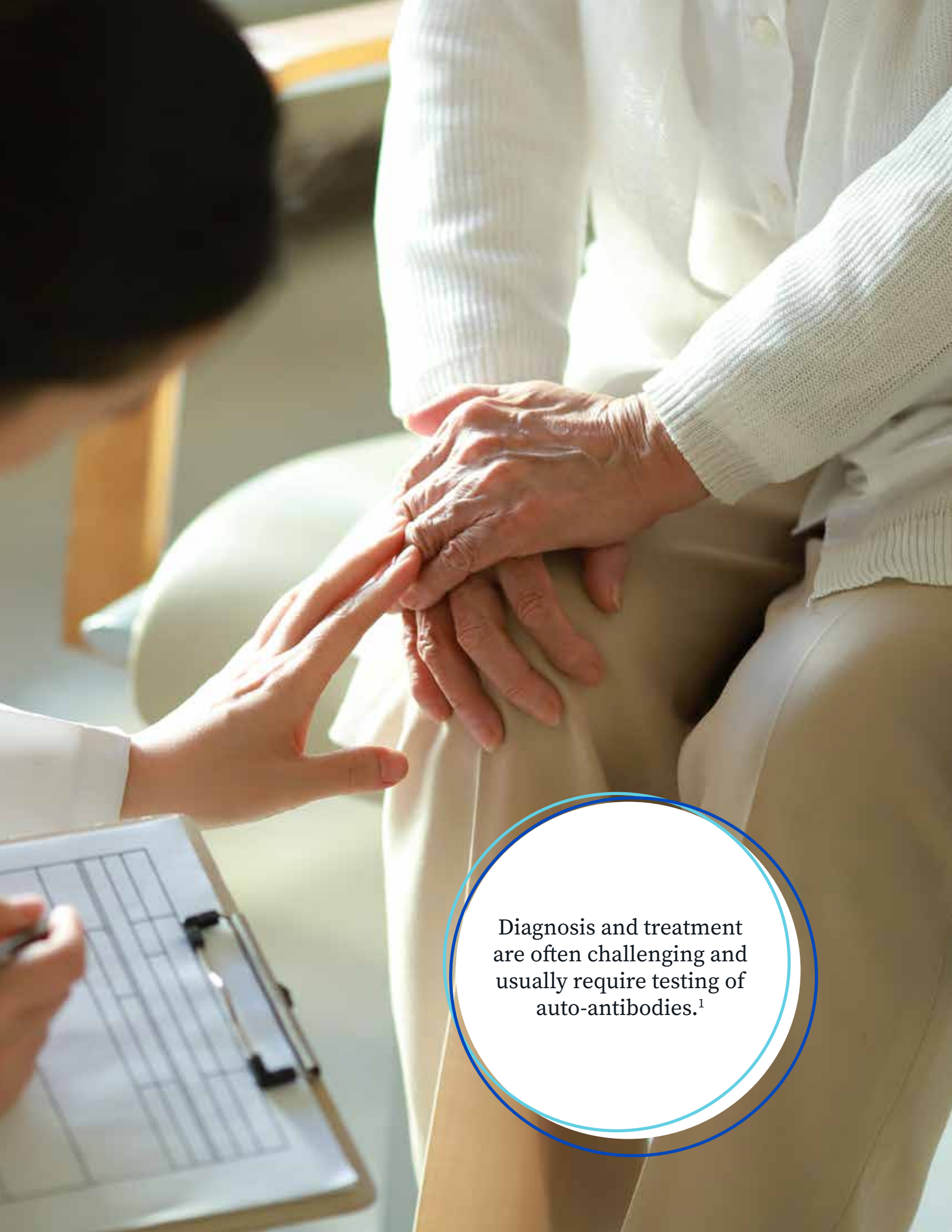
Diagnosis and treatment are often challenging and usually require testing of auto-antibodies.¹

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

Test Name	Test No.
MyoMarker® 3 Plus Profile (RDL)	520085

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)

Test Name	Test No.	Methodology	Clinical Associations
Anti-Synthetase Profile (RDL) (Anti-Jo-1; PL-7; PL-12; EJ; OJ)	520193	RIPA Gel Radiography, ELISA	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-EJ, Anti-OJ <2%. ^{1,3}
Anti-SRP Ab (RDL)	520014	RIPA Gel Radiography	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 <2% of JM. ^{4,6}
Anti-Mi-2 Ab (RDL)	520000	RIPA Gel Radiography (detects autoantibodies to both alpha and beta Mi-2 subunits)	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}
Anti-TIF-1gamma Ab (RDL) (synonymous with Anti-P155/140)	520017	ELISA	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. ^{4,6,7}
Anti-MDA-5-Ab (CADM-140) (RDL)	520002	ELISA	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. ^{4,6,7}
Anti-NXP-2 (P140) Ab (RDL)	520004	ELISA	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}
Anti-SAE1 Ab, IgG (RDL)	520011	ELISA	Very specific for DM. Cutaneous DM ² with typical skin lesions at initial disease onset progressing to myositis. 6-8% of Caucasian, 2% Asian adult PM/DM. ^{2,4}

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

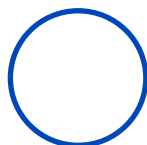
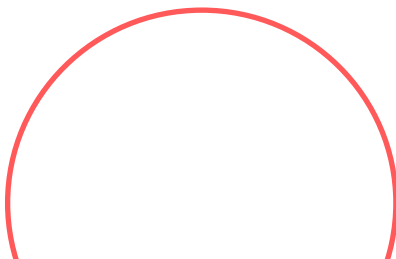
SRP: Signal Recognition Particle

SS: Sjögren's Syndrome

SSc: Systemic Sclerosis

Anti-TIF-1gamma: Transcriptional Intermediary Factor 1

UCTD: Undifferentiated Connective Tissue Disease



Myositis-Associated Antibodies (MAA)

Test Name	Test No.	Methodology	Clinical Associations
Anti-PM/Scl-100 Ab (RDL)	520007	ELISA	3-9% worldwide frequency. Approximately 20% of dSSc, 18% of overlap syndrome and 2-4% of SSs; associated with digital ulcers and lung fibrosis. ^{8,9}
Anti-Ku Ab (RDL)	520030	ELISA	Most commonly associated with SSs overlap disorders (with myositis or lupus). Common clinical features include diffuse skin changes, ILD, myositis, Raynaud's, sicca. ^{8,10}
Anti-SS-A 52kD Ab, IgG (RDL)	520015	ELISA	Reported in a variety of autoimmune diseases such as SLE, SS, RA, SSs, DM, malignancies and fibromyalgia. 5.5% of ANA-positive individuals. ¹¹
Anti-U1 RNP Ab (RDL)	520034	ELISA	Positive in 95-100% of MCTD. May also occur in SLE, IIM. ¹²
Anti-U2 RNP Ab (RDL)	520024	RIPA Gel Radiography	Commonly associated with scleroderma/myositis overlap syndrome. ¹³
Anti-U3 RNP Antibodies (Fibrillar) (RDL)	520019	RIPA Gel Radiography	Mostly found in SSs/myositis overlap. 4-10% of diffuse SSs, <2% of limited SSs. More prevalent in African Americans. Associated with pulmonary arterial hypertension (PAH), myositis, cardiac and renal involvement. ^{8,14}

Other Myositis-Specific Antibodies (MSA)

Test Name	Test No.	Methodology	Clinical Associations
Anti-HMGCR Ab (RDL)	520057	ELISA	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}
Anti-cN-1A Ab (NT5c1A) IBM (RDL)	520061	ELISA	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. ¹⁶

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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

MCTD: Mixed Connective Tissue Disease

MDA5: Melanoma Differentiation-Associated Gene 5

NXP-2: Nuclear Matrix Protein 2

PM: Polymyositis

RA: Rheumatoid Arthritis

RIPA: Radioimmunoprecipitation Assay

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SAE: Small Ubiquitin-like Modifier Activating Enzyme

SCL: Scleroderma

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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

A close-up photograph of a woman with blonde hair, wearing a white button-down shirt, holding her right hand with her left hand. The background is softly blurred. A blue circular graphic is overlaid on the image, containing white text.

Inflammatory myopathies - commonly called myositis - are rare conditions that can affect multiple organs apart from muscle and often lead to a severe impairment of the quality of life.



Labcorp now offers legacy RDL MyoMarker® 3 Plus Profile to help identify inflammatory myopathies.

References

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