MYOMARKER® PROFILE

Comprehensive, Innovative Profile and Tests to Help Identify Inflammatory Myopathies

labcorp

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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

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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 Ubiquintin-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 SSc; associated with digital ulcers and lung fibrosis.[8,9]
Anti-Ku Ab (RDL)	**520030**	ELISA	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]
Anti-SS-A 52kD Ab, IgG (RDL)	**520015**	ELISA	Reported in a variety of autoimmune diseases such as SLE, SS, RA, SSc, 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 (Fibrillarin) (RDL)	**520019**	RIPA Gel Radiography	Mostly found in SSc/myositis overlap. 4-10% of diffuse SSc, <2% of limited SSc. 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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Labcorp now offers legacy RDL MyoMarker® 3 Plus Profile to help identify inflammatory myopathies.

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