Neuberg DIAGNOSTICS

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New INSIGHTS

Myositis panel

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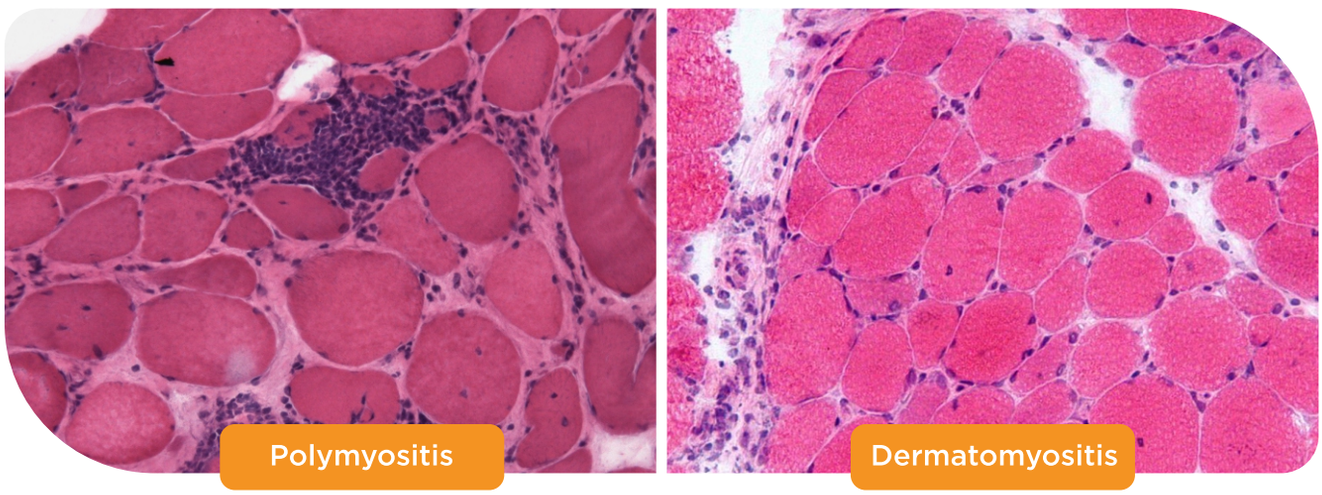
Why there is a need for myositis specific antibodies testing?



- Autoantibodies specific for idiopathic inflammatory myopathy (myositis-specific autoantibodies (MSAs)) are clinically useful biomarkers to help in the diagnosis of polymyositis/dermatomyositis (PM/DM).
- Many of these are also associated with a unique clinical subset of PM/DM, making them useful in predicting and monitoring certain clinical manifestations.
- Anti-Mi-2 is a classic marker for DM and is associated with good response to steroid treatment and good prognosis. Anti-SRP is specific for PM and is associated with treatment-resistant myopathy histologically characterized as necrotizing myopathy.
- Also, anti-MJ/nuclear matrix protein 2 (NXP-2) and anti-small ubiquitin-like modifier-1 (SUMO-1) activating enzyme (SAE) are recognized as new DM-specific autoantibodies.

Immunofluorescence ANA Pattern and Autoantibody Specificities in PM/DM

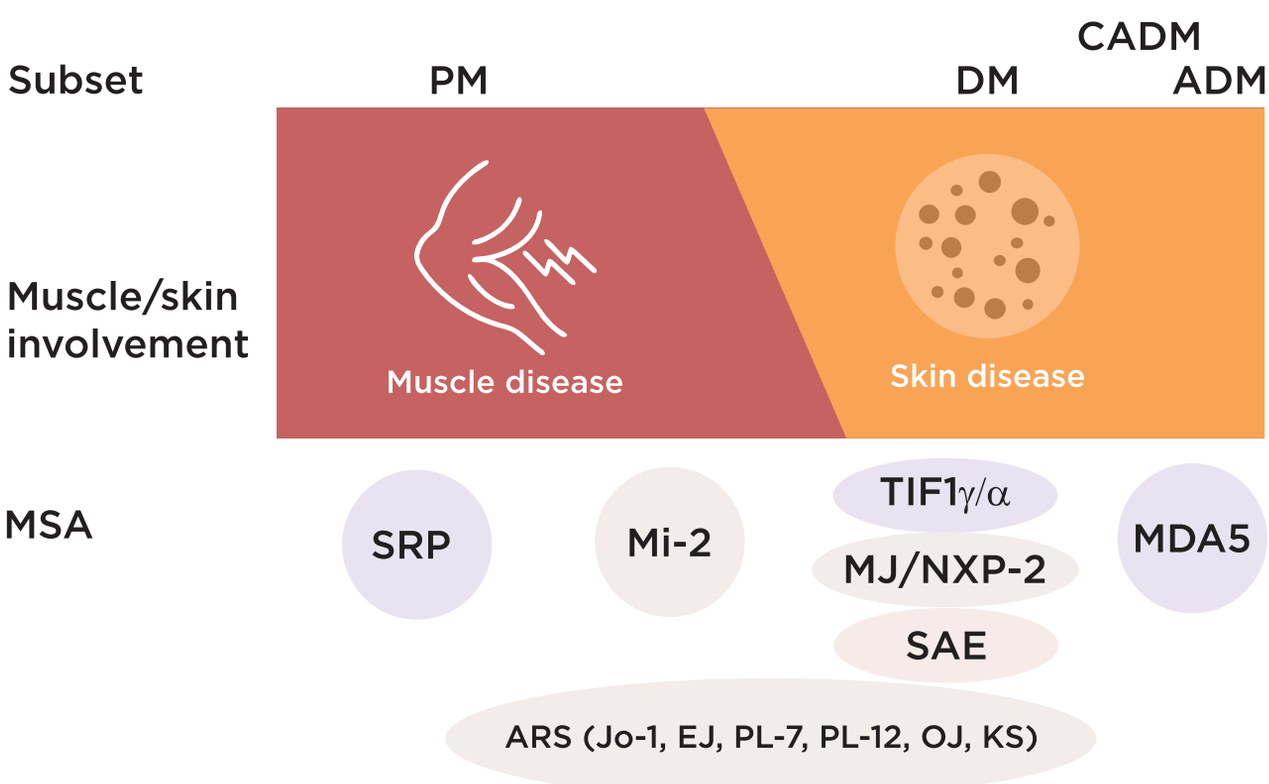


Fig.1.
A summary of the association of myositis-specific autoantibodies with the spectrum of muscle and skin involvements in different subsets of PM/DM

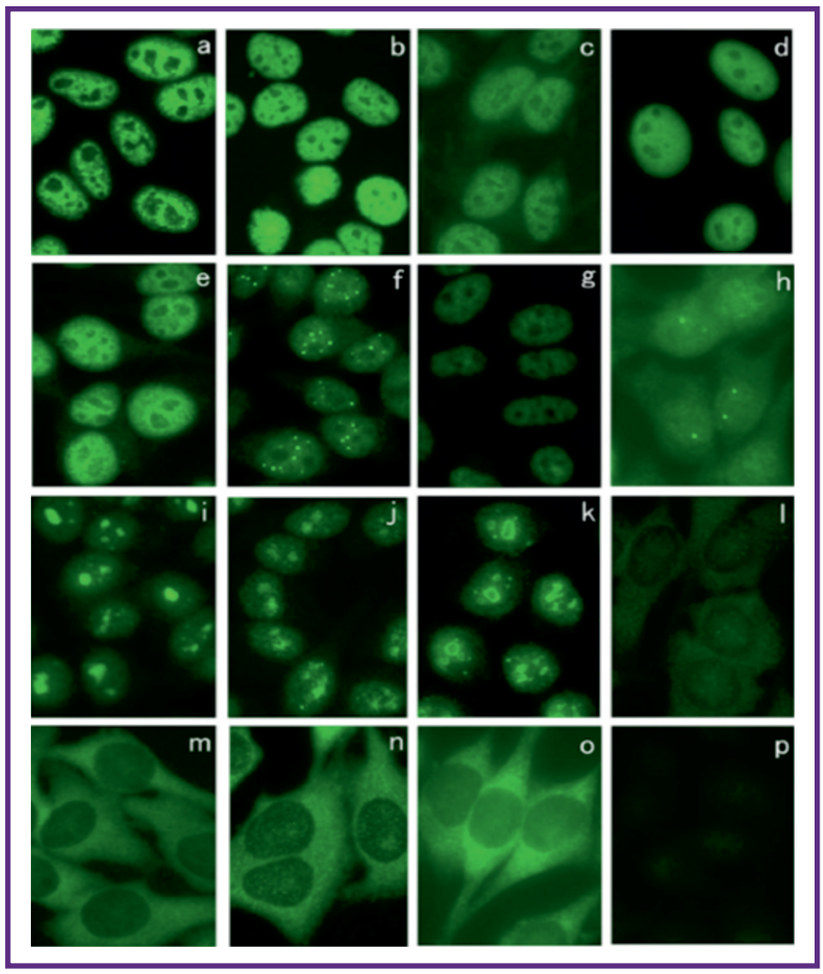


Fig.2.
Immunofluorescence antinuclear antibodies using sera from patients with PM/DM. HEp-2 ANA slides were stained using sera from patients with PM/DM. a Anti-U1RNP, b anti-Mi-2, c anti-TIF1γ/α, d anti-TIFIβ, e anti-SAE, f, g anti-MJ/NXP-2, h anti-SMN, I,j anti-PM-Scl, k anti-U3RNP, 1 anti-Jo-1, m anti-PL-7, n anti-PL-12, m anti-SRP, p anti-MDA5

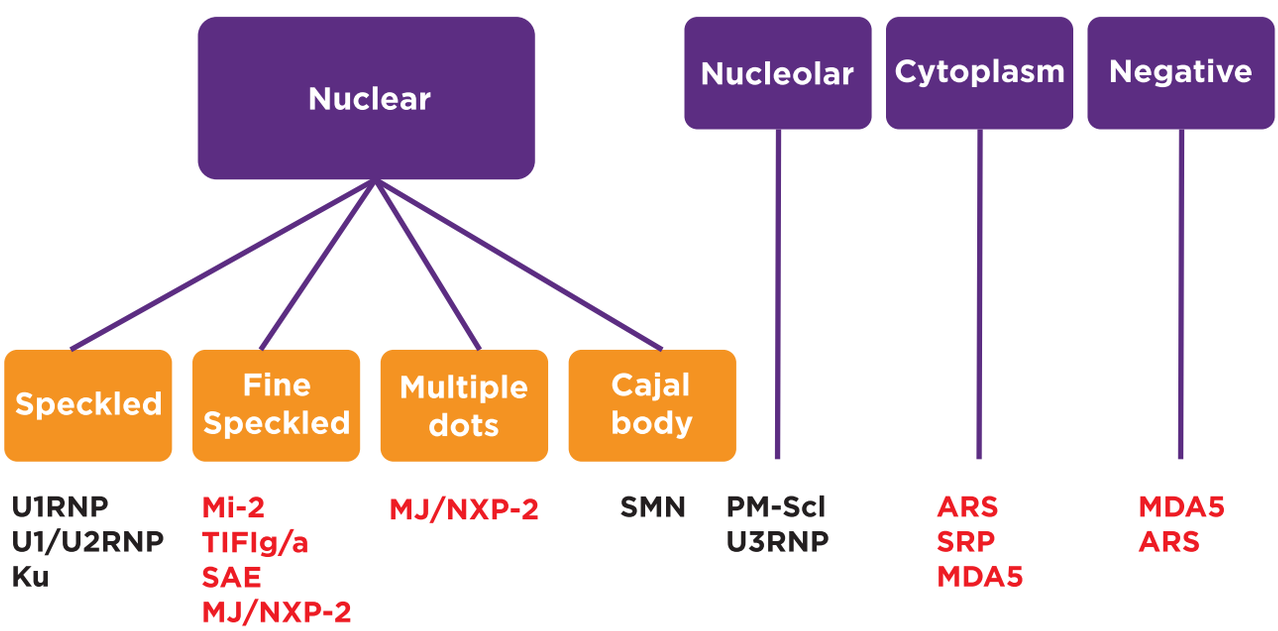


Fig.3.
Summary of HEp-2 cell immunofluorescence patterns corresponding to different autoantibody specificities in PM/DM

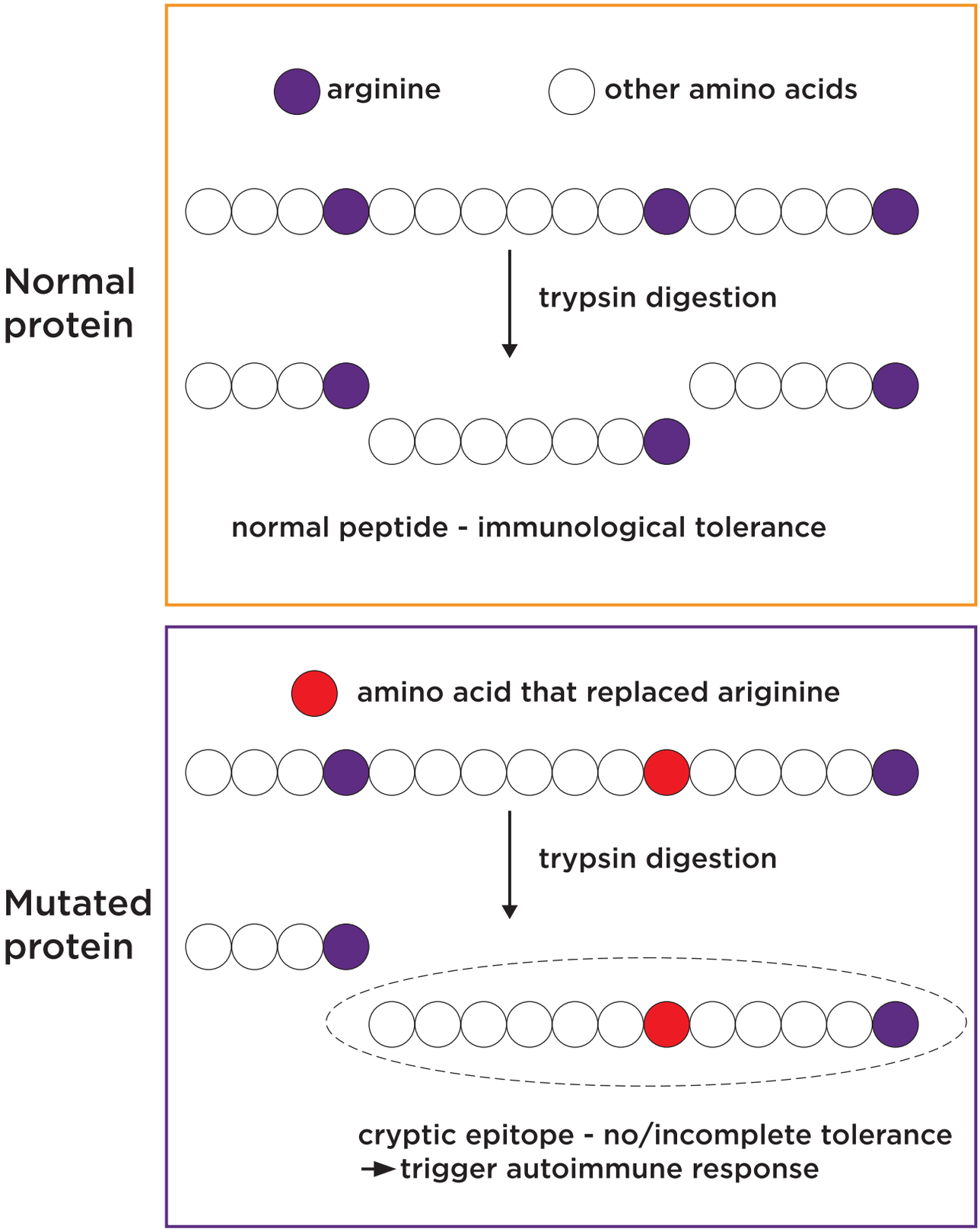


Fig.4.
Formation of cryptic epitopes via a somatic mutation. A somatic mutation that causes amino acid replacement may create cryptic epitopes, which can be recognized as non-self and trigger autoimmune response. Top; normal protein digested by trypsin makes normal peptides that are supposed to have immunological tolerance. Bottom; if ariginine is replaced by other amino acid, trypsin digeation may create cryptic epitopes that have no or incomplete immunological tolerance and trigger autoimmune response

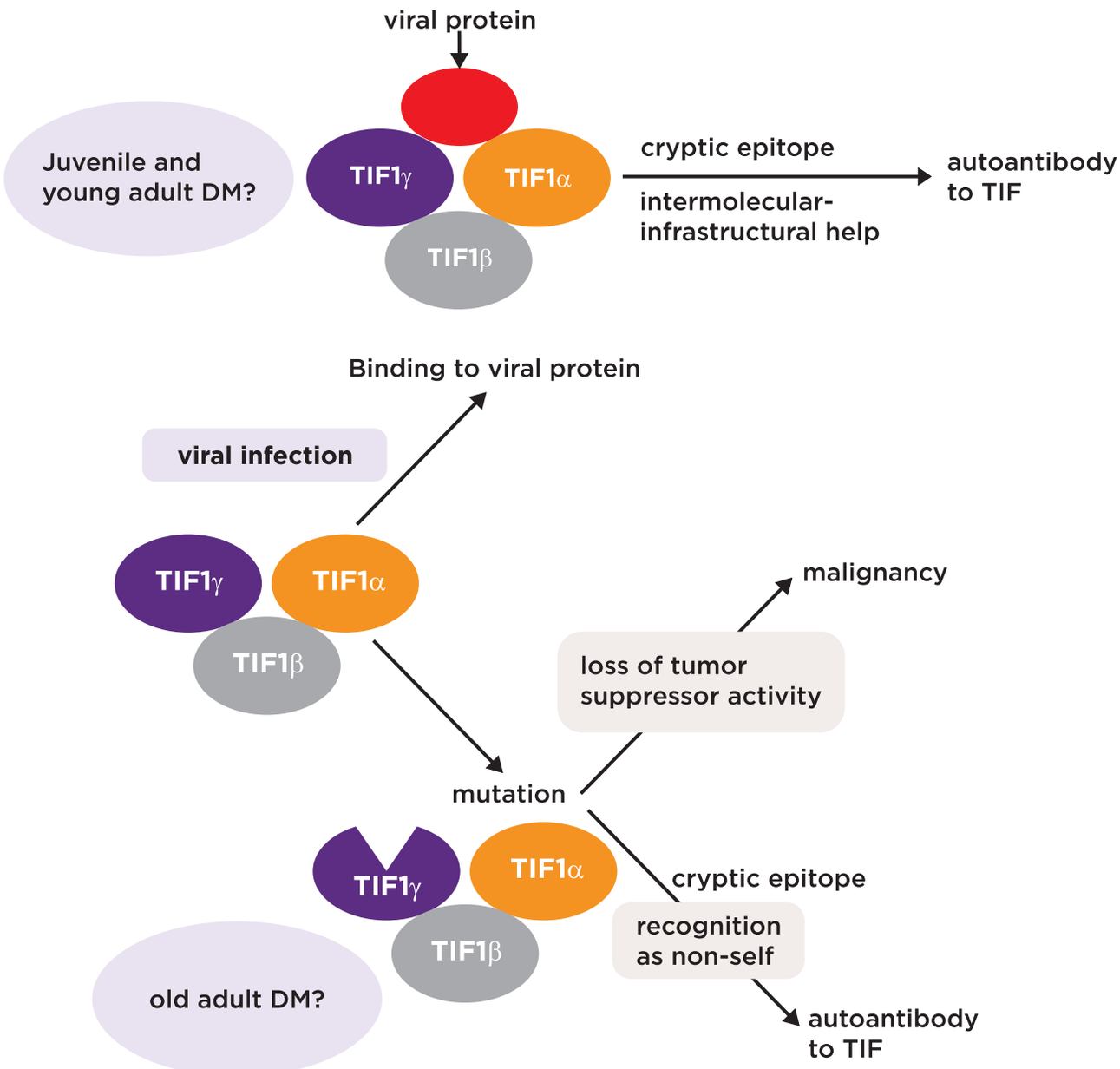


Fig.5.
Hypothesis on the production of anti-TIF1γ/α antibodies based on mutation of TIF1γ or interaction of viral proteins with TIF1. In old adult DM patients with malignancy, TIF1γ mutation may allow development of malignancy while the mutated protein may also trigger autoimmune response to TIF1γ. In JDM or young adult DM patients, interaction of viral proteins with TIF1 proteins may create cryptic epitopes, leading to the autoimmune response

Myositis-specific and myositis-associated autoantibodies

Type of autoantibodies	Myositis-specific antibodies (MSA)	Myositis-associated antibodies (MAA)	Other autoantibodies often found in myositis
Autoantibody specificities	Classic MSA; Jo-1, PL-7, PL-12, EJ, OJ, Mi-2, SRP New antibodies that can be considered MSA: KS, TIF1γ/α, TIF1β, MJ/NXP-2, MDA5/CADM-140, SAE	PM-Scl, Ku, U1RNP, U1/U2RNP, U3RNP	Ro52, Ro60, Su/Ago2
Association with SARD	PM/DM, PM/DM-overlap syndrome	PM/DM, PM/DM - overlap syndrome, Ssc, SLE	Various SARD
Detection in non-PM/DM	Uncommon (anti-ARS can be in overlap syndrome and idiopathic ILD)	Not uncommon	Often
Association with myopathy when found in non-PM/DM	Yes	Yes	No or not established
Prevalence in general population	Almost none	PM- Sci, Ku, U1/U2RNP - almost none; U1RNP, ~0.1%	Relatively common (0.5-1%)

SARD systemtic autoimmune rheumatic diseases, PM polymyositis, DM dermatomyositis, SSc scleroderma, systemic sclerosis, SLE systemic lupus erythematosus, ILD interstitial lung disease

Target autoantigens of myositis-specific autoantibodies

Autoantibodies	Target molecule	Function
Aminoacyl tRNA synthetase		
Jo -1	Histidyl tRNA synthetase	Incorporate histidine into proteins
PL-7	Threonly tRNA synthetase	Incorporate threonine into proteins
PL-12	Alanyl tRNAsynthetase	Alanine and aspartate biosynthesis and alanine incorporation into proteins
EJ	Glycyl tRNA synthetase	Glycine, serine and theonine metabolism, and aminoacyl tRNA biosynthesis
OJ	Isoleucyl tRNA synthetase	Incorporate isoleucine into proteins
KS	Asparaginyl tRNA synthetase	Glutamate, alanine and aspartate metabolism
ZO	Phenylalanyl tRNA synthetase	Incorporate phenylalanine into proteins
YRS (HA)	Tyrosyl tRNA synthetase	Incorporate tyrosine into proteins
SRP	Signal Recognition Particle	Protein maturation in the ribosome
Mi2	Helicase protein	Transcriptional regulation
MDAS (CADM140)	MDA5 (melanoma differentiation- associated gene 5)	RNA- specific helicase that mediates the antiviral response
TIF1 γ/α (p155/140, TRIM33/TRIM24)	TIF1 γ/α	Transcription and RNA metabolism
TIF1 β (TRIM28)	TIF1 β	Transcription and RNA metabolism
MJ/NXP-2	NXP2 (MORC3)	Transcriptional regulation & activation of the tumor suppressor p53
SAE	Small ubiquitin-like modifier 1 (SUMO-1) activating enzyme	Post-translational modifications

Prevalence and clinical association of myositis autoantibodies

Autoantibodies	Prevalence (%)	Disease association	Clinical association/significance
Aminoacyl tRNA synthetases			
Jo -1	15-30	PM, DM	Anti-synthetase syndrome (myositis, ILD, polyarthritis, Raynaud's phenomenon, mechanic's hands)
PL-7	<5	PM, DM	Anti-synthetase syndrome
PL-12	<5	PM, DM, CADM, ILD	Anti-synthetase syndrome, ILD, CADM
EJ	<5	PM, DM	Anti-synthetase syndrome
OJ	<5	PM, DM	Anti-synthetase syndrome, ILD
KS	<1	PM, DM, ILD	ILD
ZO	Rare	-	Myositis
YRS (HA)	Rare	-	Myositis
SRP	5	PM	Myositis (necrotizing)
Mi2	10	DM	DM with typical skin lesions and mild myositis
MDA5/CADM140	15-20	CADM/ADM	CADM, rapidly progressive ILD, severe skin manifestations
TIF1 γ/α	10-15	DM	Malignancy-associated DM
MJ/NXP2	1-5	DM	Adult and jubenile DM with severe skin disease
SAE	1	DM	DM

PM polymyositis, DM dermatomyositis, ILD interstitial lung disease, CADM clinically amyopathic dermatomyositis, ADM amyopathic dermatomyositis

Myositis Panel

(16 antigen)

Mi-2α	Mi-2β	TIF1-γ	MDA-5
NXP2	SAE-1	Ku	PM Scl-100
PM Scl-75	Jo-1	SRP	PL-7
PL-12	EJ	OJ	Ro-52

Sample type: Serum
Specimen Volume: 2 mL
Transportation Instructions: Ambient temperature

Method: Immunoblot (EIA)
TAT: Daily two batches except Sunday (11 am and 3 pm) with TAT of 4 hours

Knock out medical emergencies, before it's too late!

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- ICMR Approved COVID Tests

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