

## Autoantibodies Associated with Myositis

A great proportion of patients with myositis (polymyositis [PM], dermatomyositis [DM], anti-synthetase syndrome and overlap syndromes) have specific antibodies that occur exclusively in these conditions. There are two major types of myositis antibody groups: anti-cytoplasmic abs, of which aminoacyl-tRNA synthetases is one type, and anti-nuclear abs.

Anti-cytoplasmic abs consist of two main groups: Anti-synthetase abs which contain the following abs with similar intracellular functions: anti-Jo-1\*, anti-PL-7, anti-PL-12, anti-OJ & anti-EJ, and a non-synthetase antibody, anti-SRP\* (signal-recognition particle).

Anti-synthetase abs are considered one type of myositis-specific antibodies (MSA). Antibodies to one synthetase do not appear to cross-react with other synthetases, but other autoantibodies may occur with anti-synthetase, such as anti-SSA, anti-SSB or anti-U1RNP. Anti-synthetases have similar clinical features: myositis, interstitial lung disease (ILD), Raynaud's phenomenon, non-erosive arthritis, mechanic's hands, acute onset with fever and is found predominantly affecting young patients. These antibodies are a marker for the anti-synthetase syndrome. Anti-Jo-1 antibodies are found in 20% of adults with myositis, with lesser percentages (1 - 4%) for the other four antibodies (anti-PL-12, anti-PL-7, anti-OJ and anti-EJ). The anti-Jo-1 ab is found in two-thirds of all myositis patients with ILD, appears four to five times more frequently than other anti-synthetases and has a higher rate in PM than DM.

A second type of anti-cytoplasmic antibodies has a much different clinical picture. The anti-SRP is the key antibody which is found almost exclusively in myositis patients, occurring in approximately 4%. The clinical features associated with SRP include: more aggressive PM (found occasionally in DM and inclusion body myositis [IBM]), onset usually occurs between August and January, disease is resistant with poor response to steroids, prognosis for survival is worse than for other myositis, often disease onset is sudden with progressively severe proximal muscle weakness and severe myalgia and arthralgia, cardiac involvement, distal muscle involvement and rare ILD .

Anti-nuclear antibodies are found in myositis patients in larger numbers than anti-cytoplasmic antibodies. Over 60% of myositis patients test positive for anti-nuclear antibodies with a homogeneous, speckled or nucleolar pattern. Anti-Mi-2\* is the only anti-nuclear-associated antibody that is considered a myositis-specific antibody. The clinical associations of Mi-2 are: high rate of classic DM, a good prognosis and good response to therapy, and in some cases, rash is often classic, florid and difficult to treat.

Cancer is more prevalent in dermatomyositis and much less notable in polymyositis, but is seen.

Anti-PM/Scl\*\*, anti-Ku\*\* and anti-U2 snRNP\*\* antibodies are considered myositis-associated antibodies (MAS). The clinical features related to PM/ScI are: close association with myositis-scleroderma overlap syndrome with 25% positivity, found in 8% of all patients with myositis and most cases have had limited SSc with occasional diffuse SSc. The clinical associations of anti-Ku are: polymyositis-scleroderma overlap and other connective tissue overlaps but can be found in myositis or scleroderma alone and may be found in SLE with anti-SM. The anti-U2 snRNP is found in myositis-scleroderma overlap syndrome and in either condition alone.

\*Myositis-specific antibodies

\*\*Myositis-associated antibodies

## Myositis-Specific Abs, Myositis-Associated Abs and Their Clinical Associations

Antibody	Myositis-Specific Abs	Myositis-Associated Abs	PM	DM	Anti-Synthetase Syndrome	Overlap Syndrome	Myositis (Juvenile)	ILD	Arthritis
<b>Mi-2*</b>	4-14%	Not Applicable	Rare	Common	Not Applicable	Common	10%	Rare	Occas
<b>SRP*</b>	4%	Not Applicable	Common	Occas	Not Applicable	Not Described	Uncommon	Common	Common
<b>Jo-1*</b>	20%	Not Applicable	Common	Occas	Marker	Common	Reported	Common	Common
PL-7	1-4%	Not Applicable	Occas	Common	Marker	Common	Reported	Common	Common
PL-12	1-4%	Not Applicable	Occas	Common	Marker	Common	Reported	Common	Common
OJ	1-4%	Not Applicable	Occas	Common	Marker	Common	Reported	Common	Common
EJ	1-4%	Not Applicable	Occas	Common	Marker	Common	Reported	Common	Common
PM/Scl	Not Applicable	8%	Uncommon	Uncommon	Not Applicable	Common	Occas	Rare	Common
Ku	Not Applicable	% Unknown	Not Described	Not Described	Not Applicable	Common	Occas	Not Described	% Unknown
U2 snRNP	Not Applicable	% Unknown	Not Described	Not Described	Not Applicable	Common	Not Described	Not Described	% Unknown

**\* Mi-2, SRP and Jo-1 are most common and serve as markers for myositis.**