Myositis Profile Interpretation:

Autoantibodies of immunoglobulin class IgG against the 16 most important and most relevant of the currently known myositis-specific and myositis-associated DM/PM and overlap antigens can be detected in serum.

Mi-2

Autoantibodies against Mi-2 (against nuclear helicase) with isoforms Mi-2α (CHD3) and Mi-2β (CHD4) have a high specificity of around 95% for myositides, especially for DM with hypertrophia of the nail folds. These antibodies can be found in 15% to 30% of patients with DM. Antibodies against Mi-2 can also be detected in 8% to 12% of patients with idiopathic myositis. Some patients with anti-Mi-2 antibodies have polymyositis and, in rare cases, also inclusion body myositis. Anti-Mi-2 antibodies can usually be serologically detected in the early stage of disease. In these cases the DM (including in adolescents) often has a favorable course. However, Mi-2-positive (predominantly Mi-2β-positive) DM may also be associated with neoplasia (e.g. colon or breast carcinoma).

Mi-2α

Autoantibodies against Mi-2α, one of two isoforms of Mi-2, have largely the same serodiagnostic significance as autoantibodies against Mi-2, with a prevalence in DM of around 20%.

Mi-2β

Autoantibodies against Mi-2β (main isoform of Mi-2) are serologically detected more frequently in DM associated with neoplasia (e.g. colon or breast carcinoma).

TIF1g

TIF1-gamma autoantibodies are detected in around 15% of DM patients and only in these persons. Hence, the detection of anti-TIF1-gamma antibodies is definitive for DM. In around 58% of anti-TIF1-gamma-positive patients, DM is associated with a malignant disease (e.g. pancreatic carcinoma).

MDA5

Autoantibodies against MDA5 are detected in 13% to 26% of DM patients. They are highly specific for clinically amyopathic DM (95% of these patients are anti-MDA5-positive) or DM combined with interstitial lung disease.

NXP2

Autoantibodies against NXP2 are detected in 18% to 25% of cases of juvenile PM/DM (JDM) and in only around 1% of adult cases. This form of PM/DM is characterized by accompanying calcinosis and particularly severe and chronic disease courses. In adults the disease may be carcinoma-associated (breast, uterine or pancreatic carcinoma).

SAE (SAE1 / SAE2)

Anti-SAE1 antibodies are highly specific markers for DM in 8% of cases and for adult DM associated with interstitial lung diseases (ILD) in 5% of cases.

Ku

Autoantibodies against Ku (DNA-binding, non-histone protein) were originally described in polymyositis-scleroderma overlap syndrome. Since then, anti-Ku antibodies have been detected in other autoimmune diseases with varying frequencies (also depending on ethnic origin). They occur with a prevalence of up to 10% in systemic lupus erythematosus (SLE), a systemic autoimmune disease belonging to the group of collagenoses, which predominantly manifests with the so-called butterfly rash. 40% of patients with antibodies against Ku show symptoms of myositis or systemic sclerosis (SSc), a chronic autoimmune disease with fibrosis of the skin (scleroderma) on the joints and of inner organs such as the esophagus, lungs, heart and kidneys. Autoantibodies against Ku can also occur in Sjögren's syndrome.

PM-Scl100/PM-Scl75

Autoantibodies against the two main antigen-protein components PM-Scl100 and PM-Scl75 are classified by molecular masses. Anti-PM-Scl antibodies (antibodies against PM-Scl75 and PM-Scl100) are detected in 50% to 70% of patients with a so-called overlap syndrome. This combines the symptoms of polymyositis, dermatomyositis and systemic sclerosis (SSc). Patients with SSc exhibit mainly antibodies against PM-Scl75. Antibodies against PM-Scl75 can be detected in 3% of polymyositis cases, in 2% to 3% of patients with systemic sclerosis (SSc) and in 24% to 50% of patients with overlap syndrome. With tests that detect only anti-PM-Scl-100, the majority of SSc patients will be missed. There is no correlation between the antibody concentration and the disease activity. Due to the strong association between anti-PM-Scl antibodies and HLA class II alleles, these autoantibodies are detected exclusively in patients of Caucasian origin.

Jo-1
Autoantibodies against Jo-1 are found in polymyositis with a prevalence of 25% to 55% and a specificity of almost 100%.

SRP
Autoantibodies against SRP can be detected in around 5% of cases of polymyositis (at a specificity of around 90%). Anti-SRP antibodies are a marker for necrotising myopathy (anti-SRP syndrome). The symptoms are acute, severe, proximal, symmetrical skeletal muscle weakness, and pain in muscles, including the heart muscle. Extramuscular signs of the disease can be interstitial lung diseases.

PL-7
Autoantibodies against PL-7 occur with a prevalence of around 3% to up to 6% in patients with myositis.

PL-12
Autoantibodies against PL-12 are detected with a prevalence of up to 3% in myositis patients.

EJ
Autoantibodies against EJ are a diagnostic marker for polymyositis, occurring with a prevalence of 1% to 3%.

OJ
Autoantibodies against OJ are associated with (poly) myositis (prevalence 3%).

Ro-52
Antibodies against Ro-52 are detected in myositis patients with a prevalence of 25%. Anti-Ro-52 also occurs in some rheumatic and non-rheumatic diseases. Anti-Ro-52 autoantibodies appear to play an important role in neonatal lupus and congenital heart block. In this case, certain epitopes are probably associated with the complications during pregnancy.

cN-1A (Mup44, cN1A, NT5C1A, NT5c1A, NT5C1a, sporadic inclusion body myositis autoantigen)
The detection of anti-cN-1A antibodies enables diagnosis of the rare inclusion body myositis (IBM), a degenerative autoimmune disease of the muscles. It is delimited from the sporadically occurring, hereditary, non-inflammatory form of inclusion body myositis. Anti-cN-1A antibodies destroy structures of the muscle cells/fibers and cause inflammatory reactions with infiltration by cytotoxic T-cells. The complex pathogenesis encompasses also degenerative mechanisms. IBM is the most common chronic-inflammatory myopathy in older patients. It leads to muscle weakness and muscle atrophy of muscles near and distant of the trunk. Anti-cN-1A-positive IBM patients show especially severe courses of this autoimmune disease and increased motor impairment including the eye, facial and respiratory muscles. Prevalence of anti-cN-1A antibodies for IBM amount to 33% and up to 76%, for polymyositis 0% to 14%, for dermatomyositis 0% to 21%, for Sjögren syndrome 0% to 23% and for systemic lupus erythematosus 0% to 14% depending on the study. **While autoantibodies against cN-1A for IBM are considered diagnostic, they are clinically irrelevant for other diseases. Also, anti-cN-1A antibodies are found in 5% of healthy persons.**