DIAGNOSTIC CRITERIA

IDIOPATHIC INFLAMMATORY MYOPATHIES (2017 ACR/EULAR)

INTRODUCTION:

Idiopathic inflammatory myopathies (IIM), collectively known as myositis, are heterogeneous disorders characterized by muscle weakness and muscle inflammation. The most common subgroups in adults are dermatomyositis (DM), polymyositis (PM) and inclusion body myositis (IBM), and in children, juvenile DM (JDM). This criteria set has been approved by the European League Against Rheumatism (EULAR) Executive Committee and the American College of Rheumatology (ACR) Board of Directors as Provisional. This signifies that the criteria set has been quantitatively validated using patient data, but it has not undergone full validation based on an independent dataset, using both cases and controls. This validation step is still needed before the criteria are fully validated.

- Sensitivity with biopsies: 93%. Sensitivity without biopsies: 87%
- Specificity with biopsies: 88% Specificity without biopsies: 82%

REFERENCE VALUES:

- Not classified as IIM: score of < 5.3 (< 6.5 with muscle biopsy)
- Possible IIM: score of 5.3-5.49 (6.5 - 6.69 with muscle biopsy)
- Probable IIM: score of 5.5 - 7.49 (6.7 - 8.69 with muscle biopsy)
- Definite IIM: score of ≥ 7.5 (≥ 8.7 with muscle biopsy)

Age of onset	Points
Age of onset of the first symptom assumed to be related to the disease (years)	
≤ 17 years old	0
18-39 years old	1.3
≥ 40 years old	2.1

Muscle weakness	Points
Objective symmetric weakness, usually progressive, of the proximal upper extremities Weakness of proximal upper extremities as defined by manual muscle testing or other objective straight testing, which is present on both sides and is usually progressive over time.	0.7
Objective symmetric weakness, usually progressive, of the proximal lower extremities Weakness of proximal lower extremities as defined by manual muscle testing or other objective straight testing, which is present on both sides and is usually progressive over time.	0.8
Neck flexors are relative weaker than neck extensors Muscle grades for neck flexors are relative lower than neck extensors as defined by manual muscle testing or other objective strength testing.	1.9
In the legs, proximal muscles are relative weaker than distal muscles Muscle grades for proximal muscles in the legs are relative lower than distal muscles in the legs as defined by manual muscle testing or other objective strength testing.	0.9

Skin manifestations	Points
Heliotrope rash Purple, lilac-lilac-colored, or erythematous patches over the eyelids or in a periorbital distribution, often associated with periorbital edema.	3.1
Gottron's papules Erythematous to violaceous papules over the extensor surfaces of joints, which are sometimes scaly. May occur over the finger joints, elbows, knees, malleoli, and toes.	2.1
Gottron's sign Erythematous to violaceous macules over the extensor surfaces of joints, which are not palpable.	3.3

Other clinical manifestations	Points
Dysphagia or esophageal dysmotility Difficulty in swallowing or objective evidence of abnormal motility of the esophagus.	0.7

Laboratory measurements	Points
Anti-Jo-1 (anti-hystidyl-transfer RNA synthetase) autoantibody present Autoantibody testing in serum performed with standardized and validated test, showing positive result.	3.9
Elevated serum levels of creatine kinase (CK) or lactate dehydrogenase (LDH) or aspartate aminotransferase (ASAT/AST/SGOT) or alanine aminotransferase (ALAT/ALT/SGPT) The most abnormal test values during the disease course (highest absolute level of enzyme) above the relevant upper limit of normal.	1.3

Muscle biopsy features-presence of:	Points
Was muscle biopsy performed?	
Endomysial infiltration of mononuclear cells surrounding, but not invading, myofibers Muscle biopsy reveals endomysial mononuclear cells abutting the sarcolemma of otherwise healthy, non-necrotic fibers, but there is no clear invasion of the muscle fibers.	1.7
Perimysial and/or perivascular infiltration of mononuclear cells Mononuclear cells are located in the perrimysium and/or located around blood vessels (in either perimyseal or endomysial vessels)	1.2
Perifascicular atrophy Muscle biopsy reveals several rows of muscle fibers, which are smaller in the perifascicular region than fibers more centrally located.	1.9
Rimmed vacuoles Rimmed vacuoles are bluish by hematoxylin and eosin staining and reddish by modified Gomori trichrome stain.	3.1

REFERENCES:

Lundberg IE, Tjärnlund, Bottai M, et al. 2017 European League Against Rheumatism/ American College of Rheumatology classification criteria for adult and juvenile idiopathic inflammatory myopathies and their major subgroups. Ann Rheum Dis 2017;76: 2955-1964.