Idiopathic Inflammatory Myopathies Classification Criteria

Weighted probability-score criteria for adult/juvenile IIM, with definite/probable/possible thresholds and a subgroup classification tree.

2017 EULAR/ACR Criteria
Classification Criteria for Adult & Juvenile IIM
Lundberg IE, et al. · Arthritis Rheumatol 2017;69:2271–2282

FORMAT *Applies when no better explanation for symptoms/signs exists. Score points shown without biopsy / with biopsy.*

1 Age of Onset & Muscle Weakness

ITEM	POINTS
Onset 18–39 yr	Age of first related symptom `1.3 / 1.5`
Onset ≥40 yr	`2.1 / 2.2`
Proximal UE weakness	Symmetric, usually progressive `0.7 / 0.7`
Proximal LE weakness	Symmetric, usually progressive `0.8 / 0.5`
Neck flexors < extensors	`1.9 / 1.6`
Leg proximal < distal	`0.9 / 1.2`

2 Skin & Other Clinical Manifestations

ITEM	POINTS
Heliotrope rash	Purple/lilac periorbital patches ± edema `3.1 / 3.2`
Gottron's papules	Papules over extensor joint surfaces `2.1 / 2.7`
Gottron's sign	Non-palpable extensor macules `3.3 / 3.7`
Dysphagia/esophageal dysmotility	`0.7 / 0.6`

3 Laboratory & Muscle Biopsy

ITEM	POINTS
Anti-Jo-1 positive	`3.9 / 3.8`
↑ CK, LDH, AST, or ALT	Highest abnormal value `1.3 / 1.4`
Endomysial mononuclear infiltrate	Surrounding, non-invasive `biopsy only: 1.7`
Perimysial/perivascular infiltrate	`biopsy only: 1.2`
Perifascicular atrophy	`biopsy only: 1.9`
Rimmed vacuoles	`biopsy only: 3.1`

4 Probability Thresholds & Scoring

Definite IIM — probability ≥90%: score ≥7.5 (no biopsy) or ≥8.7 (with biopsy).

Probable IIM — probability ≥55%: score ≥5.5 (no biopsy) or ≥6.7 (with biopsy).

Possible IIM — probability 50–<55%.

Sum applicable item scores and convert to probability via the published nomogram, or use the online calculator (imm.ki.se/biostatistics/calculators/iim).

5 Subgroup Classification Tree

Adult vs. juvenile — age at onset ≥18 yr = adult; <18 yr = juvenile.

Juvenile + skin rash → Juvenile dermatomyositis (JDM).

Adult + skin findings → Dermatomyositis (DM), or amyopathic DM (ADM) if no significant weakness.

Adult, no skin findings — finger flexor weakness with no treatment response, or biopsy rimmed vacuoles → Inclusion body myositis (IBM); otherwise → Polymyositis (PM, includes IMNM).

Lundberg IE, Tjärnlund A, 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. *Arthritis Rheumatol.* 2017;69(12):2271–2282.