Subject Name:_____ Study Subject ID:__ __ __ __ __
Visit Date (MM/DD/YYYY): ____ / ____ / ____ INTERVAL (1-4):__

# 2017 European League Against Rheumatism/American College of Rheumatology Classification Criteria for Adult and Juvenile Idiopathic Inflammatory Myopathies

| When no better explanation for the symptoms and signs exist, these classification criteria can be used | | | | |
|---|---|---|---|---|
| 1. Was a muscle biopsy performed? | | ☐No | ☐Yes | |
| **CLASSIFICATION CRITERIA** | **Present in this patient?** | **Score Without Muscle Biopsy** | **Score With Muscle Biopsy** | **Points Assigned** |
| 2. Age of onset of first symptom assumed to be related to the disease ≥ 18 years and < 40 years | ☐Yes<br>☐No<br>☐Not Assessed | 1.3 | 1.5 | |
| 3. Age of onset of first symptom assumed to be related to the disease ≥ 40 years | ☐Yes<br>☐No<br>☐Not Assessed | 2.1 | 2.2 | |
| **MUSCLE WEAKNESS** | **Present in this patient?** | **Score Without Muscle Biopsy** | **Score With Muscle Biopsy** | **Points Assigned** |
| 4. Objective symmetric weakness, usually progressive, of the proximal upper extremities | ☐Yes<br>☐No<br>☐Not Assessed | 0.7 | 0.7 | |
| 5. Objective symmetric weakness, usually progressive, of the proximal lower extremities | ☐Yes<br>☐No<br>☐Not Assessed | 0.8 | 0.5 | |
| 6. Neck flexors are relatively weaker than neck extensors | ☐Yes<br>☐No<br>☐Not Assessed | 1.9 | 1.6 | |
| 7. In the legs proximal muscles are relatively weaker than distal muscles | ☐Yes<br>☐No<br>☐Not Assessed | 0.9 | 1.2 | |
| **SKIN MANIFESTATIONS** | **Present in this patient?** | **Score Without Muscle Biopsy** | **Score With Muscle Biopsy** | **Points Assigned** |
| 8. Heliotrope rash | ☐Yes<br>☐No<br>☐Not Assessed | 3.1 | 3.2 | |
| 9. Gottron´s papules | ☐Yes<br>☐No<br>☐Not Assessed | 2.1 | 2.7 | |
| 10. Gottron's sign | ☐Yes<br>☐No<br>☐Not Assessed | 3.3 | 3.7 | |

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| OTHER CLINICAL MANIFESTATIONS | Present in this patient? | Score Without Muscle Biopsy | Score With Muscle Biopsy | Points Assigned |
|---|---|---|---|---|
| 11. Dysphagia or esophageal dysmotility | ☐Yes<br>☐No<br>☐Not Assessed | 0.7 | 0.6 | |
| **LABORATORY MEASUREMENTS** | **Present in this patient?** | **Score Without Muscle Biopsy** | **Score With Muscle Biopsy** | **Points Assigned** |
| 12. Anti-Jo-1 (anti-histidyl-tRNA synthetase) autoantibody present | ☐Yes<br>☐No<br>☐Not Assessed | 3.9 | 3.8 | |
| 13. Elevated serum levels of creatine kinase (CK)* ***or*** lactate dehydrogenase (LDH)* ***or*** aspartate aminotransferase (ASAT/AST/SGOT)* ***or*** alanine aminotransferase (ALAT/ALT/SGPT)* | ☐Yes<br>☐No<br>☐Not Assessed | 1.3 | 1.4 | |
| **MUSCLE BIOPSY FEATURES- PRESENCE OF:** | **Present in this patient?** | **Score Without Muscle Biopsy** | **Score With Muscle Biopsy** | **Points Assigned** |
| 14. Endomysial infiltration of mononuclear cells surrounding, but not invading, myofibres | ☐Yes<br>☐No<br>☐Not Assessed | NA | 1.7 | |
| 15. Perimysial and/or perivascular infiltration of mononuclear cells | ☐Yes<br>☐No<br>☐Not Assessed | NA | 1.2 | |
| 16. Perifascicular atrophy | ☐Yes<br>☐No<br>☐Not Assessed | NA | 1.9 | |
| 17. Rimmed vacuoles | ☐Yes<br>☐No<br>☐Not Assessed | NA | 3.1 | |
| | | | **TOTAL SCORE** | |

* Serum levels above the upper limit of normal

**Probable IIM:** Total score ≥ 5.5, total score ≥ 6.7 if biopsy data is available.
**Definite IIM:** Total score ≥ 7.5 or more without muscle biopsy and ≥ 8.7 with muscle biopsy.
**Possible IIM:** Total score ≥ 5.3, total score ≥ 6.5 if biopsy data is available

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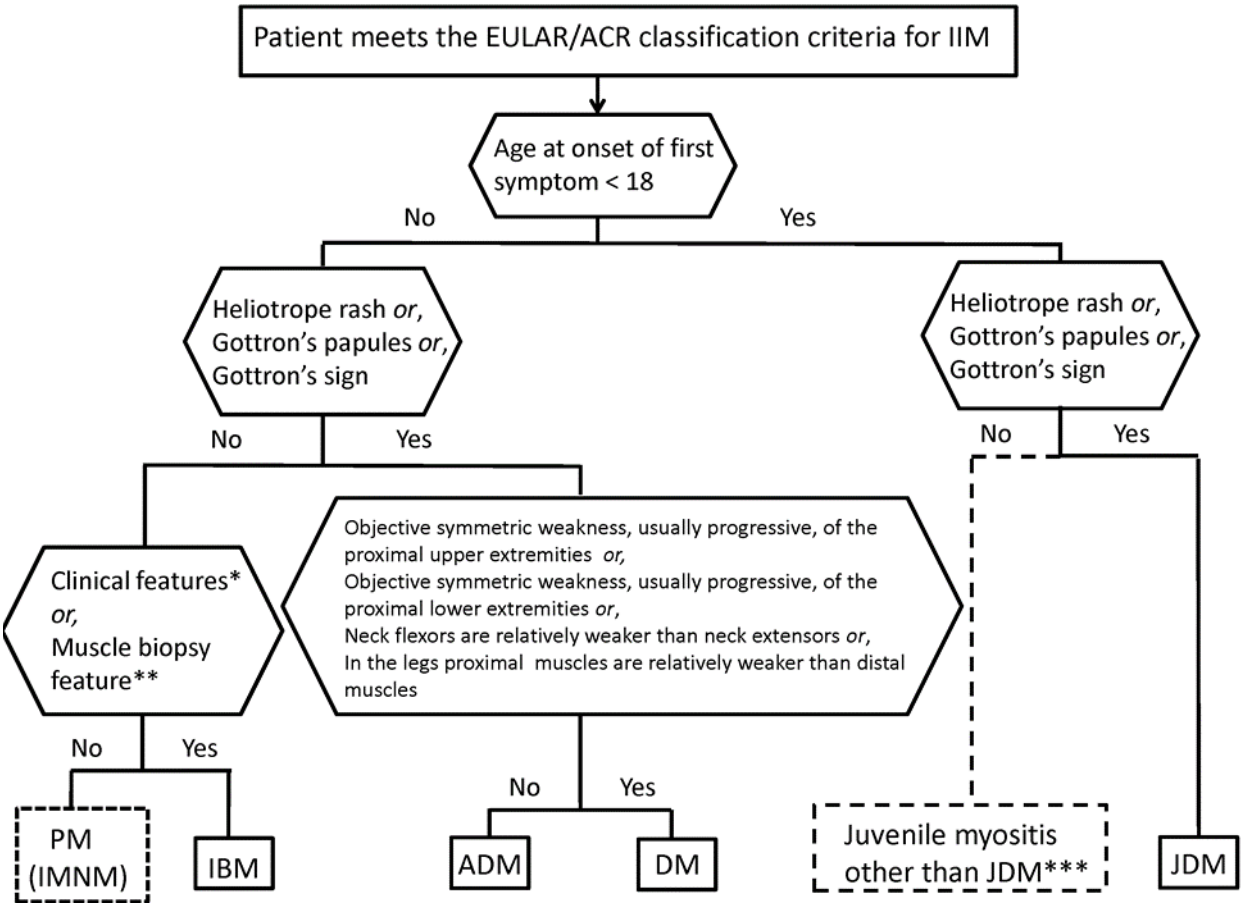
**Sub-classification:** Meet probable or definite IIM criteria + the following based on classification tree:
**Dermatomyositis (DM):** Heliotrope or Gottron's papules or Gottron's sign with any 1 pattern of weakness.
**Amyopathic DM:** any 1 of the characteristic DM rashes, without weakness
**Polymyositis (PM) (or Immune-mediated necrotizing myopathy, IMNM):** no characteristic rashes, requires muscle biopsy
**Inclusion body myositis (IBM):** No characteristic rashes. One of these 2 are required for diagnosis: Finger flexor weakness and response to treatment: not improved, *or* Muscle biopsy: rimmed vacuoles
**JDM:** Age onset < 18 years, 1 of the characteristic DM rashes
**Juvenile Myositis other than JDM:** onset <18yo without 1 of the characteristic DM rashes

| **SUBCLASSIFICATION CRITERIA** | |
|---|---|
| 1. Age of onset of first symptom assumed to be related to the disease < 18 years | ☐Yes ☐No |
| 2. Heliotrope rash *or*<br>Gottron's papules *or*<br>Gottron's sign | ☐Yes ☐No<br>☐Yes ☐No<br>☐Yes ☐No |
| 3. Objective symmetric weakness, usually progressive, of the proximal upper extremities, *or*<br>Objective symmetric weakness, usually progressive, of the proximal lower extremities, *or*<br>Neck flexors are relatively weaker than neck extensors *or*<br>In the legs proximal muscles are relatively weaker than distal muscles | ☐Yes ☐No<br>☐Yes ☐No<br>☐Yes ☐No<br>☐Yes ☐No |
| *4.* A. Finger Flexor weakness, *and*<br>B. Response to Immunosuppressive Treatment: Not improved | ☐Yes ☐No<br>☐Yes ☐No |
| 5. Muscle biopsy: rimmed vacuoles | ☐Yes ☐No |
| **SUBCLASSIFICATION: (select one)**<br>**☐PM (IMNM) ☐IBM ☐DM ☐Amyopathic DM**<br>**☐JDM ☐Juvenile myositis other than JDM** | |

# IIM Sub-classification Criteria Tree




***:** finger flexor weakness and response to treatment: not improved

****:** rimmed vacuoles

*******: Juvenile myositis other than juvenile dermatomyositis (JDM) was developed based on expert opinion.

## Definitions of the Variables

**Muscle weakness**

**Objective symmetric weakness, usually progressive, of the proximal upper extremities:** Weakness of proximal upper extremities as defined by manual muscle testing or other objective strength testing, which is present on both sides and is usually progressive over time

**Objective symmetric weakness, usually progressive, of the proximal lower extremities:** Weakness of proximal lower extremities as defined by manual muscle testing or other objective strength testing, which is present on both sides and is usually progressive over time

**Neck flexors are relatively weaker than neck extensors:** Muscle grades for neck flexors are relatively lower than neck extensors as defined by manual muscle testing or other objective strength testing

**In the legs, proximal muscles are relatively weaker than distal muscles:** Muscle grades for proximal muscles in the legs are relatively lower than distal muscles in the legs as defined by manual muscle testing or other objective strength testing

**Skin manifestations**

**Heliotrope rash**: Purple, lilac-colored or erythematous patches over the eyelids or in a periorbital distribution, often associated with periorbital edema

**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

**Gottron's sign:** Erythematous to violaceous macules over the extensor surfaces of joints, which are not palpable

**Other clinical manifestations**

**Dysphagia or esophageal dysmotility:** Difficulty in swallowing or objective evidence of abnormal motility of the esophagus

**Laboratory measurements**

**Anti-Jo-1 (anti-histidyl-tRNA synthetase) autoantibody present:** Autoantibody testing in serum performed with standardized and validated test, showing positive result

**Elevated serum levels of creatine kinase (CK) *or* lactate dehydrogenase (LD) *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

**Muscle biopsy features–presence of:**

**Endomysial infiltration of mononuclear cells surrounding, but not invading, myofibres:**
Muscle biopsy reveals endomysial mononuclear cells abutting the sarcolemma of otherwise healthy, non-necrotic muscle fibres, but there is no clear invasion of the muscle fibres

**Perimysial and/or perivascular infiltration of mononuclear cells:**
Mononuclear cells are located in the perimysium and/or located around blood vessels (in either perimysial or endomysial vessels)
**Perifascicular atrophy:** Muscle biopsy reveals several rows of muscle fibres, which are smaller in the perifascicular region than fibres more centrally located
**Rimmed vacuoles:** Rimmed vacuoles are bluish by H&E staining and reddish by modified Gomori trichrome stains.

## References

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