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 ☐ No ☐ Not Assessed	1.3	1.5	
3. Age of onset of first symptom assumed to be related to the disease ≥ 40 years	☐ Yes ☐ No ☐ 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 ☐ No ☐ Not Assessed	0.7	0.7	
5. Objective symmetric weakness, usually progressive, of the proximal lower extremities	☐ Yes ☐ No ☐ Not Assessed	0.8	0.5	
6. Neck flexors are relatively weaker than neck extensors	☐ Yes ☐ No ☐ Not Assessed	1.9	1.6	
7. In the legs proximal muscles are relatively weaker than distal muscles	☐ Yes ☐ No ☐ 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 ☐ No ☐ Not Assessed	3.1	3.2	
9. Gottron´s papules	☐ Yes ☐ No ☐ Not Assessed	2.1	2.7	
10. Gottron's sign	☐ Yes ☐ No ☐ Not Assessed	3.3	3.7	

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

OTHER CLINICAL MANIFESTATIONS	Present in this patient?	Score Without Muscle Biopsy	Score With Muscle Biopsy	Points Assigned
11. Dysphagia or esophageal dysmotility	☐ Yes ☐ No ☐ 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 ☐ No ☐ 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 ☐ No ☐ 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 ☐ No ☐ Not Assessed	NA	1.7	
15. Perimysial and/or perivascular infiltration of mononuclear cells	☐ Yes ☐ No ☐ Not Assessed	NA	1.2	
16. Perifascicular atrophy	☐ Yes ☐ No ☐ Not Assessed	NA	1.9	
17. Rimmed vacuoles	☐ Yes ☐ No ☐ Not Assessed	NA	3.1	
			TOTAL SCORE	

* Serum levels above the upper limit of normal

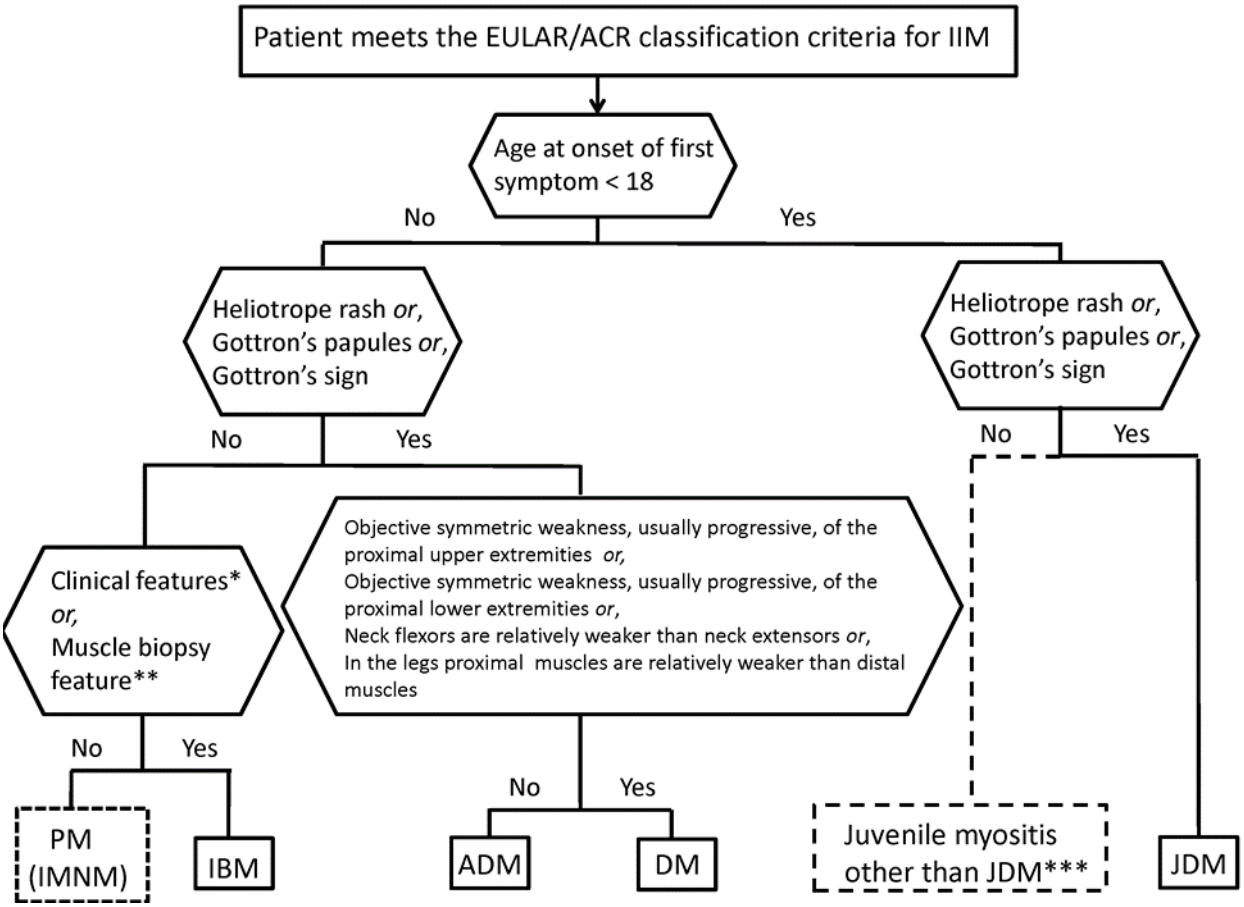
Definite IIM: Total score ≥ 7.5 or more without muscle biopsy and ≥ 8.7 with muscle biopsy.
Probable IIM: Total score ≥ 5.5, total score ≥ 6.7 if biopsy data is available.
Possible IIM: Total score ≥ 5.3, total score ≥ 6.5 if biopsy data is available
Does Not Meet Criteria for IIM: Total score < 5.3, total score < 6.5 if biopsy data is available

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

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 ☐ N/A
2. Heliotrope rash *or* Gottron's papules *or* Gottron's sign	☐Yes ☐No ☐ N/A ☐Yes ☐No ☐ N/A ☐Yes ☐No ☐ N/A
3. Objective symmetric weakness, usually progressive, of the proximal upper extremities, *or* Objective symmetric weakness, usually progressive, of the proximal lower extremities, *or* Neck flexors are relatively weaker than neck extensors *or* In the legs proximal muscles are relatively weaker than distal muscles	☐Yes ☐No ☐ N/A ☐Yes ☐No ☐ N/A ☐Yes ☐No ☐ N/A ☐Yes ☐No ☐ N/A
4. A. Finger Flexor weakness, *and* B. Response to Immunosuppressive Treatment: Not improved	☐Yes ☐No ☐ N/A ☐Yes ☐No ☐ N/A
5. Muscle biopsy: rimmed vacuoles	☐Yes ☐No ☐ N/A
SUBCLASSIFICATION: (select one) **☐ PM (IMNM) ☐ IBM ☐ DM ☐ Amyopathic DM** **☐ JDM ☐ Juvenile myositis other than JDM**	
Comment:	

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

Lundberg IE, Tjärnlund A, Bottai M, Werth VP, Pilkington C, Visser M, Alfredsson L, Amato AA, Barohn RJ, Liang MH, Singh JA, Aggarwal R, Arnardottir S, Chinoy H, Cooper RG, Dankó K, Dimachkie MM, Feldman BM, Torre IG, Gordon P, Hayashi T, Katz JD, Kohsaka H, Lachenbruch PA, Lang BA, Li Y, Oddis CV, Olesinska M, Reed AM, Rutkowska-Sak L, Sanner H, Selva-O'Callaghan A, Song YW, Vencovsky J, Ytterberg SR, Miller FW, Rider LG; International Myositis Classification Criteria Project consortium, The Euromyositis register and The Juvenile Dermatomyositis Cohort Biomarker Study and Repository (JDRG) (UK and Ireland). 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 Dec;76(12):1955-1964. doi: 10.1136/annrheumdis-2017-211468.

Lundberg IE, Tjärnlund A, Bottai M, Werth VP, Pilkington C, de Visser M, Alfredsson L, Amato AA, Barohn RJ, Liang MH, Singh JA, Aggarwal R, Arnardottir S, Chinoy H, Cooper RG, Dankó K, Dimachkie MM, Feldman BM, Garcia-De La Torre I, Gordon P, Hayashi T, Katz JD, Kohsaka H, Lachenbruch PA, Lang BA, Li Y, Oddis CV, Olesinska M, Reed AM, Rutkowska-Sak L, Sanner H, Selva-O'Callaghan A, Song YW, Vencovsky J, Ytterberg SR, Miller FW, Rider LG; International Myositis Classification Criteria Project Consortium, the Euromyositis Register, and the Juvenile Dermatomyositis Cohort Biomarker Study and Repository (UK and Ireland). 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 Dec;69(12):2271-2282.

Bottai M, Tjärnlund A, Santoni G, Werth VP, Pilkington C, de Visser M, Alfredsson L, Amato AA, Barohn RJ, Liang MH, Singh JA, Aggarwal R, Arnardottir S, Chinoy H, Cooper RG, Danko K, Dimachkie MM, Feldman BM, García-De La Torre I, Gordon P, Hayashi T, Katz JD, Kohsaka H, Lachenbruch PA, Lang BA, Li Y, Oddis CV, Olesinka M, Reed AM, Rutkowska-Sak L, Sanner H, Selva-O'Callaghan A, Wook Song Y, Vencovsky J, Ytterberg SR, Miller FW, Rider LG, Lundberg IE; International Myositis Classification Criteria Project consortium, the Euromyositis register and the Juvenile Dermatomyositis Cohort Biomarker Study and Repository (JDRG) (UK and Ireland) . EULAR/ACR classification criteria for adult and juvenile idiopathic inflammatory myopathies and their major subgroups: a methodology report. RMD Open. 2017 Nov 14;3(2):e000507. doi: 10.1136/rmdopen-2017-000507. eCollection 2017.