SUPPLEMENTARY MATERIAL

Table S1. International Myositis Classification Criteria Project questionnaire

Table S2. Clustering of the most frequent combinations of items within subgroups

Table S3. Observation frequencies for the International Myositis Classification Criteria Project questionnaire

Table S1. International Myositis Classification Criteria Project questionnaire

Item	**Alternatives**
Have you received approval from your local IRB or ethics committee for participation in this project?	□ Yes □ Exempt □ No
Has your patient been diagnosed with the diagnosis relevant for this study for more than 6 months? (A yes is required, if No select a new case)	□ Yes □ No
Center (name of university or hospital from where data is entered)	
Clinician submitting case	
Case number	
Gender	□ Female □ Male
Age (years) at onset of first symptom assumed to be related to the disease	
Age (years) at diagnosis	
Age (years) at last evaluation	
Ethnicity	□ Caucasian □ Of African descent □ Of Asian descent □ Of Native American descent □ Of Pacific Island descent □ Of Hispanic descent □ Of Mixed descent □ Unknown
Study diagnosis according to the clinician submitting the case	□ Idiopathic inflammatory myopathy (IIM) adults or children □ Not Idiopathic inflammatory myopathy (Not IIM) adults or children
Study diagnosis: Idiopathic Inflammatory Myopathy (IIM) in adults or children	□ Polymyositis □ Dermatomyositis □ Amyopathic dermatomyositis □ Hypomyopathic dermatomyositis □ Inclusion body myositis □ Immune-mediated necrotizing myopathy □ Juvenile dermatomyositis □ Juvenile polymyositis □ Other diagnosis, specify diagnosis below □ Not Idiopathic Inflammatory Myopathy (IIM)
Not Idiopathic Inflammatory Myopathy (Not IIM), adults or children, but in which the diagnosis of idiopathic myositis was considered in the differential diagnosis	□ Becker´s dystrophy □ Duchenne´s dystrophy □ Fascioscapulohumeral dystrophy □ Limb-girdle dystrophy □ Myotonic dystrophy □ Non-inflammatory inclusion body myopathy □ Other dystrophy, specify diagnosis □ Dysferlinopathy □ Acid maltase deficiency □ Allergies □ Bacterial myopathy □ Carnitine deficiency □ Celiac disease □ Crohn's disease □ Cushing syndrome □ Cysticerosis □ Diabetes mellitus □ Drug or toxin associated myopathy, specify diagnosis □ Exogenous steroid myopathy

	□ Familial periodic paralysis □ Fibromyalgia □ Filiarisis □ Glucocorticoid induced myopathy □ Gullain-Barre syndrome □ Hypercalcemia □ Hypereosinophilic syndrome □ Hypersensitivity conditions □ Hyperthyroidism □ Hypocalcemia □ Hypokalemia □ Hypothyroidism □ Immune mediated skin conditions, specify diagnosis below □ Juvenile idiopathic arthritis □ Kearns-Sayre syndrome □ Mc Ardle´s disease □ Metabolic myopathy, specify diagnosis □ Mitochondrial encephalomyopathy, lactic acidosis, stroke (MELAS) □ Mitochondrial myopathy, specify diagnosis □ Mixed connective tissue disease □ Motor neuron diseases, specify diagnosis □ Multiple sclerosis □ Myasthenia gravis □ Myoadenylate deaminase deficiency □ Myoclonic epilepsy, ragged red fibers (MERRF) □ Palmityltransferase deficiency □ Parasitic myopathy □ Phosphofructokinase deficiency □ Psoriasis □ Seborrheic dermatitis □ Statin induced myopathy □ Systemic lupus erythematosus (SLE) □ Systemic sclerosis □ Systemic vasculitis, specify diagnosis below □ Toxoplasmosis □ Trichinosis □ Trypanosoma □ Ulcerative colitis □ Verrucae vulgaris □ Viral myopathy □ Other dermatologic disease, specify diagnosis below □ Other endocrine myopathy, specify diagnosis □ Other infectious myopathy, specify diagnosis □ Other neuromuscular disease, specify diagnosis below □ Other systemic autoimmune disease, specify diagnosis below □ Other diagnosis, specify □ None applicable (Inflammatory Myopathy)
Basis for study diagnosis (check all supporting reasons)	□ Muscle weakness □ Muscle biopsy abnormalities □ Elevated muscle enzymes □ EMG abnormalities □ Rashes □ Skin biopsy □ Autoantibodies □ MRI

	□ Other, please specify
Other diagnoses in this case: (check all that apply)	□ Non applicable □ Systemic sclerosis □ Sjögren´s syndrome □ Mixed connective tissue disease □ Rheumatoid arthritis □ Systemic lupus erythematosus □ Hypothyroidism □ Hyperthyroidism □ Type I diabetes □ Juvenile idiopathic arthritis □ Malignancy □ Other, please specify
Clinical Muscle Variables – present at any time during the disease course	
1M. Objective symmetric weakness, usually progressive, of the proximal upper extremities	□ Present □ Absent □ Information not available □ Comments
2M, Objective shoulder abductor weakness	□ Present □ Absent □ Information not available □ Comments
3M. Objective elbow flexor weakness	□ Present □ Absent □ Information not available □ Comments
4M. Objective elbow extensor weakness	□ Present □ Absent □ Information not available □ Comments
5M. Wrist and finger flexors are relatively weaker than shoulder abductors on the same side	□ Present □ Absent □ Information not available □ Comments
6M. Wrist flexors are relatively weaker than wrist extensors on the same side	□ Present □ Absent □ Information not available □ Comments
7M. Objective finger flexor weakness	□ Present □ Absent □ Information not available □ Comments
8M. Objective symmetric weakness, usually progressive, of the proximal lower extremities	□ Present □ Absent □ Information not available □ Comments
9M. Objective hip flexor weakness	□ Present □ Absent □ Information not available □ Comments
10M. Objective hip abductor weakness	□ Present □ Absent □ Information not available □ Comments
11M. Objective knee extensor weakness	□ Present □ Absent □ Information not available □ Comments
12M. Knee extensors are as weak or relatively weaker	□ Present

than hip girdle muscle on the same side	□ Absent □ Information not available □ Comments
13M. Objective muscle weakness of distal lower extremities	□ Present □ Absent □ Information not available □ Comments
14M. Objective axial weakness	□ Present □ Absent □ Information not available □ Comments
15M. Objective neck flexor weakness	□ Present □ Absent □ Information not available □ Comments
16M. Neck flexors are relatively weaker than neck extensors	□ Present □ Absent □ Information not available □ Comments
17M. In the legs proximal muscles are relatively weaker than distal muscles	□ Present □ Absent □ Information not available □ Comments
18M. In the arms proximal muscles are relatively weaker than distal muscles	□ Present □ Absent □ Information not available □ Comments
19M In the legs distal muscles relatively weaker than proximal muscles	□ Present □ Absent □ Information not available □ Comments
20M In the arms distal muscles are relatively weaker than proximal muscles	□ Present □ Absent □ Information not available □ Comments
21M. Muscle tenderness	□ Present □ Absent □ Information not available □ Comments
22M. Muscle atrophy of distal forearms	□ Present □ Absent □ Information not available □ Comments
23M. Muscle atrophy of thighs	□ Present □ Absent □ Information not available □ Comments
Skin Variables – present at any time during the disease course	
1S. Heliotrope rash	□ Present □ Absent □ Information not available □ Comments
2S. Gottron´s papules	□ Present □ Absent □ Information not available □ Comments
3S. Gottron's sign	□ Present □ Absent □ Information not available

	□ Comments
4S. Erythema of the back of neck and shoulders (Shawl sign)	□ Present □ Absent □ Information not available □ Comments
5S. Erythema of the neck (V-sign)	□ Present □ Absent □ Information not available □ Comments
6S. Periorbital edema	□ Present □ Absent □ Information not available □ Comments
7S. Linear extensor erythema	□ Present □ Absent □ Information not available □ Comments
8S. Calcification	□ Present □ Absent □ Information not available □ Comments
9S. Periungual erythema or nailfold capillary abnormality	□ Present □ Absent □ Information not available □ Comments
10S. Mechanic's hands	□ Present □ Absent □ Information not available □ Comments
11S. Photodistributed violaceous erythema	□ Present □ Absent □ Information not available □ Comments
12 S. Raynaud's phenomenon	□ Present □ Absent □ Information not available □ Comments
13S. Cuticular overgrowth	□ Present □ Absent □ Information not available □ Comments
14S Poikiloderma	□ Present □ Absent □ Information not available □ Comments
Other Clinical Variables – present at any time during the disease course	
1O. Family history of autoimmune disease (see Appendix A)	□ Present □ Absent □ Information not available □ Comments
2O. Family history of muscle disease (See Appendix B)	□ Present □ Absent □ Information not available □ Comments
3Oa. Acute onset (days to 2 weeks) of symptoms	□ Present □ Absent □ Information not available □ Comments
3Ob. Subacute onset (> 2 weeks to ≤2 months) of	□ Present

symptoms	□ Absent □ Information not available □ Comments
3Oc. Insidious onset of symptoms > 2 months to years	□ Present □ Absent □ Information not available □ Comments
4O. History of episodic weakness associated with exercise or fasting	□ Present □ Absent □ Information not available □ Comments
5O. Arthritis	□ Present □ Absent □ Information not available □ Comments
6O. Polyarthralgia	□ Present □ Absent □ Information not available □ Comments
7O. Joint contractures	□ Present □ Absent □ Information not available □ Comments
8O. Unexplained Fevers	□ Present □ Absent □ Information not available □ Comments
9O. Interstitial lung disease	□ Present □ Absent □ Information not available □ Comments
1O. Dysphagia or esophageal dysmotility	□ Present □ Absent □ Information not available □ Comments
13O. Objective improvement in strength or other disease manifestation after an adequate trial of glucocorticoids and/or other immunosuppressive or immune modulating therapy for at least 8 w. Check all that apply. - prednisone ≥0.75-2 mg/kg/day (or equivalent) - methotrexate ≥10 mg/week (children: ≥0.3 mg/kg/week) - azathioprine 75 mg/d (or 2 mg/kg/day) - Other	□ Improved □ Not improved □ Unknown □ Inadequate trial □ Not used
Muscle Biopsy Variables – from any biopsy	
Muscle biopsy performed	□Yes □ No
1B. Necrosis of type I and type II muscle fibers, phagocytosis, degeneration of myofibers	□ Present □ Absent □ Information not available □ Comments
2B. Regeneration of myofibers	□ Present □ Absent □ Information not available □ Comments
3B. Endomysial infiltration of mononuclear cells surrounding, but not invading, myofibers	□ Present □ Absent □ Information not available

	□ Comments
4B. Non-necrotic fibers surrounded and invaded by mononuclear cells	□ Present □ Absent □ Information not available □ Comments
5B.Perimysial and/or perivascular infiltration of mononuclear cells	□ Present □ Absent □ Information not available □ Comments
6B. Perifascicular atrophy	□ Present □ Absent □ Information not available □ Comments
7B. Vacuolated muscle fibers	□ Present □ Absent □ Information not available □ Comments
8B. Rimmed vacuoles	□ Present □ Absent □ Information not available □ Comments
9B. Ragged red fibers, or cytochrome C oxidase-negative fibers	□ Present □ Absent □ Information not available □ Comments
10B. Many necrotic muscle fibers as the predominant feature. Inflammatory cells are sparse; perimysial infiltrate is not evident.	□ Present □ Absent □ Information not available □ Comments
11. Immunohistochemistry data available	□Yes □ No
12B. MHC Class I antigen present on scattered or more muscle fibers	□ Present □ Absent □ Information not available □ Comments
13B. Endomysial CD8+ cells surrounding myofibers with MHC Class I expression on myofibers	□ Present □ Absent □ Information not available □ Comments
14B. Membrane attack complex (MAC) depositions on small blood vessels	□ Present □ Absent □ Information not available □ Comments
15B. Reduced capillary density	□ Present □ Absent □ Information not available □ Comments
16B. MHC-1 expression of perifascicular fibers	□ Present □ Absent □ Information not available □ Comments
17B. Electron microscopy available	□Yes □ No
18B. Tubuloreticular inclusions in endothelial cells on electron microscopy	□ Present □ Absent □ Information not available □ Comments
19B. Intracellular amyloid deposits	□ Present □ Absent □ Information not available

	□ Comments
20B.15-18 nm tubulofilaments by electron microscopy (EM)	□ Present □ Absent □ Information not available □ Comments
Laboratory Variables – record the most abnormal test values during the disease course	
1L. Serum creatine kinase (CK) activity	□Value □ Upper normal limit □ Units
2L. Serum lactate dehydrogenase (LDH) activity	□Value □ Upper normal limit □ Units
3L. Serum aspartate aminotransferase (ASAT/AST/SGOT) activity	□Value □ Upper normal limit □ Units
4L. Serum alanine aminotransferase (ALAT/ALT/SGPT) activity	□Value □ Upper normal limit □ Units
5L. Serum Aldolase activity	□Value □ Upper normal limit □ Units
6L. Erythrocyte sedimentation rate (ESR)	□Value □ Upper normal limit □ Units
7L. C-reactive protein (CRP)	□Value □ Upper normal limit □ Units
Autoantibody tests available	□Yes □ No
9L. Autoantibodies ANA Anti-Jo-1 (anti-His) Anti-Mi-2 Anti-SRP Anti-Ku Anti- PL7 Anti- PL-12 Anti PM-Scl Anti-SSA Anti-Ro52/SSA Anti-Ro60/SSA Anti-La/SSB Anti-ribonucleoprotein (RNP)-70K (U1snRNP) Anti-RNP-A Anti-RNP-C Anti-Centromere B (ACA) Anti-Topoisomerase-1/Scl70, Anti-Ribosomal P antigen Anti-Sm Anti-SmB Anti-SmD RF Anti-CCP Other, please specify below	□ Present □ Absent □ Information not available □ Comments
EMG performed	□Yes □ No
1. Electromyogram (EMG) - Increased insertional and spontaneous activity in the form of fibrillation potentials, positive sharp waves, or complex repetitive	□ Present □ Absent □ Information not available

discharges	□ Comments
1L. EMG - Morphometric analysis reveals the presence of short duration, small amplitude, polyphasic motor unit action potentials (MUAPs)	□ Present □ Absent □ Information not available □ Comments
MRI of muscles performed	□Yes □ No
1. Muscle edema on STIR or T2-weighted magnetic resonance imaging (MRI)	□ Present □ Absent □ Information not available □ Comments
2. Muscle atrophy and/or increased muscle fat content on T1-weighted MRI scanning consistent with myositis	□ Present □ Absent □ Information not available □ Comments
13L. Skin biopsy compatible with dermatomyositis (or lupus)	□ Present □ Absent □ Information not available □ Comments
Other features important in making the diagnosis not listed above – please specify	
Other laboratory features important in making the diagnosis not listed above – please specify	

Table S2. Number of valid observations of all the variables contained in the International Myositis Classification Criteria Project questionnaire. Please see Table S1 for details about the meaning of the designators of each variable.

diagnosis	1602	**M1**	1561	**S1**	1576	**O3**	1528	**B1**	1140	**Ld1**	1458	**L8_1**	1342	**E2**	892
subgroups	1599	**M2**	1458	**S2**	1548	**O4**	1395	**B2**	1028	**Ld2**	1094	**L8_2**	1062	**E3**	880
center	1602	**M3**	1388	**S3**	1560	**O5**	1554	**B3**	1036	**Ld3**	1316	**L8_3**	334	**E4**	468
gendermale	1602	**M4**	1333	**S4**	1525	**O6**	1536	**B4**	979	**Ld4**	1359	**L8_4**	283	**E5**	620
adultonset	1602	**M5**	1302	**S5**	1522	**O7**	1486	**B5**	1026	**Ld5**	544	**L8_5**	308	**E6**	299
ethnicity	1602	**M6**	1210	**S6**	1491	**O8**	1519	**B6**	998	**Ld6**	1111	**L8_6**	268		
ageatonset	1565	**M7**	1310	**S7**	1463	**O9**	1549	**B7**	983	**Ld7**	1063	**L8_7**	268		
ageatdiagnosis	1594	**M8**	1520	**S8**	1496	**O10**	1549	**B8**	978			**L8_8**	375		
ageatlasteval	1505	**M9**	1453	**S9**	1448	**O11**	1016	**B9**	946			**L8_9**	1028		
basis1	1602	**M10**	1331	**S10**	1501	**O12**	573	**B10**	1003			**L8_10**	463		
basis2	1602	**M11**	1394	**S11**	1494	**O13**	228	**B12**	368			**L8_11**	426		
basis3	1602	**M12**	1280	**S12**	1518			**B13**	279			**L8_12**	1016		
basis4	1602	**M13**	1453	**S13**	1424			**B14**	263			**L8_13**	950		
basis5	1602	**M14**	1367	**S14**	1443			**B15**	190			**L8_14**	297		
basis6	1602	**M15**	1394					**B16**	285			**L8_15**	273		
basis7	1602	**M16**	1168					**B18**	127			**L8_16**	651		
basis8	1602	**M17**	1455					**B19**	118			**L8_17**	746		
basis9	1602	**M18**	1448					**B20**	119			**L8_18**	321		
		M19	1454									**L8_19**	948		
		M20	1445									**L8_20**	273		
		M21	1409									**L8_21**	270		
		M22	1399									**L8_22**	923		
		M23	1417									**L8_23**	368		

Table S3. Comparison of physician-diagnosed idiopathic inflammatory myopathies (IIM) subgroups with IIM subgroups defined according to the classification tree among patients meeting the EULAR/ACR classification criteria for IIM

Physician-diagnosed subgroups	**Classification tree subgroups***					**Total**
	JDM	DM	ADM	IBM	PM	
JDM	235	0	0	0	0	235
DM	0	191	6	2	15	214
ADM	1	1	30	0	0	32
IBM	0	0	0	66	5	71
PM	0	7	0	3	131	141
IMNM	0	0	0	0	10	10
Total	236	199	36	71	161	703

* Classification of IIM by the EULAR/ACR classification criteria for IIM, using a 55% probability cutoff for classification, followed by the classification tree for sub-classification.

JDM, juvenile dermatomyositis; DM, dermatomyositis; ADM, amyopathic dermatomyositis; IBM, inclusion body myositis; PM, polymyositis; IMNM, immune-mediated necrotizing myopathy.