SUPPLEMENTARY MATERIAL

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Supplementary 1. International Myositis Classification Criteria Project Steering Committee

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Supplementary 2. Pilot study

Category	Variable	IIM % (n) present	IIM % (n) missing	Not IIM % (n) present	Not IIM % (n) missing	Sensitivity (%)	Specificity (%)
Clinical Muscle	Weakness, Proximal UE	97 (33)	0	71 (10)	0	97	29
	Wrist or FF weakness	56 (19)	6 (2)	2 (15)	1 (7)	56	79
	Wrist/FF > shoulder abductors	18 (6)	9 (3)	7 (1)	14 (2)	18	79
	Weakness, Proximal LE	97 (33)	0	50 (7)	0	97	50
	Hip abductor weakness	88 (30)	6 (2)	43 (6)	7 (1)	88	50
	Weakness distal LE	41 (14)	0	21 (3)	14 (2)	41	64
	Knee extensors weaker than hip	18 (6)	6 (2)	7 (1)	14 (2)	18	79
	Neck flexor weakness	85 (29)	0	43 (6)	0	85	57
	Neck extensor weakness	21 (7)	21 (7)	7 (1)	21 (3)	21	71
	Symmetric weakness	85 (29)	0	57 (8)	7 (1)	85	36
	Muscle pain at rest	32 (11)	18 (6)	29 (4)	0	32	71
	Muscle tenderness	35 (12)	9 (3)	7 (1)	7 (1)	35	86
	Muscle atrophy distal forearms	18 (6)	3 (1)	0	0	18	100
	Thigh atrophy	32 (11)	3 (1)	0	0	32	100
Clinical Skin Rashes	Heliotrope	38 (13)	0	0	0	38	100
	Gottron's papules	44 (15)	0	14 (2 DM sine)	0	44	86
	Erythema extensor surfaces	35 (12)	0	29 (4)	0	35	71
	V-sign	21 (7)	0	7 (1)	0	21	93
	Shawl sign	24 (8)	0	0	0	24	100
	Calcification	9 (3)	3 (1)	0	0	9	100
	Periungual erythema, petechiae, telangiectasis, cuticular overgrowth	41 (14)	0	7 (1)	0	41	93
	Raynaud's	18 (6)	3 (1)	29 (4)	0	18	71
Clinical Other	Family history AD	26 (9)	9 (3)	28 (4)	0	26	71
	Acute onset	71 (24)	3 (1)	43 (6)	0	71	57
	Arthritis	47 (16)	0	43 (6)	0	47	57
	Polyarthralgias	44 (15)	0	50 (7)	0	44	50
	Sjogren's syndrome	3 (1)	0	7 (1)	0	3	93
	Systemic sclerosis	0	0	7 (1)	0	0	93
	MCTD	0	0	0	0	0	0
	Rheumatoid arthritis	0	3 (1)	43 (6)	0	0	57
	SLE	3 (1)	0	29 (4)	0	3	71
	Autoimmune thyroid disease	12 (4)	18 (6)	7 (1)	42 (6)	12	50
	Objective improvement strength after corticosteroid therapy	79 (27)	6 (2)	43 (6)	43 (6)	79	14
	No improvement strength after corticosteroid	3 (1)	35 (12)	14 (2)	43 (6)	3	43

Abbreviations: IIM=idiopathic inflammatory myopathies, UE=upper extremities, FF=finger flexors, LE=lower extremities, DM=dermatomyositis, AD=autoimmune disease, MCTD=mixed connective tissue disease, SLE=systemic lupus erythematosus

Supplementary 3. 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 □ Seborhheic dermatitis □ Statin induced myopathy □ Systemic lupus erythematosus (SLE) □ Systemic sclerosis □ Systemic vasculitis, specify diagnosis below □ Toxoplasmosis □ Trichinosis □ Trypanasoma □ 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 than hip girdle muscle on the same side	□ Present □ 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 symptoms	□ Present □ 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	□ Present □ Absent □ Information not available □ Comments

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	
EMG performed	□Yes □ No
1. Electromyogram (EMG) - Increased insertional and spontaneous activity in the form of fibrillation potentials, positive sharp waves, or complex repetitive discharges	□ Present □ Absent □ Information not available □ 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	

Supplementary 4. Glossary and definitions for the International Myositis Classification Criteria Project questionnaire [30, 31]

The variables in italics have been included in previous sets of criteria for inflammatory myopathies

Clinical Muscle Variables – present at any time during the disease course	**Definition**
1M. 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
2M, Objective shoulder abductor weakness	Weakness of the shoulder abductors as defined by manual muscle testing or other objective strength testing
3M. Objective elbow flexor weakness	Weakness of the elbow flexors as defined by manual muscle testing or other objective strength testing
4M. Objective elbow extensor weakness	Weakness of the elbow extensors as defined by manual muscle testing or other objective strength testing
5M. Wrist and finger flexors are relatively weaker than shoulder abductors on the same side	Muscle grades for wrist and finger flexors are relatively lower than for shoulder abductors, as defined by manual muscle testing or other objective strength testing
6M. Wrist flexors are relatively weaker than wrist extensors on the same side	Muscle grades for wrist flexors are relatively lower than for wrist extensors as defined by manual muscle testing or other objective strength testing
7M. Objective finger flexor weakness	Finger flexor weakness as defined by manual muscle testing or other objective strength testing
8M. 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
9M. Objective hip flexor weakness	Weakness of the hip flexors as defined by manual muscle testing or other objective strength testing
10M. Objective hip abductor weakness	Weakness of the hip abductors as defined by manual muscle testing or other objective strength testing
11M. Objective knee extensor weakness	Weakness of the knee extensors as defined by manual muscle testing or other objective strength testing
12M. Knee extensors are as weak or *relatively* weaker than hip girdle muscles on the same side	Muscle grades for knee extensors are comparable to or weaker than for hip girdle muscles on the same side, as defined by manual muscle testing or other objective strength testing
13M. Objective muscle weakness of distal lower extremities	Weakness of distal lower extremities as defined by manual muscle testing or other objective strength testing or functional testing (e.g., ability to walk on heals or tip toes)
14M. Objective axial weakness	Weakness of axial muscles, including neck flexors and extensors, abdominal and trunk muscles, as defined by manual muscle testing or other objective strength testing
15M. Objective neck flexor weakness	Weakness of the neck flexors as defined by manual muscle testing or other objective strength testing
16M. 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
17M. In the legs proximal muscles are	Muscle grades for proximal muscles in the legs are

relatively weaker than distal muscles	relatively lower than distal muscles in the legs as defined by manual muscle testing or other objective strength testing
18M. In the arms proximal muscles are relatively weaker than distal muscles	Muscle grades for proximal muscles in the arms are relatively lower than distal muscles in the arms as defined by manual muscle testing or other objective strength testing
19M In the legs distal muscles are relatively weaker than proximal muscles	Distal muscles in the legs are relatively weaker than proximal muscles in the legs as defined by manual muscle testing or other objective strength testing
20M In the arms distal muscles are relatively weaker than proximal muscles	Distal muscles in the arms are relatively weaker than proximal muscles in the arms as defined by manual muscle testing or other objective strength testing
21M. Muscle tenderness	Pain in any muscle induced by squeezing or palpating the muscle
22M. Muscle atrophy of distal forearms	Objective clinical evidence by physical exam of decreased distal forearm muscle mass
23M. Muscle atrophy of thighs	Objective clinical evidence by physical exam of decreased thigh muscle mass
Skin Variables – present at any time during the disease course	**Definition**
1S. Heliotrope rash	Purple, lilac-colored or erythematous patches over the eyelids or in a periorbital distribution, often associated with periorbital edema
2S. 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.
3S. Gottron's sign	Erythematous to violaceous macules over the extensor surfaces of joints, which are not palpable
4S. Erythema of the back of the neck and shoulders (Shawl sign)	Confluent erythema around the posterior base of the neck, back and upper shoulders, often in the distribution of a shawl
5S. Erythema of the neck (V-sign)	Confluent erythema around the anterior base of the neck and the upper chest, often in the shape of a "V"
6S. Periorbital edema	Swelling around the one or both orbits
7S. Linear extensor erythema	Erythema specifically located over the extensor tendon sheaths of the hands, forearms, feet and/or forelegs
8S. Calcification	Dystrophic calcium deposits, observed clinically or by imaging, which involves the skin, subcutaneous tissue, fascia or muscle
9S. Periungual erythema or nailfold capillary abnormality	Erythema proximal to the nail bed or dilatation of periungual capillaries, which may be accompanied by vessel dropout or tortuosity, and which is visible by naked eye examination or with magnification such as with otoscopy or by use of the ophthalmoscope
10S. Mechanic's hands	Scaling or cracking of the skin over the lateral or palmar aspects of the fingers or thumbs
11S. Photodistributed violaceous erythema	Erythema over the face which may be isolated malar erythema, but may include more extensive erythema including periorbital, chin, temporal, ear and frontal regions
12S. Raynaud´s phenomenon	Discoloration of fingertips or other acral areas (two or three colors) to emotion or cold

13S. Cuticular overgrowth	Enlargement or overgrowth of the cuticle onto the nailbed
14S. Poikiloderma	A fine speckled pattern of hyperpigmented and hypopigmented macules interspersed with fine teleangiectasia and cutaneous atrophy
Other Clinical Variables – present at any time during the disease course	**Definition**
1O. Family history of autoimmune disease	Patient history or documentation that one or more of the diseases listed in Appendix A were diagnosed in a blood relative.
2O. Family history of muscle disease	Patient history or documentation that one or more of the diseases listed in Appendix B were diagnosed in a blood relative
3OA. Acute onset (days to 2 weeks) of symptoms	Onset and progression, from days to 2 weeks, of the first symptoms of the syndrome to the full disease presentation
3OB. Subacute onset (> 2 weeks to ≤ 2 months) of symptoms	Onset and progression, from 2 weeks to 2 months, of the first symptoms of the syndrome to the full disease presentation
3OC. Insidious onset of symptoms > 2 months to years	Onset and progression of the syndrome to the full disease presentation over a time period of more than 2 months
4O. History of episodic weakness associated with exercise or fasting	Patient report of weakness after exercise or fasting, which is intermittent, rather than continuous
5O. Arthritis	Inflammation, including swelling, warmth, tenderness, and/or redness of one or more joints detected by physical exam
6O. Polyarthralgia	Pain in two or more joints reported by the patient
7O. Joint contractures	Fixed limitation in the normal range of motion of joints in the absence of synovitis excluding reducible deformities, avascular necrosis and deforming arthropathy.
8O. Unexplained fevers	Two or more episodes of documented body temperature of ≥ 38 degrees Celsius without obvious cause
9O. Interstitial lung disease	Radiologic (chest x-ray or chest CT scan) documentation of inflammation or scarring (fibrosis) of the parenchyma of the lung
10O. Dysphagia or esophageal dysmotility	Difficulty in swallowing or objective evidence of abnormal motility of the esophagus
11O. Objective improvement in strength or other disease manifestation after an adequate trial of glucocorticoid therapy and/or other immunosuppressive or immune modulating therapy for at least 8 weeks.	Documented increased strength after an adequate glucocorticoid treatment trial (definition: corticosteroids: – prednisone ≥0.75-2 mg/kg/day (or equivalent)) or after an adequate treatment trial with another form of immunosuppressive therapy for 8 weeks (for methotrexate, ≥10 mg/week (children: ≥0.3 mg/kg/week); for azathioprine 75 mg/d (or 2 mg/kg/day) or other (Check all that apply.)

Muscle Biopsy Variables – from any biopsy	**Definition**
1B. Necrosis of type I and type II muscle fibers, phagocytosis, degeneration of myofibers	Necrotic or degenerating fibers appear pale and loose the cross-striations associated with the contractile apparatus. Vacuolation, or myofibrillar rarefaction may

	be seen. They may be invaded by macrophages (Phagocytosis) and vary in diameter with accompanying mononuclear infiltrates
2B. Regeneration of myofibers	Fibers with focal basophilia with large nuclei
3B. Endomysial, infiltration of mononuclear cells (MNCs) surrounding but not invading, myofibers	*Muscle biopsy reveals endomysial mononuclear cells abutting the sarcolemma of otherwise healthy, non-necrotic muscle fibers, but there is no clear invasion of the muscle fibers*
4B. Non-necrotic fibers surrounded and invaded by MNCs	Muscle biopsy reveals mononuclear cells surrounding and invading otherwise healthy, non-necrotic muscle fibers.
5B. Perimysial and/or perivascular infiltration of (MNCs)	Mononuclear cells are located in the perimysium and/or located around blood vessels (in either perimysial or endomysial vessels).
6B. Perifascicular atrophy	Muscle biopsy reveals several rows of muscle fibers which are smaller in the perifascicular region than fibers more centrally located.
7B. Vacuolated muscle fibers	Muscle biopsy reveals multiple muscle fibers containing vacuoles
8B. Rimmed vacuoles	Rimmed vacuoles are bluish by Hematoxylin and Eosin staining and reddish by modified Gomori- Trichrome stains.
9B. Ragged red fibers, or cytochrome C oxidase negative fibers	Ragged red fibers: On modified Gomori-Trichrome, staining fibers may appear to contain cracks and increased red stain in the subsarcolemmal regions. These fibers may stain intensely blue with nicotinic acid adenine dinucleotide dehydrogenase (NADH) or succinate dehydrogenase (SDH) stain or have absent or diminished staining with cytochrome C oxidase stain.
10B. Many necrotic muscle fibers as the predominant feature. Inflammatory cells are sparse; perimysial infiltrate is not evident.	The major feature of the biopsy is necrotic muscle fibers. There may be phagocytosis of necrotic fibers but otherwise there is minimal inflammatory cell infiltrate evident except in the vicinity of necrotic muscle fibres and no perimysial infiltrate by routine histochemistry (Hematoxylin and Eosin or Trichrome stains)
11B Immunohistochemistry stainings available yes/no	Yes: No:
12B. MHC Class I antigen present on scattered or more muscle fibers	Immunostaining reveals expression of MHC class I on the sarcolemma of scattered or more generally on muscle fibers.
13B. Endomysial CD8+ cells surrounding myofibers with MHC Class I expression on myofibers	Immunohistochemistry of the muscle biopsy reveals CD8 + T cells surrounding otherwise healthy, non-necrotic muscle fibers that express MHC class I antigen on their sarcolemma.
14B. Membrane attach complex (MAC) depositions on small blood vessels, , or	Immunocytochemistry demonstrates deposition of membrane attack complex (MAC, C5b-9) on or around small blood vessels.
15B. Reduced capillary density	*Reduced capillary density as appreciated on quantitative analysis*
16B MHC-1 expression of perifascicular fibers	*MHC-class 1 expression is predominant on perifascicular muscle fibers*
17B. Electron microscopy information	*Yes: No:*

available y/n	
18B. Tubuloreticular inclusions in endothelial cells on electron microscopy	*Tubuloreticular inclusions are evident in endothelial cells on electron microscopy*
19B. Intracellular amyloid deposits	Intracellular amyloid deposits are evident in electron microscopy
20B.15-18 nm tubulofilaments by electron microscopy (EM)	
Laboratory Variables – record the highest values during the disease course	**Definition**
1L. Serum Creatine kinase (CK) activity	Please list the highest absolute value available and the upper limits of normal with units
2L. Serum Lactate dehydrogenase (LDH) activity	Please list absolute values and upper limits of normal with units
3L. Serum aspartate aminotransferase (ASAT/AST/SGOT) activity	Please list absolute values and upper limits of normal with units
4L. Serum alanine aminotransferase (ALAT/ALT/SGPT) activity	Please list absolute values and upper limits of normal with units
5L. Serum Aldolase activity	Please list absolute values and upper limits of normal with units
6L. Erythrocyte sedimentation rate (ESR)	Please list absolute values and upper limits of normal with units
7L. C-reactive protein (CRP)	Please list absolute values and upper limits of normal with units
8L. ***Autoantibody tests available y/n***	**Yes:** **No:**

9L. Autoantibodies	Positive	Negative	Not tested	
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				

EMG performed y/n	**Yes:**	**No:**
1.Electromyogram (EMG) - Increased insertional and spontaneous activity in the form of fibrillation potentials, positive sharp waves, or complex repetitive discharges	Increased insertional activity: upon insertion of the EMG needle there are fibrillation potentials, positive sharp waves, or complex repetitive discharges or myotonic discharges. Increased spontaneous activity: fibrillation potentials, positive sharp waves, complex repetitive discharges or pseudomyotonic discharges are seen on needle EMG even when the needle is resting in the muscle without further movement	
2.EMG - Morphometric analysis reveals the presence of short duration, small amplitude, polyphasic motor unit action potentials (MUAPs)	Analysis of at least 20 individual motor unit action potentials reveals that the average duration is short, amplitude is small, and phases are greater than 4.	
MRI performed y/n	**Yes:**	**No:**
1. Muscle oedema on STIR or T2-weighted magnetic resonance imaging (MRI)	Increased signal in muscle, often symmetric, by short tau inversion recovery (STIR)- or T-2 weighted MRI imaging, without other known cause	
2. Muscle atrophy or replacement of muscle by fat on T1-weighted MRI scanning consistent with myositis	Decreased muscle volume (i.e., muscle atrophy) or increased fat content of muscle by T1-weighted MRI imaging, without other known cause	
13L. Skin biopsy compatible with dermatomyositis (or lupus)	Biopsy findings consistent with dermatomyositis or lupus (these could include: intradermal or perivascular inflammatory cell infiltrate, liquefaction, basal cell degeneration, epidermal atrophy, hyperkeratosis, melanin incontinence, mucin deposition)	
Other features important in making the diagnosis not listed above	Any other documented clinical signs, symptoms or laboratory findings, not listed above, that were important in diagnosing the patient	

Supplementary 5. Adult comparator cases in the International Myositis Classification Criteria Project dataset

DIAGNOSIS*	n (%)
Systemic inflammatory disease	**181 (35.7)**
Systemic sclerosis	49
Systemic lupus erythematosus	41
Polymyalgia reumatica	20
Mixed connective tissue disease	15
Systemic vasculitits	15
Sjögren's syndrome	10
Rheumatoid arthritis	9
Undifferentiated connective tissue disease	6
Granulomatous myositis (sarcoidosis)	2
Polyarthritis	2
Raynaud´s syndrome	2
Crohn's disease	1
Remitting Seronegative Symmetrical Synovitis with Pitting Edema (RS3PE)	1
Granulomatous myositis	1
Ankylosis spondylitis	1
Undifferentiated spondyloarthropathy	1
Undifferentiated myopathy	1
Osteoarthritis	1
Soft tissue rheumatism	1
Panniculitis	1
Not specified	1
Dystrophy	**68 (13.4)**
Limb-girdle dystrophy	27
Fascioscapulohumeral dystrophy	11
Myotonic dystrophy	8
Dysferlinopathy	5
Becker´s dystrophy	5
Occulopharngeal muscular dystrophy	3
Welanders distal myopathy	2
Proximal myotonic myopathy (myotonic dystrophy type 2)	1
Nemaline myopathy	1
Emery-Dreifuss muscular dystrophy	1
Progressive muscular dystrophy	1
Generalized muscle pseudo-hypertrophy (Myotilinopathy)	1
Not specified	2
Drug or toxin-associated myopathy	**48 (9.5)**
Statin induced myopathy	20
Glucocorticoid induced myopathy	5
D-penicillamine	2
Acute rhabdomyolysis	5
Dropped head, related to psychiatric medication	1
Alcohol induced Neuromyopathy	1
Drug-induced rhabdomyolysis	3
Unknown cause	1
Related to anti HIV therapy	1

Rhabdomyolysis from over exertion and dehydration	1
Hydrochloroquine	1
Ranitidine induced myopathy	1
Not specified	6
Motor neuron disease / neuropathies	**44 (8.7)**
Amyotrophic lateral sclerosis (ALS)	24
Gullain-Barre syndrome	3
Myasthenia gravis	2
Kennedy´s disease	2
Multiple mononeuropathy due to systemic lupus erythematosus	1
Spinal muscle atrophy	1
Peripheral neuropathy	1
Progressive muscular atrophy	1
Multiple myeloma and chronic demyelinating inflammatory neuropathy	1
Not specified	8
Metabolic myopathy	**29 (5.7)**
Mc Ardle´s disease	6
Asymptomatic hyper-CK-emia	6
Acid maltase deficiency	4
Phosphofructokinase deficiency	2
Familial hyper-CK-emia	1
Carnitine deficiency	1
Palmityltransferase deficiency	1
Myoadenylate deaminase deficiency	1
Hypokalemia	1
Hypocalcemia	1
Sandhoff disease	1
Late onset Pompe	1
Phosphorilase B kinase deficiency	1
Metabolic Myopathy Unspecified	1
Not specified	1
Myalgias	**27 (5.3)**
Fibromyalgia	25
Myalgia	2
Dermatologic disease	**21 (4.1)**
Hypereosinophilic syndrome	5
Psoriasis	4
Psoriatic arthritis	3
Subacute cutaneous lupus erythematosus	3
Discoid lupus erthematosus	3
SLE complicated by tumid lupus and subacute cutaneous lupus erythematosus	1
Psoriatic sponyloarthropathy	1
Rosacea	1
Endocrine myopathy	**22 (4.3)**
Hypothyroidism	18

Hyperthyroidism	1
Cushing syndrome	1
Diabetes mellitus	1
Hashimoto disease, sarcoidosis and gastric chronica atrophica	1
Infectious myopathy	**20 (3.7)**
Viral myopathy	12
Parasitic myopathy	1
Tuberculous myositis & fasciitis	1
Tuberculosis	1
Tuberculosis rheumatism	1
Pneumonia	2
Whipple´s disease	1
Not specified	1
Mitochondrial myopathy	**15 (3.0)**
Myoclonic epilepsy, ragged red fibers (MERRF)	1
Encephalopathy + myopathy but normal lactate curve	1
CPEO	1
Mitochondrial cytopathy	1
Abnormal mt DNA	1
Not specified	10
Neuromuscular disease	**12 (2.4)**
Myopathy non inflammatory	2
Lambert Eatons Myastenic Syndrome (LEMS)	1
Hereditary inclusion body myopathy	1
HCV-related axonal polyneuropathy	1
Lumbar radiculopathy	1
Polyradiculopathy	1
Charcot-Marie-Tooth disease peroneal muscular atrophy	1
Chronic axonal polyneuropathy	1
Lumboischialgia	1
Central pontine myelinolysis	1
Not specified	1
Other myopathy	**12 (2.4)**
Paraneoplastic syndrome	2
Non-autoimmune myositis	2
Crural biceps myopathy, not specified	1
Myopathy secondary to neuroleptic malignant syndrome	1
Macro CK type 1	1
Oculopharyngeal myopathy	1
Necrotising myopathy	1
Paraneoplastic myopathy (neoplastic hypophosphatemia osteomalacia)	1
Distal myopathy non diagnostic	1
Undifferentiated myopathy	1
Immune-mediated skin condition	**3 (0.6)**

Allergies	2
Hypersensitivity conditions	1
Other diagnosis	**5 (1.2)**
Mixed crystal-induced myoarthopathies	1
Chronic, non-inflammatory back pain	1
Amyloidosis	1
Still diagnosed for apocamnosis, suspicion of congenital myotonic dystrophy	1
Multicentric reticulohistiocytosis	1
TOTAL	**507**

* As entered by treating physician

Abbreviations: CK=Creatine kinase, CPEO= Chronic progressive external ophthalmoplegia

Supplementary 6. Juvenile comparator cases in the International Myositis Classification Criteria Project dataset

DIAGNOSIS*	n (%)
Systemic inflammatory disease	**47 40.2)**
Systemic lupus erythematosus	16
Mixed connective tissue disease	15
Juvenile idiopathic arthritis	9
Undifferentiated connective tissue disease	3
Systemic sclerosis	2
Celiac disease	1
Crohn's disease	1
Dystrophy	**32 (27.4)**
Limb-girdle dystrophy	8
Duchenne´s dystrophy	6
Becker´s dystrophy	6
Fascioscapulohumeral dystrophy	3
Becker Muscular Dystrophy	2
Dysferlinopathy	1
Congenital muscular dystrophy positive merosin	1
Probably a mild limp girdle, FKRP mutation, L276I homozygote	1
Calpain deficient myopathy (limb-girdle)	1
Carrier Duchenne Muscular Distrophy	1
Fascioscapular Humoral Dystrophy	1
Not specified	1
Metabolic myopathy	**14 (12.0)**
Pompe´s disease	11
Acid maltase deficiency	1
Carnitine deficiency	1
Metabolic myopathy, unspecified	1
Infectious myopathy	**8 (6.8)**
Viral myopathy	4
Parasitic myopathy	1
Post - mycoplasma myositis	1
Focal myositis	1
Staphylococcus pyomyositis	1
Motor neuron disease	**4 (3.4)**
Myasthenia gravis	3
Spinal Muscular Atrophy, type III	1
Neuromuscular disease	**4 (3.4)**
Congenital myopathy, unknown origin	1
Griscillis´ syndrome with critical illness myopathy	1
Muscular dystrophy. R/o distal myopathy	1
Charcot-Marie-Tooth	1
Dermatologic disease	**2 (1.7)**
Hypereosinophilic syndrome	2

Drug or toxin-associated myopathy	**1 (0.9)**
Rhabdomyolysis	1
Endocrine myopathy	**1 (0.9)**
Hypothyroidism	1
Myalgias	**1 (0.9)**
Fibromyalgia	1
Other diagnosis	**3 (2.6)**
Myositis ossificans	1
Teratoma	1
Other diagnosis, unspecified	1
TOTAL	**117**

* As entered by treating physician

Abbreviations: NYD=x

Supplementary 7. Validation cohort from the Euromyositis register

	Patients[*] (n=592)
Sex, n (%)	
Female	412 (69.6)
Male	174 (29.4)
NA	6 (1.0)
Diagnosis[†], n (%)	
Polymyositis	256 (46.7)
Dermatomyositis	242 (45.3)
Inclusion body myositis	30 (5.5)
Juvenile dermatomyositis	16 (2.9)
Amyopathic dermatomyositis	4 (0.7)
Disease duration, median (IQR), years	6.0 (3.0-12.0)
Ethnicity[†], n (%)	
Caucasian	485 (81.9)
Hispanic	6 (1.0)
Oriental	3 (0.5)
Afro-Caribbean	1 (0.2)
Asian	3 (0.5)
NA	94 (15.9)

[*]Data from Karolinska University Hospital, Stockholm, Sweden, Prague Hospital, Prague, Czech Republic and Oslo University Hospital, Oslo, Norway

[†]As defined in the Euromyositis register

Abbreviations: NA=Information not available, IQR=Interquartile range

Supplementary 8. Validation cohort from the Juvenile dermatomyositis cohort biomarker study and repository (UK and Ireland)

	Patients (n=332)
Sex, n (%)	
Female	227 (68.4)
Male	104 (31.3)
NA	1 (0.3)
Diagnosis[*], n (%)	
Definite juvenile dermatomyositis	292 (88.0)
Probable juvenile dermatomyositis	20 (6.0)
Focal myositis	6 (1.8)
Definite polymyositis	4 (1.2)
Probable polymyositis	2 (6.0)
Other IIM	8 (2.4)
Disease duration, median (IQR), years	7.7 (3.9-11.7)
Ethnicity[*], n (%)	
White	261 (78.6)
Black-African	10 (3.0)
Black-Caribbean	11 (3.3)
Black-Other	4 (1.2)
Pakistani	11 (3.3)
Indian	10 (3.0)
Bangladeshi	4 (1.2)
Chinese	1 (0.30)
Other ethnic group	19 (5.7)
NA	1 (0.30)

[*]As defined in the Juvenile dermatomyositis cohort biomarker study and repository (UK and Ireland)
NA, information not available; IIM, idiopathic inflammatory myopathies; IQR, interquartile range.