

Development of Classification Criteria for the Idiopathic Inflammatory Myopathies and their Major Subgroups

Data Collection Form, Variable List and Glossary

In reviewing patients please use the following criteria for selecting cases for submission:

- i. The subject has been diagnosed for at least 6 months
- ii. The physician is certain of the diagnosis – only cases with known idiopathic inflammatory myopathy or, as comparators, known non-IIM cases (but in which myositis was considered in the initial differential diagnosis) are chosen
- iii. The patients in whom most complete data are available
- iv. The most recent cases are chosen first – these would likely result in more consistent evaluations and therapy

GENERAL INFORMATION

1. Have you received approval from your local IRB or ethics committee for participation in this project?

Yes

Exempt

No

Please scan and email or fax the letter of approval to Dr Ingrid Lundberg, Rheumatology Unit Karolinska University Hospital, Stockholm, Sweden. Ingrid.Lundberg@ki.se, Fax no: +46 8 5177 3080. Hereafter you will receive a study number of your center.

2. Date of data entry: (dd/month/year) _____ -

3. Center (name of university or hospital from where data is entered)

4. Clinician submitting case:

Family name _____

First name _____

5. Email _____

6. Postal address _____

7. Case identifier/number (Center number + individual case number) : _____

8. Gender: Female

Male

9A. Age (years) at onset of first symptom assumed to be related to the disease: ____ years ____ months (include years and months for children)

9B. Age (years) at diagnosis: ____ years ____ months (include years and months for children)

10. Ethnicity: (check all that apply)

- Of European descent
- Of African descent
- Of Asian descent
- Of Native American descent
- Of Pacific Island descent
- Of Hispanic descent
- Of Mixed descent
- Unknown

11. Study diagnosis according to the clinician submitting the case:

A. Idiopathic Inflammatory Myopathy (IIM), adults or children:

Myositis Onset: adult (*diagnosed at age ≥ 18*) or *childhood* (*diagnosed at age < 18 years*)
(check only one diagnosis):

- Polymyositis
- Dermatomyositis
- Amyopathic dermatomyositis
- Hypomyopathic dermatomyositis
- Inclusion body myositis
- Immune-mediated necrotizing myopathy

B. NOT Idiopathic Inflammatory Myopathy (Not IIM), adults or children, but in which the diagnosis of idiopathic myositis was considered in the differential diagnosis:

Disease Onset: adult (*diagnosed at age ≥ 18*) or *childhood* (*diagnosed at age < 18 years*)

- Non-inflammatory inclusion body myopathy
- Dystrophy, specify diagnosis
- Metabolic myopathy, specify diagnosis
- Mitochondrial myopathy, specify diagnosis
- Drug or toxin associated myopathy, specify diagnosis

- Infectious myopathy, specify diagnosis
- Endocrine myopathy, specify diagnosis
- Motor neuron diseases, please specify diagnosis
- Other neuromuscular disease, specify diagnosis
- Other rheumatic disease, specify diagnosis
- Other dermatologic disease, specify diagnosis
- Other diagnosis, specify

12. Basis for study diagnosis (check all supporting reasons):

- Muscle weakness
- Muscle biopsy abnormalities
- Elevated muscle enzymes
- EMG abnormalities
- Rashes
- Skin biopsy
- Other, please specify _____

G9. Clinician's additional comments on the case: _____

G10. Other diagnoses in this case: (check all that apply):

- Hypothyroidism
- Hyperthyroidism
- Type I diabetes
- Rheumatoid arthritis
- Juvenile idiopathic arthritis
- Systemic lupus erythematosus
- Systemic sclerosis
- Sjögren's syndrome
- Mixed connective tissue disease
- Malignancy
 - If yes, please specify type of malignancy _____
 - If yes, add age at diagnosis of malignancy _____
- Other, please specify _____
- Non applicable

Before completing the following tables, please review the Glossary of Definitions below.

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

	Present	Absent	Not available	Comments
Clinical Muscle Variables – present at any time during the disease course				
<i>1M. Objective symmetric weakness, usually progressive, of the proximal upper extremities</i>				
<i>2M. Objective shoulder abductor weakness</i>				
<i>3M. Objective elbow flexor weakness</i>				
<i>4M. Objective elbow extensor weakness</i>				
<i>5M. Wrist and finger flexors are relatively weaker than shoulder abductors on the same side</i>				
<i>6M. Wrist flexors are relatively weaker than wrist extensors on the same side</i>				
<i>7M. Objective finger flexor weakness</i>				
<i>8M. Objective symmetric weakness, usually progressive, of the proximal lower extremities</i>				
<i>9M. Objective hip flexor weakness</i>				
<i>10M. Objective hip abductor weakness</i>				
<i>11M. Objective knee extensor weakness</i>				
12M. Knee extensors are as weak or relatively weaker than hip girdle muscle on the same side				
13M. Objective muscle weakness of distal lower extremities				
<i>14M. Objective axial weakness</i>				
15M. Objective neck flexor weakness				
<i>16M. Neck flexors are relatively weaker than neck extensors</i>				
<i>17M. In the legs proximal muscles are relatively weaker than distal muscles</i>				
18M. In the arms proximal muscles are relatively weaker than distal muscles				
19M In the legs distal muscles relatively weaker than proximal muscles				
20M In the arms distal muscles are relatively weaker than proximal muscles				
21M. Muscle tenderness				
22M. Muscle atrophy of distal forearms				
23M. Muscle atrophy of thighs				
	Present	Absent	Not available	Comments
Skin Variables – present at any time during the disease course				
<i>1S. Heliotrope rash</i>				
<i>2S. Gottron's papules</i>				
<i>3S. Gottron's sign</i>				
<i>4S. Erythema of the neck (V-sign)</i>				
<i>5S. Erythema of the back of neck and</i>				

<i>shoulders (Shawl sign)</i>				
6S. Periorbital edema				
7S. Linear extensor erythema				
8S. Calcification				
9S. Periungual erythema or nailfold capillary abnormality				
10S. Mechanic's hands				
11S. Photodistributed violaceous erythema				
12 S. Raynaud's phenomenon				
13S. Cuticular overgrowth				
14S Poikiloderma				
	Present	Absent	Not available	Comments
Other Clinical Variables – present at any time during the disease course				
10. Family history of autoimmune disease (see Appendix A)				
20. Family history of muscle disease (See Appendix B)				
30. Acute onset (days to 2 weeks) of symptoms				
40. Subacute onset (> 2 weeks to ≤2 months) of symptoms				
50. Insidious onset of symptoms > 2 months to years				
60. History of episodic weakness associated with exercise or fasting				
70. Arthritis				
80. Polyarthralgia				
90. Joint contractures				
100. Unexplained Fevers				
110. Interstitial lung disease				
120. Dysphagia or esophageal dysmotility				
130. Objective improvement in strength or other disease manifestation after an adequate trial of corticosteroid and/or other immunosuppressive or immune modulating therapy				

	Present	Absent	Not available	Comments
Muscle Biopsy Variables – from any biopsy				
Muscle biopsy data available				
1B. Necrosis of type I and type II muscle fibers, phagocytosis, degeneration of myofibers				
2B. Regeneration of myofibers				
3B. Endomysial infiltration of mononuclear cells surrounding, but not invading, myofibers				
4B. Non-necrotic fibers surrounded and invaded by mononuclear cells				
5B. Perimysial and/or perivascular infiltration of mononuclear cells				
6B. Perifascicular atrophy				

7B. Vacuolated muscle fibers				
8B. Rimmed vacuoles				
9B. Ragged red fibers, or cytochrome C oxidase-negative fibers				
10B. Many necrotic muscle fibers as the predominant feature. Inflammatory cells are sparse; perimysial infiltrate is not evident.				
11. Immunohistochemistry data available if not go to xxx				
12B. MHC Class I antigen present on scattered or more muscle fibers				
13B. Endomysial CD8+ cells surrounding myofibers with MHC Class I expression on myofibers				
14B. Membrane attack complex (MAC) depositions on small blood vessels				
15B. Reduced capillary density				
16B. MHC-1 expression of perifascicular fibers				
17B. Electron microscopy available y/n				
18B. Tubuloreticular inclusions in endothelial cells on electron microscopy				
19B. Intracellular amyloid deposits or 15-18 nm tubulofilaments by electron microscopy (EM)				

Laboratory Variables – record the most abnormal test values during the disease course			
	<u>Value</u>	<u>Upper normal limit</u>	<u>Units</u>
1L. Serum creatine kinase (CK) activity			
2L. Serum lactate dehydrogenase (LDH) activity			
3L. Serum aspartate aminotransferase (ASAT/AST/SGOT) activity			
4L. Serum alanine aminotransferase (ALAT/ALT/SGPT) activity			
5L. Serum Aldolase activity			
6L. Erythrocyte sedimentation rate (ESR)			
7L. C-reactive protein (CRP)			
8L. Autoantibody tests available y/n	<u>Positive</u>	<u>Negative</u>	<u>Not tested</u>
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				
.				
	Present	Absent	Not available	Comments
<i>EMG performed y/n</i>				
<i>I . Electromyogram (EMG) - Increased insertional and spontaneous activity in the form of fibrillation potentials, positive sharp waves, or complex repetitive discharges</i>				
<i>II. EMG - Morphometric analysis reveals the presence of short duration, small amplitude, polyphasic motor unit action potentials (MUAPs)</i>				
MRI of muscles performed y/n				
1. Muscle edema on STIR or T2-weighted magnetic resonance imaging (MRI)				
2. Muscle atrophy and/or increased muscle fat content on T1-weighted MRI scanning consistent with myositis				
13L. Skin biopsy compatible with dermatomyositis (or lupus)				

Other features important in making the diagnosis not listed above – please specify:

GLOSSARY FOR THE

INTERNATIONAL MYOSITIS CLASSIFICATION CRITERIA PROJECT

This document is a GLOSSARY to be used for completing the INTERNATIONAL MYOSITIS CLASSIFICATION CRITERIA PROJECT DATASHEET. Please read this carefully prior to completing the DATASHEET and refer to it whenever questions arise as to how to best enter your data.

Clinical Muscle Variables – present at any time during the disease course	Definition
<i>1M. Objective symmetric weakness, usually progressive, of the proximal upper extremities</i>	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
<i>2M. Objective shoulder abductor weakness</i>	Weakness of the shoulder abductors as defined by manual muscle testing or other objective strength testing
<i>3M. Objective elbow flexor weakness</i>	Weakness of the elbow flexors as defined by manual muscle testing or other objective strength testing
<i>4M. Objective elbow extensor weakness</i>	Weakness of the elbow extensors as defined by manual muscle testing or other objective strength testing
<i>5M. Wrist and finger flexors are relatively weaker than shoulder abductors on the same side</i>	Muscle grades for wrist and finger flexors are relatively weaker than for shoulder abductors, as defined by manual muscle testing or other objective strength testing
<i>6M. Wrist flexors are relatively weaker than wrist extensors on the same side</i>	Muscle grades for wrist flexors are relatively weaker than for wrist extensors as defined by manual muscle testing or other objective strength testing
<i>7M. Objective finger flexor weakness</i>	Finger flexor weakness as defined by manual muscle testing or other objective strength testing
<i>8M. Objective symmetric weakness, usually progressive, of the proximal lower extremities</i>	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
<i>9M. Objective hip flexor weakness</i>	Weakness of the hip flexors as defined by manual muscle testing or other objective strength testing
<i>10M. Objective hip abductor weakness</i>	Weakness of the hip abductors as defined by manual muscle testing or other objective strength testing
<i>11M. Objective knee extensor weakness</i>	Weakness of the knee extensors as defined by manual muscle testing or other objective strength testing
<i>12M. Knee extensors are as weak or relatively weaker than hip girdle muscles on the same side</i>	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
<i>13M. Objective muscle weakness of distal lower extremities</i>	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 heels or tip toes)
<i>14M. Objective axial weakness</i>	Weakness of axial muscles, including neck flexors and extensors, abdominal and trunk muscles, as defined by manual muscle testing or other objective strength testing
<i>15M. Objective neck flexor weakness</i>	Weakness of the neck flexors as defined by manual

	muscle testing or other objective strength testing
<i>16M. Neck flexors are relatively weaker than neck extensors</i>	Muscle grades for neck flexors are relatively weaker than neck extensors as defined by manual muscle testing or other objective strength testing
<i>17M. In the legs proximal muscles are relatively weaker than distal muscles</i>	Muscle grades for proximal muscles in the legs are relatively weaker than distal muscles in the legs as defined by manual muscle testing or other objective strength testing
<i>18M. In the arms proximal muscles are relatively weaker than distal muscles</i>	Muscle grades for proximal muscles in the arms are relatively weaker 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
<i>1S. Heliotrope rash</i>	Purple, lilac-colored or erythematous patches over the eyelids or in a periorbital distribution, often associated with periorbital edema
<i>2S. Gottron’s papules</i>	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.
<i>3S. Gottron’s sign</i>	Erythematous to violaceous macules over the extensor surfaces of joints, which are not palpable
<i>4S. Erythema of the neck (V-sign)</i>	Confluent erythema around the anterior base of the neck and the upper chest, often in the shape of a “V”
<i>5S. Erythema of the back of the neck and shoulders (Shawl sign)</i>	Confluent erythema around the posterior base of the neck, back and upper shoulders, often in the distribution of a shawl
<i>6S. Periorbital edema</i>	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
3O. 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
4O. 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
5O. 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
6O. History of episodic weakness associated with exercise or fasting	Patient report of weakness after exercise or fasting, which is intermittent, rather than continuous
7O. <i>Arthritis</i>	Inflammation, including swelling, warmth, tenderness, and/or redness of one or more joints detected by physical exam
8O. <i>Polyarthralgia</i>	Pain in two or more joints reported by the patient
9O. 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.
10O. <i>Unexplained fevers</i>	Two or more episodes of documented body temperature of ≥ 38 degrees Celsius without obvious cause
11O. Interstitial lung disease	Radiologic (chest x-ray or chest CT scan) documentation of inflammation or scarring (fibrosis) of the parenchyma of the lung
12O. Dysphagia or esophageal dysmotility	Difficulty in swallowing or objective evidence of abnormal motility of the esophagus

13O. Objective improvement in strength or other disease manifestation after an adequate trial of corticosteroid therapy and/or other immunosuppressive or immune modulating therapy	Documented increased strength after an adequate corticosteroid treatment trial (definition: corticosteroids: – prednisone ≥ 0.75 -2 mg/kg/day (or equivalent) for at least 1 month , a) 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.)
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Muscle Biopsy Variables – from any biopsy	Definition
<i>1B. Necrosis of type I and type II muscle fibers, phagocytosis, degeneration of myofibers</i>	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
<i>2B. Regeneration of myofibers</i>	Fibers with focal basophilia with large nuclei
<i>3B. Endomysial, infiltration of mononuclear cells (MNCs) surrounding but not invading, myofibers</i>	<i>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</i>
<i>4B. Non-necrotic fibers surrounded and invaded by MNCs</i>	Muscle biopsy reveals mononuclear cells surrounding and invading otherwise healthy, non-necrotic muscle fibers.
<i>5B. Perimysial and/or perivascular infiltration of (MNCs)</i>	Mononuclear cells are located in the perimysium and/or located around blood vessels (in either perimysial or endomysial vessels).
<i>6B. Perifascicular atrophy</i>	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.
<i>10B. Many necrotic muscle fibers as the predominant feature. Inflammatory cells are sparse; perimysial infiltrate is not evident.</i>	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	
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	<i>Reduced capillary density as appreciated on quantitative analysis</i>
16B MHC-1 expression of perifascicular fibers	<i>MHC-class 1 expression is predominant on perifascicular muscle fibers</i>
17B. Electron microscopy information available y/n	
18B. Tubuloreticular inclusions in endothelial cells on electron microscopy	<i>Tubuloreticular inclusions are evident in endothelial cells on electron microscopy</i>
19B. Intracellular amyloid deposits or 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. 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	Autoantibodies tested, check all that apply. If tested: specify negative or positive and when possible which method was used,

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	
EMG performed y/n	
1. . <i>Electromyogram (EMG) - Increased insertional and spontaneous activity in the form of fibrillation potentials, positive sharp waves, or complex repetitive discharges</i>	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. . <i>EMG - Morphometric analysis reveals the presence of short duration, small amplitude, polyphasic motor unit action potentials (MUAPs)</i>	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	
1. Muscle edema 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.	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)
<u>Other features important in making the diagnosis not listed above</u>	Any other documented clinical signs, symptoms or laboratory findings, not listed above, that were important in diagnosing the patient

Appendix A. List of autoimmune diseases, adult or juvenile onset, to be considered for family history question 10

- Autoimmune thyroid diseases - Graves Disease or Hashimoto's Thyroiditis
- Type 1 diabetes
- Rheumatoid arthritis or juvenile rheumatoid arthritis
- Systemic Lupus Erythematosus (Lupus)
- Multiple Sclerosis
- Inflammatory bowel disease - Crohn's disease or Ulcerative colitis
- Psoriasis
- Sjogren's Syndrome
- Scleroderma (Systemic sclerosis)
- Addison's Disease
- Pernicious anemia
- Myasthenia Gravis
- Primary biliary cirrhosis
- Mixed connective tissue disease
- Vitiligo
- Autoimmune Hemolytic Anemia
- Autoimmune Thrombocytopenia
- Celiac Disease
- Pemphigus
- Alopecia Areata
- Behcet's Disease
- Myositis - Polymyositis, Dermatomyositis or Inclusion body myositis
- Goodpasture's syndrome
- Wegener's Granulomatosis
- Polyarteritis nodosa
- Henoch Schonlein Purpura.
- None
- Unknown

Appendix B. List of muscle diseases, adult or juvenile onset, to be considered for family history question 20

- Non-inflammatory inclusion body myopathy
- Muscular Dystrophy
- Metabolic myopathy
- Mitochondrial myopathy
- Drug or toxin associated myopathy
- Infectious myopathy
- Endocrine myopathy
- Motor neuron disease
- None
- Unknown
-

Appendix C. 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