# 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

**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.

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

**1.** 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 □

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

_____

_____

**3. Clinician submitting case:**
**Family name**_____

**First name**_____

**Email** _____

**Postal** address_____

**4**. **Case identifier/number**
Please fill in the study ID numbers that you have been given by the coordinating center.

Center number _____

Physician number _____

Case number _____

**5. Gender**: □ Female
□ Male

**6**. Adult onset (diagnosed at age ≥18) or childhood onset (diagnosed at age < 18 years)

Adult □ Childhood □

**7**. **Age (years) at onset** of first symptom assumed to be related to the disease: ____years and for children months: ___ months (include years and months for children)

**8. Age (years) at diagnosis**: ____years ___ months (include years and months for children)

**9. Age (years) at last evaluation**: ____years ___ months (include years and months for children)

**10. Ethnicity**: (check all that apply)

□ Caucasian

□ 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

□ Idiopathic inflammatory myopathy (IIM) adults or children

□ Not Idiopathic inflammatory myopathy (Not IIM) adults or children

**12.** Study diagnosis: Idiopathic Inflammatory Myopathy (IIM) in adults or children

Please select only one diagnosis, where requested please specify diagnosis below in the text box.

□ 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)

_____

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

Please select only one diagnosis, where requested please specify diagnosis below in the text box.

| |
|---|
| □ 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) |

If other diagnosis please specify:----------

**14. 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 _____

**14. 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
If yes, please specify type of malignancy_____

If yes, add age at diagnosis of malignancy _____

□ Other, please specify _____

***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** | **Information not available** | **Comments** |
|---|---|---|---|---|
| **Clinical Muscle Variables – present at any time during the disease course** | | | | |
| *1M. Objective symmetric weakness, usually progressive, of the proximal upper extremities* | | | | |

| | | | | |
|---|---|---|---|---|
| *2M, Objective shoulder abductor weakness* | | | | |
| *3M. Objective elbow flexor weakness* | | | | |
| *4M. Objective elbow extensor weakness* | | | | |
| *5M. Wrist and finger flexors are relatively weaker than shoulder abductors on the same side* | | | | |
| *6M. Wrist flexors are relatively weaker than wrist extensors on the same side* | | | | |
| *7M. Objective finger flexor weakness* | | | | |
| *8M. Objective symmetric weakness, usually progressive, of the proximal lower extremities* | | | | |
| *9M. Objective hip flexor weakness* | | | | |
| *10M. Objective hip abductor weakness* | | | | |
| *11M. Objective knee extensor weakness* | | | | |
| 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 | | | | |
| *14M. Objective axial weakness* | | | | |
| 15M. Objective neck flexor weakness | | | | |
| *16M. Neck flexors are* relatively *weaker than neck extensors* | | | | |
| *17M. In the legs proximal muscles are* relatively *weaker than distal muscles* | | | | |
| *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** | **Information not available** | **Comments** |
| **Skin Variables – present at any time during the disease course** | | | | |
| *1S. Heliotrope rash* | | | | |
| *2S. Gottron´s papules* | | | | |
| *3S. Gottron's sign* | | | | |
| *4S. Erythema of the back of neck and shoulders (Shawl sign)* | | | | |
| *5S. Erythema of the neck (V-sign)* | | | | |
| *6S. Periorbital edema* | | | | |
| **7**S. 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** | **Information not available** | **Comments** |
| **Other Clinical Variables – present at any time during the disease course** | | | | |
| 1O. Family history of autoimmune disease (see Appendix A) | | | | |
| 2O. Family history of muscle disease (See Appendix B) | | | | |
| 3Oa. Acute onset (days to 2 weeks) of symptoms | | | | |
| 3Ob. Subacute onset (> 2 weeks to ≤2 months) of symptoms | | | | |
| 3Oc. Insidious onset of symptoms > 2 months to years | | | | |
| 4O. History of episodic weakness associated with exercise or fasting | | | | |
| *5O. Arthritis* | | | | |
| *6O. Polyarthralgia* | | | | |
| 7O. Joint contractures | | | | |
| *8O. Unexplained Fevers* | | | | |
| 9O. Interstitial lung disease | | | | |
| 1O. Dysphagia or esophageal dysmotility | | | | |

| 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. | Improved | Not improved | Unknown | Inadequate trial | Not used |
|---|---|---|---|---|---|
| **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** | | | | | |
| Specify other immunosuppressive medication: | | | | | |

| | **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** | | | |
|---|---|---|---|
| | **Value** | **Upper normal limit** | **Units** |
| *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*** | **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 | | | | |

| | **Present** | **Absent** | **Information not available** | | **Comments** |
|---|---|---|---|---|---|
| ***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* | | | | | |
| *1L. EMG - Morphometric analysis reveals the presence of short duration, small amplitude, polyphasic motor unit action potentials (MUAPs)* | | | | | |
| MRI of muscles performed y/n | **Yes:** | | **No:** | | |
| 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) | | | N/a | Not performed | |

**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:

_____

**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** |
|---|---|
| *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 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 |
| *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 available y/n | *Yes: No:* |
| 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.<br>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 | |

**Appendix A. List of autoimmune diseases, adult or juvenile onset, to be considered for family history question 1O. Specify in the comments box.**

- Addison's Disease
- Alopecia Areata
- Autoimmune Hemolytic Anemia
- Autoimmune Thrombocytopenia
- Autoimmune thyroid diseases - Graves Disease or Hashimoto's
- Behcet's Disease
- Celiac Disease
- Goodpasture's syndrome
- Henoch Schonlein Purpura
- Inflammatory bowel disease - Crohn's disease or Ulcerative colitis
- Mixed connective tissue disease
- Multiple Sclerosis
- Myasthenia Gravis
- Myositis - Polymyositis, Dermatomyositis or Inclusion body myositis
- Pemphigus
- Pernicious anemia
- Polyarteritis nodosa
- Primary biliary cirrhosis
- Psoriasis
- Rheumatoid arthritis or juvenile rheumatoid arthritis
- Scleroderma (Systemic sclerosis)
- Sjogren's Syndrome
- Systemic Lupus Erythematosus (Lupus)
- Thyroiditis
- Type 1 diabetes
- Vitiligo
- Wegener's Granulomatosis
- Other, please specify below
- None
- No information available

Other (please specify) _____

**Appendix B. List of muscle diseases, adult or juvenile onset, to be considered for family history question 2O. Specify in the comments box.**

- Drug or toxin associated myopathy
- Endocrine myopathy
- Infectious myopathy
- Metabolic myopathy
- Mitochondrial myopathy
- Motor neuron disease
- Muscular Dystrophy
- No information available
- None
- Non-inflammatory inclusion body myopathy

Other, please specify _____