

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

## Proposed Data Collection Form and Preliminary Variable List for Discussion

### GENERAL INFORMATION

**Clinician submitting case:**

**Case number:**

**Gender:**

**Age at onset:**

**Age at diagnosis**

**Ethnicity:**

**Study Diagnosis according to the clinician:**

**Basis for Study Diagnosis (list all supporting reasons):**

**Clinician's additional comments on the case:**

**Other diagnoses in this case:**

	<b>Present</b>	<b>Absent</b>	<b>Not available</b>	<b>Comments</b>
<b>Clinical Muscle Variables</b>				
1M. Muscle weakness of proximal upper extremities				
2M. Wrist or finger flexor weakness				
3M. Relative sparing of the deltoid/shoulder abductors (e.g., wrist and/prox finger flexors weaker on MRC scale compared to shoulder abductors)				
4M. Muscle weakness of proximal lower extremities				
5M. Hip abductor weakness				
6M. Muscle weakness of distal lower extremities				
7M. Knee extensors as weak or weaker than hip girdle muscles				
8M. Neck flexor weakness				
9M. Neck extensor weakness				
10M. Symmetrical muscle weakness				
11M. Muscle pain at rest				
12M. Muscle tenderness				
13M. Muscle atrophy of distal forearms				
14M. Muscle atrophy of thighs				

	<u>Present</u>	<u>Absent</u>	<u>Not available</u>	<u>Comments</u>
<b>Skin Variables</b>				
1S. Heliotrope rash				
2S. Gottron's papules				
3S. Erythema on extensor surfaces of extremity joints				
4S. Erythema of neck (V-sign)				
5S. Erythema of back of neck and shoulders (Shawl sign)				
6S. Cutaneous or muscular calcification				
7S. Periungual erythema, petechiae, telangiectasia, or epidermal hyperplasia				
8S. Raynaud's phenomenon				
<b>Other Clinical Variables</b>				
10. First degree family history of defined autoimmune disease				
20. Acute onset (weeks to several months) of symptoms				
30. Arthritis				
40. Polyarthralgia				
50. Sjögren's syndrome				
60. Systemic sclerosis				
70. MCTD				
80. Rheumatoid arthritis				
90. Systemic lupus erythematosus				
100. Autoimmune thyroid disease				
110. Objective improvement in strength after corticosteroid therapy				
120. No objective improvement in strength after corticosteroid therapy				
130. Fevers				
140. Interstitial lung disease				
150. Dysphagia				

<b>Laboratory Variables</b>	<b><u>Value</u></b>	<b><u>Normal or expected range</u></b>	<b><u>Comments</u></b>
1L. EMG- increased spontaneous activity (fibrillation potentials, positive sharp waves, complex repetitive discharges)			
2L. Serum CK (CPK) activity			
3L. Serum LD (LDH) activity			
4L. Serum ASAT/AST/SGOT activity			
5L. Serum ALAT/ALT/SGPT activity			
6L. Serum Aldolase activity			
7L. Anti-nuclear autoantibodies (ANA)			
8L. Anti-Jo-1 autoantibodies			
9L. STIR/T2 MRI c/w inflammation			
10L. ESR			
11L. C-reactive protein			
12L. ENA autoantibodies			
13L. Other Myositis specific autoantibody			
<b>Muscle Biopsy Variables</b>	<b><u>Present</u></b>	<b><u>Absent Not available</u></b>	<b><u>Comments</u></b>
1B. Inflammatory cell infiltrates and other changes consistent with myositis (degen/regen, etc) from basic H&E staining			
2B. MHC I antigen present on scattered muscle fibers			
3B. Non-necrotic fibers surrounded and invaded by MNC			
4B. Endomysial MNC infiltrates			
5B. Perivascular MNC infiltrates			
6B. Perimysial MNC infiltrates			
7B. Perifascicular atrophy			
8B. Rimmed vacuoles			