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:

	Present	**Absent**	**Not available**	**Comments**
Clinical Muscle Variables				
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				

	Present	Absent	Not available	Comments
Skin Variables				
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				
Other Clinical Variables				
1O. First degree family history of defined autoimmune disease				
2O. Acute onset (weeks to several months) of symptoms				
3O. Arthritis				
4O. Polyarthralgia				
5O. Sjögren´s syndrome				
6O. Systemic sclerosis				
7O. MCTD				
8O. Rheumatoid arthritis				
9O. Systemic lupus erythematosus				
10O. Autoimmune thyroid disease				
11O. Objective improvement in strength after corticosteroid therapy				
12O. No objective improvement in strength after corticosteroid therapy				
13O. Fevers				
14O. Interstitial lung disease				
15O. Dysphagia				

Laboratory Variables	Value	Normal or expected range	Comments
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			
Muscle Biopsy Variables	**Present**	**Absent Not available**	**Comments**
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			