

IMACS FORM 01A: CORE PATIENT DATA

To be completed at study entry only

Subject's IMACS number _____

Assessor _____

Date of assessment (mm/dd/yy) _____

Assessment number _____

Type of Study: Therapeutic Trial _____ Natural History Study _____ Other _____

Name of Study: _____

Age at time of Enrollment: ____ Years ____ Months Gender: Male ____ Female ____

Ethnicity: ____ Hispanic or Latino ____ Not Hispanic or Latino ____ Unknown/Not reported

Race: Check all that apply:

- White or Caucasian
- African-American or Black
- Asian or Asian American, Pacific Islander
- Native American or Alaskan Native
- Other _____ (please specify)
- Unknown/Not reported

Date patient first noticed first myositis symptom (mm/yy): _____/_____/_____

Date patient first noticed muscle weakness (mm/yy): _____/_____/_____

Date diagnosis made (mm/yy): _____/_____/_____

Date of first appropriate treatment for IIM (mm/yy): _____/_____/_____

Myositis Criteria

Criteria for diagnosis of PM/DM (check all that apply):

____ Absence of other forms of myopathy, including inclusion body, metabolic, inherited or infectious forms

____ Symmetric proximal muscle weakness

____ Rash consisting of heliotrope and/or Gottron's papules/sign

____ Elevation in serum skeletal muscle enzymes. Specify maximum value (include upper limit of normal):

Enzyme	Maximum Value	Upper Limit Normal
Creatine Kinase		
Aldolase		
Lactate dehydrogenase		
Aspartate aminotransferase (AST, SGOT)		
Alanine aminotransferase (ALT, SGPT)		

____ EMG findings consistent with myositis

____ Muscle biopsy findings consistent with myositis

Criteria for IBM (check all that apply): (Completed only in IBM patients)

Characteristic Features – Inclusion Criteria

A. Clinical features:

____ 1. Duration of illness > 6 months

____ 2. Age of onset > 30 years

- ___ 3. Muscle weakness must affect proximal and distal muscles of arms and legs *and* patient must exhibit at least one of the following features:
- ___ a. Finger flexor weakness
 - ___ b. Wrist flexor > wrist extensor weakness
 - ___ c. Quadriceps muscle weakness (= or < grade 4 MRC)

B. Laboratory features:

- ___ 1. Serum creatine kinase < 12 times normal
- ___ 2. Muscle biopsy:
 - ___ a. Inflammatory myopathy characterized by mononuclear cell invasion of nonnecrotic muscle fibers
 - ___ b. Vacuolated muscle fibers
 - ___ c. Either
 - ___ (i) intracellular amyloid deposits (must use fluorescent method of identification before excluding the presence of amyloid) or
 - ___ (ii) 15 – 18-nm tubulofilaments by electron microscopy
- ___ 3. Electromyography must be consistent with features of an inflammatory myopathy (however, long duration potentials are commonly observed and do not exclude diagnosis of sporadic inclusion body myositis)

Myositis Clinical and Serologic Group

Myositis Primary Clinical Group:

apply:

- Adult OR Juvenile
- Polymyositis
 - Dermatomyositis
 - Inclusion body myositis
 - Other: please clarify _____

Does the patient have Overlap Myositis, defined by myositis plus another defined connective tissue or autoimmune disease? ___ Yes ___ No,
 If yes, which other connective tissue or autoimmune diseases? _____

Does the patient have Cancer associated myositis? (i.e., Diagnosed with cancer, excluding focal squamous cell carcinoma of the skin or focal cervical carcinoma or prostate carcinoma in situ) within 2 years of myositis diagnosis)
 ___ Yes ___ No. ; If yes, which cancer _____

Severity of Myositis at Onset:

___ 1 = mild ___ 2 = Moderate ___ 3 = Severe ___ 4 = Extremely severe

Autoantibodies Tested at Any Time During Illness Course:

Autoantibody	Negative	Positive	Not tested	Assay used*
ANA				
Jo-1				
SRP				
Mi-2				
U1RNP				
Ro				
La				
Ku				
PM-Scl				
Other _____				

*Please specify ELISA, Immunodiffusion, Immunoprecipitation, Immunofluorescence, Unknown or other

**IMACS FORM 01B: CORE PATIENT DATA
To Be Completed At Each Assessment**

Subject's IMACS number _____

Assessor _____

Date of assessment (mm/dd/yy) _____

Assessment number _____

Weight (kg): _____ Height (cm): _____

Patient's Other Diagnoses (Top 5) (Co-Morbid Conditions)

1. _____
2. _____
3. _____
4. _____
5. _____

Apparent Clinical Course (check all that apply):

____ Monocyclic: full recovery within 2 years without relapse (with or without drug therapy)

____ Chronic polycyclic: prolonged, relapsing course with one or more relapses occurring between periods of inactive disease

____ Chronic continuous: persistent disease for longer than 2 years despite drug therapy and which is never inactive

____ Undefined (illness < 2 years)

____ Other: _____

ACR Functional Status (1991 revised):

Circle worst grade ever (see definitions below): I II III IV

Circle current grade (see definitions below) I II III IV

- I. Completely able to perform usual activities of daily living (self care, vocational and avocational);
- II. Able to perform usual self-care and vocational activities, but limited avocational activities;
- III. Able to perform usual self-care activities, but limited in vocational and avocational activities;
- IV. Limited in ability to perform usual self-care, vocational and avocational activities.

If patient's disease is currently active, specify the duration of active disease

for this episode/flare of activity to present time: _____ months

Duration of active disease from diagnosis to present assessment (exclude periods of inactive disease) _____ months

If patient's myositis is currently completely inactive,

A. How many months has the patient's myositis been inactive (based on clinical and laboratory assessment), with or without medication?

_____ mo.

B. If the patient is not taking medications now, how many months has the patient's myositis been inactive (based on clinical and laboratory assessment) off all medications for myositis (i.e. in remission)? _____ mo.

Signs/Symptoms During Illness Course: Were the following present ever during the illness course?

Sign/Symptom	Ever Present?	
	Present	Absent
Pericarditis/myocarditis		
Arrhythmia		
Interstitial lung disease		
Dysphagia		
Dysphonia		
GI ulceration		
Cutaneous ulceration		
Erythroderma (extensive areas of confluent erythema, both sun exposed and non-sun exposed skin; can involve entire body)		
Calcinosis		
Arthritis		
Other thought important to prognosis <i>Specify:</i>		
Other thought important to prognosis <i>Specify:</i>		
Other thought important to prognosis <i>Specify:</i>		

<u>Medications</u>	Ever	Current	Unknown	Current Dose (mg)
Nonsteroidal Anti-Inflammatory Drugs or COX-2 inhibitors	<input type="checkbox"/>	<input type="checkbox"/>	<input type="checkbox"/>	/day
Prednisone, Oral (Prednisolone, Medrol, other corticosteroids)	<input type="checkbox"/>	<input type="checkbox"/>	<input type="checkbox"/>	/day
Intravenous methylprednisolone	<input type="checkbox"/>	<input type="checkbox"/>	<input type="checkbox"/>	/ infusion Frequency of infusion:
Topical steroids	<input type="checkbox"/>	<input type="checkbox"/>	<input type="checkbox"/>	/ day
Topical tacrolimus (Protopic) or picrolimus	<input type="checkbox"/>	<input type="checkbox"/>	<input type="checkbox"/>	/day
<i>Disease Modifying Antirheumatic Drugs (DMARDS)</i>	<input type="checkbox"/>	<input type="checkbox"/>	<input type="checkbox"/>	
Methotrexate Oral__ Subcutaneous__ IM__ IV__	<input type="checkbox"/>	<input type="checkbox"/>	<input type="checkbox"/>	/week
Hydroxychloroquine (Plaquenil)	<input type="checkbox"/>	<input type="checkbox"/>	<input type="checkbox"/>	/ day
Azathioprine (Imuran)	<input type="checkbox"/>	<input type="checkbox"/>	<input type="checkbox"/>	/day
Cyclosporin A (Sandimmune or Neoral)	<input type="checkbox"/>	<input type="checkbox"/>	<input type="checkbox"/>	/day
Tacrolimus (FK 506)	<input type="checkbox"/>	<input type="checkbox"/>	<input type="checkbox"/>	/day
Leflunomide (ARAVA)	<input type="checkbox"/>	<input type="checkbox"/>	<input type="checkbox"/>	/day
Cyclophosphamide (Cytoxan) IV	<input type="checkbox"/>	<input type="checkbox"/>	<input type="checkbox"/>	/month
Cyclophosphamide (Cytoxan) po	<input type="checkbox"/>	<input type="checkbox"/>	<input type="checkbox"/>	/day
Etanercept (Enbrel)	<input type="checkbox"/>	<input type="checkbox"/>	<input type="checkbox"/>	/week
Infliximab (Remicaide)	<input type="checkbox"/>	<input type="checkbox"/>	<input type="checkbox"/>	/infusion Frequency of infusion:
Kineret (Anakinra)	<input type="checkbox"/>	<input type="checkbox"/>	<input type="checkbox"/>	/day
Intravenous gammaglobulin (IVIG)	<input type="checkbox"/>	<input type="checkbox"/>	<input type="checkbox"/>	/month
Adalimumab (Humira)	<input type="checkbox"/>	<input type="checkbox"/>	<input type="checkbox"/>	/every other week
Mycophenolate mofetil (MMF)	<input type="checkbox"/>	<input type="checkbox"/>	<input type="checkbox"/>	/day
Rituximab (anti-CD20)	<input type="checkbox"/>	<input type="checkbox"/>	<input type="checkbox"/>	/infusion Number of infusions:
Herbal or Nutritional Supplements - specify	<input type="checkbox"/>	<input type="checkbox"/>	<input type="checkbox"/>	/day wk mth
Other drugs or biologic agents: Specify	<input type="checkbox"/>	<input type="checkbox"/>	<input type="checkbox"/>	/day wk mth
Other drugs or biologic agents: Specify	<input type="checkbox"/>	<input type="checkbox"/>	<input type="checkbox"/>	/day wk mth
Other drugs or biologic agents: Specify	<input type="checkbox"/>	<input type="checkbox"/>	<input type="checkbox"/>	/day wk mth
Other drugs or biologic agents: Specify	<input type="checkbox"/>	<input type="checkbox"/>	<input type="checkbox"/>	/day wk mth
Other drugs or biologic agents: Specify	<input type="checkbox"/>	<input type="checkbox"/>	<input type="checkbox"/>	/day wk mth
Other treatment: Specify	<input type="checkbox"/>	<input type="checkbox"/>	<input type="checkbox"/>	/day wk mth