

IMACS FORM 07b: MYOSITIS DISEASE ACTIVITY ASSESSMENT TOOL, Version 2 - 2005

Subject's IMACS number: _____ ASSESSOR: _____ Date Assessed: _____ Assessment number: _____

The clinical features recorded are based upon the previous 4 weeks and the judgment that the feature is DUE TO THE MYOSITIS DISEASE PROCESS and represents disease activity, which is potentially reversible. Please rate your overall assessment of the ongoing current disease activity for each of the systems below by drawing a vertical mark on the 10-cm. line for each system according to the following scale: left end of line = no evidence of disease activity, midpoint of line = moderate disease activity, and right end of line = extreme or maximum disease activity. Under each organ system is a set of clinical and laboratory findings that you should assess in order to determine the VAS disease activity score for each organ system.

For each individual item, also rate: A = very active, requiring disease modifying therapy; B = moderate disease activity / therapy; C = mild and stable disease activity / therapy; D = resolved (no therapy required); E = never present; NA= cannot be assessed.

Please review all instructions and the GLOSSARY for definitions prior to assessment (see pages 4 - 12).

CONSTITUTIONAL DISEASE ACTIVITY	(Absent) (Maximum) 	Maximum Value Guidelines (Examples of maximal score) Severe fatigue or malaise resulting in being bed bound and an inability to perform self care
1. Pyrexia – documented fever > 38 deg. C.		A B C D E NA
2. Weight loss – unintentional > 5%		A B C D E NA
3. Fatigue/malaise/lethargy		A B C D E NA
4. Lymphadenopathy (> 1 cm)		A B C D E NA
CUTANEOUS DISEASE ACTIVITY	(Absent) (Maximum) 	Maximum Value Guidelines (Examples of maximal score) - Ulceration to muscle, tendon or bone; - Extensive erythroderma
5. Cutaneous ulceration		A B C D E NA
6. Erythroderma		A B C D E NA
7. Erythematous rashes accompanied by vesiculobullous change, erosions or necrosis		A B C D E NA
8. Panniculitis		A B C D E NA
9. Erythematous rashes without secondary changes		B C D E NA
10. Heliotrope rash		B C D E NA
11. Gottron's papules/sign		B C D E NA
12. Periungual capillary changes		B C D E NA
13. Alopecia:		B C D E NA
• Diffuse hair loss = category B (or category C if mild and stable)		
• Focal, patchy with erythema = category C		
14. Mechanic's hands		B C D E NA
SKELETAL DISEASE ACTIVITY	(Absent) (Maximum) 	Maximum Value Guidelines (Examples of maximal score) - Severe arthritis with extreme loss of function (bedridden, inability for self care)
15. Arthritis:		A B C D E NA
• Severe active polyarthritis with major loss of function = category A		
• Moderately active arthritis with moderate loss of function = category B		
• Mild arthritis with no to mild loss of function = category C		
16. Polyarthralgias		C D E NA

Subject's IMACS number: _____ ASSESSOR: _____ Date Assessed: _____ Assessment number: _____

<u>GASTROINTESTINAL DISEASE ACTIVITY</u>	(Absent)	(Maximum)	<u>Maximum Value Guidelines (Examples of maximal score)</u>			
	----- -----		Major abdominal crisis requiring surgery or intensive care			

17. Gastrointestinal ulceration	A	B	C	D	E	NA
18. Dysphagia:	A	B	C	D	E	NA
• Moderate/severe dysphagia = category A						
• Mild dysphagia = category B (or category C if mild and stable)						
19. Abdominal pain:	A	B	C	D	E	NA
• Severe = category A						
• Moderate = category B						
• Mild = category C						

<u>PULMONARY DISEASE ACTIVITY</u>	(Absent)	(Maximum)	<u>Maximum Value Guidelines (Examples of maximal score)</u>			
	----- -----		Active interstitial lung disease or respiratory muscle weakness requiring ventilatory support			

20. Respiratory muscle weakness without intrinsic lung disease:	A	B	C	D	E	NA
♦ Dyspnea at rest = category A (or category B if moderate)						
♦ Dyspnea on exertion = category B (or category C if mild and stable)						
21. Active interstitial lung disease (ILD), WITH previously documented ILD by radiography or pathology: <i>Mark only one of category A - C, using pulmonary function tests for the primary assessment when available:</i>	A	B	C	D	E	NA
• Shortness of breath at rest <u>OR</u> FVC (Forced Vital Capacity) or DLCO (Carbon monoxide diffusion capacity) 50% predicted <u>OR</u> requirement for oxygen use or assisted ventilation = category A						
• Shortness of breath with normal activity <u>OR</u> FVC or DLCO 50 – 70% predicted = category B						
• Shortness of breath with exertional activity <u>OR</u> FVC or DLCO > 70% predicted = category C						
22. Dysphonia:		B	C	D	E	NA
• Moderate to severe = category B						
• Mild = category C						

<u>CARDIAC DISEASE ACTIVITY</u>	(Absent)	(Maximum)	<u>Maximum Value Guidelines (Examples of maximal score)</u>			
	----- -----		Myocarditis or pericarditis requiring intensive unit care			

23. Myocarditis or Pericarditis:	A	B	C	D	E	NA
• Symptomatic or with physical exam changes = category A						
• In the absence of clinical features = category B (or category C if mild and stable)						
24. Arrhythmia:	A	B	C	D	E	NA
• Severe arrhythmia = category A						
• Other arrhythmia, except sinus tachycardia = category B (or category C if mild and stable)						
25. Sinus tachycardia			C	D	E	NA

Subject's IMACS number: _____ ASSESSOR: _____ Date Assessed: _____ Assessment number: _____

<u>OTHER DISEASE ACTIVITY</u>	(Absent) _____ (Maximum)	Maximum Value Guidelines (Examples of maximal score) Extreme disease activity with major impact on function
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Specify: _____

A B C D E NA

Specify: _____

A B C D E NA

<u>GLOBAL EXTRA-SKELETAL MUSCLE DISEASE ACTIVITY</u>	(Absent) _____ (Maximum)	Overall evaluation for the totality of disease activity in all extra-skeletal muscle systems (EXCLUDING MUSCLE DISEASE ACTIVITY)
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<u>MUSCLE DISEASE ACTIVITY</u>	(Absent) _____ (Maximum)	Maximum Value Guidelines (Examples of maximal score) Severe muscle weakness resulting in being bed bound and an inability to perform self care
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26. Myositis:

- Severe muscle inflammation = category A
- Moderate muscle inflammation = category B
- Mild stable muscle inflammation = category C

A B C D E NA

27. Myalgia

C D E NA

<u>GLOBAL DISEASE ACTIVITY</u>	(Absent) _____ (Maximum)	Overall evaluation for the totality of disease activity in ALL systems, (INCLUDING MUSCLE DISEASE ACTIVITY)
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MYOSITIS DISEASE ACTIVITY ASSESSMENT TOOL, Version 2 – 2005

This is a combined tool that captures the physician's assessment of disease activity of various organ systems via the
MYOSITIS DISEASE ACTIVITY ASSESSMENT VISUAL ANALOGUE SCALES (MYOACT)

and the presence or absence of clinical features via the

MYOSITIS INTENTION TO TREAT ACTIVITY INDEX (MITAX)

MYOSITIS DISEASE ACTIVITY ASSESSMENT VISUAL ANALOGUE SCALES (MYOACT)

(Modified from the Vasculitis Activity Index, QE Whiting-O'Keefe et al. 1999, Arthritis and Rheumatism 42: 2365-71)

Please assess the following systems using your expert clinical judgement of how active the patient's myositis has been WITHIN THE PAST FOUR WEEKS. Disease activity is defined as potentially reversible clinically evident pathology or physiology resulting from the myositis disease process. **Clinical findings known or suspected to be due to another disease process or due to therapy should not be considered in this evaluation.** Please rate your overall assessment of the ongoing current disease activity for each of the systems listed by drawing a vertical mark on the 10-cm. line for each system according to the following scale: left end of line = no evidence of disease activity, midpoint of line = moderate disease activity, and right end of line = extreme or maximum disease activity. Please write in NA if the system cannot be assessed. Under each organ system is listed a set of clinical and laboratory findings that you should assess in order to determine the VAS disease activity score for each organ system.

MYOACT scoring System: The proposed scoring system for the MYOACT is the sum of the 10 cm visual analogue scale scores for each of the six individual organ systems (CONSTITUTIONAL, CUTANEOUS, SKELETAL, GASTROINTESTINAL, PULMONARY, CARDIAC) and divided by the total possible score (range = 0 - 60). If one or more organ systems were not assessed, the resulting score would be divided by the maximum possible score from those assessed items. The categories of OTHER DISEASE ACTIVITY, GLOBAL EXTRASKELETAL MUSCLE, MUSCLE and GLOBAL DISEASE ACTIVITY are not included in the MYOACT score but are scored separately.

MYOSITIS INTENTION TO TREAT ACTIVITY INDEX (MITAX)

(Modified BILAG Approach, EM Hay et al., 1993, Q J Med 86: 447-58)

The clinical features recorded are based upon:

1. the presence of clinical features at this time or symptoms **within the previous four weeks that are due to active disease, and**
2. the presumption that the feature is due to the myositis syndrome disease process

For each item of the MITAX, please choose (circle one choice) among the following options:

- A - If the feature is thought to be sufficiently active to require disease-modifying treatment
- B - If the feature is less active than A; moderate activity requiring moderate therapies
- C - If the feature stable or mild, requiring only symptomatic therapy
- D - If the feature has resolved (no medication is required)
- E - If the feature has never been present
- NA - if the feature cannot be assessed

MITAX scoring system): The scoring system is based primarily on the physician's intention to treat (Categories A – E). In some cases, a patient may meet the clinical manifestations of a particularly category, but the treatment does not match the intention to treat of that particular category; in those cases, the category matching the clinical symptoms should be marked. If more than one clinical symptom is present within a system, and different categories would be scored for each, mark the category that is most severe.

Scoring system for the MITAX:

CATEGORY A A = <u>A</u>ctive (9 points)	Denotes disease thought to be sufficiently active to require disease-modifying treatment: [High dose daily corticosteroids alone or in combination with high doses of other immunosuppressives or intravenous gammaglobulin (IVIG)]
CATEGORY B B = <u>B</u>eware (3 points)	Denotes disease that is less active than in "A"; reversible problems requiring moderate doses of corticosteroids and other immunosuppressive agents, or use of antimalarials, non-steroidal anti-inflammatory drugs (NSAIDs), topical steroids or tacrolimus. [If immunosuppressives or IVIG were used to treat signs and symptoms of Category A, the doses of at least one agent would be reduced from levels required in category A.]
CATEGORY C C = <u>C</u>ontentment (1 point)	Indicates stable mild disease, requiring only symptomatic therapy, such that immunosuppressive therapy can be reduced below the level of category B. [Doses of corticosteroids, other immunosuppressive agents, antimalarials, NSAIDs or symptomatic therapies are likely to be low.]
CATEGORY D D = <u>D</u>iscount (0 points)	System previously affected but currently inactive and no medication is required (i.e., in remission)
CATEGORY E E = <u>N</u>o evidence of this system being active now or previously (0 points)	Indicates system never involved

Each organ system of the MITAX is scored by selecting the highest category selected for any of the individual items under that organ system (i.e., the worse item carries the score for the organ system). The proposed scoring system for the MITAX is the sum of the worst category scores for each of the seven individual organ systems (CONSTITUTIONAL, CUTANEOUS, SKELETAL, GASTROINTESTINAL, PULMONARY, CARDIAC, MUSCLE) divided by the maximum possible score (range = 1 - 63). If one or more organ systems were not assessed, the score would be calculated by dividing the sum by the maximum possible score of the assessed organ systems. The categories of OTHER DISEASE ACTIVITY, GLOBAL EXTRASKELETAL MUSCLE and the GLOBAL DISEASE ACTIVITY are not included in the MITAX score but are scored separately.

APPENDIX 1: GLOSSARY for the MYOSITIS DISEASE ACTIVITY ASSESSMENT TOOL, Version 2- 2005

Please Review Glossary for definitions prior to assessment.

CONSTITUTIONAL

1. *Pyrexia*: refers to a documented fever in excess of 38°C.
2. *Weight loss*: unintentional weight loss of greater than 5% of body weight.
3. *Fatigue*: sufficiently severe to interfere with normal daily activities and not attributable to another cause.

CUTANEOUS DISEASE ACTIVITY

5. *Cutaneous ulceration*: primary injury to dermis or deeper, due to dermatomyositis.
6. *Erythroderma*: generalized widespread confluent erythema, involving both sun-exposed and non-sun-exposed skin.
7. *Erythematous rashes*: including assessment of malar rash, facial erythema, linear extensor erythema, V and shawl-sign rash, erythema in a periungual distribution, and non-sun-exposed erythema.
Secondary changes:
Necrosis: epidermal necrosis manifesting as a superficial bullous lesion whose roof is gray in color from epidermal cell death, erosions, or scale/crusts, sitting on an erythematous base. These lesions are painful when acute.
8. *Panniculitis*: tender erythematous or violaceous nodules or depressions due to inflammation in the subcutaneous fat.
10. *Heliotrope rash*: purple, lilac-colored or erythematous patches over eyelids or in a periorbital distribution, often associated with periorbital edema.
11. *Gottron's papules or sign*: erythematous to violaceous papules, plaques or macules (sign) over extensor surfaces of joints, which are sometimes scaly.
12. *Periungual capillary changes*: dilatation of periungual capillaries which may be accompanied by vessel dropout and which is visible by naked eye examination or by using additional magnification with otoscopy.
13. *Alopecia*: hair loss.
Diffuse = Nonscarring, non-erythematous widespread alopecia (telogen effluvium)
Focal = Focal patchy alopecia with erythema (localized to areas of inflammation)
14. *Mechanic's hands*: hyperkeratosis and scaling with frequent fissuring and cracking along the lateral and palmar aspects of the fingers.

SKELETAL DISEASE ACTIVITY

15. *Arthritis*: active joint inflammation marked by tenderness, warmth or swelling.
 Category A = *Severe active polyarthritis*: arthritis of 2 or more joints with clinically significant loss of the functional range of movement of the involved joints and requiring assistance with activities of daily living.
 Category B = *Moderately active arthritis*: arthritis of one or more joints with some loss of functional range of movement of the involved joints, but not requiring assistance with activities of daily living
 Category C = *Mildly active arthritis*: arthritis of one or more joints with neither loss of range of motion nor impaired activities of daily living.
16. *Polyarthralgias*: joint pain with stiffness due to active disease in 2 or more joints.

GASTROINTESTINAL DISEASE ACTIVITY

17. *Gastrointestinal ulceration*: Clinical ulceration (severe abdominal pain with GI blood loss) or documented by radiographic study (includes pneumatosis intestinalis) or by biopsy.
18. *Dysphagia*: difficulty swallowing, chewing or eating documented by clinical symptoms or by modified barium swallow examination, manometry, or other objective approaches.

If dysphagia has been stable and unchanged and is present for > 6 months, this most likely represents damage and not activity, unless there is good evidence of reversibility.

Category A = *Severe dysphagia*: accompanied by aspiration pneumonia, nasal regurgitation, or difficulty in protecting the airway;

Category B = *Moderate dysphagia*: frequent or moderate symptomatic difficulty swallowing, chewing or eating.

Category C = *Mild dysphagia*: occasional dysphagia or subclinical dysphagia noted on objective testing; can eat regular diet.

19. *Abdominal pain*: Pain in the abdominal area due to the myositis disease process

Category A = *Severe abdominal pain*, requiring hospitalization, treatment, or bowel rest with nothing per oral route (NPO)

Category B = *Moderate abdominal pain*, requiring treatment, but not hospitalization or NPO

Category C = *Mild abdominal pain* - no intervention required and does not interfere with function.

PULMONARY DISEASE ACTIVITY

20. *Respiratory muscle weakness without intrinsic lung disease*: Shortness of breath, dyspnea worsened with exertion or hypoventilation on pulmonary function testing **without** intrinsic lung disease.

21. *Interstitial lung disease*: Previously documented by radiography or pathology. Primary assessment of *lung function* should be by pulmonary function tests when available, but can be assessed by physical exam when pulmonary function testing not available. Only active features are graded, not those based on pulmonary fibrosis or irreversible features.

22. *Dysphonia*: alteration in voice quality, resonance, articulation or speech rate from normal

Moderate to severe dysphonia = persistent voice symptoms or those that interfere with communication

Mild dysphonia = intermittent voice symptoms not interfering with communication

CARDIOVASCULAR DISEASE ACTIVITY

23. *Myocarditis or pericarditis*: Inflammation of the myocardium or pericardium defined clinically or with echocardiographic or pathologic evidence.

24. *Arrhythmia*: clinical or electrocardiographic evidence of irregular heart beats

Severe arrhythmia = symptomatic and requiring therapy or other intervention, excluding sinus tachycardia.

Other cardiac arrhythmias = symptomatic, but not requiring intervention, excluding sinus tachycardia

OTHER DISEASE ACTIVITY

Please specify features not listed above felt to be due to the myositis disease process. Then on the VAS scale rate the severity of the feature(s).

MUSCLE DISEASE ACTIVITY

26. *Myositis*: Muscle inflammation based upon manual muscle strength testing, functional assessments, laboratory or other testing. In patients with stable muscle atrophy and muscle damage, only weakness attributable to active myositis is scored.

Category A = Severe muscle inflammation, requiring assistance with activities of daily living and severe loss of function

Category B = Moderate muscle inflammation: not requiring assistance with activities of daily living with moderate loss of function

Category C = Stable mild muscle inflammation: little or no loss of function

27. *Myalgia*: muscle pain or tenderness.

APPENDIX 2: MITAX SCORING A – E SYSTEM, VERSION 2- 2005

1) CONSTITUTIONAL DISEASE ACTIVITY

CATEGORY A:

Pyrexia (>38°C) **plus**
Two other constitutional features
Unintentional weight loss
Fatigue/malaise/lethargy
Lymphadenopathy (>1cm)

CATEGORY B

Any ONE constitutional feature of moderate activity requiring category B therapy (i.e., moderate doses of corticosteroids other immunosuppressive agents or use of antimalarials, NSAIDs or topical steroids or tacrolimus).

CATEGORY C

Presence of any one constitutional feature that is mild and stable, requiring only symptomatic therapy or therapy at doses below the level required in category B

CATEGORY D

Previously involved and no medication is currently required (in remission).

CATEGORY E

No previous involvement

2) CUTANEOUS DISEASE ACTIVITY

CATEGORY A

Any one of the following features with severe activity requiring category A therapy (disease modifying treatment in high dose):

1. Cutaneous ulceration
2. Erythroderma
3. Erythema accompanied by vesiculobullous or erosive changes or necrosis
4. Panniculitis

CATEGORY B

Any one of the following features with moderate activity requiring category B therapy (i.e., moderate doses of corticosteroids other immunosuppressive agents or use of antimalarials, NSAIDs or topical steroids or tacrolimus):

1. Any Category A cutaneous sign with moderate activity with therapy at doses below the level required in category A
2. Erythema without secondary changes
3. Heliotrope rash
4. Gottron's papules or sign
5. Periungual capillary changes
6. Alopecia, diffuse
7. Mechanic's hands

CATEGORY C

Any one of the following features with mild and stable activity requiring category C therapy:

1. Category A or B cutaneous signs with mild and stable activity, requiring only symptomatic therapy or therapy at doses below the level required in category B
2. Alopecia, focal patchy with erythema

CATEGORY D

Previously involved and no medication is currently required (in remission).

CATEGORY E

No previous involvement

3) SKELETAL DISEASE ACTIVITY**CATEGORY A**

Severe active polyarthritis with major loss of function (requiring assistance with activities of daily living), requiring category A therapy (disease modifying treatment in high dose).

CATEGORY B

Moderately active arthritis with moderate loss of function requiring category B therapy (i.e., moderate doses of corticosteroids other immunosuppressive agents or use of antimalarials, NSAIDs or topical steroids or tacrolimus).

CATEGORY C

Any one of:

1. Mild arthritis with no to mild loss of function, requiring only symptomatic therapy or therapy at doses below the level required in category B
2. Polyarthralgias

CATEGORY D

Previously involved and no medication is currently required (in remission).

CATEGORY E

No previous involvement

4) GASTROINTESTINAL DISEASE ACTIVITY

CATEGORY A

Any one of the following features with severe activity requiring category A therapy (disease modifying treatment in high dose):

1. Gastrointestinal ulceration (with abdominal pain or GI blood loss)
2. Moderate to severe dysphagia
3. Severe abdominal pain [requiring hospitalization, treatment, or bowel rest with nothing per oral route (NPO)].

CATEGORY B

Any one of the following features with moderate activity requiring category B therapy (i.e., moderate doses of corticosteroids other immunosuppressive agents or use of antimalarials, NSAIDs or topical steroids or tacrolimus):

1. Category A gastrointestinal signs or symptoms of moderate activity with therapy at doses below the level required in category A
2. Mild dysphagia
3. Moderate abdominal pain [requiring treatment, but not hospitalization or NPO].

CATEGORY C

Any one of the following features with mild and stable activity requiring category C therapy:

1. Category A or B gastrointestinal signs or symptoms with mild and stable activity, requiring only symptomatic therapy or therapy at doses below the level required in category B
2. Mild abdominal pain [no intervention required and does not interfere with function]

CATEGORY D

Previous involvement and no medication currently required (i.e., in remission)

CATEGORY E

No previous involvement

5) PULMONARY DISEASE ACTIVITY

CATEGORY A

Any one of the following clinical features with severe activity requiring category A therapy (disease modifying treatment in high dose):

1. Dyspnea at rest from muscle weakness
2. Active interstitial lung disease (ILD) with shortness of breath at rest or FVC (Forced Vital Capacity) or DLCO (Carbon monoxide diffusion capacity) < 50% predicted or requirement for oxygen use or assisted ventilation

CATEGORY B

Any one of the following clinical features with moderate activity requiring category B therapy (i.e., moderate doses of corticosteroids other immunosuppressive agents or use of antimalarials, NSAIDs or topical steroids or tacrolimus):

1. Category A pulmonary signs or symptoms of moderate disease activity with therapy at doses below the level required in category A
2. Dyspnea on exertion from muscle weakness
3. Active ILD with shortness of breath with normal activity or FVC or DLCO 50– 70% predicted
4. Moderate to severe dysphonia

CATEGORY C

Any one of the following clinical features with mild and stable activity requiring category C therapy:

1. Category A or B pulmonary signs or symptoms which are mild and stable activity, requiring only symptomatic therapy or therapy at doses below the level required in category B
2. Active ILD with shortness of breath with exertional activity or FVC or DLCO > 70% predicted
3. Mild dysphonia

CATEGORY D

Previous involvement and no medication currently required (i.e., in remission)

CATEGORY E

No previous involvement

6) CARDIOVASCULAR DISEASE ACTIVITY

CATEGORY A

Any one of the following clinical features with severe activity requiring category A therapy (disease modifying treatment in high dose):

1. Echocardiography or biopsy evidence of myocarditis or pericarditis with clinical symptoms
2. Severe cardiac arrhythmias

CATEGORY B

Any one of the following clinical features with moderate activity requiring category B therapy (i.e., moderate doses of corticosteroids other immunosuppressive agents or use of antimalarials, NSAIDs or topical steroids or tacrolimus):

1. Category A cardiovascular signs or symptoms with moderate activity with therapy at doses below the level required in category A
2. Myocarditis or pericarditis without clinical signs
3. Other cardiac arrhythmias

CATEGORY C

Any one of the following features with mild and stable activity requiring category C therapy:

1. Category A or B signs or symptoms with mild and stable activity, requiring only symptomatic therapy or therapy at doses below the level required in category B

2. Sinus tachycardia

CATEGORY D

Previous involvement and no therapy is currently required (i.e., in remission)

CATEGORY E

No previous involvement

7) MUSCLE DISEASE ACTIVITY

CATEGORY A

Severe muscle inflammation resulting overall in major loss of function (requiring assistance with activities of daily living) based upon manual muscle strength testing, functional assessments, laboratory or other testing.

CATEGORY B

Moderate muscle inflammation resulting overall in moderate loss of function (not requiring assistance with activities of daily living) based upon manual muscle strength testing, functional assessments, laboratory or other testing.

CATEGORY C

Any one of:

1. Stable mild muscle inflammation, resulting overall in little or no loss of function -- based upon manual muscle strength testing, functional assessments, laboratory or other testing
2. Myalgia

CATEGORY D

Previous muscle inflammation and no medication currently required (i.e., in remission)

CATEGORY E

No evidence of previous muscle inflammation.