MYOSITIS DISEASE ACTIVITY ASSESSMENT TOOL - 2009

This is a combined tool that captures the physician’s assessment of disease activity of various organ systems via the

MYOSITIS INTENTION TO TREAT ACTIVITY INDEX (MITAX)

and via the

MYOSITIS DISEASE ACTIVITY ASSESSMENT VISUAL ANALOGUE SCALES (MYOACT)

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MYOSITIS DISEASE ACTIVITY ASSESSMENT VISUAL ANALOGUE SCALES (MYOACT)
(Modified 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 disease 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 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. 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.

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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:

a) the presence of clinical features at this time or symptoms **within the previous four weeks**

b) 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:

0 - if the feature is not present

1 - if the feature is improving - clinically significant improvement in the last 4 weeks over the previous 4 weeks

2 - if the feature is the same - manifestations that have been present for the last 4 weeks and the previous 4 weeks without significant improvement or deterioration

3 - if the feature is worse - clinically significant deterioration over the last 4 weeks compared to the previous 4 weeks

4 - if the feature is new - new in the last 4 weeks (compared to the previous 4 weeks)

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.

<table>
<thead>
<tr>
<th>CATEGORY</th>
<th>Description</th>
<th>Example</th>
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<tbody>
<tr>
<td><strong>CATEGORY A</strong>&lt;br&gt;A = Active&lt;br&gt;(9 points)</td>
<td>Denotes disease thought to be sufficiently active to require disease-modifying treatment: (9 points)&lt;br&gt;[High dose daily corticosteroids alone or in combination with high doses of other immunosuppressives or intravenous gammaglobulin (IVIG)]</td>
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<tr>
<td><strong>CATEGORY B</strong>&lt;br&gt;B = Beware&lt;br&gt;(3 points)</td>
<td>Denotes disease that is less active than in “A”; reversible problems requiring moderate doses of corticosteroids, other immunosuppressive agents or use of antimalarials, non-steroidal anti-inflammatory drugs (NSAIDs), topical steroids. [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.]</td>
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<tr>
<td><strong>CATEGORY C</strong>&lt;br&gt;C = Contentment&lt;br&gt;(1 point)</td>
<td>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.]</td>
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<tr>
<td><strong>CATEGORY D</strong>&lt;br&gt;D = Discount&lt;br&gt;(0 points)</td>
<td>System previously affected but currently inactive and no medication is required (i.e., in remission)</td>
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<tr>
<td><strong>CATEGORY E</strong>&lt;br&gt;E = No evidence of this system being active now or previously&lt;br&gt;(0 points)</td>
<td>Indicates system never involved</td>
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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.
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<td><strong>D = Discount</strong>&lt;br&gt;(0 points)</td>
<td><strong>E = No evidence</strong>&lt;br&gt;(0 points)</td>
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<tr>
<td><strong>Constitutional Disease Activity</strong>&lt;br&gt;Pyrexia scoring 2, 3 or 4&lt;br&gt;+ two other clinical feature scoring &gt; 1:&lt;br&gt; - Unintentional weight loss&lt;br&gt; - Fatigue/malaise/lethargy</td>
<td>Any one clinical feature scoring 2, 3 or 4</td>
<td>Any one clinical feature scoring 1</td>
<td>No current activity but known to have been active in the past.&lt;br&gt; - ie remission</td>
<td>No current or previous activity.</td>
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<td><strong>Cutaneous Disease Activity</strong>&lt;br&gt;Any one of the following scoring 2, 3 or 4:&lt;br&gt; - Cutaneous ulceration&lt;br&gt; - Erythroderma&lt;br&gt; - Erythematous rashes with secondary changes eg. accompanied by vesiculobullous or erosive changes or necrosis&lt;br&gt; - Panniculitis</td>
<td>1) Any Category A clinical feature scoring 1&lt;br&gt; OR&lt;br&gt; 2) Any one of the following scoring 2, 3 or 4:&lt;br&gt; - Erythematous rashes without secondary changes&lt;br&gt; - Heliotrope rash&lt;br&gt; - Gottron's papules or sign&lt;br&gt; - Periungual capillary changes&lt;br&gt; - Alopecia: diffuse&lt;br&gt; - Mechanic's hands</td>
<td>1) Any Category B clinical feature scoring 1&lt;br&gt; OR&lt;br&gt; 2) Alopecia: focal - scoring 1, 2, 3 or 4</td>
<td>No current activity but known to have been active in the past.&lt;br&gt; - ie remission</td>
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<td><strong>Skeletal Disease Activity</strong>&lt;br&gt;Severe inflammatory polyarthritis scoring 2, 3 or 4</td>
<td>1) Severe inflammatory polyarthritis scoring 1&lt;br&gt; OR&lt;br&gt; 2) Moderate inflammatory arthritis scoring 2, 3 or 4</td>
<td>1) Any category B clinical feature scoring 1&lt;br&gt; OR&lt;br&gt; 2) Any one of the following scoring 1, 2, 3 or 4:&lt;br&gt; - Mild arthritis&lt;br&gt; - Arthralgia</td>
<td>No current activity but known to have been active in the past.&lt;br&gt; - ie remission</td>
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<td><strong>Gastrointestinal Disease Activity</strong>&lt;br&gt;Any one of the following scoring 2, 3 or 4:&lt;br&gt; - Moderate to severe dysphagia&lt;br&gt; - Severe abdominal pain</td>
<td>1) Any Category A clinical feature scoring 1&lt;br&gt; OR&lt;br&gt; 2) Any one of the following scoring 2, 3 or 4:&lt;br&gt; - Moderate abdominal pain&lt;br&gt; - Mild dysphagia</td>
<td>1) Any Category B clinical feature scoring 1&lt;br&gt; OR&lt;br&gt; 2) Mild abdominal pain scoring 1, 2, 3 or 4</td>
<td>No current activity but known to have been active in the past.&lt;br&gt; - ie remission</td>
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**Pulmonary Disease**
1) Ventilatory abnormalities due to muscle weakness without primary lung disease - Dyspnea at rest scoring 2, 3, or 4
   OR
2) Active ILD scoring 3 (worse) or 4 (new) in at least two of the following:
   a) dyspnea or cough due to ILD
   b) parenchymal abnormalities on CXR or HRCT and/or ground glass-shadowing
   c) PFTs - minimum of 10% change in FVC or 15% change in DLCO

1) 17a) scoring 1, OR
   Ventilatory abnormalities due to muscle weakness without primary lung disease - Dyspnea on exertion scoring 2, 3 or 4
   OR
2) Active ILD scoring 2 (same) in at least two of the following (or scoring 1 or 3 in only one category with 2 in others):
   a) dyspnea or cough due to ILD
   b) parenchymal abnormalities on CXR or HRCT and/or ground glass-shadowing
   c) PFTs - 10% change in TLC/FVC or 15% change in DLCO
   OR
3) Moderate to severe dysphonia scoring 2, 3 or 4

1) 17 b scoring 1 OR
   Active ILD scoring 1 (ie improvement) in at least two of the following:
   a) dyspnea or cough due to ILD
   b) parenchymal abnormalities on CXR or HRCT and/or ground glass-shadowing
   c) PFTs - 10% change in TLC/FVC or 15% in DLCO
   OR
2) Moderate to severe dysphonia scoring 1 OR
   Mild dysphonia scoring 1, 2, 3 or 4

No current activity but known to have been active in the past - ie remission
No current or previous activity

**Cardiovascular Disease Activity**
Any one of the following scoring 2, 3 or 4
- Myocarditis/pericarditis
- Severe cardiac arrhythmia

1) Any Category A clinical feature scoring 1
   OR
2) Any one of the following scoring 2, 3 or 4:
   - Mild/moderate cardiac arrhythmia (except sinus tachycardia)

1) Any Category B clinical feature scoring 1
   OR
2) Sinus tachycardia scoring 1, 2, 3 or 4

No current activity but known to have been active in the past - ie remission
No current or previous activity

**Muscle Disease Activity**
Myositis resulting in severe muscle inflammation scoring 2, 3 or 4

1) Severe muscle inflammation scoring 1,
   OR
2) Moderate muscle inflammation scoring 2, 3, or 4

1) Myositis resulting in moderate inflammation scoring 1,
   OR
2) Myositis resulting overall in mild inflammation scoring 1, 2, 3 or 4
   3) Myalgia scoring scoring 1, 2, 3 or 4

No current activity but known to have been active in the past - ie remission
No current or previous activity

The respiratory aspect of this tool has been validated based scoring items a-c inclusive ie using subjective and objective measures in the clinic setting. In a clinical trial when PFTs and imaging may only be performed at baseline and only symptoms may be recorded at follow-up visits then scoring will based on a stable state of symptoms for two consecutive visits. The scoring of PFTs and imaging may be carried forward for 3 months if new studies are not available.