How to assess myositis disease activity in a busy general rheumatology clinic

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Many different approaches to assessment

• **Muscle Enzymes**
  – Creatine Kinase/ lactate dehydrogenase/ ALT / AST

• **Manual muscle testing**
  – MRC/extended 13 mark MRC/extended 8 point MRC/Kendall/-4-0 scale
  – 26, 18, 16, 15, 14, 8 muscle groups assessed
  – Sum scores( 160, 140, 90, 80, undefined) or mean scores of muscles tested

• **Dynamic muscle assessment**
  – timed walk/time to arise from a chair and time to walk 30 feet
  – Repetitive testing using 1kg weight max score 56
  – Myometry 9 muscle groups
  – Hand grip strength
  – Functional Index

• **Function or disability**
  – Modified Convery Assessment Scale/modified Rankin scale
  – Short Form 36 Health Survey (SF-36)/ The Neuromuscular Symptom Score (NSS)
  – Individualised neuromuscular quality of life questionnaire
  – Health assessment questionnaire
Many different approaches to assessment

Cochrane review 2012:
Immunosuppressant and immunomodulatory treatment for dermatomyositis and polymyositis 1980-2011

10 studies included
- 10 different types function disability score
- 6 different scales of muscle strength
- >5 different sets of muscle groups tested
Proposed preliminary core set measures for disease outcome assessment in adult and juvenile idiopathic inflammatory myopathies

F. W. Miller¹, L. G. Rider², Y.-L. Chung¹, R. Cooper³, K. Danko³, V. Farewell⁴, I. Lundberg⁵, C. Morrison⁶, L. Oakley⁷, I. Oakley⁷, C. Pilkington⁸, J. Vencovsky⁹, K. Vincent¹⁰, D. L. Scott¹¹, D. A. Isenberg¹² and for the International Myositis Outcome Assessment Collaborative Study Group

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6 Disease Activity Core Set Measures

1. Physician Global Activity - Visual Analogue Scale
2. Patient Global Activity - Visual Analogue Scale
4. Functional Assessment Tools – HAQ
5. Laboratory - Muscle Enzymes
6. Extramuscular Assessment - Myositis Disease Activity Assessment Tool
International Myositis Assessment & Clinical Studies Group (IMACS)

Global Approaches To Improved Treatments And Understanding Of Myositis

News & Events

Latest News:

- 1-Jun-12: Myositis Response Criteria to be Developed

Archived News:

- News Archive

About IMACS

The International Myositis Assessment and Clinical Studies Group (IMACS) is a coalition of health care providers and researchers with experience and interest in the myositis syndromes.

The goals of IMACS are to improve the lives of children and adults who suffer from myositis by discovering better therapies through understanding the causes of this disease.
Definition of improvement

3 of any 6 of the core set measures improved by ≥20%
no more than 2 worse by ≥25% (cannot include manual muscle testing)
Validation of manual muscle testing and a subset of eight muscles for adult and juvenile idiopathic inflammatory myopathies

Lisa G. Rider, Deloris Koziol, Edward H. Giannini, Minal S. Jain, Michaele R. Smith, Kristi Whitney-Mahoney, Brian M. Feldman, Susan J. Wright, Carol B. Lindsley, Lauren M. Pachman, Maria L. Villalba, Daniel J. Lovell, Suzanne L. Bowyer, Paul H. Plotz, Frederick W. Miller and Jeanne E. Hicks

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DOI: 10.1002/acr.20035

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<table>
<thead>
<tr>
<th>Study Description</th>
<th>Intervention / Comparator</th>
<th>Status</th>
</tr>
</thead>
<tbody>
<tr>
<td>Second line agents in myositis (SELAM trial)</td>
<td>MMT(MRC) 14 muscle groups</td>
<td>Published</td>
</tr>
<tr>
<td>GB-0998 for Treatment of Steroid-resistant Polymyositis and Dermatomyositis</td>
<td>MMT (MRC) 18 muscle groups</td>
<td>Published</td>
</tr>
<tr>
<td>Rituximab in Myositis study</td>
<td>IMACs definition of improvement</td>
<td>Published</td>
</tr>
<tr>
<td>MEDI-545 in Adult Patients With Dermatomyositis or Polymyositis</td>
<td>MMT-8</td>
<td>Published</td>
</tr>
<tr>
<td>Apremilast in the Treatment of Skin Disease in Patients With Dermatomyositis</td>
<td>MMT-8</td>
<td>Terminated 2011</td>
</tr>
<tr>
<td>BAF312 in Patients With Polymyositis and Dermatomyositis</td>
<td>IMACS core set measures</td>
<td>Completed 2012</td>
</tr>
<tr>
<td>PROMETHEUS Methotrexate + Glucocorticoids Versus Glucocorticoids Alone</td>
<td>IMACS core set measures</td>
<td>Completed 2013</td>
</tr>
<tr>
<td>Abatacept Treatment in Polymyositis and Dermatomyositis (ARTEMIS)</td>
<td>IMACs definition of improvement</td>
<td>Completed 2013</td>
</tr>
<tr>
<td>Five-year Actively Controlled Clinical Trial in New Onset Juvenile Dermatomyositis</td>
<td>IMACs definition of improvement</td>
<td>Recruited</td>
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<tr>
<td>Belimumab in Myositis (BIM)</td>
<td>IMACs definition of improvement</td>
<td>Recruiting</td>
</tr>
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<td>Safety and Efficacy of BAF312 in Dermatomyositis</td>
<td>MMT-8</td>
<td>Recruiting</td>
</tr>
<tr>
<td>Tocilizumab in the Treatment of Refractory Polymyositis and Dermatomyositis</td>
<td>IMACs definition of improvement</td>
<td>Recruiting</td>
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Extramuscular Assessment - Myositis Disease Activity Assessment Tool

- Sum of the 10 cm visual analogue scale scores for each of the six individual organ systems (CONSTITUTIONAL, CUTANEOUS, SKELETAL, GASTROINTESTINAL, PULMONARY, CARDIAC)
- Divided by 60
### IMACS FORM 07a: MYOSITIS DISEASE ACTIVITY ASSESSMENT TOOL – 2005, Version 2

<table>
<thead>
<tr>
<th>Subject's IMACS number:</th>
<th>ASSESSOR:</th>
<th>Date Assessed:</th>
<th>Assessment number:</th>
</tr>
</thead>
</table>

### Constitutional Disease Activity

<table>
<thead>
<tr>
<th>(Absent)</th>
<th>(Maximum)</th>
<th>Examples of maximal score</th>
</tr>
</thead>
<tbody>
<tr>
<td></td>
<td></td>
<td>Severe fatigue or malaise resulting in being bed bound and an inability to perform selfcare</td>
</tr>
</tbody>
</table>

1. Pyrexia – documented fever > 38° Celsius
2. Weight loss – unintentional > 5%
3. Fatigue/malaise/lethargy

<table>
<thead>
<tr>
<th></th>
<th>0</th>
<th>1</th>
<th>2</th>
<th>3</th>
<th>4</th>
<th>NA</th>
</tr>
</thead>
</table>

### Cutaneous Disease Activity

<table>
<thead>
<tr>
<th>(Absent)</th>
<th>(Maximum)</th>
<th>Examples of maximal score</th>
</tr>
</thead>
<tbody>
<tr>
<td></td>
<td></td>
<td>ULCERATION TO MUSCLE, TENDON OR BONE; EXTENSIVE ERYTHEMADERMA</td>
</tr>
</tbody>
</table>

4. Cutaneous ulceration
5. Erythroderma
6. Panniculitis
7. Erythematous rashes:
   a. with secondary changes (e.g. accompanied by erosions, vesiculobullous change or necrosis)
   b. without secondary changes
8. Heliotrope rash
9. Gottron’s papules/sign
10. Periungual capillary changes
11. Alopecia:
    a. Diffuse hair loss
    b. Focal, patchy with erythema
12. Mechanics hands

<table>
<thead>
<tr>
<th></th>
<th>0</th>
<th>1</th>
<th>2</th>
<th>3</th>
<th>4</th>
<th>NA</th>
</tr>
</thead>
</table>
6 Disease Activity Core Set Measures

1. Physician Global Activity - Visual Analogue Scale
2. Patient/Parent Global Activity - Visual Analogue Scale
4. Functional Assessment Tools – HAQ
5. Laboratory - Muscle Enzymes
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**Preferred Testing Order of MMT8 Muscle Groups.**

This table provides a preferred order to the testing of muscle groups for manual muscle testing. Generally, for bilateral muscle testing, each muscle group is first tested on the right and then the left, prior to proceeding to the next muscle group in the list. Some muscle groups are listed here with anti-gravity testing, but for a weaker patient, these would be tested in a sidelying or supine position, per the table below (Testing Positions); the re-test for a weaker patient is indicated in gray scale.

Abbreviations: G.E., gravity eliminated.

<table>
<thead>
<tr>
<th>POSITION</th>
<th>ORDER OF TESTING</th>
</tr>
</thead>
<tbody>
<tr>
<td><strong>SITTING</strong></td>
<td></td>
</tr>
<tr>
<td>Deltoid middle (shoulder abductors)</td>
<td>1</td>
</tr>
<tr>
<td>Biceps brachii (elbow flexors)</td>
<td>2</td>
</tr>
<tr>
<td>Wrist extensors (extensor carpi ulnaris/radialis)</td>
<td>3</td>
</tr>
<tr>
<td>Quadriceps femoris (knee extensors)</td>
<td>4</td>
</tr>
<tr>
<td>Ankle dorsiflexors (tibialis anterior)</td>
<td>5</td>
</tr>
<tr>
<td><strong>SUPINE</strong></td>
<td></td>
</tr>
<tr>
<td>Neck flexors (scalenes, sternocleidomastoid)</td>
<td>6</td>
</tr>
<tr>
<td>Deltoid middle (G.E. test if needed)</td>
<td>-</td>
</tr>
<tr>
<td>Gluteus medius (G.E. test if needed)</td>
<td>-</td>
</tr>
<tr>
<td><strong>SIDELYING (lying on left side-right muscles tested)</strong></td>
<td></td>
</tr>
<tr>
<td>Gluteus medius (hip abductors)</td>
<td>7</td>
</tr>
<tr>
<td>Gluteus maximus (G.E. test if needed)</td>
<td>-</td>
</tr>
<tr>
<td>Biceps brachii (G.E. test if needed)</td>
<td>-</td>
</tr>
<tr>
<td>Neck flexors (G.E. test if needed)</td>
<td>-</td>
</tr>
<tr>
<td><strong>PRONE</strong></td>
<td></td>
</tr>
<tr>
<td>Gluteus maximus (hip extensors)</td>
<td>8</td>
</tr>
</tbody>
</table>
## MMT8 uses 10 point scale

<table>
<thead>
<tr>
<th>Scales of Muscle Strength</th>
<th>10-Point-Scale (Kendall FP 1993)</th>
<th>MRC-Scale</th>
</tr>
</thead>
<tbody>
<tr>
<td>No movement</td>
<td>0</td>
<td>0</td>
</tr>
<tr>
<td>Visible/palpable tension of tendons, but not visible muscle contraction</td>
<td>T</td>
<td>1</td>
</tr>
<tr>
<td>Movement in horizontal plane</td>
<td></td>
<td></td>
</tr>
<tr>
<td>Partial ROM</td>
<td>1</td>
<td>2-</td>
</tr>
<tr>
<td>Complete ROM</td>
<td>2</td>
<td>2</td>
</tr>
<tr>
<td>Movement against gravity</td>
<td>3</td>
<td>2+</td>
</tr>
<tr>
<td>Partial ROM</td>
<td></td>
<td></td>
</tr>
<tr>
<td>Gradually release from test position</td>
<td>4</td>
<td>3-</td>
</tr>
<tr>
<td>Maintains Position</td>
<td></td>
<td></td>
</tr>
<tr>
<td>Keeps test position (no additional pressure)</td>
<td>5</td>
<td>3</td>
</tr>
<tr>
<td>Keeps test position against slight pressure</td>
<td>6</td>
<td>3+</td>
</tr>
<tr>
<td>Keeps test position against slight to moderate pressure</td>
<td>7</td>
<td>4-</td>
</tr>
<tr>
<td>Keeps test position against moderate pressure</td>
<td>8</td>
<td>4</td>
</tr>
<tr>
<td>Keeps test position against moderate to strong pressure</td>
<td>9</td>
<td>4+</td>
</tr>
<tr>
<td>Keeps test position against strong pressure(normal)</td>
<td>10</td>
<td>5</td>
</tr>
</tbody>
</table>
MMT8

- Test one side only - the patient's dominant side
- Maximum score 80
MMT8

• Internal reliability 0.96
• Inter-rater reliability 0.72
• Intra-rater reliability 0.84

• Convergent construct validity
  – MMT24 0.96
  – Physician global 0.37
  – CK 0.36

Start in sitting position

1. Shoulder abduction (Deltoid)

Place the shoulder at 90° abduction
Neutral glenohumeral rotation
Start in sitting position

1. Shoulder abduction (Deltoid)
Remain in sitting position

2. Elbow flexion (Biceps)
Remain in sitting position

3. Wrist extension (wrist extensors)

Elbow in 90 flexion, forearm in pronation, wrist in extension.
Remain in sitting position

4. knee extension (Quadriceps femoris)

Knee flexed at around 5°
Remain in sitting position

5. Ankle dorsiflexion

The foot in dorsiflexion and inversion.

Arms provide external support
Move to the supine position

6. Neck flexion

Both the head and the neck are positioned into flexion
Move onto non-dominant side

7. Hip Abduction (gluteus medius)

Contralateral leg flexed at the hip and knee
Ipsilateral leg extended at the knee
Stabilizing force at the pelvis.
Move into prone position

8. Hip Extension (gluteus maximus)

90° knee flexion (if possible) and the testing surface raised to the level of the tester’s waist.
MMT8

1. Shoulder abduction (Deltoid)  6
2. Elbow flexion (Biceps)  10
3. Wrist extension (wrist extensors)  10
4. Knee extension (Quadriceps femoris)  10
5. Ankle dorsiflexion  10
6. Neck flexion  8
7. Hip Abduction (gluteus medius)  6
8. Hip Extension (gluteus maximus)  5

TOTAL  MMT-8 Score  65/80
In practice:
Patient TIF $1\gamma$ antibodies
## Disease assessment over 7 months

<table>
<thead>
<tr>
<th>Time in months</th>
<th>1</th>
<th>2</th>
<th>3</th>
<th>4</th>
<th>5</th>
<th>6</th>
<th>7</th>
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<tbody>
<tr>
<td>Physician global activity</td>
<td>50</td>
<td>60</td>
<td>47</td>
<td>13</td>
<td>40</td>
<td>3</td>
<td>30</td>
</tr>
<tr>
<td>Patient global activity</td>
<td>30</td>
<td>25</td>
<td>25</td>
<td>26</td>
<td>15</td>
<td>5</td>
<td>10</td>
</tr>
<tr>
<td>MMT8</td>
<td>69</td>
<td>68</td>
<td>73</td>
<td>71</td>
<td>74</td>
<td>77</td>
<td>78</td>
</tr>
<tr>
<td>creatine kinase</td>
<td>70</td>
<td>120</td>
<td>95</td>
<td>96</td>
<td>102</td>
<td>102</td>
<td>71</td>
</tr>
<tr>
<td>Extramuscular disease</td>
<td>40</td>
<td>20</td>
<td>10</td>
<td>3</td>
<td>0</td>
<td>5</td>
<td>10</td>
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<tr>
<td>assessment</td>
<td></td>
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<td></td>
<td></td>
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</tr>
<tr>
<td>HAQ</td>
<td>1.87</td>
<td>1.87</td>
<td>1.87</td>
<td>1</td>
<td>1.75</td>
<td>2.25</td>
<td>1.75</td>
</tr>
</tbody>
</table>
Physician global activity
Patient global activity
MMT8
Creatine kinase
Extramuscular disease assessment
HAQ
Physician global activity

Patient global activity

MMT8

Creatine kinase

Extramuscular disease assessment

HAQ
IMACs disease activity scores and instructions for their use can be found at

- OR google ‘IMACs and Myositis’