

## IMACS Core Set Measures – Case examples

### Myositis Damage Index (MDI)

The purpose of this exercise is to provide some guidelines in scoring the MDI. These cases were prepared by Fred Miller, Lisa Rider, Clarissa Pilkington, and David Isenberg. The scores provided in the 3 cases are suggestions for your consideration and there may be some differences of opinion. These cases particularly focus on scoring of the muscle systems and global activity/damage VAS scores, although the discussion presents the scoring of other involved organ systems as well.

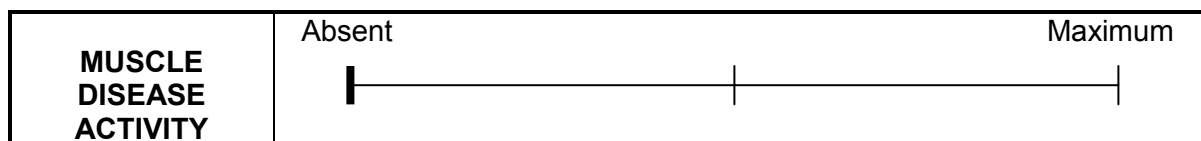
#### Case 4 - PM at Follow-up

A 52 y.o. Asian woman had the insidious onset of muscle weakness, which was likely undiagnosed for many years after onset. She underwent a full evaluation -- including a muscle biopsy showing moderate numbers of endomysial mononuclear cells without rimmed vacuoles or extreme variation of myofiber size -- which resulted in a diagnosis of definite polymyositis. She refused treatment with corticosteroids or other therapies but took herbal remedies for an additional 3 years. Two years ago, when her muscles were very painful, she was confined to a wheelchair and unable to swallow solids. Her family convinced her to seek medical treatment and she was found to have very active polymyositis. She received intravenous pulse solumedrol and was begun on high dose oral prednisone in divided doses and methotrexate therapy.

While on high dose oral prednisone, she developed brittle insulin-dependent diabetes that never resolved, avascular necrosis of the left hip requiring joint replacement, and myocarditis with subsequent congestive heart failure, which also limits her function. She never regained much strength and at first was confined to a wheelchair, but has been bed-bound for a year, requiring assistance for almost all activities of daily living (HAQ = 2.8). Her myalgias resolved, but her swallowing continued to be a worsening problem, eventually requiring a nasogastric feeding tube to prevent aspiration. She develops severe shortness of breath with any activity, and sometimes at rest, requiring intermittent nasal oxygen. On exam she had severe muscle atrophy of all limbs and 2-6/10 strength (Unilateral MMT-8 = 28 out of 80). Her CPK is 65 (normal range 15-250 U/l), serum creatinine is 0.3 (normal range 0.7 – 1.2 mg/dl), and the rest of her laboratory testing, including CXR, is normal. She is essentially unchanged from 6 months ago.

### MYOSITIS DISEASE ACTIVITY and DAMAGE ASSESSMENT TOOL VALUES

#### Muscle Disease Activity Assessment: VAS = 0 cm



Example of Max score: Severe muscle weakness resulting in being bed bound and an inability to perform self care



#### **Discussion of Case 4:**

The patient has no evidence of disease activity, but has severe damage of many organ systems. She has severe muscle damage resulting in fixed muscle weakness with extensive atrophy, severe functional impairment and a bed-bound state (**MDI Muscle VAS = 10 cm**). Other damaged organ systems include: skeletal with avascular necrosis requiring hip replacement (**MDI Skeletal VAS = 8 cm**); GI system requiring a nasogastric tube (**MDI GI VAS = 9 cm**); pulmonary system with impaired respiratory musculature (**MDI Pulmonary VAS = 8 cm**); cardiac involvement with congestive heart failure and functional impairment (**MDI Cardiovascular VAS = 7 cm**) and endocrine system involvement with steroid-induced, brittle insulin-dependent diabetes (**MDI Endocrine VAS = 6 cm**). Her extremely limited function (**HAQ = 2.8 out of 3.0 maximum**) and multiple systems with high degree of damage place her **MDI Global Damage score at 9 cm**, but her ability to breath and swallow without external support systems keep her from attaining a MDI Global Damage VAS score of 10 cm.

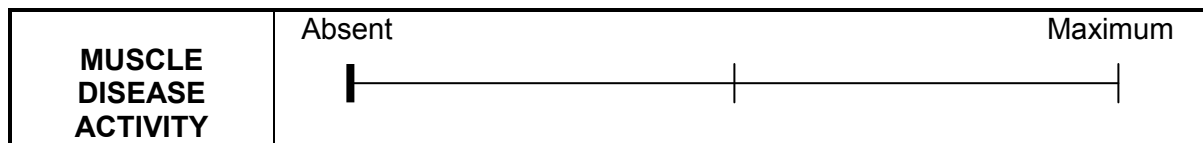
**Case 5 – PM at Follow-up**

A 69-year-old Latino male presents with a vague history of 2-3 years of increasing fatigue with muscle weakness in the upper and lower extremities, unexpected falling and slowly increasing dysphagia with solid but not liquid foods. He denies alcohol or drug use, muscle pain and weight loss. His past medical history is unremarkable and family history is negative for any neuromuscular or autoimmune diseases except for SLE in a younger sister. He was seen by you a year ago and a detailed workup then only revealed muscle weakness, proximal > distal, in the 6-7/10 range of the upper and lower extremities (Unilateral MMT-8 = 52 out of 80) and moderate atrophy of the shoulder, arm and thigh muscles. His HAQ was 2.0 and he reportedly was unable to climb stairs without assistance and could not keep up with family members when walking. Laboratories were remarkable for a CK of 325 (normal range 20 to 250) with 100% MM, a creatinine of 0.8 (normal 0.9-1.4 mg/dl) and a positive ANA at 1:640, speckled pattern. All of his other extensive laboratory and autoantibody tests were negative or unremarkable. A barium swallow examination showed moderate esophageal dysfunction but no evidence of aspiration. His CXR and EKG were normal, but his EMG showed evidence of active muscle inflammation in the upper and lower extremities. A thigh muscle biopsy showed variation in myofiber size, degeneration and regeneration, rare scattered endomysial mononuclear cell infiltrations, fatty replacement of myofibers and increased interstitial spaces, but no evidence of inclusion bodies or other abnormalities. An extensive cancer work up was negative. You suggested he begin using a cane for ambulation to minimize the risk of falling.

A year ago, he initially refused all therapy, especially corticosteroids given his sister’s negative experience with them, but after further explanations, finally agreed to take 40 mg prednisone daily and was told to return in 1 month for follow up; he returned 6 months later with a normal CK but no change in muscle strength or swallowing. You begin a slow steroid taper over the next 6 months to no prednisone today and he remains completely unchanged in all respects from your initial evaluation 12 months ago except for normalization of all muscle enzymes and a creatinine of 0.7 (normal 0.9-1.4 mg/dl).

**MYOSITIS DISEASE ACTIVITY and DAMAGE ASSESSMENT TOOL VALUES**

**Muscle Disease Activity Assessment: VAS = 0 cm**



Example of Max score: Severe muscle weakness resulting in being bed bound and an inability to perform self care

- 25. Myositis:
  - a) Severe muscle inflammation **OR** 0 1 2 3 4 NA
  - b) Moderate muscle inflammation **OR** 0 1 2 3 4 NA
  - c) Mild muscle inflammation 0 1 2 3 4 NA
- 26. Myalgia 0 1 2 3 4 NA



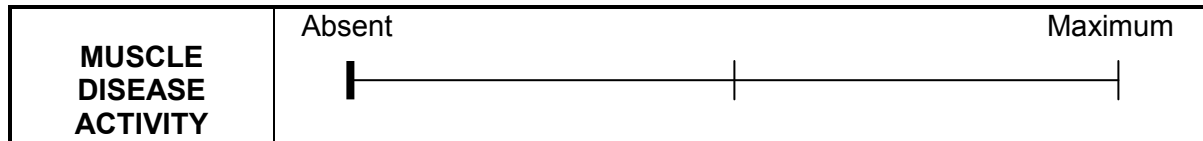
**Case 6 – JDM at Follow-up**

During the one-and-a-half years after Case 3 (see Case 3 in Global Disease Activity handout) presented, the child received oral corticosteroids, photoprotection and physical therapy and was treated as an outpatient; no additional therapy was needed. Prednisone was eventually tapered and discontinued because she developed moderate transient hypertension requiring treatment and bilateral cataract formation, which impaired her vision. Her strength showed gradual improvement but she complains of not being back to normal, having difficulty keeping up with classmates at recess, and of getting tired more easily than before her illness started.

Today she appears well with normal vital signs; there are mild atrophic skin changes over the 2nd and 3rd knuckles, a mild heliotrope rash and slightly enlarged periungual capillaries along with 2 small superficial plaques of calcinosis over her elbows. Abdominal strength is grade 8/10, neck and arm flexors have mild atrophy and are grade 8/10, but strength is otherwise normal (MMT-8=76 out of 80). CK is 110 (normal 20 to 215 U/l), aldolase is 4 (normal 1 to 9 U/l), and creatinine is 0.7 (normal 0.7 – 1.2 mg/dl). Essentially, she is unchanged from 6 months ago.

**MYOSITIS DISEASE ACTIVITY and DAMAGE ASSESSMENT TOOL VALUES**

**Muscle Disease Activity Assessment: VAS = 0 cm**



Example of Max score: Severe muscle weakness resulting in being bed bound and an inability to perform self care

25. Myositis:

- a) Severe muscle inflammation **OR** 0 1 2 3 4 NA
- b) Moderate muscle inflammation **OR** 0 1 2 3 4 NA
- c) Mild muscle inflammation 0 1 2 3 4 NA

26. Myalgia

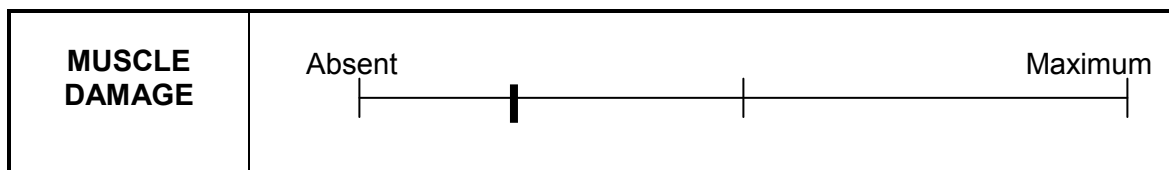
- 0 1 2 3 4 NA

**Global Disease Activity Assessment: VAS = 1 cm**



Overall evaluation for the totality of disease activity in **ALL** systems (**including** muscle)

**Myositis Damage Index (MDI) Muscle Damage Assessment: VAS = 2 cm**

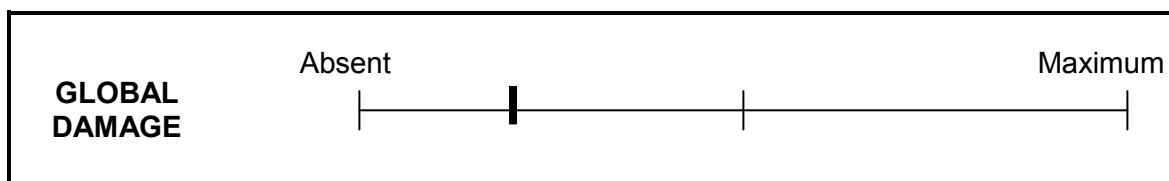


Example of Max score: Severe muscle atrophy or weakness resulting in being bed bound and an inability to perform self care

1. Muscle atrophy (clinical)	0	1	NA
2. Muscle weakness not attributable to active muscle disease	0	1	NA
3. Muscle dysfunction: decrease in aerobic exercise capacity	0	1	NA
4. <u>Muscle atrophy assessed by radiographic methods</u>	0	1	NA
5. <u>Low serum creatinine</u>	0	1	NA

Creatinine value = 0.7 mg/dL; lower limit normal value = 0.7mg/dL

**Myositis Damage Index (MDI) Global Damage Assessment: VAS = 2 cm**



Examples of Max score: None

**Discussion of Case 6**

One-and-a-half years later her disease is improved, with normalization of muscle enzymes and without evidence of muscle disease activity (**Muscle disease VAS = 0 cm**), but there is mildly active skin disease based upon the finding of a persistent heliotrope rash and periungual capillary changes (Cutaneous MDAAT numerical score = 2 [feature is the same], Cutaneous VAS activity = 2 cm); and continued mild fatigue (Constitutional MDAAT numerical score = 2 [feature is the same], Constitutional VAS activity = 1 cm). Her **Global disease activity VAS = 1 cm**).

Evidence of damage is present. She has stable, persistent muscle weakness with atrophy, as well as aerobic exercise impairment, without other known cause (such as steroid myopathy) and no evidence of muscle disease activity (**MDI Muscle damage VAS = 2 cm**). Regarding her skin, she has atrophic Gottron's lesions from prior active disease and several superficial plaques of calcinosis (**MDI Cutaneous VAS = 2 cm**). She also has bilateral cataract formation with impaired vision (**MDI Ocular VAS = 5 cm**). She has no other evidence of damage, giving her a **MDI Global Damage Assessment VAS = 2 cm**.