IMACS FORM 08: MYOSITIS DAMAGE INDEX (MDI) - 2001

Please see the instructions and Myositis Damage Index Glossary of Terms Prior to Assessment (pp. 5-8).

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

MUSCLE DAMAGE	(Absent) (Maximum)	**Maximum Value Guidelines (Examples of maximal 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. Muscle atrophy assessed by radiographic methods	*0*	*1*	*NA*
5. Low serum creatinine	*0*	*1*	*NA*

Creatinine value = ____ mg/dL or umol/L; lower limit normal value = _____mg/dL or umol/L

SKELETAL DAMAGE	(Absent) (Maximum)	**Maximum Value Guidelines (Examples of maximal score)**
		-Life threatening fractures from osteoporosis -Avascular necrosis requiring arthroplasty

6. Joint contractures	0	1	NA
7. Osteoporosis with fracture or vertebral collapse (excluding avascular necrosis)	0	1	NA
8. Avascular necrosis	0	1	NA
9. Deforming arthropathy (including reducible deformities, excluding avascular necrosis and contractures)	0	1	NA
10. Osteoporosis without clinical fracture, requiring treatment of osteoporosis	*0*	*1*	*NA*
11. Limitation of motion (Passive Joint ROM in degrees):			
a. Elbow extension (degrees): R_____L_____	*0*	*1*	*NA*
b. Hip flexion (degrees): R_____L_____	*0*	*1*	*NA*
c. Knee extension (degrees): R_____L_____	*0*	*1*	*NA*
d. Ankle dorsiflesion (degrees) R_____L_____	*0*	*1*	*NA*

CUTANEOUS DAMAGE	(Absent) (Maximum)	**Maximum Value Guidelines (Examples of maximal score)**
		Calcinosis with extensive subcutaneous exoskeleton resulting in extreme loss of function (bedridden, inability for self care)

12. Calcinosis:	0	1	NA
a. Calcinosis, superficial plaques or nodules	*0*	*1*	*NA*
b. *Calcinosis, tumoral*	*0*	*1*	*NA*
c. Calcinosis, planar	*0*	*1*	*NA*
d. Calcinosis, exoskeleton/calcinosis universalis	*0*	*1*	*NA*
13. Alopecia	0	1	NA
14. Cutaneous scarring or atrophy	0	1	NA
15. Poikiloderma	0	1	NA
16. Lipodystrophy	0	1	NA

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

GASTROINTESTINAL DAMAGE	**(Absent)** — **(Maximum)**	**Maximum Value Guidelines (Examples of maximal score)**
		Extreme dysfunction requiring total parenteral nutrition

17. Dysphagia	0	1	NA
18. Gastrointestinal dysmotility, constipation, diarrhea or abdominal pain	0	1	NA
19. Infarction or resection of bowel or other gastrointestinal organs	0	1	NA
20. Steatosis	*0*	*1*	*NA*

PULMONARY DAMAGE	**(Absent)** — **(Maximum)**	**Maximum Value Guidelines (Examples of maximal score)**
		- Fibrosis requiring ventilatory support; or -Pulmonary hypertension resulting in extreme loss of function (bedridden, inability for self care)

21. Dysphonia	0	1	NA
22. Impaired lung function due to respiratory muscle damage	0	1	NA
23. Pulmonary fibrosis	0	1	NA
24. Pulmonary hypertension	0	1	NA
25. Diminished lung function:			
a. DL-CO/Va (Carbon monoxide diffusing capacity corrected for alveolar volume)(% Predicted =_____%)	*0*	*1*	*NA*
b. FEV1 (forced expiratory volume in 1 second) (% predicted = _____%)	*0*	*1*	*NA*

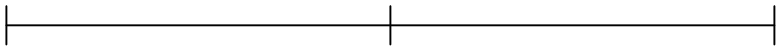
CARDIOVASCULAR DAMAGE	**(Absent)** — **(Maximum)**	**Maximum Value Guidelines (Examples of maximal score)**
		--Myocardial dysfunction resulting in extreme loss of function (bedridden, inability for self care)

26. Hypertension requiring treatment > 6 months	0	1	NA
27. Ventricular dysfunction / cardiomyopathy	0	1	NA
<u>Assessed in Adult Patients > = 18 years of age:</u>			
28. Angina or coronary artery bypass	0	1	NA
29. Myocardial infarction	0	1	NA

PERIPHERAL VASCULAR DAMAGE	**(Absent)** — **(Maximum)**	**Maximum Value Guidelines (Examples of maximal score)**
		- Limb loss; or -Thrombosis requiring intensive care unit (ICU) care; or-Severe disease resulting in extreme loss of function (bedridden, inability for self care)

30. Tissue or pulp loss	0	1	NA
31. Digit or limb loss or resection	0	1	NA
32. Venous or arterial thrombosis with swelling, ulceration or venous stasis	0	1	NA
<u>Assessed in Adult Patients > = 18 years of age:</u>			
33. Claudication	0	1	NA

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

	(Absent) (Maximum)	Maximum Value Guidelines (Examples of maximal score)
ENDOCRINE DAMAGE		Extreme disease resulting in ICU care or life threatening complications, such as ICU stay for seizures or DKA; renal failure, amputation

Assessed in Patients with diagnosis before 18 years of age:

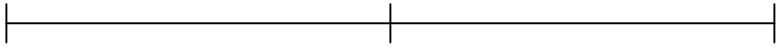
34. Growth failure	0	1	NA
35. Delay in development of secondary sexual characteristics	0	1	NA

Assessed in Both Pediatric and Adult Patients:

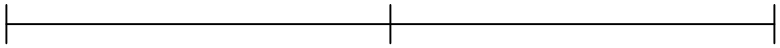
36. Hirsutism or Hypertrichosis	0	1	NA
37. Irregular menses	0	1	NA
38. Primary or Secondary Amenorrhea	0	1	NA
39. Diabetes	0	1	NA
40. Hyperlipidemia (regardless of treatment)	*0*	*1*	*NA*

Assessed in Adult Patients (> = 18 years of age and adolescent patients when applicable)

41. Infertility (female or male)	0	1	NA
42. Sexual dysfunction	0	1	NA

	(Absent) (Maximum)	Maximum Value Guidelines (Examples of maximal score)
OCULAR DAMAGE		Complete loss of vision in both eyes

43. Cataract resulting in visual loss	0	1	NA
44. Visual loss, other, not secondary to cataracts	0	1	NA

	(Absent) (Maximum)	Maximum Value Guidelines (Examples of maximal score)
INFECTION		Infection resulting in septic shock or life threatening complications

45. Chronic infection	0	1	NA
Specify:_____			
Specify:_____			
46. Multiple infections	0	1	NA
Specify:_____			
Specify:_____			

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

MALIGNANCY (Absent) ——— (Maximum)

Maximum Value Guidelines (Examples of maximal score)

Malignancy resulting in ICU care or life threatening complications

47. Any form of cancer, Specify type, grade and stage:_____ 0 1 NA

OTHER DAMAGE, specify (Absent) ——— (Maximum)

Maximum Value Guidelines (Examples of maximal score)

Extreme disease damage resulting in ICU care or life threatening complications

48. *Death* *0 1 NA*
*Cause and date:*_____

49. Specify:_____

50. Specify:_____

GLOBAL DAMAGE (Absent) ——— (Maximum)

Maximum Value Guidelines (Examples of maximal score)

None

IMACS MYOSITIS DAMAGE INDEX (MDI) - 2001
(Modification of the SLICC/ACR Damage Index, Brit J Rheum 1996; 35: 248-54)

Damage is defined as persistent changes in anatomy, physiology, pathology or function, which are present for at least 6 months. Damage may be the result of prior active disease (causing scarring, fibrosis and atrophy), complications of therapy, co-morbid conditions, or other events. A portion of disease damage is disease chronicity. Features of damage are ascertained by clinical assessment and must be present for at least 6 months (or the pathology that led to the feature must have been present for at least 6 months) despite prior immunosuppressive or other therapy, including exercise and rehabilitation. Only items present since date of diagnosis should be included. Damage is often permanent and cumulative. Damage scores most often increase over time, but in some cases may decline (i.e., a manifestation which was previously present which has currently resolved would receive a score of 0 in the present assessment).

For each organ system, please assess the severity and extent of damage exhibited by the patient at this time. To assess the severity, please rate your overall assessment of the current disease damage for each of the systems below by drawing a vertical mark on the 10-cm. line according to the following scale:

-Left end of line = no evidence of disease damage,
-Midpoint of line = moderate disease damage, and
-Right end of line = extreme or maximum disease damage.
-Please write in NA if the system cannot be assessed.

To assess the extent of damage in that organ system, please indicate the following for the specific items that are assessed:

-A score of 0 indicates the item has never been present.
-A score of 1 indicates the damage manifestation at a single point in time that has been present for at least six months.
-NA = cannot be assessed.

At the end of each organ system section, items in italics represent specialised objective testing items that are optional for the assessment and will be scored separately in an extended score. When absolute values are requested, please provide these even if they are within normal limits.

IMACS MYOSITIS DAMAGE INDEX Scoring System

The MDI has three proposed scores: an **extent of damage score**, a **severity of damage score** and an **extended damage score.**

The proposed scoring system for the **MDI extent of damage score** is the sum of all 0 or 1 scores for the eleven individual organ systems (MUSCLE, SKELETAL, CUTANEOUS, GASTROINTESTINAL, PULMONARY, CARDIOVASACULAR, PERIPHERAL VASCULAR, ENDOCRINE, OCULAR, INFECTION, MALIGNANCY) divided by the total possible score (range = 0 – 35 in children, 0-37 for adolescents and 0-38 in adults). If one or more items were not assessed, the resulting score would be divided by the maximum possible score of the assessed items. The categories of OTHER DAMAGE and GLOBAL DAMAGE are not included in the MDI extent of damage score but are scored separately.

The proposed **MDI severity of damage score** is the sum of the 10 cm visual analogue scale scores for each of the eleven individual organ systems (MUSCLE, SKELETAL, CUTANEOUS, GASTROINTESTINAL, PULMONARY, CARDIOVASACULAR, PERIPHERAL VASCULAR, ENDOCRINE, OCULAR, INFECTION, MALIGNANCY) divided by the total possible score (range = 0 – 110). If one or more organ systems were not assessed, the resulting score would be divided by the maximum possible score of the assessed items. The categories of OTHER DAMAGE and GLOBAL DAMAGE are not included in the MDI severity score but are scored separately.

The proposed **MDI extended damage score** is the sum of the optional items listed in italics under the systems MUSCLE, SKELETAL, GASTROINTESTINAL, PULMONARY, ENDOCRINE divided by the total possible score. Each optional item is scored 0 or 1, providing a range of 0 – 16 for the extended damage score. If one or more items were not assessed, the resulting score would be divided by the maximum possible score assessed.

IMACS MYOSITIS DAMAGE INDEX Glossary of Terms

MUSCLE DAMAGE

1. *Muscle atrophy*: decreased muscle mass assessed by clinical exam
2. *Muscle weakness not attributable to active muscle disease:* weakness present for at least 6 months, demonstrated on clinical examination, not thought to be due to active muscle inflammation based on assessments of clinical and laboratory measures, such as serum muscle enzymes, magnetic resonance imaging, or repeat muscle biopsies.
3. *Muscle dysfunction*, decrease in aerobic exercise tolerance by clinical history **or** assessed by aerobic exercise testing, due to muscle damage and not attributable to cardiac, pulmonary, psychologic or other factors.
4. *Muscle atrophy:* assessed by radiographic methods, including T1 MRI, CT scan, DEXA scan (body composition) (OPTIONAL ASSESSMENT

SKELETAL DAMAGE

5. *Joint contractures*: fixed limitation in the normal range of motion of joints, in the absence of synovitis, excluding reducible deformities, avascular necrosis and deforming arthropathy.
6. *Osteoporosis with fracture or vertebral collapse (excluding avascular necrosis):* demonstrated by any imaging technique.
7. *Avascular necrosis:* demonstrated by any imaging technique.
8. *Osteoporosis without clinical fracture:* requiring treatment of osteoporosis beyond calcium and vitamin D (prophylactic) therapy, demonstrated by any imaging technique (OPTIONAL ASSESSMENT)

CUTANEOUS DAMAGE

9. *Calcinosis:* dystrophic calcification, observed clinically or radiologically in the skin, subcutaneous tissue, fascia, or muscle. Calcinosis can be clinically or radiographically distinguished into 4 subtypes (Blane CE et al, 1984, AJR, 142: 397-400): (*the 4 subtypes are part of the OPTIONAL ASSESSMENT*)
    a. *Superficial plaques or nodules:* circumscribed lesions confined to the cutaneous or subcutaneous tissue
    b. *Tumoral:* large circumscribed nodules, which are intramuscular. Can ulcerate if subcutaneous.
    c. *Planar or fascial:* linear accumulations of calcinosis that are along the fascial plane of subcutaneous tissue or muscles.
    d. *Exoskeleton/calcinosis universalis:* widespread sheets of calcium in the muscle, fascia and subcutaneous tissue, often encasing the patient. Can ulcerate when subcutaneous.
10. *Alopecia:* hair loss with scarring present
11. *Cutaneous scarring or atrophy:* dermal or epidermal, with or without telangiectasia
12. *Poikiloderma:* fine speckled pattern of hyper- and hypopigmented macules interspersed with telangiectasias and cutaneous atrophy *(requires all 3 features)* usually in areas of photosensitivity
13. *Lipodystrophy:* loss of subcutaneous fat (localised or widely distributed) observed clinically or radiologically.

GASTROINTESTINAL DAMAGE

14. *Dysphagia:* persistent difficulty swallowing by history or persistent changes documented by radiography or other objective measures.
15. *Infarction or resection of bowel or other GI organs*, by history.
16. *Steatosis (OPTIONAL ASSESSMENT):* persistent fatty changes of liver, documented by ultrasound, CT scan or biopsy on at least 1 occasion, with persistent changes on reexamination

PULMONARY DAMAGE

17. *Dysphonia:* persistent alteration in voice quality, resonance, articulation or speech rate from normal.
18. *Impaired lung function due to respiratory muscle damage:* shortness of breath not thought to be due to active muscle inflammation or intrinsic pulmonary disease

19. *Pulmonary fibrosis:* shortness of breath **or** rales on physical exam for at least 6 months, with previously documented abnormal chest radiograph, computed tomography scan, or biopsy evidence of interstitial lung disease (ILD)/pulmonary fibrosis.
20. *Pulmonary hypertension:* Right ventricular prominence, **or** loud P2, **or** by direct measurement of pulmonary pressures (greater than 10% above upper limit of normal).

CARDIOVASCULAR DAMAGE

21. *Hypertension:* Diagnosed by *b*lood pressure > 95% of upper limits of normal for age and gender, requiring treatment > 6 months
22. *Ventricular dysfunction/Cardiomyopathy:* ventricular dysfunction documented clinically or by echocardiography.

Assessed only in adult patients:

23. *Angina:* episodes of angina present for a period of at least 6 months.
24. *Myocardial infarction*: documented by electrocardiogram and enzymes.

PERIPHERAL VASCULAR DAMAGE

25. *Tissue or pulp loss:* tissue loss such as pulp space loss or loss less than entire digit.

Assessed only in adult patients:

26. *Claudication:* by history.

ENDOCRINE DAMAGE

Assessed only in Patients < 18 years of age:

27. *Growth failure:* Two of the following three features*:*
    a. Less than 3rd percentile height for age
    b. Growth velocity over 6 months less than 3rd percentile for age
    c. Crossing at least 2 centiles (5%, 10%, 25%, 50%, 75%, 95%) on growth chart
    (See http://www.cdc.gov/nchs/about/major/nhanes/growthcharts/clinical_charts.htm to obtain growth charts for U.S. patients)
28. *Delay in development of secondary sexual characteristics:* greater than 2 standard deviations beyond mean for age in Tanner staging (see Harriet Lane Handbook: A Manual for Pediatric House Officers, Ed. G.K. Sidberry, **or** Tanner, JM and Davies, PS, *J Pediatr* 1985; 107:317-29 for age-defined mean values for North American children).

Assessed in both pediatric and adult patients:

29. *Hirsutism:* excessive terminal hair growth in an adult male distribution, i.e., lip, chin, chest, back. This is to be assessed only in girls, women and pre-pubertal boys. *Hyptertrichosis:* Generalised increase in body hair.
30. *Irregular menses:* Missing more than one ovulatory menstrual cycle (i.e., 24 – 37 days in duration) in one year, assessed at least 3 years after menarche or before menopause.
31. *Primary amenorrhea:* the absence of any menstruation within 4.5 years of reaching puberty (i.e., in a girl without genital malformations who has reached her complete morphological development, including a bone age of at least 15 years, breasts for at least 2 years, and adult type pubic hair)
32. *Secondary amenorrhea*: cessation of menstrual periods before menopause, after initially menstruating
33. *Diabetes mellitus:* fasting glucose > 140 mg/dl or 2 hour glucose in Oral Glucose Tolerance Test > 200 mg/dl
34. *Hyperlipidemia (OPTIONAL ASSESSMENT):* Cholesterol or lipoprotein levels >95% of age appropriate reference ranges
35. References: *Summary of the Second Report of the National Cholesterol Education Program (NCEP) Expert Panel on Detection, Evaluation, and Treatment of High Blood Cholesterol in Adults (Adult Treatment Panel II). JAMA. 1993; 269:3015-3023*; *"Report of the expert panel on blood cholesterol levels in children and adolescents" by the National Cholesterol Education program (NCEP), NIH Publication No 91-2732, September 1991.*

Assessed in adult patients and adolescents when applicable:

36. *Infertility (female or male):* no pregnancy after one year attempting to conceive via regular sexual intercourse
37. *Sexual dysfunction:* patient dissatisfaction with sexual function (female or male)

OCULAR DAMAGE

38. *Cataract:* lens opacity in either eye, ever, whether primary or secondary to steroid therapy, documented by ophthalmoscopy, and resulting in visual loss.
39. *Visual loss, other*: legal blindness, with vision less than or equal to 20/200.

INFECTION

40. *Chronic infection:* Infection (presumptive diagnosis with or without positive culture) requiring > 6 months of antimicrobial treatment, OR persisting with or without clinical symptoms > 6 months and associated with disability (absence from school, work, day-care)
41. *Multiple infections:* > 3 infections (at same or different sites, with same or different organisms) requiring antimicrobial treatment or disability (absence from school, work, day-care) over a 6 month period

MALIGNANCY

42. Documented by pathology, excluding dysplasias, including myositis associated malignancies.

OTHER

43. **Death** should be recorded under OTHER, and the date and cause of death noted (OPTIONAL ASSESSMENT)

44. Any other features of myositis damage not listed but felt to be of importance to the patient or physician. Please specify the feature in as much detail as possible. (These are not scored)

GLOBAL DAMAGE ASSESSMENT

45. Your expert clinical judgement of the totality of disease damage in all systems