

IMACS FORM 07A: MYOSITIS DISEASE ACTIVITY ASSESSMENT TOOL (MDAAT) – 2005, VERSION 2

General Guidelines for Completion:

This is a combined tool that captures the physician's assessment of disease **activity** of various organ systems using (1) the 0-4 scale described below and (2) a visual analog scale (VAS). Please assess the clinical features (items 1-26) of each organ system based upon:

- a) The presence of clinical features or symptoms **within the previous 4 weeks that are due to active disease** (i.e. use your clinical judgment to determine how active the myositis-associated clinical feature has been **within the previous 4 weeks**)
- b) The judgment that the feature is due to the myositis disease process (i.e. clinical findings known or suspected to be due to another disease process or due to therapy should **NOT** be considered in this evaluation)
- c) The concept that disease activity is defined as a potentially reversible finding
- d) A clinical, functional, and laboratory assessment for each organ system:
 - NA = Cannot be assessed
 - 0 = Not present in the last 4 weeks
 - 1 = Improving - clinically significant improvement in the last 4 weeks compared to the previous 4 weeks
 - 2 = The same - manifestations that have been present for the last 4 weeks without significant improvement or deterioration compared to the previous 4 weeks
 - 3 = Worse - clinically significant deterioration over the last 4 weeks compared to the previous 4 weeks
 - 4 = New - in the last 4 weeks (compared to the previous 4 weeks)

Also, rate your overall (global) assessment of the ongoing disease activity over the past 4 weeks for each organ system on the 0-10cm VAS scale (which precedes the listed clinical features) by drawing a **vertical** mark on the 10cm line according to the following guidelines:

- left end of line = no evidence of disease activity
- midpoint of line = moderate disease activity
- right end of line = extreme or maximum disease activity

Please review the glossary as you score each listed clinical feature. The VAS score for each organ system integrates the severity of activity based upon all of the clinical features listed for that particular organ system.

NOTE: The “Extramuscular Global Assessment” is very important as this is a Core Set Measure encompassing an overall evaluation for the disease activity in all the extramuscular organ systems and excludes muscle disease activity.

Guidelines for scoring mild, moderate, severe:

First, identify the category of mild-severe **using the glossary as a guide**. Then score what has happened in the last 4 weeks compared to the previous 4 weeks. Note that with worsening (3) or new (4) activity in the designated category, the same degree of activity should be ascribed in the items that are "less severe." For example:

- In a patient developing new moderate muscle inflammation (see glossary for definition) in the last 4 weeks, “moderate muscle inflammation” (25b) would score a 4 as would “mild muscle inflammation” (25c)
- If “severe muscle inflammation” worsened in the last 4 weeks, then the severe (25a), moderate (25b) and mild (25c) muscle inflammation categories would all score a 3

If a patient had severe muscle inflammation at last visit one month ago and improves to a moderate category over the past 4 weeks (based on the glossary definition), then score the severe category (25a) as a 1 (improving) and score moderate (25b) and mild (25c) as either a 1 or 2 (this would depend on just how much improvement has occurred over the last month so the glossary should be reviewed for this). If one month later the symptoms have further improved, then score the severe category (25a) as a 0 and the moderate (25b) and mild (25c) categories as a 1.

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

Constitutional Disease Activity	(Absent)	(Maximum)	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° Celsius	0	1	2	3	4	NA
2. Weight loss – unintentional > 5%	0	1	2	3	4	NA
3. Fatigue/malaise/lethargy	0	1	2	3	4	NA

Cutaneous Disease Activity	(Absent)	(Maximum)	Examples of maximal score				
	----- -----		- Ulceration to muscle, tendon or bone; - Extensive erythroderma				

4. Cutaneous ulceration	0	1	2	3	4	NA
5. Erythroderma	0	1	2	3	4	NA
6. Panniculitis	0	1	2	3	4	NA
7. Erythematous rashes:						
a. with secondary changes (e.g. accompanied by erosions, vesiculobullous change or necrosis)	0	1	2	3	4	NA
b. without secondary changes	0	1	2	3	4	NA
8. Heliotrope rash	0	1	2	3	4	NA
9. Gottron's papules/sign	0	1	2	3	4	NA
10. Periungual capillary changes	0	1	2	3	4	NA
11. Alopecia:						
a. Diffuse hair loss	0	1	2	3	4	NA
b. Focal, patchy with erythema	0	1	2	3	4	NA
12. Mechanics hands	0	1	2	3	4	NA

Skeletal Disease Activity	(Absent)	(Maximum)	Examples of maximal score			
	----- -----		Severe arthritis with extreme loss of function (bedridden, inability for self care)			

13. Arthritis:						
a. Severe active polyarthritis	0	1	2	3	4	NA
b. Moderately active arthritis	0	1	2	3	4	NA
c. Mild arthritis	0	1	2	3	4	NA
14. Arthralgia	0	1	2	3	4	NA

Gastrointestinal Disease Activity	(Absent)	(Maximum)	Examples of maximal score			
	----- -----		Major abdominal crisis requiring surgery or intensive care			

15. Dysphagia:						
a. Moderate/severe dysphagia	0	1	2	3	4	NA
b. Mild dysphagia	0	1	2	3	4	NA
16. Abdominal pain related to the myositis disease process:						
a. Severe	0	1	2	3	4	NA
b. Moderate	0	1	2	3	4	NA
c. Mild	0	1	2	3	4	NA

Pulmonary Disease Activity	(Absent)	(Maximum)	Examples of maximal score			
	----- -----		Active interstitial lung disease or respiratory muscle weakness requiring ventilatory support			

17. Respiratory muscle weakness without interstitial lung disease (ILD):						
a. Dyspnea at rest	0	1	2	3	4	NA
b. Dyspnea on exertion	0	1	2	3	4	NA
18. Active reversible ILD (i.e. not just ventilatory abnormalities due to pulmonary fibrosis): <i>Read glossary for scoring pulmonary function tests and score each item below (a,b and c).</i>						
a. Dyspnea or cough due to ILD	0	1	2	3	4	NA
b. Parenchymal abnormalities on chest x-ray or high resolution CT scan (HRCT) and/or ground glass shadowing on HRCT	0	1	2	3	4	NA
c. Pulmonary Function Tests: ≥ 10% change in FVC or DLCO	0	1	2	3	4	NA
19. Dysphonia:						
a. Moderate to severe	0	1	2	3	4	NA
b. Mild	0	1	2	3	4	NA

Cardiovascular Disease Activity	(Absent)	(Maximum)	Examples of maximal score				
	----- ----- _____ cm		Myocarditis, pericarditis or severe arrhythmia requiring intensive care unit				

20. Pericarditis	0	1	2	3	4	NA
21. Myocarditis	0	1	2	3	4	NA
22. Arrhythmia:						
a. Severe arrhythmia	0	1	2	3	4	NA
b. Other arrhythmia, except sinus tachycardia	0	1	2	3	4	NA
23. Sinus tachycardia	0	1	2	3	4	NA

Other Disease Activity	(Absent)	(Maximum)	Examples of maximal score				
	----- ----- _____ cm		Extreme disease activity with major impact on function				

24. Specify: _____	0	1	2	3	4	NA
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Extramuscular Global Assessment	(Absent)	(Maximum)	Examples of maximal score				
	----- ----- _____ cm		Overall evaluation for disease activity in all extramuscular systems (EXCLUDING MUSCLE DISEASE ACTIVITY)				

Muscle Disease Activity	(Absent)	(Maximum)	Examples of maximal score				
	----- ----- _____ cm		Severe muscle weakness resulting in being bed bound and an inability to perform self care				

25. Myositis:						
a. Severe muscle inflammation	0	1	2	3	4	NA
b. Moderate muscle inflammation	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	(Absent)	(Maximum)	Examples of maximal score				
	----- ----- _____ cm		Overall evaluation for the totality of disease activity in ALL systems, (INCLUDING MUSCLE DISEASE ACTIVITY)				