

GLOSSARY FOR THE MYOSITIS DISEASE ACTIVITY ASSESSMENT TOOL (MDAAT)

CONSTITUTIONAL

1. Pyrexia: refers to a **documented** fever in excess of 38°C
2. Weight loss: unintentional weight loss of greater than 5% in body weight
3. Fatigue: sufficiently severe to interfere with normal daily activities not attributable to another cause

CUTANEOUS DISEASE ACTIVITY

4. Cutaneous ulceration: extensive injury to dermis or deeper due to dermatomyositis
5. Erythroderma: generalized, widespread confluent erythema involving both sun-exposed and non sun-exposed skin with >50% of body surface area involved
6. Panniculitis: tender erythematous or violaceous nodules or depressions due to inflammation in the subcutaneous fat
7. Erythematous rashes (includes malar rash, facial erythema, linear extensor erythema, V-sign, shawl sign, periungual erythema, sun-exposed or non sun-exposed erythema): (a) **Secondary change**: *erosions*: slightly depressed lesions with denuded epithelium; *vesiculobullous*: fluid-filled lesions ≤ 0.5cm (vesicles) or ≥ 0.5cm (bullae); *necrosis*: dead or devitalized tissue manifested as a black eschar, ulceration or wet or dry gangrene. (b) **Without secondary change**: erythema without accompanying changes to epidermis or dermis
8. Heliotrope rash: purple, lilac-colored or erythematous patches over eyelids or in a periorbital distribution, often associated with periorbital edema
9. Gottron's papules or sign: erythematous to violaceous papules, plaques or macules (sign) over extensor surfaces of joints, which are sometimes scaly
10. Periungual capillary changes: dilatation of periungual capillaries which may be accompanied by vessel dropout and which is visible by naked eye examination or by using additional magnification with otoscopy
11. Alopecia:
 - a) Diffuse: non-scarring, non-erythematous widespread alopecia
 - b) Focal: patchy alopecia with scaling and erythema localized to areas of inflammation (i.e. scalp rash of dermatomyositis)
12. Mechanic's hands: hyperkeratosis and scaling with frequent fissuring and cracking along the lateral and palmar aspects of the fingers

SKELETAL DISEASE ACTIVITY

13. Arthritis: active joint inflammation marked by tenderness, warmth or swelling
 - a) **Severe**: arthritis of **two or more** joints with clinically significant loss of the functional range of movement and requiring assistance with activities of daily living
 - b) **Moderate**: arthritis of **one or more** joints with some loss of functional range of movement, but not requiring assistance with activities of daily living
 - c) **Mild**: arthritis of **one or more** joints with neither loss of range of motion nor impaired activities of daily living
14. Arthralgia: joint pain with or without stiffness but due to an inflammatory process in two or more joints

GASTROINTESTINAL DISEASE ACTIVITY

15. Dysphagia: difficulty swallowing, chewing or eating documented by clinical symptoms or by barium swallow examination, manometry, or other objective measure

If dysphagia has been stable and unchanged and is present for > 6 months, this is most likely to represent a damage item and should be scored as such unless there is good evidence of reversibility.

- a) **Severe**: accompanied by aspiration pneumonia, nasal regurgitation, or difficulty in protecting the airway
 - b) **Moderate**: frequent or moderate symptomatic difficulty swallowing, chewing or eating
 - c) **Mild**: occasional dysphagia or asymptomatic dysphagia noted on objective testing; can eat regular diet
16. Abdominal pain: pain in the abdominal area related to the myositis disease process
 - a) **Severe**: requiring hospitalization, treatment, or bowel rest with nothing per oral route (NPO)
 - b) **Moderate**: requiring treatment, but not hospitalization or NPO
 - c) **Mild**: no intervention required and does not interfere with function

PULMONARY DISEASE ACTIVITY

17. Respiratory muscle weakness without interstitial lung disease (ILD): Shortness of breath worsened with exertion or hypoventilation on pulmonary function testing in the absence of intrinsic lung disease

18. Active reversible ILD: Previously documented by radiography or pathology. Only active features are graded, not those based on pulmonary fibrosis or irreversible features.
- In patients with new or significant deterioration in symptoms a complete evaluation with pulmonary function testing (PFTs) and radiography (chest x-ray or high resolution CT scan) is required. If further investigation is necessary to elucidate the cause of symptoms, then defer scoring until results are available.
 - 18a can always be answered clinically, but 18b and 18c may not be answered if radiography and PFTs (respectively) have not been recently performed.
 - If there has been no significant change since last visit - score 2. If results normalize - score 0. If patient is clinically better but no recent radiography/PFTs are available, score as NA.

Any percentage change in PFTs is based on a change in the absolute value. As a guide a significant change in PFTs is defined as a $\geq 10\%$ change in FVC (minimum 200 ml for adults) or $\geq 15\%$ in DLCO (minimum 3ml/min/mmHg for adults). However, in some patients smaller changes in FVC or DLCO may be suggestive of worsening disease in the context of worsening symptoms or imaging studies.

19. Dysphonia: alteration in voice quality, resonance, articulation or speech rate from normal
- a) *Moderate to severe*: persistent voice symptoms or those that interfere with communication
 - b) *Mild*: intermittent voice symptoms not interfering with communication

CARDIOVASCULAR DISEASE ACTIVITY

20. Pericarditis: Inflammation of the pericardium defined clinically or by electrocardiogram (EKG) or echocardiogram
21. Myocarditis: Inflammation of the myocardium defined clinically or with echocardiographic or other objective evidence
22. Arrhythmia: clinical or electrocardiographic evidence of irregular heart beat
- a) *Severe arrhythmia*: symptomatic and requiring therapy or other intervention, excluding sinus tachycardia
 - b) *Other cardiac arrhythmias*: symptomatic, but not requiring intervention, excluding sinus tachycardia
23. Sinus tachycardia: resting heart rate > 100 beats per minute in an adult patient or greater than upper limit of age-appropriate normal value in a pediatric patient

OTHER DISEASE ACTIVITY

24. Specify feature that is felt to be due to the myositis disease process. Then on the VAS rate the severity of this feature: 'max' would be severe involvement with requirement for intensive care in the case of a systemic feature, or extensive/generalized cutaneous involvement.

EXTRAMUSCULAR GLOBAL ASSESSMENT

Overall evaluation for the disease activity in all extramuscular systems (**excluding** muscle disease activity). **This is an IMACS Core Set Measure.**

MUSCLE DISEASE ACTIVITY

25. Myositis: Muscle inflammation based upon manual muscle strength testing, functional assessments, laboratory or other testing. **In patients with stable muscle atrophy and damage only weakness attributable to active myositis is scored**
- a) *Severe muscle inflammation*: requiring assistance with activities of daily living and severe loss of function
 - b) *Moderate muscle inflammation*: not requiring assistance with activities of daily living with some loss of function
 - c) *Mild muscle inflammation*: little or no loss of function
26. Myalgia: muscle pain or tenderness

GLOBAL DISEASE ACTIVITY

Physician judgment of overall disease activity based on and laboratory assessments. **This is an IMACS Core Set Measure.**