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) **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 ≥10% change in FVC (minimum 200 ml for adults) or ≥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 all clinical and laboratory assessments. **This is an IMACS Core Set Measure.**