Cutaneous Assessment Tool for Myositis- Binary Method

Mark all lesions that are present. Scores for each lesion that is present are shown in parentheses. Absent lesions are scored as 0. The CAT activity and damage scores are obtained by adding the respective item scores. Activity and Damage are assessed independently. Active lesions are assumed to be related to reversible inflammation or vasculopathy. Damage lesions are assumed to be persistent, irreversible and related to prior active disease or therapy.

CUTANEOUS MANIFESTATIONS OF ACTIVITY

| Lesions 1 - 7 include erythematous lesions with or without secondary changes (dyspigmentation, scaling, crusting, erosions, vesicles, bullae, ulceration, necrosis) in a distribution of: | |
|---|-----|
| 1. Gottron's papules/sign | (1) |
| 2. Heliotrope rash | (1) |
| 3. Malar or facial erythema | (1) |
| 4. Linear extensor erythema | (1) |
| 5. "V" sign rash | (1) |
| 6. "Shawl" sign rash | (1) |
| 7. Non-sun exposed erythema | (1) |
| 8. Erythroderma | (1) |
| 9. Livedo reticularis | (1) |
| 10. Ulceration | (1) |
| 11. Mucous membrane lesions | (1) |
| 12. Periungual capillary loop changes | (1) |
| 13. Mechanic's hands | (1) |
| 14. Cuticular overgrowth | (1) |
| 15. Subcutaneous edema | (1) |
| 16. Panniculitis | (1) |
| 17. Alopecia | (1) |
| CAT Activity Score (Sum of activity lesions) | /17 |

CUTANEOUS MANIFESTATIONS OF DAMAGE

Lesions 1 - 7 include atrophy or

| dyspigmentation without erythema in a distribution of: | | |
|--|-----|--|
| 1. Gottron's papules/sign | (1) | |
| 2. Heliotrope rash | (1) | |
| 3. Malar or facial erythema | (1) | |
| 4. Linear extensor erythema | (1) | |
| 5. "V" sign rash | (1) | |
| 6. "Shawl" sign rash | (1) | |
| 7. Non-sun exposed erythema | (1) | |
| | | |
| 18. Poikiloderma vasculare atrophicans | (1) | |
| 19. Calcinosis | (1) | |
| 20. Lipoatrophy | (1) | |
| 21. Depressed scar >0.5 cm | (1) | |
| | | |
| | | |
| | | |

CAT Damage Score

(Sum of damage lesions)

Glossary

Definitions for Activity Lesions

- 1. **Gottron's papules/sign:** erythematous to violaceous papules and small plaques over the extensor surfaces of large or interphalangeal joints. May include secondary changes of scale, crusting, erosions, vesicles, bullae, ulceration or necrosis, as well as dyspigmentation with blanchable erythema. Atrophic lesions and dyspigmented lesions without blanchable erythema should be scored as damage.
- 2. **Heliotrope rash:** violaceous to erythematous periorbital skin eruption, confined to the upper eyelids, with or without edema. May include dyspigmentation with blanchable erythema. Atrophic lesions and dyspigmented lesions without blanchable erythema should be scored as damage.
- 3. **Malar/facial erythema:** erythema over the face which may be isolated malar erythema, but may include more extensive erythema including perioral temporal, ear and frontal regions. May include secondary changes of scale, crusting, erosions, vesicles, bullae, ulceration or necrosis, as well as dyspigmentation with blanchable erythema. Atrophic lesions and dyspigmented lesions without blanchable erythema should be scored as damage.
- 4. **Linear extensor erythema:** erythema specifically located over the extensor tendon sheaths of the hands, forearms, feet and/or forelegs, which spares the skin over the extensor joint surfaces. May include secondary changes of scale, crusting, erosions, vesicles, bullae, ulceration or necrosis, as well as dyspigmentation with blanchable erythema. Atrophic lesions and dyspigmented lesions without blanchable erythema should be scored as damage.
- 5. **"V" sign rash:** discrete confluent and erythematous macular eruption over the lower anterior neck and upper anterior chest. May include secondary changes of scale, crusting, erosions, vesicles, bullae, ulceration or necrosis, as well as dysrpigmentation with blanchable erythema. Atrophic lesions and dyspigmented lesions without blanchable erythema should be scored as damage.
- 6. "Shawl" sign rash: discrete confluent and erythematous macular eruption in a shawl distribution over the upper back, posterior neck, and shoulders, sometimes extending to the lateral arms. May include secondary changes of scale, crusting, erosions, vesicles, bullae, ulceration or necrosis, as well as dyspigmentation with blanchable erythema. Atrophic lesions and dyspigmented lesions without blanchable erythema should be scored as damage.
- 7. **Non-sun exposed erythema:** erythema in areas not exposed to sun i.e. usually covered by clothing or in protected areas such as under the chin, flexural areas, palms, soles, trunk, groin, etc; found in addition to or exclusive of erythema described in items 3-6. May include secondary changes of scale, crusting, erosions, vesicles, bullae, ulceration or necrosis, as well as dyspigmentation with blanchable erythema. Atrophic lesions and dyspigmented lesions without blanchable erythema should be scored as damage.
- 8. **Erythroderma:** extensive areas of confluent erythema, involving both sun exposed and non-sun exposed skin; can involve the entire
- 9. **Livedo reticularis:** a fixed peripheral vascular condition characterized by a bluish-reddish netlike mottling of the skin that is found on the trunk or the extremities. The discoloration persists after the skin has been warmed.
- 10. **Ulceration:** injury to dermis, subcutaneous or deeper soft tissue, secondary to vascular insufficiency, trauma, infection, or unknown etiology. This category is for isolated ulceration. Other types of ulceration are in other categories: i.e., Gottron's ulceration (#1), ulceration with erythema (#3-6), mucous membrane ulceration (#12), ulceration with panniculitis (#18), and ulceration with calcinosis (#21).
- 11. **Mucous membrane lesions:** macules, dilated gingival capillaries, erosions/apthae, or ulceration (injury to submucosa or deeper) involving oral, nasal, gingival, or genital regions.

- 12. **Periungual capillary loop changes:** dilatation of capillaries which may be accompanied by vessel dropout (with or without magnification).
- 13. **Mechanic's hands:** lesions on the palmar or lateral aspects of the digits which can include fissuring, cracking, hyperkeratosis, scaling, hyperpigmentation.
- 14. Cuticular overgrowth: enlargement and overgrowth of the cuticle onto the nailbed.
- 15. Subcutaneous edema: swelling of skin and soft tissue which may be localized or generalized.
- 16. **Panniculitis:** painful erythematous or violaceous subcutaneous nodules or depressions due to inflammation in the subcutaneous fat (biopsy documentation preferable). These lesions may ulcerate.
- 17. **Alopecia:** hair loss. Should be non-scarring, but may be localized or diffuse, and may be with or without erythema.

Definitions for damage lesions

- 1. **Gottron's papules/sign:** atrophic lesions (epidermal or dermal) with or without telangiectasia OR dyspigmentation without blanchable erythema in a distribution of Gottron's papules or Gottron's sign.
- 2. **Heliotrope rash:** atrophic lesions (epidermal or dermal) with or without telangiectasia OR dyspigmentation without blanchable erythema in a distribution of heliotrope rash.
- 3. **Malar/facial erythema:** atrophic lesions (epidermal or dermal) with or without telangiectasia OR dyspigmentation without blanchable erythema in a distribution of malar or facial erythema.
- 4. **Linear extensor erythema:** atrophic lesions (epidermal or dermal) with or without telangiectasia OR dyspigmentation without blanchable erythema in a distribution of linear extensor erythema.
- 5. **"V" sign rash:** atrophic lesions (epidermal or dermal) with or without telangiectasia OR dyspigmentation without blanchable erythema in a distribution of "V" sign rash.
- 6. **"Shawl" sign rash:** atrophic lesions (epidermal or dermal) with or without telangiectasia OR dyspigmentation without blanchable erythema in a distribution of "shawl" sign rash.
- 7. **Non-sun exposed erythema:** atrophic lesions (epidermal or dermal) with or without telangiectasia OR dyspigmentation without blanchable erythema in a distribution of non-sun exposed erythema.
- 18. **Poikiloderma vasculare atrophicans:** a fine speckled pattern of hyperpigmented and hypopigmented macules interspersed with fine telangiectasia and cutaneous atrophy. Usually occurs in areas of photosensitivity eruption. May arise de novo.
- 19. **Calcinosis:** dystrophic calcification in the subcutaneous tissue, muscle, interfascial planes, or across ioints
- 20. **Lipoatrophy:** loss of subcutaneous fat which may be localized or more widely distributed. May be accompanied by hepatomegaly, hirsutism, acanthosis nigricans, hyperlipidemia, hyperinsulinemia.
- 21. **Depressed scar > 0.5 cm:** end stage of lesions due to vascular occlusion or vascular insufficiency manifested as depressions due to atrophy or scarring. May be the end stage of ulceration, purpura, vascular insufficiency seen in Raynaud's, panniculitis, or infection healing with atrophy, depression and scarring (this may include scarring alopecia).