

Cutaneous Assessment Tool for Myositis

The Cutaneous Assessment Tool (CAT) is designed to catalogue and grade the severity of the cutaneous lesions found in the adult and juvenile idiopathic inflammatory myopathies. A wide spectrum of both classical and more obscure cutaneous lesions is listed with definitions, followed by a list of descriptors which will be used to grade the severity of the lesion. Grading is based on: (a) absence or presence of a given lesion; (b) presence of a primary finding without secondary changes; and (c) presence of a primary lesion associated with different degrees of secondary changes. Some lesions are graded by absence or presence, with severity depending upon the extent and/or site of the lesion. By choosing from the list associated with a lesion, an observer can describe the major characteristics of the lesion in a given patient. Scores were assigned a priori by the investigators based on expert opinion on the relative importance of individual lesions and their degree of activity or damage.

To complete the questionnaire, assess each lesion by checking all items that describe it. If the lesion is absent, indicate this. Every item should be assessed. More than one item may be chosen for each lesion (e.g. item 2 heliotrope rash may include edema and hypo/hyperpigmentation with erythema, or either descriptor alone). Complete the assessment in the order specified. After completing the questionnaire, rate the activity and damage of the skin both by 10 cm visual analogue scales and by 5 point Likert scales (page 9).

The CAT is organized to catalogue active lesions, and irreversible lesions or cumulative damage. Activity is associated with underlying inflammation or vasculopathy. Active lesions include those that are characteristic, erythematous vasculopathic, hand changes and other (panniculitis and alopecia). Persistent manifestations can be thought of in two ways: a) persistent lesions that have a low level of continued activity, yet demonstrate residual changes (such as poikiloderma vasculare atrophicans or calcinosis) and b) irreversible lesions such as scars. Damage constitutes lesions that persistent for at least 6 months that are which result from prior active disease or therapy. Changes associated with damage are often post inflammatory, cumulative, or irreversible.

Index of Lesions

ACTIVE CUTANEOUS MANIFESTATIONS	
Characteristic Lesions:	Vasculopathic Lesions:
<u>1A. Gottron's papules or Gottron's sign</u>	<u>9A. Livedo reticularis</u>
<u>2A. Heliotrope rash</u>	<u>10A. Ulceration</u>
Erythematous Lesions:	<u>11A. Mucous membrane lesions</u>
<u>3A. Malar or facial erythema</u>	<u>12A. Periungual capillary loop changes</u>
<u>4A. Linear extensor erythema</u>	Hand Lesions:
<u>5A. "V" sign rash</u>	<u>13A. Mechanic's hands</u>
<u>6A. "Shawl" sign rash</u>	<u>14A. Cuticular overgrowth</u>
<u>7A. Non-sun exposed erythema</u>	Other Active Lesions:
<u>8A. Erythroderma</u>	<u>15A. Subcutaneous edema</u>
	<u>16A. Panniculitis</u>
	<u>17A. Alopecia</u>
CUTANEOUS MANIFESTATIONS OF DAMAGE	
<u>1D – 7D. Atrophy or dyspigmentation without blanchable erythema in a distribution of the stated lesion</u>	
<u>18D. Poikiloderma vasulare atrophicans</u>	
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<u>21D. Depressed scar</u>	

Example of how to complete the CAT:

A patient has some Gottron's papules which are hyperpigmented with blanching on compressing the lesions and many Gottron's papules which are erythematous-violaceous with secondary changes. Each of these descriptors would be checked on the form. All other descriptors for this lesion, which are not present in this patient, would be left blank.

1. Gottron's papules OR Gottron's sign: erythematous to violaceous papules and small plaques over the extensor surfaces of large or interphalangeal joints.

- absent
- hypo/hyperpigmentation with erythema (blanching occurs on compression of the lesion and/or erythema is distinguishable at the advancing border): resolution stage
- erythematous-violaceous papules, or hyperpigmented papules in a dark-skinned person
- erythematous-violaceous papules, or hyperpigmented papules in a dark-skinned person, accompanied by secondary changes of scale, crusting, or erosions
- erythematous-violaceous papules, or hyperpigmented papules in a dark-skinned person, with vesicles, bullae, ulceration, or necrosis

Each of the 21 items would be similarly assessed.

CUTANEOUS ASSESSMENT TOOL (CAT)

Instructions:

Assess each lesion by checking all descriptors that describe it. If the lesion is absent, indicate this. Every item should be assessed. More than one box may be chosen for each lesion. Complete the questionnaire in the order specified. Also rate the global activity and damage of the skin disease using the 10 cm visual analogue scales and the 5 point Likert scales.

ACTIVE CUTANEOUS MANIFESTATIONS: Lesions 1A – 7A, 8A – 17A

Characteristic lesions:

1. Gottron's papules OR Gottron's sign: erythematous to violaceous papules and small plaques over the extensor surfaces of large or interphalangeal joints.

1A. Activity:

- absent
- hypo/hyperpigmentation with erythema (blanching occurs on compression of the lesion and/or erythema is distinguishable at the advancing border): resolution stage
- erythematous-violaceous papules, or hyperpigmented papules in a dark-skinned person
- erythematous-violaceous papules, or hyperpigmented papules in a dark-skinned person, accompanied by secondary changes of scale, crusting, or erosions
- erythematous-violaceous papules, or hyperpigmented papules in a dark-skinned person, with vesicles, bullae, ulceration, or necrosis

1D. Damage:

- absent
- atrophy (epidermal or dermal) with or without telangiectasia in a distribution of Gottron's papules or Gottron's sign
- hypo/hyperpigmentation without blanchable erythema in a distribution of Gottron's papules or Gottron's sign

2. Heliotrope rash: violaceous to erythematous periorbital skin eruption, confined to the upper eyelids, with or without edema.

2A. Activity:

- absent
- hypo/hyperpigmentation with erythema (blanching occurs on compression of the lesion and/or erythema is distinguishable at the advancing border): resolution stage
- faint erythema or faint violaceous rash, or faint hyperpigmentation in a dark-skinned person
- intense erythema or intense violaceous rash, or intense hyperpigmentation in a dark-skinned person
- edema

2D. Damage:

- absent
- atrophy (epidermal or dermal) with or without telangiectasia in a distribution of heliotrope rash
- hypo/hyperpigmentation without blanchable erythema in a distribution of heliotrope rash

Erythema: erythematous discrete and confluent macular eruption.

Erythematous lesions are numbered 3-7:

3. Malar OR 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.

3A. Activity:

- absent
- hypo/hyperpigmentation with erythema (blanching occurs on compression of the lesion and/or erythema is distinguishable at the advancing border): resolution stage
- erythema, or hyperpigmentation in a dark-skinned person, without secondary changes (no scale, crust, erosions)
- erythema, or hyperpigmentation in a dark-skinned person, accompanied by secondary changes of scale, crusting or erosions.
- erythema, or hyperpigmentation in a dark-skinned person, accompanied by vesicles, bullae or ulceration.

3D. Damage:

- absent
- atrophy (epidermal or dermal) with or without telangiectasia in a distribution of malar or facial erythema
- hypo/hyperpigmentation without blanchable erythema in a distribution of malar or facial erythema

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.

4A. Activity:

- absent
- hypo/hyperpigmentation with erythema (blanching occurs on compression of the lesion and/or erythema is distinguishable at the advancing border): resolution stage
- erythema, or hyperpigmentation in a dark-skinned person, without secondary changes (no scale, crust, erosions)
- erythema, or hyperpigmentation in a dark-skinned person, accompanied by secondary changes of scale, crusting or erosions.
- erythema, or hyperpigmentation in a dark-skinned person, accompanied by vesicles, bullae or ulceration.

4D. Damage:

- absent
- atrophy (epidermal or dermal) with or without telangiectasia in a distribution of linear extensor erythema
- hypo/hyperpigmentation without blanchable erythema in a distribution of linear extensor erythema

5. "V" sign rash: discrete confluent and erythematous macular eruption over the lower anterior neck and upper anterior chest.

5A. Activity:

- absent
- hypo/hyperpigmentation with erythema (blanching occurs on compression of the lesion and/or erythema is distinguishable at the advancing border): resolution stage
- erythema, or hyperpigmentation in a dark-skinned person, without secondary changes (no scale, crust, erosions)
- erythema, or hyperpigmentation in a dark-skinned person, accompanied by secondary changes of scale, crusting or erosions.
- erythema, or hyperpigmentation in a dark-skinned person, accompanied by vesicles, bullae, or ulceration.

5D. Damage:

- absent
- atrophy (epidermal or dermal) with or without telangiectasia in a distribution of "V" sign rash
- hypo/hyperpigmentation without blanchable erythema in a distribution of "V" sign rash

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.

6A. Activity:

- absent
- hypo/hyperpigmentation with erythema (blanching occurs on compression of the lesion and/or erythema is distinguishable at the advancing border): resolution stage
- erythema, or hyperpigmentation in a dark-skinned person, without secondary changes (no scale, crust, erosions)
- erythema, or hyperpigmentation in a dark-skinned person, accompanied by secondary changes of scale, crusting or erosions.
- erythema, or hyperpigmentation in a dark-skinned person, accompanied by vesicles, bullae or ulceration.

6D. Damage:

- absent
- atrophy (epidermal or dermal) with or without telangiectasia in a distribution of "shawl" sign rash
- hypo/hyperpigmentation without blanchable erythema in a distribution of "shawl" sign rash

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.

7A. Activity:

- absent
- hypo/hyperpigmentation with erythema (blanching occurs on compression of the lesion and/or erythema is distinguishable at the advancing border): resolution stage
- erythema, or hyperpigmentation in a dark-skinned person, without secondary changes (no scale, crust, erosions)
- erythema, or hyperpigmentation in a dark-skinned person, accompanied by secondary changes of scale, crusting or erosions.
- erythema, or hyperpigmentation in a dark-skinned person, accompanied by vesicles, bullae or ulceration.

7D. Damage:

- absent
- atrophy (epidermal or dermal) with or without telangiectasia in a distribution of non-sun exposed erythema
- hypo/hyperpigmentation without blanchable erythema in a distribution of non-sun exposed erythema

8A. Erythroderma: extensive areas of confluent erythema, involving both sun exposed and non-sun exposed skin; can involve the entire

- absent
- present

Vasculopathic Lesions: Items 9 - 12

9A. 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.

- absent
- present, occupying 1-10% body surface area
- present, occupying 10-25% body surface area
- present, occupying > 25% body surface area

10A. 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).

- absent
- ulcer(s) with depth to dermis/subcutis (fat)
- ulcer(s) with depth to muscle, tendon, or bone

11A. Mucous membrane lesions: macules, dilated gingival capillaries, erosions/aphthae, or ulceration (injury to submucosa or deeper) involving oral, nasal, gingival, or genital regions.

- absent
- macular lesions
- gingival erythema and/or swelling involving dilated gingival capillaries
- erosions/aphthae
- ulceration

12A. Periungual capillary loop changes: dilatation of capillaries which may be accompanied by vessel dropout.

- absent
- periungual erythema
- capillary loop changes present: detected only by otoscopy using additional magnification aids (oil or water)
- capillary loop changes present: visible to naked eye

Hand changes: Items 13-14

13A. Mechanic's hands: lesions on the palmar or lateral aspects of the digits which can include fissuring, cracking, hyperkeratosis, scaling, hyperpigmentation.

- absent
- superficial scale, dryness, or hyperpigmentation only
- cracking and fissuring without bleeding
- cracking and fissuring with bleeding

14A. Cuticular overgrowth: enlargement and overgrowth of the cuticle onto the nailbed.

- absent
- cuticular overgrowth present in 1 - 4 digits
- cuticular overgrowth present in ≥ 5 digits

Other:

15A. Subcutaneous edema: swelling of skin and soft tissue which may be localized or generalized.

- absent
- present, occupying 1-10% body surface area
- present, occupying 10-25% body surface area
- present, occupying $> 25\%$ body surface area

16A. Panniculitis: painful erythematous or violaceous subcutaneous nodules or depressions due to inflammation in the subcutaneous fat (biopsy documentation preferable). These lesions may ulcerate.

- absent
- tender nodules or plaques
- tender nodules or plaques with ulceration

17A. Alopecia: hair loss.

Diffuse: Nonscarring, non-erythematous widespread alopecia (telogen effluvium)

Focal: Focal patchy alopecia with erythema (localized to areas of inflammation)

- absent
- diffuse
- focal/patchy

Other active manifestation: _____

CUTANEOUS MANIFESTATIONS OF DAMAGE: Lesions 1 D– 7D, 18D – 21D.

Atrophy or dyspigmentation without blanchable erythema in a distribution of lesions 1D – 7D (see pages 3 - 6)

18D. 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.

- absent
- poikiloderma occupying 1-10% body surface area
- poikiloderma occupying 10-25% body surface area
- poikiloderma occupying > 25% body surface area

19D. Calcinosis: dystrophic calcification in the subcutaneous tissue, muscle, interfascial planes, or across joints

- absent
- superficial plaques or nodules
- calcinosis circumscripta (large tumorous deposits) or calcinosis universalis (intermuscular fascial plane deposition)
- extensive subcutaneous exoskeleton or calcinosis with ulceration

20D. Lipoatrophy: loss of subcutaneous fat which may be localized or more widely distributed and accompanied by hyperlipidemia.

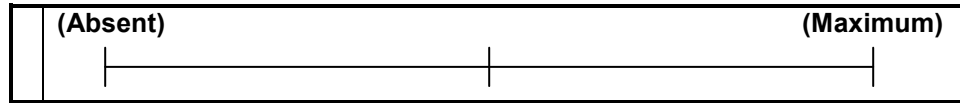
- absent
- localized: focal areas of subcutaneous fat loss
- partial: involving face, upper limbs and shoulder, possibly accompanied by complement abnormalities
- total: widespread accompanied by some of the following: hepatomegaly, hirsutism, acanthosis nigricans, hyperlipidemia, hyperinsulinemia.

21D. 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).

- absent
- present

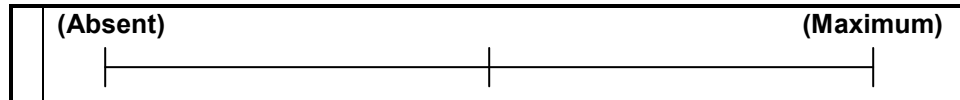
Other manifestation of damage: _____

After completing the assessment tool (items 1-21), rate the activity and damage of the skin disease on the following 10 cm visual analogue scales and 5 point Likert scales



| no evidence of Skin disease activity | extreme skin disease activity

- _____ 0 = no evidence of skin disease activity
- _____ 1 = mild skin disease activity
- _____ 2 = moderate skin disease activity
- _____ 3 = severe skin disease activity
- _____ 4 = extremely active skin disease



no evidence of skin disease damage | extreme skin disease damage

- _____ 0 = no evidence of skin disease damage
- _____ 1 = mild skin disease damage
- _____ 2 = moderate skin disease damage
- _____ 3 = severe skin disease damage
- _____ 4 = extreme skin disease damage

CUTANEOUS ASSESSMENT TOOL (CAT) (SCORING)

For each lesion, check all boxes that apply. (more than one box may be marked per lesion)

ACTIVE CUTANEOUS LESIONS: Lesions 1A – 7A, 8A – 17A.

Characteristic lesions:

1. Gottron's papules OR Gottron's sign: erythematous to violaceous papules and small plaques over the extensor surfaces of large or interphalangeal joints.

1A. Activity:

- 0 absent
- 1 hypo/hyperpigmentation with erythema (blanching occurs on compression of the lesion and/or erythema is distinguishable at the advancing border): resolution stage
- 2 erythematous-violaceous papules, or hyperpigmented papules in a dark-skinned person
- 3 erythematous-violaceous papules, or hyperpigmented papules in a dark-skinned person, accompanied by secondary changes of scale, crusting, or erosions
- 4 erythematous-violaceous papules, or hyperpigmented papules in a dark-skinned person, with vesicles, bullae, ulceration, or necrosis

1D. Damage:

- 0 absent
- 0 for Activity (A), 1 for Damage (D) atrophy (epidermal or dermal) with or without telangiectasia in a distribution of Gottron's papules or Gottron's sign
- 0 for A, 1 for D hypo/hyperpigmentation without blanchable erythema in a distribution of Gottron's papules or Gottron's sign

2. Heliotrope rash: violaceous to erythematous periorbital skin eruption, confined to the upper eyelids, with or without edema.

2A. Activity:

- 0 absent
- 1 hypo/hyperpigmentation with erythema (blanching occurs on compression of the lesion and/or erythema is distinguishable at the advancing border): resolution stage
- 2 faint erythema or faint violaceous rash, or faint hyperpigmentation in a dark-skinned person
- 3 intense erythema or intense violaceous rash, or intense hyperpigmentation in a dark-skinned person
- 1 edema

2D. Damage:

- 0 absent
- 0 for A, 1 for D atrophy (epidermal or dermal) with or without telangiectasia in a distribution of heliotrope rash
- 0 for A, 1 for D hypo/hyperpigmentation without blanchable erythema in a distribution of heliotrope rash

Erythema: erythematous discrete and confluent macular eruption.

Erythematous lesions:

3. Malar OR 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.

3A. Activity:

- 0 absent
- 1 hypo/hyperpigmentation with erythema (blanching occurs on compression of the lesion and/or erythema is distinguishable at the advancing border): resolution stage
- 2 erythema, or hyperpigmentation in a dark-skinned person, without secondary changes (no scale, crust, erosions)
- 3 erythema, or hyperpigmentation in a dark-skinned person, accompanied by secondary changes of scale, crusting or erosions.
- 4 erythema, or hyperpigmentation in a dark-skinned person, accompanied by vesicles, bullae or ulceration.

3D. Damage:

- 0 absent
- 0 for A, 1 for D___ atrophy (epidermal or dermal) with or without telangiectasia in a distribution of malar or facial erythema
- 0 for A, 1 for D___ hypo/hyperpigmentation without blanchable erythema in a distribution of malar

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.

4A. Activity:

- 0 absent
- 1 hypo/hyperpigmentation with erythema (blanching occurs on compression of the lesion and/or erythema is distinguishable at the advancing border): resolution stage
- 2 erythema, or hyperpigmentation in a dark-skinned person, without secondary changes (no scale, crust, erosions)
- 3 erythema, or hyperpigmentation in a dark-skinned person, accompanied by secondary changes of scale, crusting or erosions.
- 4 erythema, or hyperpigmentation in a dark-skinned person, accompanied by vesicles, bullae or ulceration.

4D. Damage:

- 0 absent
- 0 for A, 1 for D___ atrophy (epidermal or dermal) with or without telangiectasia in a distribution of linear extensor erythema
- 0 for A, 1 for D___ hypo/hyperpigmentation without blanchable erythema in a distribution of linear extensor erythema

5. "V" sign rash: discrete confluent and erythematous macular eruption over the lower anterior neck and upper anterior chest.

5A. Activity:

- 0 absent
- 1 hypo/hyperpigmentation with erythema (blanching occurs on compression of the lesion and/or erythema is distinguishable at the advancing border): resolution stage
- 2 erythema, or hyperpigmentation in a dark-skinned person, without secondary changes (no scale, crust, erosions)

- 3 erythema, or hyperpigmentation in a dark-skinned person, accompanied by secondary changes of scale, crusting or erosions.
- 4 erythema, or hyperpigmentation in a dark-skinned person, accompanied by vesicles, bullae, or ulceration.

5D. Damage:

- 0 absent
- 0 for A, 1 for D atrophy (epidermal or dermal) with or without telangiectasia in a distribution of "V" sign rash
- 0 for A, 1 for D hypo/hyperpigmentation without blanchable erythema in a distribution of "V" sign rash

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.

6A. Activity:

- 0 absent
- 1 hypo/hyperpigmentation with erythema (blanching occurs on compression of the lesion and/or erythema is distinguishable at the advancing border): resolution stage
- 2 erythema, or hyperpigmentation in a dark-skinned person, without secondary changes (no scale, crust, erosions)
- 3 erythema, or hyperpigmentation in a dark-skinned person, accompanied by secondary changes of scale, crusting or erosions.
- 4 erythema, or hyperpigmentation in a dark-skinned person, accompanied by vesicles, bullae or ulceration.

6D. Damage:

- 0 absent
- 0 for A, 1 for D atrophy (epidermal or dermal) with or without telangiectasia in a distribution of "shawl" sign rash
- 0 for A, 1 for D hypo/hyperpigmentation without blanchable erythema in a distribution of "shawl" sign rash

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.

7A. Activity:

- 0 absent
- 1 hypo/hyperpigmentation with erythema (blanching occurs on compression of the lesion and/or erythema is distinguishable at the advancing border): resolution stage
- 2 erythema, or hyperpigmentation in a dark-skinned person, without secondary changes (no scale, crust, erosions)
- 3 erythema, or hyperpigmentation in a dark-skinned person, accompanied by secondary changes of scale, crusting or erosions.
- 4 erythema, or hyperpigmentation in a dark-skinned person, accompanied by vesicles, bullae or ulceration.

7D. Damage:

- 0 absent

- 0 for A, 1 for D atrophy (epidermal or dermal) with or without telangiectasia in a distribution of non-sun exposed erythema
- 0 for A, 1 for D hypo/hyperpigmentation without blanchable erythema in a distribution of non-sun exposed erythema

8A. Erythroderma: extensive areas of confluent erythema, involving both sun exposed and non-sun exposed skin; can involve the entire

- 0 absent
- 8 present

Vasculopathic Lesions:

9A. 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.

- 0 absent
- 2 present, occupying 1-10% body surface area
- 3 present, occupying 10-25% body surface area
- 4 present, occupying > 25% body surface area

10A. Ulceration: injury to dermis or deeper, 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).

- 0 absent
- 15 ulcer(s) with depth to dermis/subcutis (fat)
- 30 ulcer(s) with depth to muscle, tendon, or bone

11A. Mucous membrane lesions: macules, dilated gingival capillaries, erosions/apthae, or ulceration (injury to submucosa or deeper) involving oral, nasal, gingival, or genital regions.

- 0 absent
- 1 macular lesions
- 2 gingival erythema and/or swelling involving dilated gingival capillaries
- 3 erosions/apthae
- 4 ulceration

12A. Periungual capillary loop changes: dilatation of beginning, middle and end of capillaries which may be accompanied by vessel dropout.

- 0 absent
- +1 periungual erythema
- 2 capillary loop changes present: detected only by otoscopy using additional magnification aids (oil or water)
- 3 capillary loop changes present: visible to naked eye

Hand Lesions:

13A. Mechanic's hands: lesions on the palmar or lateral aspects of the digits which can include fissuring, cracking, hyperkeratosis, scaling, hyperpigmentation.

- 0 absent
- 1 superficial scale, dryness, or hyperpigmentation only
- 2 cracking and fissuring without bleeding
- 3 cracking and fissuring with bleeding

14A. Cuticular overgrowth: enlargement and overgrowth of the cuticle onto the nailbed.

- 0 absent
- 1 cuticular overgrowth present in 1 - 4 digits
- 2 cuticular overgrowth present in ≥ 5 digits

Other:

15A. Subcutaneous edema: swelling of skin and soft tissue which may be localized or generalized.

- 0 absent
- 1 present, occupying 1-10% body surface area
- 2 present, occupying 10-25% body surface area
- 3 present, occupying $> 25\%$ body surface area

16A. Panniculitis: painful erythematous or violaceous nodules or depressions which may or may not ulcerate, due to inflammation in the subcutaneous fat (biopsy documentation preferable).

- 0 absent
- 4 tender nodules or plaques
- 8 tender nodules or plaques with ulceration

17A. Alopecia: hair loss.

Diffuse: Nonscarring, non-erythematous widespread alopecia (telogen effluvium)

Focal: Focal patchy alopecia with erythema (localized to areas of inflammation)

- 0 absent
- 1 diffuse
- 2 focal/patchy

Other active manifestation: Not scored _____

CUTANEOUS MANIFESTATIONS OF DAMAGE: Lesions 1D – 7D, 18D – 21D

Atrophy or dyspigmentation without blanchable erythema in a distribution of lesions 1 – 7 (see pages 11 - 13)

18D. 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.

- 0 absent
- 1 poikiloderma occupying 1-10% body surface area
- 2 poikiloderma occupying 10-25% body surface area
- 3 poikiloderma occupying > 25% body surface area

19D. Calcinosis: dystrophic calcification in the subcutaneous tissue, muscle, interfascial planes, or across joints

- 0 absent
- +1 superficial plaques or nodules
- +2 calcinosis circumscripta (large tumerous deposits) or calcinosis universalis (intermuscular fascial plane deposition)
- +3 extensive subcutaneous exoskeleton or calcinosis with ulceration

20D. Lipoatrophy: loss of subcutaneous fat which may be localized or more widely distributed and accompanied by hyperlipidemia.

- 0 absent
- 1 localized: focal areas of subcutaneous fat loss
- 2 partial: involving face, upper limbs and shoulder, possibly accompanied by complement abnormalities
- 3 total: widespread accompanied by some of the following: hepatomegaly, hirsutism, acanthosis nigricans, hyperlipidemia, hyperinsulinemia.

21D. 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).

- 0 absent
- 1 present

Other chronic manifestation: Not scored_____