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.

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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/apthae, 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/apthae
- 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.
17A. Alopecia: hair loss.
   Diffuse: Non-scarring, 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 1D – 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 hyperpigmentated and hypopigmentated 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 circumspecta (large tumerous 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

<table>
<thead>
<tr>
<th>(Absent)</th>
<th>(Maximum)</th>
</tr>
</thead>
<tbody>
<tr>
<td></td>
<td></td>
</tr>
<tr>
<td>no evidence of</td>
<td>extreme skin</td>
</tr>
<tr>
<td>Skin disease activity</td>
<td>disease activity</td>
</tr>
</tbody>
</table>

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

<table>
<thead>
<tr>
<th>(Absent)</th>
<th>(Maximum)</th>
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<tbody>
<tr>
<td></td>
<td></td>
</tr>
<tr>
<td>no evidence of</td>
<td>extreme skin</td>
</tr>
<tr>
<td>skin disease damage</td>
<td>disease damage</td>
</tr>
</tbody>
</table>

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/aphthae, 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/aphthae
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.

- **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 hyperpigmentated and hypopigmentated 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 circumspecta (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_________________________________________________________