

Supplemental Online Content

Sun Q, Asch S, Bayart C, et al. Development and initial validation of a novel system to assess ichthyosis severity. *JAMA Dermatol*. Published online February 16, 2022. doi:10.1001/jamadermatol.2021.5917

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This supplemental material has been provided by the authors to give readers additional information about their work.

eFigure 1. Anatomical definitions. Specific delineations for each body region scored were provided for all raters

Head and neck

- **Face:** extends from the forehead to the chin and includes the mouth, nose, cheeks, eyes and ears. It excludes the neck. (1/3)
- **Scalp:** extends from the junction of forehead skin to the occiput (back of head). It excludes the face. (1/3)
- **Neck:** extends from the mandible to the attachment of the sternocleidomastoid at the clavicle. It also includes the back of the neck, which extends from the C1 vertebrae to the trapezius muscle. (1/3)

Torso – Includes back, chest, abdomen, and groin

- **Chest and abdomen:** extends from one anterior axillary fold to the other and from the clavicles to the anterior iliac spine
- **Back:** extends laterally from one posterior axillary fold to the other and from T1-S1. It excludes posterior arms and neck.
- **Groin:** Extends from anterior superior iliac spine (ASIS) supero-laterally and includes the lower portion of the anterior abdominal wall, ending at the pubic tubercle medially.

Upper extremities: extends from the acromial process to the fingers and includes the anterior axillary fold and antecubital fossa. (*It excludes the elbow and any portion of the trunk that may be visible.*)

Lower extremities: extends from the superior iliac spine to the toes and includes the buttocks. It excludes knees and soles, which are assessed separately.

Elbows and knees

- **Elbows:** Includes attachment of the humerus to the proximal radius and ulna. It excludes antecubital fossa.
- **Knees:** Includes attachment of the femur to the tibia and patella. It excludes the popliteal fossa.

Palms: extends from the tip of the phalanges to the carpal bones (wrist joint) and includes the area between the five phalanges. It includes glabrous skin.

Soles: extends from the tip of the toes to the calcaneus (heel). It includes glabrous skin.

By contrast, because erythema can be obscured by scale, the erythema score for a particular region should be the worst score for that region.

eTable 1. Score calculations for (a) individuals <8 years old and (b) individuals ≥8 years old*

(a)

Body region	Scale (0-4)	Erythema (0-4)	Multiplier	Score per region
Head				
Scalp	(+)		X 0.067	
Face	(+)		X 0.067	
Neck	(+)		X 0.067	
Trunk				
Chest/Back Abdomen	(+)		X 0.297	
Groin	(+)		X 0.003	
Upper extremities				
Palms	(+)		X 0.004	
Elbows	(+)		X 0.004	
Rest of upper extremities	(+)		X 0.192	
Lower extremities				
Knees	(+)		X 0.012	
Soles	(+)		X 0.009	
Rest of lower extremities	(+)		X 0.279	
Final score (Range: 0-8)				Final score:

(b)

Body region	Scale (0-4)	Erythema (0-4)	Multiplier	Score per region
Head				
Scalp	(+)		X 0.033	
Face	(+)		X 0.033	
Neck	(+)		X 0.033	
Trunk				
Chest/Back Abdomen	(+)		X 0.297	
Groin	(+)		X 0.003	
Upper extremities				
Palms	(+)		X 0.004	

Elbows	(+)	X 0.004	
Rest of upper extremities	(+)	X 0.192	
Lower extremities					
Knees	(+)	X 0.016	
Soles	(+)	X 0.012	
Rest of lower extremities	(+)	X 0.372	
Final score (Range: 0-8)					Final score:

*The final score was calculated by first multiplying the sum of scale (0-4) and erythema (0-4) by the multiplier for each body region (which differs for those younger than 8 years and those 8 years and older) and then adding the scores for all 10 regions

eFigure 2. Instructions. Specific instructions on how to score the test photo set were provided for each rater.

Scoring overview

Scores will grade: 1) **scale severity** and 2) **erythema**.

Scoring for scale should be done for the entire body site. It should be an **average** of affected areas within that site, not the worst score for that body site.

Half-points are allowed. 2 limbs=one body site.

Erythema score for a particular site should be the **worst** score for that site because erythema can be obscured by scale.

You will have one month to:

1. Score the **average** scale severity (0-4, half points allowed) for each body site. Use the standard photos and descriptors to guide you.
2. Score erythema severity (0-4, half points allowed) for each site. Use the standard photos and descriptors to guide you.
3. Enter your scores for each patient into the scoring sheet.
4. After you have completed scoring for all test patients, please mail back all photos and your scoring sheet.
5. *Please note that while the ISS takes into account scoring for the groin and buttocks, you will not be able to score these sites since we do not have test photos of these areas. You will be able to score these sites in person in clinic if necessary.

Body regions for each patient (each site is scored separately):

1. Head and neck:

Scalp, face, neck

2. Trunk:

Groin and chest/abdomen/back

3. Upper extremities:

Elbows, palms, rest of arms

4. Lower Extremities

Knees, soles, rest of legs

eFigure 3. Standard photographs. Standards of the torso, elbows, knees, palms, soles and erythema were provided for each level of severity. Raters scored the test photo set using these standards as guidelines.

TORSO SCALE SEVERITY STANDARDS

(Severity score 0: Normal skin; no perceptible scale, and no loss of normal skin markings)



Severity score 1: Normal skin intermixed with small scales or areas of minimally shiny/waxy/thickened skin and/or partial loss of normal skin markings



Severity score 2: Several areas of scaling upon a background of mildly thickened skin often with some loss of normal skin markings



Severity score 3: Confluent scales with or without focal areas of thick, piled scales and moderately thickened skin



Severity score 4: Extensive areas of confluent, primarily thick, piled scale and severely thickened skin

ELBOW SCALE SEVERITY STANDARDS

(Severity score 0: Normal skin; no perceptible scale, and no loss of normal skin markings)



Severity score 1: Normal skin intermixed with small scales or areas of minimally shiny/waxy/thickened skin and/or partial loss of normal skin markings



Severity score 2: Several areas of scaling upon a background of mildly thickened skin often with some loss of normal skin markings



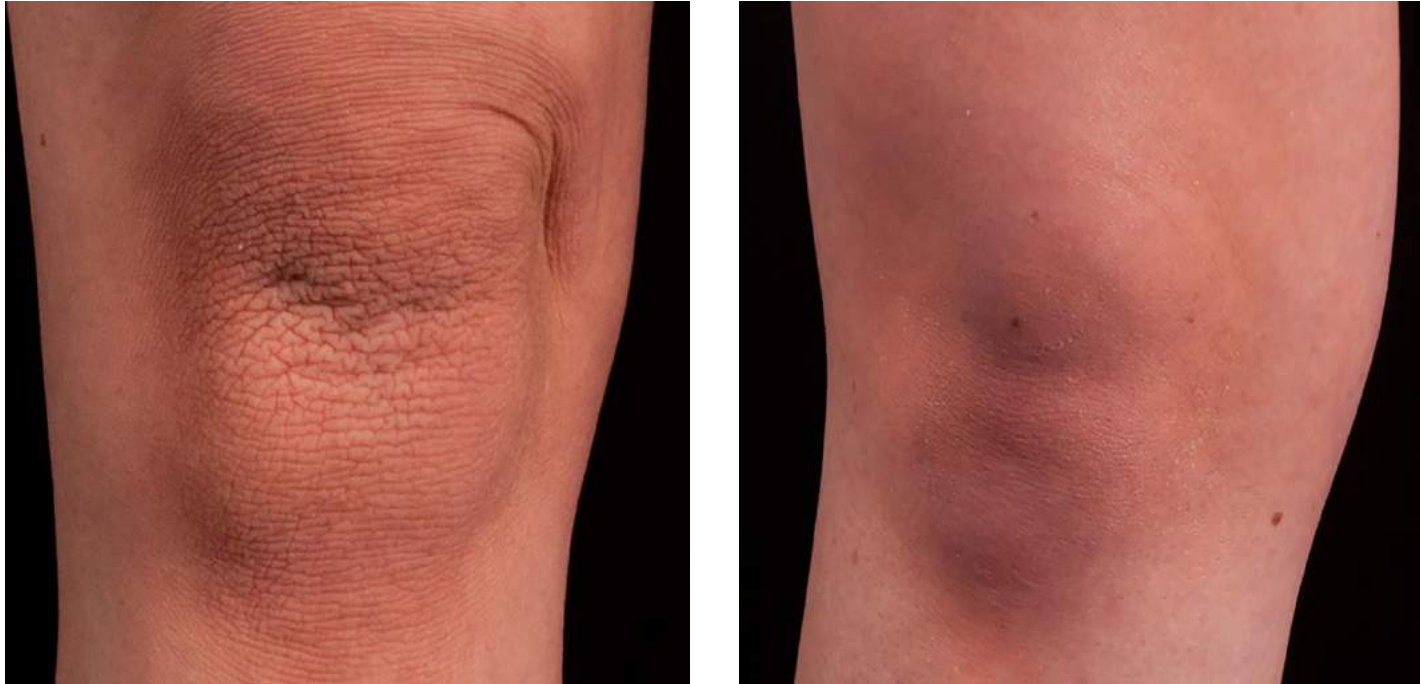
Severity score 3: Confluent scales with or without focal areas of thick, piled scales and moderately thickened skin



Severity score 4: Extensive areas of confluent, primarily thick, piled scale and severely thickened skin

KNEE SCALE SEVERITY STANDARDS

(Severity score 0: Normal skin; no perceptible scale, and no loss of normal skin markings)



Severity score 1: Normal skin intermixed with small scales or areas of minimally shiny/waxy/thickened skin and/or partial loss of normal skin markings



Severity score 2: Several areas of scaling upon a background of mildly thickened skin often with some loss of normal skin markings



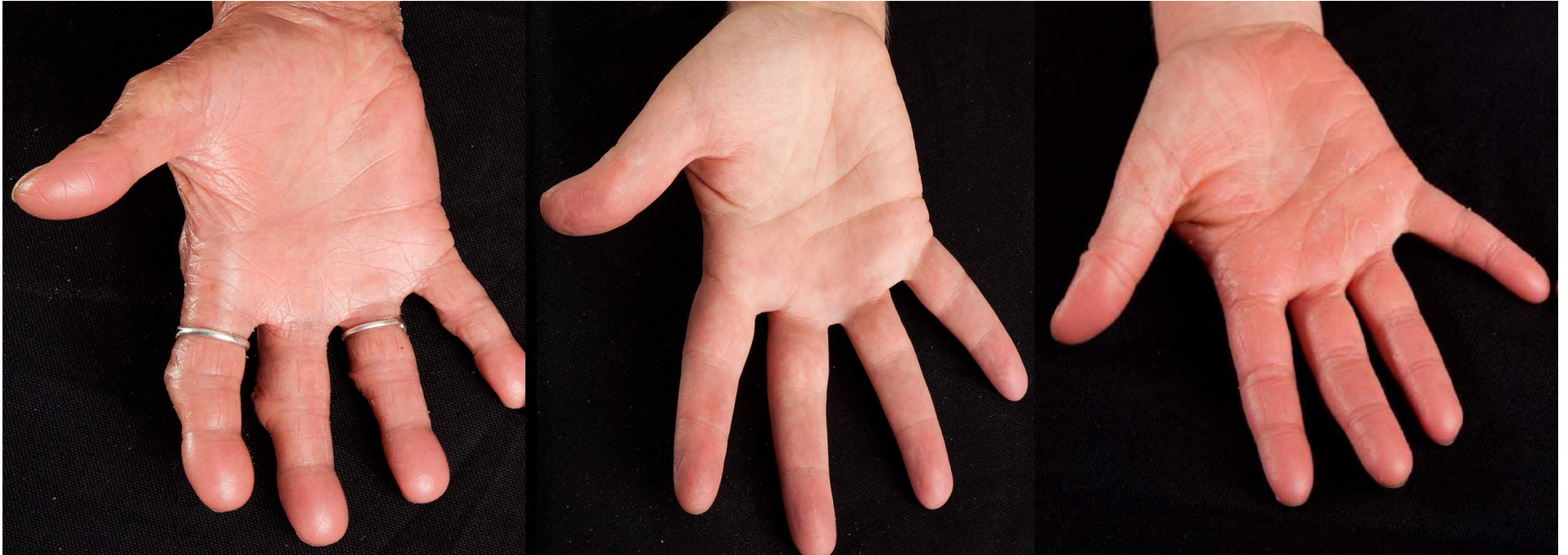
Severity score 3: Confluent scales with or without focal areas of thick, piled scales and moderately thickened skin



Severity score 4: Extensive areas of confluent, primarily thick, piled scale and severely thickened skin

PALM SCALE SEVERITY STANDARDS

(Severity score 0: Normal skin; no perceptible scale or accentuated skin markings)



Severity score 1: Minimal, confluent or focal thickening with continued visibility of normal skin lines. May have minimal desquamative scale.



Severity score 2: Mild, confluent or focal thickening that at least partially obscures skin lines. May have mild desquamative scale, some accentuation of coarse skin lines and/or mild fissuring.



Severity score 3: Moderate, confluent or focal piled or desquamative scale or yellow thickening. Often with accentuated coarse skin lines with or without fissures.



Severity score 4: Thick, confluent or focal yellow piled scale and/or severe desquamative scale with or without fissures.

SOLE SCALE SEVERITY STANDARDS

(Severity score 0: Normal skin; no perceptible scale or accentuated skin markings)



Severity score 1: Minimal, confluent or focal thickening with continued visibility of normal skin lines. May have minimal desquamative scale.



Severity score 2: Several areas of scaling upon a background of mildly thickened skin often with some loss of normal skin markings



Severity score 3: Moderate, confluent or focal piled or desquamative scale or yellow thickening. Often with accentuated coarse skin lines with or without fissures.



Severity score 4: Thick, confluent or focal yellow piled scale and/or severe desquamative scale with or without fissures.

ERYTHEMA SEVERITY STANDARDS
(Severity score 0: No erythema)



Severity score 1:
Mild: Barely perceptible pink



Severity score 2:
Moderate: Pink to red



Severity score 3:
Severe: Bright red



Severity score 4:
Very severe: Deep red-purple

eFigure 4. Descriptors. Descriptions for each level of scale and erythema severity. Palms and soles demonstrate distinctive scale variation and require a separate set of descriptors from other body sites.

Scale descriptors for all sites but the palms and soles	
0	Normal skin; no perceptible scale, and no loss of normal skin markings
1	Normal skin intermixed with small scales or areas of minimally shiny/waxy/thickened skin and/or partial loss of normal skin markings
2	Several areas of scaling upon a background of mildly thickened skin often with some loss of normal skin markings
3	Confluent scales with or without focal areas of thick, piled scales and moderately thickened skin
4	Extensive areas of confluent, primarily thick, piled scale and severely thickened skin

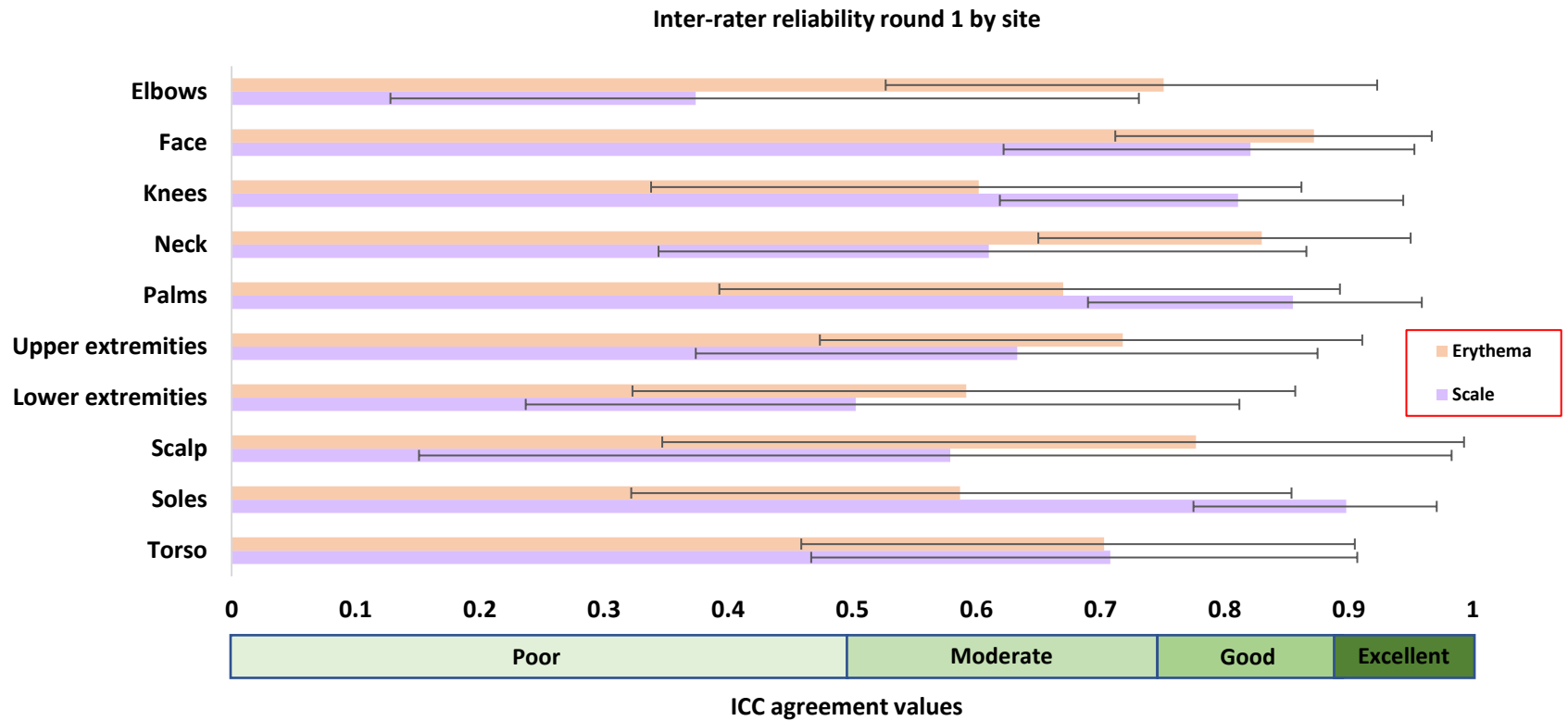
Scale descriptors for the palms and soles	
0	Normal skin; no perceptible scale or accentuated skin markings
1	Minimal, confluent or focal thickening with continued visibility of normal skin lines. May have minimal desquamative scale.
2	Mild, confluent or focal thickening that at least partially obscures skin lines. May have mild desquamative scale, some accentuation of coarse skin lines and/or mild fissuring.
3	Moderate, confluent or focal piled or desquamative scale or yellow thickening. Often with accentuated coarse skin lines with or without fissures.
4	Thick, confluent or focal yellow piled scale and/or severe desquamative scale with or without fissures.

Erythema descriptors	
0	No erythema
1	Barely perceptible pink
2	Pink
3	Red
4	Deep red-purple

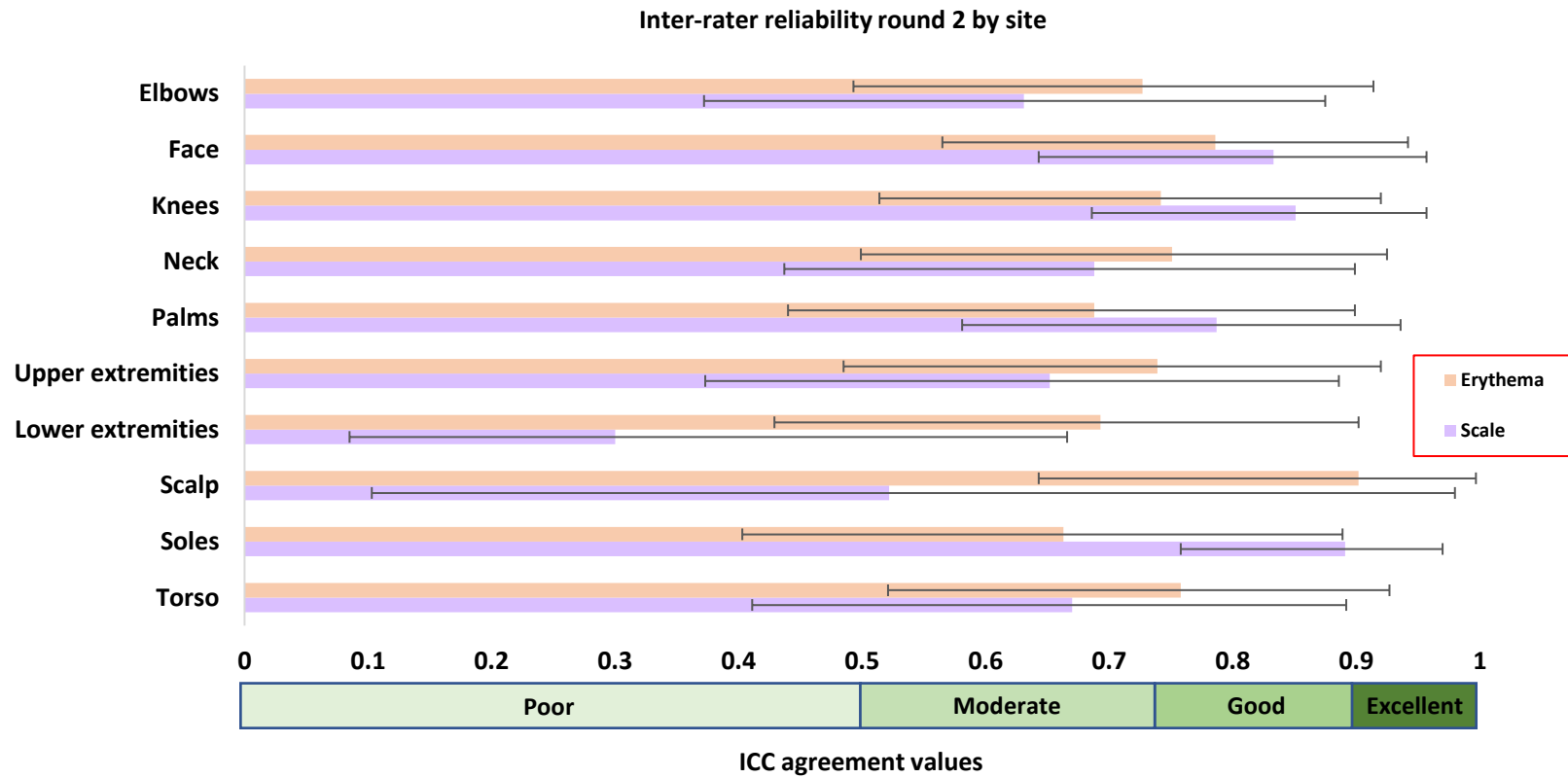
eFigure 5. First cohort: Inter-rater reliabilities for photographic testing.

(a) round 1. All body sites exhibited moderate—good reliabilities for scale except elbows. All body sites exhibited moderate – good reliabilities for erythema. **(b)** round 2. All body sites exhibited moderate—good reliabilities for scale except lower extremities. All body sites exhibited moderate – good reliabilities for erythema.

(a)



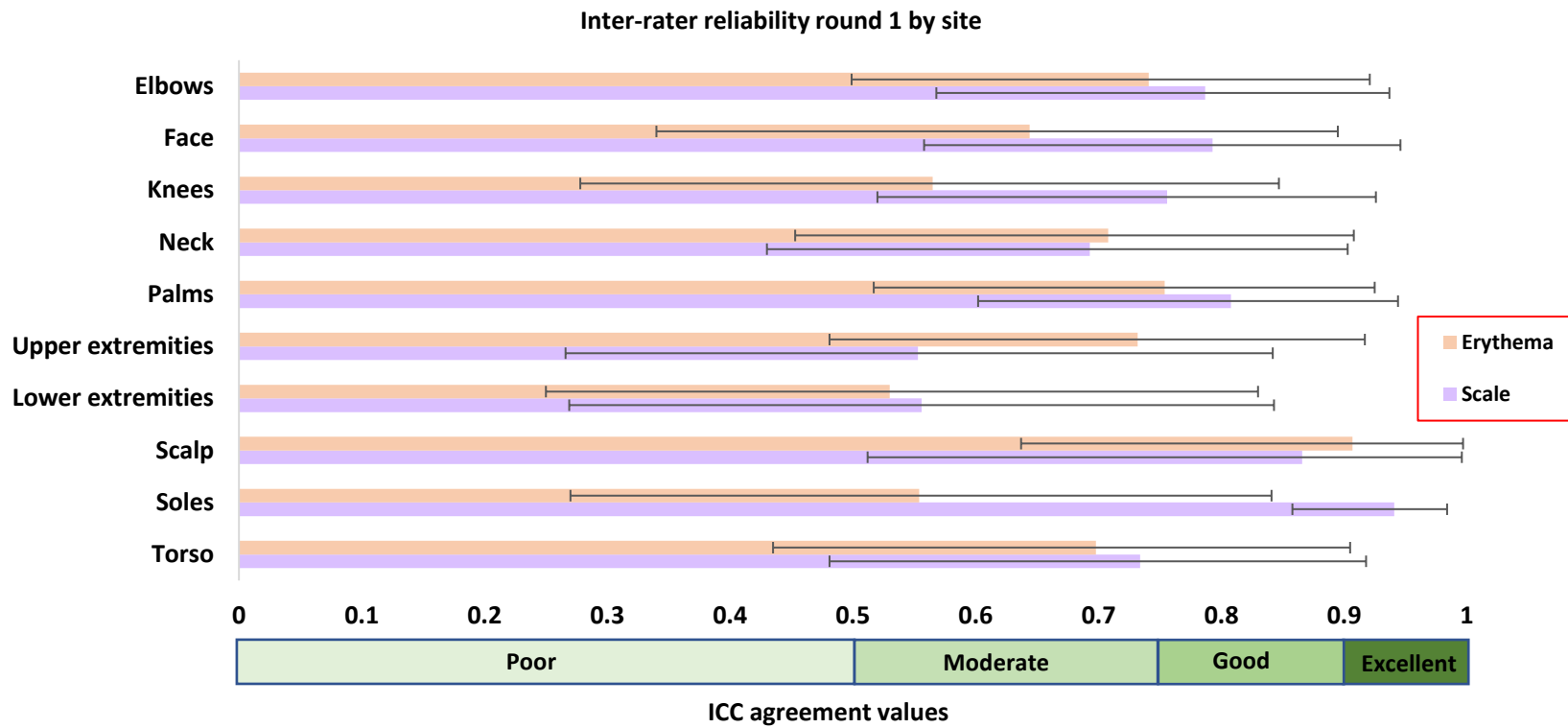
(b)



eFigure 6. Second cohort: Inter-rater reliabilities for photographic testing.

(a) round 1. All body sites exhibited moderate—excellent reliabilities for scale. All body sites exhibited moderate – excellent reliabilities for erythema. (b) round 2. All body sites

(a)



(b)

