

CLASI: Cutaneous Lupus Erythematosus Disease Area and Severity Index

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Slide 1. We are going to talk about how to use the CLASI, the Cutaneous Lupus Erythematosus Disease Area and Severity Index. The purpose of this training video is to familiarize you with the history of the CLASI, the different sections of the CLASI and how to apply each part. You can pause the presentation at any time and return to the video at any time during the test. After each section please press play to advance to the next slide.

Background

- Not enough trials for LE
- FDA Concept Paper: SLE (9-16-2003)
 - Measure of disease activity
 - Measure of disease induced damage
 - Measure of response as determined by the patient, "a patient global response"
 - Measure of health related quality of life

Slide 2. The FDA has offered guidance on how to measure systemic lupus in clinical studies, including separate measures of disease activity, disease damage, patient global response, and health related quality of life.

Consensus

- Need surrogate markers as endpoint for studies
- Single organ studies adequate to receive FDA approval for new drugs

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Slide 3. There is a consensus that there is a need for surrogate markers as endpoints for studies. In addition, there is FDA interest in single organ studies as being adequate to receive FDA approval for new drugs.

Development of the Cutaneous Lupus Area and Severity Score (CLASI)

- Need for a new CLE index
- Distinguish between damage and activity
 - To differentiate
 - To avoid paradoxical stability as activity resolves and damage increases

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Slide 4. There was agreement that there was a need for a new cutaneous lupus index that would distinguish between damage and activity. Separate damage and activity scores would allow differentiation of these scores and avoid the paradoxical stability that might happen as activity resolves and damage increases.

CLASI

- Separation between Activity and Damage
- Activity
 - Erythema
 - Scale/Hypertrophy
 - Acute Hair loss/Non-scarring Alopecia
 - Mucous membrane lesions
- Damage
 - Scarring and Scarring Alopecia
 - Dyspigmentation

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Slide 5. The CLASI separates measures of activity and damage. Activity is measured as degree of erythema, presence of scale/hypertrophy, acute hair loss or nonscarring alopecia, and the presence of mucous membrane lesions. Damage is measured as scarring of skin, scarring alopecia., and dyspigmentation .

Cutaneous LE Disease Area and Severity Index (CLASI)

Select the score in each anatomical location that describes the most severely affected cutaneous lupus-associated lesion

Examiner	activity		damage		Anatomical Location
	Erythema	Scale/ hyperpigmentation	Dyspigmentation	Scarring/ Atrophy/ Panniculitis	
	0-absent 1-pink, faint erythema 2-red 3-dark red, papulovesicular/ crusted hemorrhagic	0-absent 1-scale 2-verruccous hyperplasia	0-absent 1-dyspigmentation	0-absent 1-scarring 2-severely atrophic scarring or panniculitis	
	Scalp			See below	Scalp
	Face				Face
	Nose (not malar area)				Nose (not malar area)
	Rest of the face				Rest of the face
	V-area neck (frontal)				V-area neck (frontal)
	Post. neck &/or shoulders				Post. neck &/or shoulders
	Chest				Chest
	Abdomen				Abdomen
	Back, buttocks				Back, buttocks
	Arms				Arms
	Hands				Hands
	Legs				Legs
	Feet				Feet

Mucous membrane	Dyspigmentation
Mucous membrane lesions (examine if patient confirms involvement)	Report duration of dyspigmentation after active lesions have resolved (visual report to patient - risk appropriate type)
0-absent 1-lesion ulceration	0 Dyspigmentation usually lasts less than 12 months (dyspigmentation weak above normal) 1 Dyspigmentation usually lasts at least 12 months (dyspigmentation weak is bluish)

Alopecia

Recent hair loss within the last 30 days / as reported by patient

0-none
1-few
2-many

Divide the scalp into four quadrants as shown. The dividing line between right and left is the midline. The dividing line between frontal and occipital is the line connecting the highest points of the ear ribs. A quadrant is considered affected if there is a lesion within the quadrant.

Alopecia (primarily not obviously scarring)

0-absent
1-diffuse, non-inflammatory
2-focal or patchy in one quadrant
3-focal or patchy in more than one quadrant

Scarring of the scalp (judged clinically)

0-absent
1-in one quadrant
2-two quadrants
3-three quadrants
4-affects the whole scalp

Total Activity Score
(On the activity score please add up the scores of the left side i.e. for Erythema, Scale/Hyperpigmentation, Mucous membrane involvement and Alopecia)

Total Damage Score
(On the damage score please add up the scores of the right side i.e. for Dyspigmentation, Scarring/Atrophy/Panniculitis and Scarring of the Scalp)

Albrecht and Werth,
JID 125:889, 2005

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Slide 6. This slide shows the CLASI. It is a one-page instrument that measures activity on the left half of the form and damage on the right half. The entire lower portion of the instrument relates to alopecia. At the bottom of the form are two boxes, one on the left to tally the total activity score and the other on the right to report the total damage score.

Select the score in each anatomical location that describes the most severely affected cutaneous lupus-associated lesion

		activity		damage		
E x t e n t	Anatomical Location	Erythema	Scale/ Hypertrophy	Dyspigmentation	Scarring/ Atrophy/ Panniculitis	Anatomical Location
			0-absent 1-pink; faint erythema 2- red; 3-dark red; purple/violaceous/ crusted/ hemorrhagic	0-absent; 1-scale 2-verrucous/ hypertrophic	0-absent, 1-dyspigmentation	0 ... absent 1 ... scarring 2 ... severely atrophic scarring or panniculitis
	Scalp				See below	Scalp
	Ears					Ears
	Nose (incl. malar area)					Nose (incl. malar area)
	Rest of the face					Rest of the face
	V-area neck (frontal)					V-area neck (frontal)
	Post. Neck &/or shoulders					Post. Neck &/or shoulders
	Chest					Chest
	Abdomen					Abdomen
	Back, buttocks					Back, buttocks
	Arms					Arms
	Hands					Hands
	Legs					Legs
	Feet					Feet

Mucous membrane		Dyspigmentation	
Mucous membrane lesions (examine if patient confirms involvement)		Report duration of dyspigmentation after active lesions have resolved (verbal report by patient ... tick appropriate box)	
0-absent; 1-lesion or ulceration		<input type="checkbox"/> Dyspigmentation usually lasts less than 12 months (dyspigmentation score above remains) <input type="checkbox"/> Dyspigmentation usually lasts at least 12 months (dyspigmentation score is doubled)	

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Slide 7. This shows the top half of the CLASI instrument. The columns at the left and right sides detail the anatomical locations to be measured for activity and damage. The left half of the CLASI relates to activity.. The first column rates erythema, which is measured as 0- absent; 1 – pink, light erythema; 2 – red; or 3- dark red, purple/violaceous/crusted/hemorrhagic. The worst degree of erythema in any given body area is the one that is scored.


The next column over measures scale/hypertrophy. It is measured as 0 – absent; 1 – scale; 2- verrucous/hypertrophic. The worst degree of scale/hypertrophy in any given body area is the one that is scored.

On the damage half (right side) of the CLASI measurements of dyspigmentation are either 0 – absent; or 1 – dyspigmentation, which refers to hypopigmentation or hyperpigmentation.

At the right bottom of this top half of the CLASI form there is a box where you must check the length of time the dyspigmentation persists. If it usually lasts less than 12 months, then the top box on the right bottom panel must be checked. If dyspigmentation usually lasts at least 12 months, then the bottom

box on the right second box must be checked. When lesions are typically present less than 12 months, the total dyspigmentation score is the sum of each of the anatomical scores. When dyspigmentation typically last 12 months or longer, then the total dyspigmentation score is doubled.

The last column of the top part of the CLASI instrument is titled scarring/atrophy/panniculitis. This is used to rate the same anatomical locations rated previously for activity and dyspigmentation. If scarring is absent, then that is scored a 0. If there is scarring, with a loss of normal skin surface markings, then scarring is scored a 1. If the scarring is depressed below the skin surface or has lipoatrophy because of resolved panniculitis, then the scarring is scored a 2 for severely atrophic scarring or panniculitis. All scarring related to the scalp is recorded in the scarring alopecia portion of the CLASI at the right bottom of the entire CLASI instrumer, not in the upper half of the CLASI. This allows capturing more detail about scarring alopecia than is needed for the other anatomical areas. The left lower corner of the upper half of the CLASI evaluates mucous membrane lesions. If the patient reports that they have lesions in their mouth, then you should check to make sure they are active and that you attribute them to lupus. Lesions can be hidden, leading to inter-rater variability depending on how carefully the mouth is examined. The mouth is examined only if the patient reports lesions.



Professional Online Instrument Training

Alopecia

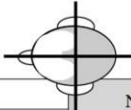
Recent Hair loss (within the last 30 days / as reported by patient)	
1-Yes	
0-No	

Divide the scalp into four quadrants as shown. The dividing line between right and left is the midline. The dividing line between frontal and occipital is the line connecting the highest points of the ear lobe. A quadrant is considered affected if there is a lesion within the quadrant.

Alopecia (clinically not obviously scarred)	Scarring of the scalp (judged clinically)
0-absent 1-diffuse, non-inflammatory 2-focal or patchy in one quadrant; 3-focal or patchy in more than one quadrant	0-absent 3- in one quadrant 4- two quadrants 5- three quadrants 6- affects the whole skull

Total Activity Score
(For the activity score please add up the scores of the left side i.e. for Erythema, Scale/Hypertrophy, Mucous membrane involvement and Alopecia)

Total Damage Score
(For the damage score, please add up the scores of the right side, i.e. for Dyspigmentation, Scarring/Atrophy/Panniculitis and Scarring of the Scalp)



NB: if scarring and non-scarring aspects seem to coexist in one lesion, please score both

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Slide 8. This is the lower half of the CLASI instrument. It is devoted completely to alopecia, with evaluation of activity alopecia on the left side of the form and damage alopecia on the right. The scalp is divided into quadrants, as depicted in the picture at the top. There is a line down the middle of the scalp vertically and another line across the ears, dividing the scalp into quadrants that will be assessed further down in the instrument.

If the patient has more than their usual amount of hair loss in the last 30 days, and attributes this loss to lupus, then “1” for “Yes” is answered in the upper box on recent hair loss. If there is just a normal amount of hair loss, then the recent hair loss is scored a 0. Just below the assessment for recent hair loss is the rating for activity alopecia. If there is no erythema or scale with associated hair loss, then this area is rated 0, for absent. If there is diffuse, non-inflammatory alopecia, as would be seen in a patient with systemic lupus having a flare, that would be a scored “1”. Patients with erythema and scale attributed to cutaneous lupus with associated hair loss would be scored “2” if there is focal or patchy hair loss in one quadrant of the scalp and “3” if there is focal or patchy hair loss in more than one quadrant.

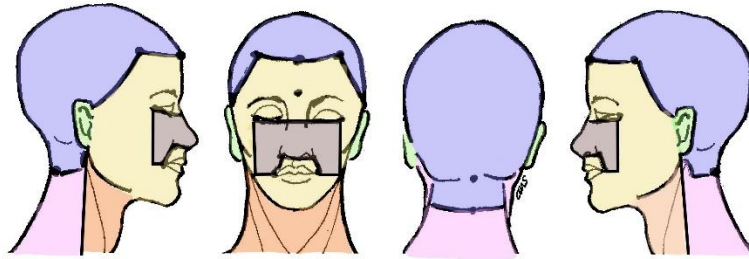
The assessment of damage alopecia is done on the right side of the CLASI form in the section on scarring of the scalp. If there is no scarring, this is scored a 0. If there is a

8

loss of hair follicles and associated loss of hair in lesions located in just one quadrant, that is scored a 3, two quadrant involvement is scored a 4, three quadrant involvement is a 5, and four quadrant involvement is a 6.

It is possible for patients to have both activity and damage alopecia at the same time in the same lesions. A patient may have scarring in a lesion, with loss of hair follicles. If there is superimposed erythema or scale attributed to lupus, that would be considered activity alopecia in addition to the scarring, since it is often hard to tell if the hair loss with both activity and damage lesions superimposed is from activity or damage. If the erythema and scale resolve, and the patient has scarring alopecia only, then the activity alopecia would be scored 0 and the only alopecia score would related to alopecia related to damage.

The total activity score is calculated as the sum of all the scores for erythema and scale/hypertrophy. The total damage score is calculated by adding the scores on the damage side of the CLASI. If dyspigmentation is typically present 12 months or more, then the dyspigmentation score is doubled and added to the scarring scores of the CLASI.

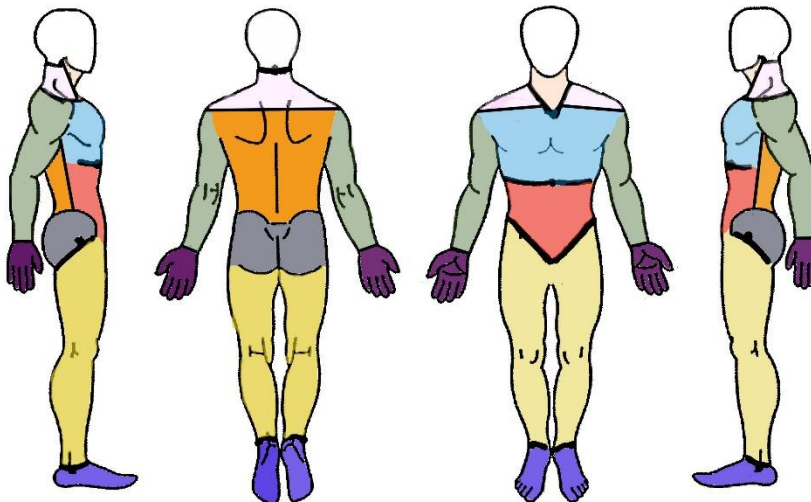


CLASI Anatomic Boundaries

- | | |
|--------------|--------------|
| Scalp | Rest of face |
| Ears | V-area neck |
| Nose (malar) | Post. neck |

CLASI Anatomic Boundaries

- | | |
|------------------------|----------|
| V-area neck (frontal) | Buttocks |
| Post. Neck & shoulders | Arms |
| Chest | Hands |
| Abdomen | Legs |
| Back | Feet |



Scalp: The natural hairline (follicles present) OR if bald, the frontal border begins 8 cm from the midpoint of the glabella and extends 8 cm left and right from that midpoint. For the back of the head, the theoretical hairline would end at 8cm below the external occipital protuberance.

Ears: The entire pinnae and ear canal, including the dorsal surface (posterior portion of the ear).

Nose (including malar area): The entirety of the nose, including from the outer corners of the eyelids straight down until the commissural fold of the lips (outer corners) is reached. From the corners of the lips to the lesser ala of the nose, following the nasolabial fold. The malar area is superiorly bound by the inner eyelids.

Rest of face: Superiorly bound by the hairline/scalp, laterally bound by the hairline and the ears, inferiorly bound by the jawline. Excludes the malar area.

V-area neck (frontal): Superiorly bound by the jawline, laterally bound by the tip of the earlobes. The lateral boundaries converge into a “v-area” that terminates at the 2nd rib or sternal angle. The submental area extending into the neck is included into the V-area neck. Further photo review of lupus patients V-area is needed.

Posterior neck: extends up between the hairline and the ear. If bald, posterior neck begins 8 cm below external occipital protuberance.

Shoulders: laterally bound by the glenohumeral joint. Dorsal inferior boundary is the line drawn between the spines of the scapula. Excludes the frontal neck area. Ventral inferior boundary is from the glenohumeral joint, following the clavicle until the frontal neck area border is met.

Chest: Laterally bound by the mid axillary lines (MAL). Inferior border is from the tip of the xiphoid process extending to the MAL. Excludes the V-area and shoulders.

Abdomen: Laterally bound by the mid axillary lines. Superior border is from the tip of the xiphoid process extending to the MAL. Inferior border is the inguinal folds.

Back: Superiorly bound by the spines of the scapula. Laterally bound by the MAL. Inferiorly bound by the PSIS (posterior superior iliac spine).

Buttocks: Superiorly bound by the PSIS. Anteriorly bound by a line from the anterior superior iliac spine (ASIS) to the greater trochanter. Inferiorly bound by the gluteal folds.

Arms: The border between the shoulders and arms is the glenohumeral joint.

Hands: circumferentially bound by the ulnar and radial protuberances

Legs: superiorly bound by the inguinal folds (ventral) and the gluteal folds (dorsal), laterally bound by the ASIS to the greater trochanter, and inferiorly bound by the medial malleolus.

Feet: circumferentially bound by the inferior edge of the medial malleolus

Groin: Included in the abdomen unless lesions are on mucosal surfaces.

Validation of the CLASI

Interrater Reliability:

Activity: ICC 0.86 (95% CI 0.72 to 0.99)

Damage: ICC 0.92 (95% CI 0.85 to 1.00)

Intrarater Reliability:

Activity: Spearman's rho 0.96 (95% CI 0.89 to 1.00)

Damage: Spearman's rho 0.99 (95% CI 0.97 to 1.00)

Albrecht and Werth, JID 125:889, 2005

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Slide 9. There have been a number of studies done to validate the CLASI. The first looked at inter-rater and intra-rater reliability by having multiple raters evaluate the same patients and also the same rater re-rating the same patient at a later time. Both the inter-rater and intra-rater reliability were in the excellent range.

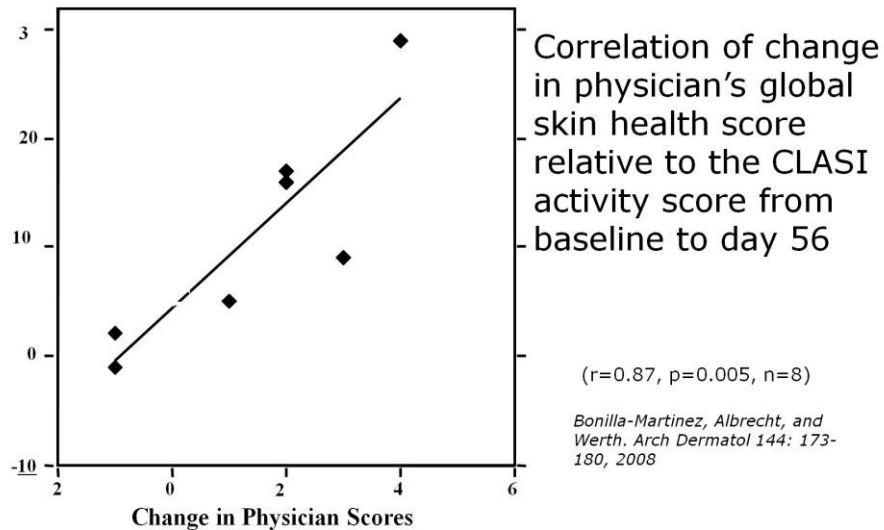
Extension of CLASI Validation to Rheumatology

- Separate study to examine inter-rater and intra-rater validation with rheumatologists
- Excellent correlation in CLASI findings between and within both dermatologists and rheumatologists

Krathen M, Dunham J, Kolasinski SL, Kwan-Morley J, Von Feldt J, and Werth VP, Arthritis Care & Research 59:338-344, 2008

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Slide 10. The CLASI validation was then extended to rheumatology in a separate study that involved a collaboration between rheumatologists and dermatologists. There were excellent correlations in CLASI findings between and within both dermatologists and rheumatologists.



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Slide 11. Another study looked at the responsiveness over time of the CLASI after beginning a new therapeutic intervention. In this study, the correlation of the change in physician's global skin health score relative to the change in CLASI activity score was examined from baseline to day 56, and showed an excellent linear correlation. As the physician global skin score improved, so did the CLASI score.

Clinical response in activity and damage over time (SCLE)



Baseline

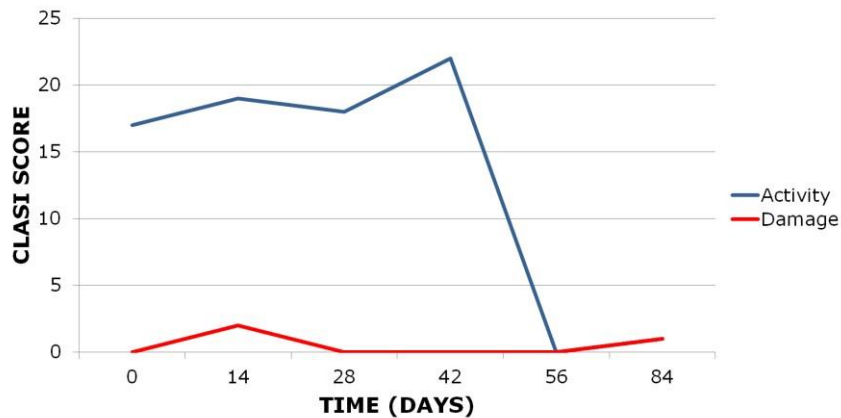


Day 56

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Slide 12. The clinical response in activity and damage was measured at baseline and day 56 in a patient with SCLE who was treated with methotrexate. You can see the skin largely cleared by day 56.

Clinical Response in Activity and Damage Over time in SCLE



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Slide 13. This shows the CLASI score assessments over time for the previous patient, with the activity score in blue and the damage score in red. The activity score decreased to 0 by day 56, after the methotrexate dose was titrated up and started to work. The amount of damage, measured as dyspigmentation or scarring, in SCLE is minimal, and is reflected in the low CLASI damage scores that are close to 0 during the time of the study.

Clinical response in activity and damage over time (DLE)



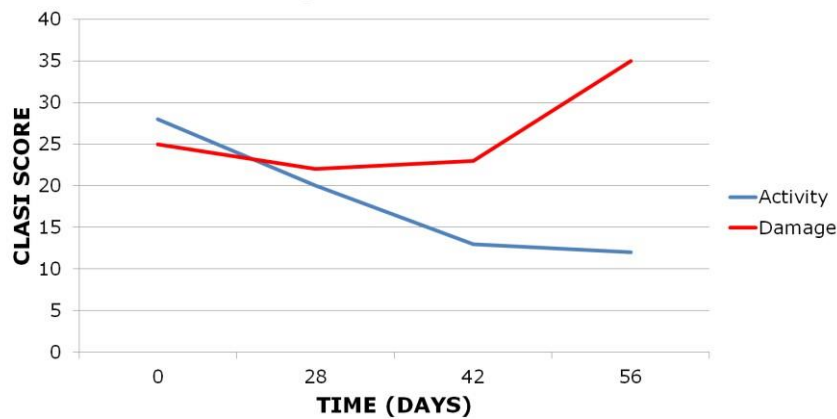
Baseline

Day 56

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Slide 14. This shows another patient from the CLASI responsiveness study. This patient had discoid lupus, and at baseline had activity, manifested by erythema in the skin lesions, but also damage manifested by dyspigmentation. By day 56 on antimalarials, the patient had less erythema but more dyspigmentation. This demonstrates that while erythema improves as the activity decreases, the dyspigmentation may actually worsen.

Clinical Response in Activity and Damage Overtime in DLE



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Slide 15. This graph is of the previous discoid lupus patient, showing that the activity, shown in blue, decreases over time, but the damage, shown in red, actually increases. Thus if you added the activity and damage scores together, you would actually have a paradoxical stability of the combined score at day 0 and day 56. This demonstrates the importance of having the activity and damage scores recorded separately, so you can demonstrate that the activity is actually improving in this patient.

Mycophenolate Mofetil Trial

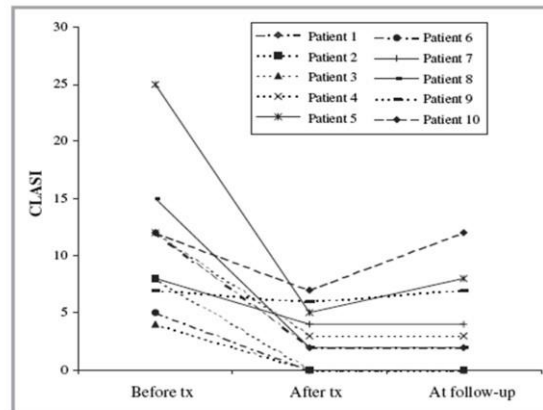


Kreuter et al. *Br J Dermatol* 156:1321, 2007

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Slide 16. This is a picture of a patient before, on the left, and after treatment, on the right, with mycophenolatemofetil. This study was an open label study of 10 patients with SCLC treated with the mycophenolatemofetil.

Mycophenolate Mofetil Trial



Kreuter et al. *Br J Dermatol*
156:1321, 2007

Fig 3. Course of Cutaneous Lupus Erythematosus Disease Area and Severity Index (CLASI) in patients with subacute cutaneous lupus erythematosus treated with mycophenolate sodium. tx, treatment.

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Slide 17. This figure shows the response of the CLASI before and after treatment with mycophenolatemofetil in these 10 patients with SCLE, demonstrating that many, but not all of the patients experienced improvement in their skin activity after beginning the medication.

Lessons: Surface area

- Surface area assessment does not work:
 - In general (*Tilling-Grosse et Ress, BJD 1993; 128:69*)
 - For atopic eczema (*Charman et al, ArchDerm 2000;136:763, Charman et al, BJD 1999;140:109*)
 - For psoriasis (*Ramsay et al, BJD 1991;124:565*)
- Lesion counting is not necessarily better:
 - For acne (*Lucky et al, JAAD 1996;35:559*)

Slide 18. In developing the CLASI, there were several concepts considered prior to determining the system of rating the worst area in a body surface. First, surface area assessment often does not work well, as demonstrated in studies of eczema and psoriasis. Diseases like cutaneous lupus that may affect only a small surface area make it even more difficult to determine the actual body surface area involved and to demonstrate a change in the context of treatment. Lesion counting is not necessarily better, since as a lesion heals it may actually break up into several smaller lesions that would result in a higher lesion count.

Surface area

- Ends up with minimal percentages (head incl scalp 9% BSA)
- What % would the lesion on the cheek be?



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Slide 19. To demonstrate this concept, this picture shows a patient with involvement of around 2% of her skin with discoid lupus lesions. It would be difficult to determine the actual % of the lesion on the cheek in a reproducible way and even harder to determine improvement based on the % area involve.

Surface area is conceptually impossible



Slide 20. This photograph also demonstrates the difficulty in assessing surface area. This slide shows a patient with subacute cutaneous lupus. There is clearing in the center of the lesion, and it would again be very difficult to accurately assess the surface area involved here. Where does the lesion begin and end?

Surface Area and Score:

Distribution of the Unweighted CLASI Score

Area	CLASI (activity and damage)	PASI	Rule of 9
Head	33%	10%	9%
Breast	14%	5%	9%
Abdomen	7%	5%	9%
Back	14%	10%	18%
Legs	14%	40%	36%
Arms/hands	14%	30%	18%
Mucous membrane	1%		

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Slide 21. This table shows the surface area and distribution of the unweighted CLASI score. It shows that with the CLASI there is more weighting for the head, an area more frequently involved in cutaneous lupus, than is seen with other measures, such as the PASI and the Rule of 9. This increases the sensitivity to detect changes affecting an area that is frequently involved in cutaneous lupus.

Surface area and CLASI score

- More weight given to the head
- Less to trunk and extremities
- Reflects the distribution and impact of CLE

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Slide 22. Thus, the CLASI gives more weight to the head, less to the trunk and extremities. It reflects the distribution and impact of cutaneous lupus.

Erythema

Most relevant aspect of the score:

- Very reactive
- Very prominent
- High total score

- Subjective assessment, but has been shown to correspond to doppler flow meter, reflectance colorimetry, video analysis

(Lahti et al. Arch Derm Res 1993;285:278, Quinn ActaDermVenereol 1993;73:21, Ormerod et al. JAAD 1997;37:51)

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Slide 23. We will now discuss Erythema, which is the most relevant aspect of the score and is very reactive, prominent, and gives a high total score. Although a subjective assessment, it has been shown to correspond to measurement with Doppler flow, reflectance colorimetry, and video analysis.

Erythema

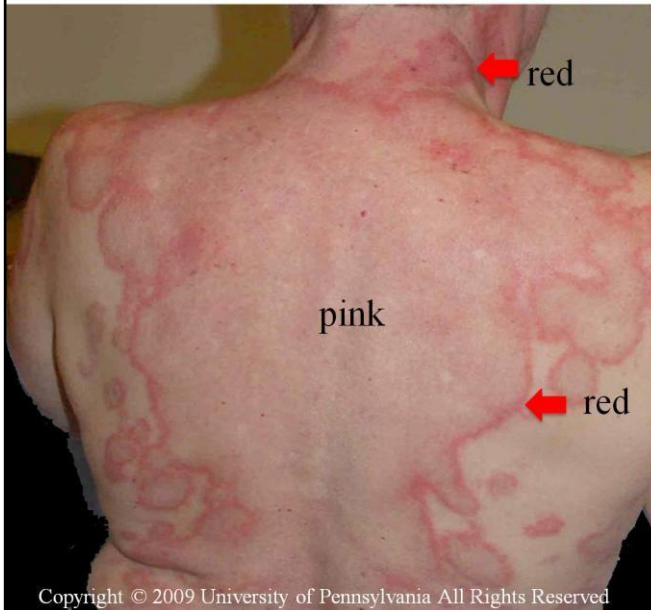


Erythema

Erythema resolved

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Slide 24. This shows erythema on the left, captured with the CLASI, that is resolved after therapy, on the right.



Scoring Erythema

Rate worst area, which is red

Slide 25. This shows erythema that is pink in some areas in the center, but red at the border. With the CLASI the worst area in an anatomic area is rated, so in this particular patient the worst area on the posterior neck &/or shoulders and the back is red, rated as a 3 for erythema.



Slide 26. This slide shows scale and hypertrophic lesions. Since the worst areas are hypertrophic, this would be rated as a "2" under scale/hypertrophy.

Dyspigmentation

- Prominent and disfiguring symptom
- Frequently transient
- Rated in reference to the last 12 months



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Slide 27. We will now discuss dyspigmentation; dyspigmentation is a prominent and disfiguring symptom. It can be transient and it is rated in reference to the how long it is present. When lesions are typically present less than 12 months, then the total dyspigmentation score is the sum of each of the anatomical scores. When dyspigmentation typically lasts 12 months or longer, then the total dyspigmentation score is doubled

Dyspigmentation



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Slide 28. This slide shows hyperpigmentation at the borders of the lesion of hypertrophic DLE.

Subtle Dyspigmentation

- Bridge of the nose LE associated dyspigmentation
- Chin questionable
- What is normal in patients with this type of skin color?



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Slide 29. This slide shows dyspigmentation over the bridge of the nose. The patient reports having a butterfly rash there previously attributed to lupus, then resolved, leaving the hyperpigmentation. The chin is hyperpigmented, but this could be a normal variant for this skin type. It would be helpful to know if the patient had a lesion on her chin previously attributed to lupus that then resolved, leaving the hyperpigmentation.

Alopecia

- Conceptually most difficult symptom
- More than one type of alopecia can be present
- Differential diagnosis can be challenging
 - i.e. traction alopecia or female pattern baldness

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Slide 30. We will now discuss Alopecia. Alopecia is conceptually the most difficult symptom. More than one type of alopecia can be present. The differential diagnosis can be challenging, since there are many causes of alopecia, including traction alopecia or female pattern baldness.

Alopecia

- Erythema in scalp, associated with activity alopecia (area above rectangle)
- Localized scarring alopecia in lower portion of rectangle
- Shows both activity and damage in the same area



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Slide 31. This slide shows erythema in the scalp associated with activity alopecia in the area above the rectangle, as well as in the upper half of the rectangle. There is localized scarring with loss of hair follicles and hair in the lower portion of the rectangle, representing an element of damage alopecia. This shows activity and damage alopecia in the same quadrant of the scalp, and both should be rated. If there is just one quadrant involved, then the score would be 2 for activity alopecia on the left bottom of the alopecia assessment and a 3 for scarring alopecia on the right bottom of the alopecia assessment.

Alopecia

Traction?



Slide 32. This is the scalp of a patient with flaring SLE. She reports more thinning of her hair in the past month, which she attributes to her SLE flare. She also has an element of traction alopecia due to tight pulling of her braids. However, since it was judged that a portion of the hair loss was due to her lupus, but there is no erythema or scale and the loss is diffuse, this was given a score of "1" for diffuse noninflammatory alopecia.

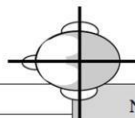
Alopecia Classification

- Alopecia related to activity
 - Diffuse alopecia related to acute disease
 - Patchy non-scarring alopecia related to lesions
- Alopecia related to damage
 - Scarring alopecia

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Slide 33. In summary alopecia related to activity is either diffuse alopecia related to acute disease or patchy non-scarring alopecia related to cutaneous lupus lesions. Alopecia related to damage is scarring alopecia.

Alopecia



Recent Hair loss (within the last 30 days / as reported by patient)		NB: if scarring and non-scarring aspects seem to coexist in one lesion, please score both	
1-Yes			
0-No			
Divide the scalp into four quadrants as shown. The dividing line between right and left is the midline. The dividing line between frontal and occipital is the line connecting the highest points of the ear lobe. A quadrant is considered affected if there is a lesion within the quadrant.			
Alopecia (clinically not obviously scarred)		Scarring of the scalp (judged clinically)	
0-absent		0-absent	
1-diffuse, non-inflammatory		3-in one quadrant	
2-focal or patchy in one quadrant;		4-two quadrants	
3-focal or patchy in more than one quadrant		5-three quadrants	
		6-affects the whole skull	

Total Activity Score

(For the activity score please add up the scores of the left side i.e. for Erythema, Scale/Hypertrophy, Mucous membrane involvement and Alopecia)

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Total Damage Score

(For the damage score, please add up the scores of the right side, i.e. for Dyspigmentation, Scarring/Atrophy/Panniculitis and Scarring of the Scalp)

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Slide 34. To summarize, the activity alopecia score is written on the left side and the damage alopecia score is on the right side. The maximum activity alopecia score is 3, if more than one quadrant has lesions of erythema, scale, and hair loss. The maximum damage alopecia is 6, if there are lesions of scarring in all quadrants in the scalp, leading to loss of hair follicles and the hair. It is possible to have erythema and scale leading to hair loss that counts as activity alopecia in the same area as there are damage alopecia lesions.

Scarring

- Quintessential damage of the skin
- Scarring rated 2 if severely atrophic or associated with panniculitis
- Scarring of the scalp causes alopecia

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Slide 35. We will now discuss scarring; scarring represents quintessential damage of the skin. Scarring is rated in the worst area in a geographic location. It is rated 1 if there is a loss of skin markings but not depression and a 2 if scarring is depressed (severely atrophic) or associated with a resolved panniculitis, leading to lipoatrophy. Scarring of the scalp can result in permanent alopecia.

Mucous membrane lesions

- Sign of clinical activity
- Can be painful or painless
- May be hidden
- To achieve good inter-rater reliability the score was based on the patient's response followed by examination

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Slide 36. Mucous membrane lesions are a sign of clinical activity. They can be painful or painless and may be hidden. To achieve good inter-rater reliability, the score was based on the patient's response that they had oral lesions, followed by an examination to confirm that there are lesions attributable to lupus.

Mucous membrane



Slide 37. This slide shows a patient who reported she had oral lesions. Subsequent examination of her mouth showed the presence of active oral lesions that were attributed to lupus.

**Attribution to CLE is Critical: Differential
Diagnosis: Acne, Rosacea, CLE**

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Slide 38. When using the CLASI it is very important to make sure that skin lesions are attributed to SLE and not other factors. In this example, the patient has acne and thus the CLASI should not be used in rating these lesions.

**Attribution to CLE is Critical:
Differential Diagnosis: Rosacea, CLE**



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Slide 39. In this example, this patient has malar erythema due to acne rosacea, with follicular erythematous papules. Her lesions should not be rated with the CLASI.

Attribution to CLE is Critical: Differential Diagnosis: Tinea, CLE



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Slide 40. In this slide, the patient has a fungal infection due to tinea on her chin, with a scaly elevated border. KOH analysis of the scale was positive and showed fungal hyphae. The CLASI should not be used to assess this patient.

Select the score in each anatomical location that describes the most severely affected cutaneous lupus-associated lesion

Extent	activity			damage	
	Anatomical Location	Erythema	Scale/ Hypertrophy	Dyspigmentation	Scarring/ Atrophy/ Panniculitis
		0-absent 1-pink; faint erythema 2- red; 3-dark red; purple/violaceous/ crusted/ hemorrhagic	0-absent; 1-scale 2-verrucous/ hypertrophic	0-absent, 1-dyspigmentation	0 ... absent 1 ... scarring 2 ... severely atrophic scarring or panniculitis
	Scalp				See below
	Ears				
	Nose (incl. malar area)				Nose (incl. malar area)
	Rest of the face				Rest of the face
	V-area neck (frontal)				V-area neck (frontal)
	Post. Neck &/or shoulders				Post. Neck &/or shoulders
	Chest				Chest
	Abdomen				Abdomen
	Back, buttocks				Back, buttocks
	Arms				Arms
	Hands				Hands
	Legs				Legs
	Feet				Feet

Mucous membrane		Dyspigmentation
Mucous membrane lesions (examine if patient confirms involvement)		Report duration of dyspigmentation after active lesions have resolved (verbal report by patient ... tick appropriate box)
0-absent; 1-lesion or ulceration		<input type="checkbox"/> Dyspigmentation usually lasts less than 12 months (dyspigmentation score above remains) <input type="checkbox"/> Dyspigmentation usually lasts at least 12 months (dyspigmentation score is doubled)

Slide 41. We will now practice using the upper portion of the CLASI to evaluate activity and damage in the next few examples.

**Anatomical location:**

- Arms, legs and back
- Not v of neck,
- Arms, legs and back
- Not v of neck, chest, or abdomen

Erythema:

- Neither dark or purple, nor pink or faint
- Red (2)
- Equally strong on the arms and legs
- The deepest red on the buttocks is as red as on the arms

Hypertrophy, Dyspigmentation, Scarring:

- No evidence (0)

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Slide 42. This is a patient with SLE on the arms, legs and back, but with no involvement of the v of the neck, chest, or abdomen. There is red erythema on the arms, legs, and buttocks that is equally red in the worst areas in all three involved anatomic areas. There is no hypertrophy, dyspigmentation, or scarring.

**DLE****Anatomical location:**

Nose

Erythema:

Red (2)

Scale/Hypertrophy:

Scale (1)

Dyspigmentation:

Yes (1)

Scarring/Atrophy:

yes (1)

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Slide 43. This is a discoid lupus lesion on the nose. There is red erythema in several areas on the right side of this lesion. Thus the erythema is rated as red (2), even though many areas of the lesion are pink. The areas of scale are not in the areas that are red. There is scale, but not hypertrophy, so this is rated a 1. There is scarring with loss of surface markings, but no depression of the scar, so scarring is rated as 1.



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SCLE

Anatomical location:

Posterior neck &
shoulders, back

Erythema:

Red (2)

Scale/Hypertrophy

None (0)

**Dyspigmentation,
Scarring:**

Absent (0)

Slide 44. This is subacute cutaneous lupus on the posterior neck & shoulders, as well as the back. The worst erythema in both anatomic areas is red, so erythema is rated a 2. The contrast between red on the periphery and pink in the center of the lesion is helpful in training the eye to rate pink or red in other lesions of lupus. There is no scale/hypertrophy, dyspigmentation, or scarring.



DLE

Anatomical location:

Rest of Face and Malar Area
including nose

Erythema:

Red (2)

Scale/Hypertrophy:

Absent (1)

Dyspigmentation:

Present (1)

Scarring:

Atrophic scarring (2)

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Slide 45. This is a case of DLE involving the nose/malar area and the rest of face. The rest of face is determined by a line that extends from the lateral eyebrow down and involves areas lateral to the cheek, the forehead, under the nose, and the chin. There is red erythema in both areas, rated as a 2. The worst areas show scale, thus rate 1, in both the nose/malar area and the rest of face. There is dyspigmentation in both areas. There are also depressed scars on the chin and rest of face that would be rated as a 2 for severely atrophic scarring.



DLE

Anatomical location:

Rest of Face, Ears and Post. Neck

Erythema:

Pink (1)

Scale/Hypertrophy:

Absent (0) in neck, ear
Hypertrophic (2) rest of face

Dyspigmentation

Present (1)

Scarring(1)

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Slide 46. This is a case of DLE involving the rest of face, ears, and posterior neck. There pink erythema (rated 1) in all areas. There is one hypertrophic lesion anterior to the ear, located on the rest of face. The ears and posterior neck have no scale and are nonhypertrophic, so are rated 0 for scale/hypertrophy. There is dyspigmentation present (rated 1), and scarring with loss of skin markings, but no depression (rated 1).



Anatomical location:	Arms
Erythema:	Pink (1)
Scale/Hypertrophy:	Hypertrophic (2)
Dyspigmentation:	Present (1)
Scarring:	Present (1)

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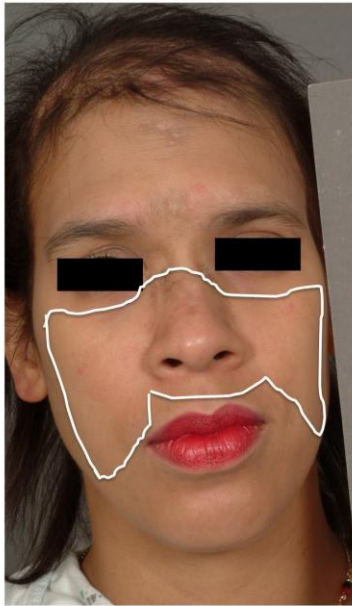
Slide 47. This shows lesions of hypertrophic lupus on the arm. There is pink erythema and scale/hypertrophy (rated 2), dyspigmentation (rated 1), and scarring that is not depressed (rated 1).



Anatomical location:	Malar area (including nose), Rest of Face
Erythema (nose):	Red (2)
Erythema (rest of face):	Pink (1)
Scale/Hypertrophy:	Malar area: Absent (0)
Rest of Face:	Scale (1)
Dyspigmentation:	Absent (0)
Scarring:	Absent (0)

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Slide 48. There are lesions on both the nose/malar area and rest of face. The worst areas on the nose/malar area are red (rated 2, red arrow), while on the rest of face they are pink (rated 1, pink arrow). There is no scale in the nose/malar area, but the pink arrow points to a lesion with scale. There is no dyspigmentation or scarring.



DLE/SLE

Anatomical location:

Nose/malararea and rest of face

Erythema:

Patches of fainterythema (1)

Scale/Hypertrophy:

Absent (0)

Dyspigmentation:

Present (1)

Scarring:

Present (1)

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Slide 49. This is a case of DLE showing involvement to the nose/malar area and rest of face. There is pink erythema (rated 1) in both areas. There is no scale/hypertrophy (rated 0). There is dyspigmentation (rated 1). There is scarring in both areas that is not depressed (rated 1). The malar area is defined as a vertical line from the area below the lateral-most area of the eyebrows to the nasolabial fold, as depicted in the photo.



DLE

Anatomical location:

Arm

Erythema:

Absent (0)

Scale/Hypertrophy:

Absent (0)

Dyspigmentation:

Present (1)

Scarring:

Present (1)

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Slide 50. This is a discoid lupus on the arm that shows no erythema (rated 0), no scale/hypertrophy (rated 0), dyspigmentation (rated 1), and scarring with loss of skin markings but no depression (rated 1).



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DLE

Anatomical location:

Scalp

Erythema:

Absent (0)

Scale/Hypertrophy:

Absent (0)

Dyspigmentation:

Present (1)

Alopecia:

- No recent hair loss (0)
- Non scarring alopecia in more than one quadrant (0)
- Scarring alopecia
- of the whole scalp (6)

Slide 51. This is a picture of DLE involving the scalp with no erythema (rated 0), no scale/hypertrophy (rated 0), dyspigmentation (rated 1), and damage alopecia. The patient reports no increased hair loss in the last month (0), there is no activity alopecia (rated 0), and there is scarring alopecia of all 4 quadrants in the scalp (rated 6).

SCLE**Anatomical location:**

Scalp

Erythema:

Pink (1)

Scale/Hypertrophy:

Absent (0)

Dyspigmentation:

Absent (0)

Scarring:

Not applicable

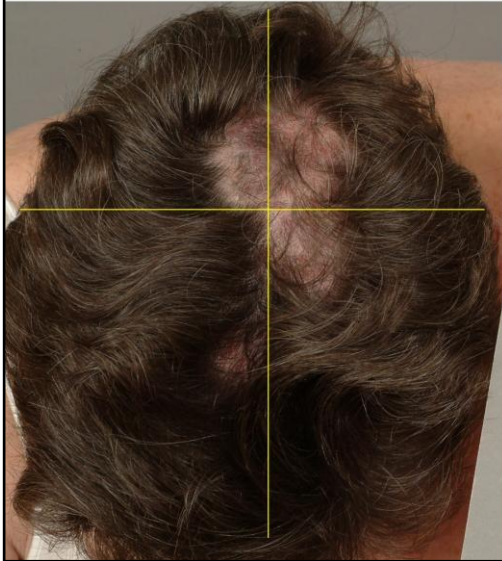
Alopecia:

Recent hair loss: ?

Patchy nonscarring alopecia in one quadrant (2)

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Slide 52. This shows SCLE in the scalp. There is pink erythema (rated 1), no scale/hypertrophy (rated 0), no dyspigmentation (rated 0), activity alopecia with hair loss associated with the pink erythema in one quadrant (rated 2), and no damage alopecia (no scarring associated with permanent loss of hair follicles leading).



SCLE

Anatomical location:

Scalp

Erythema:

Pink (1)

Scale/Hypertrophy:

Absent (0)

Dyspigmentation

Absent (0)

Current hairloss: ?**Alopecia:**

- Patchy alopecia in more than one quadrant (3)
- Scarring of the scalp (0)

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Slide 53. This shows SCLE in the scalp. There is pink erythema (rated 1) no scale/hypertrophy (rated 0), no dyspigmentation (rated 0), patchy activity alopecia in more than one quadrant (rated 3), and no scarring alopecia (rated 0).



SLE/DLE

Anatomical location:

Nose/malar area, rest of face

Erythema:

Red (2)

Scale/Hypertrophy:

Absent (0)

Dyspigmentation

Present (1)

Scarring:

Present (0)

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Slide 54. This shows SLE/DLE on the nose/malar area and rest of face. There is red erythema (rated 2), no scale/hypertrophy (rated 0), dyspigmentation (rated 1), and no scarring (rated 0).



DLE

Anatomical location:

Back

Erythema:

Pink, faint erythema (1)

Scale/Hypertrophy:

Absent (0)

Dyspigmentation

Present (1)

Scarring:

Present (1)

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Slide 55. This is DLE on the back. There is pink erythema in the worst areas (rated 1), no scale/hypertrophy (rated 0), dyspigmentation (rated 1), and scarring with loss of skin surface markings (rated 1).

**DLE/SLE****Anatomical location:**

Scalp

Erythema:

Pink (1)

Scale/Hypertrophy:

Absent (0)

Dyspigmentation

Absent (0)

Current hair loss: ?**Alopecia:**

- Patchy alopecia in more than one quadrant (3)
- Scarring of the scalp:
- The whole skull (6): see arrows showing absence of hair follicles

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Slide 56. This is DLE on the scalp. There is pink erythema (rated 1), no scale/hypertrophy (rated 0), no dyspigmentation (rated 0), patchy alopecia in more than one quadrant (rated 3), and scarring of all 4 quadrants (see arrows), with loss of hair follicles (rated 6).



SCLE

Anatomical location:

Malararea (incl. Nose), rest of face

Erythema:

Red (2)

Scale/Hypertrophy:

Absent (0)

Dyspigmentation

Absent (0)

Scarring:

Not applicable

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Slide 57. This is SCLE on the face in the nose/malar area and rest of face. The worst area in both the nose/malar area and rest of face is red. The two areas labeled on the left cheek are in the nose/malar area and are red. The lesion on the left rest of face is pink, but the lesion on the upper eyelid is considered rest of face and is red. Since the worst area on rest of face is red, the erythema for rest of face should be scored a 2. There is no sale/hypertrophy (scored 0), no dyspigmentation (scored 0), and no scarring (scored 0).



SLE

Anatomical location:

Malar area (incl. Nose),
Rest of Face

Erythema:

Hemorrhagic/ crusted(3)

Scale/Hypertrophy:

Absent (0)

Dyspigmentation:

Absent (0)

Scarring:

Absent (0)

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Slide 58. This is acute lupus erythematosus on the face, with involvement of the nose/malar area and rest of face. There is hemorrhagic/crusted lesions (rated 3), no scale/hypertrophy (rated 0), no dyspigmentation (rated 0), and no scarring (rated 0). This represents I activity, but not damage, due to cutaneous lupus.

Summary

- CLASI is a validated score for evaluating lupus-specific skin lesions
 - Inter-rater and intra-rater reliability for use by dermatologists and rheumatologists
 - Demonstrates responsiveness in the context of treatment
 - Used in a number of prospective studies

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Slide 59. In summary, the CLASI is a validated score for evaluating lupus-specific skin lesions. It has been shown to have excellent inter-rater and intrarater reliability for use by dermatologists and rheumatologists and demonstrates responsiveness in the context of treatment. The CLASI has been used in a number of prospective studies.

Remember that you can refer back to any part of the video at any time during the test. You may keep the video open during the test and do not need to close it to resume the test.