

Round 3 - Vulvar Lichen Sclerosus Severity Scale

Welcome to the Adult Vulvar Lichen Sclerosus Severity Scale Survey

Thank you so much for continuing to participate in this survey. By this point, you should have received an email with both your individual responses, as well as a summary of the responses provided by your colleagues so far. Please take a look at this data before you proceed in this survey.

As already mentioned in the email you were sent, we have made some changes in this survey.

The things that we have changed are as follows:

1. For remaining items which have not yet reached consensus (see below), we have provided 2 options: "Include" or "Exclude" from the severity scale with the intention of ensuring that we come to a consensus.
2. In terms of measuring symptoms, 72% of you have agreed that a 5-point severity scale should be used for this purpose. Given that this was by far the most popular option, we have accepted this consensus and now ask whether frequency of symptoms should be measured as well.
3. In terms of measuring symptoms for which validated scales exist (e.g. quality of life and changes to sexual function), we have selected the top 3 options and provided only those to you with the intention of ensuring that we come to a consensus.
4. In terms of the timeline over which to measure symptoms, we have selected the top 3 options and provided only those to you with the intention of ensuring
5. In terms of measuring signs, we have selected the top 3 options and provided only those to you with the intention of ensuring that we come to a consensus.

The things that we have added are as follows:

1. We have added "hyperpigmentation" in the signs category
2. For some items that have reached consensus (see below) including: excoriations, erosions, ulcers, sclerosis, and lichenification, there has been concern that we may not be united on the definitions of these items. Therefore, we have asked you again about each of these items (whether to include or exclude) along with an accepted definition of that term.
3. "Narrowing of the introitus" has reached consensus but we have an additional question asking whether this includes both narrowing and formation of posterior commissure bands/fourchette webs.
4. We have added the item "posterior commissure bands" in the architectural changes category

Addressing some of the comments we received:

1. For the question regarding the timeline over which to measure symptoms (e.g. in the past month/3 months have you experienced itch?) there were many comments suggested that we ask about symptoms from the onset or ask over various time periods. From a research perspective, when we test the validity of this scale in the future, we will need a set timeline that we can use for all patients to directly compare. Of course clinically, understanding what the symptoms were like from the onset may be important but for research purposes we need to define a particular time

period.

2. Participants have asked that there be separate research criteria and clinical criteria. We were hoping for feedback from other participants as to this point and ask that you comment at the end in the comments section.

The following are the items that have yet to reach consensus:

1. Symptoms

- a. Irritation
- b. Dryness
- c. Difficulty urinating (dribbling, having to stand up to void)
- d. Difficulty with bowel movements (constipation)
- e. Bleeding with scratching
- f. I don't have any symptoms
- g. Mood changes (depression/anxiety)

2. Signs

- a. Telangiectasia
- b. Petechiae
- c. Ecchymosis
- d. Colour (normal vs. abnormal)
- e. Pallor
- f. Hypopigmentation
- g. Erythema
- h. Induration
- i. Edema
- j. Atrophy
- k. Whether disease is symmetrical or asymmetrical

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The items that have reached consensus and will be included in the final scale have been removed for the next round.

Items for which >75% consensus was reached (>75% assessed the time as being very important or essential towards determining disease severity):

1. Symptoms

- a. Itch
- b. Pain unrelated to intercourse (burning, soreness, discomfort, etc.) at rest or during activity
- c. Pain with intercourse
- d. Bleeding and pain with intercourse
- e. Skin tearing with intercourse
- f. Quality of life
- g. Changes/decrease in sexual function

2. Signs

- a. Fissures
- b. Whitening
- c. Crinkly/fine wrinkling of skin/parchment-like skin
- d. Extent of disease (figure of eight Vs. localized to labia, localized to clitoris, localized to the perineum or combined)
- e. Erosions
- f. Ulcerations
- g. Hyperkeratosis
- h. Excoriations
- i. Lichenification
- j. Elasticity (or loss of elasticity)
- k. Sclerosis
- l. Architectural changes
 - i. Clitoral hood fusion
 - ii. Labial fusion/resorption
 - iii. Narrowing of the introitus
 - iv. Anterior changes (fusion anteriorly below the clitoris, causing urethral occlusion at its extreme)
 - v. Perianal involvement

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* 1. Please assess the following patient-reported symptoms and determine whether they should be included or excluded in the severity scale:

Please note that the following items have already reached consensus to be included:

- Itch
- Pain unrelated to intercourse (burning, soreness, discomfort, etc.) at rest or during activity
- Pain with intercourse
- Bleeding and pain with intercourse
- Skin tearing with intercourse.

	Include	Exclude
Irritation	<input type="radio"/>	<input type="radio"/>
Dryness	<input type="radio"/>	<input type="radio"/>
Difficulty urinating (dribbling, having to stand up to void)	<input type="radio"/>	<input type="radio"/>
Difficulty with bowel movements (constipation)	<input type="radio"/>	<input type="radio"/>
Bleeding with scratching	<input type="radio"/>	<input type="radio"/>
I don't have any symptoms	<input type="radio"/>	<input type="radio"/>
Other (please specify)	<input type="text"/>	

* 2. 72% consensus has now been reached regarding the method by which to measure symptoms (5-point Likert scale).

Should **frequency** of symptoms be measured as well?

For example how often is a patient itchy: every day, >once per week, >once every 2 weeks, >once per month, <once per month.

- Yes
- No

* 3. Please assess the following patient-reported symptom and determine whether it should be included or excluded in the severity scale:

Please note that the following items have already reached consensus to be included:

-Quality of Life

-Changes/decrease in sexual function

	Include	Exclude
Mood changes (depression/anxiety)	<input type="radio"/>	<input type="radio"/>

* 4. How should these other symptoms (quality of life, sexual function, and mood) be measured? (note: "quality of life" and "change/decreased sexual function" have already gained consensus but there is not yet consensus on how to measure these items.)

- Presence or absence of symptoms (e.g. Yes/No)
- If patient screens positive then... 5 point severity scale (e.g. 1 no impact, 2 barely affected, 3 slightly affected, 4 moderately affected, 5 strongly affected)
- If patient screens positive then... Use previously validated scale for severity (e.g. female sexual function index, dermatology quality of life index or a variation of it, etc.)

* 5. When measuring patient-reported symptoms, is the duration of symptoms important in measuring lichen sclerosis **severity** (assuming that there are symptoms)?

- Yes
- No

* 6. Patients should be asked about their symptoms over what time period i.e. over the past week/month/year? An example would be: "Over the past month, please rate how often you feel the following: itch"

Please note: specifying a time period is necessary in order to use the scale being developed for research now or in the future. Testing this scale for validity will require a standardized time period.

- Month
- 3 months
- 6 months

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* 7. Please assess the following signs on clinical exam and determine whether they should be included or excluded in the severity scale:

(Note: the following items have already reached consensus to be included:

- Fissures
- Whitening
- Crinkly/fine wrinkling of skin/parchment-like skin
- Extent of disease (figure of eight vs. localized to labia, localized to clitoris, localized to the perineum or combined)
- Erosions
- Ulcerations
- Hyperkeratosis
- Excoriations
- Lichenification
- Elasticity (or loss of elasticity)
- Sclerosis

Also Note: if you are unclear as to the meaning of a specific term - please take this into consideration when rating its importance in determining disease severity.

Also note: this list was generated based on our comprehensive review of the literature, all items included are due to our desire to be as all-inclusive as possible to get a consensus on which of these items are important, not due to our own opinion that these items are important.)

	Include	Exclude
Telangiectasia	<input type="radio"/>	<input type="radio"/>
Petechiae	<input type="radio"/>	<input type="radio"/>
Ecchymosis	<input type="radio"/>	<input type="radio"/>
Colour (normal vs abnormal)	<input type="radio"/>	<input type="radio"/>
Pallor	<input type="radio"/>	<input type="radio"/>
Hypopigmentation	<input type="radio"/>	<input type="radio"/>
Hyperpigmentation	<input type="radio"/>	<input type="radio"/>
Erythema	<input type="radio"/>	<input type="radio"/>
Induration	<input type="radio"/>	<input type="radio"/>
Edema	<input type="radio"/>	<input type="radio"/>

Include

Exclude

Atrophy (*Thinning or depression of skin due to reduction of underlying tissue*)

Whether the disease is symmetrical vs. asymmetrical

Other (please specify)

* 8. How should these clinical signs be measured? **Please see the email with the draft scale** to understand how this would look and take this into consideration when making your selection below.

- Presence or absence of signs (e.g. Yes/No)
- If patient screens positive then... Classify sign subjectively as mild, moderate, or severe.
- If patient screens positive then... Classify sign as mild, moderate, or severe depending on percentage of vulva affected (mild - <30%, moderate - 30-50%, severe - >50%)

* 9. The following signs have reached consensus in the first 2 rounds of the survey to be included in the scale. We are now providing you these signs again along with their definitions. Based on these definitions, do you think that this sign should be included or excluded in the severity scale?

	Include	Exclude
Excoriations (<i>Surface disruption (notably excoriations) occurring as a result of the "itch-scratch cycle"</i>)	<input type="radio"/>	<input type="radio"/>
Erosions (<i>A shallow defect in the skin surface; absence of some, or all, of the epidermis down to the basement membrane; the dermis is intact</i>)	<input type="radio"/>	<input type="radio"/>
Ulcers (<i>Deeper defect; absence of the epidermis and some, or all, of the dermis</i>)	<input type="radio"/>	<input type="radio"/>
Sclerosis (<i>Hardening of the tissue</i>)	<input type="radio"/>	<input type="radio"/>
Lichenification (<i>Thickening of the tissue and increased prominence of skin markings. Scale may or may not be detectable in vulvar lichenification. Lichenification may be bright-red, dusky-red, white, or skin colored in appearance</i>)	<input type="radio"/>	<input type="radio"/>

* 10. "Narrowing of the introitus" has previously gained consensus. Should the definition include formation of posterior commissure bands/fourchette webs?

- No, the definition should only include introital narrowing
- Yes, the definition should include formation of posterior commissure bands/fourchette webs

* 11. Should formation of posterior commissure bands/fourchette webs be included or excluded as an individual item in the category of architectural changes?

Note: in the category of architectural changes the following items have already reached consensus to be included:

-Clitoral hood fusion

-Labial fusion/resorption

-Narrowing of the introitus

-Anterior changes (fusion anteriorly below the clitoris, causing urethral occlusion at its extreme)

-Perianal involvement

Include

Exclude

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* 12. How should architectural changes/anatomical distortion be measured? Please see the email with the draft scale to understand what this would look like in practice.

- Presence or absence of that change (e.g. Yes/No)
- If patient screens positive then... Classify subjectively as Mild, Moderate, or Severe
- If patient screens positive then... Classify extent of change as slight, partial, or complete (e.g. slight, partial, or complete labial fusion)
- If patient screens positive then... Classify according to percentage of architecture impacted (e.g. <30% labial fusion, 30-50% labial fusion, >50% labial fusion)

* 13. Would you like your name to be acknowledged in publication(s) that result from this project?

- No
- Yes, Please enter your name below (Name to appear in acknowledgements in any publication arising from this work):

* 14. Would you like to participate in this project in the future in any of the following? (Please choose all that apply)

- No
- Yes, in further surveys regarding refining the scale
- Yes, in an in-person meeting at the next world congress
- Yes, in testing how the scale works (reliability, validity), etc

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15. Thank you so much for participating!

Please feel free to provide us comments.