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Outcome Measures in Adult Vulvar Lichen Sclerosus: A Case Series of Women Diagnosed as Juveniles

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Objectives: Studies on the consequences of juvenile vulvar lichen sclerosus (JVLS) in adulthood are limited. A number of measuring tools are available for analyzing adult vulvar lichen sclerosus (VLS), but these have not been applied in studies on JVLS. The aim is to study physical findings, quality of life, sexual well-being, and self-image in adult women with a history of juvenile VLS.

Materials and Methods: Adult women with a biopsy proven history of JVLS were recruited to be examined and surveyed using available standardized measurement tools. This took place in an outpatient setting by physicians who were not involved in the treatment of participants.

Results: Twenty-seven women (median age 29 years) with a history of JVLS and median time since biopsy of 19.5 years were recruited. Of these women, 59% currently had symptoms, 63% had signs of active disease, and 85% had moderate to severe architectural changes. Despite these residual signs, vulvar specific-quality of life and vulvar self-image scored favorably while generic health-related quality of life was somewhat effected.

Conclusions: JVLS has consequences in adulthood involving physical findings and vulvar quality of life. The use of standardized outcome measures for clinical practice and research purposes facilitates a better understanding of the sequelae to JVLS.

Key Words: girl, juvenile, lichen sclerosus, quality of life, self-image, vulva (*J Low Genit Tract Dis* 2024;28: 295–299)

Vulvar lichen sclerosus (VLS) is a chronic skin disease primarily affecting the anogenital region.¹ Clinical manifestations at any age may include itching, pain, bleeding due to fissures, and permanent loss of vulvar architecture.¹ The disease is most often diagnosed in postmenopausal women, but up to 15% of cases are prepubertal girls.² Juvenile VLS (JVLS) has an estimated prevalence of at least 1:900 girls.³ The majority of cases of VLS in childhood or adolescence do not resolve at puberty even if symptoms are well-controlled.⁴ Studies about how VLS in adults affects sexual well-being, self-image, and other aspects of quality of life (QoL) have been performed, but there is limited research on the consequences in adulthood of juvenile onset VLS.^{5,6}

The aim of this study is to investigate the effect of VLS in adult women with a history of JVLS on physical findings, vulva-specific QoL, and self-image.

MATERIALS AND METHODS

Participants were recruited from our previous study of women with biopsy proven JVLS who had completed dermatology and sexuality questionnaires online.⁶ Eligible participants were invited by email and letter to visit the outpatient department. Informed consent was requested. Participants had never been under the care of members of the research team. Between June and October 2022 the participants underwent a physical exam and filled in questionnaires on-site. Travel expenses were reimbursed.

Symptoms and Physical Examination

Two gynecologists (BM, MtKB) and one dermatologist (CvH or KW) performed the vulvar examination. Disease severity was measured with the Adult Vulvar Lichen Sclerosus Severity Scale (AVLSSS),⁷ the Clinical Lichen Sclerosus Score (CLISSCO),⁸ and additional items, each with a four-point scale 0–3: none-mild-moderate-severe analogous to the description given for the items in the AVLSSS and CLISSCO (Supplement A1, <http://links.lww.com/LGT/A348>).

Questionnaires

RAND-36.v2,⁹ a general health-related quality of life questionnaire, consisting of physical and mental health components, was modified to include the entire mental health component and only the domain ‘general health’ from the physical component. Four domains of the physical health component were deemed not relevant in this study of young women in good general health.

The *Vulvar Quality of Life Index (VQLI)* evaluates the impact of vulvar diseases on QoL.^{*10}

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TABLE 1. Demographics and History of 27 Adults With a History of JVL

Item	Number (percentage)
Age	Median 29 (IQR: 25–35, range 20–49)
Age at biopsy	Median 9 (IQR: 6–15, range 4–18)
Highest completed education	University 7 (26%)/college 9 (33%)/vocational/other 11 (41%)
Ever pregnant	10 (37%)
Number of pregnancies	18
Caesarean sections	4 (22.2%)
Sexually active	Now 21 (77.8%)/previously 5 (18.5%)/never 1 (3.7%)
Age at first sexual encounter	Median 16.5 (IQR: 16–18.5, range 14–26)
LS affected first sexual encounter	6 (22.2%)
Preferred sexual partner	Male 21 (77.8%)/female 2 (7.4%)/both 1 (3.7%)
Negative sexual experience in childhood	4 (14.8%)
Was not aware of diagnosis LS at initial recruitment (2021)	5 (18.5%)
Under surveillance at online survey	13 (48.1%)
Under surveillance at onsite visit	12 (44.4%)
Uses maintenance treatment	3 (11.1%)
Uses medicine on demand only	10 (37.2%)
No current medical treatment	14 (51.9%)
Complaints past year	16 (59.3%)
Recent complaints	No 13 (48.1%)/mild 3 (11.1%)/moderate 8 (29.6%)/severe 3 (11.1%)
Pain during sexual contact	9 (33.3%)
LS in family	8 (29.6%)
Autoimmune disease in family	5 (18.5%)
Vulvar cancer in family	1 (3.7%)

The *Female Genital Self Image Scale (FGSIS)* evaluates a woman's appreciation of her own genitals.^{7,11}

Data was stored using Lime Survey and Gems Tracker software. Descriptive statistics were calculated using Microsoft Excel 365 version 16.0.16212.42301 and IBM-SPSS-Statistics-version-28. IRB approval was granted on March 1, 2022. Participants gave written informed consent.

RESULTS

Twenty-seven women were recruited from the group of 78 eligible participants.⁶ Median age was 29 years. At the time of the visit, 21 women (78%) were sexually active. Five women (18.5%) had not been aware of the diagnosis VLS prior to recruitment for

the online study in 2021, and 14 (52%) were not currently under surveillance. Three women (11%) used maintenance therapy, 10 (37%) used medicinal ointment on-demand, and 14 (52%) used no medicinal treatment. Sixteen women (59%) had experienced complaints in the past year (see Table 1).

Symptoms and Physical Examination

Participants were symptomatic in 59% (AVLSSS) and in 56% (CLISSCO). Eighteen women (63%) had signs of active disease, and 33% had moderate to severe active disease. Architectural changes were seen in 93%, while 85% had moderate to severe architectural changes (see Figure 1). Table 2 and Supplement A1, <http://links.lww.com/LGT/A348>, give complete results. The median scores

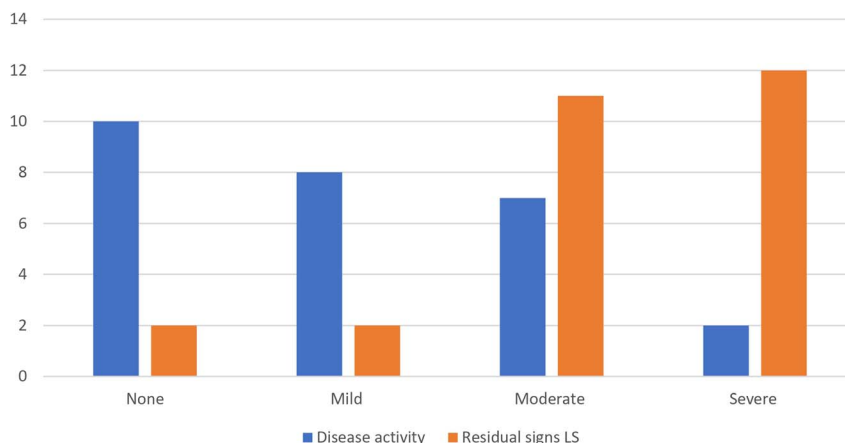


FIGURE 1. Disease activity and residual signs of LS rated by three experts in 27 women with a history of JVL.

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TABLE 2. Symptoms, Signs, and Architectural Changes Following the Adult Vulvar Lichen Sclerosus Severity Scale in 27 Women With a History of JVLS

	Absent (%)	Mild (%)	Moderate (%)	Severe (%)
Symptoms				
^a Itch	18 (66.7)	2 (7.4)	4 (14.8)	3 (11.1)
^a Pain unrelated to intercourse at rest or during activity	19 (70.4)	5 (18.5)	2 (7.4)	1 (3.7)
Changes/decrease in sexual function	25 (92.6)	2 (7.4)	0 (0)	0 (0)
^a Dyspareunia	18 (66.7)	3 (11.1)	6 (22.2)	0 (0)
^a Dysuria/pain with defecation	24 (88.9)	2 (7.4)	1 (3.7)	0 (0)
Quality of life impairment	19 (70.4)	5 (18.5)	2 (7.4)	1 (3.7)
Signs				
^a Fissures	16 (59.3)	8 (29.6)	2 (7.4)	1 (3.7)
^a Whitening	4 (14.8)	7 (25.9)	5 (18.5)	11 (40.7)
Crinkly/fine wrinkling/parchment-like skin	12 (44.4)	6 (22.2)	6 (22.2)	3 (11.1)
Erosions	24 (88.9)	1 (3.7)	2 (7.4)	0 (0)
Ulcerations	27 (100)	0 (0)	0 (0)	0 (0)
Hyperkeratosis	23 (85.2)	3 (11.1)	1 (3.7)	0 (0)
Excoriations	23 (85.2)	2 (7.4)	2 (7.4)	0 (0)
Lichenification	17 (63.0)	5 (18.5)	2 (7.4)	3 (11.1)
Sclerosus	17 (63.0)	2 (7.4)	6 (22.2)	2 (7.4)
Telangiectasia	25 (92.6)	2 (7.4)	0 (0)	0 (0)
^a Petechiae/ecchymosis	24 (88.9)	2 (7.4)	1 (3.7)	0 (0)
Architectural changes				
^a Clitoral hood fusion	7 (25.9)	9 (33.3)	9 (33.3)	2 (7.4)
^a Labial fusion/resorption	2 (7.4)	0 (0)	12 (44.4)	13 (48.1)
^a Narrowing of the introitus	24 (88.9)	2 (7.4)	1 (3.7)	0 (0)
^a Anterior changes (fusion anteriorly below the clitoris, causing urethral occlusion at its extreme)	25 (92.6)	1 (3.7)	1 (3.7)	0 (0)
^a Perianal involvement	17 (63.0)	7 (25.9)	1 (3.7)	2 (7.4)
^a Formation of posterior commissure bands (fourchette web)	17 (63.0)	6 (22.2)	2 (7.4)	2 (7.4)
^bOverall score				
Active lichen sclerosis disease	10 (37.0)	8 (29.6)	7 (25.9)	2 (7.4)
Residual changes due to lichen sclerosis	2 (7.4)	2 (7.4)	11 (40.7)	12 (44.4)

^aThe CLISSCO is composed of these symptoms, signs, and architectural changes.

^bAs scored by visual evaluation and consensus of the three vulvar specialists.

for whitening and labial fusion or resorption were moderate. The median scores of crinkly/fine wrinkling/parchment-like skin and clitoral hood fusion were mild.

Questionnaires

RAND 36.v2. A median of 100 (no impact) (interquartile range [IQR]: 88–100) was found in the social functioning domain. Me-

dian of the other domains were as follows: role-emotional domain median 75 (IQR: 67–92), mental health domain median 70 (IQR: 65–80), vitality domain median 62.5 (IQR: 50–75), and the general health domain median 70 (IQR: 60–80). Complete results are shown in Supplement A2, <http://links.lww.com/LGT/A348>.

VQLI-scores ranged from 0 to 16 with a median score of 7.00 (IQR: 4.00–9.00) (Supplement A3, <http://links.lww.com/LGT/A348>). Thirteen participants experienced minimal or no impact

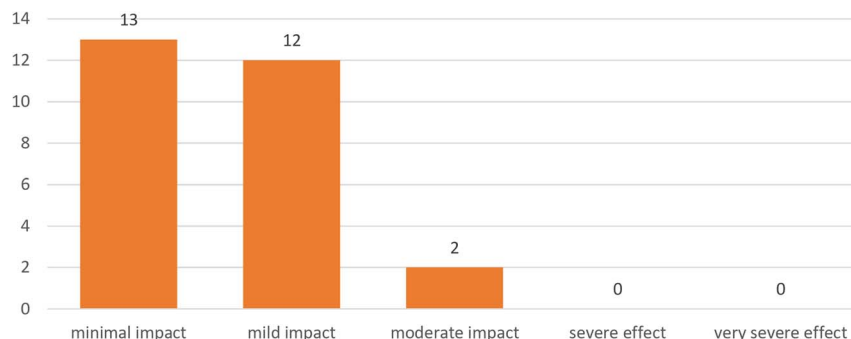


FIGURE 2. Effect of lichen sclerosis on vulvar quality of life using Vulvar Quality of Life Index scores in 27 women with a history of JVLS.

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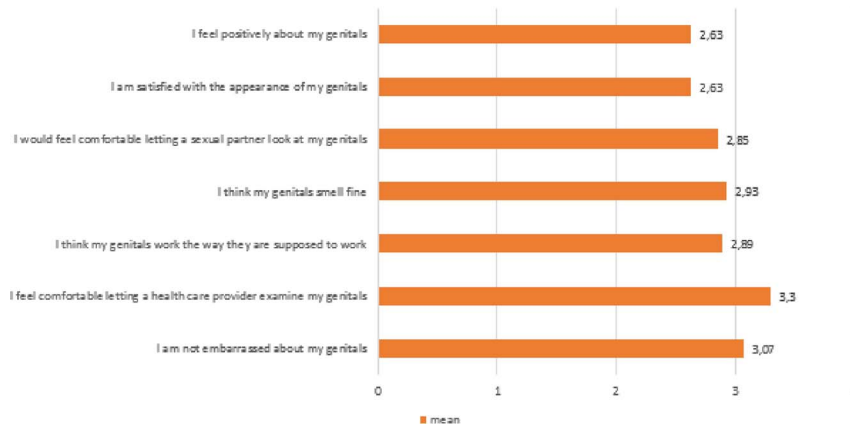


FIGURE 3. Self-image as scored using the FGSIS in 27 women with a history of JVLS.

on vulvar QoL, 12 participants reported mild impact and 2 expressed moderate impact (see Figure 2). A median score of ‘minimal’ was found in the items: “itchy, painful, burning or irritated vulvar skin”, “dysuria, heat intolerance, discharge, or wetness” and “been worried about long-term health implications of vulvar skin condition.” For all other items, median impact was zero.

FGSIS scores ranged from 13 to 28 with a median score of 20.00 (IQR: 18.00–23.00) (Supplement A4, <http://links.lww.com/LGT/A348>). On all items, a median score of 3 (“I agree with statement”) was found (see Figure 3).

DISCUSSION

We report on the results of a study of 27 women with a history of JVLS using available measurement tools for physical findings, vulvar QoL, and vulvar self-image.

In the current series of young women (mean age 29 years), many (59%) were symptomatic. Other studies show similar results though few girls have been followed into adulthood, with one study reporting that 40% remained symptomatic after menarche.¹² We note that despite 85% of women showing moderate to severe architectural changes, just about 10% had narrowing of the introitus or any urethral occlusion, possibly related to their premenopausal status or to the fact that the women were sexually active. Despite the fact that architectural changes were often observed (in 25 of 27 participants) some items were rarely present (eg, hyperkeratosis seen four times, erosions three times or ulcers never seen), resulting in low overall scores on physical findings. These results do not predict whether or not these young women will develop vulvar cancer much later in life.

As only 11% of the women in this study applied any maintenance therapy while the vast majority had moderate to severe irreversible architectural changes, we conclude that it is important that juvenile patients, their caregivers, and health care providers are made aware of current insights regarding the importance of maintenance therapy for juveniles.¹³

Using the RAND-36.v2, we found that four of the five domains tested showed some impact of the disease on health-related quality of life. There was no impact found in the domain *social functioning*, possibly due to adequate coping mechanism developed in childhood. The results of the VQLI show little effect on vulvar-QoL and, thus, seem more favorable than those of generic QoL as scored with the RAND-36.v2. The differences in vulvar-QoL scores in our participants as compared to other studies on VLS, often studying middle aged women, may reflect either generational differences or coping mechanism developed in childhood.¹⁴

We agree with Sheinis et al. that LS severity cannot be assessed by visual inspection alone and needs to include symptoms and QoL assessment as well.¹⁵ Genital self-image is an important aspect of sexual well-being. While no cutoff value for the FGSIS has been established, median score in the present study was lower than that found in the control group in a study on VLS.¹⁶

After data collection was completed a Delphi study by Simpson et al. on core outcome domains for VLS was published, which proposed three core outcome domains to be measured in all clinical trials: ‘symptoms’, ‘clinical (visible) signs,’ and ‘QoL – LS specific’, aspects of LS investigated in our series.¹⁷

A limitation is the lack of a control group. In addition, because our study group was small studying potential correlations between individual item scores such as use of maintenance and current complaints or scarring was not possible. Furthermore, there are no established normal values for the VQLI or FGSIS to help interpret the results. The strength of the present study lies in the use of standardized outcome measures for simultaneous evaluation of physical findings, QoL, and vulvar self-image in women with a history of JVLS, making a comparison to other age groups with VLS, to women with other vulvar diseases and to non-affected women possible.

CONCLUSIONS

A consensus on standardized measurement tools for juvenile and adult VLS in the clinical and research setting is essential. A first step would be performing a systematic review on available measurement tools for VLS based on the recently established core outcome domains.¹⁷ Timely diagnosis, adequate therapy, follow-up and maintenance treatment should be mainstays in the care of girls with JVLS, in the hope that this will lead to a reduction in permanent architectural changes and a reduction in the negative impact on quality of life for these patients.

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[‡]Reference companion manuscript.¹⁷

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