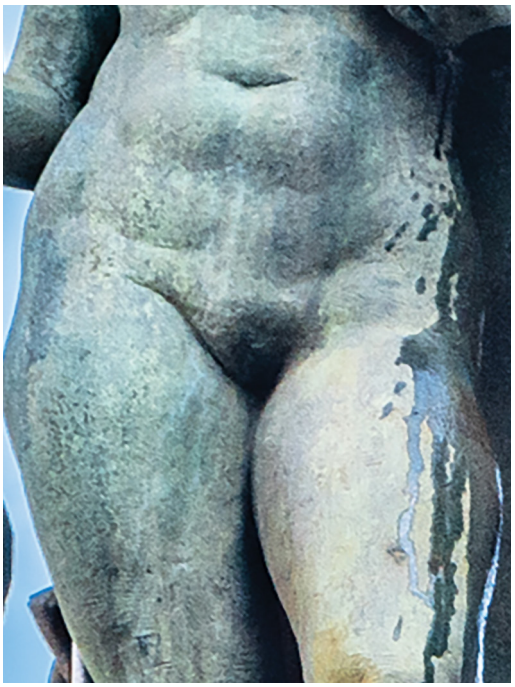
A practical reference for the management of stable vulval lichen sclerosus in general practice



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Background
Vulval lichen sclerosus (LS) is a chronic inflammatory dermatosis affecting the genital skin in women. Vulval LS can cause significant symptoms, architectural changes and increased risk of squamous cell carcinoma. Effective long-term management relies on accurate diagnosis, sustained disease suppression and at least annual clinical review.

Objective
This article provides a practical framework for general practitioners to support lifelong management of vulval LS.

Discussion
Key elements include patient education on self-monitoring, assessment of symptoms, recognition of clinical features and therapy adjustments. Attention should also be given to sexual function, mental health and medical comorbidities. Consistent long-term management of vulval LS improves quality of life, optimises treatment adherence and reduces negative sequalae of the condition.

LICHEN SCLEROSUS (LS) is a chronic inflammatory dermatosis that typically affects the genital skin. Prevalence has been estimated at 1–3%, but this likely underrepresents the true disease burden because of underdiagnosis.[1,2] Disease onset can occur in childhood, prior to menopause and post menopause.[1] Female genital LS can affect the vulva, perineum, anus, buttocks and thighs.[3]

Symptoms of LS can precipitate distress, leading to significant adverse outcomes and poor quality of life.[4] Commonly, women experience pruritus, pain, dyspareunia, decreased orgasm quality and distress regarding genital self-image.[4] Symptoms are heterogenous and range from asymptomatic to severe.

The clinical signs of LS include porcelain white papules and plaques, crinkled skin texture, ecchymoses, fissures and erosions. Disease progression can lead to architectural change of the vulva due to scarring, including labia minora resorption, clitoral burying, adhesions and introitus stenosis.[2,4,5] LS is also associated with a 5–6% lifetime risk of squamous cell carcinoma.[6]

LS is diagnosed either by characteristic clinical appearance (eg pallor or whitening of the skin, architectural changes of the vulva) or skin biopsy. It is a chronic, lifelong condition that is generally treatable.
The mainstay of long-term therapy is topical corticosteroid (TCS) treatment, usually with ultrapotent TCSs. Goals of treatment include symptom relief, halting of the progression of architectural change and reduction of the risk of squamous cell carcinoma (SCC).[1,7] Patients with stable LS require lifelong clinical monitoring.

This article aims to provide a practical reference for the monitoring and management of LS in general practice.

Diagnosis

Diagnosis of LS can be made clinically when hallmark features are seen. These include pallor (sometimes observed in a 'figure-of-8' pattern), typical vulvar architectural changes, and improvement of symptoms with a TCS and/or reversal when the TCS is withdrawn.[1] However, skin biopsy is important when diagnosis is not clear. There are several differential diagnoses that can resemble LS but do not cause architectural change; these include lichen simplex chronicus, contact dermatitis, psoriasis and vitiligo.[8] Similarly, there are conditions that can alter vulvar architecture, including erosive vaginal lichen planus, autoimmune bullous disease, severe drug reactions, Crohn's disease and malignancy.[9–12] Adding to the diagnostic complexity, multiple conditions can co-exist. Furthermore, up to one-third of women with LS are asymptomatic.[13] Referral to a tertiary vulval unit or vulvar specialist is encouraged when diagnosis is in doubt.

Stable disease

Women with LS who are asymptomatic with objectively suppressed skin disease require lifelong 12-monthly examinations[1] to assess for:

- symptoms
- signs of disease activity
- architectural changes
- malignancy
- comorbid conditions.

Symptom control alone is not a reliable indicator of adequate treatment, necessitating regular examination.[13] Regular visits also allow the clinician opportunity to engage with patient progress, ensure treatment adherence and provide encouragement regarding ongoing TCS use.

Vulval cancer and human papillomavirus status

Vulval SCC can arise from two main pathways: human papillomavirus (HPV)-dependent and HPV-independent (HPV-I), the latter of which is associated with LS.[14] The precursor to HPV-I SCC is termed differentiated vulval intraepithelial neoplasia or HPV-I vulval intraepithelial neoplasia (dVIN or HPV-I VIN). The HPV-dependent precursor is termed high-grade squamous intraepithelial lesion (HSIL) or usual-type VIN.[15] Immunohistochemistry staining for p16 and p53 proteins on lesional tissue is important in determining HPV association, as HPV-I SCC is associated with treatment resistance, higher recurrence and higher risk of death when compared with HPV-dependent disease.[14]

A 2022 systematic review showed a HPV-I SCC risk of 2.2% and a dVIN risk of 1.2% in patients with LS.[7] Importantly, there was also a significant reduction in malignancy risk for those who were adequately managed with TCSs, in line with cohort studies.[16–18] Patients with LS should be counselled on SCC risk, with an emphasis on the importance of adherence to TCS therapy.

Examination

Examination should focus on skin changes and architectural changes. Skin changes can include pallor (whitening), hyperkeratosis, ecchymoses, skin fissures and atrophy, and the presence of these features can suggest LS activity (Figures 1 and 2).

Architectural changes are typically gradual and can include: clitoral burying, labial resorption/agglutination and introital stenosis. Clinical photography is considered the gold standard to monitor

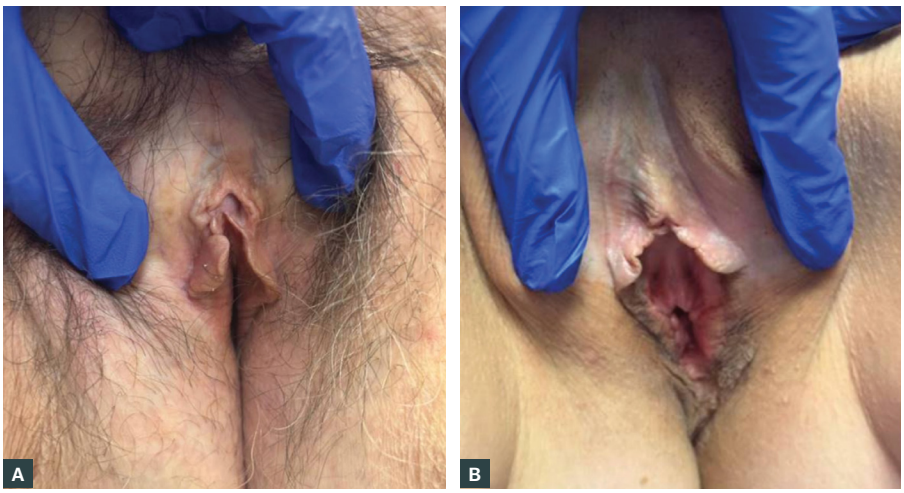

Figure 1. Clinical images of two patients with mild lichen sclerosus. **A.** Inactive/stable lichen sclerosus with resorption of the inferior aspect of the right labia minora. **B.** Active lichen sclerosus with pallor to interlabial sulci, flattening of the clitoral hood and asymmetrical labia minora.

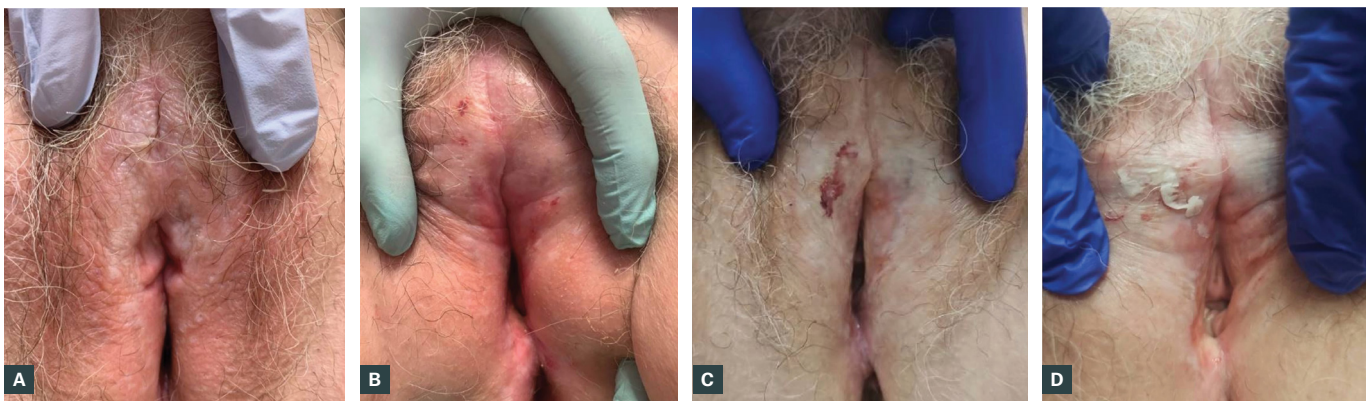

Figure 2. Clinical images of one patient with active advanced vulval lichen sclerosus demonstrating varying clinical signs over 2 years. **A.** Architectural changes demonstrated are clitoral burying and anterior midline fusion, with resorption of the labia minora. There is pallor to the superior bilateral labia. **B.** Six months later, pallor is more subtle to the labia and more pronounced to the perianal region. There are small ecchymoses to the vulva. **C.** Twelve months later, there is widespread pallor to bilateral vulva and a prominent ecchymoses to the right superior vulva. **D.** Six months later, there is hyperkeratosis to the right vulva and introital stenosis.

for architectural change.[19] There is no consensus on a standardised scoring system for severity of LS.[5] A persistent (steroid-resistant) plaque, patch or erosion should raise concern for an alternative diagnosis or malignancy.[1]

Self-examination, where feasible, is helpful for women to identify changes in their own skin, allows for as-needed escalation to medical review, assists with precise treatment application and affords a sense of control over the therapeutic journey.[20] Clinical photography can also be used as an instructive tool for self-examination and TCS application. The International Society for the Study of Vulvovaginal Disease (ISSVD) has published a step-by-step self-examination guide for patients, accessible in several languages,[21] as well as a guide for healthcare professionals.[22]

Table 1. Commonly used topical corticosteroid ointments for vulval lichen sclerosus

Potency	Generic name	Brand name examples	Listed on the Pharmaceutical Benefits Scheme
Mild	• Hydrocortisone 0.5–1%	Cortic-DS	Yes
Moderate	• Methylprednisolone aceponate 0.1%	Advantan Fatty Ointment Supriad	Yes
	• Mometasone furoate 0.1%	Elocon Novasone	Yes
Potent	• Betamethasone dipropionate 0.05%	Diprosone Eleuphrat	Yes
Ultrapotent	• Betamethasone dipropionate 0.05% in optimised vehicle	Diprosone OV	No
	• Clobetasol propionate 0.05%	Xobet	Yes
	• Clobetasol propionate 0.1%	Compounded	No

Treatment

The mainstay of LS treatment is with a TCS, usually potent or ultrapotent. Management consensus in Australia is lifelong suppression therapy rather than on-demand use for symptom control.[1] Topical calcineurin inhibitors, such as tacrolimus, have some evidence for efficacy, although meta-analyses suggest TCSs are superior.[23] Recalcitrant cases of LS (control not achieved on TCS alone) are sometimes managed with systemic agents, usually at tertiary vulval units.[1]

Ointment preparations of TCSs are preferred over creams for vulval skin because of better penetration, better barrier function and lower allergen risk.[23,24] Commonly used preparations in Australia are shown in Table 1. Notably, clobetasol propionate 0.05% ointment is now available on the Pharmaceutical Benefits Scheme as Xobet; formerly it was only available as a compounded preparation.

Initial management of active LS depends on disease activity, patient preference, clinician experience and availability of products. Patients are typically commenced on an ultrapotent or potent TCS. Initial treatment and treatment for acute flares of LS are often managed with a 'blitz' dose-tapering regimen, such as clobetasol propionate 0.05% daily for 1 month, alternate days for 1 month, then 2–3 times per week. Units of

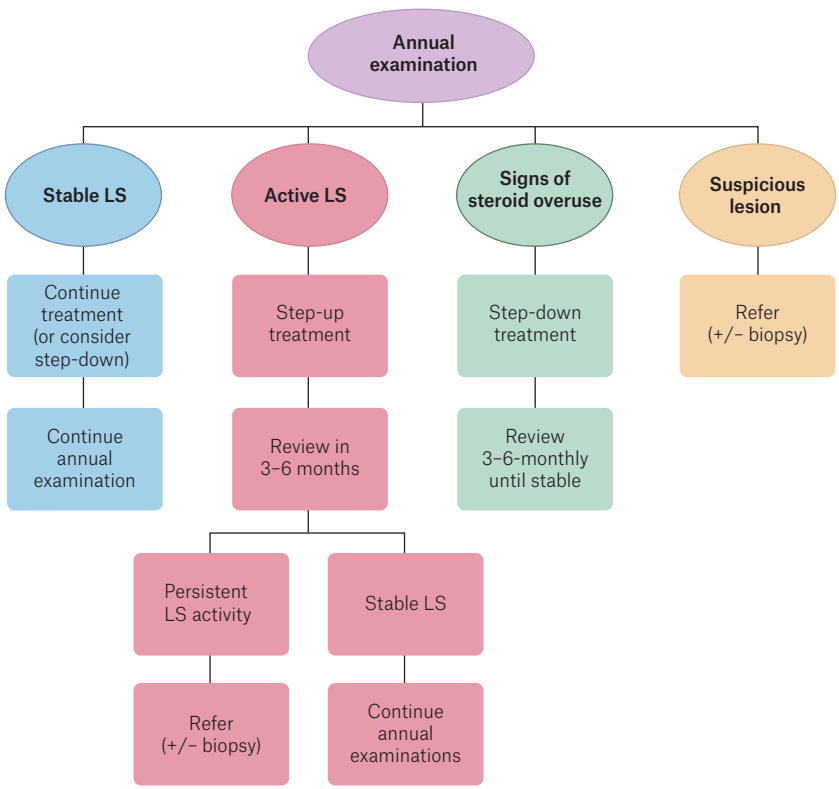

Figure 3. Management guide for women with stable vulval LS on topical corticosteroid therapy.
LS, lichen sclerosus.

application are often described by fingertip unit, or by 'lentil' (1/4 fingertip unit) or 'pea' (1/2 fingertip unit).[1]

Maintenance treatment is required lifelong to maintain symptom suppression and minimise the risk of progressive anatomical change and malignancy (Figure 3).[18,25] Patients should be counselled on the importance of continuing maintenance treatment even when asymptomatic.
LS severity can vary greatly; therefore, for some women, ultrapotent TCS application is required twice-daily, whereas for others, a moderately potent TCS twice-weekly is adequate. Where disease activity varies by anatomical site, the use of different topical corticosteroid potencies may be appropriate, for example, a mildly potent TCS for the perianal skin and a potent TCS for the vulva.[1]

TCS application with a mirror is encouraged, especially when specific anatomical areas are being targeted because of LS activity or if there is uneven ointment application. Hair-bearing skin should be avoided if unaffected by LS (Figure 4). When clinical photography is incorporated into medical reviews, it may be a useful adjunct to treatment counselling for both clinicians and patients.

The following is an example of patient instructions for TCS application:

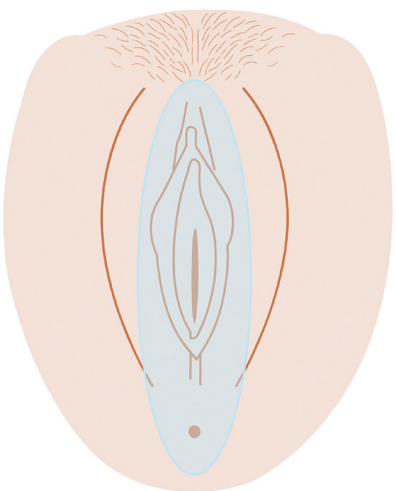

Figure 4. General treatment field for corticosteroid application in vulval lichen sclerosus demonstrated by the blue shaded area.

- The prescribed TCS ointment may be applied in the morning or evening, or twice daily if directed.
- Using a hand-held mirror, identify the specific areas of affected skin requiring treatment. Some patients find it helpful to rest one foot on a stool or elevated surface to improve visual access.
- Gently separate the labia and apply a small amount of ointment (eg a half–pea-sized quantity) directly to the affected skin. This may include the area from the clitoris down towards the vaginal opening and perianal region.
- Gently spread the ointment in a thin layer over the affected areas only, taking care to avoid application to hair-bearing skin.

Other measures important for vulval skin barrier care include:

- regularly using emollient such as white soft paraffin
- using soap-free skin cleanser, ideally an emulsifying agent
- wearing loose underwear composed of natural fibres
- avoiding sanitary pad or liner use
- avoiding overwashing or bidet use.

Safety of topical corticosteroids

Adverse effects from treating LS with TCSs are uncommon. Steroid rosacea or steroid dermatitis, especially on hair-bearing vulva, can indicate TCS overuse.[1] It can present as poorly demarcated red to purple patches (Figure 5) and might be accompanied by burning or discomfort.[1] Other findings of TCS overuse include telangiectasia, epidermal cysts and atrophy. Management may include a temporary reduction in steroid potency; however, rebound vasodilation to the skin may produce discomfort and itch. Regular application of barrier creams and cautious titration of steroid strength may help mitigate symptoms.[1]

Multiple cohort studies of vulval LS show that the benefits of long-term TCS therapy clearly outweigh the risks, with no irreversible local or systemic adverse effects reported.[26] Despite this, steroid phobia is common and represents a significant barrier to treatment adherence.[27] Common patient concerns include fear of overuse, reliance and ambiguous labelling such as 'use sparingly'.[28] Pharmacists also play an important part in patient education; however, variation in messaging regarding long-term TCS use may inadvertently reinforce patient uncertainty.[29] Clinicians should proactively address steroid phobia, provide clear and consistent counselling, and prepare patients for potentially conflicting advice. The Melbourne Sexual Health Centre and ISSVD have published patient information leaflets that reinforce the safety of TCSs in LS.[30,31]

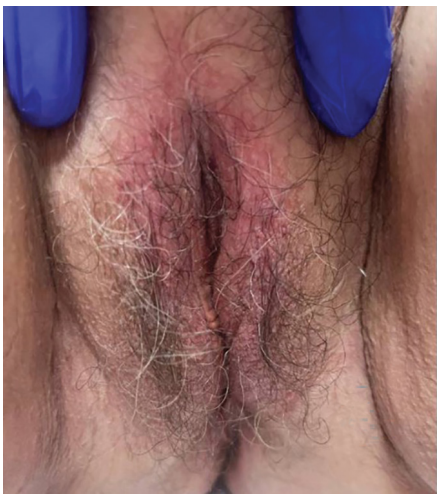

Figure 5. Steroid rosacea in a patient with lichen sclerosus. Poorly demarcated red patches can be seen on the labia majora.

Multifactorial vulval itch

Pruritus is the most common symptom of active LS, but it should not be assumed to reflect LS activity without careful history and examination.[32] LS frequently coexists with, or is difficult to distinguish from, other causes of vulvar itch. This includes vulvovaginal candidiasis, oestrogen deficiency (genitourinary syndrome of menopause), contact dermatitis and inflammatory dermatoses such as eczema and psoriasis.[8] Vulvovaginal candidiasis is common and may cause itch and discomfort without typical discharge, particularly in women using oestrogen replacement.[33] Persistent or worsening itch in the absence of objective examination findings attributable to LS should prompt reassessment for an alternative or concurrent diagnosis.

Sexual function

Sexual dysfunction is common in patients with LS, affecting up to 50%.[4] Commonly, women experience dyspareunia, decreased orgasm quality and distress regarding genital self-image. Furthermore, sexual activity can cause symptom flare, contributing to fear or distress about anticipated sexual activity.[4] Psychosexual counselling has been shown to increase quality-of-life scores for women with LS and should be considered for those experiencing sexual dysfunction.[34] Pelvic floor physiotherapy is also a minimally invasive tool that is useful to address sexual dysfunction and pelvic floor pain.[1]

Comorbidities

The exact pathogenesis of LS is unknown, but current research hypothesises a combination of factors that have genetic, immune, hormonal and environmental bases.[1] There is a correlation between LS and some autoimmune conditions, including thyroid disease, pernicious anaemia, vitiligo and psoriasis.[11,35–37] An association has also been described between LS and immune checkpoint inhibitor therapy.[38] With regard to mental health, several studies have published an association between anxiety and depression and LS, reporting prevalence rates of 40% and 58% for depression and 27% for anxiety.[34,39] Factors that can complicate management, such as urinary incontinence, should also be optimally managed.[1]

Conclusion

It is important to be aware of long-term management recommendations for patients with vulval LS. Optimal monitoring focuses on holistic care, improves treatment outcomes and reduces the risk of negative sequelae. The aim of this article was to outline practical considerations for managing patients with stable vulval LS in the primary care setting.

Key points

- Lifelong topical corticosteroids are the mainstay of treatment for vulval LS.
- Women with stable vulval LS should be examined every 12 months.
- Examination should focus on skin changes and architectural (anatomy) changes.
- Referral or biopsy is necessary for suspicious or treatment-resistant lesions.
- Sexual dysfunction is common, and a multidisciplinary approach is recommended.

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