

# Lichen sclerosis: Review of a vulvar dystrophy with malignant potential

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by ERIC D. SWISHER, MD

**T**he lichenoid vulvar disorders are a group of inflammatory dystrophic conditions with some overlapping clinical features but unrelated pathophysiology. Lichen planus, lichen sclerosis, and lichen simplex chronicus are distinct conditions within this group of dermatoses. Lichen sclerosis is an acquired chronic dermatosis that has a 6- to 10-fold predilection for women, so the disorder is especially relevant to the gynecologic clinician's practice. There is an apparent bimodal distribution with the disorder affecting prepubertal girls and menopausal women.<sup>1,2</sup> This age-related distribution may warrant reconsideration as 40% of women with the disorder report onset of symptoms

prior to menopause and, contrary to previous perceptions, a majority of childhood cases do not resolve following puberty.<sup>3,4</sup> Historically, the disorder had a widely variable nomenclature that has evolved significantly and may cause confusion. The International Society of Vulvar Disease established the term *lichen sclerosis* as the standard and eliminated previous terms including *lichen sclerosis et atrophicus*, *Bowen's disease*, *Kraurosis vulvae*, *leukoplakic vulvitis*, and *hyperplastic vulvitis* among others.<sup>5</sup> Lichen sclerosis is significantly debilitating, with vulvar pain and pruritus compounded by scarring, stenosis, and phimosis, which contribute to dyspareunia and other functional impairments. The

potential for malignant transformation in the chronically inflamed tissues is an additional sequela, which may benefit from therapy and requires long-term informed clinical follow-up.<sup>3</sup>

## Incidence

The precise incidence of lichen sclerosis is uncertain, as many cases are asymptomatic or remain undiagnosed. The US gynecologic population has a 1.7% to 3% prevalence of biopsy proven lichen sclerosis.<sup>6,7</sup> Consolidation of the nomenclature and improving detection rates may have affected the apparent increase in incidence identified in a 20-year review of the disease.<sup>8</sup>

Lichen sclerosis has been associated with squamous cell carcinoma of the



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vulva at a cumulative rate of 6.7%.<sup>8</sup> The potential for malignant transformation of lichen sclerosus ranges from 2% to 6%. This malignant transformation is by a differentiated vulvar intraepithelial neoplasia (VIN) pathway and arises without the effect of HPV.<sup>8</sup> The risk increases with age and duration of disease; therefore, lichen sclerosus warrants lifelong surveillance.

### Pathophysiology

The precise pathogenesis of lichen sclerosus is not well established, though it is generally thought to be of an autoimmune etiology. In fact, autoantibodies to extracellular matrix protein are present in 74% of women with lichen sclerosus.<sup>9</sup> Patients with lichen sclerosus also are commonly affected by other autoimmune disorders such as thyroiditis, which coexists in 12% to 16% of cases.<sup>3,10</sup> There appears to be a role for genetic predispositions as evidenced by monozygotic twin case studies and the increased prevalence among family members of those affected.<sup>2,11-13</sup> Infectious etiologies have been postulated to include *Borrelia burgdorferi* and human papillomavirus, with little data to suggest a causative association. Local factors, currently undefined, appear to contribute to lichen sclerosus. This has been demonstrated by grafting affected vulvar that normalizes elsewhere, and normal skin grafted to the vulva becomes affected in patients with existing disease.<sup>11</sup>

The role of estrogen deficiency as a singular or meaningful etiology is contradicted by the persistence of the disease in prepubertal girls following menarche and the ineffectiveness of topical estrogen or androgen therapy.<sup>14-16</sup>

Histologic changes of vulvar lichen sclerosus include epidermal atrophy with loss of rete ridges and homogenization of

the dermis with collagen. There is typically a band of T-lymphocyte infiltrate near the basement membrane.<sup>3,17</sup> These changes may regress to a more nonspecific inflammatory pattern or approach normalcy following therapy; however, the structural anatomic alterations on gross inspection will be permanent.

### Clinical Presentation

The most common complaints among patients diagnosed with lichen sclerosus are pruritis, vulvar irritation, and vulvar pain. Despite the severity of symptoms associated with many cases of lichen sclerosus, some women may actually remain asymptomatic. Fissures may be reported or observed. Pain with elimination may develop in more severely affected patients and dyspareunia is common. As the inflammatory changes progress, dysuria and dyspareunia become more prevalent as stenosis may develop. The “parchment paper” or porcelain white presentation of vulvar tissues affected by lichen sclerosus appears thin and atrophic, though histology demonstrates collagenous thickening of the dermis. There is regression of the normal architecture including narrowing of the introitus, phimosis of the clitoral hood, and, atrophy of the labia minora (**Figure 1**). The common distribution is a figure 8 involving the vulvar and the anovaginal tissues of the posterior fourchette, though early in the disease the affected tissues may be more focal (**Figure 2**). These preliminary focal white atrophic plaques will eventually coalesce into a more generally affected vulva.<sup>2</sup>

The differential diagnosis should include atrophy, candidiasis, contact dermatitis, lichen planus, lichen simplex chronicus, pemphigus, atopic dermatitis, vitiligo, and psoriasis,



**FIGURE 1.** Lichenification and flattening of the vulvar architecture with phimosis of the clitoral hood. Photo credit: Dr. Nathan Webb, Associate Professor Department of Obstetrics and Gynecology, VCU Medical Center.

among others. Some trial and error may be warranted in atypical cases, but biopsy is always more definitive and remains the gold standard. The other lichenoid vulvar disorders are usually differentiated by exam, and, despite the shared nomenclature, these are entirely unrelated pathologic abnormalities. Lichen planus presents as an erythematous vestibule with erosions and vaginal or oral involvement, which is not found in lichen sclerosus. Lichen simplex chronicus is a hypertrophic response to an intractable itch-scratch cycle with edema, leathery inflammatory thickening, and excoriations. Fissures may be present in both conditions, but scarring is atypical of lichen simplex.<sup>1</sup> The

shiny, inflamed appearance of a vulvar dystrophy may be mistaken for vulvar candidiasis and if so, the absence of a prompt response to antifungal therapy should lead to the reconsideration of lichen sclerosus. Biopsy for diagnosis is a valuable adjunct to clinical findings in some cases, but a necessity when the clinical findings are atypical or vulvar intraepithelial neoplasia is suspected.

In children, the mean age of onset was 5 years with the actual diagnosis being delayed an additional 1-2 years.<sup>18, 20</sup> Similar to the untreated course in adults, failure to treat pediatric and adolescent cases leads to progressive scarring and loss of architecture.<sup>20, 21</sup> These changes are irreversible and a risk of carrying a long-term disease burden into adulthood for the 75% of adolescent cases that persist.<sup>19, 20</sup> The purpura sometimes associated with these dystrophic vulvar tissues in pediatric patients may also be misidentified as evidence of physical abuse.<sup>20</sup>

## Therapy

The expectation of therapy is symptom control and tissue preservation through remission rather than resolution, thus lichen sclerosus management should be lifelong. Topical steroids remain the cornerstone of therapy. Tissue atrophy often associated with extended topical steroids does not appear to be a complication of steroid use in lichen sclerosus patients. Therefore, adequate steroid therapy for clinical effect and long-term therapy for maintenance should not be limited by the concern for steroid complications unless identified on exam. Therapy is typically initiated with a superpotent topical corticosteroid (clobetasol propionate 0.05% or fluocinonide 0.1%) once or twice daily for 2 weeks and then nightly for



**FIGURE 2.** Figure 8 distribution of dermatosis changes typical of lichen sclerosus. Photo credit: Dr. Nathan Webb, Associate Professor, Department of Obstetrics and Gynecology, VCU Medical Center.

4 weeks (**Table 1**).<sup>20</sup> Clinical trials have generally provided continuous therapy up to 12 weeks, so a longer duration of this initial intense steroid use is not discouraged. Three to 6 months of continuous daily therapy may be required to induce remission in some patients, so consistent re-evaluation is the best guide to therapy.<sup>20, 22</sup> Milder cases may be managed with an intermediately potent steroid such as betamethasone dipropionate 0.05% or triamcinolone acetonide 0.5% in a similar twice-daily pattern until tapering to nightly application. Once remission is achieved, maintenance therapy 2 nights per week is advisable, and this treatment is typically indefinite.<sup>3, 11</sup> Some patients will not be compliant long term, but cessation of therapy typically leads to

recurrence of symptoms and further tissue dystrophy. Lasting remission without maintenance therapy is less common, and that subgroup of patients is difficult to predict.<sup>11</sup>

The steroid is best provided in ointment form and dosage should be measured in fingertip units (FTU). One FTU is the bead of ointment from a 5-mm nozzle extending from the fingertip to the most distal crease of the index finger (**Figure 3**). A typical application is 0.5 FTU, and demonstrating this instruction to patients will minimize overuse. This dosage volume should equate to depletion of a 30-gram tube of steroid during a 6-month period of therapy.<sup>11</sup>

Reevaluation at 4 to 6 weeks confirms the response to therapy and guides the future steroid regimen for remission.<sup>20</sup> Adherent patients who fail to respond should be promptly reevaluated for alternative diagnoses, as the steroid responsiveness of lichen sclerosus is highly reliable. There should be a low threshold to biopsy unresponsive or atypical patients. An 18% risk of malignancy with lichen sclerosus concomitant with VIN vs the 3% risk for lichen sclerosus alone demonstrates that VIN is a significant gauge of risk. Patients with suspected or confirmed VIN should be liberally evaluated by biopsy.<sup>8</sup> In addition to ameliorating pruritus and vulvar pain, a prospective trial now confirms that steroid therapy does in fact reduce the long-term risk of malignancy.<sup>20, 22</sup> Patients should be followed semiannually or annually once adequately treated. Subspecialty referral may be indicated for patients with complicated presentations, steroid-resistant clinical findings, and VIN or carcinoma, depending on the comfort and experience of the provider.<sup>11</sup>

Although the corticosteroids are the

only therapy to have been prospectively shown to reduce the risk of lichen sclerosis associated malignancy, alternatives to corticosteroid therapy have been used historically and in clinical trials. The use of testosterone for the treatment of lichen sclerosis has no place in current gynecologic practice. Estrogen replacement therapy does address the concomitant vulvar atrophy present in postmenopausal patients but has no significant therapeutic effect on lichen sclerosis. Studies comparing calcineurin inhibitor immunosuppressant therapy such as tacrolimus have failed to exceed the response to topical corticosteroid therapy. Data suggest there is little difference between pimecrolimus and clobetasol propionate in relieving symptoms, though clobetasol propionate appears more effective by examination criteria.<sup>16</sup> Among the newest modalities and a novel approach to the disease, platelet rich plasma, showed initial promise but failed to demonstrate adequate efficacy in a placebo-controlled clinical trial.<sup>23,24</sup>

There are several energy-based approaches to therapy under investigation, including high-intensity focused ultrasound, fractional laser, and photodynamic therapy. Photodynamic therapy and UV phototherapy demonstrate

efficacy but currently have not demonstrated superior results compared with steroids. The impact on risk of malignancy also is not yet established for these modalities. Superficial ablation using fractional CO<sub>2</sub> laser shows promise in small preliminary investigations, but prospective randomized trials are incomplete. High intensity focused ultrasound (HIFU) improved symptoms in 47% to 90% of patients treated in clinical trials, but energy-related cutaneous complications affected up to 10%. Histologic evaluation does show significant promise for HIFU therapy, which may surpass the efficacy of topical steroids.<sup>3</sup> The logistics of therapy for the energy-based approaches affects these modalities as some patients may not accept repetitive weekly visits for therapy. Equipment costs will preclude availability in lower-volume clinics. Thermal complications also are a factor unique to these modalities. Further study will likely define a role for those energy-based therapies proven to be successful as economics guide practical applicability for an individual practice or institution.

Though uncommonly deployed in current practice, surgical management of clitoral phimosis may be beneficial for some patients and success rates are very promising. Dissection by blunt or sharp release of the clitoral hood may address pain, pseudocyst accumulation, and anorgasmia.<sup>3,11</sup> Patients affected by stenosis of the introitus may improve with dilator therapy, but surgical management may provide an efficacious alternative. Case series evaluating perineoplasty with vaginal mucosal advancement as well as simple surgical release of the introital scar in unresponsive patients have demonstrated significant success. Postoperative steroid therapy still is recommended, and larger trials will be

**TABLE 1. EXAMPLES OF COMMON TOPICAL STEROIDS FOR VULVAR THERAPY**

**Super High Potency Steroids**

- Fluocinonide 0.1%
- Clobetasol Propionate 0.05%
- Halobetasol Propionate 0.05%

**High Potency Steroids**

- Fluocinonide 0.05%
- Clobetasol Propionate 0.025%
- Halobetasol Propionate 0.01%

**Medium Potency Steroids**

- Triamcinolone Acetonide 0.025%
- Fluocinolone Acetonide 0.025%

required to establish the role of surgical intervention.<sup>3</sup>

**Conclusion**

Lichen sclerosis is a debilitating and premalignant vulvar condition that benefits from long-term follow up and maintenance steroid therapy. Early and ongoing intervention may prevent irreversible loss of anatomic architecture. Long-term annual or semi-annual evaluation even with patients treated to remission is prudent due to the risk of recurrence and the risk of malignancy. Biopsy should be promptly considered for patients unresponsive to corticosteroid therapy, suspected vulvar intraepithelial neoplasia, or atypical clinical presentations. Some patients may require management in a vulvar specialty clinic, but the typical patient will do well in a general gynecologic practice with long-term steroid maintenance and regular examinations to monitor for malignant transformation. ■

**FOR REFERENCES VISIT**  
[contemporaryobgyn.net/lichensclerosis](http://contemporaryobgyn.net/lichensclerosis)



**FIGURE 3.** One fingertip unit as a guide to dosing topical steroid.