IMPORTANCE  Adult vulvar lichen sclerosis (VLS) may be complicated by loss of vulvar structure and vulvar carcinoma. There is a lack of evidence as to the ideal method to maintain long-term remission and prevent complications.

OBJECTIVES  To determine whether long-term preventive topical corticosteroid (TCS) treatment of VLS, with a target outcome of induction and maintenance of normal skin texture and color, reduces the risk of vulvar carcinoma, relieves symptoms, improves function, and preserves vulvar architecture, and to evaluate the adverse effects of treatment.

DESIGN, SETTING, AND PARTICIPANTS  A prospective longitudinal cohort study was conducted in 507 women with biopsy-proved VLS from January 2, 2008, through September 26, 2014, in the private practice of a dermatologist and a gynecologist in Sydney, Australia.

INTERVENTIONS  Preventive treatment using TCSs of various potencies, adjusted to meet a target outcome of normal skin color and texture, with regular long-term follow-up by a dermatologist or gynecologist.

MAIN OUTCOMES AND MEASURES  Symptoms or signs of VLS, scarring, development of malignant neoplasms, and adverse effects.

RESULTS  The mean age at presentation was 55.4 years (range, 18-86 years); duration of symptoms at presentation, 5.0 years (range, 0.1-40.0 years); and duration of follow-up, 4.7 years (range, 2.0-6.8 years). Remission was induced with a potent TCS, followed by regular preventive TCS treatment of a potency titrated to achieve the target outcome. Patients were followed up at least annually. A total of 150 patients (29.6%) did not carry out the advised treatment and were considered partially compliant. A total of 357 patients (70.4%) adhered to treatment instructions and were considered compliant. Biopsy-proved squamous cell carcinoma or vulvar intraepithelial neoplasia occurred during follow-up in 0 of the compliant patients vs 7 (4.7%) of the partially compliant patients (P < .001). Suppression of symptoms occurred in 333 (93.3%) compliant patients vs 87 (58.0%) partially compliant patients (P < .001). Adhesions and scarring occurred during follow-up in 12 (3.4%) compliant patients and 60 (40.0%) partially compliant patients (P < .001). Reversible TCS-induced cutaneous atrophy occurred in 4 (1.1%) compliant patients and 3 (2.0%) partially compliant patients.

CONCLUSIONS AND RELEVANCE  This prospective, single-center, longitudinal cohort study of adult patients with VLS suggests that individualized preventive TCS regimens that achieve objective normality of skin color and texture and are used by compliant patients who attend regular long-term follow-up visits may modify the course of the disease. There was a significant difference in symptom control, scarring, and occurrence of vulvar carcinoma between compliant and partially compliant patients. The adverse effects of TCSs were minimal.
Vulvar lichen sclerosus (VLS) is an uncommon skin disease that is frequently complicated by the loss of normal vulvar architecture and less commonly by vulvar squamous neoplasia (VSN), including vulvar intraepithelial neoplasia and invasive squamous cell carcinoma. Before the finding in 1991 that VLS could be suppressed by superpotent topical corticosteroids (TCSs), rates of vulvar squamous cell carcinoma among patients with VLS were reported to be about 5%.2-4

The current accepted management guidelines for VLS advocate the use of superpotent TCSs as first-line treatment to achieve remission. There is good evidence to support this approach.1,5 Less is known about the best practice to maintain remission, the ideal duration of follow-up, and whether long-term management can prevent complications. In a previous study, 2 of us recommended long-term treatment and surveillance for all patients.6 However, other authors have contended that only patients with difficult-to-control VLS should be regularly followed up and have expressed concern that long-term follow-up is a burden on the health care system.7-10

This prospective, single-center cohort study of 507 women with biopsy-proved VLS aims to answer the following questions about long-term management: whether individualized TCS regimens that treat to a target outcome of continued maintenance of normal skin color and texture improve function, achieve preservation of the vulvar architecture, and reduce the risk of VSN without significant adverse effects.

Methods

A prospective, single-center cohort study of women with biopsy-proved VLS who were treated with TCSs was conducted from January 2, 2008, through September 26, 2014. Patients were considered compliant if they self-reported that they followed treatment instructions “most of the time” or “all of the time” and partially compliant if they self-reported that they followed treatment instructions “some of the time,” “little of the time,” or “none of the time,” either in terms of frequency of application and/or potency of TCS. Our study compared these 2 groups. All patients were evaluated in the private practice of the authors (J.B. and G.F.) in Sydney, Australia. Inclusion criteria were age older than 18 years, biopsy-proved VLS, and having been followed up for a minimum of 2 years. Using previously published studies,2-4 we assumed a 5% risk of VSN. Based on an initial audit of patients attending our practice, only 90 (67.0%) patients were compliant. The other 45 (33.0%) patients were partially compliant. To detect a decrease to 1.0% incidence of VSN in the compliant group compared with the partially compliant group with 80% power at 5% significance, we required a total of 504 patients. At the time of study completion, we had included 357 compliant and 150 partially compliant patients (compliance rate, 70.4%) for a total of 507 patients.

This study was approved by the Human Research and Ethics Committee of the Northern Sydney Local Health District. Written or oral consent was obtained from all patients.

Data were systematically collected and recorded in a computerized database that was established in 2008 (Genie Solutions). The following characteristics were recorded for all patients:

1. Historical features: age, ethnicity, menopausal status (premenopausal, postmenopausal without hormone therapy, or postmenopausal with hormone therapy), duration of symptoms, and previous treatment.
2. Symptoms: itching, vulvar pain, and dyspareunia.
3. Clinical features: distribution (figure of 8, localized on an area of the vulva, clitoris, or perineum, or symmetrical involvement of the vulva), fissuring, telangiectasia, pigmentation, labial fusion (none, anterior, posterior, or both), clitoral hood fusion, and VSN (squamous cell carcinoma, vulvar intraepithelial neoplasia, or none); 73% of patients consented to clinical photographic monitoring. For those who did not consent, detailed clinical descriptions were used.
4. Severity of disease for the purposes of selection of potency of TCSs: defined by degree of hyperkeratosis: mild (1+), moderate (2+), severe (3+), or very severe (4+) (Figure 1). Scarring was not used as a severity marker because it does not change with treatment.
5. Adverse effects: stinging or irritation caused by topical therapy; atrophy evidenced by skin fragility, telangiectasia, and visible veins; and corticosteroid dermatitis evidenced by development of erythema with burning or soreness.

Initial treatment regimens were individualized, with the target outcome being an objective return of the vulvar skin to normal color and texture. Patients were initially treated with a single TCS agent, applied daily, to achieve symptom control. The most commonly used agent in 325 (64.1%) patients was betamethasone dipropionate, 0.05%, in optimized vehicle ointment, a superpotent TCS, followed by methylprednisolone acetonate ointment, 0.1%, a midpotency TCS that was used in 156 (30.8%) patients. Clobetasol propionate ointment, 0.05%, an ultrapotent TCS, was used in 17 (3.4%) patients and hydrocortisone ointment, 1%, was used in 9 (1.8%) patients (Table 1). The decision regarding which TCS with which to initiate treatment was made according to the degree of severity of hyperkeratosis (Figure 1): very severe (4+), clobetasol propionate ointment; moderate to severe (2+ or 3+), betamethasone dipropionate ointment; and mild (1+), methylprednisolone acetonate ointment. A small number (9 [1.8%]) of patients had very mild “burnt out” disease and were treated with hydrocortisone ointment.

Once disease and symptom suppression had been achieved, long-term preventive management was initiated. A gradual reduction of TCS potency, titrated to the clinical response, was attempted in all patients. Treatment was outcome based, with the target being as close as possible to normal skin color and texture (Figure 2). As long as there were no adverse effects, this treatment was maintained. If atrophy or corticosteroid dermatitis developed, the potency of the TCS was reduced. If hyperkeratosis returned, the potency of the TCS was increased. Patients used the treatment at least 3 times per week. For patients with very severe disease, a potent to superpotent TCS was used daily.

Follow-up was conducted every 3 to 6 months for the first 2 years and then at least yearly to ensure that treatment was...
adequate to maintain the target outcome and to encourage compliance. Patients were not instructed to treat themselves as required, and it was emphasized that treatment should be continued preventively even when asymptomatic. Patients were informed that possible outcomes of poor compliance included cancer and scarring.

At follow-up visits, the following features were recorded: self-reported compliance regarding frequency and potency of the TCS used, subjective symptomatic response to treatment, objective clinical response to treatment, maintenance therapy required, progress or onset of adhesions or scarring, adverse effects, and development of squamous cell carcinoma or vulvar intraepithelial neoplasia.

At the completion of the 7-year observational period, all data collected from the 507 patients were entered into an Excel 2011 spreadsheet (Microsoft Corporation). Statistical analysis was conducted using SPSS, version 20.0 (SPSS Inc). Descriptive statistics are presented, and Fisher exact tests, Pearson χ², or independent, unpaired, 2-tailed t tests were conducted, as appropriate, to compare outcome rates between those who were compliant and partially compliant.

### Results

#### Demographic Data

The mean age at presentation was 55.4 years (range, 18-86 years), and the mean duration of symptoms before presentation was 5.0 years (range, 0.1-40.0 years). A total of 158 (31.2%) patients were premenopausal, 307 (60.6%) were postmenopausal and not using hormone therapy, and 42 (8.3%) were postmenopausal and using either topical or systemic hormone therapy. The mean duration of follow-up for all patients was 4.7 years (range, 2.0-6.8 years). Most patients (476 [93.9%]) were white. The mean age at presentation of patients who developed VSN was 57.8 years (range, 29-76 years), with a mean duration of symptoms before presentation of 9.4 years (range, 1-30 years).

#### Clinical Features

At presentation, most patients (491 [96.8%]) were symptomatic and most (311 of 414 sexually active patients [75.1%]) experienced dyspareunia. Nearly all women (471 [92.9%]) expe-
Table 1. Suggested Guidelines for Initial Management

<table>
<thead>
<tr>
<th>Degree of Hyperkeratosis</th>
<th>Initial Management</th>
<th>Frequency</th>
</tr>
</thead>
<tbody>
<tr>
<td>0+ (Burnt out; very mild)</td>
<td>Mild-potent TCS (in our study, hydrocortisone ointment, 1.0%)</td>
<td>Once daily</td>
</tr>
<tr>
<td>1+ (Mild)</td>
<td>Midpotent TCS (in our study, methylprednisolone aceponate ointment, 0.1%)</td>
<td>Once daily</td>
</tr>
<tr>
<td>2+ (Moderate)</td>
<td>Superpotent TCS (in our study, betamethasone dipropionate, 0.05%, in optimized vehicle ointment)</td>
<td>Once daily</td>
</tr>
<tr>
<td>3+ (Severe)</td>
<td>Superpotent TCS (in our study, betamethasone dipropionate, 0.05%, in optimized vehicle ointment)</td>
<td>Twice daily</td>
</tr>
<tr>
<td>4+ (Very severe)</td>
<td>Ultrapotent TCS (in our study, clobetasol propionate ointment, 0.05%)</td>
<td>Twice daily</td>
</tr>
</tbody>
</table>

Abbreviation: TCS, topical corticosteroid.

Figure 2. Suggested Long-term Topical Corticosteroid (TCS) Management of Vulvar Lichen Sclerosis

Initial TCS treatment for 6 weeks based on degree of hyperkeratosis
- Very severe (4+), ultrapotent TCS
- Moderate to severe (2-3+), superpotent TCS
- Mild (1+), midpotent TCS
- Burnt out (0+), mild-potent TCS

Normal skin color and texture
- Skin color and texture not normalized
- Reduce TCS strength
- Continue initial TCS treatment
- Reassess every 3 months until skin color and texture are normal and TCS strength and regimen are stable
- Review every 6-12 months long term

Treatment guideline based on the degree of hyperkeratosis of vulvar lichen sclerosus.

Table 2. Clinical Characteristics of Partially Compliant Patients Who Developed VSN

<table>
<thead>
<tr>
<th>Patient No./Age, Decade</th>
<th>Prior Documented Duration of Disease, mo</th>
<th>Stage of Carcinoma</th>
<th>Dyspareunia</th>
<th>Itching Distribution</th>
<th>Degree of Hyperkeratosis</th>
<th>Fissuring</th>
<th>Structural Change</th>
<th>Labial Fusion</th>
<th>Follow-up Duration at Time of VSN Diagnosis, mo</th>
<th>Symptom Resolution</th>
</tr>
</thead>
<tbody>
<tr>
<td>1/40s</td>
<td>36</td>
<td>Differentiated VIN</td>
<td>No</td>
<td>Yes</td>
<td>2+ (Moderate)</td>
<td>No</td>
<td>None</td>
<td>18</td>
<td>Yes</td>
<td></td>
</tr>
<tr>
<td>2/70s</td>
<td>36</td>
<td>Invasive SCC</td>
<td>Not sexually active</td>
<td>Yes</td>
<td>Figure of 8</td>
<td>3+ (Severe)</td>
<td>No</td>
<td>Anterior and posterior</td>
<td>18</td>
<td>No</td>
</tr>
<tr>
<td>3/60s</td>
<td>120</td>
<td>Invasive SCC</td>
<td>Yes</td>
<td>Yes</td>
<td>Figure of 8</td>
<td>3+ (Severe)</td>
<td>No</td>
<td>Anterior and posterior</td>
<td>68</td>
<td>Yes</td>
</tr>
<tr>
<td>4/70s</td>
<td>48</td>
<td>Differentiated VIN</td>
<td>Yes</td>
<td>Yes</td>
<td>Figure of 8</td>
<td>2+ (Moderate)</td>
<td>No</td>
<td>None</td>
<td>36</td>
<td>No</td>
</tr>
<tr>
<td>5/50s</td>
<td>180</td>
<td>Differentiated VIN</td>
<td>Yes</td>
<td>Yes</td>
<td>Figure of 8</td>
<td>1+ (Mild)</td>
<td>No</td>
<td>None</td>
<td>24</td>
<td>No</td>
</tr>
<tr>
<td>6/20s</td>
<td>12</td>
<td>Differentiated VIN</td>
<td>Yes</td>
<td>No</td>
<td>Figure of 8</td>
<td>1+ (Mild)</td>
<td>No</td>
<td>None</td>
<td>18</td>
<td>No</td>
</tr>
<tr>
<td>7/60s</td>
<td>360</td>
<td>Invasive SCC</td>
<td>Yes</td>
<td>Yes</td>
<td>Figure of 8</td>
<td>3+ (Severe)</td>
<td>Yes</td>
<td>None</td>
<td>36</td>
<td>Yes</td>
</tr>
</tbody>
</table>

Abbreviations: SCC, invasive squamous cell carcinoma; VIN, vulvar intraepithelial neoplasia; VSN, vulvar squamous neoplasia.
who had an outcome of normal skin color and texture, the mean time to reach this outcome was 4.9 months (compliant, 4.8 months; partially compliant, 5.3 months). A further 26 (7.3%) compliant and 20 (13.3%) partially compliant patients had significant improvement in skin color and texture without achieving complete normality. Only a small percentage (5 [1.4%]) of compliant patients were resistant to TCSs.

A total of 357 (70.4%) patients self-reported as compliant with our instructions. The partially compliant group (150 [29.6%]) was characterized by patients who applied treatment only when symptomatic, forgot to use their treatment, used it less often than recommended, or refused to use any TCS that was more potent than hydrocortisone ointment, 1.0%, because of a fear of adverse effects. Most partially compliant patients used significantly less TCS than compliant patients, with some using none yet still attending follow-up visits.

During the study, VSN occurred in 0 compliant patients and 7 partially compliant patients (P < .001). This outcome presented as development of treatment-resistant hyperkeratotic plaques and nodules and was confirmed by biopsy before referral for surgical excision. Of the 7 women who developed VSN, there was no defining clinical feature to differentiate them from those who did not other than poor compliance with treatment (Table 2).

During regular follow-up, 333 (93.3%) compliant patients became consistently free of itching and pain compared with 87 (58.0%) partially compliant patients (P < .001). Of those who were sexually active and presented with dyspareunia (n = 311), there were 207 compliant and 104 partially compliant patients. Of the compliant patients, 194 (93.7%) had complete or near-complete resolution compared with 68 (65.4%) partially compliant patients (P < .001). Of the 311 sexually active patients with dyspareunia, 102 were premenopausal and 209 were postmenopausal. Resolution of dyspareunia rates were similar between the premenopausal and postmenopausal groups. Progress of adhesions and scarring occurred in 12 (3.4%) compliant patients and 60 (40.0%) partially compliant patients (P < .001). Cutaneous atrophy was observed in 4 (1.1%) compliant and 3 (2.0%) partially compliant patients. In all patients, this outcome was reversed after a reduction of TCS potency. The only other adverse effect encountered was corticosteroid dermatitis, which presented as redness and symptoms of burning or soreness observed in 8 (2.2%) compliant patients and 6 (4.0%) partially compliant patients (Table 3); this outcome is analogous to periorificial dermatitis of the face. This outcome was twice as common as atrophy and reversed after reduction of TCS potency.

### Discussion

This study describes a cohort of 507 women with biopsy-proved VLS who were prospectively followed up for a minimum of 2 years and for whom standardized data were collected. Objective remission was maintained with regular preventive TCS treatment that was individualized to achieve a clinical target outcome rather than prescribed according to a standard treatment regimen. Although superpotent to potent TCSs were used in most patients to achieve remission, we found that a mild to moderate potency of TCS, if used regularly and preventively, was effective in maintaining objective and symptomatic remission in most patients with minimal adverse effects.

The original study by Dalziel et al, which demonstrated the efficacy of topical clobetasol propionate ointment, 0.05%, in inducing remission in VLS, has been validated by others. However, guidelines for long-term maintenance of VLS remission are based on a few studies that have not been adequately powered to reach firm conclusions regarding best practice.

A prospective study by Renaud-Vilmer et al had a mean follow-up of 4.7 years, but the sample size (83 patients) made it difficult to draw statistically significant conclusions. Nonetheless, the study suggested that there may be a protective effect from continued treatment and observed that carcinoma developed only in women who were not treated or who were irregularly treated. In this study, it was noted that withdrawal of treatment resulted in a high relapse rate of 85%.

It is a basic premise in dermatological practice that chronic inflammatory skin diseases frequently require ongoing suppression. This fact, in addition to the high rates of relapse in patients who stop using TCSs after achieving remission, has been the basis for our recommendation to continue maintenance treatment even when patients are asymptomatic.

To our knowledge, the largest previous long-term study of TCS-treated women with VLS to date was a retrospective cohort study of 253 women with a mean follow-up of 5.5 years. It showed that topical clobetasol propionate ointment completely reversed cutaneous changes in 23% of patients and provided symptomatic relief. Most patients were given a TCS for "intermittent maintenance self-treatment" after the initial

### Table 3. Compliance and Treatment Outcomes in Compliant vs Partially Compliant Patients

<table>
<thead>
<tr>
<th>Outcome</th>
<th>No./Total No. (%)</th>
<th>Compliant</th>
<th>Partially Compliant</th>
<th>P Value</th>
</tr>
</thead>
<tbody>
<tr>
<td>SCC or VIN</td>
<td>0/357 (0.0)</td>
<td>0/150 (0.0)</td>
<td></td>
<td>.001</td>
</tr>
<tr>
<td>Progression of adhesions or scarring</td>
<td>12/357 (3.4)</td>
<td>60/150 (40.0)</td>
<td></td>
<td>&lt;.001</td>
</tr>
<tr>
<td>Symptom resolution</td>
<td>333/357 (93.3)</td>
<td>87/150 (58.0)</td>
<td></td>
<td>&lt;.001</td>
</tr>
<tr>
<td>Improvement in dyspareunia in sexually active women</td>
<td>194/207 (93.7)</td>
<td>68/104 (65.4)</td>
<td></td>
<td>&lt;.001</td>
</tr>
<tr>
<td>Surgical division of adhesions</td>
<td>10/357 (2.8)</td>
<td>7/150 (4.7)</td>
<td></td>
<td>.29</td>
</tr>
<tr>
<td>Adverse effects</td>
<td></td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>Corticosteroid dermatitis</td>
<td>8/357 (2.2)</td>
<td>6/150 (4.0)</td>
<td></td>
<td>.37</td>
</tr>
<tr>
<td>Atrophy</td>
<td>4/357 (1.1)</td>
<td>3/150 (2.0)</td>
<td></td>
<td>.43</td>
</tr>
</tbody>
</table>

Abbreviations: SCC, invasive squamous cell carcinoma; VIN, vulvar intraepithelial neoplasia.
treatment period. Of this cohort, 10 patients developed a malignant neoplasm (4%). The authors acknowledged that their study was not adequately powered to determine whether treatment could decrease the risk of malignant transformation but speculated that scarring might be prevented by early treatment. Our research supports this finding, with the proviso that treatment should then continue on a preventive basis.

A Cochrane review regarding treatment of VLS stated that, to determine whether treatment can reduce the risk of developing malignant neoplasms, a randomized clinical trial of at least 984 treated and untreated patients of adequate duration would be required. Given that the efficacy of TCSs in VLS is proven and the outcome of lack of treatment includes the possibility of scarring and carcinoma, we believe it would not be ethical to conduct such a study. However, the question of whether treatment can change the course of the disease is critical to management and advice to patients long term.

This current study shows a statistically significant difference in disease outcomes in patients who used preventive treatment regularly when asymptomatic and those who, despite strong advice to the contrary, chose not to. This outcome included development of scarring and malignant neoplasms as well as objective disease suppression. For most patients, the objective clinical response correlated with symptom control and resolution of dyspareunia, which was achieved in more than 90% of compliant patients.

In Australia, clobetasol propionate ointment only became available during the past decade; previously, it was necessary to use other alternatives. This lack of availability enabled us to discover that other types of TCSs were as effective in all but the most severe cases of VLS.

In 2010, the British Association of Dermatology published a guideline for management of VLS. This guideline and the Cochrane review stated that there have been no comparative clinical trials with which to make recommendations on the most suitable TCS for treating VLS. In fact, most original studies have used clobetasol propionate ointment, with only 3 short-term studies using mometasone furoate, 0.1%. Our study, however, suggests that there is no single TCS regimen that should be applied to VLS and that the regimen itself is less important than the response to treatment. We suggest that physicians who treat VLS familiarize themselves with the relative potencies of TCSs available in their country and select the most appropriate based on the severity of hyperkeratosis. Based on our experience, the broad recommendations for long-term maintenance treatment are shown in Figure 2. To ensure that patients’ treatment is at a potency that matches their disease severity, regular follow-up is required. Our principle of management is that as long as the skin color and texture remain normal, with no sign of atrophy or erythema, there is no need to reduce TCS potency; however, if hyperkeratosis returns, potency should be increased (Figure 2).

There continue to be widespread concerns about atrophy with long-term use of regular TCSs. However, in this study, with supervised use of TCSs at a potency titrated to disease severity, this outcome was not an issue. Calcineurin inhibitors were not used by our group because they lack long-term safety data. Furthermore, our data support an excellent safety profile of long-term TCS use that obviates the need for calcineurin inhibitors.

Most important, carcinoma did not develop in any of the 357 compliant patients compared with 7 of 150 (4.7%) partially compliant patients (P < .001) during follow-up. The risk of cancer in the partially compliant group is close to the risk of 5% to 6% of untreated women in previous studies. This finding suggests that as-required treatment that controls symptoms only may not reduce the risk of developing cancer and supports a previous study suggesting that compliance with treatment could potentially reduce the incidence of cancer. It seems that, in compliant patients who maintain normal skin color and texture, the risk of VSN is greatly reduced.

We consider a 5% risk of cancer to be significant enough to recommend long-term follow-up by a medical professional for all patients regardless of disease or symptom severity. Our data demonstrate that the development of VSN is unpredictable and is not limited to patients with severe disease. The only predictor we have identified is a lack of consistent preventive treatment. Vulvar lichen sclerosis does not affect large numbers of women, and we contend that it would not be a burden to the health system to monitor these patients regularly to ensure ongoing control and compliance with topical TCS therapy, which we have shown to be safe, high in treatment efficacy, and low in cost. When considering the cost burden of long-term follow-up, a comparison should be made between the cost of following up patients to ensure they do not develop cancer and the cost of treating cancer in patients with poorly controlled disease.

Our data suggest that symptom control is not an adequate end point for treatment. Many partially compliant patients were largely asymptomatic in our study, using treatment only when itchy. Common among this group was a persistent belief that being asymptomatic rendered treatment unnecessary, yet this behavior predisposed them to continued disease activity, scarring, and malignant neoplasms. Indeed, 3 of the 7 women who developed VSN reported being asymptomatic (Table 2).

Our study did not encompass quality-of-life data on patients with scarring, but it did indicate that scarring is preventable. Once established, it is irreversible and likely to have significant effects. Although scarring was more likely in patients with severe hyperkeratosis, almost half of patients with mild disease had developed scarring on presentation. Thus, the pathogenic mechanisms of epidermal and dermal disease in VLS may be independent of each other; despite this fact, both respond to TCS treatment, which arrested the progression of scarring in most compliant patients.

The limitations of our study are that it was not randomized owing to ethical concerns and that compliance rates relied on self-reporting. Other factors that may have introduced bias are unlikely because the compliant and partially compliant groups had similar demographic characteristics.

Conclusions

This longitudinal prospective cohort study observed treatment outcomes of biopsy-proved VLS that was managed with regular, long-term, individualized TCS regimens. Treatment
regimens matched the potency and duration of TCS treatment to the objective disease severity, and TCSs were used preventively and regularly once patients had achieved clinical remission. The long-term target outcome was preservation of normal skin color and texture rather than symptom control alone. This regimen produced good results compared with earlier studies that used clobetasol propionate ointment as needed. In comparing compliant and partially compliant patients, we have demonstrated that, in this cohort, preventive long-term treatment not only improved function and relieved symptoms but also reduced development or progression of scarring and eliminated the risk of cancer. We did not encounter significant adverse effects; in particular, there were few cases of reversible cutaneous atrophy. Our data lead us to recommend this treatment strategy based on objective disease suppression and symptom control, with regular follow-up to optimize compliance, adjust treatment potency, and monitor for complications.

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Study concept and design: Bradford, Fischer.

Acquisition, analysis, or interpretation of data: All authors.


Critical revision of the manuscript for important intellectual content: All authors.

Statistical analysis: Lee, Bradford.

Administrative, technical, or material support: All authors.

Study supervision: Fischer.

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