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Diagnosis and Management of Vulvar Skin Disorders

Vulvar skin disorders include a variety of inflammatory conditions of the vulva that also may affect the extragenital area. Pruritus and pain are two of the most common presenting symptoms in vulvar clinics (1). Vulvovaginal symptoms often are chronic and can adversely affect sexual function and sense of well-being. The purpose of this Practice Bulletin is to provide updated diagnostic and management recommendations for the most common vulvar skin conditions associated with inflammation: contact dermatitis, lichen simplex chronicus, lichen sclerosus, and lichen planus. Other vulvovaginal disorders such as vaginitis, vulvar low-grade squamous intraepithelial lesions and vulvar high-grade squamous intraepithelial lesions (previously termed vulvar intraepithelial neoplasia), genitourinary syndrome of menopause (vulvovaginal atrophy), and vulvar pain (vulvodynia) are addressed in other documents from the American College of Obstetricians and Gynecologists (2–6).

Background

Contact Dermatitis

Contact dermatitis of the vulva is a nonscarring inflammatory reaction characterized by chronic itching or burning, and can present at any age (7). Approximately half of individuals who present with chronic vulvovaginal pruritus have contact dermatitis (8). Irritant contact dermatitis occurs when the skin is in contact with moisture, sweat, and urine. Other irritants include cleansers, fragrances, lubricants, and other topical products that may exacerbate symptoms. Allergic contact dermatitis is defined as a type IV delayed hypersensitivity reaction, in which an individual develops an allergy to a product applied topically. Products may include fragrance, antibiotics, local anesthetics, and components of some topical treatments (9). Many seemingly innocuous activities such as bathing, the use of sanitary or incontinence pads, or the use of feminine hygiene products (Box 1) can initiate this inflammatory process (10).

Lichen Simplex Chronicus

Vulvar lichen simplex chronicus is a chronic, nonscarring inflammatory disease of the vulvar skin characterized by intense and unrelenting itching and scratching. The most commonly reported symptom of lichen simplex chronicus is chronic or intermittent pruritus accompanied by vigorous scratching or rubbing. Pruritus occurs most frequently in the evening or during the night, leading to sleep disturbances (11). In vulvar specialty clinics, lichen simplex chronicus accounts for 10–35% of patients evaluated (11). It is both one of the most common primary causes of vulvar pruritus and a secondary complication of any pruritic vulvar condition (12), including environmental factors (eg, heat, excessive sweating, and irritation from clothing or topically applied products) and dermatologic diseases (eg, candidiasis, contact dermatitis, and lichen sclerosus) (13). Lichen simplex chronicus occurs primarily in middle to late adult life. Up to 75% of patients with lichen simplex chronicus have a personal or an immediate family history of seasonal allergies, asthma, or childhood eczema (11), which supports the



Box 1. Common Vulvar Irritants and Allergens

- Adult or baby wipes
- Antiseptics (eg, povidone iodine, hexachlorophene)
- Body fluids (eg, semen or saliva)
- Colored or scented toilet paper
- Condoms (lubricant or spermicide containing)
- Contraceptive creams, jellies, foams, nonoxynol-9
- Personal lubricants
- Dyes (eg, chemically treated clothing, hygiene products, perfume)
- Synthetic nylon underwear and tight clothing or undergarments
- Emollients (eg, lanolin, jojoba oil, glycerin)
- Laundry detergents, fabric softeners, and dryer sheets
- Shaving cream and shaving (in general)
- Rubber products (including latex)
- Sanitary products, including tampons and pads
- Soaps, bubble bath, bath salts, shampoos, and conditioners
- Tea tree oil
- Topical agents:
 - anesthetics (eg, benzocaine, lidocaine, dibucaine)
 - antibacterials (eg, neomycin, bacitracin, polymyxin)
 - antimycotics (eg, imidazoles, nystatin)
 - corticosteroids
 - medications, including trichloroacetic acid, 5-fluorouracil, podofilox, or podophyllin
- Vaginal hygiene products, including vaginal sprays, washes, douches, perfumes, and deodorants

idea that lichen simplex chronicus is a localized variant of atopic dermatitis.

Lichen Sclerosus

Lichen sclerosus is a chronic scarring dermatologic disorder that most commonly affects the anogenital skin of postmenopausal women and prepubertal girls (14–16). Prevalence estimates range from 1 in 32 among nursing home residents, to almost 1 in 60 among patients seen in a general gynecologic practice, to 1 in 900 among patients in a pediatric vulvar clinic (17–19). The exact prevalence of lichen sclerosus is difficult to determine because the condition often is asymptomatic and goes unrecognized by physicians (20, 21). Although the exact etiology of this condition is unclear, autoimmune processes and genetic factors are believed to play a role in the pathogenesis (20, 22, 23).

Lichen Planus

Lichen planus is a scarring inflammatory disorder of the skin, oral mucosa, and vulvovaginal area (24). Although the exact etiology is unknown, lichen planus is believed to be associated with dysfunction of cell-mediated immunity (9), and concomitant autoimmune disorders have been seen in approximately one third of patients with lichen planus (25). Its estimated prevalence in the general population is less than 1% (24), and it most commonly affects perimenopausal and menopausal women (20, 26). Up to 70% of individuals with erosive vulvovaginal lichen planus will have oral involvement (7, 27–29).

Differential Diagnosis

In assessing vulvar pruritus and pain, the common causes of vulvovaginitis (candidiasis, bacterial vaginosis, and trichomoniasis), particularly in their more complicated forms, should be included in the differential diagnosis (Box 2). For example, non-*albicans* candida infections typically present with burning rather than itching, and minimal evidence of inflammation. Genital HPV infection is common among young women and patients who are immunosuppressed, and it is associated with various vulvar epithelial disorders, including genital warts, vulvar low-grade squamous intraepithelial lesions, vulvar high-grade squamous intraepithelial lesions, and some types of vulvar carcinoma (3). Paget disease of the vulva is a rare form of intraepithelial neoplasia characterized by pale vacuolated cells, and it accounts for less than 2% of vulvar neoplasms (30). Most patients who present with Paget disease have a preinvasive disease state; however, at times, an underlying adenocarcinoma is present (30). Chronic vulvar pruritus also may be caused by systemic diseases with vulvar manifestations such as Crohn disease (31, 32) and hidradenitis suppurativa (33). Genitourinary syndrome of menopause (which includes vulvovaginal atrophy) can cause vulvar pain and vulvar pruritus (4, 5, 34). Vulvodynia also should be included in the differential diagnosis, and is characterized by discomfort and pain that occur in the absence of visible findings or a specific, clinically identifiable cause (6, 35).

Clinical Considerations and Recommendations

► What is the initial approach to patients with vulvovaginal symptoms?

The initial evaluation of patients with vulvovaginal symptoms should include a comprehensive medical history, a physical examination, and evaluation of abnormal vaginal discharge if an infectious etiology is suspected (7).



Box 2. Conditions Commonly Associated With Vulvar Pruritus and Vulvar Pain

Vulvar Pruritus	Vulvar Pain
<p>Infections</p> <ul style="list-style-type: none"> • Tinea cruris • Trichomoniasis • Vulvovaginal candidiasis • Molluscum contagiosum • Infestations, including scabies and pediculosis <p>Dermatoses</p> <ul style="list-style-type: none"> • Atopic and contact dermatitis • Lichen sclerosus, lichen planus, lichen simplex chronicus • Psoriasis <p>Neoplasia</p> <ul style="list-style-type: none"> • Paget disease • Vulvar LSIL or HSIL • Vulvar cancer <p>Vulvar manifestations of systemic disease</p> <ul style="list-style-type: none"> • Crohn disease • Hidradenitis suppurativa <p>Hormonal deficiencies</p> <ul style="list-style-type: none"> • Genitourinary syndrome of menopause (vulvovaginal atrophy) 	<p>Infections</p> <ul style="list-style-type: none"> • Recurrent vulvovaginal candidiasis • Herpes <p>Dermatoses</p> <ul style="list-style-type: none"> • Lichen sclerosus • Lichen planus • Immunobullous disorders <p>Neoplasia</p> <ul style="list-style-type: none"> • Paget disease • Vulvar LSIL or HSIL • Vulvar cancer <p>Neurologic</p> <ul style="list-style-type: none"> • Postherpetic neuralgia • Nerve compression or injury • Neuroma <p>Trauma</p> <ul style="list-style-type: none"> • Female genital cutting or female genital mutilation • Obstetric complications <p>Iatrogenic</p> <ul style="list-style-type: none"> • Postoperative • Chemotherapy • Radiation <p>Hormonal deficiencies</p> <ul style="list-style-type: none"> • Genitourinary syndrome of menopause (vulvovaginal atrophy) • Lactational amenorrhea

Abbreviations: LSIL, low-grade squamous intraepithelial lesion; HSIL, high-grade squamous intraepithelial lesion. Adapted from Bornstein J, Goldstein AT, Stockdale CK, Bergeron S, Pukall C, Zolnoun D, et al. 2015 ISSVD, ISSWSH, and IPPS consensus terminology and classification of persistent vulvar pain and vulvodynia. Consensus Vulvar Pain Terminology Committee of the International Society for the Study of Vulvovaginal Disease (ISSVD), the International Society for the Study of Women's Sexual Health (ISSWSH), and the International Pelvic Pain Society (IPPS). *J Low Genit Tract Dis* 2016;20:126–30.

The medical history should include questions about the onset, duration, location, and nature of vulvar symptoms; the relationship of symptoms to the patient's menstrual cycle; and any possible precipitating or known risk factors. In evaluating vulvar pruritus, separately grouping patients with acute and chronic symptoms can be helpful to guide the differential diagnosis (Fig.1). Patients who present with pain first should be evaluated to rule out an underlying clinically identifiable disorder, including inflammatory conditions, neoplasias, infections, or neurologic disorders. When other causes are ruled out, the diagnosis of vulvodynia can be made, and attention can be directed to explaining the nature

of the pain disorder (6). It should be noted that patients may have both a specific disorder (eg, lichen sclerosus) and vulvar pain (7, 35).

Vulvar skin disorders can cause changes in the color, texture, and architecture of the vulvar anatomy. Clinicians should be familiar with normal vulvar anatomy and variations, and should examine the entire vulva from clitoris to below the anus to confirm there are no changes in architecture. For example, the clitoral hood should be mobile and easily retracted. The entire vulva should be examined for changes in color and texture and to detect the presence of fissures, excoriations, erosions, ulcers, and other lesions. A speculum examination, if tolerated,



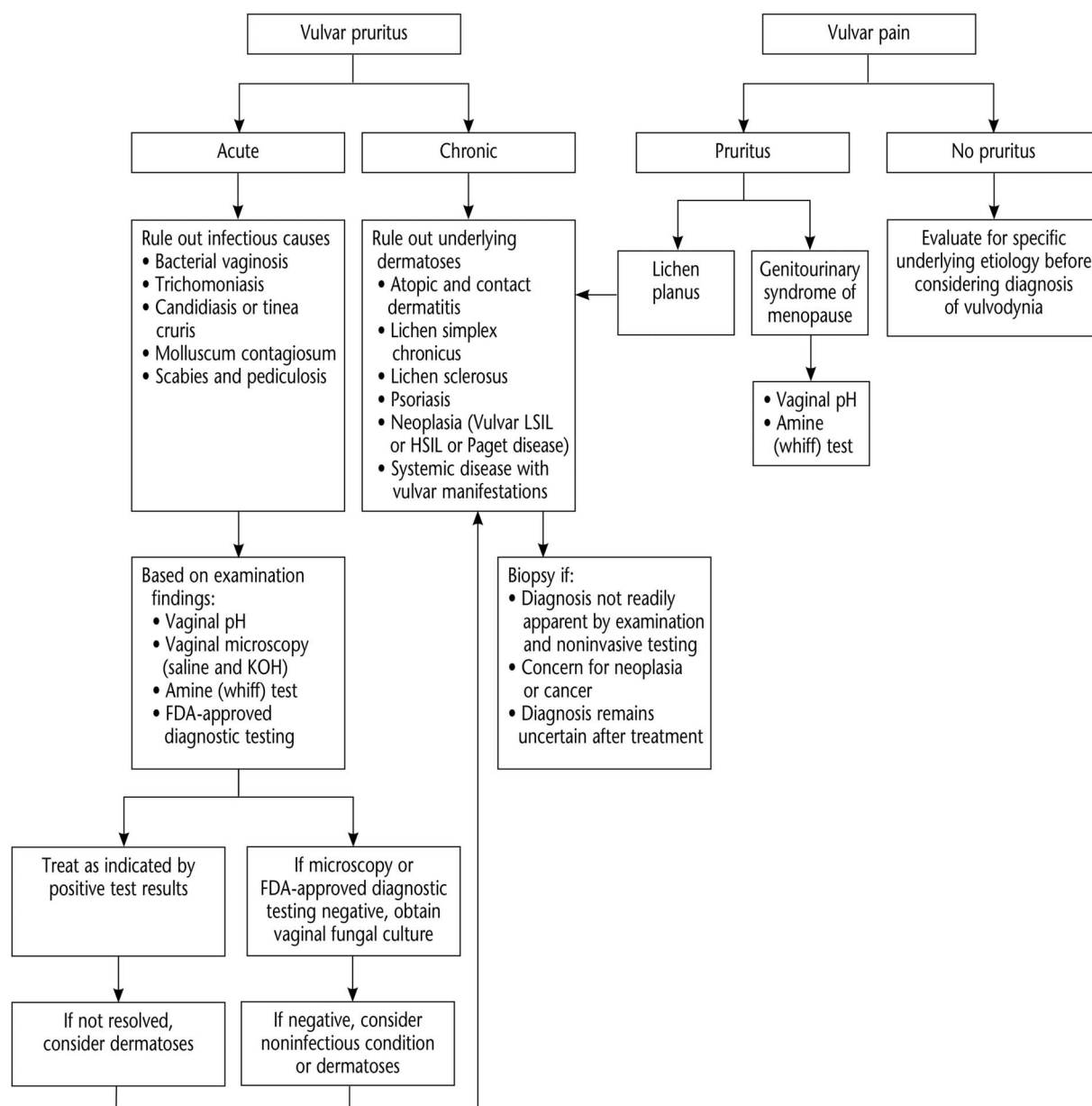


Figure 1. Vulvar dermatoses diagnostic algorithm. Abbreviations: FDA, U.S. Food and Drug Administration; HSIL, high-grade squamous intraepithelial lesion; KOH, potassium hydroxide; LSIL, low-grade squamous intraepithelial lesion. Modified from Stockdale CK, Boardman L. Diagnosis and treatment of vulvar dermatoses. *Obstet Gynecol* 2018;131:371–86.

should be performed to look for erythema, erosion, ulceration, synechiae, and discharge (20). In addition to a complete medical history and physical examination of the vulva and vagina, an evaluation of abnormal vaginal discharge (pH testing, a potassium hydroxide “whiff test,” microscopy, U.S. Food and Drug Administration [FDA]-approved diagnostic tests) is recommended for the initial evaluation of patients with vaginitis symptoms (2, 36). Vaginal fungal culture and testing for suspected sexually transmitted infections should be performed as

indicated by Centers for Disease Control and Prevention guidelines (36).

► *When and how should a vulvar biopsy be performed?*

Vulvar biopsy is recommended for visible lesions in any of the following circumstances (3, 36):

- lesions are atypical (eg, new pigmentation, indurated, affixed to underlying tissue, bleeding, or ulcerated)



- there is concern for malignancy
- lesions in a patient who is immunocompromised (including those infected with human immunodeficiency virus [HIV])
- the diagnosis is uncertain
- lesions do not respond to standard therapy
- the disease worsens during therapy

Changes on the vulva often are subtle and can be overlooked. Findings such as thickening, pebbling, hypopigmentation, or thinning of the epithelium indicate a possible dermatologic process, and biopsy will aid in diagnosis and management. Biopsy is indicated to rule out malignancy for visible lesions with atypical or changing vascular patterns; stable lesions that rapidly change in color, have irregular borders, or increase in size; or unresolving lesions (3). Although information regarding the evaluation of vulvar lesions in patients with HIV and other immunocompromised conditions is limited, the threshold for biopsy should be low in these patients given that individuals infected with HIV have higher rates of vaginal, vulvar, and perianal neoplasia compared with the general population (3, 37–39).

The biopsy can be performed as a punch, shave, or excisional biopsy, depending on the characteristics of the lesion, physician preference, and equipment availability (40). Pigmented lesions that raise the possibility of a melanoma should be biopsied using a full thickness technique (eg, elliptical or punch excision) including the most abnormal or atypical part of the lesion (40, 41). For sclerotic lesions or ulcerative areas, it is preferable to biopsy the edge of the lesion including a border with normal skin; when sampling hyperpigmented areas, biopsy of the thickest region is recommended (40, 42). Multicentric lesions or lesions with multiple morphologies may warrant multiple biopsies. Small lesions often can be completely excised, and lesions involving the submucosal or subcutaneous tissue can be adequately sampled. Lesions that are raised, superficial, or have an irregular surface—such as bullous lesions or lesions associated with lichen sclerosus or lichen planus—can be sampled with a shave technique or a variation called the snip biopsy, which uses curved iris scissors in place of a number 15 scalpel blade (40).

► *How is contact dermatitis diagnosed and treated?*

Diagnosis

Clinical features of contact dermatitis range from mild erythema, swelling, and scaling to marked erythema, excoriations, fissures, erosions, and ulcers (9, 43). Diagnosis of contact dermatitis can be made based on history and physical examination findings (7, 9, 26, 43–45).

Treatment

Recommended treatment of contact dermatitis includes patient counseling regarding vulvar care and removal of vulvar irritants and allergens (Box 1 and Box 3) and symptom management with a topical corticosteroid ointment and an oral antipruritic medication as needed (7, 9). The severity and chronicity of contact dermatitis will determine the need for and the potency of the topical corticosteroid chosen for treatment (Box 4). A topical corticosteroid ointment should initially be applied once or twice daily (depending on condition severity, corticosteroid potency, and patient preference) until the lesions heal (7, 9). A topical corticosteroid ointment is preferable to other formulations (cream, gel, lotion), which often contain alcohol or preservatives that can cause or exacerbate irritation (7, 10, 20). Pruritus and associated scratching and rubbing can be reduced by using a combination of nonsedating antihistamines (eg, cetirizine, loratadine) for daytime use and sedating antihistamines (eg, hydroxyzine) or a tricyclic antidepressant with antihistamine properties (eg, doxepin) at night if needed (9, 26, 44). Topical antipruritic medications should be avoided because they can cause allergic contact dermatitis (44).

► *How is lichen simplex chronicus diagnosed and treated?*

Diagnosis

Clinically, lichen simplex chronicus appears as one or more erythematous, scaling, or lichenified plaques. Various degrees of excoriation are often visible. In

Box 3. Patient Counseling for Prevention and Control of Vulvar Disorders

- Avoid vulvar irritants and allergens.
- Use mild soaps, but avoid use on the vulva.
- Cleanse the vulva with water only.
- Gently pat the vulva dry after bathing.
- Apply preservative-free, unscented or fragrance-free emollient to hold moisture in the skin and improve barrier function.
- Use PeriCare bottle to rinse after urination.
- Use 100% cotton, unscented or fragrance-free menstrual pads.
- Use adequate lubrication for intercourse (unscented or fragrance-free silicone-based lubricants recommended).

Modified from Stockdale CK, Boardman L. Diagnosis and treatment of vulvar dermatoses. *Obstet Gynecol* 2018;131:371–86.



Box 4. Common Topical Corticosteroid Ointments for Vulvar Skin Disorders*

High Potency

- Betamethasone dipropionate 0.05%
- Clobetasol propionate 0.05%
- Halobetasol propionate 0.05%
- Fluocinonide 0.05%
- Triamcinolone acetonide 0.5%

Medium Potency

- Desoximetasone 0.05%
- Hydrocortisone butyrate 0.1%
- Hydrocortisone valerate 0.2%
- Mometasone furoate 0.1%
- Triamcinolone acetonide 0.025%, 0.1%

Low Potency

- Desonide 0.05%
- Hydrocortisone 1%, 2.5%

*A topical steroid ointment is preferable to other formulations (cream, gel, lotion), which often contain alcohol or preservatives that can cause or exacerbate irritation.

Data from Stockdale CK, Boardman L. Diagnosis and treatment of vulvar dermatoses. *Obstet Gynecol* 2018;131:371–86; Thorstensen KA, Birenbaum DL. Recognition and management of vulvar dermatologic conditions: lichen sclerosus, lichen planus, and lichen simplex chronicus. *J Midwifery Womens Health* 2012;57:260–75; and Ference JD, Last AR. Choosing topical corticosteroids. *Am Fam Physician* 2009;79:135–40.

long-standing disease, the skin appears thickened, leathery, or “bark-like.” Poorly demarcated, thick plaques with scale and areas of hyperpigmentation and hypopigmentation also may be present (26). Erosions and ulcers also can develop, most commonly from chronic scratching.

The diagnosis of lichen simplex chronicus can be made clinically, and biopsy is rarely necessary (see *When and how should a vulvar biopsy be performed?* earlier in this section) (7, 9, 26). A vaginal fungal culture should be considered to determine the presence of underlying vulvovaginal candidiasis (7, 20).

Treatment

Recommended treatment of lichen simplex chronicus includes patient education on how to stop the “itch-scratch cycle,” information regarding vulvar care and hygiene (Box 1 and Box 3), a medium-potency or

high-potency topical corticosteroid ointment (Box 4), and oral antipruritic medication as needed (7, 9, 26). As with contact dermatitis, the general approach to therapy is multimodal and is focused on vulvar self-care (Box 3), control of itching and scratching, and treating inflammation with topical corticosteroids (see *How is contact dermatitis diagnosed and treated?* earlier in this section). In patients with daytime itching or those who cannot tolerate medications with antihistaminic (eg, hydroxyzine) or sedative (eg, amitriptyline) properties, treatment with selective serotonin reuptake inhibitor antidepressants (such as fluoxetine, paroxetine, sertraline, or citalopram) can be considered (7, 20). A topical corticosteroid ointment (medium-potency or high-potency for patients with severe disease) should be applied once or twice daily. Patients using high-potency topical corticosteroids should be followed up 4 weeks after initiation of treatment (9). Additional follow-up visits and subsequent use of steroids (frequency and potency) should be based on the patient’s response to initial therapy.

Patients with lichen simplex chronicus benefit from counseling directed at early recognition of symptoms and prompt treatment. Treatment failures often result from use of low-potency topical corticosteroids, shorter-than-recommended courses of moderate-potency or high-potency topical corticosteroids, or failure to control other factors (eg, nighttime scratching) (12). Although resolution can be obtained with appropriate treatment, recurrences are common (20).

► How is lichen sclerosus diagnosed and treated?

Diagnosis

Patients presenting with lichen sclerosus most commonly report vulvar pruritus, irritation, burning, dyspareunia, and tearing (9). On examination, typical lichen sclerosus lesions appear as porcelain-white papules and plaques, often with areas of ecchymosis (or purpura) (7, 20, 21). The skin commonly appears thinned, whitened, and crinkled (hence the description, “cigarette paper” or “cellophane appearance”). Although the genital mucosa is largely spared with lichen sclerosus, involvement of the mucocutaneous junctions may lead to introital narrowing. Involvement of the superior vulva to the perianal tissue can create the classic “figure of eight” or hourglass shape (9). Other findings include fusion of the labia minora, phimosis of the clitoral hood, and fissures (46).

A biopsy should be considered to confirm the diagnosis of lichen sclerosus (21). Although lichen sclerosus usually can be diagnosed clinically if characteristic signs are present, a biopsy often is advised because other vulvar diseases can mimic lichen sclerosus (21, 20), and



patients with lichen sclerosis, especially untreated lichen sclerosis, are at increased risk of vulvar squamous cell carcinoma, with estimates of risk ranging from 2% to 5% (21, 47–50).

Treatment

Treatment is aimed at controlling symptoms and preventing disease progression (7). Patients should be counseled that although lichen sclerosis is a chronic condition, long-term management is possible with maintenance therapy and routine follow-up (7).

Initial Treatment

A medium-potency or high-potency topical corticosteroid ointment is recommended for the initial treatment of lichen sclerosis. Current evidence and expert guidelines support the use of clobetasol propionate 0.05% or mometasone furoate 0.1% (21, 51–54). Although additional research is needed to identify the optimal dosing, potency, and duration of use, expert guidelines such as those from the British Association of Dermatologists recommend clobetasol propionate 0.05% ointment once daily at night for 4 weeks, then alternate nights for 4 weeks, and then twice weekly for 4 weeks (21). Monitoring at 3 months following initial therapy for lichen sclerosis is recommended to assess the patient's response to therapy and to ensure proper application of the medication (9, 21). For patients with well-controlled disease, it is reasonable to perform a second assessment 3 to 6 months later (7, 21). For those with poorly controlled disease, more frequent visits are indicated (7, 21, 50). Patients should be advised to return for visits if persistent ulcerations or new growths appear (21). Biopsy of such lesions, and of erosions, hyperkeratotic or hyperpigmented areas, is important to exclude intraepithelial neoplasia or invasive squamous cell cancer.

Long-Term Maintenance Therapy

Once remission is achieved, long-term, individualized topical corticosteroid therapy is recommended to maintain normality of skin color and texture and to prevent scarring in patients with lichen sclerosis (9, 50, 55, 56). Maintenance treatment should be titrated to the lowest dose needed to maintain symptom resolution (55). Although chronic topical corticosteroid therapy raises concerns over skin changes, secondary infection, and risk of systemic absorption, studies have shown that long-term maintenance with medium-potency or high-potency topical corticosteroid therapy is effective and does not appear to lead to atrophy, telangiectasia, striae, or secondary infection (50, 56, 57). A randomized study of 27 patients with lichen sclerosis found that a twice-

weekly maintenance regimen with mometasone furoate 0.1% ointment for 56 weeks prevented lichen recurrence with no reported adverse effects (56). A prospective cohort study of 507 patients followed for a minimum of 2 years demonstrated that adherence (defined as all or most of the time) to an individualized long-term topical corticosteroid therapy regimen conferred multiple benefits over partial adherence (defined as some, little, or none), including a significant difference in symptom control (93.3% vs. 58.0%; $P < .001$), adhesions and scarring (3.4% vs. 40.0%; $P < .001$), and occurrence of vulvar carcinoma (0% vs. 4.7%; $P < .001$) (50).

Treatment Failure

If treatment with topical corticosteroids fails, the most important next step is to check that the diagnosis of lichen sclerosis is correct. If not obtained previously, a biopsy should be performed (55). Issues with treatment adherence also should be ascertained (21). In addition, careful attention to the role of urinary incontinence, the possibility of superimposed bacterial or fungal infection, or the development of contact dermatitis or lichen simplex chronicus is important and may explain worsening of symptoms despite correct use of medication (21).

In patients with lichen sclerosis that is poorly controlled or is resistant to topical corticosteroids, intralesional corticosteroid injections can be considered (7, 21, 58–60). The British Association of Dermatologists guidelines suggest 10–20 mg of intralesional triamcinolone in patients whose condition does not respond to topical corticosteroid treatment and who have had a biopsy to exclude malignancy (21). There is no consensus on repeat dosing of intralesional corticosteroid injections.

For patients with a confirmed diagnosis of lichen sclerosis that does not respond to topical or intralesional corticosteroids, or for patients at risk of skin atrophy, topical calcineurin inhibitors (eg, topical tacrolimus or pimecrolimus) can be considered (7, 9, 52, 61). Because data on the long-term safety of these topical calcineurin inhibitors are not currently available, both pimecrolimus and tacrolimus remain second-line agents for the treatment of dermatologic disorders. Patient counseling and documentation should address the unknown long-term safety profile and the FDA black box warning about the potential risk of cancer associated with these agents (62, 63).

Referral

When improvement is not seen with therapy and other causes have been ruled out, patients should be referred to a vulvar dermatoses specialist, who may prescribe alternative therapies (eg, retinoids, phototherapy)



(64–66). Surgery, although not curative, is reserved exclusively for the treatment of malignancy and postinflammatory sequelae (eg, release of labial adhesions, introital stenosis) (48, 67).

► *How is lichen planus diagnosed and treated?*

Diagnosis

Lichen planus can affect the vulva and vagina and has several clinical variants. Symptoms commonly reported include dyspareunia, burning, soreness, itching, and increased vaginal discharge (9). Classical (papulosquamous) lichen planus presents as white, reticulate, lacy, or fernlike striae (Wickham striae). Dusky pink, poorly demarcated papules also may be present (7, 26). On occasion the skin may appear uniformly white, and thus lichen planus can be confused with lichen sclerosus (7, 20). Hypertrophic lichen planus is the least common subtype and presents as white, thick, warty plaques (9, 24). In erosive lichen planus, deep, painful, erythematous erosions appear in the posterior vestibule and often extend to the labia minora, resulting in agglutination and resorption of the labial architecture. The vaginal epithelium can become erythematous, eroded, acutely inflamed, and denuded of epithelium. Erosive patches, if present, are extremely friable. Over time these eroded surfaces may adhere, resulting in synechiae and eventually, complete obliteration of the vaginal space (9, 68, 69). Because oral involvement is common among patients with erosive lichen planus, evaluation of the oral cavity is recommended (7, 27–29). Referral to an oral specialist (periodontist) is recommended if oral disease is present.

Examination of vaginal discharge shows a predominance of inflammatory cells and immature parabasal and basal epithelial cells, which are small and round with relatively large nuclei. The vaginal pH is increased, usually in the range of 5–6 (24). Although purulent discharge may be seen with vaginal involvement, its absence does not rule out vaginal disease (24). Biopsy results may be relatively nonspecific because of the complete loss of the vaginal epithelium, but biopsy should be performed if indicated (see *When and how should a vulvar biopsy be performed?* earlier in this section). Direct immunofluorescence should be considered if there is potential for immunobullous diseases, such as cicatricial pemphigoid, bullous pemphigoid, and pemphigus vulgaris, which may mimic lichen planus (9, 14, 70).

Treatment

The prognosis for spontaneous remission of vulvovaginal lichen planus is poor. Patients should be counseled that

complete control of symptoms is not typical in patients with vulvovaginal lichen planus (14, 20, 69, 71). Based on limited evidence and expert opinion, the recommended initial treatment for lichen planus is a high-potency topical corticosteroid ointment (Box 4) (9, 27, 72, 73). A common regimen is twice-daily use with subsequent tapering, both in frequency and potency as the patient's condition improves (9, 27). As recommended for other inflammatory dermatoses, vulvar comfort care and hygiene may be helpful for patients with lichen planus (7, 9, 20, 24). Initial follow up at 2–3 months after initiation of therapy is recommended to assess treatment response. Although stable disease can be evaluated annually by the patient's primary care physician, erosive disease requires long-term specialized follow-up (9).

In patients with lichen planus that is resistant to topical corticosteroid therapy, topical calcineurin inhibitors can be considered (7, 9, 74). Patient counseling and documentation should address the unknown long-term safety profile and the FDA black box warning about the potential risk of cancer associated with these agents (62, 63).

For erosive lichen planus with vaginal involvement, treatment with intravaginal corticosteroids should be considered in addition to topical vulvar treatment (7, 9, 24, 75). Hydrocortisone acetate suppositories (25 mg) can be inserted intravaginally twice daily and then slowly tapered off to a symptom-free maintenance dose (eg, once or twice weekly). Using this regimen, 81% of patients reported substantial improvement in burning, itching, pruritus, and vaginal discharge (75). Substantial improvement in erythema and erosive changes was also noted, although vaginal stenosis remained unchanged and there was no comparator (75). In patients with lichen planus, regular use of graded vaginal dilators in conjunction with topical intravaginal corticosteroid therapy is recommended to help prevent vaginal scarring, synechiae, and complete obliteration of the vaginal vault (7).

Consultation and Referral

Collaboration with multidisciplinary experts to address emotional support, sexual dysfunction, and the need for long-term treatment should be included as part of the therapeutic program for patients with poorly controlled disease. When improvement is not seen with initial therapy and other causes have been ruled out, referral to a specialist in the treatment of vulvar dermatoses is recommended. For more extensive vaginal disease, compounded high-dose (100 mg) hydrocortisone inserts may be prescribed by a trained specialist (7, 20). Several alternative therapies have been studied, although the data supporting their use is limited. Mycophenolate mofetil and methotrexate have been studied in small cohorts, and a tapering regimen of oral steroids has been



successfully used for severe flares (9, 76–78). Retinoid acitretin can be helpful in hypertrophic cases (9). There is little evidence for the use of systemic agents such as azathioprine, dapsone, griseofulvin, chloroquine, and minocycline (9). Surgery may be required to restore the ability to have intercourse. In one series, 6 of 11 patients (55%) who underwent lysis of vulvovaginal adhesions followed by long-term vaginal dilation were able to have intercourse, although approximately 50% continued to fear pain with sex (79).

Summary of Recommendations

The following recommendation is based on good and consistent scientific evidence (Level A):

- ▶ A medium-potency or high-potency topical corticosteroid ointment is recommended for the initial treatment of lichen sclerosis.

The following recommendations are based on limited or inconsistent scientific evidence (Level B):

- ▶ Long-term, individualized topical corticosteroid therapy is recommended to maintain normality of skin color and texture and to prevent scarring in patients with lichen sclerosis.
- ▶ For patients with a confirmed diagnosis of lichen sclerosis that does not respond to topical or intralesional corticosteroids or for patients at risk of skin atrophy, topical calcineurin inhibitors (eg, topical tacrolimus or pimecrolimus) can be considered.
- ▶ Because oral involvement is common among patients with erosive lichen planus, evaluation of the oral cavity is recommended.
- ▶ The recommended initial treatment for lichen planus is a high-potency topical corticosteroid ointment.
- ▶ In patients with lichen planus that is resistant to topical corticosteroid therapy, topical calcineurin inhibitors can be considered.

The following recommendations are based primarily on consensus and expert opinion (Level C):

- ▶ The initial evaluation of patients with vulvovaginal symptoms should include a comprehensive medical history, a physical examination, and evaluation of abnormal vaginal discharge if an infectious etiology is suspected.
- ▶ Vulvar biopsy is recommended for visible lesions in any of the following circumstances:

- lesions are atypical (eg, new pigmentation, indurated, affixed to underlying tissue, bleeding, or ulcerated)
- there is concern for malignancy
- lesions in a patient who is immunocompromised (including those infected with HIV)
- the diagnosis is uncertain
- lesions do not respond to standard therapy
- the disease worsens during therapy
- ▶ Recommended treatment of contact dermatitis includes patient counseling regarding vulvar care and removal of vulvar irritants and allergens and symptom management with a topical corticosteroid ointment and an oral antipruritic medication as needed.
- ▶ The severity and chronicity of contact dermatitis will determine the need for and the potency of the topical corticosteroid chosen for treatment.
- ▶ Recommended treatment of lichen simplex chronicus includes patient education on how to stop the “itch-scratch cycle,” information regarding vulvar care and hygiene, a medium-potency or high-potency topical corticosteroid ointment, and oral antipruritic medication as needed.
- ▶ Monitoring at 3 months following initial therapy for lichen sclerosis is recommended to assess the patient’s response to therapy and to ensure proper application of the medication.
- ▶ In patients with lichen sclerosis that is poorly controlled or is resistant to topical corticosteroids, intralesional corticosteroid injections can be considered.
- ▶ For erosive lichen planus with vaginal involvement, treatment with intravaginal corticosteroids should be considered in addition to topical vulvar treatment.
- ▶ In patients with lichen planus, regular use of graded vaginal dilators in conjunction with topical intravaginal corticosteroid therapy is recommended to help prevent vaginal scarring, synechiae, and complete obliteration of the vaginal vault.

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The MEDLINE database, the Cochrane Library, and the American College of Obstetricians and Gynecologists' own internal resources and documents were used to conduct a literature search to locate relevant articles published between January 2000–February 2020. The search was restricted to articles published in the English language. Priority was given to articles reporting results of original research, although review articles and commentaries also were consulted. Abstracts of research presented at symposia and scientific conferences were not considered adequate for inclusion in this document. Guidelines published by organizations or institutions such as the National Institutes of Health and the American College of Obstetricians and Gynecologists were reviewed, and additional studies were located by reviewing bibliographies of identified articles. When reliable research was not available, expert opinions from obstetrician–gynecologists were used.

Studies were reviewed and evaluated for quality according to the method outlined by the U.S. Preventive Services Task Force:

- I Evidence obtained from at least one properly designed randomized controlled trial.
- II-1 Evidence obtained from well-designed controlled trials without randomization.
- II-2 Evidence obtained from well-designed cohort or case–control analytic studies, preferably from more than one center or research group.
- II-3 Evidence obtained from multiple time series with or without the intervention. Dramatic results in uncontrolled experiments also could be regarded as this type of evidence.
- III Opinions of respected authorities, based on clinical experience, descriptive studies, or reports of expert committees.

Based on the highest level of evidence found in the data, recommendations are provided and graded according to the following categories:

Level A—Recommendations are based on good and consistent scientific evidence.

Level B—Recommendations are based on limited or inconsistent scientific evidence.

Level C—Recommendations are based primarily on consensus and expert opinion.

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