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Patient education: Lichen sclerosus (Beyond the Basics)

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INTRODUCTION

Lichen sclerosus (LS) is a skin disorder that causes the skin to become thin, whitened, and wrinkled, and can cause itching and pain. LS usually occurs in postmenopausal women, although men, children, and premenopausal women may be affected. It can develop on any skin surface, but in women it most commonly occurs near the clitoris, on the labia (the inner and outer genital lips), and in the anal region (figure 1). In men, it most commonly occurs on the glans (head) of the penis and the foreskin. In 15 to 20 percent of patients, LS lesions develop on other skin surfaces, such as the thighs, breasts, wrists, shoulders, neck, and even inside the mouth.

It is not clear exactly how many people have LS. Estimates for LS involving the female genitals vary from 1 in 30 older adult women seen in general gynecology offices to 1 in 300 to 1000 patients referred to dermatologists.

LICHEN SCLEROSUS CAUSES AND RISK FACTORS

The cause of lichen sclerosus (LS) is not clear; health care providers suspect that a number of

factors may be involved.

Genetic factors — LS seems to be more common in some families. People who are genetically predisposed to LS may develop symptoms after experiencing trauma, injury, or sexual abuse.

Disorders of the immune system — LS in females may be an autoimmune disorder, in which the body's immune system mistakenly attacks and injures the skin. Women with LS are at greater risk of developing other autoimmune disorders, such as some types of thyroid disease, anemia, diabetes, alopecia areata, and vitiligo [1].

Infections — Researchers have not been able to clearly demonstrate any relationship between infections and LS. LS is not contagious.

Hormones — LS is more common in prepubertal girls and postmenopausal women, suggesting that hormonal changes influence the disease. However, treatments such as hormone replacement therapy or the application of testosterone or progesterone have not been shown to be effective for females with LS.

Urine — There is evidence that urine may contribute to male LS, in that microscopic droplets of urine may pool between the glans penis and the foreskin, contributing to LS in uncircumcised men.

LICHEN SCLEROSUS (LS) SIGNS AND SYMPTOMS

Features of genital LS in women — Some women with genital LS feel dull, painful discomfort in the vulva, while other women have no symptoms. The most common symptoms include:

- **Vulvar itching** – The most common symptom of LS is itching. It may be so severe that it interferes with sleep.
- **Anal itching, fissures, bleeding, and pain** – (See "Patient education: Anal fissure (Beyond the Basics)".)
- **Painful sexual intercourse (dyspareunia)** – This can occur as a result of repeated cracking of the skin (fissuring) or from narrowing of the vaginal opening due to scarring.

Typically, women with genital LS have thin, white, wrinkled skin on the labia, often extending down and around the anus (figure 1). Purple-colored areas of bruising may be seen. Cracks (also known as fissures) may form in the skin in the area around the anus, the labia, and the clitoris. Relatively minor rubbing or sex may lead to bleeding due to the fragility of the involved skin.

Genital LS may progress and change the appearance of the genital area as the outer and inner

lips of the vulva fuse (stick together) and cover the clitoris. The opening of the vagina can become narrowed, and cracks, fissures, and thickened, scarred skin in the genital and anal area can make sexual intercourse or genital examination painful. LS does not affect the inner reproductive organs, such as the vagina and uterus.

Features of genital LS in men — In men, LS may appear on the head of the penis. Men who develop LS are usually uncircumcised (they have not had the foreskin of the penis removed), and the foreskin can become tight, shrunken, and scarred over the head of the penis. Men with LS may also have problems pulling back the foreskin and may experience decreased sensation at the tip of the penis, painful erections, or problems with urination.

Features of LS in other areas — LS may also cause lesions to occur in areas outside the genitals, especially the upper body, breasts, and upper arms. These lesions tend to be white, flat, slightly wrinkly, and not as itchy as the affected skin of the genitals and anus.

LICHEN SCLEROSUS DIAGNOSIS

Providers typically use the following methods to diagnose lichen sclerosus (LS).

History and physical examination — A medical history and physical examination of the vulvar and anal areas will be done, looking for the signs and symptoms of LS. A general skin examination may also be performed to exclude LS elsewhere on the body.

Biopsy — To confirm a suspected diagnosis of LS, a biopsy is recommended. A small piece of the affected skin will be removed and sent to a pathologist to be examined with a microscope.

Excluding other conditions — Tests may be done to exclude other conditions that could cause symptoms similar to those of LS, such as:

- Lichen planus (a skin disease that can also cause itching and fusing of genital skin). Lichen planus can occur together with LS.
- Low estrogen level (a lack of the hormone estrogen can rarely cause fusing of genital skin but is often the cause of painful intercourse). (See "Patient education: Vaginal dryness (Beyond the Basics)".)
- Vitiligo (a disorder that can cause white skin patches similar to those of LS). Vitiligo can occur together with LS.
- Pemphigoid (a blistering skin disorder that also causes scarring of the vulva) is extremely rare.

- Infections can cause similar symptoms but usually do not cause the typical skin changes of LS. However, infection can occur together with LS.

LS and cancer — Women with LS affecting the vulva are at a slightly increased risk for developing squamous cell skin cancer of the vulva.

Diagnosing genital LS early, treating it effectively, and biopsying any abnormal areas may help to reduce the risk of developing or missing a diagnosis of skin cancer. A once-yearly examination of the skin of the vulva is recommended, and women should examine themselves regularly for lumps or sores that do not heal. A biopsy should be performed if there are areas that do not improve with treatment. There is early evidence to suggest that good control of LS may reduce the risk of vulval cancer.

LS lesions outside the genital area do not have an increased risk of cancer. Men with LS that affects the skin of the penis have an increased risk of squamous cell skin cancer of the penis.

LS and painful sexual intercourse — LS can lead to constriction of the vaginal opening and pain during sexual intercourse. Women who experience pain during sex first require treatment to suppress any active disease. Once the disease is controlled, some clinicians may recommend an estrogen cream to help to soften the skin around the vaginal opening. Devices called vaginal dilators, which patients can use at home, also may be used to slowly stretch the skin.

Pain with intercourse can also occur from other causes. Patients who notice pain during intercourse should discuss their symptoms with their health care providers.

LICHEN SCLEROSUS TREATMENT

The goals of treatment of lichen sclerosus (LS) are to relieve bothersome symptoms and to prevent the condition from worsening. A clinician may recommend medication for the physical symptoms, and may refer the patient for support and therapy for other issues associated with the condition, such as problems with sex.

All patients with genital LS, even those without noticeable symptoms, need to use medication on a regular and ongoing basis. Patients also should see a health care provider for reevaluation of the disease at least once or twice yearly.

Patients who are diagnosed with genital LS should talk to their clinician about:

- The lifelong and potentially progressive nature of LS; appropriate treatment can stop the condition from worsening.

- Ways to manage the condition.
- The slightly increased risk of vulvar cancer and the need for ongoing monitoring.
- How to keep the genital area healthy and avoid scratching (table 1).
- Persistent pain with intercourse.
- Good vulval hygiene, including avoidance of irritant products (eg, soaps, douches, and body washes) and the use of a bland emollient (moisturizer).

Depending on the severity of the condition, a health care provider may recommend one or more of the following treatments for genital LS:

- Steroid ointments are recommended to reduce inflammation and itching. They are the treatment of choice for genital LS. Strong steroid ointments (eg, clobetasol propionate) are the mainstay of treatment for genital LS and are effective in the majority of women. Initial treatment usually requires daily application of the ointment for one to three months to resolve the symptoms and reduce inflammation. After the initial course, most women require "maintenance" therapy with either less frequent application of the strong steroid ointment or a switch to a less potent steroid. Although there may be warnings on the product about the use of topical steroids on genital skin, it is important to use an adequate amount to bring the disease under control. The health care provider will provide guidance about the amount to use and frequency of application.
- Steroid injections, especially if steroid ointments are not effective.

Another class of topical medications are the calcineurin inhibitors (eg, tacrolimus or pimecrolimus), which are sometimes prescribed for patients who respond poorly to steroids or cannot tolerate steroid treatment.

Although it is not approved by the US Food and Drug Administration (FDA) for this use, an oral medication called acitretin has also been used for the treatment of LS in some patients. Because it has many side effects, including a risk for liver damage, the drug is used primarily in patients who have not been helped by other treatments. Acitretin can cause severe birth defects, and women should not get pregnant during treatment or for three years after taking the drug. For this reason, acitretin usually is not recommended for women of child-bearing age.

Some women with LS may develop abnormal fusion of the labia and/or scarring. Vaginal dilators can be used in this situation to stretch the skin to help restore normal function. Surgery may also be used in this situation. It is important to continue medical treatment (with corticosteroids) and

dilators after surgery to prevent the recurrence of scarring.

Men who have genital LS are generally treated with circumcision, which removes the foreskin of the penis. After circumcision, LS does not usually come back.

Vulvar pain sometimes persists despite treatment with topical steroids. Treatment approaches for this include self-massage and dilator therapy to reduce pain with sexual intercourse. Sometimes oral medications are recommended.

WHAT TO EXPECT

The good news for patients who have been diagnosed with lichen sclerosus (LS) is that treatments such as topical steroid ointments are very effective. Thus, early treatment of LS with topical steroids can prevent scarring. Follow-up is important throughout the lifetime.

WHERE TO GET MORE INFORMATION

Your health care provider is the best source of information for questions and concerns related to your medical problem.

This article will be updated as needed on our website (www.uptodate.com/patients). Related topics for patients, as well as selected articles written for health care professionals, are also available. Some of the most relevant are listed below.

Patient level information — UpToDate offers two types of patient education materials.

The Basics — The Basics patient education pieces answer the four or five key questions a patient might have about a given condition. These articles are best for patients who want a general overview and who prefer short, easy-to-read materials.

Patient education: Lichen sclerosus (The Basics)
Patient education: Vulvar itching (The Basics)
Patient education: Lichen planus (The Basics)
Patient education: Vulvar pain (The Basics)

Beyond the Basics — Beyond the Basics patient education pieces are longer, more sophisticated, and more detailed. These articles are best for patients who want in-depth information and are comfortable with some medical jargon.

Patient education: Anal fissure (Beyond the Basics)
Patient education: Menopausal hormone therapy (Beyond the Basics)

Professional level information — Professional level articles are designed to keep doctors and other health professionals up-to-date on the latest medical findings. These articles are thorough, long, and complex, and they contain multiple references to the research on which they are based. Professional level articles are best for people who are comfortable with a lot of medical terminology and who want to read the same materials their doctors are reading.

Vulvar dermatitis
Vulvar lesions: Differential diagnosis based on morphology
Epidemiology and risk factors for cutaneous squamous cell carcinoma
Vulvar cancer: Epidemiology, diagnosis, histopathology, and treatment
Vulvar intraepithelial neoplasia
Vulvar lichen planus
Vulvar lichen sclerosus
Treatment of vulvodynia (vulvar pain of unknown cause)
Overview of vulvovaginal complaints in the prepubertal child
Cutaneous squamous cell carcinoma (cSCC): Clinical features and diagnosis
Extragenital lichen sclerosus

The following organizations also provide reliable health information.

- National Institute of Arthritis and Musculoskeletal and Skin Diseases

  (www.niams.nih.gov/Health_Info/Lichen_Sclerosus/default.asp)

- The Association for Lichen Sclerosus and Vulval Health

  (www.lichensclerosus.org)

- National Vulvodynia Association

  (www.nva.org)

- The International Society for the Study of Vulvovaginal Disease

  (www.issvd.org)

- CareDownThere

  (www.caredownthere.com.au)

[1-10]

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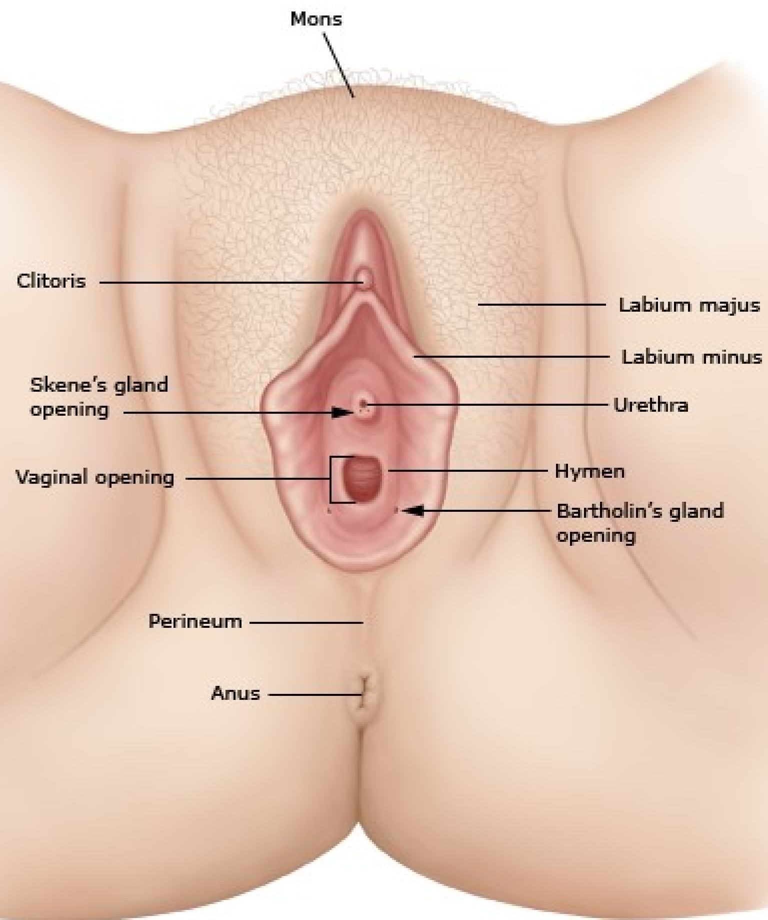
Topic 8413 Version 13.0

GRAPHICS

Adult female external genitalia



This drawing shows the different parts of the genitals.

Graphic 53704 Version 9.0

Healthy vulval hygiene practices

Do not use	Instead use
Pantyhose	Stockings with a garter belt
	Thigh high or knee high stockings
Synthetic underwear	Cotton underwear or no underwear
Jeans and other tight pants	Loose pants, skirts, dresses
Swimsuits, leotards, thongs, or lycra clothes	Loose-fitting cotton clothes
Pantyliners	Tampons or cotton pads
Scented soaps or shampoos	Fragrance free pH neutral soap (such as Basis, Neutrogena, Dove soap)
Bubble bath	Tub baths in the morning and at night with nothing added to the water and at a comfortable temperature
Scented detergents	Unscented detergents
Washcloths	Use fingertips for washing
Feminine sprays, douches, powders	These are not necessary. Do not use them.
Dyed toilet paper and other products	Toilet paper and other products without dyes
Hair dryers to dry vulva skin without contact	Dry vulva by gentle patting with a clean towel. Do not rub dry.

Graphic 58896 Version 3.0

Contributor Disclosures

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Conflict of interest policy