

Pediatric Vulvar Lichen Sclerosus

Lichen sclerosus—pronounced "LIKE-in skler-O-sus"—is a skin disease that mainly involves the genital area. It can occur in girls, boys, women and men but is most common in young girls before puberty and in women after menopause. When it occurs in children, it is often called "Pediatric Lichen Sclerosus". Lichen sclerosus in young girls mainly involves the skin of the vulva (the area surrounding the opening of the vagina) but may occasionally involve the anus as well. While you may have never heard of lichen sclerosus, it is actually a commonly diagnosed skin condition of the vulva in young girls. It is also common to have symptoms for months or years before the condition is diagnosed. Because lichen sclerosus involves the genital area, it can be both physically and emotionally distressing.

Lichen sclerosus is fairly easy to diagnose by clinicians familiar with the disease and good treatment is available. However, lichen sclerosus can sometimes be difficult to manage and may have complications. The best way to keep lichen sclerosus under control is to learn about the disease and to work with a health care provider experienced in managing it.

Typical lichen sclerosus symptoms

The most common things that girls with lichen sclerosus feel are vulvar itching and soreness. These symptoms can range from mild to severe. Girls may react to or express their discomfort in different ways. For example, some girls may simply complain that their "private area" hurts or itches while others may respond by holding their genital area or rubbing or scratching it. The degree of discomfort may not be proportional to the amount of disease present.

Lichen sclerosus can also cause painful urination and even constipation. Painful urination happens when urine passes over vulvar skin, which may be irritated and inflamed. Constipation occurs from stool withholding due to painful fissures (small tears) in the skin near the anus. Since it is painful to have a bowel movement, some girls will withhold their stool and develop constipation. Constipation from lichen sclerosus may be mistaken for a gastrointestinal problem or a behavioral problem.

Some girls with lichen sclerosus will have a discharge that is noticed on their underpants. The discharge may be bloody due to small spots of vulvar bleeding from skin irritation and scratching. This sort of bloodstained discharge is *not* dangerous.

Since there are other causes of these symptoms besides lichen sclerosus, it is important that a knowledgeable health care professional perform a careful physical examination. It is also possible for some girls with lichen sclerosus to have no symptoms at all. In this case, the skin disease is only discovered during a routine check-up.

The examination of the patient>

Examining the vulva of young girl with suspected lichen sclerosus is crucial for making the diagnosis and for following her response to treatment. It involves a careful inspection or look at the skin and at the opening to the vagina. This does NOT include the "internal" part of the adult pelvic exam, where a metal or plastic instrument is put inside the vagina.

Parents may have concerns that a gynecologic examination could be emotionally or physically traumatic to their infant, toddler, or preteen daughter. It may be helpful to remember that examination of the vulva is a normal part of a girl's routine pediatric check-up. This examination is quite different than the adult examination and is tailored specifically for a child. It is conducted in a supportive environment that is intended to remove fears and insure the child's comfort. In addition, you will be able to be present during the examination.

This examination is usually done with the girl lying down on her back with her legs placed in the "frog-leg" position. The examiner uses care to gently separate the labia (lips) so that the entire vulva may be clearly observed. There may be a brief moment of discomfort during the exam since the vulva can be sore, but the examiner will take care to minimize the discomfort. Older girls can place their feet in the footrests used for the adult gynecologic examination since this may allow the examiner to better visualize the vulva and the area around the anus in the older girl. The examiner may use a special light or magnifier to help examine the area.

You can help your daughter during the examination by reassuring her and/or distracting her with talk. Try to prepare her for the examination beforehand, even if it is a follow-up examination. For younger girls, bring along a favorite stuffed animal or another object for her to hold.

What your daughter's examiner may see

Lichen sclerosus can have different features in different girls. The disease can occur on some or all of these areas of a girl's anatomy: labia minora (inner lips), labia majora (outer lips), clitoris, introitus (opening to the vagina), and perianal area (area around the anus).

Your daughter's examiner may see a variety of changes of the skin including lightening or thinning of the skin, small tears, bruises or bleeding. Lichen sclerosus usually looks the same on both sides around the entrance to the vagina and the perianal area; this gives it what is known as an "hourglass" or "figure-of-eight" look. In severe cases, scarring may cause a shrinking appearance to the vulvar tissues. An important goal of taking care of lichen sclerosus is to make the diagnosis early, before scarring has occurred, and to treat promptly to help prevent scarring. Once scarring has occurred, it cannot be reversed.

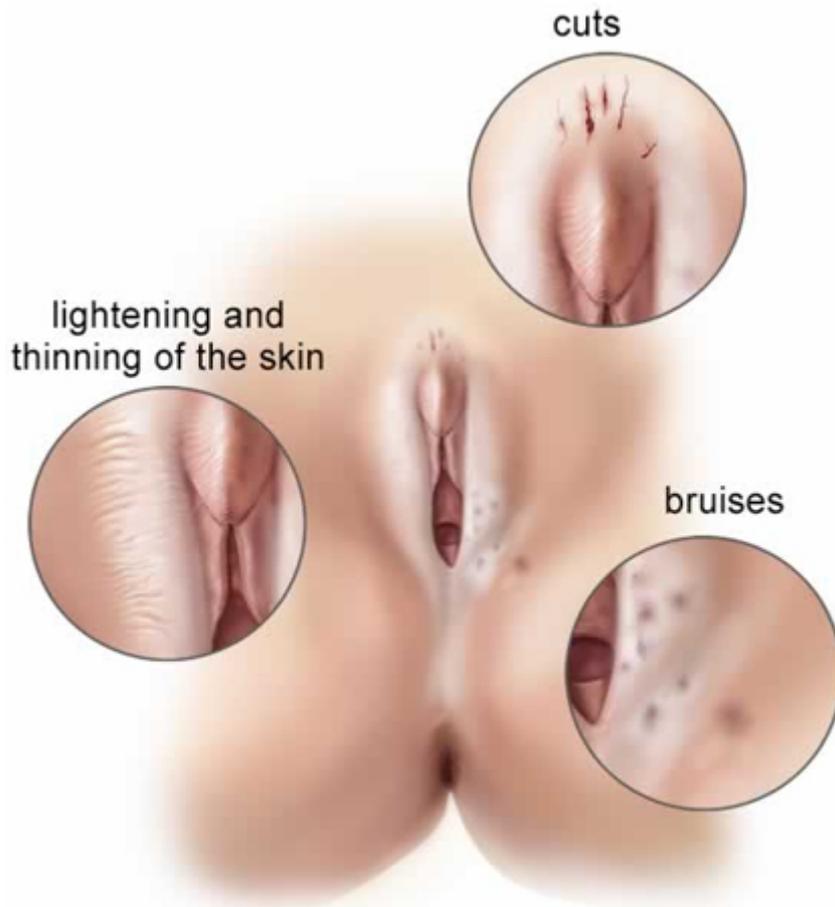


Illustration by Lisa Clark

"Tests" for lichen sclerosus

An experienced clinician can usually make the diagnosis of lichen sclerosus by examination alone. In that case treatment can be started right away.

A skin biopsy is usually not needed to diagnose lichen sclerosus since the condition has a typical look. However, if the diagnosis is not certain, or if another vulvar skin disease might be present, a skin biopsy may be recommended to help make the diagnosis. A skin biopsy is done in the office using a local anesthetic to numb the skin. A very small sample of skin (about an eighth of an inch or less) is removed and sent to the laboratory for examination.

Cultures for bacterial, fungal, and viral infections are sometimes done to make sure that an infection is not what's causing a girl's vulvar symptoms in the first place, or just to make sure an infection is not occurring along with the lichen sclerosus. Cultures are done by gently touching the area with saline-moistened swabs.

Blood tests are not needed to make the diagnosis of lichen sclerosus, but may be rarely suggested to look for other associated diseases related to lichen sclerosus.

The cause of lichen sclerosus

The cause of lichen sclerosus is simply not known. Research has focused on possible causes including:

- **Genetics.** Sometimes lichen sclerosus does run in families but there is no predictable way that it gets passed on. Most daughters or sisters of women with lichen sclerosus do not have lichen sclerosus. However, if one of your daughters has lichen sclerosus, it's a good idea to have all of your daughters examined since they may have lichen sclerosus and not know it.
- **Autoimmunity.** Some girls with lichen sclerosus also have an autoimmune disease like vitiligo (white patches on the skin), thyroid disease, alopecia areata (spots of hair loss on the scalp), or a rheumatologic disease (arthritis, rashes). However, lichen sclerosus itself is not considered an autoimmune disease.
- **Infection.** No specific infection has been shown to cause lichen sclerosus.
- **Local factors.** Local irritation of the vulva (due to warmth, moisture, tight underwear, trauma from bike riding and masturbation) may make symptoms worse in some girls.

Concerns about sexual abuse>

When a young girl has soreness of her vulva with bruising and scarring, it is natural for any parent and health care provider to be concerned about sexual abuse. A careful history and physical examination by an experienced clinician may differentiate between findings of lichen sclerosus and sexual abuse.

It is important to be open to questions about possible sexual abuse. It can be frustrating to be asked these questions, especially since many girls with lichen sclerosus have seen more than one doctor about their symptoms before the disease is diagnosed and these questions may get asked on more than one occasion.

It is also important to realize that lichen sclerosus and sexual abuse can both occur in the same girl.

Treatment of lichen sclerosus>

Medication

The most effective treatment for lichen sclerosus in young girls is the topical application of one of the prescription steroid creams. The improvement in a girl's pain and itching is dramatic as is the healing of her vulva. Even if the disease is causing her little or no symptoms it should be treated to help prevent scarring.

The medication is typically applied twice a day for 6-12 weeks; then, based on how the girl is doing, the medication is reduced to a few times a week. The goal is to use the least amount of medicine necessary to keep the disease under control.

Signs that the disease is under good control are:

- Decreased vulvar itching
- Decreased vulvar soreness
- A more normal appearing vulva with little or no bruising or fissures

The typical thinning of vulvar skin also improves with treatment but the lightening of the skin may remain even with good treatment. Side effects are uncommon and will be checked for by your daughter's clinician with regular follow-up. Non-steroid topical medications are being studied for the treatment of lichen sclerosus and may hold promise as a way of treating lichen sclerosus without the potential side effects of topical steroids. Other topical and/or oral medicines may be recommended if your daughter's clinician suspects she also has a vulvar skin infection (such as a bacterial, fungal, or viral infection) or a local allergic reaction.

Vulvar hygiene

In addition to using medicines for lichen sclerosus, good vulvar hygiene is important to keeping your daughter comfortable. A few suggestions to help cut down on local irritation of her vulvar skin are to have her:

- Wear loose fitting cotton underpants during the day and no underpants at night
- Avoid wearing nylon tights, leotards, or other tight clothing
- Use an unscented, non-irritating cleanser (such as Cetaphil^R Gentle Skin Cleanser)
- Avoid using irritants such as bubble baths and perfumed or deodorant soaps
- Use cleansing products (soaps, shampoos) at the end of her baths to avoid long contact of the cleanser with the vulva
- Avoid staying for long periods of time in a wet bathing suit, gym clothes, or damp leotard
- Avoid the use of fabric softeners and dryer sheets

The prognosis of lichen sclerosus - or what to expect in the future>

The prognosis of pediatric lichen sclerosus is **usually very good**. Most girls will improve significantly with treatment. However, the signs and symptoms of lichen sclerosus may come and go over several years. Girls may have occasional flare-ups of the disease with the usual vulvar soreness and itching and the vulvar changes they had before. Topical medication will need to be used periodically to calm down the flare-ups.

The main complications of lichen sclerosus have to do with scarring. Scarring is caused by chronic (long-term) inflammation of the vulvar skin and may lead to a shrinking of the vulvar tissue. To minimize these scarring complications, it is important to start treatment early, before scarring has occurred, and to treat any flare-ups right away. In rare cases, surgery is needed to correct complications of scarring such as narrowing of the vaginal opening. Scarring and shrinkage of the clitoris and labia minora cannot be reversed.

Lichen sclerosus does not cause skin cancer. However, skin that is scarred by lichen sclerosus in adulthood may be more likely to develop a particular skin cancer called squamous cell carcinoma. Because it is not known whether or not young girls with lichen sclerosus have an increased risk of developing vulvar squamous cell carcinoma later in life, it makes sense that all girls with lichen sclerosus be monitored for life by an experienced clinician.

Sexual functioning later in life>

Parents may have concerns about how lichen sclerosus will affect their daughter's sexual functioning later in life. Since treatment is so effective, young girls with lichen sclerosus who do not have significant scarring can be expected to have normal sexual functioning. If a young girl has or develops significant scarring, it is possible for her to have reduced sensation and sexual pleasure later on.

Psychosocial concerns>

Having a long-term, itchy, painful, disease involving the genital area may be very upsetting to a young girl. Girls with lichen sclerosus may need a great deal of emotional support and encouragement to deal with their disease.

As a girl with lichen sclerosus goes through adolescence, it is natural for her to have concerns about how her skin disease will affect her ability to have sexual pleasure, have sexual intercourse, and reproduce. These concerns should be expected and talked about. When to discuss these issues depends on the age and maturity of the individual girl.

A few final thoughts>

Learning that your daughter has a chronic vulvar skin disease can be very distressing. We hope that this pamphlet will help give you the knowledge and encouragement needed to team-up with your daughter and her health care professional to take care of her condition.

Like any other long-term disease, lichen sclerosus will have its ups and downs. There will be times that the disease won't bother your daughter at all and times that it will cause her pain and worry. There will be times that she will be good about using her medication and times that she may not be. Your support is critical to help her deal with these ups and downs.

Resources

National Institute of Arthritis and Musculoskeletal and Skin Diseases:
www.niams.nih.gov/hi/topics/lichen/lichen.htm