Anogenital Pruritus:
Lichen Sclerosus in Children

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Lichen sclerosus (LS) is a chronic, mucocutaneous inflammatory disorder of unknown etiology. It has been referred to as lichen sclerosus et atrophicus, lichen albus, balanitis xerotica obliterans, and kraurosis vulvae. Recent attempts to standardize nomenclature within and between specialties have resulted in the all-encompassing term lichen sclerosus.\(^1\)\(^2\) LS is a fairly common problem, classically affecting pre-pubertal females and post-menopausal women. Reports list the prevalence as 1 in 300 to 1 in 1,000 women\(^2\)\(^3\) and 1 in 900 girls,\(^4\) and it may be under-reported and underrecognized. Approximately 7% to 15% of all cases are found in pre-pubertal females.\(^1\)\(^3\)\(^5\)\(^6\) Nevertheless, some cases involve males, with the

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**CME EDUCATIONAL OBJECTIVES**

1. Identify the clinical manifestation of lichen sclerosus in children and adolescents.

2. Evaluate the complications that children with lichen sclerosus can anticipate during the course of their disease.

3. Determine the most effective evidenced-based treatment options for lichen sclerosus in children and adolescents.

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Figure 1. Whitish, atrophic plaques of the perivaginal and perianal region.
reported female-to-male ratio ranging from 10 to 1 to 6 to 1.1,2 LS can present as young as 6 months in both sexes, with the female mean age of developing symptoms at 4.0 years6 to 5.0 years4 and the mean age at diagnosis of 5.2 years6 to 6.7 years.4 This 1-year average of delay in diagnosis reveals a significant problem for patients. Therefore, LS is an important condition for pediatricians to recognize and consider in the differential diagnosis of anogenital conditions in children. Some clinicians believe it to be one of the most common chronic vulvar conditions.4,7

**Clinical**

Childhood LS most commonly affects the genital region, with fewer than 10% of patients having lesions elsewhere on the body.4 The most common clinical presentation in females is vulvar or perianal pruritus. Powell et al found that 80% of patients presented with itching or soreness. Other symptoms included purpura (20%), dysuria (16%), constipation (12%), and genital erosions (6%). Only 7% were asymptomatic. Maronn et al emphasized the high incidence of constipation with anogenital LS.6 In the latter study, 78% of patients identified pruritus as their most common symptom, but 67% had severe constipation. Eighty-nine percent of the patients had at least one gastrointestinal symptom, including constipation, pain on defecation, perianal fissuring, fecal impaction, and soiling. Berth-Jones et al also found symptoms of dysuria and pain with defecation in 67% of patients.8 Additionally, their patients reported a high incidence of soreness (67%) and pruritus (53%).

Less commonly, patients had recurrent bleeding and/or blistering (30%). Therefore, young females presenting to the pediatrician’s office with anogenital itching, soreness, or gastrointestinal problems may have LS (see Table).

In females, LS can affect the vulva, perianal region, clitoris, internal surface of the labia majora, labia minora, and the vaginal introitus. Patients with significant involvement have a figure-8 pattern encircling the vagina and anus (see Figure 1, page 785). On exam, females have white polygonal papules with central induration, which over time coalesce into plaques. These plaques develop a smooth, atrophic, porcelain-white, cigarette-paper surface. Edema, telangiectasia, purpura, and fissures may be seen at any time. More severe inflammation leads to blistering with hemorrhagic bullae or erosions.1,3,8,9 If untreated, vaginal architecture can be altered. Gibbon et al reported two cases, ages 3 and 4, who presented with labial fusion due to LS.10 Typically, the labia minora becomes reabsorbed and the clitoris is entrapped, revealing an overall atrophic, shiny, white vulva missing normal anatomy. The vaginal introitus also becomes stenotic and narrowed, which in adults leads to chronic pain and dyspareunia.

For boys, phimosis, or inability to retract the foreskin, is a common presentation. LS is believed to be one of the major causes of scarring phimosis leading to circumcision. Chalmers et al found LS in 14% of patients requiring circumcision for medical reasons, with a median age of 7 years.11 Forty-percent of patients with phimosis had LS in a study by Kiss, et al with the highest incidence in boys 9 to 11 years.12 Even more striking, 60% of patients with acquired phimosis and 30% with congenital phimosis had LS in the study by Mattioli, et al.13 Patients also present with adhesions to the glans, decreased sensation of the glans, dysuria, meatal stenosis, urinary obstruction, and urethral discharge.1,3 Mattioli et al also found that 15% of their cases with hypospadias had LS.13 Males rarely have perianal involvement, which is distinct from females.14 Examination reveals a diagnostic sclerotic white ring at the tip of the prepuce, leading to difficulty retracting the foreskin. Boys can also less commonly have a mottled appearance to the glans with consequential meatal stenosis and urinary symptoms.

Koebner phenomenon occurs when lesions develop in areas of trauma or at previous surgery sites and is well known in association with LS. For example, LS has been found adjacent to vulvectomy sites.3 It has been postulated that recurrent low grade infection around the vulva, tight clothing, trauma from bicycle riding and superficial radiation therapy for pruritus have all contributed to cases of LS.1 Recurrent trauma or infection may also lead to LS on the penis after surgery for squamous cell carcinoma, correction of gonadal dysgenesis, and correction of hypospadias. Historically, LS in women was corrected with vulvotomies, but the failure rate was quite high, most likely because of this phenomenon.
DIFFERENTIAL DIAGNOSIS

Although LS has a characteristic clinical presentation in both males and females, definitive diagnosis can be obtained with a skin biopsy from the affected site. Although the histologic features can vary somewhat depending on the lesion’s duration and associated rubbing of the skin, LS has a distinctive microscopic appearance. Skin biopsies are recommended for the full evaluation of LS. However, in children, a clinical diagnosis is usually sufficient until the age when skin biopsy is more feasible. Delay of biopsy is possible because the clinical history and appearance is usually characteristic.

The differential diagnosis for LS can be broad. It is important to first consider any chronic infections causing discomfort or skin changes. This can include candidiasis, superficial bacterial infections, and viral infections such as herpes simplex. Cultures and treatment of these infections is recommended if the history or exam is suggestive. Additionally, chronic diarrhea from any number of causes can lead to irritant dermatitis in this region. Irritant or allergic dermatitis can also be caused by lotions, powders, over-the-counter anti-itch creams, diaper, soaps, or detergents.15 Any of these problems can be mistaken for LS or co-exist with LS.

Sexual abuse is commonly a consideration when patients with LS are examined. The skin affected by LS in prepubescent females is more easily damaged by mild trauma.16 The erosions, fissures, hematomas, bleeding, and scarring that accompany LS raise concerns for possible abuse. There have been numerous case reports of patients with the classic presentation of LS undergoing extensive evaluation for sexual abuse.16-18 In the report by Marron, et al, 50% of patients who were first misdiagnosed as having been sexually abused before the correct diagnosis of LS was made.6 Young et al reported on three cases of LS among 469 prepubescent females referred for evaluation of sexual abuse.6 Powell et al mentioned that sexual abuse had been considered in more than 70% of their cohort of 72 girls with LS.19 Since LS exhibits koebnerization and can develop at sites of trauma, it has been theorized that these patients are more likely to exhibit injuries after abuse.8,9 Although the heightened awareness of sexual abuse in many cases has resulted in appropriate referrals by pediatricians to specialty clinics for the proper diagnosis of LS to be made at a younger age, the added emotional trauma to the family can be unnecessary.19 Nevertheless, if sexual abuse is suspected, the two conditions can certainly occur in the same patient, and at times a full inquiry into sexual abuse may still be appropriate. In one review, 12 of 42 patients (29%) with a diagnosis of LS were also sexually abused.20

Other considerations in the differential diagnosis for LS are morphea (a form of localized scleroderma) and lichen planus.13,14,21 As with LS, both conditions have unknown etiology with slightly different clinical presentation and histologic differences. LS is more commonly found on the genitalia than morphea. Morphea also forms a smooth, shiny oval plaque yet feels more firm and indurated with a violaceous border.21 Lichen planus can affect the genitalia as papules or erosions, but typically has other areas on the body involved as well. In genital lichen planus, purple, flat-topped polygonal papules are found in clusters. Patients with lichen planus also have Wickham’s striae on their buccal mucosa and vulva, appearing as white, lacy, thin plaques. All of these conditions are believed to be related to autoimmunity and may be a spectrum of the same disease. Farrell et al reported on patients found to have both LS and morphea and one with all three diagnoses.21 With reports of patients with coexistent lesions, it is unclear if morphea and LS are the same disease, distinct diseases occurring coincidentally, or distinct diseases with similar etiology.1 Nevertheless for the pediatrician, if there is any doubt as to the appropriate diagnosis, a skin biopsy or referral to a specialty clinic should be sufficient to establish the diagnosis.

Lastly, LS can also be misdiagnosed as vitiligo, since both conditions cause depigmentation in the genitals of both males and females. Vitiligo frequently is found on other areas of the body such as face, elbows, knees, and digits. Additionally, vitiligo does not have the associated pruritus, pain, superficial atrophy, or scarring seen with LS.

ETIOLOGY

The etiology of LS is unknown, although a relationship to autoimmune disease is currently the most accepted theory. There have been many reports of familial cases among first degree relatives, so there seems to be a genetic role in the pathogenesis.1,4,14 Also, many patients with LS have positive autoantibodies or associated autoimmune diseases. The strongest associations seem to be with concomitant thyroid disease, vitiligo, alopecia areata, aplastic anemia, and diabetes mellitus, but there are reports of associations with many other autoimmune diseases as well.1 In the study by Powell et al of 70 girls with LS, 15% had an associated autoimmune disease, which included vitiligo, thyroid disease, alopecia areata, and rheumatic symptoms.2 Sixty-five percent of patients had a family history of autoimmune disease.
as well. In a study of 350 adult women with LS, 21.5% had an autoimmune disease, 42% had positive autoantibody titers, and 21% had a first-degree relative with an autoimmune-related disease. Thyroid disease alone was found in 30% of 211 women with LS by Birenbaum et al. There are few studies evaluating the relationship of autoimmune disease in males. In a study of 29 men with LS, 21% had associated autoimmune disease, including thyroid disease, alopecia areata, diabetes mellitus, pernicious anemia, and liver disease. Most recently, Oyama et al found autoantibodies against extracellular matrix type I in 20 of 30 (67%) of female patients with LS, suggesting a direct pathologic mechanism by which LS could occur through an autoimmune mechanism.

Many autoimmune diseases have strong relationship to certain inherited HLA types. For this reason, HLA typing has been performed on many patients with LS. The HLA class II antigen HLA-DQ7 has the strongest association with lichen sclerosus. Most of these studies evaluated adults with LS. Powell et al reported on 30 children with LS and found 66% of the patients had HLA-DQ7, which is higher than in the control population. The higher incidence of concomitant autoimmune diseases, family history of autoimmune disease, and associations with HLA antigens in groups of patients with LS suggest autoimmunity as a likely cause of this disorder. Nevertheless, there is currently no definitive laboratory test or known pathogenic antigen.

Other potential causes mentioned in the literature include possible associations with Borrelia burgdorferi infections, viral illnesses, and sex hormonal factors. Although investigations into an association with B. burgdorferi have been abundant, there are conflicting data with regard to its association with LS, and this theory has fallen out of favor. For many years, LS was believed to be related to sex hormonal influences, given the observation that most cases occur in premenstrual females and postmenopausal women, both with low estrogen states. Nevertheless, treatments with topical hormones have proven to be ineffective.

**PROGNOSIS**

LS is a relapsing and remitting disease. In adults, it rarely resolves completely, causing significant morbidity. In the past it was believed that childhood LS resolved at puberty. However, there are now many reports of childhood LS persisting into adulthood. Powell et al followed 21 girls with LS after puberty. Although most had improvement, 75% of them still had symptoms of LS and vulvar squamous cell carcinoma, which on average occurs 10 years after being diagnosed with LS. There are sporadic reports of women with vulvar carcinoma who first developed LS in childhood. One 32-year-old woman with well-documented LS beginning before puberty died of vulvar squamous cell carcinoma. In men, the incidence of squamous cell carcinoma with LS has been less well studied, with most reports ranging from 2% to 8%. However, one report of 20 cases of squamous cell carcinoma found evidence of LS in 50% on histopathologic examination. Chronic inflammation and scarring is the most likely cause of this conversion to a malignancy in both men and women. Human papillomavirus (HPV) may also contribute to the risk of malignancy in some cases. Occasionally, oncogenic HPV types have been found in genital LS.

**In females, lichen sclerosus can affect the vulva, perianal region, clitoris, internal surface of the labia majora, labia minora, and the vaginal introitus.**

**EVALUATION AND TREATMENT**

In evaluating a new patient with LS, it is important to obtain a good history. In addition to a thorough physical examination with possible skin biopsy. Patients with symptoms of common autoimmune diseases or with a family history of autoimmune disease may benefit from systemic evaluation for these conditions. Evaluation and elimination of any viral, bacterial, or yeast co-infection as well as potential topical irritants is essential. In children, chronic exposure to moisture related to diapers leads to an irritant dermatitis, which must be managed. General perineal skin care measures to avoid irritation include avoiding soaps, detergents, lotions, or perfumes while keeping the area clean and dry. Washing only with tepid water and applying plain petrolatum afterward while the skin is moist is preferred.

LS has no cure. Historically, LS was treated surgically with vulvotomies. Despite this drastic approach, LS commonly recurred, perhaps due to the koechnerization phenomenon. Later, topical
testosterone, progesterone, and estrogen were used in an attempt to correct possible local hormonal balances, but these approaches were also without success.2,6

Currently, the use of ultrapotent topical corticosteroids is the first-line therapy, as mentioned in the British guidelines for management of LS,30 with topical immunomodulators also playing a significant role. Ultrapotent topical steroids are extremely effective in reducing both the signs and symptoms of LS in both girls and women in more than 90% of cases within 4 to 12 weeks.31-34 In a study of 36 girls, 72% were symptom free with complete resolution in 22% on examination after at least 3 months.32 Topical corticosteroids improve the atrophy, erosions, and anatomic changes that occur with LS, reversing histopathologic changes (see Figure 2). Most authorities recommend using a strong topical steroid (clobetasol propionate 0.05%) in the form of an ointment once or twice a day for 4 to 8 weeks. At this point, most patients experience dramatic improvement in both their symptoms and the clinical signs of LS. Once symptoms are under control, a maintenance regimen is initiated using the same potency or less potent ointment a few times a week or as needed. Since LS is a relapsing condition, reapplication of the ointment on a twice-daily basis during flares is recommended. Ultrapotent topical steroids are tolerated well by patients with rare minimal side effects, which can include a burning sensation, irritation, erythema, allergic dermatitis, and secondary yeast infections.33,34 Although atrophy is a common concern of many physicians when using topical corticosteroids, intermittent use of these medications for this chronic condition actually improves the atrophy caused by LS.32,33,34 Treating patients aggressively not only improves signs and symptoms of disease, but also improves quality of life and may help to decrease the risk of malignancy.3

Most recently, topical calcineurin inhibitors (tacrolimus and pimecrolimus) have been studied for the treatment of LS. As immunomodulators, these agents decrease inflammation by inhibiting T lymphocyte activity. Despite not being indicated for LS, there have been many reports of the efficacy of topical immunomodulators in children.35-39 A multicenter trial on the treatment of LS with topical tacrolimus was completed in 2006. In this study, 84 patients (49 women, 32 men, 3 girls) were followed over 24 weeks. While using topical tacrolimus 0.1% ointment twice daily, 16% of patients had complete clearance of active LS on exam by week 16, and 43% had complete clearance by week 24. An additional 34% had partial resolution by week 24. Thirty-five percent of patients had reduced symptoms within 2 weeks, and 9% had a recurrence by 3 months.36 The most common side effects included burning sensation (25%), itching (20%), erythema (13%), pain (7%), soreness (7%), and flu-like symptoms (5%). Use of these medications on small body surface areas twice a day does not seem to lead to significant systemic absorption.36,37 Nevertheless, long-term safety has not been determined, especially with regard to risk of malignancy.

For males, topical corticosteroids or topical calcineurin inhibitors can improve phimosis in 70% to 80% of boys and should be used as a first-line treatment.30 If unsuccessful and phimosis persists, then circumcision is in-
Topical treatments are also required for persistent LS after circumcision and when it affects the glans. Males should be monitored as well for possible urinary difficulties and malignancy development.

In general, both ultrapotent topical steroids and topical calcineurin inhibitors are effective and safe at treating LS, but recurrences are still common. The actual recurrence rates are difficult to determine given the lack of large study groups and lack of extended long-term follow-up. Additionally, it has yet to be determined if topical treatment can limit the risk of developing malignancy in pediatric anogenital LS. Although topical corticosteroids and/or immunomodulators are standard first-line treatment for LS, additional reports have included surgical treatments, oral retinoids, cyclosporine, oral antibiotics, and phototherapy.

**SUMMARY**

Lichen sclerosus is a chronic mucocutaneous inflammatory condition affecting both adults and children in a bimodal age distribution. Children can present as young as 6 months of age but average around 5 years. Females present with vulvar itching, soreness, dysuria, or gastrointestinal complaints, while males tend to have difficulty retracting the foreskin leading to phimosis. On examination, white smooth atrophic plaques are found in the anogenital region with atrophy and possible distortion of anatomy. LS in children has been commonly misdiagnosed as sexual abuse, leading to delay in appropriate diagnosis and unnecessary turmoil for families. It is a chronic relapsing and remitting condition possibly due to autoimmunity. When evaluating patients, a complete history can help guide which patients may benefit from a systemic evaluation for autoimmune disease. Although some patients have spontaneous resolution during puberty, many do not. There is a significant risk of squamous cell carcinoma developing in genital LS in adults possibly from chronic inflammation, delay in diagnosis, and delay in appropriate treatment. The risk of squamous cell carcinoma in pediatric onset LS is undefined. It is also unclear if effective control of cutaneous inflammation can decrease the risk of malignant transformation. Treatment is aimed at decreasing symptoms and returning involved skin to its normal appearance. Relapses are common. Ultrapotent topical corticosteroids are first line for the treatment of LS and can be used intermittently for years for flares. Topical tacrolimus or pimecrolimus are also good treatment options at controlling inflammation. Patients need to be monitored every 6 to 12 months even when asymptomatic because of the potential for development of malignancy. Given the distressing nature of LS, support groups and a multidisciplinary approach are recommended.

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