ABSTRACT. Objective. To call attention to constipation as a frequent sign of lichen sclerosus (LS) in girls.

Methods. A focused questionnaire was sent to parents of 24 girls with anogenital LS seen in the pediatric dermatology clinic at the Children’s Hospital of Wisconsin between January 2001 and May 2004.

Results. Eighteen of 24 questionnaires were completed and returned. The average age of onset of LS was 4.2 years, but average age at diagnosis was 5.2 years; only 1 patient was diagnosed correctly by her primary care physician. Itching was the most common symptom (78%). Severe constipation was reported in 67% of patients, and 89% had at least 1 gastrointestinal complaint (bleeding with bowel movements, fissuring, soiling, fecal impaction, or constipation).

Conclusions. Childhood anogenital LS often presents with recalcitrant constipation or other gastrointestinal complaint. Primary care physicians need to consider the diagnosis of LS and perform a thorough examination by looking for anogenital lesions when a female pediatric patient presents with unexplained constipation or other severe gastrointestinal complaints. Pediatrics 2005; 115:e230–e232. URL: www.pediatrics.org/cgi/doi/10.1542/peds.2004-1544; constipation, dermatology.

ABBREVIATIONS. LS, lichen sclerosus; GI, gastrointestinal.

Lichen sclerosus (LS) is an uncommon inflammatory disorder that can occur at any age, but has a predilection for the vulvar skin of prepubertal girls and postmenopausal women (Fig 1). It is an eruption, the typical appearance of which is an ivory-white, sharply demarcated figure-8 pattern encircling the vagina and anus. It begins as papules containing a central depression and often coalesces into plaques. The skin surface may appear finely wrinkled or have purpuric spots, fissures, and erosions. Although less common, lesions may also present extragenitally. An often-unappreciated sign of LS, extremely distressing to these children, is severe constipation, which is unrelied by standard treatment measures.

During the past 2.5 years we have seen 24 girls with LS in the pediatric dermatology clinic at the Medical College of Wisconsin (Milwaukee, WI). A significant number of their parents have expressed considerable anger and frustration due to the failure of their physicians to recognize the association between severe constipation and LS, resulting in a long delay in diagnosis. The English-language literature, including major textbooks of medicine, pediatrics, obstetrics and gynecology, and dermatology, fail to mention constipation as a feature of LS. By reporting our experience with these patients, we hope to bring this association to the attention of primary care physicians.

METHODS

A focused questionnaire was developed by us and approved by the Children’s Hospital of Wisconsin Institutional Review Board. It was sent to parents and/or guardians of girls with the diagnosis of genital LS seen in the pediatric dermatology clinic at the Children’s Hospital of Wisconsin between January 2001 and May 2004. We asked about timing of complaints in relation to actual diagnosis of LS. The questionnaire also focused on specific signs and symptoms, diagnosis, and specialists consulted for the problem.

RESULTS

Of the 24 surveys mailed, 18 (75%) were completed and returned.

The patients ranged in age from 4 to 17 years (mean: 8 years). The average age at onset of symptoms was 4.2 years, but the average age at diagnosis was 5.2 years. There was a time lag of ~1 year from onset of signs and symptoms until diagnosis of LS, the longest interval being 3 years. Only 1 patient was diagnosed by a primary care physician (a family practitioner). Two children were diagnosed in a hospital pediatric gastrointestinal (GI) clinic, and a pediatric dermatologist diagnosed all others. Efforts to obtain a diagnosis also prompted families to make multiple visits to pediatricians, gynecologists, and pediatric general and vascular surgeons.

Table 1 lists the signs and symptoms of LS in our patients and indicates those that were most bothersome. Itching was the most common complaint, but GI-related complaints were also quite prevalent. Twelve of 18 patients had constipation, and 9 had perianal fissuring. Eleven of 18 patients had pain on defecation and bleeding with bowel movements, and a smaller number had problems with soiling and fecal impaction. Thirteen of 18 cited a GI-related complaint as one of their most distressing symptoms.

Many patients were misdiagnosed before the correct diagnosis of LS was made. Nine of 18 patients were wrongly suspected of having been sexually abused by a caregiver, 7 were misdiagnosed with a yeast infection, and 3 were thought to have a primary process causing constipation. Several other in-
correct diagnoses were made: fell off bicycle, hurt doing gymnastics, chronic inflammation, hemangioma, and dermatitis.

The patients were treated by other physicians with numerous topical agents including antifungal creams, antibiotics, estrogen, moisturizers, and corticosteroids of various potencies. Most of these treatments were ineffective. With 1 exception, all patients finally had alleviation of symptoms when prescribed a class 1 topical corticosteroid. Eight of 18 patients experienced side effects from the different treatments, including cutaneous atrophy, pubic hair growth, burning, discoloration, and secondary infection.

**DISCUSSION**

Childhood LS, which occurs most frequently in the anogenital region, represents ~15% of total cases of the disorder and is seen in a 10:1 ratio of females to males. One study of pediatric vulvar LS reported a prevalence of 1:900, with a mean age at symptom onset of 5.0 years but a mean age at diagnosis of 6.7 years. Our patients had a somewhat younger mean age of onset at 4.2 years, with a mean age at diagnosis of 5.2 years. Our patients had a delay of ~1 year from symptom onset until diagnosis, as compared with 1.7 years reported in the aforementioned study.

Anogenital LS can present with a variety of signs/symptoms (itching, soreness, purpura, dysuria, constipation, pain on defecation, soiling, perianal fissures, bleeding) and without symptoms. Vulvar itching and soreness are the most commonly reported symptoms. A recent study of 70 pediatric patients reported itching and soreness in 80%. Our study also found itching to be the most frequent complaint; it was reported by 14 (78%) of the 18 subjects. However, constipation and other GI-related complaints were also prevalent. Twelve of the 18 (67%) patients reported being constipated, and 16 of 18 (89%) had at least 1 other GI complaint (bleeding with bowel movements, pain on defecation, fissuring, soiling, fecal impaction, or constipation).

LS has been mistaken frequently for child abuse because of its appearance on the genitalia and associated symptoms. Our findings supported these data, because half our 18 subjects were wrongly suspected of having been sexually abused before the establishment of the correct diagnosis.

Estimates of the frequency of constipation in pediatric LS patients varies considerably. Powell and Wojnarowska found constipation to be a symptom in only 12% of their 70 subjects, whereas the study by Clark and Mueller showed 29% of patients with constipation, and Loening-Baucke reported 82% of children had constipation. In our study, 67% of the patients suffered from constipation. In Loening-Baucke’s series, constipation was not only a symptom of LS but also the chief complaint in all patients. She suggests possible reasons for her patients having higher rates of constipation compared with previous studies as: (1) either feces covered LS lesions; or (2) the skin changes were wrongly attributed to chronic irritation from feces. Difficulties with defecation in children with vulvar LS are likely related to the presence of open, tender areas of fissuring and may be mistaken for a primary GI or behavioral problem.

There are cases reported of children with vulvar LS and a chief complaint of constipation who were inappropriately diagnosed and treated. For example, 1 was referred to a psychiatrist and treated for Senna addiction, and another was admitted to the hospital for multiple enema treatments.

The prognosis of childhood vulvar LS remains unknown. It was once widely believed that LS would resolve after a child had gone through puberty. Powell and Wojnarowska followed 21 girls through puberty. Sixteen patients reported improvement of symptoms, but 11 still required topical corticoste-
roids to treat pruritus. Sixteen of the 21 still had physical signs of LS including pallor, atrophy, labial resorption, and purpura. In another report by Powell and Wojnarowska,2 18 children were followed through puberty and only 2 had complete remission. These studies concluded that LS may improve symptomatically but may not resolve entirely at puberty.2,8

For many years LS was believed to be a “hormonal” disorder and was treated with topical estrogen, testosterone, or progesterone. Not only were these agents ineffective, but masculinization and other side effects were troublesome also. Numerous other ineffective therapeutic modalities have been tried, including radiation, tissue extracts, retinoids, vitamins, and even surgery.1,4,9 However, superpotent class 1 topical corticosteroids, including betamethasone, clobetasol, diflorasone, and halobetasol, have been considered the preferred treatment and standard of care for the past several years. Nevertheless, concerns regarding possible thinning of the skin, causing excessive absorption of steroids and resulting in local and systemic side effects, have prompted a continued search for an optimal therapy. Several reports have been published recently claiming resolution of LS in response to applications of tacrolimus, an immunomodulator recently developed for patients with atopic eczema.9–11 Should tacrolimus prove to be as effective as a class 1 super-potent topical steroid, it will offer a tremendous advantage because of the absence of side effects. However, additional trials of tacrolimus in LS patients and careful surveillance of their course are needed to determine if this drug will cure, maintain remission, or only be effective for short-term use.

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Pediatrics 2005;115:e230
DOI: 10.1542/peds.2004-1544 originally published online January 3, 2005;

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*Pediatrics* 2005;115;e230

DOI: 10.1542/peds.2004-1544 originally published online January 3, 2005;

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