

## Lichen Sclerosus

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Lichen sclerosus (LS) is a chronic, inflammatory skin condition that primarily affects the genital and perianal regions. It is an uncommon autoimmune condition characterised by skin atrophy and hypopigmentation. While the exact cause remains unclear, several factors, including genetic predisposition, autoimmune responses, hormonal influences, and chronic irritation, are believed to contribute to its development. The condition can lead to significant discomfort, pain, and psychosexual implications. <sup>1, 2, 3</sup>

### Epidemiology

Lichen sclerosus can affect individuals of all ages, with increasing incidence with age. In females, two peaks of onset are during prepubertal and perimenopausal/postmenopausal ages. The exact prevalence in the general population is unknown, but estimates suggest a range between 1 in 300 to 1 in 1,000 individuals. Prevalence may be underestimated because many patients are asymptomatic, and it is frequently misdiagnosed. It is more common in women than men with a female to male ratio varying from 1:1 to 10:1. Lichen sclerosus, both genital and extragenital, has no known racial predilection. A genetic predisposition, based on family clustering, is apparent. Of patients with vulvar lichen sclerosus, 8-39% report a family history of the condition, while only 1% of male genital lichen sclerosus patients have a family history. <sup>1, 4, 5, 9</sup>

### Pathophysiology

The pathophysiology of lichen sclerosus is complex and not fully understood. It is believed to involve various genetic, immunological, hormonal, and environmental factors. LS is considered an autoimmune-mediated condition. The exact trigger for this immune response is not well-defined but involves a combination of genetic susceptibility and environmental factors. Genetic predisposition is suggested by the increased risk in families with affected members. Specific genetic markers and associations with certain human leukocyte antigen (HLA) types have been reported in some studies, indicating a potential hereditary predisposition to the condition. <sup>1, 2, 4, 9</sup>

Lichen sclerosus is characterised by the infiltration of immune cells, particularly T-lymphocytes, into the affected skin. These immune cells release inflammatory mediators, such as cytokines, chemokines, and growth factors, leading to chronic inflammation and tissue damage. The exact triggers that initiate the autoimmune response are not fully understood but may involve the recognition of self-antigens as foreign or the activation of autoreactive immune cells. <sup>1, 2, 4, 9</sup>

Hormonal factors may play a role in the development and exacerbation of lichen sclerosus, as the condition is more prevalent in postmenopausal women and prepubertal girls. Oestrogen

has been implicated in modulating the immune response and influencing tissue repair processes. Fluctuations in hormone levels during puberty and menopause may contribute to LS development or exacerbation. <sup>1, 2,4, 9</sup>

Chronic irritation and microtrauma to the affected skin have been proposed as potential triggers for LS, especially in the anogenital region. Repeated mechanical trauma or friction can disrupt the skin barrier and trigger an inflammatory response, leading to LS development. <sup>1, 2,4</sup>

Lichen sclerosis is associated with alterations in the extracellular matrix, particularly the accumulation of collagen and elastin fibres, leading to sclerosis and atrophy of the skin. These changes in the connective tissue architecture can contribute to the characteristic symptoms and appearance of LS. <sup>1, 2,4, 9</sup>

In some cases, infections, such as human papillomavirus (HPV), have been suggested as possible triggers for lichen sclerosis. HPV infection can induce changes in the local immune environment and promote chronic inflammation, potentially contributing to LS development in susceptible individuals. <sup>1, 2,4, 9</sup>

### **Symptoms and Presentation**

Lichen sclerosis most commonly affects the genitals and less often the extragenital area. Lesions typically begin as a sharply demarcated erythema that becomes thin, hypopigmented, ivory-white, porcelain-like, and sclerotic plaques. Plaques may later become thickened due to repeated excoriations. Itching is the main symptom and is usually worse at night. Other lesions may include telangiectasias, purpura, fissures, ulcerations, and oedema. <sup>1, 9</sup>

Lichen sclerosis typically presents with pruritus, which is often severe and persistent. Symptoms commonly include pruritus, a local burning sensation, pain, and painful defecation, however, lesions can be asymptomatic. In females' genital lesions begin around the periclitoral hood. The affected area varies from a small and single area to large area involving the entire region of the vulva, perineum, and peri-anus. Lichen sclerosis usually spares the vagina and cervix. Male genital LS occurs in the foreskin, glans penis, and the coronal sulcus penis. <sup>7</sup> Extragenital lesions occur on any part of the skin and are usually asymptomatic. The most involved areas are inframammary areas, neck, wrists, thighs, upper back, and shoulders. The involvement of the oral mucosa appears clinically as bluish-white papules on the buccal mucosa or under the tongue. <sup>1, 3,4</sup>

### **Diagnosis**

Diagnosis of lichen sclerosis is clinical, based on a careful medical history including autoimmune diseases in the patient and family, examination of the mucosa's, extragenital skin and a gynaecological exam. Workup should include investigation of thyroid function, and according to symptoms, investigation of the other autoimmune diseases. <sup>1, 9</sup>

Due to LS resemblance to other dermatological conditions, such as lichen planus and other inflammatory skin disorders, a biopsy may be necessary for confirmation. Biopsies should be

performed in case of atypical clinical presentation; suspected malignancy; and nonresponse to recommended first-line treatment. Histopathological examination reveals epidermal atrophy, sclerosis, hyalinisation, and a lymphocytic inflammatory infiltrate.<sup>1</sup>

### **Complications**

Lichen sclerosus is a chronically relapsing condition and if untreated leads to a potential scarring process that may evolve to a complete loss of standard vulvar architecture in women including introital stenosis, fusion, and resorption of the labia minora, and urethral strictures in males. Tissue adhesion and sclerosis lead to tearing and the loss of sexual function, in addition to dysuria, constipation, itching, and soreness. Vulvar lichen sclerosus may evolve to vulvar squamous cell carcinoma in the affected area with an estimated risk up to 5 %, however, its association with penile SCC is not clear. Extragenital lesions are not associated with the risk of transformation. Melanoma and basal cell carcinoma have been reported.<sup>1,8</sup>

### **Treatment**

Treatment of lichen sclerosus aims to alleviate symptoms and prevent complications such as atrophy, scar formation, anatomical distortion, as well as malignant transformation, and improve quality of life. Management strategies can vary depending on the severity of symptoms, age, and gender. Patients should be educated about the condition and encouraged to avoid the use of irritating products such as soap in the area, and to use emollients to break the itch-stretch cycle. LS may lead to scarring and loss of standard genital architecture and education relating to sexual dysfunction and dyspareunia may be required. Patients with genital lichen sclerosus should also be educated on what changes (e.g., ulceration) might indicate malignant transformation and necessitate an immediate re-evaluation.<sup>1,4,5,9</sup>

For genital LS, the gold standard treatment is three months application of high potency topical steroids for example clobetasol propionate. Apply once daily, at night, for 4 weeks, then on alternate nights for 4 weeks, and then twice weekly for a further 4 weeks, before review One fingertip unit amount should be used for each application, with no more than 10 g used monthly. A 30g tube of Dermovate should last at least 12 weeks.<sup>10</sup>

Second-line therapies include topical calcineurin inhibitors and imiquimod. For postmenopausal women, oestrogen therapy may be beneficial in managing LS symptoms and preventing disease progression. In men, early circumcision may be recommended. Surgery is indicated only for the treatment of complications associated with lichen sclerosus. Narrowband ultraviolet B (NB-UVB) phototherapy has shown promising results in treating LS, particularly in cases resistant to topical therapies. For extragenital LS, therapeutic modalities are limited and include phototherapy, ultrapotent topical steroids, tacrolimus ointment 0, 1%, and systemic steroids or methotrexate.<sup>1,,4,6,8</sup>

Given the potential for malignant transformation to squamous cell carcinoma, patients with anogenital lichen sclerosus should receive long-term follow-up.<sup>1</sup>

## Prognosis and Outlook

Prognosis can vary depending on several factors, including the age of onset, severity of symptoms, and response to treatment. In general, lichen sclerosis is a chronic condition with a variable course, and prognosis is generally considered to be good with appropriate management. Prognosis is good for more acute genital cases, especially for those in the paediatric age group, in whom it may resolve spontaneously. Lichen sclerosis tends to be a progressive condition. However, with proper treatment and management, disease progression can be slowed or halted, and symptom relief can be achieved. If it is not well-managed, LS can lead to scarring in the affected areas, particularly in the anogenital region. Scarring may cause narrowing or fusion of the genital structures, potentially leading to sexual dysfunction or difficulty with urination. Early and consistent treatment can help prevent or minimise these complications. <sup>8,9</sup>

Lichen sclerosis can cause significant discomfort, itching, pain, and psychosexual implications. With appropriate treatment, symptom relief is achievable in many cases, leading to an improved quality of life. Although the risk is relatively low, individuals with LS have a slightly increased risk of developing squamous cell carcinoma in the affected skin. Regular monitoring and follow-up are essential to detect any signs of malignancy promptly. <sup>8,9</sup>

Prognosis is generally better for individuals who respond well to treatment. Topical corticosteroids, the mainstay of treatment, are effective in managing symptoms and controlling inflammation for most patients. However, some individuals may require alternative therapies or a combination of treatments to achieve optimal results. The prognosis may differ based on gender and age. Lichen sclerosis in postmenopausal women may be more challenging to manage, and symptoms can be more persistent. In contrast, LS in prepubertal girls may spontaneously resolve with hormonal changes during puberty. Lichen sclerosis can have a psychological impact on affected individuals due to its location and the potential impact on sexual health. Psychological support and counselling can be beneficial for individuals dealing with LS-related emotional distress. <sup>8,9</sup>

Early diagnosis, appropriate management, and regular follow-up with healthcare professionals are key factors in achieving a good prognosis. With proper care, many individuals with lichen sclerosis can effectively control symptoms, and recent advances in understanding the pathophysiology have improved diagnostic and management strategies. Early recognition and appropriate treatment are essential to prevent complications and improve the quality of life for affected individuals. Further research is needed to determine the exact aetiology of lichen sclerosis and explore novel treatment modalities. <sup>1, 8, 9</sup>

## References

1. Chamli, A., Souissi, A. (2023). Lichen Sclerosus. StatPearls Publishing. Available from: <https://www.ncbi.nlm.nih.gov/books/NBK538246/>
2. Murphy, R., White, S. (2018). Lichen Sclerosus. *Dermatol Clin.* 2018;36(3):277-285.
3. Kirtschig, G. (2016). Lichen Sclerosus-Presentation, Diagnosis and Management. *Dtsch Arztebl Int.* 2016 May 13;113(19):337-43. doi: 10.3238/arztebl.2016.0337. PMID: 27232363; PMCID: PMC4904529.
4. Vahabi, F., Rezaei N., Moradi, S., Alikhan, A., *et al.* (2018). Lichen Sclerosus: Diagnostic and therapeutic update. *Curr Opin Pediatr.* 2018;30(4):482-487.
5. Birenbaum, D., Young, R. (2019). Lichen Sclerosus. *J Am Acad Dermatol.* 2019;80(3):669-675.
6. Phan, K., Ramachandran, V., Sebaratnam, D. (2019). Narrowband ultraviolet B phototherapy for the treatment of lichen sclerosus. *Australas J Dermatol.* 2019;60(1):51-56.
7. Lee A, Fischer G. (2015). Lichen Sclerosus in males: Review of literature and treatment recommendations. *J Urol.* 2015;194(5):1247-1253.
8. De Luca, D., Papara, C., Bieber, K., *et al.* (2023). Lichen Sclerosus: The 2023 Update. *Frontiers.* Available at: <https://www.frontiersin.org/articles/10.3389/fmed.2023.1106318/full>
9. Pappas-Taffer, K. (2020). Lichen Sclerosus. *Medscape.* Available at: <https://emedicine.medscape.com/article/1123316-overview?form=fpf>
10. NI Formulary (2023). Lichen Sclerosus. Available at: <https://niformulary.hscni.net/formulary/7-0-contraception-gynaecology-and-urinary-tract-disorders/7-0-d-lichen-sclerosus/>