Lichen Sclerosus

Synonyms: *lichen sclerosus et atrophicus*, *balanitis xerotica obliterans*, *lichen albus*, *white spot disease*, *Csillag’s disease*, *kraurosis vulvae*

Lichen sclerosus (LS) is a chronic inflammatory dermatosis which usually affects the skin of the anogenital region in women, and the glans penis and foreskin in men - *balanitis xerotica obliterans* (BXO). It occurs less commonly in extragenital areas. It does not cause any systemic disease outside the skin.

**Aetiology**

The cause is unknown:

- Many findings obtained in recent years point increasingly towards an autoimmune-induced disease in genetically predisposed patients and further away from an important impact of hormonal factors.\[1\]
- Preceding infections may play a provocative part.
- The role for borrelial infection is still controversial.
- Trauma and an occlusive moist environment may act as precipitating factors.
- There is an association with other autoimmune diseases.

**Epidemiology**

The true incidence of LS is unknown and is probably underestimated. It is thought to affect around 1 in 1,000 women and 1 in 100,000 men.\[2\]

- Women: there are two peaks of incidence - in prepubertal girls and postmenopausal women.\[3\]
- Men: it is most common in boys aged 9 to 11 years. It also more commonly affects white men after the third decade of life.
- One study has shown that BXO was present in nearly 35% of boys who had a circumcision for phimosis.\[4\]

**Presentation**

The lesions are white thickened patches (porcelain-white papules and plaques). These may progress to crinkled white patches (like cigarette paper). Active lesions may have areas of ecchymosis, hyperkeratosis or bullae.
Women
Symptoms:
- Itch - can be severe and disturb sleep, as it is usually worse at night. This is usually the first symptom.
- Pain can occur if there are fissures or erosions, leading to dyspareunia.
- Perianal lesions are common (about 30%) and may cause constipation.
- May be asymptomatic and found incidentally.

Signs:
- White lesions as above. These may be patchy, or in a figure-of-eight area around the vulva and anus.
- Destructive scarring may cause shrinking of the labia, narrowing of the introitus, or the clitoris may be obscured by adhesions. Genital involvement does not occur; the vagina and cervix are always spared.
- Perianal lesions occur in around 30% of cases.
- In girls, the signs may be mistaken for sexual abuse, as ecchymosis often occurs and can be very striking.

Men
Lesions are usually on the prepuce, glans penis and coronal sulcus.

Symptoms:
- Soreness, haemorrhagic blisters.
- Itching is not usually a common symptom in men.
- Dyspareunia, painful erections due to phimosis.
- If there is meatal scarring, poor urinary stream or dysuria.

Signs:
- White patches on the glans or prepuce.
- Haemorrhagic vesicles or purpura.
- Rarely, blisters or ulcers.
- If scarring has occurred - phimosis, wasting of the prepuce, meatal narrowing/thickening.
- Perianal involvement rarely (if ever) occurs.
- May be asymptomatic.

Other sites
- LS elsewhere is unusual; it is most often on the upper trunk, axillae, buttocks and lateral thighs.
- Oral lesions are extremely rare, but can affect sites where there is cornified stratified squamous epithelium - eg, tongue, gingiva and hard palate.

Diagnosis and investigation
- The diagnosis is usually made clinically.
- Biopsy:
  - Is indicated only when there is diagnostic uncertainty or suspected malignancy.[5]
  - Is not always practical - eg in children. It may be preferable to start treatment and to monitor response.
  - Is essential if lesions do not respond to adequate treatment.

- Swabs are not required routinely but may be necessary in cases where there is erosive disease to exclude infection such as candidiasis or herpes simplex.
- Blood tests:
  - Consider autoimmunity screen and thyroid function tests if symptoms are present. There is no evidence to support testing for autoantibodies without a clinical indication.
Differential diagnosis

- In children, signs may mimic those of child sexual abuse. Note that a diagnosis of LS does not automatically exclude sexual abuse.
- Various other skin, genital or mucosal conditions, including:
  - Vitiligo.
  - Localised scleroderma.
  - Lichen planus.
  - Leukoplakia.
  - Vulval intraepithelial neoplasia.
  - Bowen's disease (squamous cell carcinoma in situ (SCCIS)); if on the penis, this is Queyrat's erythroplasia.
  - Graft-versus-host disease.

Management[3]

Where should patients be treated and followed up?

- LS can be managed by a GP, dermatologist or gynaecologist, depending on local expertise and protocols. Referral may be required to confirm the diagnosis.
- Specialist advice on treatment may be needed - eg, if a woman does not appear to respond to treatment. Vulval clinics and urology clinics have a role, particularly if there are complications. The British Society of Dermatologists suggests follow-up visits at three and six months, to ensure response to treatment and that patients are confident in managing their condition.
- Long-term follow-up is needed for patients with poorly controlled LS.
- Patients who respond well to treatment and need only small amounts of topical steroids should be reviewed annually - this can be in primary care. Give patients clear instructions to report immediately any persistent new lumps, skin changes, erosions or ulcers.

Female anogenital LS

The current evidence demonstrates the efficacy of clobetasol propionate, mometasone furoate, and pimecrolimus in treating genital lichen sclerosus.[6] Clobetasol has been demonstrated to be more effective than pimecrolimus.[7]

A reducing course of clobetasol propionate is the usual treatment:

- The usual regimen is - once-daily (at night) use for one month, alternate nights for one month, then twice-weekly for one month with review at three months.
- If the patient's symptoms return during reduction of treatment, go back up to the frequency that was effective.
- A 30 g tube of clobetasol propionate should last 12 weeks; the patient should then be reviewed.
- If the treatment has been successful, the hyperkeratosis, ecchymoses, fissuring and erosions should have resolved but the atrophy and colour change remain.
- Maintenance treatment may be required - either with less potent steroid preparations or less frequent use of very potent steroids.[8]
- Ointment bases are less allergenic, but the choice of base will depend on patient preference.
- For children, betamethasone dipropionate can be used.
- Between 4% and 10% of women with anogenital LS will have steroid-resistant disease. The recommended second-line treatment is topical tacrolimus or pimecrolimus under the supervision of a specialist clinic.
- Detailed information should be given to patients which includes information about using topical steroids.

Although some studies have demonstrated some benefit of topical tacrolimus and pimecrolimus, long-term safety of these drugs is not established and there are concerns about an increased risk of malignancy with their use in this condition, which already has a premalignant potential. Therefore, these medications should not be used as first-line treatment.[3]

NB: oestrogen or testosterone creams should not be used to treat LS. Testosterone is no better than petroleum jelly and there may be adverse effects. It must not be used in children.

Male anogenital LS

- Use potent topical steroids (eg, clobetasol propionate or betamethasone valerate), applied once-daily until remission, then gradually reduced.
- May need intermittent use (eg, once weekly) to maintain remission.
- Topical mometasone furoate has been used successfully in children with LS of the glans penis.

Other treatments for anogenital LS

- Treat any secondary infection.
- Advice to patients:
  - Wash with bland emollients - eg, aqueous cream; avoid topical irritants and tight clothing; use lubricants if necessary; give details of support groups.
  - Warn patients to seek medical attention if there are possible signs of malignancy, ie if the area develops a persistent lump, change in texture of the skin (such as thickening) or a non-healing ulcer/erosion (see 'Complications and their treatment', below).
  - If relevant, advise which creams/ointments may be used with condoms.
If there is apparent treatment failure, consider:

- Compliance: for example, patients may be deterred by side-effect warnings on steroid preparations; elderly patients may have difficulty applying the creams.
- Whether the diagnosis is correct. There may be an additional problem - eg, infection or allergy to the preparation.
- Whether there is a complication. (See 'Complications and their treatment', below.)

**Extragenital LS**

Shave excision and CO₂ laser have been used successfully, treating symptoms and appearance. UVA-1 is the most successful phototherapy for LS.

**Asymptomatic patients**

Treatment is recommended if patients have features of active disease - eg, ecchymosis, hyperkeratosis or progressive atrophy.

**Complications and their treatment**

- **Scarring:**
  - This is common and may cause urinary symptoms or sexual dysfunction.
  - May require surgery - eg, circumcision, meatal dilatation or vulval surgery.
  - One-stage or staged repairs using oral mucosa grafts are the most recommended procedures for the treatment of LS urethral strictures in men.

- **Constipation due to perianal fissures** - prescribe softening laxatives.

- **Squamous cell carcinoma (SCC):**
  - There is a small risk of SCC of the vulva (2-4% lifetime risk).
  - It has been estimated that between 4% and 8% of men with this condition will develop SCC of the penis. However, it is unclear if BXO itself causes the development of SCC or if it is due to co-existent infection with human papillomavirus.
  - Long-term follow-up is advisable.
  - Warn patients about signs of malignancy and biopsy any suspicious lesions.
  - Extragenital lesions do not appear to have any increased risk of malignancy.

- **Dysaesthesia:**
  - Vulvodynia or penile dysaesthesia can occur following inflammatory conditions of the genitalia. This is a neuropathic type of pain.
  - Topical local anaesthetics (eg, 5% lidocaine ointment) can be given to those with vestibulodynia and vulvodynia or penile dysaesthesia.

- **Sexual dysfunction:**
  - Be aware this may be a complication; offer referral.

**Prognosis**

- Symptom remission can be achieved in 98% of compliant and 75% of non-compliant women by using potent topical steroids.
- In males, a course of steroids may prevent the need for circumcision.
- Most men are either cured by topical treatment with ultrapotent steroid (50-60%) or by circumcision (>75%).
- However, in some cases LS recurs. More complex cases may be an overlap syndrome with lichen planus, and may be more difficult to treat.

**Further reading & references**

5. The Management of Vulval Skin Disorders; Royal College of Obstetricians and Gynaecologists (February 2011)
9. Guidelines for the management of lichen sclerosus; British Association of Dermatologists (2010)

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