Lichen sclerosus

Kate Dalziel, Sarah Shaw

Lichen sclerosus is a chronic inflammatory skin disease usually involving the anogenital skin. It is most common in postmenopausal women but occurs in both sexes at all ages. The cause is unknown, but lichen sclerosus is strongly associated with autoimmune disorders, particularly thyroid disease, in almost 30% of patients.

Why is it missed?
Lack of familiarity with the condition and failure to examine the genital skin properly can lead to long delays in diagnosis. Reticence and embarrassment on the part of patient and doctor may hinder the taking of a full history and examination. Common mistaken diagnoses in women are candida infection and postmenopausal vulval atrophy. Candida vulvovaginitis is usually confined to women of childbearing age and is unusual in older women unless there are additional risk factors such as diabetes. The delay to diagnosis in one case series of 327 female patients was 2.2 years in children and 5.3 years in adults.

Lack of familiarity with the condition probably explains why lichen sclerosus is rarely diagnosed as a cause of severe constipation in girls. Terminology has been confusing in the past, when terms such as vulval dystrophy and balanitis xerotica obliterans have been used instead during invasive or 64 slice (or more) computed tomography coronary angiography. Reversible myocardial ischaemia is found during non-invasive functional imaging.

Overcoming barriers
This guideline will enhance the understanding of the clinical assessment and investigation of patients with suspected acute coronary syndrome. Healthcare professionals should particularly note the recommendations for urgent referral for hospital assessment and the timing of diagnostic testing and ensure these can be implemented. In patients with suspected stable angina, the guideline emphasises the importance of a detailed clinical history to inform an initial triage of whether pain might be cardiac in origin, and mechanisms should be in place to ensure that this is accurately recorded. In patients with non-anginal pain, further diagnostic testing is not generally recommended. In patients with symptoms of typical or atypical angina who require further diagnostic testing, the estimated likelihood of coronary artery disease will determine which tests to use. Healthcare providers should ensure appropriate and timely access to high quality diagnostic testing and interpretation and have systems in place to audit their use.

Contributors: AC, AT, and JS contributed to a first draft that followed the template agreed by the BMJ and NICE and they shared in the subsequent editing. AC is guarantor.

Funding: The National Collaborating Guideline Centre for Acute and Chronic Conditions was commissioned and funded by the National Institute for Health and Clinical Excellence to write this summary.

Competing interests: All authors have completed the Unified Competing Interest form at www.icmje.org/coiDisclosure.pdf (available on request from the corresponding author) and declare that (1) AC, AT, and JS have support from NICE for work on guideline development. AC works in the National Collaborating Guideline Centre for Acute and Chronic Conditions funded by NICE. JS receives travel expenses to attend guideline development group and other relevant meetings and payment to her NHS trust to fund her time as clinical adviser for the guideline. AT has support from an NIHR biomedical research unit grant to fund the cardiovascular imaging department; (2) AC, AT, and JS have had no relationships with companies that might have an interest in the submitted work in the previous three years; (3) their spouses, partners, or children have no financial relationships that may be relevant to the submitted work; and (4) they have no non-financial interests that may be relevant to the submitted work.

Provenance and peer review: Commissioned; not externally reviewed.


bmj.com: recent summaries of NICE guidance
- Reducing the risk of venous thromboembolism in patients admitted to hospital (2010;340:c95)
- Depression in adults, including those with a chronic physical health problem (2009;339:b4108)
- When to suspect child maltreatment (2009;339:b2689)
- Early management of persistent non-specific low back pain (2009;338:b1805)

EASILY MISSED?

Lichen sclerosus
of lichen sclerosus. The introduction of internationally agreed terminology and the advent of multidisciplinary vulval clinics have improved the recognition of lichen sclerosus and the use of correct nomenclature.

**Why does this matter?**
Lichen sclerosus is highly symptomatic. Vulval, penile, and perianal disease can cause severe intractable itching and soreness. Scarring may follow, leading to loss of vulval anatomy, clitoral phimosis, and vulval stenosis in women. Sexual function may be severely impaired. Phimosis and anterior urethral obstruction can occur in men and boys. The development of squamous cell carcinoma on genital lichen sclerosus is a well recognised complication in both sexes and is estimated to occur in about 5% of women. Sexual abuse is sometimes suspected in children with lichen sclerosus and, although the condition does not exclude abuse, which must always be considered, this has led to mistaken diagnosis.

**How is it diagnosed?**

**Clinical features**

**Adult women**
The commonest presenting symptom of lichen sclerosus is severe vulval itching, and many patients will already have self medicated with over the counter antifungal or anti-itch creams. Typical changes of lichen sclerosus include areas of white skin that may be small and numerous or confluent over larger areas affecting the labia minora, labia majora, and adjacent skin of the perineum and groin creases. Perianal disease is common, giving a “figure of eight” pattern of affected skin around the vulva and perianal region. The white skin often looks thin, wrinkled and fragile, with red or purple areas of bleeding into the skin. Excoriations are common. Sometimes there are thickened plaques or even warty areas. In a series of 253 women with lichen sclerosus, 177 had atrophic change and 94 had hyperkeratotic (warty) areas.

In the patients with scarring (about 70% of women) symptoms can include difficulty in passing urine, especially in men, and in women, dyspareunia, apareunia and splitting of the skin on attempted intercourse. Women may notice the change in their vulva with loss of the labia minora (fig 1) and fusion of the labia over the clitoris. Occasionally the first presentation of lichen sclerosus will be when squamous carcinoma develops: a plaque, ulcer or nodule can arise and enlarge very quickly, sometimes in a matter of weeks.

**Adult men**
Itching and soreness of the glans and prepuce are common. White areas and purpura can be seen on the glans and foreskin but may be hidden by a tight phimosis (fig 2). Squamous cell carcinoma can arise in the affected area, with features similar to those described above for women.

**Children**
Prepubertal girls with lichen sclerosus often present with vulval and perianal itching and show the typical skin changes as described for adult women, but they may also present with intractable constipation, soiling, anal fissures, and bleeding. Boys tend to present with phimosis, and this may hide the typical skin changes.

**Differential diagnosis**
Other chronic itchy skin diseases need to be considered. Various types of eczema that affect the perineal and perianal areas, such as contact dermatitis and seborrhoeic eczema, are common. Another auto-immune skin condition, lichen planus, can also show a predilection for genital skin. It too may cause scarring and squamous carcinoma. Plaques of vulval intraepithelial neoplasia may be white, but the other typical features of lichen sclerosus will be absent.

---

**Fig 1 | Vulval lichen sclerosus showing white plaques and scarring with loss of the labia minora**

**Fig 2 | Lichen sclerosus of glans penis in adult showing whiteness and purpura**
Lichen sclerosus is often diagnosed by the clinical appearance, but if uncertainty exists then it is essential that a biopsy is done before very potent topical steroids are used. Check thyroid function, and remember that a patient with lichen sclerosus is also more likely to have other autoimmune disorders.

Consider using topical steroids to treat phimosis caused by lichen sclerosus as this may avoid the need for circumcision.

Advise patients about the small (5%) risk of malignant change and to report immediately any change, such as an ulcer or lump.

Investigations

No diagnostic biochemical or immunological investigations exist, but a skin biopsy will often provide the diagnosis. The histology of lichen sclerosus is distinctive, and biopsy will help to differentiate it from other skin diseases that cause anogenital itching and scarring. However, use of a very potent topical steroid (such as clobetasol propionate) may completely correct the clinical and histological changes, so such a steroid must not be used before a definitive diagnosis is made. Accurate diagnosis is important because patients with lichen sclerosus will need follow-up and education about ongoing treatment and the risk of cancer development.

Check thyroid function in all adult patients with lichen sclerosus, given the association with autoimmune thyroid disease.

How is it managed?

Evidence based guidelines for the management of lichen sclerosus state that if clinical doubt exists then a biopsy should be performed to confirm the diagnosis. The general practitioner may wish to refer the patient to a specialist with an interest in vulval disease or to an established vulval clinic (based in dermatology, genitourinary medicine, or gynaecology departments). Pending the definitive diagnosis, emollients can be used as soap substitutes and to help repair the skin barrier, and a moderate potency topical steroid such as clobetasol butyrate ointment can be used sparingly. After the definitive diagnosis is made, accurate diagnosis is important because patients with lichen sclerosus will need follow-up and education about ongoing treatment and the risk of cancer development.

Check thyroid function in all adult patients with lichen sclerosus, given the association with autoimmune thyroid disease.

Always take a careful history

Unusually for Lyme Regis, on England’s south coast, there had been a severe episode of heavy snow that lasted for several days and I was snowbound at home. I lived a mile or so from the town centre, and my practice, at the top of a steep hill. Everything was at a standstill, and I could not get my car on the road.

On the third day I was surprised to see that the milkman had arrived. He had managed to obtain chains to fix to the wheels of his van. He told me of the difficulties he had encountered on his round and mentioned that “poor old Mrs Jones has had water trouble.” She was one of my patients, so I donned suitable clothing and boots, grabbed my medical case, and trudged and slithered the mile down hill in a blizzard to poor old Mrs Jones’s house.

On my arrival, Mrs Jones welcomed me with great surprise: “Fancy you turning out on a day like this, doctor. Do come in from the cold.” I explained that the milkman had advised me of her water trouble. She nodded and pointed up to the ceiling, where a large patch of damp was to be seen. However, the plumber was on his way, so I commiserated, and we had a cup of tea and a chat. I then trudged back up the hill, treating some children on the way by joining in with a snowball fight.

Alexander Fernandez retired general practitioner, Lyme Regis alexfernandez33@hotmail.com

Cite this as: BMJ 2010;340:c418