Lichen Sclerosis in Boys
by Karl Becker

SUMMARY

Background: Lichen sclerosus (LS) is a sclerosing skin disease. When it appears in boys, it nearly always affects the penis and usually causes phimosis requiring surgical treatment. The clinical significance of this disease in boys is inadequately recognized.

Methods: The etiology, clinical manifestations, diagnosis, and treatment of LS in boys are presented in the light of a review of selected literature. We also present our own experience with this disease in an ambulatory pediatric surgery practice.

Results: LS has long been recognized as a disease of the prepubertal male genitalia (in such cases, the condition is also called “balanitis xerotica obliterans”). It is thought to be the main cause of acquired phimosis, and it can also involve the meatus and urethra as it progresses. Its possible association with squamous cell carcinoma of the penis remains unclear. Its etiology is unknown; its pathophysiological mechanism involves T-lymphocyte-mediated inflammation. The treatment of choice is complete circumcision. There is still controversy regarding the conservative treatment of LS with topical steroids.

Conclusion: LS is much more common in boys than is generally assumed. Lichen sclerosus should be suspected in any case of acquired phimosis. Treatment with complete circumcision does not necessarily bring about a definitive cure. Further research on the pathogenesis of this disease is needed.

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that the foreskin had previously been retractable and had then become non-retractable (secondary phimosis). No patient had LS on any part of the body other than the penis. Among the affected boys were three pairs of identical twins and one pair of non-twin brothers. 169 of 225 patients received clinical follow-up.

Primary involvement of the meatus with clinically relevant stenosis was present in 6 boys (2.7%). In 18 cases (10.7%), clinically relevant meatal stenosis requiring surgery was present after the lichenoid changes had healed (in general, the frequency of meatal stenosis after circumcision without LS is less than 1%). Among 10 patients who underwent partial circumcision, five (50%) suffered a recurrence.

There was only one recurrence after total circumcision. This patient was an obese boy with a so-called buried penis. The skin around the penis developed lichen sclerosus after circumcision, leading to recurrent phimosis.

The anterior portion of the urethra was found to be involved in a single case.

In all of the cases that were followed up clinically, complete healing was observed.

**Epidemiology**

Systematic studies have shown that 10% to 40% of all surgically treated cases of phimosis are due to LS (6, 8). The corresponding percentage among the author’s own patients was 15%. Assuming that 1% of boys develop phimosis requiring surgery after they have outgrown the stage of physiological phimosis (e1), we can estimate the prevalence of prepubertal LS among all boys in the German population at 0.1% to 0.4%. On the other hand, a large-scale epidemiological study that was performed at an army hospital in Texas yielded a prevalence figure of 0.07% among boys under age 10 (9). This value seems relatively low, perhaps because boys in the USA are commonly circumcised shortly after birth, and perhaps because of a selection effect of the military environment (10). The prevalence of LS in girls has been estimated at 0.1% to 0.3% (7, 10). Thus, close analysis reveals that the condition is about equally common in boys and girls (11, e2). As there have been no large-scale epidemiological studies to date of persons of both sexes with LS, the condition is often discussed—particularly in the dermatological literature—as if it predominantly affected girls and women (10, 12).

LS is generally considered responsible for most cases (80% to 90%) of acquired (secondary) phimosis. The author’s own findings bear this out as well. Thus, any case of secondary phimosis in childhood should arouse the clinical suspicion of LS (6, 8).

The reported frequency of involvement of the meatus and urethra varies from 2% to 40% and depends on the severity of disease course and, above all, on the duration of the sclerosing process before adequate treatment is begun (13, 14, e3).

LS is considerably more common in boys with hypospadias (5, 13, 15).
**Etiology**

Lichen sclerosus is a lymphocyte-mediated chronic inflammatory disease of the skin. There is increasing evidence for an underlying autoimmune mechanism (12, 16). An infectious cause has often been postulated, but never proved.

There have been multiple reports of co-occurrence of lichen sclerosus with autoimmune diseases including vitiligo, Hashimoto’s thyroiditis, and type 1 diabetes (17).

The twin age peaks (prepubertal and postmenopausal) for LS in women suggest a hormonal influence on the disease, but attempts at hormonal therapy have been abandoned after failing to yield any clear benefit (18).

The association of LS with HLA-DQ7 and the appearance of familial cases (7, 17) imply a genetic predisposition to LS in female patients. The literature contains little evidence of a familial predisposition to LS in boys (13). LS in (female) twins has been reported, and, as described above, the author has treated three pairs of identical twin boys and one pair of non-twin brothers with LS. It is noteworthy that the disease tends to manifest itself simultaneously, or nearly so, in affected siblings (17).

In boys, the warm and moist local environment under the foreskin seems to promote the development of LS (13, 19).

**Clinical manifestations**

The clinical examination on initial presentation often already reveals a whitish, porcelain-like sclerotic scarring of the distal portion of the prepuce, causing progressive phimosis and typically appearing as a whitish ring (Figure 2) (8). In many boys, however (23.8% in the author’s series), the disease cannot be seen on clinical examination (6). The affected boys rarely complain of symptoms; in severe cases, there may be dysuria. In the author’s series, the disease was generally present for about three to six months. It progresses at a variable rate. Some patients can recall having had a small injury or infection at the onset of the condition; this is consistent with Köbner’s phenomenon, in which an influence of this kind serves as a trigger for the inflammatory process of LS (17).

At surgery, the foreskin is generally found to be thick and coarse. Inspection after dorsal incision of the foreskin often reveals a surprising picture, as the degree of external involvement does not reliably predict the degree of involvement of the glans. In addition to the preputial changes, one nearly always finds mother-of-pearl-colored perifrenular involvement of the glans, causing a sclerotic frenulum breve, with a greater or lesser degree of spread in the direction of the meatus (Figure 3). This implies that the process begins near the frenulum, rather than near the meatus (20). Often, the lips of the meatus already display delicate sclerotic changes, but meatal stenosis is rare in the initial phase (6, 13). In clinically severe cases, which do not necessarily go hand in hand with marked histopathological changes (6), the inner surface of the prepuce is involved.
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all the way to the sulcus, and the glans is completely covered by a white stratum corneum.

Mild meatal stenosis often develops postoperatively within four months. Functionally relevant meatal stenosis arose in 10.7% of the author’s patients who were circumcised because of LS (Figure 4). Bale et al. reported on a comparable percentage of postoperative meatal stenosis (5), which they considered to be a specific sequela of the disease (e4), while Kiss did not observe any meatal stenosis in 471 patients with LS (6).

It is worthy of note that meatal stenosis can arise years afterward, even in the absence of demonstrably persistent or recurrent lichen sclerosus (e5). This is perhaps explained by lessened perfusion of the meatus resulting from sclerosis of the frenulum region (e6).

In the author’s experience, progressive involvement of the urethra occurs only in a minority of boys, and only when the disease has been present for a good deal longer than one year (14). Among men aged 30 and older, squamous cell carcinoma (SCC) of the penis is a rare finding (3% to 4%) in the disease’s later course (21, e7). It is unclear whether, and to what extent, lichen sclerosus in childhood might increase the risk of later development of SCC (22). Human papilloma virus 16 (HPV-16) has been postulated as a possible precipitating factor (23); if this is confirmed, the consequence might be a recommendation for vaccination of all LS patients against HPV.

Treatment

On the basis of a study on the topical treatment of phimosis (e8), a British guideline recommends the initial treatment of boys with LS with a highly potent corticosteroid (18). In the study concerned, however, not one patient with LS responded to conservative treatment. Likewise, the prospective randomized double-blind trial of Lindhagen on the conservative treatment of phimosis revealed that the patients who did not respond to treatment had lichen sclerosus (25).

In a small number of studies dealing specifically with LS, improvement after treatment with clobetasol was reported, depending on the stage of the disease (24, 25). The available data remain inadequate to support the efficacy of any particular type or duration of conservative treatment. In the author’s experience, however, conservative, topical treatment with clobetasol has been effective in very early cases of LS. These boys need meticulous long-term follow-up, as their risk of recurrent LS is unknown. Thus, until randomized controlled studies with long-term results are available, conservative treatment should be provided only in carefully selected cases by physicians with adequate experience in the treatment of LS.

Total circumcision yields the most durable clinical benefit and is usually curative (6, 8, 13, 15). Whenever there is the least suspicion of LS on the basis of the history or physical examination, the resected foreskin must be examined histopathologically. Partial circumcision, leaving residual foreskin behind, is often followed by recurrent disease (in 50% of the author’s cases). On the other hand, recurrence is very rare after total circumcision: it occurred in only one of the author’s cases, an obese boy with buried penis due to the large amount of subcutaneous fat in the pubic area, like the patients described by Gargollo and Depasquale (13, 19) (Figure 5).

If the postoperative follow-up examination reveals clinically relevant meatal stenosis (uroflowmetry with a plateau phase of less than 10 mL/sec), meatotomy is indicated. In order to avoid the risk of recurrence, however, meatotomy should only be performed after lichen sclerosus is completely healed (14). Bougie dilatation of the meatus is less effective (13).

Lichen sclerosus in girls

The main aspects of lichen sclerosus in girls will be described briefly here to round out the discussion of the disease and to highlight the differences in its appearance in girls and boys.
The lichenoid changes begin on either the vulva or the anus (Figure 6) and then become confluent to form a perineal figure eight. When the diagnosis is in doubt, histopathological examination is helpful. The question of potential sexual abuse requires careful attention. On the one hand, it is not rare for lichen sclerosus to be misdiagnosed as evidence of sexual abuse. On the other hand, sexual abuse does promote the development of LS through the isomorphic effect (the Köbner phenomenon).

Extragenital involvement is somewhat more common in girls than in boys, and the correlation with autoimmune disease is stronger. Symptoms can arise in the form of itching, local wound pain, and difficulty defecating due to anal fissures, leading to constipation (18).

A course of ultra-high-potency corticosteroid therapy (clobetasol propionate 0.05%) for two to three months is successful in over 90% of cases (22). Afterward, milder corticosteroid ointments (or, alternatively, calcineurin antagonists or UVA phototherapy) and local care are provided in an attempt to achieve a prolonged remission (18, e9, e10).

Although the condition does improve spontaneously at puberty in a small number of cases, the current evidence indicates that LS in girls is a chronic disease. Thus, a properly functioning physician-patient relationship, with due attention to all of the relevant psychosocial aspects, is very important over the course of the disease. Centers for the treatment of girls with LS, as have been developed by Powell et al., would be a welcome development in Germany as well (7).

**Overview**

In summary, LS is still underappreciated as a cause of (usually acquired) phimosis in boys, even though its importance in this respect was already recognized 50 years ago (4–6, 21). Only 10% of the author’s patients came to be treated with the suspected diagnosis of LS, even though the disease was clinically evident in 80%. The treatment of boys with this condition is very successful when compared to that of female patients or of adult males.

LS is a chronically progressive process. Early diagnosis and treatment lessen the risk of recurrence and of meatal and urethral involvement (19) and can even prevent the need for surgery in rare cases. The fact that the process begins near the frenulum suggests that it has a relevant vasculitic component (16).

Long-term studies will be needed to show whether boys with LS stand to profit from vaccination against HPV (e9).

It would be desirable to have an authoritative statement of the current medical-scientific understanding and observations regarding lichen sclerosus. The creation of a German guideline would be a welcome first step in this direction.

The author would like to supplement the published protocol of Depasquale (13) with the following further recommendations:

- Lichen sclerosus (LS) is a progressive inflammatory disease of the skin that leads to phimosis in boys. It can be cured by total circumcision.
- The clinical suspicion of LS should be aroused, in particular, by any case of acquired phimosis.
- Potential postoperative sequelae include meatal stenosis and, after partial circumcision or in obese patients, recurrent LS.
- LS in girls is a chronic disease that is treated topically with immunosuppressive or immune-modulating agents.
- Definitive establishment of the diagnosis by histopathological examination should be a goal in every case of suspected LS.

- Cicatricial phimosis that appears in childhood and arouses the suspicion of LS should be presented to a pediatric surgeon or urologist who is familiar with the clinical picture of this disease.
- Acquired phimosis always demands special attention.
- If LS is clinically suspected, total circumcision is the treatment of choice.
- Any slight changes that may be visually apparent at surgery, e.g., in the perifrenular area, necessitate histological examination of the resected foreskin.
- The affected boys should have long-term clinical follow-up and, when indicated, topical treatment after surgery.
- Suspected meatal stenosis should be objectified by uroflowmetry.
- Meatotomy should be performed only after the lichenoid changes have fully resolved in order to avoid recurrence.
- The child’s parents should be informed about the nature of the disease and reassured that its overall course is favorable.

**Conflict of interest statement**

The author declares that he has no conflict of interest as defined by the guidelines of the International Committee of Medical Journal Editors.

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**REFERENCES**


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