Treatment and follow-up of genital lichen sclerosus in male children: multidisciplinary management at a tertiary care center

Alessia Paganelli,[1] Paolo Viscardo Fabbri,[2] Filippo Ghidini,[3] Laura Bigi,[1] Claudia Lasagni,[1] Pier Luca Ceccarelli[3]

[1]Section of Dermatology, Rare Diseases Outpatient Clinic, Modena; [2]Department of Anatomic Pathology; [3]Department of Pediatric Surgery, Modena University Hospital, Italy

Dear Editors,

Lichen sclerosus (LS) is a chronic inflammatory disorder affecting mostly – but not exclusively – the genital area.[1] Old-fashioned pseudonyms include balanitis xerotica obliterans and kraurosis vulvae.[2] LS is typically characterized by the presence of hypopigmented atrophic plaques, often accompanied by itching and/or pain.[3] Other possible LS-associated signs include genital discharge, bruises, erosions and scarring. Possible complications encompass urinary and/or sexual dysfunction and the occurrence of squamous cell carcinomas.[1,3] Known risk factors for LS include familial history, metabolic syndrome and concomitant autoimmune disorders (*e.g.* alopecia areata, autoimmune thyroiditis).[4,5] The diagnosis is mostly clinical; however, histological confirmation is sometimes required.[6] LS-specific dermoscopic criteria have also recently been proposed for LS.[7] Possible therapeutic strategies include emollients, topical corticosteroids (CS) and/or topical calcineurin inhibitors. Systemic immunosuppression is only required in more severe and/or refractory cases.[8] LS is classically described as being more frequent in women and having two main peaks of incidence, in prepubertal age and late adulthood.[1] LS was also traditionally considered to be a rare disease (*Supplementary Table 1*),[2] despite current evidence suggesting such LS prevalence to possibly be underestimated, especially in male subjects and/or children.[9,10] The aim of the present paper is to give an insight in the impact of LS on the male pediatric population, through retrospective evaluation of the casuistry collected at a tertiary referral center in the last 10 years. Only male subjects, aged 18 or under, histologically diagnosed with LS between 2013 and 2023 were included. Patients without at least one dermatological and/or urological follow-up visit were excluded from the study population.

In total, 187 cases were considered in our study (Table 1). Mean follow-up duration was 1.9 years (range 2 weeks to 8.2 years). Mean age at diagnosis was 10. Affected areas mainly included the foreskin (Figure 1), the external urethral orifice (EUO) and the glans penis. No perianal or perineal involvement was recorded. Approximately 90% of the patients only presented with phimosis, in the absence of further disease localization. Complete remission of genital LS after circumcision was achieved in the majority of cases even in the complete absence of systematic application of any topicals (Table 1). Only one patient experienced post-operative scar contracture of the corona of glans penis, which subsequently resolved with topical CS therapy. Active LS was present in the glans area in 12 subjects (6.4%) in the form of erythematous (n=5), hypochromic/achromic (n=3) or dyschromic (n=4) plaques. These subjects were commonly prescribed intermittent courses of topical CS. Significant EUO stenosis, with clinical indication to undergo urological intervention (either meatoplasty, meatotomy or meatal dilation), was detected in 8 cases (4.2%). None of these children recurred after surgical treatment and chronic topical steroidal therapy was not needed in most cases (75%). Despite clinically relevant EUO stenosis being quite uncommon, EUO was found to have reduced dimensions (diameter of approximately 1-2 mm) in 29 cases (15.5%) during dermatological FUP. These patients were also referred to the pediatric urology department for specific evaluation but did not show any urological and/or urodynamic complication. A significant bias in the interpretation of our results resides in the lack of systematic histological analysis of foreskin specimens after circumcision. In fact, only around one third of the circumcision specimens were sent for pathological examination at our center. Of these, LS turned out to account for approximately two thirds of the diagnoses (60.85%). Our data therefore possibly still underestimate LS real incidence, suggesting systematic foreskin histological examination could be necessary in order to gain more precise estimates of real LS incidence in the pediatric population. Not only the presented results clearly indicate a shift from the classical paradigm considering LS as a morbid condition affecting prevalently post-menopausal women, but also suggest a prevalence of LS in male children far above the threshold for being classified as a rare disease (>1/2000, see *Supplementary Table 1*). In fact, LS appears to be relatively

Correspondence: Alessia Paganelli, Dermatology Unit, Arcispedale Santa Maria Nuova, viale Risorgimento 80, 42123 Reggio Emilia (RE), Italy. E-mail: alessia.paganelli@ausl.re.it

Key words: lichen sclerosus, skin inflammation, rare cutaneous disease, dermatologic surgery, pediatric dermatology.

Contributions: the authors contributed equally.

Conflict of interest: the authors declare no potential conflict of interest.

Data availability: data are available from the authors upon reasonable request.

Ethical approval and consent to participate: the patients in this manuscript have given written informed consent to publication of their case details. All the procedures described were performed in accordance with the principles of the Helsinki Declaration.

Received: 19 June 2023.
Accepted: 2 July 2023.

Dermatology Reports 2023; 15:9774
doi:10.4081/dr.2023.9774

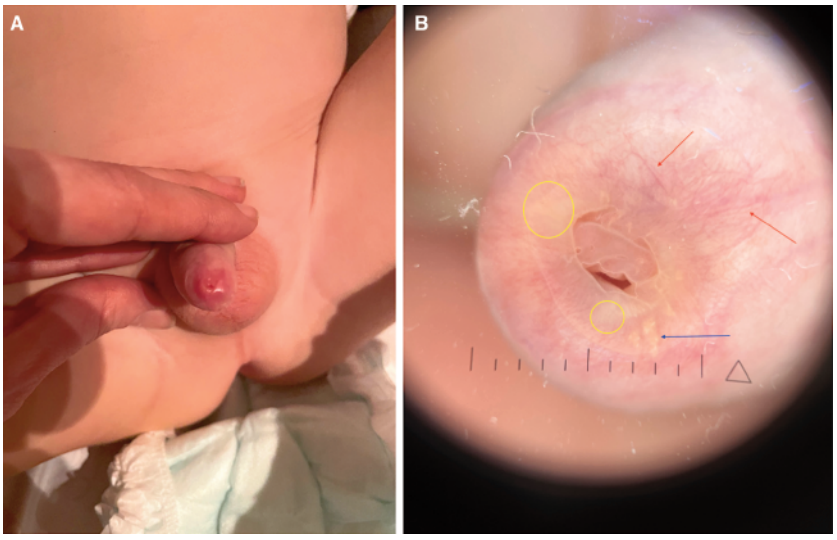

Figure 1. Clinical (**A**) and dermoscopic (**B**) images of genital lichen sclerosus in a 2-year-old boy. Clinically, erythema, skin stiffness and a shiny appearance of the preputial area can be observed. White structureless areas (yellow circles), linear vessels (red arrows) and white chrysalis-like structures (blue arrow) can be observed in dermoscopy.

common in males in the pediatric population, confirming previous literature possibly being biased by misdiagnosis of the disease.[6,9] Moreover, we also observed chronic CS therapy not to be required after circumcision and/or meatal stricture surgical treatment. These findings suggest surgery to possibly be the first-choice therapeutic option for an inflammatory disorder.[2,9,10]

The availability of definitive curative strategies implicitly call into doubt the definition of LS as a chronic disorder, changing the way not only patients and their doctors, but also healthcare systems, should conceive this peculiar disease. However, longer-term follow-up of these patients and broader casuistries are needed to confirm our results.

Table 1. Pediatric patients histologically diagnosed of lichen sclerosus (LS) at our center between January 2013 and January 2023. Results are indicated either as absolute counts (n) and percentages (%). Topical therapy is here not stratified for the type of LS localization.

Patient characteristics	N	%
Phimosis		
Yes	185	98.9
No	2	1.1
Glans		
Erythematous	5	2.7
Hypochromic	3	1.6
Dyschromic	4	2.1
External urethral orifice		
Normal	150	80.2
Reduced	29	15.5
Stenosis	8	4.2
Topical therapy		
None	75	40.1
Emollients/vit E only	31	16.6
CS as needed only	29	15.5
Emollient/vit E + CS as needed	48	25.7
Prolonged CS*	4	2.1

vit E, vitamin E; CS, corticosteroid. *Patients requiring prolonged CS therapy included: 1 post-surgical stenosis of corona glans penis, 2 external urethral orifice stenosis, 1 glans involvement.

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Supplementary Material.
Table 1. A) List of the local and national registries used for the assessment of epidemiological data. B) Preliminary epidemiological data.