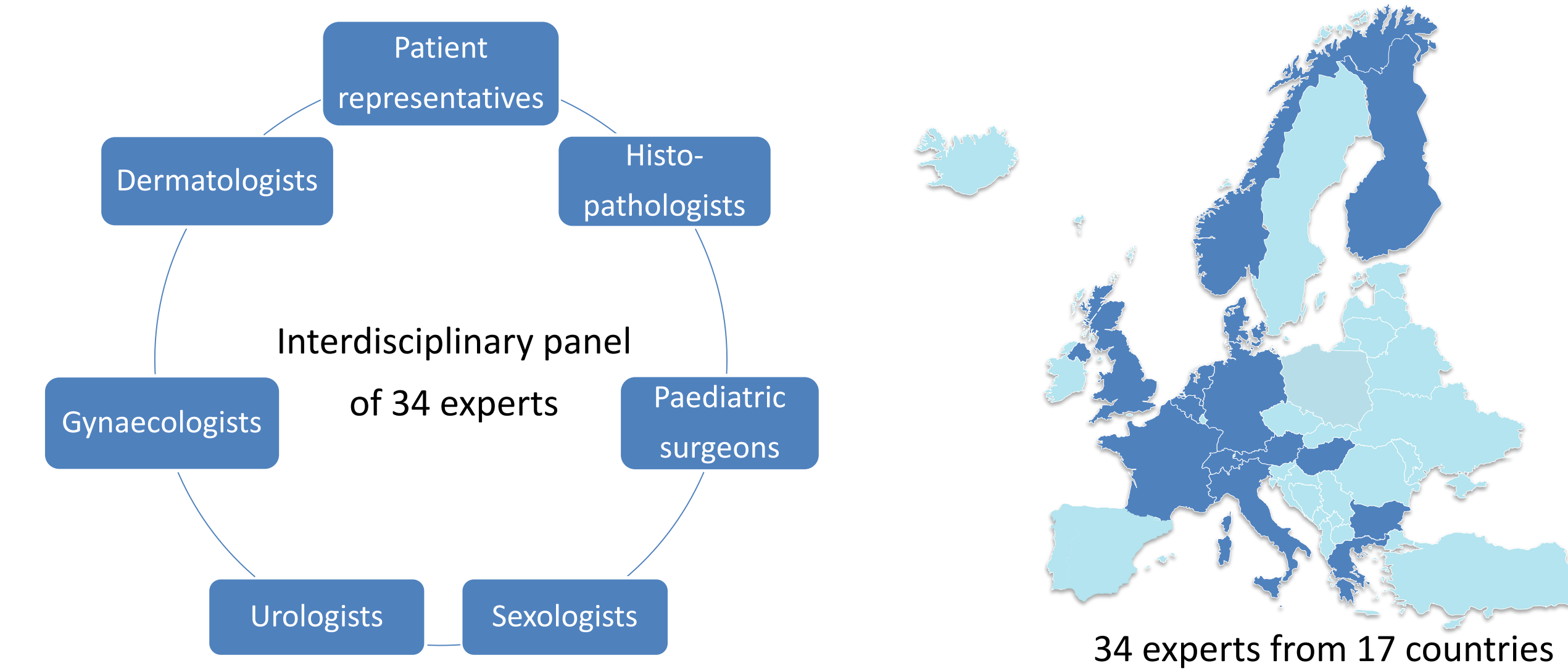


EuroGuiDerm Guideline for Lichen sclerosis

G Kirtschig¹, M Kinberger², A Kreuter³, R Simpson⁴, A Günther⁵, C van Hees⁶, K Becker⁷, MJ Ramakers⁸, M Corazza⁹, S Müller¹⁰, S von Seitzberg¹¹, MJ Boffa¹², R Stein¹³, G Barbagli¹⁴, CC Chi^{15,16}, JN Dauendorffer¹⁷, B Fischer¹⁸, M Gaskins², E Hiltunen-Back¹⁹, A Höfinger¹⁸, NH Köllmann¹⁸, H Kühn²⁰, HK Larsen²¹, M Lazzeri²², W Mendling²³, AF Nikkels²⁴, M Promm²⁵, KK Rall²⁶, S Regauer²⁷, M Sárdy²⁸, N Sepp²⁹, T Thune³⁰, A Tsiogka³¹, S Vassileva³², L Voswinkel²⁰, L Wölber³³, RN Werner²

1 Medbase Health Centre, Frauenfeld, Switzerland. 2 Department of Dermatology, Venereology and Allergology, Division of Evidence-Based Medicine (dEBM), Charité - Universitätsmedizin Berlin, corporate member of Freie Universität Berlin and Humboldt-Universität zu Berlin, Berlin, Germany. 3 Department of Dermatology, Venereology, and Allergology, HELIOS St. Elisabeth Hospital Oberhausen, Oberhausen, Germany. 4 Centre of Evidence Based Dermatology, University of Nottingham, Nottingham, UK. 5 Gynäkologisches Tumorzentrum St. Anna, Lucerne, Switzerland. 6 Department of Dermatology, Erasmus University Medical Center, Rotterdam, the Netherlands. 7 Office for Paediatric surgery, Bonn, Germany. 8 CenSeRe (Centre for Psychological, Relational, Sexual Health), Voorschoten, the Netherlands. 9 Section of Dermatology and Infectious Diseases, Department of Medical Sciences, University of Ferrara, Ferrara, Italy. 10 Department of Dermatology, University Hospital Basel, Basel, Switzerland. 11 The Danish Lichen Sclerosus Association, Denmark. 12 Department of Dermatology, Mater Dei Hospital, Msida, Malta. 13 Center for Pediatric, Adolescent and Reconstructive Urology, Medical Faculty Mannheim, University of Medical Center Mannheim, Heidelberg University, Mannheim, Germany. 14 Centro Chirurgico Toscano, Arezzo, Italy. 15 Department of Dermatology, Chang Gung Memorial Hospital, Linkou, Taoyuan, Taiwan. 16 College of Medicine, Chang Gung University, Taoyuan, Taiwan. 17 Department of Dermatology, Centre for genital and sexually transmitted diseases, University Hospital Saint Louis, Paris. 18 The Swiss Lichen Sclerosus Association, Switzerland / Verein Lichen Sclerosus e.V., Switzerland. 19 Department of Dermatovenereology, Helsinki University Hospital, Helsinki, Finland. 20 The German Lichen Sclerosus Association, Germany. 21 Department of Dermatology and Venereology, Copenhagen University Hospital, Bispebjerg Hospital, Copenhagen, Denmark. 22 Department of Urology, IRCCS Humanitas Research Hospital, Rozzano (MI), Italy. 23 German Center for Infections in Gynecology and Obstetrics, at Helios University Hospital Wuppertal- University Witten/Herdecke, Germany. 24 Department of Dermatology, University Medical Center of Liège, Liège, Belgium. 25 Department of Paediatric Urology and Clinic St. Hedwig, University Medical Centre of Regensburg, Regensburg, Germany. 26 Department of Women's Health, Women's University Hospital Tuebingen, Tuebingen, Germany. 27 Diagnostic and Research Institute of Pathology, Medical University Graz, Graz, Austria. 28 Department of Dermatology, Venereology and Dermatocology, Semmelweis University, Budapest, Hungary. 29 Department of Dermatology and Venereology, Ordensklinikum Linz Elisabethinen, Linz, Austria. 30 Department of Dermatology, Haukeland University Hospital, Bergen, Norway. 31 National and Kapodistrian University of Athens, Faculty of Medicine, 1st Department of Dermatology-Venereology, Andreas Sygros Hospital, Athens, Greece. 32 Department of Dermatology and Venereology, University Hospital "Alexandrovska", Medical University - Sofia, Sofia, Bulgaria. 33 Department of Gynaecology, University Medical Centre Hamburg-Eppendorf and Centre for Colposcopy and Vulvovaginal disease Jersusalem Hospital Hamburg, Hamburg, Germany.



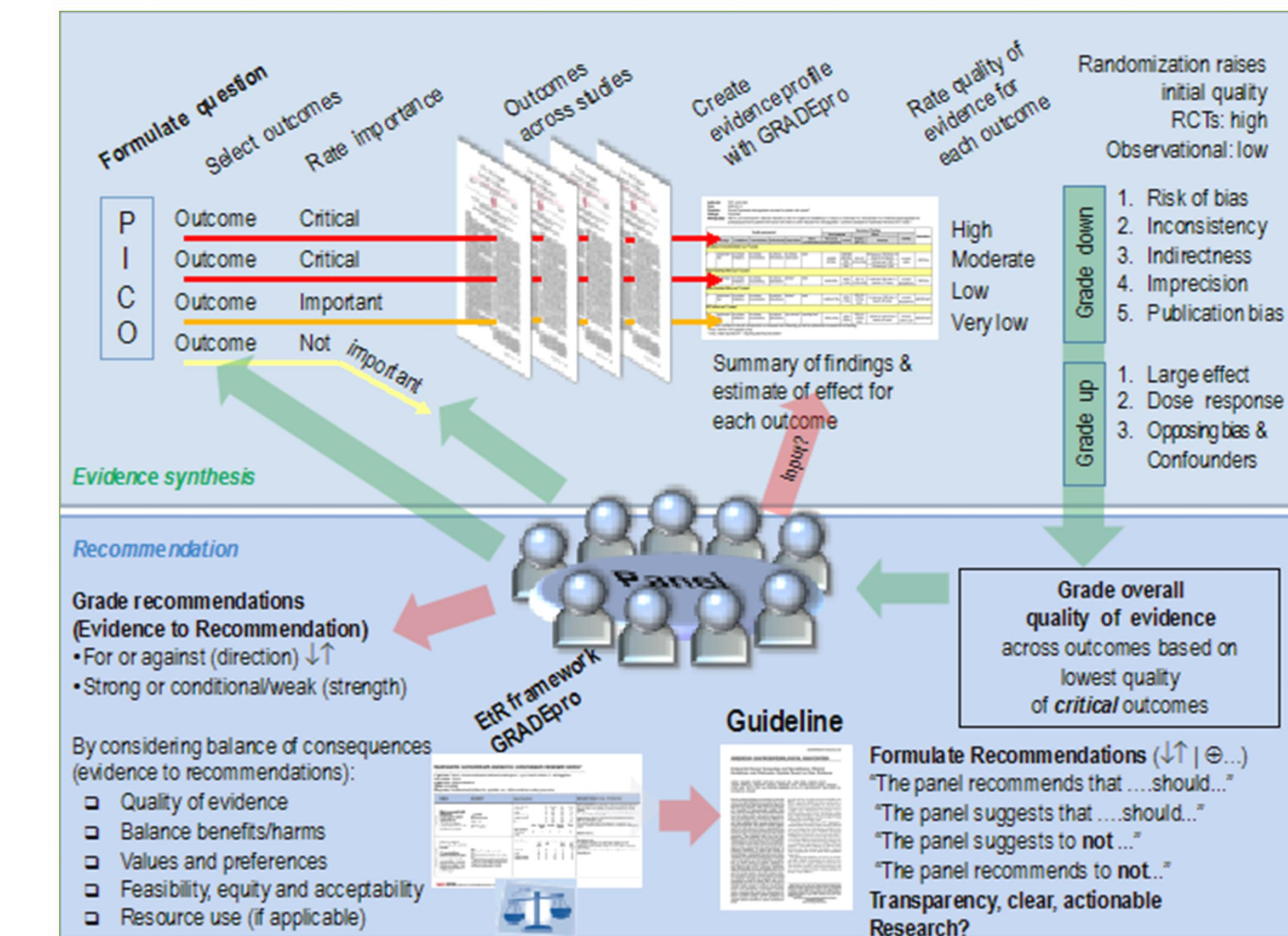
Lichen sclerosis – Main facts

- Inflammatory skin disease
- Involves typically the anogenital site
- Symptoms: e.g. itching, soreness, sexual and urinary dysfunction
- Signs: e.g. whitening of the genital skin, redness, oedema, fissuring, scarring, shrinkage, fusion of structures
- Associated with an increased risk of genital cancer
- Huge impact on the quality of life
- Underdiagnosed and undertreated



Methods

- Developed in accordance with the EuroGuiDerm Methods Manual v1.3¹
- Interdisciplinary panel of experts
- Systematic review of the evidence in collaboration with the British Association of Dermatologists (BAD)
- Development of recommendations using the GRADE (Grading of Recommendations Assessment, Development, and Evaluation) methodology²



Overview of the GRADE methodology (adapted from²)

Consensus-based chapters and recommendations		Evidence- and consensus-based chapters and recommendations	
Definition of disease / Histopathology / Genetic predisposition / Immunological findings / Associated diseases		Topical treatment with emollients	UV therapy
Epidemiology	Lichen sclerosis in pregnancy	Topical and intralesional corticosteroids	Photodynamic therapy
Clinical presentation and sequelae of disease	Pain in lichen sclerosis	Topical calcineurin inhibitors	Laser therapy
Trigger factors	Follow-up	Topical retinoids	Cryotherapy
Diagnosis / Differential diagnoses	Patient education programs	Topical hormone preparations	Systemic treatment
Aims of treatment / Assessment of the treatment success	Interdisciplinary management / Improvement of care	Platelet rich plasma	Surgical interventions
Skin care and basic therapy	Future research / Upcoming treatments		

Main recommendations from the treatment chapters

Women with genital lichen sclerosis

We recommend ultrapotent or potent topical corticosteroids	↑↑
We recommend co-treatment with emollients	↑↑
We suggest topical calcineurin inhibitors as second choice or as an additional treatment if topical corticosteroids are contraindicated or insufficient (off label)	↑
We suggest intralesional corticosteroids for the treatment of topical steroid-resistant hyperkeratotic lesions (provided malignancy has been excluded)	↑
We suggest UVA-1 therapy as a second choice treatment, taking into account carcinogenicity and practicality	↑
We suggest acitretin, taking into account teratogenicity, if systemic therapy is needed (off label)	↑
We suggest de-adhesion / synechiolysis / perineoplasty in women who have a persistent introital stenosis that causes mechanical problems in voiding or sexual intercourse, despite guideline-conform treatment with topical steroids	↑

Girls with genital lichen sclerosis

We recommend ultrapotent or potent topical corticosteroids	↑↑
We recommend co-treatment with emollients	↑↑
We suggest topical calcineurin inhibitors as second choice or as an additional treatment if topical corticosteroids are contraindicated or insufficient (off label)	↑

Men with genital lichen sclerosis

We recommend ultrapotent or potent topical corticosteroids	↑↑
We suggest co-treatment with emollients	↑
We suggest circumcision, preferably removing the complete foreskin, if guideline-conform treatment with e.g. steroids in men with phimosis caused by lichen sclerosis fails	↑
We suggest frenuloplasty in combination with intralesional triamcinolone or alternatively, a complete circumcision if guideline-conform treatment with e.g. steroids in men with scarring or shortening of the frenulum caused by lichen sclerosis fails	↑
We suggest urethroplasty using oral mucosa grafts in men with urethral stricture due to lichen sclerosis causing mechanical problems in voiding or sexual intercourse	↑
We suggest topical calcineurin inhibitors as second choice or as an additional treatment if topical corticosteroids are contraindicated or insufficient (off label)	↑
We suggest acitretin if systemic therapy is needed (off label)	↑

Strength	Wording	Symbols	Implications
Strong recommendation for the use of an intervention	"We recommend..."	↑↑	We believe that all or almost all informed people would make that choice.
Weak recommendation for the use of an intervention	"We suggest..."	↑	We believe that most informed people would make that choice, but a substantial number would not.

Boys with genital lichen sclerosis

We recommend ultrapotent or potent topical corticosteroids	↑↑
We suggest co-treatment with emollients	↑
We suggest circumcision, preferably removing the complete foreskin if guideline-conform treatment with e.g. steroids in boys with phimosis caused by lichen sclerosis fails	↑
We suggest frenuloplasty in combination with intralesional triamcinolone, or alternatively, a complete circumcision if guideline-conform treatment with e.g. steroids fails in boys with scarring or shortening of the frenulum caused by lichen sclerosis	↑
We suggest topical calcineurin inhibitors as second choice or as an additional treatment if topical corticosteroids are contraindicated or insufficient (off label)	↑

Patients with extragenital lichen sclerosis

We recommend UV therapy	↑↑
We suggest ultrapotent or potent topical corticosteroids	↑
We suggest methotrexate, taking into account teratogenicity if systemic treatment is needed in adult patients (off label)	↑
We suggest co-treatment with emollients	↑

References

- 1 European Center for Guidelines Development, European Dermatology Forum. EuroGuiDerm Guideline and Consensus Statement Development Manual. Version 1.3; February 2020.
- 2 Schünemann H, Brozek J, Guyatt G, Oxman A. GRADE handbook for grading quality of evidence and strength of recommendations. Updated October 2013 ed. The GRADE Working Group; 2011.