

# **EuroGuiDerm Guideline for Lichen sclerosus**

G Kirtschig<sup>1</sup>, M Kinberger<sup>2</sup>, A Kreuter<sup>3</sup>, R Simpson<sup>4</sup>, A Günthert<sup>5</sup>, C van Hees<sup>6</sup>, K Becker<sup>7</sup>, MJ Ramakers<sup>8</sup>, M Corazza<sup>9</sup>, S Müller<sup>10</sup>, 5 von Seitzberg<sup>11</sup>, MJ Boffa<sup>12</sup>, R Stein<sup>13</sup>, G Barbagli<sup>14</sup>, CC Chi<sup>15,16</sup>, JN Dauendorffer<sup>17</sup>, B Fischer<sup>18</sup>, M Gaskins<sup>2</sup>, E Hiltunen-Back<sup>19</sup>, A Höfinger<sup>18</sup>, NH Köllmann<sup>18</sup>, H Kühn<sup>20</sup>, HK Larsen<sup>21</sup> M Lazzeri<sup>22</sup>, W Mendling<sup>23</sup>, AF Nikkels<sup>24</sup>, M Promm<sup>25</sup>, KK Rall<sup>26</sup>, S Regauer<sup>27</sup>, M Sárdy<sup>28</sup>, N Sepp<sup>29</sup>, T Thune<sup>30</sup>, A Tsiogka<sup>31</sup>, S Vassileva<sup>32</sup>, L Voswinkel<sup>20</sup>, L Wölber<sup>33</sup>, RN Werner<sup>2</sup>

1 Medbase Health Centre, Fravenleid, Switzerisand 2 Department of Dermatology, Venereology and Allergology, Division of Evidence, Based Medicine (dEBM), Charack - Universitat media in Berlin, and Humbolds Universitat zu Berlin, Berlin, Germany 3 Department of Dermatology, Venereology, and Allergology, HELIOS St. Elizabeth Hospital Oberhausen, Oberhause Permatology, University of Nottingham, Nortingham, Nortingham, Nortingham, Nortingham, Nortingham, Nortingham, Nortingham, Nortingham, Nortingham, UK. 5 Gynāldologisches Jumorzenturum St. Anna, Lucrene, Switzerland. 6 Department of Medical Sciences, University of Nortingham, Office for Pacifications, Sexual Health), Voorschoteen, the Netherlands. 9 Section of Dermatology, Enaburum St. Anna, Lucrene, Switzerland. 6 Department of Medical Sciences, University of Nortingham, Office for Pacifications, Sexual Health), Voorschoteen, the Netherlands. 9 Section of Dermatology, Enaburum St. Anna, Lucrene, Switzerland. 6 Department of Medical Sciences, University of Nortingham, No of Ferrara, Herrara, Finland. 20 The German Lichen Sclerosus Association, German Lichen Sclerosus Association, German, 21 Department of Unology, IACCS Humanitas Research Hospital, Bispebjerg Hospital, Bozano (Mi), Italy, 23 German Center for Infections in Gynecology and Obstetutes, at Helios University Hospital Wappertal—University Witten/Herdecke, Germany, 24 Department of Dermatology, IACCS Humanitas Research Hospital, Bozano (Mi), Italy, 23 German Center for Infections in Gynecology and Obstetutes, at Helios University Hospital Wappertal—University Witten/Herdecke, Germany, 24 Department of Dermatology, IACCS Humanitas Research Hospital, Bozano (Mi), Italy, 23 German Center for Infections in Gynecology and Obstetutes, at Helios University Hospital Wappertal—University Witten/Herdecke, Germany, 24 Department of Dermatology, IACCS Humanitas Research Hospital, Bozano (Mi), Italy, 23 German Center for Infections in Gynecology and Obstetutes, at Helios University Hospital Wappertal—University Witten/Herdecke, Germany, 24 Department of Dermatology, IACCS Humanitas Research Hospital, Bozano (Mi), Italy, 23 German Center for Infections in Gynecology and Obstetutes, at Helios University Hospital Wappertal—University Witten/Herdecke, Germany, 24 Department of University Hospital, Bozano (Mi), Italy, 23 German Center for Infections in Gynecology and Obstetutes, at Helios University Hospital, Bozano (Mi), Italy, 23 German Center for Infections in Gynecology and Obstetutes, at Helios University Hospital, Bozano (Mi), Italy, 24 German Center for Infections in Gynecology and Obstetutes, at Helios University Hospital, Bozano (Mi), Italy, 24 German Center for Infections in Gynecology and Obstetutes, at Helios University Hospital, Bozano (Mi), Italy, 24 German Center for Infections in Gynecology and Obstetutes, at Helios University Hospital, Bozano (Mi), Italy, 24 German Center for Infections in Gynecology and Obstetutes, at Helios University Hospital, Bozano (Mi), Italy, 24 German Center for Infections in Gynecology and Obstetutes, at Helios Uni University Medical Center of Liège, Urège, Belgium. 25 Department of Pacidistric University Medical Contrar of Pegensburg, Germany. 27 Diagnostic and Research Institute of Pathology, Medical University Graz, Graz, Austria. 28 Department of Dermatology, Venercology and Clinic St. Hedwey. Budapear, Hungary, 29 Department of Dermatology and Venereology, Orderskinskum Linz Elisabethinen, Linz, Austria. 30 Department of Dermatology Housesty Hospital, Bergen, Norway, 31 National and Kapodistrian University Hospital, Palexandrovska', Medical University - Sofia, Sofia, Bulgaria: 33 Department of Gynaecology, University Medical Centre Hamburg, Eppendorf and Centre for Colposcopy and Vulvoyaginal disease Jersualem Hospital Hamburg, Hamburg, German



| 34 experts | from 17 | countrie |
|------------|---------|----------|
|------------|---------|----------|

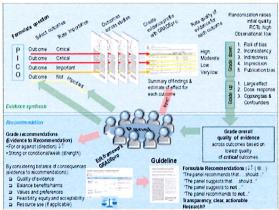
| Consensus-based chapters and recommendations  Definition of disease / Histopathology / Genetic predisposition / Immunological findings / Associated diseases |  | Evidence- and consensus-based chapters and recommendations |                        |  |
|--|--|--|------------------------|--|
|  |  | Topical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

## Lichen sclerosus - 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



- Developed in accordance with the EuroGuiDerm Methods Manual v1.3<sup>1</sup>
- · Interdisciplinary panel of experts
- Systematic review of the evidence in collaboration with the British Association of
- Development of recommendations using the GRADE (Grading of Recommendations Assessment, Development, and Evaluation) methodology<sup>2</sup>



Overview of the GRADE methodology (adapted from<sup>2</sup>)

### Main recommendations from the treatment chapters

#### Women with genital lichen sclerosus

| 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)  | 1 |
| We suggest intralesional corticosteroids for the treatment of topical steroid-resistant hyperkeratotic lesions (provided malignancy has been excluded)   | 1 |
| We suggest UVA-1 therapy as a second choice treatment, taking into account carcinogenicity and practicality  | 1 |
| We suggest acitretin, taking into account teratogenicity, if systemic therapy is needed (off label)  | 1 |
| We suggest de-adhesion / synechlolysis / 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 | 1 |

#### Girls with genital lichen sclerosu

| Chris With Bellitan Herien Selenous   |    |
|---|----|
| We recommend ultrapotent or potent topical corticosteroids  | 个个 |
| 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) | 1  |

#### Men with genital lichen sclerosus

| We recommend ultrapotent or potent topical corticosteroids   |   |
|--|---|
| We suggest co-treatment with emollients  |   |
| We suggest circumcision, preferably removing the complete foreskin, if guideline-conform<br>treatment with e.g. steroids in men with phimosis caused by lichen sclerosus fails   | 1 |
| We suggest frenuloplasty in combination with intralesional triamcinolone or alternatively, a<br>complete circumcision if guideline-conform treatment with e.g. steroids in men with scarring or<br>shortening of the frenulum caused by lichen sclerosus fails | 1 |
| We suggest urethroplasty using oral mucosa grafts in men with urethral stricture due to lichen<br>sclerosus causing mechanical problems in volding or sexual intercourse   | 1 |
| We suggest topical calcineurin inhibitors as second choice or as an additional treatment if topical corticosteroids are contraindicated or insufficient (off label)  | 1 |
| We suggest acitretin if systemic therapy is needed (off label)   | 1 |

| Strength                   | Wording       | Symbols | Implications   |
|----------------------------|---------------|---------|--|
| Strong recommendation for  | We recommend' | 22      | We believe that all or almost all informed people would make that choice.      |
| the use of an intervention |               | 4.4     |  |
| Wesk recommendation for    | We suggest'   |         | We believe that most informed people would make that choice, but a substantial |
| the use of an intervention |               |         | number would not.  |

#### Boye with gonital lichon colorogu

| obys with genital fictien scienosus  |   |
|--|---|
| We recommend ultrapotent or potent topical corticosteroids   |   |
| We suggest co-treatment with emollients  | 1 |
| We suggest circumcision, preferably removing the complete foreskin if guideline-conform treatment with e.g. steroids in boys with phimosis caused by lichen sclerosus fails  | 1 |
| 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 sclerosus |   |
| We suggest topical calcineurin inhibitors as second choice or as an additional treatment if topical corticosteroids are contraindicated or insufficient (off label)  | 1 |

### Patients with extragenital lichen sclerosus

| We recommend UV therapy   | 个个 |
|---|----|
| We suggest ultrapotent or potent topical corticosteroids  | 1  |
| We suggest methotrexate, taking into account teratogenicity if systemic treatment is needed in adult patients (off label) |    |
| We suggest co-treatment with emollients   | 1  |