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# The Management of Vulval Skin Disorders



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This is the first edition of this guideline.

## 1. Purpose and scope

The aim of this guideline is to review the diagnosis and management of common vulval dermatoses seen in general gynaecological practice and the role of specialist training and support.

## 2. Introduction and background

Symptoms and signs of vulval skin disorders are common and include pruritus, pain and changes in skin colour and texture. Community-based surveys indicate that about one-fifth of women have significant vulval symptoms. In the hospital setting, common causes are dermatitis, lichen simplex, vulval candidiasis, lichen sclerosus and lichen planus. Lichen sclerosus accounts for at least 25% of the women seen in dedicated vulval clinics, with estimates of incidence quoted as one in 300 to one in 1000 of all patients referred to dermatology departments. This guideline aims to provide an evidence-based framework for improving the initial assessment and care of women with vulval disorders. This guidance is intended for the general gynaecologist, with advice on when to refer to the specialist multidisciplinary team as the most efficient link to other services, such as a patch test clinic, psychosexual counsellors or reconstructive surgeons. Details of individual skin conditions can be found in Appendix 2. Advice on general care of the vulval skin can be found in Appendix 3 and on training in Appendix 4.

## 3. Identification and assessment of evidence

The Cochrane Library and the Cochrane Register of controlled trials were searched for relevant randomised controlled trials, systematic reviews and meta-analyses. A search of Medline from 1980 to January 2010 was also carried out. The database was searched using the MeSH terms 'pruritus vulvae', 'lichen simplex', 'lichen sclerosus', 'lichen planus', 'vulva and dermatosis', 'vulva and candidiasis', 'vulva and disease and diagnosis' and 'vulvar intraepithelial neoplasia', including all subheadings. This search was combined with a keyword search using the terms 'disease and diagnosis' and limited to 'English language and women and humans'. Databases searched were the Cochrane Database of Systematic Reviews, DARE, EMBASE, Medline, Pubmed, the National Library of Health and the National Guidelines Clearing House. Other related guidelines were available from the British Association of Sexual Health and HIV, the National Institute for Health and Clinical Excellence, the Association of Physicians in Obstetrics and Gynecology and the British Association of Dermatologists.

The definition of the type of review used in this guideline was adult women referred to secondary care with symptoms and/or signs of vulval skin disorders, and the interventions to be studied were specified as the role of history taking, examination and investigations in the diagnosis and management of vulval skin disorders, the use of ultrapotent steroids and alternatives in vulval dermatoses and the provision of surveillance in secondary care for women with vulval disorders.

The research questions were developed and submitted for peer review. Two independent reviewers assessed the literature generated by the review to identify recommendations and the supporting evidence. Evidence was graded according to RCOG Clinical Governance Advice No. 1: *Development of RCOG Green-top Guidelines: Policies and Processes*.<sup>1</sup>

#### 4. What information needs to be included in history taking and examination when women are referred to the gynaecology clinic with symptoms and/or signs of a vulval skin disorder to aid investigation and management?

**A standard gynaecological history will not identify all relevant symptoms. The history taken from a women presenting with vulval symptoms needs to explore symptoms at other skin sites, medical and drug history and family history.**



Non-specific symptoms such as pruritus and pain are the common presenting symptoms in vulval skin disorders, but they are non-specific. Possible causes include vulval dermatoses, infection, contact dermatitis, hormone deficiency and systematic skin disorders. Self-medication or previous inadequate or inappropriate treatments may contribute to symptoms. It is essential to explore a wide relevant history to elucidate possible causal or contributing factors. This will include a past history of abnormal cervical cytology, cigarette smoking and immune deficiency for women with suspected usual type vulvar intraepithelial neoplasia (VIN). Vulval skin often comes into contact with potential allergens and the use of a questionnaire at the clinic can help identify potential allergens and irritants (see Appendix 5).

Evidence level 4

**The history should include details of any personal or family history of autoimmune conditions.**



Women with lichen sclerosus and erosive lichen planus are at increased risk of a personal or family history of autoimmune disorders. Circulating autoantibodies are more frequent in women with erosive lichen planus. The most common autoimmune conditions in women with lichen sclerosus are thyroid disorders, alopecia areata, pernicious anaemia, type 1 diabetes and vitiligo. The reported prevalence of autoimmune conditions in first-degree relatives is around 30%.<sup>2</sup> However, vesiculobullous autoimmune diseases at anogenital sites are uncommon.

Evidence level 2+

**The history should include details of any personal or family history of atopic conditions (hay fever, asthma, eczema).**



The appearance of vulval dermatitis is often non-specific and the characteristic appearance seen at other skin sites may not be seen in anogenital skin. In a case series of 38 women attending a vulval disorders clinic with vulval dermatitis, 97% had a history of atopy and/or seborrhoeic dermatitis.<sup>3</sup>

Evidence level 3

**The history should include any symptoms of urinary or faecal incontinence.**



Urine and faeces are very irritating and can cause damage to skin barrier function. Women should be asked specifically about any urinary or faecal incontinence that damages the vulval skin either directly or indirectly by the use of sanitary products. A small case series suggested that allergies to sanitary products are rare, but dermatitis may be exacerbated by moisture, temperature and friction.<sup>4</sup>

Evidence level 3

**Women with vulval skin disorders require systematic examination of the anogenital region and other skin and mucosal sites.**



It is important to systematically examine the vulva with adequate light and exposure. This is best achieved by using an examination couch where the woman's legs can be raised into a modified lithotomy position with a good light source. Colposcopy is not necessary. It is important to ask the woman to identify the symptomatic area. Some women can struggle to identify the area and some women may never self-examine or be familiar with vulval anatomy. If VIN is suspected, it is important to examine other lower genital tract sites including the vagina,

Evidence level 4

cervix and perianal skin. It is important to examine the rest of the body, including the mouth, for signs of lichen planus and the scalp, elbows, knees and nails for psoriasis. Eczema may be seen at any site.

Evidence level 4

## 5. Which investigations are useful in the investigation of a woman with a vulval skin disorder?

**In the initial assessment of a woman with vulval symptoms, consider testing for thyroid disease, diabetes and sexually transmitted infections if clinically indicated.**

D

There is limited evidence for many routine tests, but the routine investigations listed above can help clarify the diagnosis by identifying associated conditions or conditions that are contributing to vulval symptoms.<sup>3</sup> Under-investigation of women with symptoms suggestive of sexually transmitted infections has been reported in retrospective case series.<sup>5</sup>

Evidence level 4

**Skin biopsy is not necessary when a diagnosis can be made on clinical examination. Biopsy is required if the woman fails to respond to treatment or there is clinical suspicion of VIN or cancer.**

D

Common vulval dermatoses can often be diagnosed on clinical grounds, although a diagnostic biopsy can be helpful when there is clinical uncertainty or failure to respond to treatment. VIN is a histological diagnosis and a biopsy must be taken.<sup>6</sup> On excision, 19–22% of cases of VIN have unrecognised invasion detected.<sup>7</sup> All atypical or suspicious areas must be biopsied to exclude invasive disease. Skin biopsies can safely be taken in the outpatient setting under local anaesthetic. If the biopsy site requires a suture, an undyed vicryl suture should be used.

Evidence level 4

**Women suspected of having lichen sclerosus or lichen planus should be investigated for other autoimmune conditions if there are clinical symptoms or signs.**

C

Women with lichen sclerosus and erosive lichen planus are at increased risk of having or going on to develop another autoimmune disorder.<sup>2</sup> However, no evidence has been identified to support testing for autoantibodies without a clinical indication.<sup>8</sup>

Evidence level 2+

**Serum ferritin should be checked in women with vulval dermatitis.**

C

Correction of iron-deficiency anaemia or low serum ferritin can relieve vulval symptoms. In a case series of 38 women with vulval dermatitis, 20% were found to have iron-deficiency anaemia.<sup>3</sup>

Evidence level 3

## 6. What is the role of skin patch testing in the investigation and management of women with vulval dermatoses?

**Skin patch testing should be performed for women seen with vulval dermatitis.**

D

Specific allergic reactions are often identified in women with pruritus vulvae. It is difficult to generalise the results from case series owing to limited numbers and varying patient selection, but most studies have confirmed that 26–80% of women referred with vulval symptoms have at least one positive result on patch testing.<sup>3,9–12</sup> The most common relevant allergens are cosmetics, medicaments and preservatives.<sup>9–11</sup> Others include fragrances, preservatives in topical treatments, rubber and textile dyes. Washing powder, fabric conditioners, sanitary towels or panty liners and synthetic underwear may also be irritants. Secondary sensitisation to multiple products is common.<sup>3</sup> Factors associated with contact allergy appear to be vulval dermatitis, self-reporting of severe pruritus and the use of multiple topical treatments.<sup>12</sup> It is essential to test for any potential contact allergens that the woman is exposed to in addition to the British Contact Dermatitis Society's standard series.

Evidence level 3

## 7. How should lichen sclerosus and lichen planus be managed?

**Ultrapotent steroids are important in the management of women diagnosed with lichen sclerosus and lichen planus. The patient and her general practitioner require clear advice on the management regime (Appendix 6 describes a suitable management regime).**

C

Corticosteroids have anti-inflammatory and immunosuppressive properties by altering lymphocyte differentiation and function and inhibiting cytokine production. Clobetasol propionate is the most potent topical corticosteroid available. Response rates reported from large case series of women diagnosed with lichen sclerosus are high, with either complete or partial resolution of symptoms in 54–96% of women.<sup>13–17</sup> Improvement in vulval skin texture and colour is seen less often.<sup>13</sup> Evidence on the use of clobetasol propionate comes from a number of case series and non-randomised prospective studies. A 20-year prospective study of the use of clobetasol propionate reported that the probability of remission was significantly associated with age.<sup>18</sup> Women under the age of 50 years had the highest response rates. Relapse is common: 84% of women experience a relapse within 4 years. Higher response rates are seen with longer regular use before returning to ‘as required’ use. Clobetasol propionate appears to be effective and safe in premenarchal girls.<sup>17</sup> The effect of age may be related to disease evolution and/or ability to comply with treatment instructions. Women must have clear instructions on the treatment regime, including the amount applied, timing and frequency of application and application site.

Evidence level 2+

**Approximately 4–10% of women with anogenital lichen sclerosus will have symptoms that do not improve with topical ultrapotent steroids (steroid-resistant disease). The recommended second-line treatment is topical tacrolimus under the supervision of a specialist clinic.**

D

Tacrolimus and pimecrolimus belong to the class of immunosuppressant drugs known as calcineurin inhibitors. Their mode of action differs from that of corticosteroids, mainly reducing inflammation by suppressing T-lymphocyte responses. Tacrolimus and pimecrolimus have both been shown to be effective at controlling a number of vulval dermatoses including lichen sclerosus and lichen planus.<sup>19–24</sup> In a phase II study, maximal effects were seen after 10–24 weeks of treatment and 77% of women had a full or partial response.<sup>25</sup> Calcineurin inhibitors are well tolerated and their use avoids the adverse effects of steroids. However, use of calcineurin inhibitors in anogenital lichen sclerosus is off-licence and should only be undertaken in a specialist clinic. The long-term safety of topical calcineurin inhibitors is not established; however, based on reports of extensive use, safety would appear to be low. While awaiting long-term data, use for longer than 2 years is not recommended owing to concerns about potential malignant transformation. A number of other oral and topical therapies for second-line treatment have been reported in small case series, but there is not sufficient evidence to recommend these agents at present.<sup>26–29</sup>

Evidence level 3

**Surgery and CO<sub>2</sub> laser vaporisation are not recommended for the treatment of symptoms of lichen sclerosus. However, these treatments have a role in restoring function impaired by agglutination and adhesions such as urinary retention or narrowing of the vaginal introitus that affect sexual function or body image.**

D

CO<sub>2</sub> lasers have not been shown to be useful in the management of lichen sclerosus, although laser surgery may be useful in treating the sequelae of scarring secondary to lichen sclerosus.<sup>30</sup>

Evidence level 3

## 8. How should VIN be managed?

**The gold standard for the treatment of VIN is local surgical excision. C**

C

Women undergo treatment of VIN to relieve symptoms of severe pruritus, to exclude invasive disease and to reduce the risk of developing invasive cancer. Simple vulvectomy and radical vulvectomy are inappropriate surgical treatments owing to their adverse effects on sexual function and body image. Local excision is adequate with the same recurrence rates and provides a specimen for histological diagnosis.<sup>31-33</sup> Twelve to seventeen percent of women undergoing excision of VIN have clinically unrecognised invasion diagnosed on histology.<sup>7,34-36</sup> If surgical treatment is not undertaken, adequate biopsy sampling is required to reduce the risk of unrecognised invasive disease. Complete response rates are higher with excision than with ablative or medical treatment techniques.<sup>7</sup> The risk of recurrence is lower with surgical margins free from disease. However, the risk of residual or recurrent disease is not sufficient to recommend re-excision in the absence of clinical disease.<sup>36,37</sup> Even with uninvolved surgical margins, there is still a residual risk of recurrence.

Evidence  
level 3

**Women undergoing surgical excision of VIN should have access to reconstructive surgery. D**

D

Primary closure following excision of small lesions of VIN can produce good results without tension, scarring or disruption to normal anatomy. However, with larger lesions, multifocal lesions or certain anatomical sites, scarring and tension can result in pain and psychosexual morbidity. It is important that women are offered reconstructive surgery.<sup>38-41</sup> Two small case series have shown good sexual function after using different reconstructive techniques following excision of VIN.<sup>40,41</sup>

Evidence  
level 4

**Non-surgical treatments are accepted as an alternative to surgery, but women require regular, long-term follow-up. B**

B

Medical or non-surgical treatments avoid the complications of surgery and spare the vulval anatomy. Topical imiquimod cream is licensed for the treatment of genital warts, but there are a number of case series and small randomised controlled trials with short- and medium-term outcome showing a 15-81% clinical and histological response rate to the imiquimod regime used two to three times per week.<sup>42-48</sup> Adverse effects include pain, erythema and swelling and can result in non-compliance. Cidofovir is also used in the treatment of genital warts and a small case series has shown clinical and histological responses in women with VIN.<sup>49,50</sup> A prospective randomised trial of imiquimod and cidofovir is currently recruiting in the UK. Long-term clinical outcomes and the risk of invasion are not yet known, although there is one report of a case of invasion diagnosed 7 months after imiquimod treatment of VIN. Laser ablation has been shown to be effective in case series. Treatment failure rates are in the order of 40%, but laser ablation is not suitable for hair-bearing skin owing to the involvement of skin appendages.<sup>51</sup> Laser therapy is most useful where tissue conservation is important, such as in the glans and hood of the clitoris, or when surgery is contraindicated. Small case series have looked at the use of cavitron ultrasonic surgical aspiration,<sup>52</sup> photodynamic therapy,<sup>53-55</sup> interferon<sup>56</sup> and therapeutic human papillomavirus (HPV) vaccine,<sup>57-59</sup> but there is insufficient evidence to recommend any of these for treatment in routine clinical care at present.

Evidence  
level 1

## 9. What non-specific measures and advice are useful in the control of vulval symptoms?

**A key part of management is general care of the vulval skin and avoidance of any potential irritants that may worsen vulval irritation. D**

D

Emollients are widely recognised as having a key role in protecting the skin and restoring skin barrier function. General vulval care includes avoiding potential irritants that may worsen vulval symptoms. Uncontrolled studies have shown that these measures reduce symptoms and resolve contact dermatitis and lichen simplex chronicus. Vulval skin is sensitive and may react both to irritants and to allergens. Irritants are commonly encountered and include underwear, sanitary protection, textile dyes, soaps and detergents<sup>9</sup> (see Appendix 3). Avoiding soap and detergents and using soap substitutes can be soothing and protective to the skin. The combined use of emollients and soap substitutes helps maintain symptom relief and is safe and inexpensive. A small, prospective, open trial of maintenance with an emollient following steroid therapy showed that a proportion of women can maintain symptom relief and reduce the use of topical corticosteroids.<sup>60</sup>

Evidence level 3

**It is important to enquire about over-the-counter preparations that may aggravate skin conditions.**



Women may delay seeking a medical opinion and self-medication may contribute to secondary problems such as allergic contact dermatitis, irritant contact allergy or secondary infection. In a large patient survey, adverse self-management was identified as being associated with longer duration of vulval symptoms.<sup>61</sup>

Evidence level 4

## 10. Do women with vulval skin disorders need to remain under long-term surveillance at the gynaecology clinic?

**Women with VIN need to be seen on a regular basis for vulvoscopy or careful clinical assessment and biopsy of any suspicious area. C**

C

Both types of VIN have a well recognised risk of developing into a squamous cell cancer. Small cohort studies have found cancer to develop within 8 years of diagnosis; the reported risk of progression to cancer varies widely, but appears to be in the order of 40–60%.<sup>31,33,34,36,62–67</sup> The risk is higher in untreated women. The risk of unrecognised invasion means a low threshold should be set for biopsy. Women treated surgically for VIN still have a residual risk of developing invasive cancer in the order of 4%. These women should be seen on an annual basis at least for inspection of vulval skin and receive information regarding signs and symptoms (such as pain or ulceration) that would prompt an earlier review.

Evidence level 2

**Women who have been treated for VIN are at risk of intraepithelial neoplasia at other sites. Colposcopy examination should be available at follow-up. C**

C

Four percent of women diagnosed with VIN will have intraepithelial neoplasia at other lower genital tract sites. Women with usual type VIN with multifocal/multicentric disease are at higher risk of recurrence and should be seen at a specialist colposcopy service to ensure inspection of other lower genital tract sites, including the cervix, vagina and vulval and perianal skin.<sup>32,67</sup> Women eligible for cervical screening should comply. Most dermatologists refer women with VIN to a gynaecology service. Evidence level 3

Evidence level 2

**There is no evidence that follow-up of women with lichen sclerosus needs to be hospital based.**



Women with lichen sclerosus and lichen planus appear to have a lifetime risk of developing invasive vulval cancer in the order of 2–4%.<sup>68,69</sup> Estimating the risk of squamous cell cancer among women with vulval dermatoses is difficult and constrained by case selection, limited follow-up and coexisting risk factors. There is conflicting evidence on whether or not risk is independent of successful treatment.<sup>13,14,69,70</sup> Because of the low individual risk, women are not required to be seen for review at a hospital-based clinic, but women need to be given information regarding the risk of invasive disease. Women with lichen sclerosus and differentiated VIN are recognised as a high-risk group and should be kept under specialist review.

Women should be referred back if there are suspicious changes or symptom control cannot be sustained. It has been suggested that women with genital lichen planus also have a risk of vulval cancer; however, the number of case reports is too low to confirm if there is a risk associated with this condition per se and best practice is to recommend self-surveillance.

Evidence level 4

### 11. How should sexual problems associated with vulval skin disorders be identified and, if identified, what is the most effective approach to their management?

**Women should be asked about the impact of their vulval disorder on sexual function and appropriate advice and care should be available.**

D

Vulval disorders often result in poor sexual function or body image. Women may not volunteer information about sexual function, but this can be a very distressing aspect of vulval disorders. Studies that have identified high rates of sexual dysfunction have used specific tools or questionnaires, and their use at initial assessment may be helpful to identify psychosexual problems.<sup>71–75</sup> Advice on sexual behaviour should include the importance of arousal and the use of lubrication.<sup>61</sup> Psychosexual counselling is a specialist service and referral should be available. It is also important to recognise that treatment of vulval disorders can have a negative as well as a positive impact on sexual function.

Evidence level 4

### 12. What is the role of self-examination and what information should women be given on this?

**Women with vulval symptoms should be encouraged to perform self-examination to monitor their skin condition and any suspicious areas.**

D

There are no clinical trials on self-examination or the frequency or duration of monitoring. However, patient support groups and specialist societies do advocate self-examination to detect any suspicious areas.<sup>76–79</sup> Self-examination techniques are provided by these groups. Self-examination may have a role in follow-up. A small audit of a group of patients discharged from the vulval clinic for follow-up of lichen sclerosus reported that 38% had not seen their general practitioner 12 months after discharge. Of those who were seen, 17% were not examined.<sup>80</sup>

Evidence level 4

### 13. What training should general gynaecologists have in the management of vulval disorders?

**According to the RCOG core curriculum, obstetrics and gynaecology trainees must have knowledge and experience of the management of common vulval disorders as a training requirement.**

D

Core training in common vulval disorders will allow many women with vulval symptoms to be diagnosed and managed within the general gynaecology service. For a specialist role, the RCOG has an ATSM training programme in vulval disease (see Appendix 4).



A service evaluation of a specialist vulval clinic demonstrated that most women attending this specialist service had common vulval disorders such as lichen sclerosus, lichen simplex chronicus, lichen planus and VIN.<sup>81</sup> However, almost half of the women referred to the specialist clinic had no diagnosis made at the time of referral or received a diagnosis but no treatment or inadequate treatment was implemented. Most women with lichen sclerosus and lichen simplex will respond to general advice on vulval care and topical steroids. Apart from women with VIN, only those with failure of standard treatment, steroid-resistant disease or intractable symptoms require specialist input.

Evidence  
level 4

#### 14. What is the most effective model for care provision for the investigation and management of women with vulval skin disorders?

**Women with complex or rare vulval skin disorders or who do not respond to standard treatment should be seen at a specialist vulval clinic.**

D

Women who have difficulty with symptom control should be referred to a specialist clinic.<sup>81,82</sup> This includes women who require frequent or prolonged use of ultrapotent topical steroids. Such women may require additional support to use first-, second- or third-line therapy. They require biopsy of any suspicious or resistant areas.

Evidence  
level 4

Two retrospective reviews of specialist vulval clinics have shown that 38% of women need to be seen by more than one specialist including a gynaecologist, dermatologist, genitourinary physician and psychosexual counsellor.<sup>5,83</sup> The multidisciplinary team reduces the risk of incorrect or inadequate treatment, facilitates communication between specialties and provides a one-stop service for the appropriate diagnosis, investigation and management of patients. If the development of a multidisciplinary team is not feasible, effective communication links are required with other specialties.

Evidence  
level 4

Women are concerned about the delay and uncertainty in the diagnosis and management of their vulval disorder. This highlights the need for adequate training in the core curriculum and the role of a specialist service for women with uncommon or unresponsive conditions.

Evidence  
level 4

#### 15. Recommendations for audit

Recommended audit for vulval disorder services:

- number of new referrals, the presenting symptoms and the diagnosis made
- documentation of the response rate to first-line treatment, including symptom control and quality of life
- documentation that information is provided, including contact at a patient support group.

#### 16. Topics for further research

Recommended research topics are:

- a clinical trial of the first-line management of women with lichen sclerosus comparing topical ultrapotent corticosteroids versus topical tacrolimus or pimecrolimus
- a clinical trial of medical management versus surgical treatment of VIN
- large long-term large cohort studies on the risk of malignancy in all common vulval skin disorders.

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## Appendix 1

### EXECUTIVE SUMMARY

Pruritus vulvae and vulval pain are very common complaints and most women initially self-medicate. Although it is often self-limiting, chronic vulval pruritus suggests an underlying vulval dermatosis. Careful and systemic examination is fundamental to making a diagnosis. Skin biopsies are not always necessary but are essential if VIN or invasive disease is suspected or if the condition does not respond to treatment.

General care of vulval skin is a fundamental component of treatment. Avoidance of potential irritants will benefit most conditions. The mainstay of the management of lichen sclerosus is topical ultrapotent steroids. Women require clear advice on the appropriate treatment regimes. Women with VIN require a biopsy to confirm disease. Long-term surveillance is necessary, particularly when a medical or conservative approach to management is taken. All gynaecological trainees require experience in the management of common skin disorders, but a specialist service improves care for women by improving the accuracy of diagnosis and the implementation of adequate and appropriate treatment.

## Appendix 2 VULVAL SKIN DISORDERS

### Common vulval skin disorders

#### *Lichen sclerosus*

Anogenital lichen sclerosus can present at any age, but is more commonly seen in postmenopausal women. It causes severe pruritus, which may be worse at night. The whole vulval perianal area may be affected in a figure-of-eight distribution. Uncontrollable scratching may cause trauma with bleeding and skin splitting and symptoms of discomfort, pain and dyspareunia.

Lichen sclerosus is not linked to female hormone changes, contraceptives, hormone replacement therapy or the menopause. Evidence suggests that it is an autoimmune condition, with around 40% of women with lichen sclerosus having or going on to develop another autoimmune condition.

Pruritus is related to active inflammation with erythema and keratinisation of the vulval skin. Hyperkeratosis can be marked with thickened white skin. The skin is often atrophic, classically demonstrating subepithelial haemorrhages (ecchymoses), and it may split easily. Continuing inflammation results in inflammatory adhesions. Often there is lateral fusion of the labia minora, which become adherent and eventually are completely reabsorbed. The hood of the clitoris and its lateral margins may fuse, burying the clitoris. Midline fusion can produce skin bridges at the fourchette and narrowing of the introitus. Occasionally, the labia minora fuse together medially, which also restricts the vaginal opening and can cause difficulty with micturition and even urinary retention.

#### *Lichen planus*

Lichen planus is a common skin disease which may affect the skin anywhere on the body. Lichen planus usually affects mucosal surfaces and is more commonly seen in oral mucosa. Lichen planus presents with polygonal flat-topped violaceous purpuric plaques and papules with a fine white reticular pattern (Wickham striae). However, in the mouth and genital region it can be erosive and is more commonly associated with pain than with pruritus. Erosive lichen planus appears as a well demarcated, glazed erythema around the introitus. The aetiology is unknown, but it may be an autoimmune condition. It can affect all ages and is not linked to hormonal status.

#### *Lichen simplex chronic or chronic vulval dermatitis*

Women with sensitive skin, dermatitis or eczema can present with vulval symptoms, which can result in lichen simplex chronic, a common inflammatory skin condition. This presents with severe intractable pruritus, especially at night. Non-specific inflammation often involves the labia majora but can extend to the mons pubis and inner thighs. There may be erythema and swelling with discrete areas of thickening and lichenification, especially with scratching. These symptoms can be exacerbated by chemical or contact dermatitis and are sometimes linked to stress or low body iron stores. The mainstay of treatment is general care of the vulva (Appendix 3), avoiding potential irritants and use of emollients and soap substitutes. Antihistamines or antipruritics may be helpful, especially if sleep is disturbed. However, moderate or ultrapotent topical steroids may be necessary to break the itch-scratch cycle.

#### *VIN*

VIN is divided into two types depending on its histopathological characteristics.

#### *Usual type VIN*

Nearly all VIN is of usual type: warty, basaloid and mixed (warty and basaloid). Usual type VIN is more common in women aged 35–55. It is associated with HPV (especially HPV-16), intraepithelial neoplasia of the cervix and vagina, perianal, anal and natal cleft skin and mucosa, cigarette smoking and chronic

immunosuppression. Clinically, it may be multifocal and multicentric. The appearance of usual type VIN varies widely: red, white or pigmented; plaques, papules or patches; erosions, nodules, warty or hyperkeratosis. Usual type VIN is associated with warty or basaloid squamous cell carcinoma.

#### *Differentiated type VIN*

Differentiated type VIN is rarer than usual type and is seen in older women aged 55–85. Some cases are associated with lichen sclerosus. Differentiated type VIN is not related to HPV and does not appear to have a long intraepithelial stage. It is linked to keratinising squamous cell carcinomas of the vulva. Clinically, it tends to be unifocal in the form of an ulcer or plaque. The risk of progression appears to be higher than in usual type VIN.

The symptom of pruritus can be intractable, although the use of emollients or a mild topical steroid may help. The gold standard for VIN is local surgical excision. Women undergo treatment of VIN to relieve symptoms, to confirm histology and to exclude invasive disease. Local excision is adequate surgical treatment and allows a specimen to be taken for histological diagnosis. Twelve to sixteen percent of women undergoing an excision have unrecognised invasion and, if conservative or medical treatment is undertaken, care must be taken to ensure adequate biopsy sampling to avoid unrecognised invasive disease.

#### *Vulval candidiasis*

Vulval candidiasis tends to present with irritation and soreness of the vulva and anus rather than discharge. Diabetes, obesity and antibiotic use may be contributory. Vulval candidiasis may become chronic and a leading edge of inflammation with satellite lesions extending out from the labia majora to the inner thighs or mons pubis. Prolonged topical antifungal therapy may be necessary to clear a skin infection with oral or topical preparations.

#### *Vulval psoriasis*

Psoriasis can involve the skin of the vulva but not vaginal mucosa. The appearance of vulval psoriasis differs from the typical scale of non-genital sites: it often appears as smooth, non-scaly red or pink discrete lesions. Scratching may cause infection, dryness and skin thickening. Examination of other sites including nails and scalp may be helpful in making a diagnosis. Emollients, soap substitutes, topical steroids and calcipotriene are useful for symptom control, but cold tar preparations should not be used in genital sites.

### **Rare vulval skin disorders**

#### *Beçhet's disease*

Beçhet's disease is a chronic multisystem disease characterised by recurrent oral and genital ulcers. In women, ulcers can involve the cervix, vulva or vagina. The ulcers are usually recurrent and painful and can leave scarring. Treatment to control flare-ups and reduce symptoms is based on topical or systemic immunosuppressants.

#### *Extramammary Paget's disease*

Extramammary Paget's disease of the vulva is a rare vulval condition seen in postmenopausal women. The main symptom is pruritus. On examination, lesions have a florid eczematous appearance with lichenification, erythema and excoriation. Extramammary Paget's disease can be associated with an underlying adenocarcinoma. The gastrointestinal and urinary tracts and the breasts should be checked. Surgical excision is recommended to exclude adenocarcinoma of a skin appendage. Photodynamic therapy and topical imiquimod have been used with some success. Despite obvious clinical features, surgical margins are difficult to achieve owing to subclinical disease, and recurrence is common.

#### *Plasma cell (Zoon's) vulvitis*

Zoon's vulvitis is a rare benign chronic inflammatory condition of the vulva that presents with symptoms of pruritus, burning, dyspareunia and dysuria. It usually presents in postmenopausal women. Zoon's vulvitis

is diagnosed histologically and is characterised by dermal infiltration with plasma cells, vessel dilatation and haemosiderin deposition. The aetiology of this condition is unknown; one theory is that it is an autoimmune disorder. There have been case reports favouring successful treatment with topical ultrapotent steroids

#### *4. Vulval Crohn's disease*

Crohn's disease is a chronic inflammatory bowel disorder. It can involve the vulva by direct extension from involved bowel or 'metastatic' granulomas. Rarely, it is seen without known bowel disease or preceding the presentation of bowel disease. The vulva is often swollen and oedematous with granulomas, abscesses, draining sinuses and ulceration. Surgery can result in sinus and fistula formation and tissue breakdown and therefore should be avoided. Treatments include metronidazole and oral immunomodulators.



## Appendix 3

### GENERAL CARE OF VULVAL SKIN

Most women with a vulval disorder will benefit from advice on general care of vulval skin and avoiding potential irritants.

- Washing with water only causes dry skin and makes itching worse. Use a soap substitute to clean the vulval area. Use a small amount of the cream or ointment with water to wash your skin. This will stop the skin from getting as dry and irritated as it would if you used soap or water alone. The cream/ointment is safe to use frequently.
- Shower rather than bath and clean the vulval area only once a day. Overcleaning can aggravate vulval symptoms. If you use a bath, it is helpful to add a bath emollient.
- Avoid using sponges or flannels to wash the vulva. These can irritate your skin. Instead, wash your vulva using aqueous cream or another soap substitute with just your hand. Gently dab the vulval area dry with a soft towel or use a hairdryer on a cool setting held well away from the skin.
- Wear loose-fitting silk or cotton underwear. Close-fitting clothes such as tights, cycling shorts, leggings or tight jeans should be avoided. Wear loose-fitting trousers or skirts and replace tights with stockings. At home, you may find it more comfortable to wear long skirts without underwear.
- Sleep without underwear.
- Avoid fabric conditioners and biological washing powders. You may want to wash your underwear separately in a non-biological washing powder/gel.
- Avoid soaps, shower gel, scrubs, bubble baths, deodorants, baby wipes or douches in the vulval area.
- Some over-the-counter creams including baby or nappy creams, herbal creams (e.g. tea tree oil, aloe vera) and 'thrush' treatments may include possible irritants.
- Avoid wearing panty liners or sanitary towels on a regular basis.
- Avoid antiseptic (as a cream or added to bath water) in the vulval area.
- Wear white or light colours of underwear. Dark textile dyes (black, navy) may cause an allergy; if you wash new dark underwear a few times before wearing it, it will be less likely to cause a problem.
- Avoid coloured toilet paper.
- Avoid wearing nail varnish on finger nails if you tend to scratch your skin.

#### Use of emollients to protect your skin

- Emollients can be used as moisturisers throughout the day. These products can be bought in 500 g tubs or in 100 g tubes over the counter or on prescription from your family doctor.
- Using one of these moisturisers every day can help relieve symptoms. Even when you do not have symptoms, using a moisturiser will protect the skin and can prevent flare-ups.
- It is important to find the moisturiser that suits you best. If the first one you try does not work well, it is well worth trying another one.
- If your skin is irritated, aqueous cream can be kept in the fridge and dabbed on to cool and soothe the skin as often as you like.

## Appendix 4

The RCOG has an Advanced Training Skills Module (ATSM) training programme in vulval disease. This is available on the RCOG website at <http://www.rcog.org.uk/files/rcog-corp/ED-ATSM-Vulval-Disease.pdf>.

This ATSM is designed to provide training in all aspects of vulval disease. Successful completion of the ATSM will equip an individual to develop this aspect of care in their future practice. It is also a component of knowledge for those doctors wishing to work in community-based gynaecology or a gynaecological oncology service in the future.

## Appendix 5

### EXAMPLE OF A PATIENT QUESTIONNAIRE

You have been referred to this clinic with a skin problem. It would be helpful if you could complete this questionnaire before you are seen. This will help to identify any factors that may be causing or aggravating your skin problem. This will be discussed with the doctor, but you may want to add additional notes if you feel it will help you to remember any important information.

Do you take a bath?	Yes	No	Sometimes
Do you take a shower?	Yes	No	Sometimes
Do you wash your hair in the bath or shower?	Yes	No	Sometimes
What do you wash over all with?			
What do you wash the vulva area with?			
Do you use moist skin wipes in the vulval area?	Yes	No	Sometimes
Do you use talcum powder in the vulval area?	Yes	No	Sometimes
Do you use antiseptic in the bath?	Yes	No	Sometimes
What do you use to wash your clothes?			
Do you use a fabric softener/conditioner?	Yes	No	Sometimes
What type/material underwear do you usually wear?			
Do you wear dark-coloured underwear?	Yes	No	Sometimes
What colour toilet paper do you use?			
Do you use tampons?	Yes	No	Sometimes
Do you use sanitary towels?	Yes	No	Sometimes
Do you use panty liners?	Yes	No	Sometimes
Do you use incontinence pads?	Yes	No	Sometimes
Do you use condoms?	Yes	No	Sometimes

The following questions relate to your own health.

Do you have any of the following conditions?

Diabetes	Yes/No	
Thyroid disease (over- or underactive thyroid gland)	Yes/No	
Alopecia (hair loss)	Yes/No	
Pernicious anaemia (treated by monthly vitamin B injections)	Yes/No	
Vitiligo (patches of white skin)	Yes/No	
Rheumatoid arthritis	Yes/No	
Hayfever /asthma	Yes/No	
Do you have any allergies?	Yes/No	
Do you have any other skin conditions (e.g. eczema, psoriasis, vitiligo, dry skin, sensitive skin, flaky scalp)?	Yes/No	If yes, please write them down
Are you on any medicines? This includes prescribed/herbal/over the counter/HRT /contraception.	Yes/No	If yes, please write them down

If you have already tried treatments for your skin problem, please note them below.

Name of treatment	How long did you use it for?	Effects

## Appendix 6

### PATIENT INFORMATION ON THE USE OF CLOBETASOL PROPRIONATE 0.05% CREAM OR OINTMENT

You should apply your clobetasol cream/ointment sparingly (this means half to one finger tip) to the affected area(s). These are the areas where you notice itch/discomfort or changes in the skin. Apply the cream:

- once daily for 1 month
- then on alternate days for 1 month
- then twice a week for 1 month
- then once a week for 1 month
- then gradually reduce this until you can use it occasionally or not at all.

One 30 g tube of clobetasol cream should last at least 3 months. This amount should not cause you to have adverse effects on the treated skin or elsewhere in the body.

If symptoms return after the 4-month course, you can use the clobetasol cream/ointment every night for 2 weeks to treat the flare-up and then try to reduce the frequency, as above.

If symptoms keep coming back quickly when you stop using the cream, you may prefer to use the cream regularly once or twice a week long term. Long-term use is safe as long as one 30 g tube lasts at least 3 months. More than this may cause skin thinning.

It is normal to notice stinging for a few minutes after applying the cream. However, if you notice stinging in the area for more than 1–2 hours after applying the cream, you may have become sensitive to one of the ingredients. There are several alternative creams and you should contact your clinic for advice.

## Appendix 7

Clinical guidelines are: 'systematically developed statements which assist clinicians and patients in making decisions about appropriate treatment for specific conditions'. Each guideline is systematically developed using a standardised methodology. Exact details of this process can be found in Clinical Governance Advice No.1: *Development of RCOG Green-Top Guidelines* (available on the RCOG website at <http://www.rcog.org.uk/womens-health/clinical-guidance/development-rcog-green-top-guidelines-policies-and-processes>). These recommendations are not intended to dictate an exclusive course of management or treatment. They must be evaluated with reference to individual patient needs, resources and limitations unique to the institution and variations in local populations. It is hoped that this process of local ownership will help to incorporate these guidelines into routine practice. Attention is drawn to areas of clinical uncertainty where further research may be indicated.

The evidence used in this guideline was graded using the scheme below and the recommendations formulated in a similar fashion with a standardised grading scheme.

Classification of evidence levels	Grades of recommendations
1++ High-quality meta-analyses, systematic reviews of randomised controlled trials or randomised controlled trials with a very low risk of bias	<b>A</b> At least one meta-analysis, systematic review or randomised controlled trial rated as 1++ and directly applicable to the target population; or  A systematic review of randomised controlled trials or a body of evidence consisting principally of studies rated as 1+ directly applicable to the target population and demonstrating overall consistency of results
1+ Well-conducted meta-analyses, systematic reviews of randomised controlled trials or randomised controlled trials with a low risk of bias	<b>B</b> A body of evidence including studies rated as 2++ directly applicable to the target population, and demonstrating overall consistency of results; or  Extrapolated evidence from studies rated as 1++ or 1+
1- Meta-analyses, systematic reviews of randomised controlled trials or randomised controlled trials with a high risk of bias	<b>C</b> A body of evidence including studies rated as 2+ directly applicable to the target population and demonstrating overall consistency of results; or  Extrapolated evidence from studies rated as 2++
2++ High-quality systematic reviews of case-control or cohort studies or high-quality case-control or cohort studies with a very low risk of confounding, bias or chance and a high probability that the relationship is causal	<b>D</b> Evidence level 3 or 4; or  Extrapolated evidence from studies rated as 2+
2+ Well-conducted case-control or cohort studies with a low risk of confounding, bias or chance and a moderate probability that the relationship is causal	
2- Case-control or cohort studies with a high risk of confounding, bias or chance and a significant risk that the relationship is not causal	
3 Non-analytical studies, e.g. case reports, case series	
4 Expert opinion	
	<b>Good practice point</b>   Recommended best practice based on the clinical experience of the guideline development group

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The guideline review process will commence in 2014 unless evidence requires earlier review.

#### DISCLAIMER

The Royal College of Obstetricians and Gynaecologists produces guidelines as an educational aid to good clinical practice. They present recognised methods and techniques of clinical practice, based on published evidence, for consideration by obstetricians and gynaecologists and other relevant health professionals. The ultimate judgement regarding a particular clinical procedure or treatment plan must be made by the doctor or other attendant in the light of clinical data presented by the patient and the diagnostic and treatment options available.

This means that RCOG guidelines are unlike protocols or guidelines issued by employers, as they are not intended to be prescriptive directions defining a single course of management. Departure from the local prescriptive protocols or guidelines should be fully documented in the patient's case notes at the time the relevant decision is taken.