# Vulval problems

The vulva contains skin, mucous membrane and glands. There is enormous potential for pathology, including psychosomatic problems. The scope is so wide that this article aims to give just a general overview with frequent links to other articles that cover aspects in greater detail.

The vulva is affected by lack of oestrogen after the menopause. Pruritus vulvae and irritation are common symptoms in a postmenopausal woman. Examination of the vulva should exclude finding ulceration or a mass that may accompany these, as they may also be indicative of infection, inflammation, or malignancy.

Consider infective causes, dermatological conditions, hormonal deficiency, systemic disease and malignant or pre-malignant change. The likely spectrum of disease varies considerably according to the age of the patient.

Vaginismus and sexual dysfunction in women are covered elsewhere - see the separate Vaginismus article. As many as one fifth of women have significant vulval symptoms.

## Presentation

### Symptoms

Note first the age of the patient. Get as much history as possible before examination:

- Are symptoms sudden or gradual in onset?
- Is there pain, discomfort, irritation, or itching, or just 'something which has been found'? Dyspareunia has its own separate article.
- Is there any discharge?
- Is it a vulval or a vaginal problem? Many women are not very good at accurate localisation of that part of their anatomy.

- Is there anything to see? Some women will have made close inspection with a hand mirror whilst others would not contemplate looking down there.
- Is there dysuria? If so, is there urinary frequency too? There may be no urinary tract infection but it is painful to pass urine through an inflamed area.
- If she is sexually active? Does her partner have any problems?
- Are there any other current problems such as psoriasis, chickenpox or recent use of antibiotics?

### Signs

Signs are as variable as the differential diagnosis and so they will be considered together. Generally, examination will be limited to inspection with gentle parting of the labia. Vaginal examination is often unnecessary and, with the current condition, it may be too painful to be reasonable.

## Investigations

Some conditions can be diagnosed simply on inspection but others may require swabs and possibly viral culture or even biopsy to confirm the nature.

## Congenital anomalies

Female genital abnormalities are uncommon and often do not present until, or well after, puberty. See the separate Female Genital Abnormalities article for more information.

## Infections

### Candidiasis

Candidal infection can occur at all ages.

- It may cause vaginitis and a white, curdy discharge as well as vulvitis.
- There is usually pruritus and a red rash.
- Topical antifungal creams are usually adequate. Vaginal infection may need treating too.

### Varicella

Girls with chickenpox sometimes get vesicles around the vulva. This can be very painful or itchy. Treatment is symptomatic with tepid baths, soothing lotions and occasionally topical anaesthetic creams.

### Genital herpes

See the separate Genital Herpes Simplex article.

Genital herpes is caused equally in the UK by the herpes simplex viruses types 1 and 2 (HSV-1 and HSV-2) and it is spread by direct contact. There are painful, fluid-filled vesicles around the genital area. Virus can be cultured from the fluid.

- All sexual contacts must be notified to seek advice. Ideally referral to a genitourinary medicine (GUM) clinic should be made for diagnosis, treatment and contact tracing.
- The vesicles leave tender ulcers that may take two to four weeks to heal the first time they occur.
- Typically, another outbreak can appear weeks or months after the first, but it is almost always less severe and is shorter than the first outbreak. Although the infection can lie dormant indefinitely, the number of outbreaks tends to decrease over a period of years.
- Management involves salt baths, analgesics, loose underwear, antiviral therapy and abstinence from sexual intercourse until all ulcers are healed

### Genital warts

Genital warts are caused by human papillomavirus (HPV), usually types 6 and 11.

- Transmission is usually sexual.
- Warts can appear one to eight months after infection.
- There may be pain, bleeding and pruritus.
- Screening for other sexually transmitted infections is usually advisable.

- Podophyllin paint compound BP destroys the affected skin cells so the warts shrink or disappear. It must be applied accurately and should not be used in pregnancy.
- Other treatments include imiquimod, cryotherapy and electrocautery.

### Shingles

Herpes zoster does not often affect the genital region but the lesions are characteristic. The anterior two thirds of the labia majora are innervated from L1 and the posterior third from S2/S3/S4.

### Infestations

Infestations such as scabies and pubic lice may affect the area. Treatment is with an insecticide such as malathion or permethrin. For pubic lice, screening for other sexually acquired infections is advised, and infected individuals should be advised to notify sexual partners. For scabies all household members should be treated simultaneously.

## Dermatological conditions

### Nappy rash

- Nappy rash is an irritant contact dermatitis.
- The skin in contact with nappies is exposed to friction and excessive hydration, has a higher pH than other skin, and is repeatedly soiled with faeces containing enzymes with high irritation potential for the skin [1] .
- The combination of these factors frequently results in skin damage, leading to visible erythematous lesions that can be irritating and painful to the child.
- The vulva may be bright red and there may be little patches away from the main part, suggesting candidal infection too.
- Note whether the child looks cared for or neglected.
- Advise about frequent nappy changing and also leaving the child without a nappy for as long as possible.
- Advise use of a barrier preparation at each nappy change [2] .

- Topical hydrocortisone 0.5-1% once a day for up to a week may be considered to reduce inflammation [3] . This should be applied sparingly, then followed a few minutes later by application of the barrier preparation.
- If there is suspicion of candidal infection, a combination of hydrocortisone and an antifungal is required.
- Consider oral antibiotics for secondary bacterial infection.

### Vulval dermatitis

- Dermatitis may be irritant (for example, from wetness, incontinence, vigorous cleansing) or allergic contact dermatitis. Specific allergic reactions (eg, to perfume or rubber) may cause pruritus vulvae. Patch testing may be useful.

Almost any skin disorder may also affect the vulva but a few are of special note:

- Lichen planus has a very unpleasant variation that causes painful erosive vulvitis:
    - It usually affects women over the age of 45, and the vestibular area and lower vaginal skin can (rarely) be involved [4] .
    - Examine the mouth which may also be involved.
    - There is intense erythema, oedema and superficial ulceration.
    - It leads to scarring and introital narrowing resembling chronic lichen sclerosus.
    - Biopsy will distinguish it from other ulcerative disorders, including pemphigus, pemphigoid and erythema multiforme.
    - Systemic steroids are often required.

- Psoriasis is not usually itchy, but it can be on the vulva:
    - It does not affect the vaginal mucosa.
    - It may be necessary to take scrapings to exclude tineal infection.
    - There will usually be evidence of the disease elsewhere.
    - Treatment is as for psoriasis elsewhere.
- Behçet's syndrome is a disease of unknown aetiology:
    - It is characterised by recurrent aphthous ulcers, possibly ulcers of the vulva. There is often associated uveitis.
    - These usually occur on a cyclical basis and often are related to the menstrual cycle.
    - They are sometimes associated with arthritis, usually of the knees.

## Swelling

Swelling or oedema of the vulva can be due to venous or lymphatic obstruction:

- Secondary to malignancy in the pelvis.
- Dependent oedema with prolonged sitting in bed.
- Pregnancy, where varicosities may appear - usually resolving at the end of the pregnancy.
- Haematoma suggesting trauma (may have been of a sexual nature).

## Vulval ulcers

When a patient presents with a vulval ulcer the following need to be excluded with culture and/or biopsy with colposcopy:

### Sexually transmitted infection

- HSV.

- Chancroid.
- Granuloma inguinale.
- Lymphogranuloma venereum.
- Syphilis. **NB**: this should not be forgotten, as the number of cases in the UK is increasing.

### Malignancy

- Vulval cancer.
- Vulval intraepithelial neoplasia (VIN).
- Paget's disease of the vulva.

### Other ulcerative conditions

- Behçet's disease.
- Systemic lupus erythematosus.

## Potentially pre-malignant conditions

### Lichen sclerosus

- Most cases of lichen sclerosus are in postmenopausal women, although it can occur in prepubescent girls and young women. It can be familial and may affect the male prepuce too.
- The risk of developing invasive disease is around 4% [5] .
- There is an association with other autoimmune diseases, usually a thyroid disorder.
- Lichen sclerosus may present with pruritus vulvae, vulvodynia, superficial dyspareunia, or visible lesions. It has an appearance called 'cigarette paper' skin as it is thin, white and crinkly. The introitus may shrink with fusion of the labia minora.
- Treatment is with potent topical corticosteroids.

### Vulval intraepithelial neoplasia (VIN)

- VIN is a pre-malignant condition which can arise in pre-existing vulval disorders, or independently.
- It usually presents with pruritus. There may be red, white or raised areas of skin.
- It is a histological condition and a biopsy must be taken.
- Treatment is usually by local excision. Imiquimod can be effective as an alternative or adjunct to surgery.
- Some women may not have active treatment.

# Malignant disease of the vulva

85% of cancers of the vulva are squamous and the remaining are of various histological types, including melanomas. Vulval cancer may present with a vulval lump, vulval bleeding due to ulceration, pruritus or pain. See the separate Vulval Cancer and Vulval Intraepithelial Neoplasia article for further information.

### Paget's disease of the vulva [6]

- Extra-mammary Paget's disease is a rare form of superficial skin cancer. However, the most common site of involvement is the vulva.
- It is seen mainly in postmenopausal white women.
- A woman with Paget's disease of the vulva may present with pruritus and weeping or bleeding of the lesion.
- The lesion appears to have an eczematous or velvet-like surface.
- Surgical excision is the gold-standard treatment; however, recurrence rates are high and extensive excisions can produce long-lasting cosmetic and functional defects.
- There is increasing evidence for the safety and efficacy of 5% imiquimod [7] .
- Other treatments include photodynamic therapy, laser therapy, radiotherapy or chemotherapy.

### Melanoma

- Melanoma should be considered if there are pigmented lesions on the vulva.
- They are suspicious if they are blue-black in colour, have a jagged or fuzzy border, are raised or ulcerated, or are larger than approximately 1 cm.
- Melanomas may be misdiagnosed as undifferentiated squamous carcinoma, particularly if they are amelanotic.
- Most melanomas are located on the labia minora or clitoris and prognosis is related to the size of the lesion and the depth of invasion.

## Referral

Consider a suspected cancer pathway referral (for an appointment within two weeks) for vulval cancer in women with an unexplained vulval lump, ulceration or bleeding [8] .

- When a woman presents with vulval symptoms, a vulval examination should be offered.
- Vulval cancer can also present with vulval bleeding due to ulceration. A patient with these features should be referred urgently.
- A patient who presents with pruritus or pain may be reasonably managed with a period of 'treat, watch and wait'.
- This should include active follow-up until symptoms resolve or a diagnosis is confirmed.
- If symptoms persist, the referral may be urgent or non-urgent, depending on the symptoms and the degree of concern about cancer.
- Other paths of referral may include a GUM clinic or gynaecology or dermatology, based on appearance and suspicions of the examining GP.

## Sexual abuse

See the separate articles Safeguarding Children - How to Recognise Abuse or a Child at Risk and Safeguarding Children - Referral and Management of an Abused or At-risk Child for detailed information.

Sexual abuse can occur at any age (including in the elderly) but the problem is particularly well documented in children.

If a child presents with a condition that is usually sexually transmitted (such as genital warts or HSV), sexual activity needs to be considered, but it is not the only cause. Evidence of trauma, especially with a spurious explanation, is also suggestive. Other features may include behavioural disorders and inappropriate sexuality of behaviour.

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