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Assessment and management of vulval pain

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The vulva refers to the female external genitalia comprising the mons pubis, clitoris, labia majora, labia minora, and perineum (figure). Vulval pain can be divided into two categories: pain secondary to a specific, identifiable, underlying disorder and idiopathic pain with no recognisable underlying disease. Women use the term pain to include a variety of unpleasant symptoms including burning, soreness, and throbbing, and some women insist that they do not have pain but describe the sensation with one of these words, which can be misleading for clinicians.

The exact prevalence of vulval pain in adult women is not known, but a recent population based survey from the United States found that 16% of women had at some time experienced chronic burning, knife-like pain, or pain on contact with the vulva that lasted for at least three months.[1] Seven per cent of women reported current vulval pain or discomfort.

Vulval pain has many causes (box 1),[2] and it is not always straightforward to make a definitive diagnosis. Small cohort studies have shown that chronic vulval pain that is inadequately treated may have a detrimental effect on sexual and social functioning.[3-7] This article aims to cover the assessment and management of women with vulval pain.

SOURCES AND SELECTION CRITERIA

We searched for papers that had been published up to August 2011 using appropriate index terms (vulval pain, vulvodynia, vestibulodynia, lichen planus, and lichen sclerosus) from the National Library computerised search service (Medline, PubMed). Most studies were observational but a few randomised controlled trials were available.

Who gets vulval pain and what causes it?

Vulval pain can affect almost any woman of any age, and it is difficult to develop a profile of a woman "at risk." As outlined in box 1, vulval pain may be caused by infective, inflammatory, or neoplastic disease, where there are usually identifiable vulval changes. In the absence of clinical findings and where pain or discomfort is persistent, a diagnosis of vulvodynia should be considered. Vulvodynia is defined as "vulval discomfort, most often described as burning pain, occurring in the absence of relevant visible findings, or a specific, clinically identifiable neurological disorder."[2]

To aid the diagnosis and management of women with vulval pain, it is helpful to consider the acute and chronic causes of this symptom.

Acute vulval pain

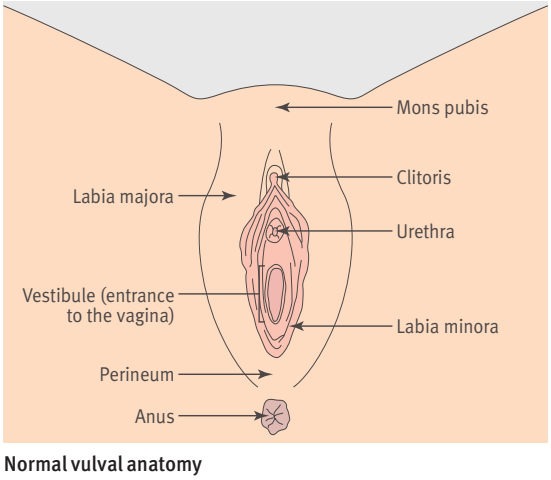
Common causes of acute vulval pain include infection (such as vulvovaginal herpes and candidiasis, which can cause excoriation and fissures in the skin through scratching) and skin disease (such as vulval dermatitis (eczema)). Skin disease often causes acute pain from excoriated and fissured skin, which can become secondarily infected and result in an acute flare of symptoms. During these flares, signs can often be seen on examination, although conditions such as herpes may not be apparent at the time of the consultation if the lesions have already resolved. Contact dermatitis is not uncommon, and vulval skin is often exposed to a variety of irritants and sensitisers, including urine, fragranced wash products, and topical drugs, such as steroids, lidocaine products, and clotrimazole. It is important to recognise that patients with a diagnosis of vulvodynia can also present with acute exacerbations of pain because of flares ups or concurrent infection.

Chronic vulval pain

Any of the acute presentations listed above can lead to chronic pain if left untreated. Chronic vulval discomfort

SUMMARY POINTS

- Vulvodynia is a diagnosis of exclusion; it may be generalised or localised, unprovoked or provoked, and it often presents as chronic superficial pain in the genital region
- Patients with generalised skin conditions may have painful involvement of the vulval area
- Cultures from vulval swabs (or findings on a wet preparation) may identify a treatable infective cause
- A punch biopsy may be a useful diagnostic tool if abnormal lesions are seen on examination
- Refer patients with vulvodynia or difficult to treat disease to specialist care—ideally to a multidisciplinary service
- Treat unprovoked vulvodynia according to principles of chronic pain management (for example, tricyclic antidepressants or gabapentin); women with provoked vulvodynia may respond to physical and psychosexual therapy aimed at desensitisation



Normal vulval anatomy

Box 1 | International classification of vulval pain[2]

Vulval pain related to a specific disorder (often acute but may be chronic)
Infectious (such as vulval candidiasis and herpes)
Inflammatory (such as lichen sclerosus and lichen planus)
Neoplastic (such as vulval intraepithelial neoplasia and squamous cell carcinoma)
Neurological (such as herpes neuralgia)

Vulvodynia (not related to an underlying disorder, often chronic)
Generalised
Provoked (sexual, non-sexual, or both)
Unprovoked
Mixed (provoked and unprovoked)

Localised (such as vestibulodynia, clitorodynia, and hemivulvodynia)
Provoked (sexual, non-sexual, or both)
Unprovoked
Mixed (provoked and unprovoked)

Box 2 | Questions to ask when taking a history

Pain
What is the pain score (out of 10) on a good to average day and a bad day? (10 is the worst pain ever)
What is the nature of the pain? Does it radiate? Are there any aggravating or relieving factors?
Do you have any other pain, such as back pain?
Is the pain relieved by standing or lying flat (pudendal neuralgia)?
If sexual intercourse is painful is the discomfort at the entrance to the vagina or deep inside?

Functioning
What do you miss as a result of the pain?
How does the pain affect your life?

Sexual functioning (if appropriate)
Is vaginismus (involuntary tightening of the pelvic floor muscles on penetration) a problem? Do you have difficulties having sexual intercourse (vaginismus, dyspareunia)?
How do you feel about sex (avoidance, loss of libido)?
Is a lack of lubrication (poor arousal) a problem?

Previous failed treatments
What was adherence like?
What were the side effects?
How long was the treatment used for?

Other
Do you have any past or current gynaecological problems?
Do you have any skin problems such as eczema or psoriasis?
Is there a relevant medical or surgical history, such as thyroid dysfunction?

associated with skin diseases, such as dermatitis or psoriasis, is often associated with vulval itching. In these conditions, acute-on-chronic flare ups can occur as a result of skin fissuring. Other causes of chronic discomfort include recurrent candidiasis and idiopathic vulval fissuring. Lichen sclerosus and lichen planus can also cause discomfort. Women may also exhibit a combination of problems, such as symptoms resistant to treatment and psychosexual dysfunction, and in these complex cases or where the cause is unclear, referral to secondary care is recommended.

Vulvodynia is a chronic pain syndrome that can be considered after other causes have been excluded and discomfort remains. The pain of vulvodynia can be localised or generalised; it can also be spontaneous or provoked by touch or pressure on the vulva—for example, during the insertion of sanitary protection, sexual intercourse, or even just the wearing of clothes. The term vestibulodynia describes provoked pain localised to the vestibule region, with pain experienced on sexual and non-sexual touch. Clinically, this can be demonstrated by applying gentle pressure to the vestibule area with a cotton tipped swab. The maximal area of tenderness is experienced over the entrance to the Bartholin's glands.[8]

Many patients have both provoked and unprovoked symptoms and may benefit from combined treatment strategies.[9]

How should patients with vulval pain be evaluated?

History: which questions are important to ask?

It is important to clarify early on that the woman has discomfort of the external genitalia and the entrance of the vagina rather than pain deep in the vagina or deep dyspareunia, because these symptoms are managed differently. She may use the terms burning, stinging, swelling, or discomfort rather than pain. A detailed pain history is important including the location and severity of the pain, any radiation, and factors that aggravate or relieve the pain (box 2). It is also important to assess how it affects her daily life and functioning. Inquire about treatments she might have tried, about how these were used, and her adherence to treatment. If a woman reports difficulties with intercourse, it is important to determine whether pain on intercourse is superficial, at the point of penetration, which is likely to be related to a vulval problem or deep inside (deep dyspareunia), which is not caused by vulval disease. Ask specifically about vaginismus (tightening of the pelvic floor muscles) and whether there is a lack of vaginal lubrication—some women with a vulval condition develop a phobia about sex and get involuntary vaginismus because of the pain. This problem can remain once the primary cause of vulval pain has been treated. It is important to acknowledge what effect any sexual dysfunction has on her life and relationships. To develop an overall profile of the woman's problem, take a brief gynaecological history—ask about menstruation, fertility, and urinary problems. Conditions such as pudendal neuralgia, an entrapment nerve syndrome, can present with symptoms similar to vulvodynia. Patients with this condition usually experience pain on sitting that is relieved on standing or lying down, and they may have a sensation of a lump or fullness in the vagina.[10] The two conditions must be distinguished to allow for appropriate management.

Also ask about the presence or history of skin conditions and about exposure to potential irritants and allergens such as over the counter remedies, creams, and scented sanitary pads or panty liners.[w1] Ask about urinary incontinence because urine can be a vulval irritant.

Examination of a woman with vulval pain

When examining the vulva it is important to be systematic; use a good light source; and examine the mons pubis, clitoris, labia majora, labia minora, and perineum (figure). Many of the conditions associated with vulval pain are intermittent and repeated examination may be necessary to identify a lesion.

Look specifically for changes in anatomical shape,

TIPS FOR NON-SPECIALISTS

If the patient has a history of dermatitis (eczema) or psoriasis in other areas of the body, these disorders may affect the genital region and cause discomfort

Always carry out a careful examination of the anogenital area with a good light source so that small fissures in the interlabial sulci and the perineal body are not missed

Perform a speculum examination because this can show vaginal pathology, such as erosive lichen planus or desquamative inflammatory vaginitis

Advise the woman to substitute soap and specialised feminine products with bland emollients to reduce irritation and sensitisation

Always refer a patient with chronic vulval pain to a specialised vulval service if you are unclear about the diagnosis

skin colour, induration, and ulcerated and eroded areas. Neoplastic lesions are usually evident on examination and rarely present with pain. Premalignant lesions (vulval intraepithelial neoplasia), however, can cause pain, and these can present as pigmented, ulcerated, or indurated skin lesions.

Be mindful of the patient's response to examination of the vulva. Patients with vulvodynia commonly show touch sensitivity (allodynia). If tolerated by the patient, digital palpation of the levator muscles is useful to assess muscle symmetry and hypertonicity. A Q-tip cotton wool bud applied gently to the vulva in the interlabial sulci, the introitus, and around the clitoris can elicit tenderness in the absence of clinically visible changes such as redness and fissures in the skin and mucosa. This Q-tip test is useful in eliciting provoked vulvodynia. In a recent study, 61% of patients referred to a specialist clinic with refractory vulvodynia were found to have a clinically relevant vulval dermatosis such as lichen planus, dermatitis, or lichen sclerosus.[11]

Vaginitis, erosive vaginal lichen planus, or pemphigus vulgaris may be seen on speculum examination of the vagina.[12 13] Speculum examination also allows the cervix to be assessed and swabs to be taken if appropriate.

Examination of a woman with vulval pain should also include a general examination of skin surfaces and the oral cavity to look for systemic diseases that might account for the pain, such as lichen planus.[13]

What other investigations may be needed?

A diagnosis of vulvodynia can be made on history and examination, and further investigation may not be necessary. However, it is important to exclude other treatable causes before this diagnosis is made. If symptoms are intermittent, a diagnosis of recurrent infection is likely, and a full screen for infections (including sexually transmitted infections) can be offered to look for infective causes of discomfort such as candida and herpes.

Refer women with a vulval lesion of uncertain diagnosis on examination for a 4 mm punch biopsy of the vulva. This procedure can be performed in an outpatient setting under local anaesthetic. Colposcopy is useful in assessing any cervical or upper vaginal lesions that are noted on examination.

In women with suspected contact dermatitis, skin patch testing can be useful to identify the cause.

Expert consensus states that if no other pathology can be found, it is reasonable to diagnose vulvodynia without further investigation.[14-16]

A PATIENT'S STORY

I have lived with vulval pain for more than 25 years. I began to experience vulval itching and soreness in my early teens, as well as burning pain when I tried to insert tampons. My general practitioner did a vaginal swab, which showed that I had thrush, but despite repeated use of the topical antifungals prescribed my symptoms failed to clear and began to worsen. The soreness and pain had a detrimental impact on all aspects of my life—physical and emotional—for many years, often affecting my ability to concentrate at work and to sleep properly, and penetrative sex was excruciatingly painful. It wasn't until my early 20s that I was diagnosed with vulvodynia and vestibulodynia, and in my 30s I was also diagnosed with lichen sclerosus. The treatments that I found most useful were vaginal biofeedback physiotherapy for the vestibulodynia, washing with emulsifying ointment, and keeping the lichen sclerosus under control with steroid creams. I also met a wonderful partner who helped get my sex life back on track, received invaluable practical and emotional support from the London Vulval Pain Support Group, and reduced my stress levels with exercise and meditation. I still have bad flare ups of symptoms from time to time, but I also enjoy long phases of remission when I am able to have pain-free sex and lead a normal life.

How can vulval pain be managed?

Management options in primary care

The management of vulval pain depends on the cause. However, most vulval problems can be managed adequately in primary care, and basic treatment can be of great benefit to patients. Reassurance is important. Many patients respond to simple interventions such as the regular use of emollients. Advice about not washing the genital area with soaps and scented products is likely to reduce irritation.

A diagnosis of acute vulvovaginal candidiasis can be made in women with a short history of vulval pain and itching and vulval erythema, oedema, and a white curd-like discharge on examination. Prescribe antifungal drugs immediately without waiting for confirmatory swabs. Guidelines for the management of acute and chronic candidiasis have been developed by the British Association for Sexual Health and HIV.[17]

The British Association of Dermatologists has produced evidence based guidance on the management of lichen sclerosus that includes the use of ultrapotent topical corticosteroids and emollients.[18]

Referral to secondary care

Referral to specialist care is indicated if the diagnosis is uncertain; if the woman responds poorly to treatment; or she has a suspicious vulval lesion that is pigmented, white, ulcerated, or eroded (in which case, urgent referral is indicated because the lesion may be neoplastic).[19]

Refer patients with confirmed inflammatory skin disease whose symptoms cannot be controlled to a dermatologist, gynaecologist, or genitourinary medicine specialist, depending on local services. Patients with an optimally treated inflammatory skin disease who still have vulval pain may need to be referred to a multidisciplinary specialist service.

Ideally, secondary level care should be team based because different aspects of vulval disease may need to be dealt with by different specialties.[19] This team can comprise representatives from gynaecology, dermatology, genitourinary medicine, physiotherapy, psychosexual therapy, clinical psychology, and pain management. Ideally, women with refractory vulval pain should be referred to multidisciplinary clinics that specialise in vulval disease.

How is vulvodynia managed?

The British Society for the Study of Vulval Diseases has recently published guidelines for the management of vulvodynia that focus on the multidisciplinary approach to patients and the importance of a clinical "lead" to assess, diagnose, and triage patients to the different supportive specialties.[14] The guidelines are based on the best available evidence from clinical trials.

ADDITIONAL EDUCATIONAL RESOURCES

Resources for healthcare professionals

British Society for the Study of Vulval Diseases (www.bssvd.org)—UK multidisciplinary specialist society for vulval disease

Nunns D, Mandal D. Vulvodynia guidelines. 2010 www.onlinelibrary.wiley.com/doi/10.1111/j.1365-2133.2010.09684.x/abstract;jsessionid=50678EFF08892EA8964A4B104B784BB0.d02t04. Evidence based guidelines produced by the UK British Society for the Study of Vulval Diseases guidelines group

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Clinical Effectiveness Group, British Association for Sexual Health and HIV. UK guidelines on the management of vulvovaginal candidiasis. 2007. www.bashh.org/documents/1798

Royal College of Obstetricians and Gynaecologists. Vulval skin disorders, management (Green-top 58). www.rcog.org.uk/womens-health/clinical-guidance/management-vulval-skin-disorders-green-top-58. Guidance intended for the general gynaecologist, with advice on when to refer to the specialist multidisciplinary team

Genital disorders modules in the dermatology elearning project (www.e-lfh.org.uk/projects/dermatology/index.html)—Online training course that is free to UK doctors

University of Hertfordshire vulval disease course (www.health.herts.ac.uk/uhpgms/documents/vulval%20disorders2011%20Flyer.pdf)—Two day short course for healthcare professionals keen to know more about vulval problems (March each year)

Resources for patients

Vulval Pain Society (www.vulvalpainsociety.org)—Provides support and information for women with vulvodynia; contains information on workshops for women and links to patient support groups

British Pain Foundation (www.britishpainsociety.org/patient_publications.htm)—Provides useful information for women with vulval pain

UK Lichen Planus (www.uklp.org.uk/)—Provides support and information for women and men with lichen planus including non-genital LP

National Vulvodynia Association (www.nva.org/)—US non-profit organisation that aims to improve the lives of women with vulvodynia; provides education, networking, support, awareness, and advocacy

Worldwide Lichen Sclerosus Support (www.lichensclerosus.org/)—Provides support and information for women and men with lichen sclerosus

QUESTIONS FOR FUTURE RESEARCH

What is the true prevalence of vulval pain?

What is the best multidisciplinary model to manage vulvodynia?

Which drugs work best to treat unprovoked vulvodynia?

What is the best therapeutic approach for managing provoked vulvodynia?

Drug based treatments

A large body of evidence suggests that women who have unprovoked pain can be managed in a similar way to those with other neuropathic pain conditions.[20 21] The National Institute for Health and Clinical Excellence (NICE) recommends the use of a tricyclic antidepressant or pregabalin (or both) for neuropathic pain, although it does not list unprovoked vulvodynia in the list of neuropathic conditions.[22] Tricyclic antidepressants, gabapentin, and pregabalin have shown benefit in the treatment of unprovoked vulvodynia in some small cohort studies, however.[20 21 23] A 47% complete response (no pain at six months) rate has been reported.[24] The findings of a qualitative study suggest that a combination of different treatment strategies, such as drugs and physical therapies, improves outcomes.[9 24]

For patients with provoked (sexual) pain, the benefit of drugs such as tricyclic antidepressants and gabapentin is less clear, and these treatments would not be considered first line.

Topical local anaesthetics such as 5% lidocaine gel or ointment are commonly prescribed for women with vulvodynia, but few controlled studies are available, so the effectiveness of these treatments is not clear. One randomised controlled trial found a better response rate for placebo than for topical lidocaine in women with vestibulodynia (33% *v* 20%), but this was not significant.[25] In addition, local anaesthetics are potent sensitisers and should be used with caution on the vulva because they can lead to allergic contact dermatitis.

Psychological and psychosexual therapy

Women with provoked pain may have a problem with hypersensitivity to touch (allodynia), which leads to superficial dyspareunia and tampon intolerance. The British Society for the Study of Vulval Diseases guidelines recommends psychosexual and physical therapy techniques as first line treatments to desensitise the vulva.[14] It is unclear exactly how these techniques work, but they do help reverse an apparent hypertonicity in the levator ani (pelvic floor muscles) and help patients overcome a phobia of genital touch.[26] A variety of physical therapies has been reported to show benefit including pelvic floor muscle biofeedback, self massage of the vulva, vaginal TENS (transcutaneous electrical nerve stimulation), and the use of vaginal trainers.[26 w2-w5] No studies have reported an optimal technique, and success depends on several factors, including the therapist, degree of patient support, and duration and number of sessions. Biofeedback therapy has been used successfully to help overcome pelvic floor muscle dysfunction in women with provoked vulvodynia, with response rates of 52%.[26]

Patients with provoked pain may benefit from a psychological approach initially. In one randomised controlled trial, 78 women with provoked pain were randomised to one of three arms: group cognitive behavioural therapy (for 12 weeks), pelvic floor biofeedback (for 12 weeks), and vestibulectomy.[27] At six months' follow-up, all patients reported improvements in pain scores, although no significant difference was seen between groups. Surgery produced the greatest improvement in sexual functioning, but the high number of participants in the surgery arm who declined to be included in the study was of concern. The study supported both forms of non-surgical treatment and suggested that patients prefer a behavioural approach to treatment than a surgical one.

Surgery

Surgical excision of the vestibule (vestibulectomy) may benefit a minority of patients with provoked pain and is per-

formed more commonly in the United States.[28] The procedure that yields the best result is the modified vestibulectomy, in which a horseshoe shaped area of the vestibule and inner labial fold is excised, followed by advancement of the posterior vaginal wall.[28 w6 w7] Patients with provoked pain may have complex psychosexual and psychological problems, and outcomes after surgery can be poor if these problems are not dealt with.[29] Surgery should not be the first line treatment for women with provoked vulvodynia, and those who do undergo surgery will need psychosexual support preoperatively and postoperatively to ensure the best outcome.[30] Surgery for women with unprovoked vulvodynia is not appropriate because it does not deal with the complex chronic pain issues that these women experience.

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Patient consent obtained.

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ANSWERS TO ENDGAMES, p 50

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PICTURE QUIZ

A 79 year old man with a lesion on his cheek

1 Basal cell carcinoma.
2 Malignant.
3 The main risk factor for basal cell carcinoma is exposure to ultraviolet light. Genetic risk factors include having fair skin, blue eyes, or red hair and having a family history of skin cancer. Acquired risk factors include exposure to ultraviolet light, radiation injury, immunosuppressive drugs, and trauma.
4 This is a high risk lesion because it is on the central face, one of the areas identified by the National Cancer Peer Review Programme as having a high risk of recurrence after treatment.
5 This basal cell carcinoma should be treated by surgery (wide local excision or Mohs micrographic surgery).

CASE REPORT

A woman with ketoacidosis but not diabetes

1 Alcoholic ketoacidosis.
2 Isopropanol toxicity, ethylene glycol toxicity, and diabetic ketoacidosis.
3 Intravenous normal saline and dextrose.

STATISTICAL QUESTION

Confidence intervals and statistical significance

Statement *b* is true, whereas *a* is false.