# Vulvodynia

Vulvodynia literally means vulval (vulvar) pain. It can be felt in the skin of the mons pubis, clitoris, labia majora, labia minora and the mucosa of vestibule of the vagina. Pain may also involve the perineum, perianal skin and extend to the inguinal creases and medial thighs.

Vulval pain has many causes. It may be secondary to infectious, inflammatory or neoplastic conditions of the vulva, or may be referred from myofascial or skeletal structures, or from pelvic organs.

Vulvodynia is the term used when a nociceptive cause of local and referred pain has been excluded. It may sometimes be preceded by an episode of painful vulval infection or skin disease or arise de novo.

Understanding and therefore managing vulvodynia requires knowledge of the current concepts of pain pathophysiology. Vulvodynia may be loosely classified as a type of neuropathic pain but does not affect a named sensory peripheral nerve. It also exhibits some features of a complex regional pain syndrome.

A thorough history is the most important requirement for diagnosis. Examination is limited and usually normal. Extensive and complicated investigations are usually not required but most women will need assessment by a pelvic floor physiotherapist and some women will require referral to specialists in regional centres.

The International Society for the Study of Vulvovaginal Disease (ISSVD) has classified vulval pain into vulval pain related to a specific disorder, and vulvodynia (see Box 1 below).

Box 1: International Society for the Study of Vulvovaginal Disease Terminology and Classification of Vulvar Pain

Category A: Vulvar pain related to a specific disorder

- Infectious
- Inflammatory
- Neoplastic
- Neurologic

Cathegory B: Vulvodynia

1. Generalized
- provoked - sexual or non sexual
- unprovoked
- mixed
2. Localized – (eg, vestibulodynia, clitorodynia)
- provoked
- unprovoked
- mixed

**Category A : Vulvar pain related to a specific disorder**

Category A relates to secondary causes of vulval pain that can be attributed to a specific or visible disorder. In these cases pain is a symptom. For example, in inflammatory dermatoses pain is due to a breach of the epithelium of the skin or mucosa such as an excoriation, fissure, erosion or ulcer. Conditions such as dermatitis (eczema), psoriasis and lichen sclerosus usually present with itch but can cause pain if there is epithelial damage.

Lichen planus can be itchy if it involves the keratinized epithelium of the vulva but commonly involves the vaginal mucosa where it causes painful erosions. Infection with herpes simplex virus causes direct stimulation of nerve endings as well as erosions. Candidiasis is predominantly pruritic, but can cause pain if fissures are present.

Neoplastic conditions such as vulval intraepithelial neoplasia, squamous cell carcinoma or extra-mammary Paget's disease can cause pain, usually because of epithelial damage.

Pudendal nerve compression causes true neuropathic pain. As this is a specific diagnosis involving a named nerve this would not be classified as vulvodynia. Vulval pain can also be secondary to myofascial pelvic floor dysfunction or referred from musculoskeletal structures in the lower back, sacrum and pelvis, or from pelvic organs.

Psychiatric conditions (somatoform and conversion disorders, Munchausen's syndrome) should also be considered but are rare.



Home truth: History will provide most of the information needed for diagnosis

**Category B: Vulvodynia**

Vulvodynia is a primary pain condition. There is no identifiable nociceptive cause and pain is the diagnosis rather than a symptom. In these cases, neuroplasticity is maladaptive. Sensitization and amplification of neural stimuli occur both peripherally where pain is felt, and centrally in the spinal cord and brain.

Peripheral nociceptors become sensitized to noxious stimuli. Type C nerve fibres which transmit pain discharge more easily at lower levels of stimulation and at lower thresholds, and even spontaneously. Resulting pain memory leads to pain sensitization where pain signals are transmitted unnecessarily, long after any original source of pain has healed, and becomes chronic. Persistent stimulation in pain fibres can elicit chemical and anatomic reorganization in spinal cord neurons and brain.

Psychological factors alter the responsiveness of the CNS and amplify incoming signals, with frequent effects on cognition and behavior. There is often associated anxiety, fear and depression. Fortunately, neuroplasticity also means these changes are not irreversible.

In vulvodynia, myosfascial structures of the pelvic floor are often hypertonic. This induces tissue ischaemia, inflammation, and nerve activation leading to release of inflammatory mediators and neurotransmitters causing neurogenic inflammation.

There may be symptoms arising from other organs such as bladder (interstitial cystitis/painful bladder syndrome), lower bowel (irritable bowel syndrome) and muscles and joints (fibromyalgia). This multisystem presentation can be explained by shared innervation between organs. Thus a vicious cycle of pain amplification ensues.

## Making the diagnosis

Nociceptive pain can be described specifically and localized fairly accurately. There is a visible, although sometimes subtle abnormality causing the pain. By contrast neuropathic pain is poorly localized and adjectives used for description are vague. Symptoms are often out of proportion to examination findings.

Women presenting with vulvodynia be divided in to those who present with generalized burning (generalized unprovoked vulvodynia ) or with pain localized to vaginal introitus triggered by penetrative sexual intercourse or insertion of tampons (localized provoked vestibulodynia). A less common presentation is localized unprovoked clitorodynia.

**History**

Ask the patient to describe the pain. Neuropathic pain is often described as a rawness, soreness, or burning. In generalized vulvodynia pain is diffuse, poorly localized, and may radiate to the medial thighs. It often increases in intensity throughout the day and worsens with sitting rather than standing or lying down. Use of a pain chart or Visual Analogue Scale can be helpful.

Pain is localized to the vaginal introitus but may be felt in the posterior fourchette or circumferentially in women with localized provoked vestibulodynia. Patients often report a tearing sensation with intercourse. Stinging with application of topical treatment especially topical corticosteroids is common.

Depending on symptoms, other diagnoses to consider include pudendal neuropathy, atrophic vaginitis, pelvic pathology, referred pain or a somatoform disorder. Systems review should include bladder and bowel symptoms. Ask about associated sleep disturbance, fatigue, appetite or mood changes and the effect on relationship or work

Take an obstetric, gynaecological and sexual history, including Pap smear results. Ask about vulval skin care routine, especially application of topical agents to vulval skin and whether these are helpful or aggravate symptoms. Review all medications.

**Examination**

The skin usually appears normal except for erythema due to vasodilatation (attributed to autonomic dysfunction and neurogenic inflammation). There may be hypersensitivity to light touch. Look carefully with magnification and good illumination for subtle signs of dermatological disease. If possible gently stretch skin to examine interlabial sulci and the posterior fourchette to look for fissuring.

Mild lichenification may manifest as altered skin texture with increased skin markings, and possible hyper- or hypo- pigmentation. If there is erythema in the

vaginal introital mucosa, look for erosions suggesting lichen planus, or the glazed petechial appearance seen in plasma cell vulvitis. Speculum examination should be considered if deep vaginal involvement is suspected but is often not possible because of associated vaginismus. If pelvic pathology is suspected gentle bimanual examination may be attempted.

**Investigations**
Take a low vaginal swab looking for any evidence of infection. If fissures or erosions are present also perform a viral swab for HSV PCR. Pelvic ultrasound is indicated if pelvic pathology is suspected.

Biopsy is not usually helpful when only non specific diffuse erythema is present. However localized erythema, especially with a greyish border, may be suggestive of lichen planus and biopsy warranted. Always biopsy any erosion or ulcer using a 3mm punch biopsy from the edge of lesion. Topical anaesthetic cream prior to infiltrative local anaesthetic may be helpful in reducing injection pain. A suture is not always required if a topical haemostatic agent such as silver nitrate is used.

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## Management

A pelvic floor assessment by a pelvic floor physiotherapist is helpful in most patients and early referral should be considered. If you feel you cannot exclude a subtle dermatosis, a dermatologist referral is recommended. Similarly refer to a gynaecologist or gastroenterologist if you suspect pelvic organ pathology.

If pudendal neuropathy is suspected refer the patient to a women's hospital with a specialized clinic, or if specific spinal nerve root involvement refer to an appropriate surgeon for consideration of MRI. In rare cases where you suspect a true psychosis such as somatoform disorder, offer referral to a psychiatrist, although in practice this is rarely accepted.

**General treatment**

Women with vulvodynia need empathy and support. Education about anatomical terms and the pathogenesis of chronic pain will aid understanding. The aim of treatment is to manage and live with the pain: it should not be expected to completely or rapidly resolve. Aim for an improved quality of life achieved in small steps by realistic goal setting.

Stop all topical treatment especially corticosteroid and antifungal creams. Advise the woman to wear loose clothing and use a non-soap cleanser or emulsifying ointment diluted with tap water to wash. Give attention to sleep, diet and exercise, limiting alcohol and caffeine intake. Treat any coexisting constipation.

## Specific treatments

Physical
Whether vulvodynia is primary or secondary there is usually a component of increased tone in the pelvic floor muscles contributing to, if not the cause of, the pain. Early referral to physiotherapist is important. Most women will accept this suggestion, and often find the physiotherapist plays an important supportive role in their treatment. Frequent visits are not usually required.

Treatment includes education and retraining of pelvic floor muscles to decrease tone and sensitivity. Physiotherapists can also instruct about the use of dilators which are helpful not only for muscle retraining but also to for reduction of hyperasesthesia which assists desensitization.

Psychological
Specific referral to a psychologist is not necessary for all women with vulvodynia. Explanation and reassurance which allows self help may be sufficient. Many women have been told that the pain is 'all in their heads'. Try to put a positive spin on this statement by explaining the pathophysiology of chronic pain.

Depending on patient preference you may suggest meditation, relaxation, mindfulness or yoga (see Resources p x). However many patients still benefit from referral to a psychologist or, if dyspareunia is a significant symptom, or if vulvodynia is causing sexual difficulties consider referral to a sexual counselor.

Pharmacological
Treatment uses a multimodal approach. Medication alone can be expected to improve pain by about 50% in a quarter to a third of women. Topical treatment (see below) can be used alone if pain is mild, or as an adjunct to other strategies, or if oral medication is refused or contraindicated due to drug interaction or comorbidities.

Emulsifying ointment diluted with tap water can be soothing as a barrier and allows desensitization. Local anaesthetic (lignocaine 2% gel or 5% ointment) or amitryptilne 2% cream (available through compounding pharmacies) should be applied twice daily rather than 'as needed". Allow 4-6 weeks for the onset of action and continue for at least three months if effective.

Resistance to the suggestion of oral medication is commonly encountered because of fear of side effects or perceived stigma about taking 'antidepressants'. Preempt this by explaining that their primary use is for pain management and that they are rarely used now for treatment of depression. Encouragement and support is needed to continue therapy, especially initially and when doses are increased. Although many women are able to cease oral medication when they have improved, this is usually not for at least several months.

Start with the a tricyclic antidepressant (TCA), either amitryptiline or nortryptiline. The most common side effect, sedation, usually becomes less troublesome with time. Start with a low dose (5-10mg) and take at dinner time not bedtime. Advise patient to expect sedation in the first 3-4 days that should improve with time. Increase the dose by 5-10mg nocte every 2-3 weeks to a dose of 30-50mg nocte. Allow 6-8 weeks for onset of action but maximum efficacy make take 2-3 months or more. Withdraw medication over several weeks.

Adverse effects (dry mouth, blurred vision, constipation) are uncommon at the low doses used. In older women consider effect on cognition and cardiac conduction – nortryptiline is safer in this circumstance. TCAs are contraindicated in patients with narrow angle glaucoma and it is important to check drug interactions.

The "anticonvulsants" (alpha-2–delta ligands) gabapentin and pregabolin can be used if TCAs are contraindicated, cause side effects or are ineffective. At low doses they can also be used in combination with low dose TCAs. For pregabolin start at 25mg nocte and increase as tolerated titrating against side effects and efficacy. The maximum dose is 300mg twice daily but doses in excess of 150mg bd are commonly associated with side effects such as sedation, dizziness and weight gain.

Serotonin-Noradrenaline reuptake inhibitors (SNRIs) such as duloxetine, venlafaxine and desvenlafaxine are third line drugs. Their use in chronic pain is 'off label' and non PBS listed. They can be used in combination with the 'anticonvulsants". Opiates are rarely if ever indicated.

Surgery is rarely required. However a gynaecological opinion about Fenton's procedure and vestibulectomy can be sought in cases where localized introital pain and vaginismus persist. Botulinum toxin is used in tertiary referral centres and may be of benefit in some women. Women with refractory vulvodynia despite the measures listed above may benefit from referral to a multidisciplinary pain clinic.

Telehealth consultations may be useful for review of patients but the first assessment requires a face to face consultation. The sensitive nature of vulval examination is not suited to telemedicine and also subtle dermatological signs will be missed.

Gems

- Examination is usually normal in patients with vulvodynia
- Early referral to a pelvic floor physiotherapist should be considered in most cases
- Extensive investigations will usually not add to diagnosis if the history or examination are non contributory
- A multimodal approach is best for management