



The lower vagina and vulva are richly supplied with peripheral nerves and are, therefore, sensitive to pain, particularly the region of the hymeneal ring. Although the pudendal nerve (arrow) courses through the area, it is an uncommon source of vulvar pain.

FIRST OF 3 PARTS

VULVAR PAIN SYNDROMES

Your first challenge is making the correct diagnosis

➡ Many cases of generalized vulvodynia and localized vulvodynia (vestibulodynia) are mistakenly attributed to yeast infection, pudendal neuralgia, and other entities. Avoid those pitfalls by using a reliable roadmap for evaluation, differentiation, and identification of the various forms of vulvar pain.

Neal M. Lonky, MD, MPH, moderator; Libby Edwards, MD, Jennifer Gunter, MD, and Hope K. Haefner, MD, panelists

Although the incidence of vulvar pain has increased over the past decade—thanks to both greater awareness *and* increasing numbers of affected women—the phenomenon is not a recent development. As early as 1874, T. Galliard Thomas wrote, “[T]his disorder, although fortunately not very frequent, is by no means very rare.”¹ He went on to express “surprise” that it had not been “more generally and fully described.”

Despite the focus Thomas directed to the issue, vulvar pain did not get much attention until the 21st century, when a number of studies began to gauge its prevalence. For example, in a study in Boston of about 5,000 women, the lifetime prevalence of chronic vulvar pain was 16%.² And in a study in Texas, the prevalence of vulvar pain in an urban, largely minority population was estimated to be 11%.³ The Boston study also reported that “nearly 40% of women chose not to seek treatment, and, of those who did, 60% saw three or more doctors, many of whom could not provide a diagnosis.”²

Clearly, there is a need for comprehensive information on vulvar pain and its causes, symptoms, diagnosis, and

treatment. To address the lack of guidance, OBG MANAGEMENT Contributing Editor Neal M. Lonky, MD, assembled a panel of experts on vulvar pain syndromes and invited them to share their considerable knowledge. The ensuing discussion, presented in three parts, offers a gold mine of information.

In this opening article, the panel focuses on causes, symptomatology, and diagnosis of this common complaint. In Part 2, which will appear in the October issue of this journal, the focus is the bounty of treatment options. Part 3 follows in November, when the discussion shifts to vestibulodynia.

Common diagnoses—and misdiagnoses

Dr. Lonky: What are the most common diagnoses when vulvar pain is the complaint?

Dr. Gunter: The most common cause of chronic vulvar pain is vulvodynia, although lichen simplex chronicus, chronic yeast infections, and non-neoplastic epithelial disorders, such as lichen sclerosus and lichen planus, can also produce irritation and pain. In postmenopausal women, atrophic vaginitis can

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The OBG MANAGEMENT expert panel



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also cause a burning pain, although symptoms are typically more vaginal than vulvar. Yeast and lichen simplex chronicus typically produce itching, although sometimes they can present with irritation and pain, so they must be considered in the differential diagnosis. It is important to remember that many women with vulvodynia have used multiple topical agents and may have developed complex hygiene rituals in an attempt to treat their symptoms, which can result in a secondary lichen simplex chronicus.

That said, there is a high frequency of misdiagnosis with yeast. For example, in a study by Nyirjesy and colleagues, two thirds of women who were referred to a tertiary clinic for chronic vulvovaginal candidiasis

were found to have a noninfectious entity instead—most commonly lichen simplex chronicus and vulvodynia.⁴

Dr. Edwards: The most common “diagnosis” for vulvar pain is vulvodynia. However, the definition of vulvodynia is pain—i.e., burning, rawness, irritation, soreness, aching, or stabbing or stinging sensations—in the absence of skin disease, infection, or specific neurologic disease. Therefore, even though the usual cause of vulvar pain is vulvodynia, it is a diagnosis of exclusion, and skin disease, infection, and neurologic disease must be ruled out.

In regard to infection, *Candida albicans* and bacterial vaginosis (BV) are usually the first conditions that are considered when a patient complains of vulvar pain, but they are not common causes of vulvar pain and are never causes of chronic vulvar pain. Very rarely they may cause recurrent pain that clears, at least briefly, with treatment.

Candida albicans is usually primarily pruritic, and BV produces discharge and odor, sometimes with minor symptoms. Non-albicans *Candida* (e.g., *Candida glabrata*) is nearly always asymptomatic, but it occasionally causes irritation and burning.

Group B streptococcus is another infectious entity that very, very occasionally causes irritation and dyspareunia but is usually only a colonizer.

Herpes simplex virus is a cause of recurrent but not chronic pain.

Chronic pain is more likely to be caused by skin disease than by infection. Lichen simplex chronicus causes itching; any pain is due to erosions from scratching.

Dr. Haefner: Several other infectious conditions or their treatments can cause vulvar pain. For example, herpes (particularly primary herpes infection) is classically associated with vulvar pain. The pain is so great that, at times, the patient requires admission for pain control. Surprisingly, despite the known pain of herpes, approximately 80% of patients who have it are unaware of their diagnosis.

Although condyloma is generally a painless condition, many patients complain of pain following treatment for it, whether treatment

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TABLE 1 Terminology and classification of vulvar pain from the International Society for the Study of Vulvovaginal Disease

A. Vulvar pain related to a specific disorder	
1.	Infectious (including candidiasis, herpes)
2.	Inflammatory (lichen planus, immunobullous disorders)
3.	Neoplastic (Paget’s disease, squamous cell carcinoma)
4.	Neurologic (herpes neuralgia, spinal nerve compression)
B. Vulvodynia	
1.	Generalized
a.	Provoked (sexual, nonsexual, or both)
b.	Unprovoked
c.	Mixed (provoked and unprovoked)
2.	Localized (including vestibulodynia, clitorodynia, hemivulvodynia)
a.	Provoked (sexual, nonsexual, or both)
b.	Unprovoked
c.	Mixed (provoked and unprovoked)

SOURCE: Moyal-Barracco and Lynch.⁵ Reproduced with permission from the *Journal of Reproductive Medicine*.

“Skin diseases that affect the vulva are usually pruritic—pain is a later sign.”
—Libby Edwards, MD

involves topical medications or laser surgery.

Chancroid is a painful vulvar ulcer. Trichomonas can sometimes be associated with vulvar pain.

Dr. Lonky: What terminology do we use when we discuss vulvar pain?

Dr. Haefner: The current terminology used to describe vulvar pain was published in 2004, after years of debate over nomenclature within the International Society for the Study of Vulvovaginal Disease.⁵ The terminology lists two major categories of vulvar pain:

- **pain related to a specific disorder.** This category encompasses numerous conditions that feature an abnormal appearance of the vulva (**TABLE 1**).
- **vulvodynia**, in which the vulva appears normal, other than occasional erythema, which is most prominent at the duct openings (vestibular ducts—Bartholin’s and Skene’s).

As for vulvar pain, there are two major forms:

- **hyperalgesia** (a low threshold for pain)
- **allodynia** (pain in response to light touch).

Some diseases that are associated with vulvar pain do not qualify for the diagnosis of vulvodynia (**TABLE 2**, page 54) because they are associated with an abnormal appearance of the vulva.

What needs to be ruled out for a diagnosis of vulvodynia?

Dr. Lonky: What skin diseases need to be ruled out before vulvodynia can be diagnosed?

Dr. Edwards: Skin diseases that affect the vulva are usually pruritic—pain is a later sign. Lichen simplex chronicus (also known as eczema) is pruritus caused by any irritant; any pain that arises is produced by visible excoriations from scratching.

Lichen sclerosus manifests as white epithelium that has a crinkling, shiny, or waxy texture. It can produce pain, especially dyspareunia. The pain is caused by erosions that arise from fragility and introital narrowing and inelasticity.

Vulvovaginal lichen planus is usually erosive and preferentially affects mucous membranes, especially the vestibule; it sometimes affects the vagina and mouth, as well.

Desquamative inflammatory vaginitis is most likely a skin disease that affects only the vagina. It involves introital redness and a clinically and microscopically purulent vaginal discharge that also reveals parabasal cells and absent lactobacilli.

Dr. Lonky: You mentioned that neurologic diseases can sometimes cause vulvar pain. Which ones?

Dr. Edwards: Pudendal neuralgia, diabetic neuropathy, and post-herpetic neuralgia are the most common specific neurologic causes of vulvar pain. Multiple sclerosis can also produce pain syndromes. Post-herpetic neuralgia follows herpes zoster—not herpes simplex—virus infection.

Dr. Lonky: Any other conditions that can cause vulvar pain?

Dr. Haefner: Aphthous ulcers are common and are often flared by stress.

Non-neoplastic epithelial disorders are also seen frequently in health-care providers’

TABLE 2 Conditions other than vulvodynia that are associated with vulvar pain

Acute irritant contact dermatitis (e.g., erosion due to podofilox, imiquimod, cantharidin, fluorouracil, or podophyllin toxin)
Aphthous ulcer
Atrophy
Bartholin's abscess
Candidiasis
Carcinoma
Chronic irritant contact dermatitis
Endometriosis
Herpes (simplex and zoster)
Immunobullous diseases (including cicatricial pemphigoid, pemphigus vulgaris, linear immunoglobulin A disease, etc.)
Lichen planus
Lichen sclerosus
Podophyllin overdose (see above)
Prolapsed urethra
Sjögren's syndrome
Trauma
Trichomoniasis
Vulvar intraepithelial neoplasia

“Sixty-five percent to 75% of women referred to my clinic with chronic yeast actually have lichen simplex chronicus or vulvodynia.”

—Jennifer Gunter, MD

offices; many patients who experience them report pain on the vulva.

It is always important to consider cancer when a patient has an abnormal vulvar appearance and pain that has persisted despite treatment.

What are the most common vulvar pain syndromes?

Dr. Lonky: If you were to rank vulvar pain syndromes according to their prevalence, what would the most common syndromes be?

Dr. Gunter: Given the misdiagnosis of many women, who are told they have chronic yeast infection, as I mentioned, it's hard to know which vulvar pain syndromes are most prevalent. I suspect that lichen simplex chronicus

is most common, followed by vulvodynia, with chronic yeast infection a distant third.

My experience reflects what Nyirjesy and colleagues⁴ found: 65% to 75% of women referred to my clinic with chronic yeast actually have lichen simplex chronicus or vulvodynia. In postmenopausal women, atrophic vaginitis is also a consideration; it's becoming more common now that the use of systemic hormone replacement therapy is decreasing.

Dr. Lonky: What about subsets of vulvodynia? Which ones are most common?

Dr. Edwards: There is good evidence of marked overlap among subsets of vulvodynia. The vast majority of women who have vulvodynia experience primarily provoked vestibular pain, regardless of age. However, I find that almost all patients also report pain that extends beyond the vestibule at times, as well as occasional unprovoked pain.

The diagnosis requires the exclusion of other causes of vulvar pain, and the subset is identified by the location of pain (that is, is it strictly localized or generalized or even migratory?) and its provoked or unprovoked nature.

Localized clitoral pain and vulvar pain localized to one side of the vulva are extremely uncommon, but they do occur. And although I rarely encounter teenagers and prepubertal children who have vulvodynia, I do have patients in both age groups who have vulvodynia.

Dr. Lonky: Are there racial differences in the prevalence of vulvodynia?

Dr. Edwards: Although several good studies show that women of African descent and white patients are equally likely to experience vulvodynia, the vast majority (99%) of my patients who have vulvodynia are white. My patients of African descent consult me primarily for itching or discharge.

My local demographics prevent me from judging the likelihood of Asians having vulvodynia, and our Hispanic population has limited access to health care.

In general, I don't think that demographics are useful in making the diagnosis of vulvodynia.

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Do women who have vulvar pain tend to have comorbidities?

Dr. Lonky: Do your patients who have vulvodynia or another vulvar pain syndrome tend to have comorbidities? If so, is this information helpful in establishing the diagnosis and planning therapy?

Dr. Haefner: Women who have vulvodynia often have other medical problems as well. In my practice, when new patients who have vulvodynia complete their intake survey, they often report a history of headache, irritable bowel syndrome, interstitial cystitis, fibromyalgia,⁶ chronic fatigue syndrome, back pain, and temporomandibular joint (TMJ) disorder. These comorbidities are not particularly helpful in establishing the diagnosis of vulvodynia, but they are an important consideration when choosing therapy for the patient. Often, the medications chosen to treat one condition will also benefit another condition. However, it's important to check for potential interactions between drugs before prescribing a new treatment.

Dr. Gunter: A significant number of women who have vulvodynia also have other chronic pain syndromes. For example, the incidence of bladder pain syndrome–interstitial cystitis is 68% to 82% among women who have vulvodynia, compared with a baseline rate among all women of 6% to 11%.^{7–10} The rate of irritable bowel syndrome is more than doubled among women who have vulvodynia, compared with the general population (27% versus 12%).⁸ Another common comorbidity, hypertonic somatic dysfunction of the pelvic floor, is identified in 10% to 90% of women who have chronic vulvar pain.^{8,11,12} These women also have a higher incidence of non-genital pain syndromes, such as fibromyalgia, migraine, and TMJ dysfunction, than the general population, as Dr. Haefner noted.^{8,12,13}

Many studies have evaluated psychological and emotional contributions to chronic vulvar pain. Pain and depression are intimately related—the incidence of depression among all people who experience chronic pain ranges from 27% to 54%, compared with 5% to 17% among the general

population.^{14–16} The relationship is complex because chronic illness in general is associated with depression. Nevertheless, several studies have noted an increase in anxiety, stress, and depression among women who have vulvodynia.^{17–19}

I screen every patient for depression using a Patient Health Questionnaire (PHQ-9); I also screen for anxiety. I find that a significant percentage of patients in my clinic are depressed or have an anxiety disorder. Failure to address these comorbidities makes treatment very difficult. I typically prescribe citalopram (Celexa), although there is some question whether it can safely be combined with a tricyclic antidepressant. We also offer stress-reduction classes, teach every patient the value of diaphragmatic breathing, offer mind-body classes for anxiety and stress, and provide intensive programs where the patient can learn important self-care skills, such as pacing (spacing activities throughout the day in a manner that avoids aggravating the pain), and address her anxiety and stress in a more guided manner. We also have a psychologist who specializes in pain for any patient who may need one-on-one counseling.

Dr. Edwards: The presence of comorbidities is somewhat useful in making the diagnosis of vulvodynia. I question my diagnosis, in fact, when a patient who has vulvodynia does *not* have headaches, low energy, depression, anxiety, irritable bowel syndrome, constipation, fibromyalgia, chronic fatigue, sensitivity to medications, TMJ dysfunction, or urinary symptoms.

How common is pudendal neuralgia?

Dr. Lonky: How prevalent is a finding of pudendal neuralgia?

Dr. Edwards: The prevalence and incidence of pudendal neuralgia are not known. Those who specialize in this condition think it is relatively common. I do not identify or suspect it very often. Its definitive diagnosis and management are outside the purview of the general gynecologist, but the general

“Women who have vulvodynia often have other medical problems as well.”

—Hope K. Haefner, MD

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TABLE 3 Nantes Criteria for pudendal neuralgia by pudendal nerve entrapment**Essential criteria**

- Pain in the territory of the pudendal nerve: from the anus to the penis or clitoris
- Pain is predominantly experienced while sitting
- The pain does not wake the patient at night
- Pain with no objective sensory impairment
- Pain relieved by diagnostic pudendal nerve block

Complementary diagnostic criteria

- Burning, shooting, stabbing pain; numbness
- Allodynia or hyperpathia
- Rectal or vaginal foreign body sensation (sympathalgia)
- Worsening of pain during the day
- Predominantly unilateral pain
- Pain triggered by defecation
- Presence of exquisite tenderness on palpation of the ischial spine
- Clinical neurophysiology findings in men or nulliparous women

Exclusion criteria

- Exclusively coccygeal, gluteal, pubic, or hypogastric pain
- Pruritus
- Exclusively paroxysmal pain
- Imaging abnormalities able to account for the pain

Associated signs not excluding the diagnosis

- Buttock pain on sitting
- Referred sciatic pain
- Pain referred to the medial aspect of the thigh
- Suprapubic pain
- Urinary frequency or pain on a full bladder, or both
- Pain occurring after ejaculation
- Dyspareunia or pain after sexual intercourse, or both
- Erectile dysfunction
- Normal clinical neurophysiology

SOURCE: Labat et al.²¹ Reproduced with permission from *Neurology and Urodynamics*.

gynecologist should recognize the symptoms of pudendal neuralgia and refer the patient for evaluation and therapy.

Dr. Lonky: What are those symptoms?

Dr. Haefner: Pudendal neuralgia often occurs following trauma to the pudendal nerve. The pudendal nerve arises from sacral nerves, generally sacral nerves 2 to 4. Several tests can be utilized to diagnose this condition, including quantitative sensor tests, pudendal nerve motor latency tests, electromyography

(EMG), and pudendal nerve blocks.²⁰

Nantes Criteria allow for making a diagnosis of pudendal neuralgia (**TABLE 3**).²¹

Initial treatments for pudendal neuralgia should be conservative. Treatments consist of lifestyle changes to prevent flare of disease. Physical therapy, medical management, nerve blocks, and alternative treatments may be beneficial.

Pudendal nerve entrapment is often exacerbated by sitting (not on a toilet seat, however) and is reduced in a standing position. It tends to increase in intensity throughout the day.²² The final treatment for pudendal nerve entrapment is surgery if the nerve is compressed. By this time, the generalist is not generally the provider who performs the surgery.

Dr. Gunter: I believe pudendal neuralgia is sometimes overdiagnosed. EMG studies of the pudendal nerve, often touted as a diagnostic tool, are unreliable (they can be abnormal after vaginal delivery or vaginal hysterectomy, for example). In my experience, bilateral pain is less likely to be pudendal neuralgia; spontaneous *bilateral* compression neuropathy at exactly the same level is not a common phenomenon in chronic pain.

I reserve the diagnosis of pudendal neuralgia for women who have allodynia in the distribution of the pudendal nerve with severe pain on sitting, and who have exquisite tenderness when pressure is applied over the pudendal nerve (at the level of the ischial spine on vaginal examination). Typically, the vaginal sidewall on the affected side is very sensitive to light touch. I do see pudendal nerve pain after vaginal surgery when there has been some compromise of the pudendal nerve or the sacral plexus. This is typically unilateral pain.

Dr. Lonky: Thank you all. We'll continue our discussion, with a focus on treatment, in the October 2011 issue. 📧

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Examining the **EVIDENCE** CONTINUED FROM PAGE 15

“evidence-based medicine” undermines the very foundation of scientific investigation.

More questions than answers

The major conclusion of this study is that EFM protects against early neonatal death. So why is there no information about cause of death? These data should be readily available from a linked birth/death certificate data set. Such information might help to determine whether the excess early neonatal deaths were related to EFM or, more likely, to other variables surrounding or related to the delivery, such as the inability to

perform an emergency cesarean, if indicated, or the lack of providers skilled in neonatal resuscitation. ❌

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