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## Vulvar Pain Syndromes; Epithelial Ovarian Cancer

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## Table of Contents

### Chapter 1—Vulvar Pain Syndromes . . . . .2

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# Chapter 1—Vulvar Pain Syndromes

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## I. INTRODUCTION

Vulvodynia, or vulvar pain syndrome, is a condition that every gynecologist can expect to encounter. However, many diagnostic and therapeutic dilemmas are associated with this group of disorders, including a differential diagnosis that overlaps greatly among the disorders. Identifying the principle cause of vulvar pain is the first step in selecting appropriate therapy. This article provides a review of vulvar anatomy and describes diagnosis and management of the 5 most common disorders including vulvar vestibulitis, dysesthetic (“essential”) vulvodynia, vulvar dermatosis and dermatitis, vaginismus, and vulvar papillomatosis. A case study illustrating clinical features of one of the disorders that cause vulvodynia is presented, and a summary of key points in the diagnosis and treatment of the 5 most common disorders is also provided.

The term *vulvodynia* was first used in 1983 at the 7th congress of the International Society for the Study of Vulvar Disease (ISSVD); currently, the definition of the term is still evolving. At the ISSVD congress, vulvodynia was defined as chronic vulvar discomfort, further characterized by burning, stinging, irritation, or rawness. In addition, the ISSVD Committee on Vulvodynia allowed for the definition of subsets in order to individualize patient evaluation and treatment. Although this approach is conceptually satisfying (ie, vulvar discomfort must have different etiologies in different patients), in actual practice distinguishing signs and symptoms often overlap in individual patients. The degree of overlap has led some reviewers to conclude that the range of disorders associated with chronic vulvar discomfort are best characterized, not by subcategories, but by the generic term *vulvar pain syndrome*.<sup>1</sup>

### VULVAR ANATOMY

The external female genitalia include the mons pubis; labia majora and minora; clitoris, with its prepuce and frenulum; and vulvar vestibule. The Skene’s para-urethral glands, Bartholin’s (major vestibular) glands,

minor vestibular glands, and urethra all open into the vulvar vestibule (**Figure 1**). The vestibule is delineated superiorly by the clitoris, inferiorly by the posterior fourchette, by the hymen at its innermost boundary (which separates it from the vagina), and externally by a line described by D.B. Hart in 1882. Hart’s line, which can easily be seen at the inner aspect of the labia minora, marks a transition from (lateral) pigmented epithelium to (medial) nonpigmented epithelium.

Embryologically, structures external to Hart’s line are of ectodermal origin; structures internal to it—including the vestibule, urethra, and bladder—develop from the anterior portion of the cloaca and, therefore, are of endodermal origin, constituting the lowermost portion of the urogenital sinus. This common origin has led to the suggestion (supported by prevalence rates) that an etiologic association exists among vulvodynia, interstitial cystitis, and irritable bowel disease.<sup>2,3</sup>

Microscopically, the vulva is covered by a keratinized stratified squamous epithelium. The stratum corneum becomes progressively thinner toward the hymen; on the inner aspect of the hymen, the covering is a nonkeratinized mucous membrane rich in glycogen. In addition to perspiration from apocrine and eccrine sweat glands, significant transdermal water loss occurs because of the stratum corneum’s relative inefficiency. A moist environment supportive of microbial growth can be found even on external, nonmucosal surfaces. Therefore, careful assessment of changes in microbial flora is imperative in patients with vulvar pain.

### DIAGNOSTIC CATEGORIES

Patients presenting with vulvar discomfort frequently have physical findings—or an absence of findings—that provide the basis for diagnosing a subtype of vulvodynia. Generally, when a distinct subtype can be identified, a specific treatment can be shown to be effective. McKay<sup>4</sup> first defined the commonly encountered subtypes of vulvodynia (in order of decreasing frequency) as *vulvar dermatoses*, *cyclic vulvitis/candidiasis*, *squamous papillomatosis*, *vulvar vestibulitis*, and *essential vulvodynia*. Since McKay’s report was published, various reviewers have suggested