Chapter 1

The Basics

Patients with vulval problems have often spent many years in fruitless pursuit of a diagnosis and effective treatment. The reasons for this are as follows:

- Most vulval conditions are chronic dermatological diseases that cannot be cured and must therefore be managed. Such conditions on the vulva often look and behave differently from the same conditions on other parts of the skin; furthermore, management strategies must often be modified in order to be effective on the vulva.
- Dermatological disease of the vulva has far more importance for the patient than the same disease on less emotionally significant areas of the body. Patients frequently present not with symptoms of the vulval disease but with its sexual or relationship consequences. It is therefore no wonder that in the past women with vulval disorders have been unfairly labelled as 'psychosomatic'.
- The vulva and vagina are in the centre of the lower pelvis, are closely related to other pelvic organs and are bound to them by the myofascial structure of the pelvic floor (see Figures 1.1–1.3). Referred vulval pain from other pelvic viscera, and even from the spine and hip joints, is therefore an important concept in understanding vulvovaginal disorders.
- Vulval disease comes with a significant emotional overlay. Embarrassment commonly prevents patients from seeking help. Doctors are only human, and embarrassment can affect them too. Frequently, we hear from patients that they were given a prescription without being examined. It is often difficult for doctors to elicit an adequate history because of the intimate nature of a woman's symptoms and the need to take a detailed sexual and environmental history. Eliciting such histories takes patience and empathy. Patients may either avoid saying what is really on their mind or, alternatively, pour out huge amounts of disorganised, emotionally charged information. It is essential to help them to organise their thoughts. Start at the beginning and get them to think back on how their complaint evolved and how they came to be in your consulting room.

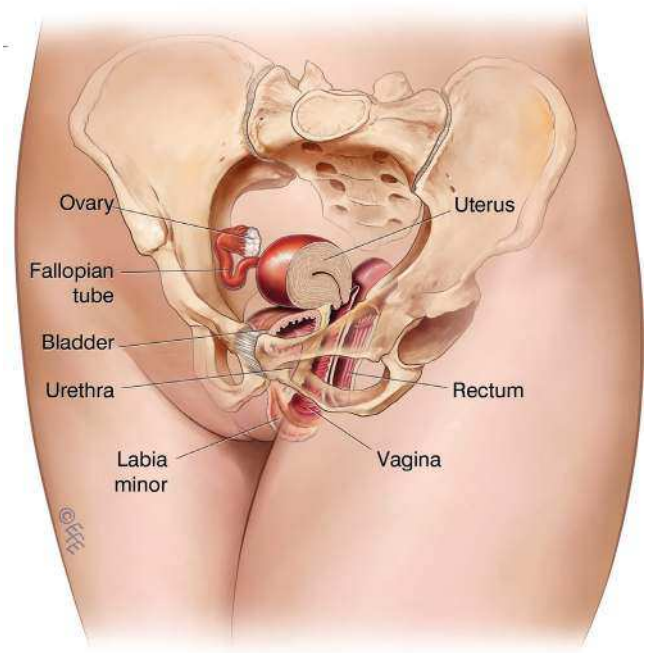

Figure 1.1 The female reproductive tract. With permission from Dr Levent Efe, CMI.

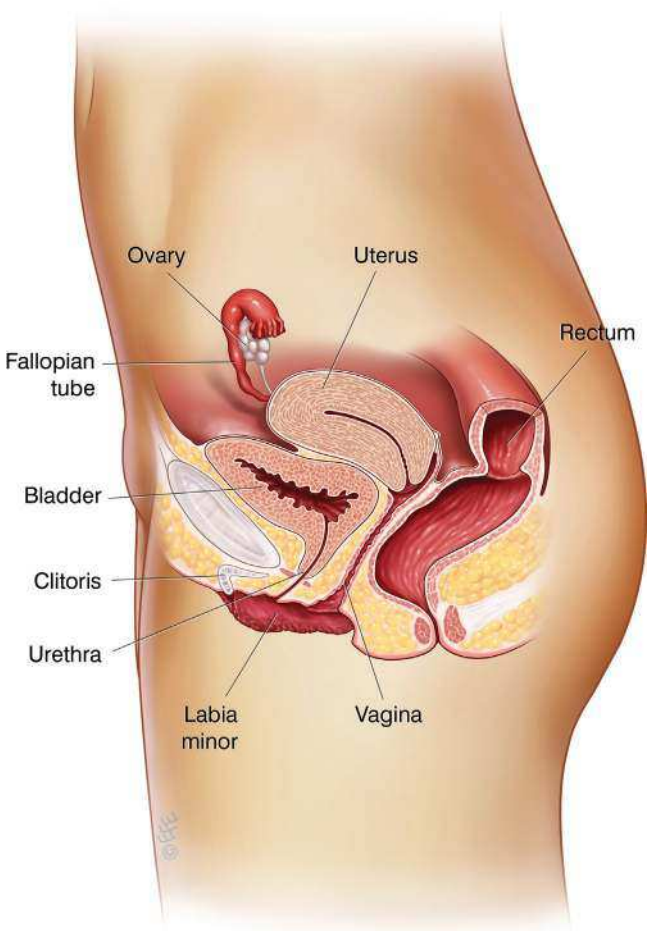

Figure 1.2 The female reproductive tract: side view. With permission from Dr Levent Efe, CMI.

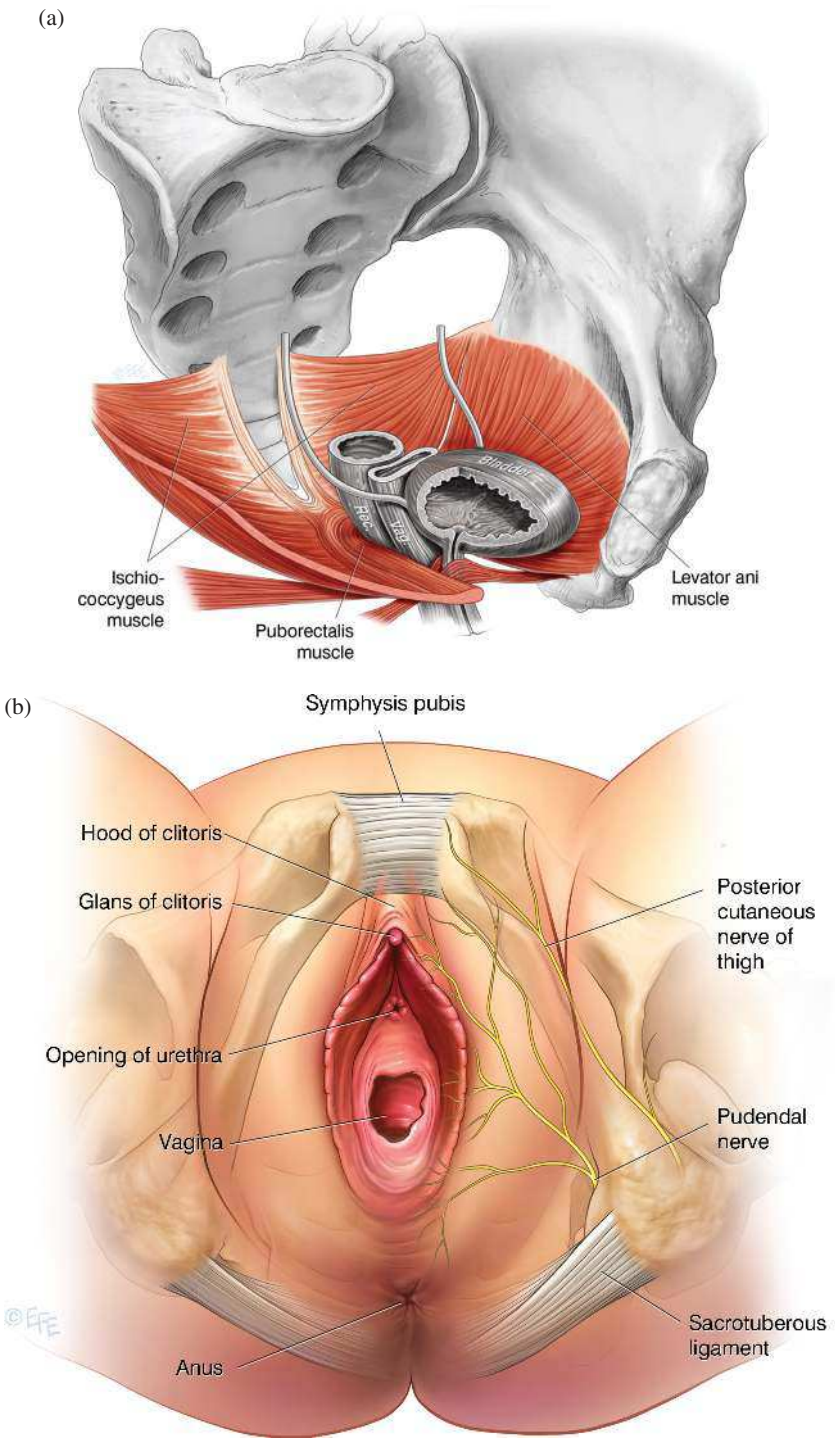

Figure 1.3 (a) The pelvic floor. (b) View of the vulval area. With permission from Dr Levent Efe, CMI.

An emerging issue in the management of vulval disease is the transgender patient. Male-to-female patients with a neovagina and female-to-male patients who have retained a vagina that has been changed by exposure to androgen form a unique group with their own special needs.

Understanding Vulval Conditions

The management of patients with vulval disease fundamentally requires an understanding of dermatological diagnosis and therapy, especially those skin diseases with a predilection for this part of the body. However, dermatological knowledge is not enough. An understanding of gynaecology, gastroenterology, urology, vaginal bacteriology, spinal function and dysfunction, and finally sexual medicine are all essential for optimal management.

Another challenge is the group of patients who appear to defy diagnosis. We believe that this group is very small indeed and that it is possible to classify virtually all patients. However, achieving this relies on very thorough history taking and on combining many different medical disciplines. The more difficult vulval cases are always multifactorial. The term 'vulvodynia' (vulval pain of unknown origin) is not used in this book, as we believe that a rational diagnosis can eventually be found for almost all vulval symptoms.

The purpose of this handbook is to introduce these concepts and to provide practical management recommendations. These are based on our 50 years of collective experience of helping patients with vulval disease, as well as published research by ourselves and others.

We hope this book will give clinicians the tools to approach vulval patients with confidence, and that this will in turn improve the lives of many women.

Anatomy

The Vulva

The vulva is not only part of the skin but is also the entrance to the genital tract. It is essential to understand that vulval skin extends all the way into the hymen. This means that rashes within the vaginal introitus are still classified as vulval, and the patient must be instructed to apply any treatment far enough inside to adequately control these rashes.

The vulva is exposed to many potential irritants, which can result in dermatological symptoms. These include:

- Vaginal discharge, both normal and abnormal
- Menstrual fluid
- Semen
- Urine
- Faeces
- Sweat
- Tight clothes
- Lubricants
- Perfumed products including toilet paper, wet wipes and feminine hygiene products
- Medications
- Pads and panty liners
- Hair-removal practices

The Vagina

The vagina is the conduit between the uterus and the vulva. Its mucosa is prone to similar diseases as in the mouth. Located in the middle of the lower pelvis (see Figure 1.1), its anatomical relationships include the following:

- Bladder and urethra
- Cervix
- Rectum and anus
- Uterovesical and rectovaginal peritoneal pouches
- Sacrum and coccyx

The Pelvic Floor

The pelvic floor is a myofascial structure that encompasses the entire pelvis (Figure 1.3). It is divided functionally into upper and lower parts. The upper part supports the bladder neck, cervix and upper rectum. The lower part supports the urethra, vagina, lower rectum and anus.

The pelvic floor acts as a conduit for pain referral throughout the pelvis. It is closely related to the muscles of the lower back and hip, and may be involved in lumbosacral dysfunction.

Innervation

The innervation of the lower vagina, vulva and anus is from sacral nerve roots S2, S3 and S4 via the pudendal nerve. The anterior vulva is supplied by the genital branch of the genitofemoral nerve (L1–L2) and the ilioinguinal nerve (L1). Thus, lumbosacral, coccygeal and even lower thoracic spinal disorders may produce referred vulval pain.

Clinical Presentation

The majority of patients presenting with chronic vulval symptoms have a skin disease such as eczema, psoriasis or lichen sclerosus, or are suffering from chronic vulvovaginal candidiasis. Many will have a personal or family history of the same condition or will have evidence of it elsewhere on their skin. Patients with eczema are usually atopic. This historical information can provide very helpful clues to a possible diagnosis.

When a patient presents with a vulval complaint, she usually complains of one or more of the following symptoms. Patients sometimes have trouble communicating their thoughts. It can be helpful to run through this list with them, in order to better delineate their real story:

- Itch
- Pre-menstrual or post-menstrual exacerbation of symptoms
- Irritation
- Soreness
- Pain
- Dyspareunia
- Burning
- Stinging
- Stabbing
- Crawling sensations (formication)

- Awareness of the vulva
- Dysuria

The duration of symptoms, any precipitating and exacerbating factors, and previous treatments should be recorded. Bladder, menstrual and bowel function also need to be recorded, as vulvovaginal disease is frequently associated with dysfunction in these systems. Spinal and hip joint disease and dysfunction in voiding may play a significant role in vulval symptoms and should be recorded.

What Do the Symptoms Mean?

Itch and *irritation* are usually due to a non-eroded inflammatory skin condition. *Burning*, *stinging*, *stabbing*, *formication* and *vulval awareness* are usually due to a neuromuscular dysfunction. *Soreness* and *pain* are often due to erosions or fissures, either part of a skin condition or secondary to excoriations produced by scratching.

Dyspareunia

Dyspareunia means pain with sexual intercourse. It is categorised into abdominal and vulvovaginal types. *Abdominal dyspareunia* usually relates to disease or dysfunction at or above the level of the cervix. The pain is experienced in the lower abdomen, as it relates to the upper pelvic floor. *Vulvovaginal dyspareunia* is caused by disease or dysfunction at the level of the lower pelvic floor. The pain is experienced at the vaginal entrance, or further up the vagina. This pain usually relates to a vulvovaginal skin condition, which produces vulval or introital tenderness, splitting or fissuring. It can also relate to dysfunction of the pelvic floor.

Important points to elicit in a history of dyspareunia are:

- Site of entry (or vaginal) dyspareunia (with a mirror if necessary)
- Onset of pain:
  - with foreplay (or masturbation)
  - during vaginal intercourse, or
  - after intercourse is concluded, and
  - whether gradual or sudden
- Duration of pain after intercourse
- Nature of the pain: tearing or splitting, dull or sharp
- What relieves the pain
- Whether the pain is severe enough to result in apareunia
- If the pain is experienced also in a non-sexual context, particularly on tampon insertion or with pressure on the vulva, for example when riding a bicycle

History Taking

The Dermatological History

The following factors in a patient's dermatological history may be relevant to the vulva:

- Atopic disease: eczema, hay fever, asthma
- Psoriasis
- Autoimmune conditions: systemic lupus erythematosus, Sjogren's syndrome, autoimmune thyroiditis, pernicious anaemia

- Allergic reactions to drugs or topical therapy
- Lichen planus

The Gynaecological History

Menstrual disturbance often results in more frequent use of menstrual protection, leading to more contact irritation.

Oestrogen status is important. It is low in post-menopausal and lactating women, and of course in pre-pubertal girls. In general, vaginal candidiasis does not occur in a low-oestrogen environment, and so a post-menopausal woman who does not use systemic or vaginal oestrogen should be assumed not to have candidiasis, unless proven otherwise.

Gynaecological surgery including laser surgery, even of a very minor nature, may cause or worsen vulval disorders.

Patients often assume that their symptoms are due to sexually transmissible infections. It is important to assess this possibility, but the investigation is frequently negative.

Herpes simplex virus infection of the vulva rarely by itself causes chronic vulval symptomatology, but it can be the precipitating factor for chronic vulval dermatitis, entry dyspareunia, neuropathic pain or, very occasionally, anxiety or obsessive–compulsive behaviour.

The Urological History

Urinary incontinence has a strong association with vulval disorders, partly due to simple maceration but also because neuromuscular dysfunction that affects the bladder may also affect the vulva.

Vulvovaginal disorders often result in bladder dysfunction disorders, either infective or non-infective. Many patients, however, will present with the secondary bladder symptoms only, and it will become apparent only after careful history taking that the real culprit is in the vagina or vulva.

Urological surgery, even diagnostic cystoscopy, may cause vulval symptoms.

Very occasionally, symptoms originating from the bladder may be experienced in the vulva and vagina without obvious bladder symptomatology.

The Gastroenterological History

Bowel disturbances, especially diarrhoea, may cause or worsen vulvovaginal disorders.

Haemorrhoids make anal cleansing after defecation more difficult.

Faecal incontinence must always be asked about in women who have had vaginal deliveries. It is surprising how frequently this occurs.

The presence of diseases that produce problems with absorption, most commonly coeliac disease, may result in reduced effectiveness of medications.

Crohn's disease may rarely directly affect the vulva.

The Musculoskeletal History

It is essential to enquire about the following:

- Back injuries (motor vehicle accidents, falls onto the coccyx, heavy lifting, falls causing back injury)
- Sciatica
- Hip joint pain, arthritis and injury

- Lumbosacral osteoarthritis with/without disc protrusion
- Spinal surgery
- Exercise routines
- Weight gain

The Environmental History: Secret Women's Business

It is very likely that your patient has her personal hygiene beliefs, practices and rituals. These are often cherished and difficult to change. You need to find out and you will not unless you ask. Ask specifically about possible irritants and allergens including:

- Washing routines: frequency, use of soap, bubble baths and perfumed oils
- Sanitary pads, incontinence pads, liners and tampons
- Lubricants
- Condoms
- Shaving and waxing
- Douching
- Underwear, G-strings
- Over-the-counter and home remedies
- Exercise routines, including clothing worn
- Sports, particularly cycling and horse riding
- Swimming, saunas and spa baths

The Psychological History

Although this need not be exhaustive, it is important to determine whether:

- The patient is still able to enjoy intercourse
- Her partner is sympathetic
- Her problem has ended any previous sexual relationships
- She is suffering from depression, shame or anxiety independent of, or related to, her problem
- She has had any traumatic sexual experiences, either recently or as a child
- If the patient is a child, sexual abuse has been considered and how has this affected the family
- She has beliefs about her condition that are related to misleading information, often obtained from the internet
- She is angry with the medical profession regarding previous treatment failures

Patient Beliefs

It is important to find out what patients believe is responsible for their symptoms and also their attitude to your possible treatments. Examples of beliefs that may impact on your therapeutic strategies include:

- The assumption that vulval symptoms are due to thrush (although this involves 20% at most)
- Fear that treatment with oestrogen will predispose to breast cancer
- Fear that the use of tampons will result in toxic shock
- A belief that symptoms are due to genital herpes, even when there is no objective evidence

- Fear of the use of any form of corticosteroid (it will 'thin the skin')
- Fear that their skin condition is transmissible
- Fear of cancer
- A belief that their condition is the result of a sexual encounter
- A belief that their condition was transmitted from contact with a fomite (such as a toilet seat)

Summary of History Taking

- Symptoms
- Cycling of symptoms
- Duration of problem
- Previous treatment and whether it has helped, even briefly
- Personal habits
- Dermatological personal and family history
- Atopic disease
- Dyspareunia
- Effect on sexual relationships
- Gynaecological history
- Gastrointestinal history
- Urological history
- General medical history
- Psychological history
- Medications including over-the-counter ones
- Allergies
- Secret women's business

Examination

The vulva and vagina display a high level of anatomical and colour variation. Some of this is congenital, some is age related and much is due to vaginal childbirth. Female genital mutilation will present from time to time, and there is also the increasingly common phenomenon of cosmetic reduction labioplasty. The clinician's ability to define what is 'normal' on examination will therefore be determined by their clinical experience in women's medicine.

Although most vulval problems are fundamentally dermatological, the typical appearance of most skin diseases is very different when they occur on genital skin, and may be quite subtle. A good light and adequate access to the genital skin by careful patient positioning is essential. A couch with foot rests is ideal; however, many patients are humiliated and stressed by having their feet placed in stirrups, and the latter is not necessary for an adequate vulval examination.

Inspect the groin and pubic area first, then the external labia majora, the inter-labial sulcae, and then the vaginal introitus. The clitoral hood should be gently retracted to inspect the glans clitoris. Include the perianal area and natal cleft with your patient lying on her side. It may then be helpful to perform a general skin examination to look for clues that will help with diagnosis, for example with possible psoriasis or lichen planus. Include the buccal mucosa, as this may give a clue to lichen planus.

It is very helpful to use a hand mirror to allow the patient to demonstrate the area of her concern, and so that the clinician can show the patient the areas that actually require

application of any topical treatments. Many women have never inspected their own vulva, and it is important to familiarise them with their own anatomy and what the various parts are called.

Common Variations of the Vulva

Common variations of the vulva include:

- Pigmentation: non-Caucasians commonly demonstrate hyperpigmentation of the labia minora; however, patients of all races may develop vulval 'freckling'
- Size of the labia minora: pre-pubertal girls have very small labia minora, but virtually all normal adult pre-menopausal women possess labia minora; after the menopause, these may reduce significantly in size
- Asymmetry of the labia minora
- Size of vaginal opening
- Degree of rugosity of the mucosal surface of the labia minora and vaginal mucosa
- Vulval papillomatosis (tiny projections from the inner surface of the labia minora, a normal variant)
- Prominence of sebaceous glands ('Fordyce spots')
- Erythema: in some patients the sulcus between the labia minora and majora is persistently red in the absence of any pathology
- Length and density of pubic hair
- Clitoral size
- Prominence of gland openings (these may form obvious pits on the mucosal surface)
- Amount of normal discharge
- Apparent webbing at the base of the fourchette

Finding Abnormalities During the Physical Examination

Vulval rashes can be subtle, and initial examination may suggest a normal vulva. Look carefully for:

- Increased erythema of the labia minora, or the sulcus between the labia minora and majora
- Fissuring (skin splits): you may have to gently stretch the skin to find these
- Textural change: lichenification of labia majora and perianal skin, 'cigarette paper wrinkling'
- Evidence of erosions or ulcers
- Mucosal petechiae
- Colour change: light or dark
- Atrophy of mucosa
- Presence of any unusual lesions

Do not be surprised if there are no abnormalities. Many patients with significant vulval symptoms are normal on examination. This finding means that either they have a skin disease that is episodic or that their symptoms are not caused by a dermatological problem.