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

- Most vulval conditions are chronic dermatological diseases that cannot be cured. They must be managed. Such conditions on the vulva often look and behave differently from the same conditions on other parts of the skin. Management strategies that are effective for non-genital skin must often be modified in order to be effective on the vulva.
- The vulva and vagina are in the centre of the lower pelvis and are closely related to other pelvic organs, bound to them by the myo-fascial structure of the pelvic floor (see Figures 1.1, 1.2 and 1.3). Referred vulval pain from other pelvic viscera and from the lumbosacral spine and hip joints is an important concept in understanding vulvo-vaginal disorders.
- 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 of its sexual or relationship consequences. It is therefore no wonder that in the past, women with vulval disorders have been unfairly told ‘it’s all in your head’.
- 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: patients often tell us that they were not examined. History taking is difficult because of the intimate nature of a woman’s symptoms. A detailed sexual and environmental history is essential, and 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 important to help them to organise their thoughts. Start at the beginning and get them to

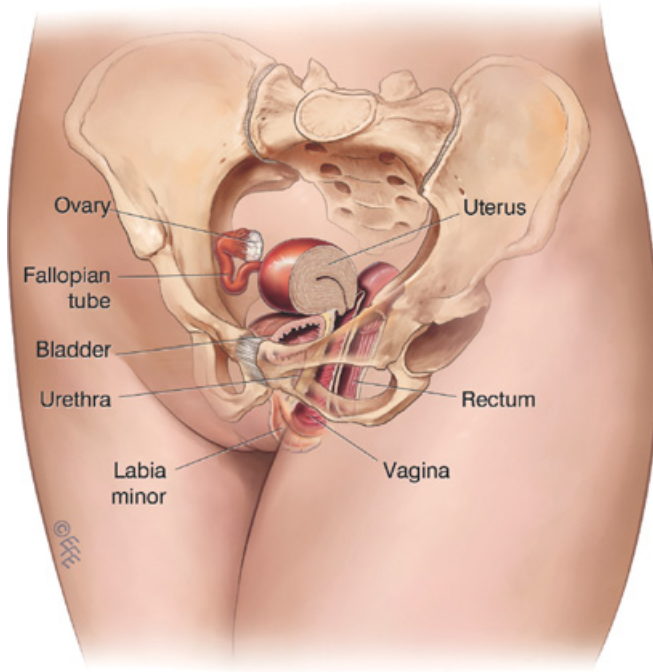


Figure 1.1 The female reproductive tract. With permission from Dr Levent Efe, CMI

think back on how their complaint evolved and how they came to be in your consulting room.

- 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.

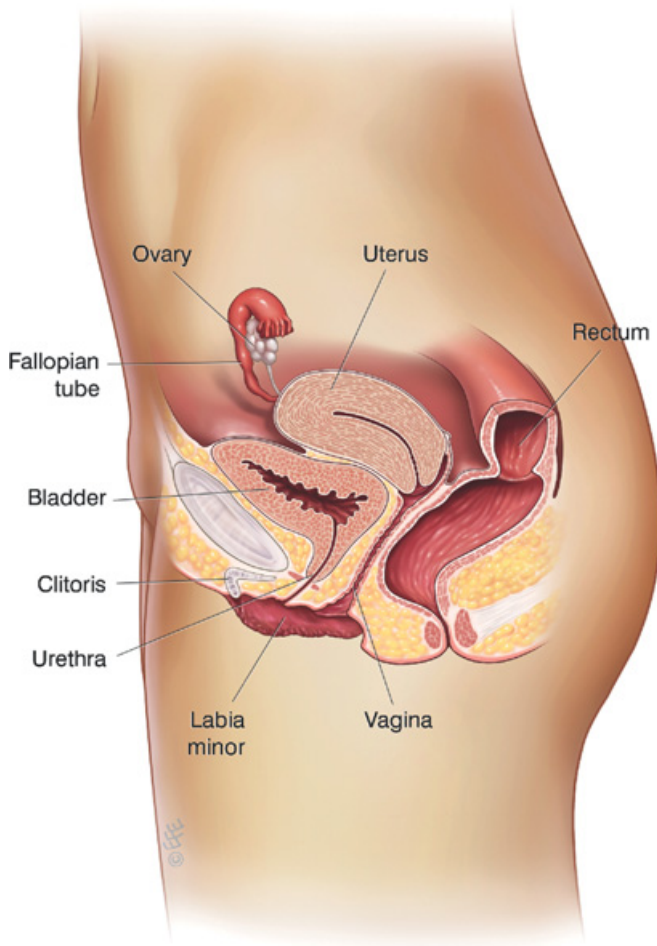


Figure 1.2 The female reproductive tract: side view. With permission from Dr Levent Efe, CMI

The purpose of this handbook is to introduce these concepts and to provide practical management recommendations. These are based on our 60 years' collective experience of helping patients with vulval disease, as well as published research by ourselves and others. We hope that 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 also the entrance to the genital tract. It is essential to understand that vulval skin extends all the way to 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.

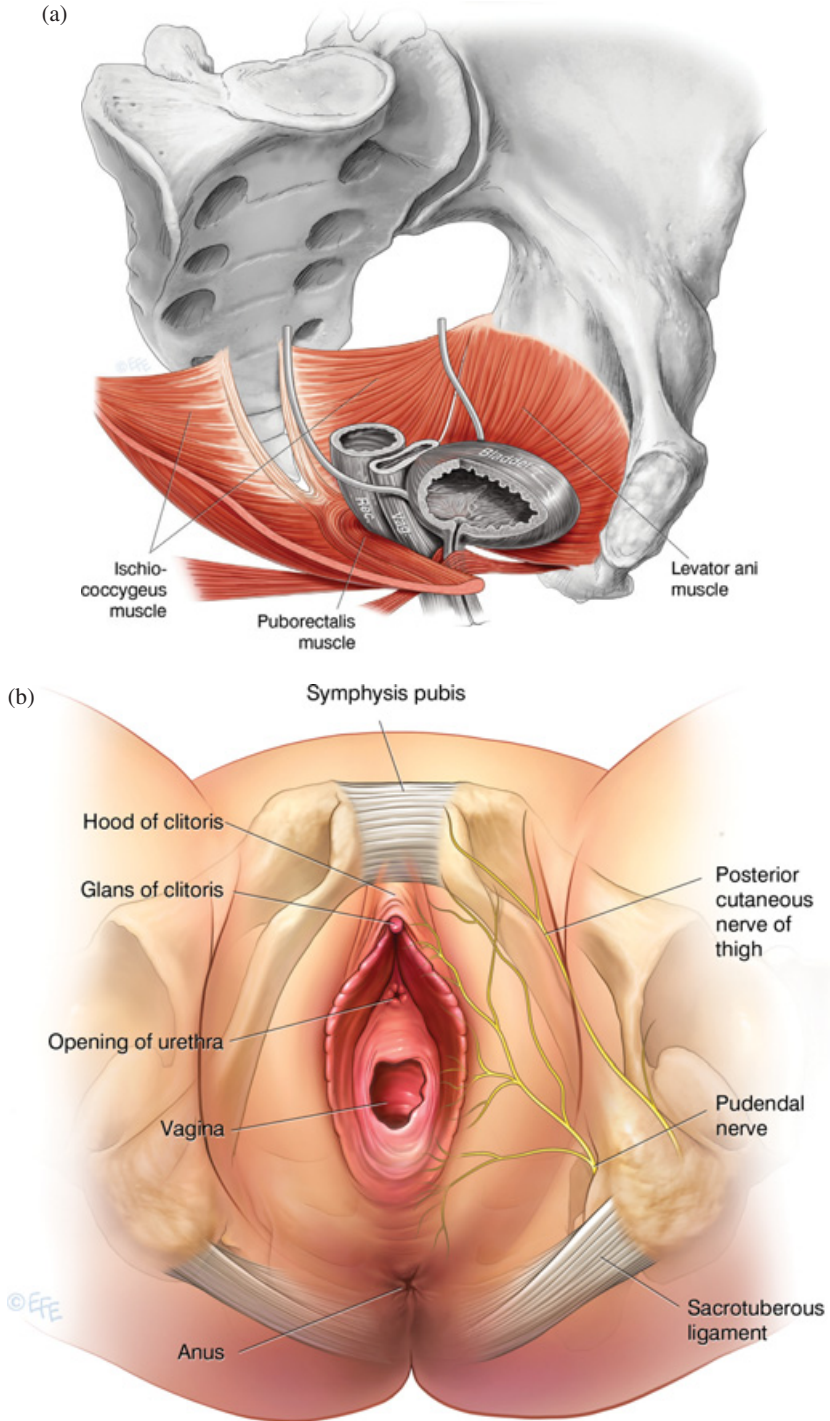


Figure 1.3 (a) The pelvic floor. (b) View of the vulval area. With permission from Dr Levent Efe, CMI

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

- menstrual fluid
- urine
- faeces
- sweat
- vaginal discharge, both normal and abnormal
- semen
- tight clothes
- lubricants
- perfumed products including toilet paper, wet wipes and feminine hygiene products
- medications, both vaginal and oral
- 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 relations include:

- bladder and urethra
- cervix
- rectum and anus
- utero-vesical and recto-vaginal peritoneal pouches
- sacrum and coccyx.

The Pelvic Floor

The pelvic floor is a complex myo-fascial structure that encompasses the entire pelvis. It 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 affected by lumbo-sacral or hip dysfunction.

One way of understanding functional pelvic floor anatomy is to think of it as having 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. When considering the lower pelvic floor, it is helpful to think of the attached structures arranged posterior to anterior: lumbo-sacral spine and hip joints, rectum and anus, vulva and vagina, and urethra and bladder. This is because referral of pain and dysfunction in the lower pelvis tends to be from posterior to anterior (see Chapter 8 on pain).

The Innervation

The innervation of the lower vagina, vulva and anus is from sacral nerve roots S2, 3 and 4 via the pudendal nerve. The anterior vulva is supplied by the genital branch of the genito-femoral nerve (L1 and L2) and the ilio-inguinal nerve (L1). Thus lumbo-sacral, coccygeal and even lower thoracic spinal disorders may produce referred vulval pain (see Chapter 8 for a discussion on pain and dysfunction referral in the pelvis).

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 vulvo-vaginal candidiasis. Many have a personal or family history of the vulval skin condition or have evidence of it elsewhere on their skin, so a general skin examination is very helpful. 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 needs to be recorded, as vulvo-vaginal 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. **Soreness and pain** are often due to erosions or fissures, either caused by a skin condition or secondary to excoriations produced by scratching. They can less often be due to neuro-muscular disorders. Burning, stinging, stabbing, formication and vulval awareness are usually due to a neuro-muscular dysfunction.

Dyspareunia

Dyspareunia means pain during sexual intercourse. We find it helpful to categorise dyspareunia into abdominal and vulvo-vaginal types.

Abdominal dyspareunia is sexual pain experienced in the lower abdomen, as it relates to the upper pelvic floor. It usually relates to disease or dysfunction at or above the level of the cervix, for example endometriosis.

Vulvo-vaginal dyspareunia is experienced at the vaginal entrance, or further up into the vagina proper. It is caused by disease or dysfunction at the level of the lower pelvic floor. This pain is usually caused by a vulval and/or vaginal skin condition. It can also be caused by disease or dysfunction of the bowel, anus, the lumbo-sacral spine and hip joints.

Important points to elicit in a history of dyspareunia are:

- Is the onset of pain:
 - with foreplay (or masturbation)?
 - during vaginal intercourse?
 - after intercourse is concluded?
 - gradual or sudden?
- Is the duration of pain
 - at intromission, and then improves?
 - at intromission and then relieves rapidly after withdrawal?
 - throughout intercourse then continues for a variable time after intercourse has ceased?
- Where is the site of entry (or vaginal) dyspareunia (with a mirror if necessary)?
- What is the nature of the pain: tearing, splitting, dull or sharp?
- What relieves the pain?
- Is the pain severe enough to result in apareunia?
- Is the same pain also experienced in a non-sexual context, particularly tampon insertion or with pressure on the vulva (especially with tight clothes)?

History Taking

The Dermatological History

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

- atopic disease (e.g., eczema, hay fever or asthma)
- psoriasis
- autoimmune conditions (e.g., systemic lupus erythematosus, Sjogren's syndrome, autoimmune thyroiditis or pernicious anaemia)
- allergic reactions to drugs or topical therapy
- lichen planus, particularly oral.

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 (STIs). It is important to assess this possibility, but investigation is frequently negative.

Herpes simplex infection of the vulva is an acute event that is rarely by itself responsible for chronic vulval symptomatology. However, 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. This is partly due to simple vulval maceration caused by contact with urine and pads, but also because vulval skin disorders often cause or worsen bladder dysfunction via the pelvic floor.

Vulvo-vaginal 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. This is especially true of urethral disorders.

The Gastroenterological History

Bowel disturbances may cause or worsen vulvo-vaginal disorders. Diarrhoea often results in peri-anal and vulval irritant contact dermatitis. Constipation tends to 'wind up' the posterior pelvic floor, and may lead to vaginal dyspareunia of neuro-muscular origin.

Haemorrhoids make anal cleansing after defecation more difficult, and often causes dermatitis due to excessive cleaning.

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 and may do so in the absence of active gastro-intestinal disease.

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
- lumbo-sacral 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 own personal hygiene beliefs, practices and rituals. These are often cherished and difficult to change. You need to find these out – and you won’t 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, consider possible sexual abuse and how this could affect the family
- she has beliefs about her condition that are related to misleading information, often 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 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 (e.g., a toilet seat).

Summary of History Making

- 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
- gastro-intestinal history
- urological history
- general medical history
- psychological history
- medications, including over the counter
- 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 age-related and much due to vaginal childbirth. Female genital mutilation will present from time to time, and there is also the increasingly common phenomenon of cosmetic reduction labiaplasty. 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 this is not essential for an adequate vulval examination.

Inspect the groins 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 clitoridis. Include the peri-anal area and natal cleft with the patient lying on her side. It may then be helpful to perform a general skin and oral examination to look for clues that will help with diagnosis, for example, of possible psoriasis or lichen planus. Include the buccal mucosa as this may give a clue to lichen planus, which is not always symptomatic.

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 the names of the components.

Common Physiological Variations

Common physiological variations of the vulva include:

- pigmentation: Non-Caucasians commonly demonstrate hyper-pigmentation of the labia minora. However, patients of all races may develop vulval hyper-pigmentation, which becomes more apparent with advancing age
 - size of labia minora: pre-pubertal girls have very small labia minora, however, virtually all normal adult pre-menopausal women possess labia minora. After menopause, these may reduce significantly in size but absence of labia minora, particularly if it is asymmetrical, should be cause for concern regarding possible scarring vulval conditions such as lichen sclerosus and lichen planus
 - asymmetry of the labia minora
 - size of the vaginal opening
 - degree of rugosity of the mucosal surface of 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 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 labia minora, and the sulcus between minora and majora
- wet desquamation (scale), especially in the inter-labial sulcus
- fissuring (skin splits): you may have to gently stretch the skin to find these
- textural change: lichenification of labia majora and peri-anal 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.

The Speculum Examination

Many vulval skin diseases involve the vulva only, and there is no need for a speculum examination. The diseases that do extend into the vagina are often so uncomfortable that speculum examination is very difficult and may need to be postponed. While there is a place for examination under anaesthesia, initial treatment of the vulval problem may make speculum examination in the consulting room possible at a subsequent visit.

If when you examine the vulva the introitus is involved and there appears to be extension into the vagina, attempt a speculum exam using a small, straight-bladed instrument. Remember, you are not taking a Pap test, and therefore a complete view of the cervix is not essential. Your aim is to visualise enough of the vaginal wall to determine the degree of intravaginal inflammation.

Look for:

- erythema, and whether it is confluent or patchy; refer to the oral mucosa as a guide to what is normal
- erosions
- petechiae
- degree, colour and type of discharge.

In the rare case of intra-vaginal adhesions, a one-finger digital examination will usually give more information than a speculum examination.

Investigations

The most commonly performed investigations in patients with vulval disease are:

- low vaginal swab for *Candida* (or rarely bacterial) infection
- viral swab for polymerase chain reaction testing for herpes simplex if suspected. This must be taken from an ulcer or erosion and the lesion should be firmly swabbed. This will be uncomfortable. Warn the patient of this prior to performing the test.
- vulval swab for *Candida* or bacterial infection
- scraping of vulval skin for dermatophyte (tinea) infection: this is done with a scalpel blade into a specimen jar (check with your pathologist)
- skin biopsy.

All patients should have a low vaginal swab taken for culture. A high vaginal swab taken during a speculum examination is unnecessary, and may not reflect the environment of the lower vagina. Furthermore, passing a speculum past inflamed vulval skin is often exquisitely painful.

A vulval swab or scraping is performed if infection of the hair-bearing skin is suspected clinically.

A skin biopsy is performed if there is a visible rash or lesion that is diagnosable by biopsy, particularly for suspected malignancy. It should never be performed on clinically normal skin. Specific situations that always require biopsy are:

- a white lesion
- a unilateral lesion

- suspicion of malignancy: persistent ulcer, nodule, fissure, pigmented lesion, white hyperkeratotic lesion
- a lesion that has not adequately responded to initial treatment.

Other tests that may be relevant include:

- mid-stream urine for microscopy and culture if urological symptoms are present
- patch testing if a contact allergy is suspected
- serum follicle-stimulating hormone (FSH) may in some cases be required to assist in confirming menopause
- tests for systemic autoimmune disease may be relevant in patients with conditions such as lichen sclerosis.

Colposcopy is usually unnecessary in the diagnosis of vulvo-vaginal disorders, except when vulval intraepithelial neoplasia is suspected. The use of acetic acid on the vulva is unnecessary in benign skin diseases and usually produces significant stinging that patients find traumatic.

Patient Categorisation

At the end of your history and examination, you will find that patients with vulval disease fall into the following broad categories:

- patients with a rash and non-cycling symptoms
- patients with a rash and cyclical symptoms
- patients with a rash or lesion and no symptoms
- patients with no rash but symptoms.

This will be discussed more later in the book.

Summing Up: Common Things Occur Commonly

The majority of patients presenting with chronic vulval symptoms have a vulval presentation of a common skin disease such as eczema, psoriasis or lichen Sclerosus or are suffering from chronic vulvo-vaginal candidiasis.

Many will have a personal or family history of the same condition elsewhere on the skin. Many patients with eczema are atopic. This historical information can provide very helpful clues to a possible diagnosis.