

Vulval diseases

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GP CME

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Outline of this presentation

- Inflammatory skin diseases
- Neoplastic skin diseases
- Diagnosis and management

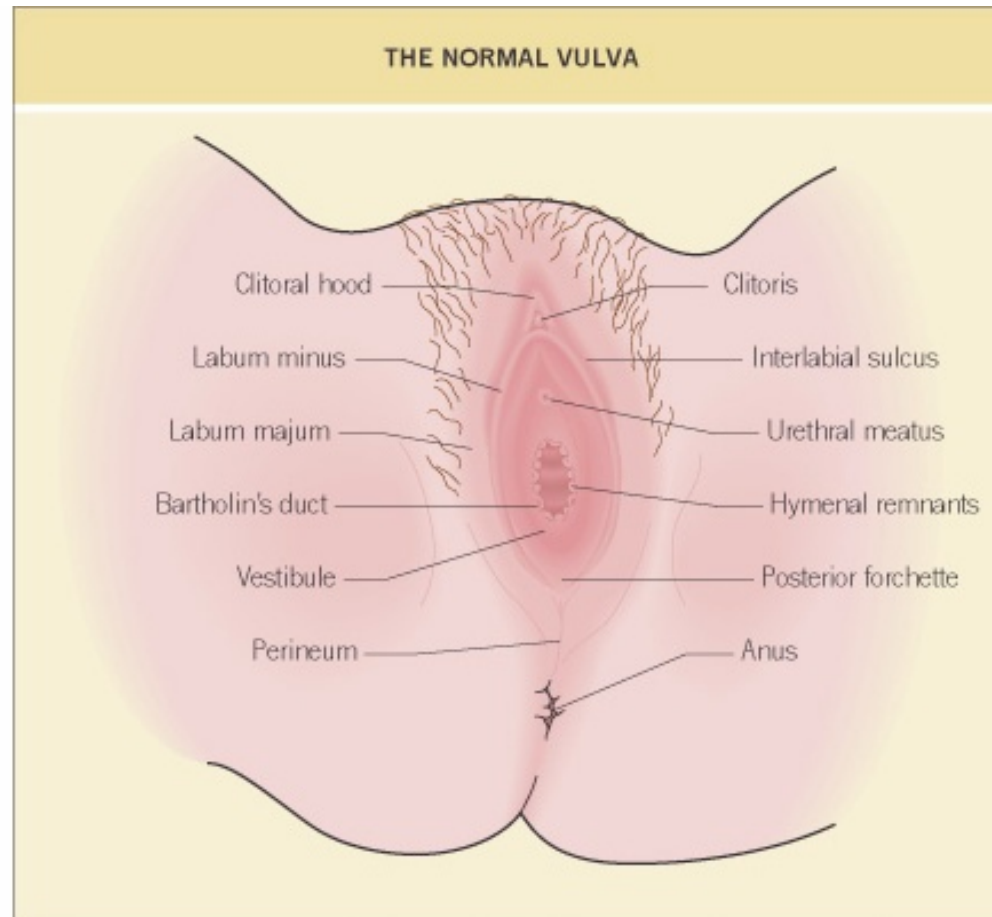
Inflammatory skin diseases: symptoms

- Pruritus
- Soreness
- Dyspareunia
- Asymptomatic signs

Inflammatory skin diseases: signs

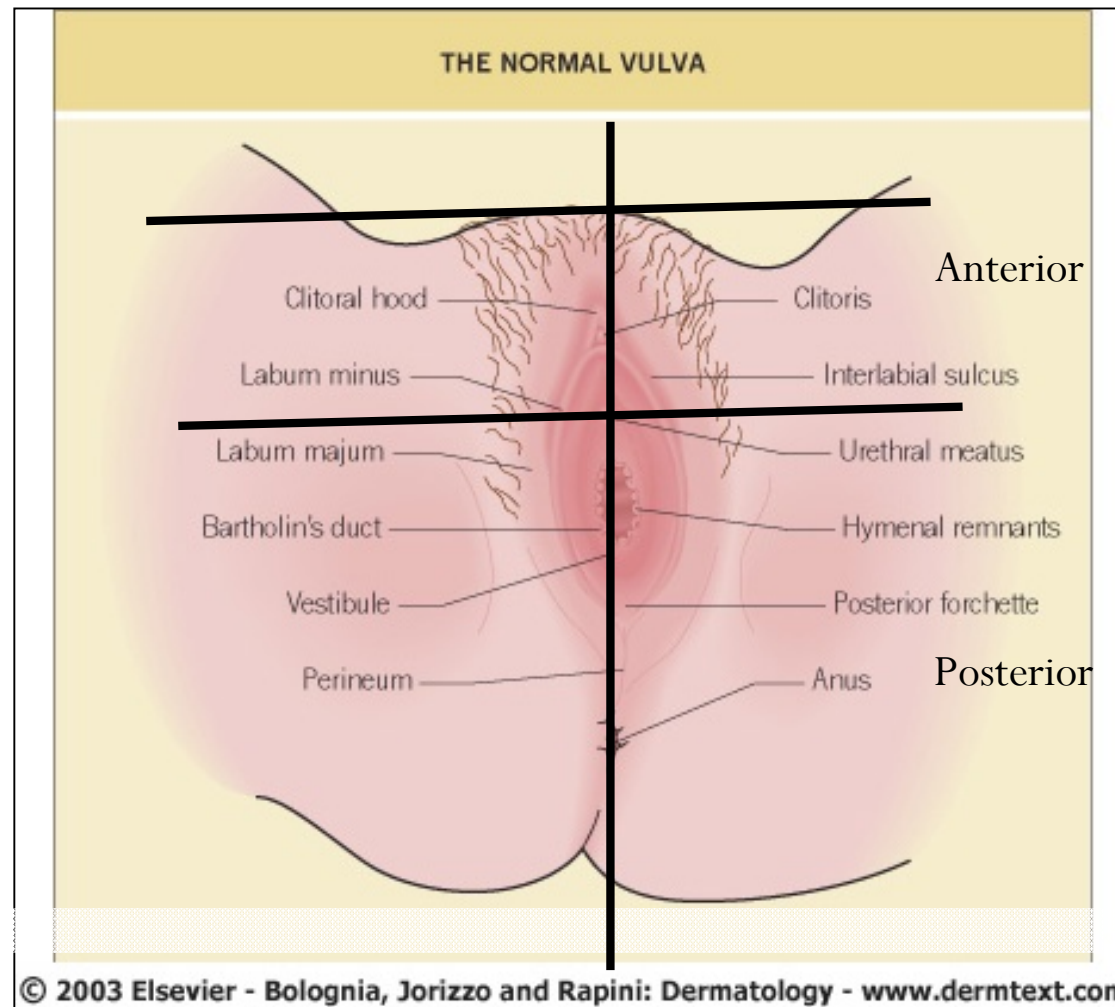
- None
- What site is affected?
- Any anatomic abnormality?
- Colour: red, white
- Primary lesions: macules, papules, plaques etc
- Secondary lesions: fissures, excoriations, haemorrhage, crusting etc.

Learn anatomic terminology



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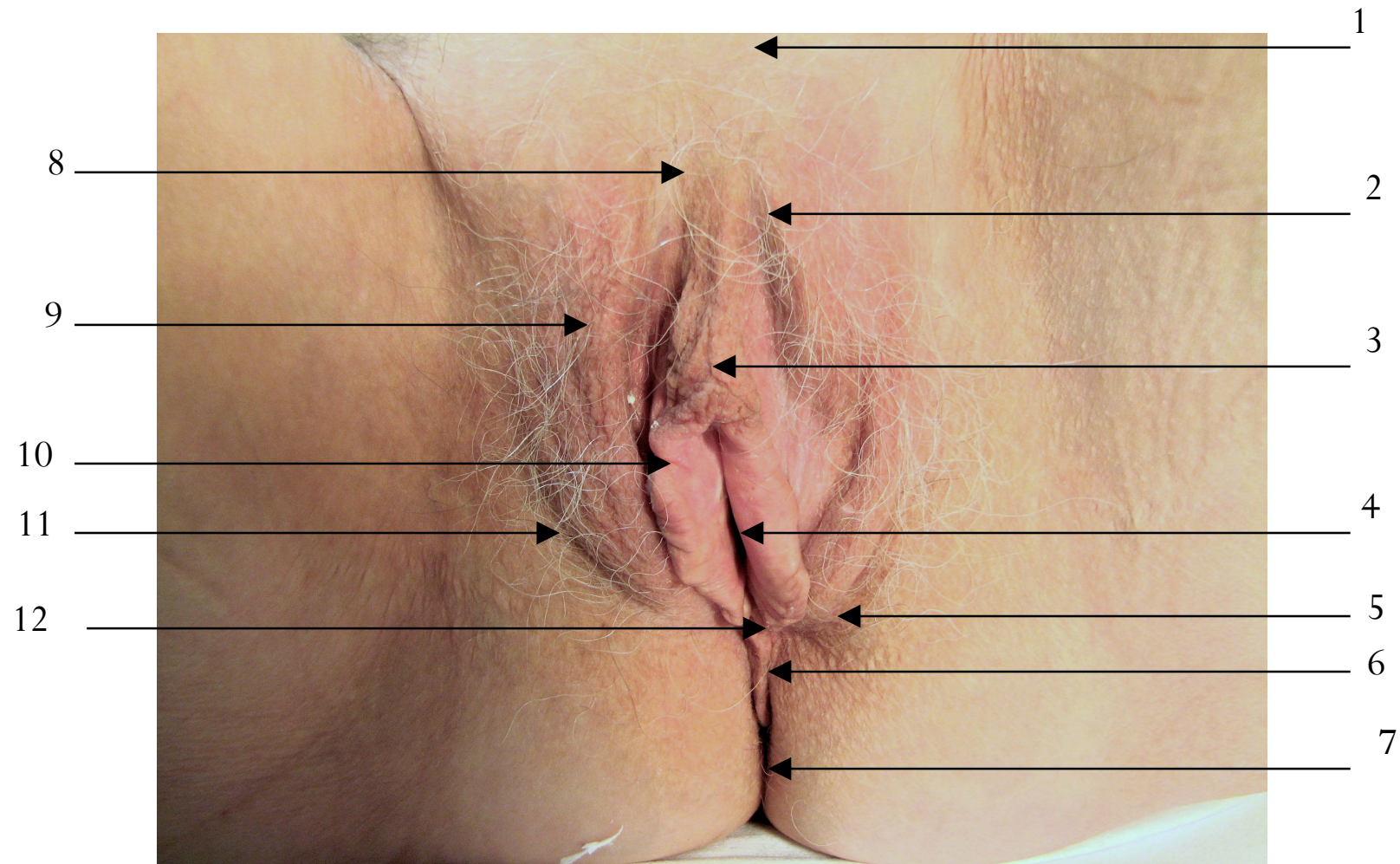
Examination of the vulva



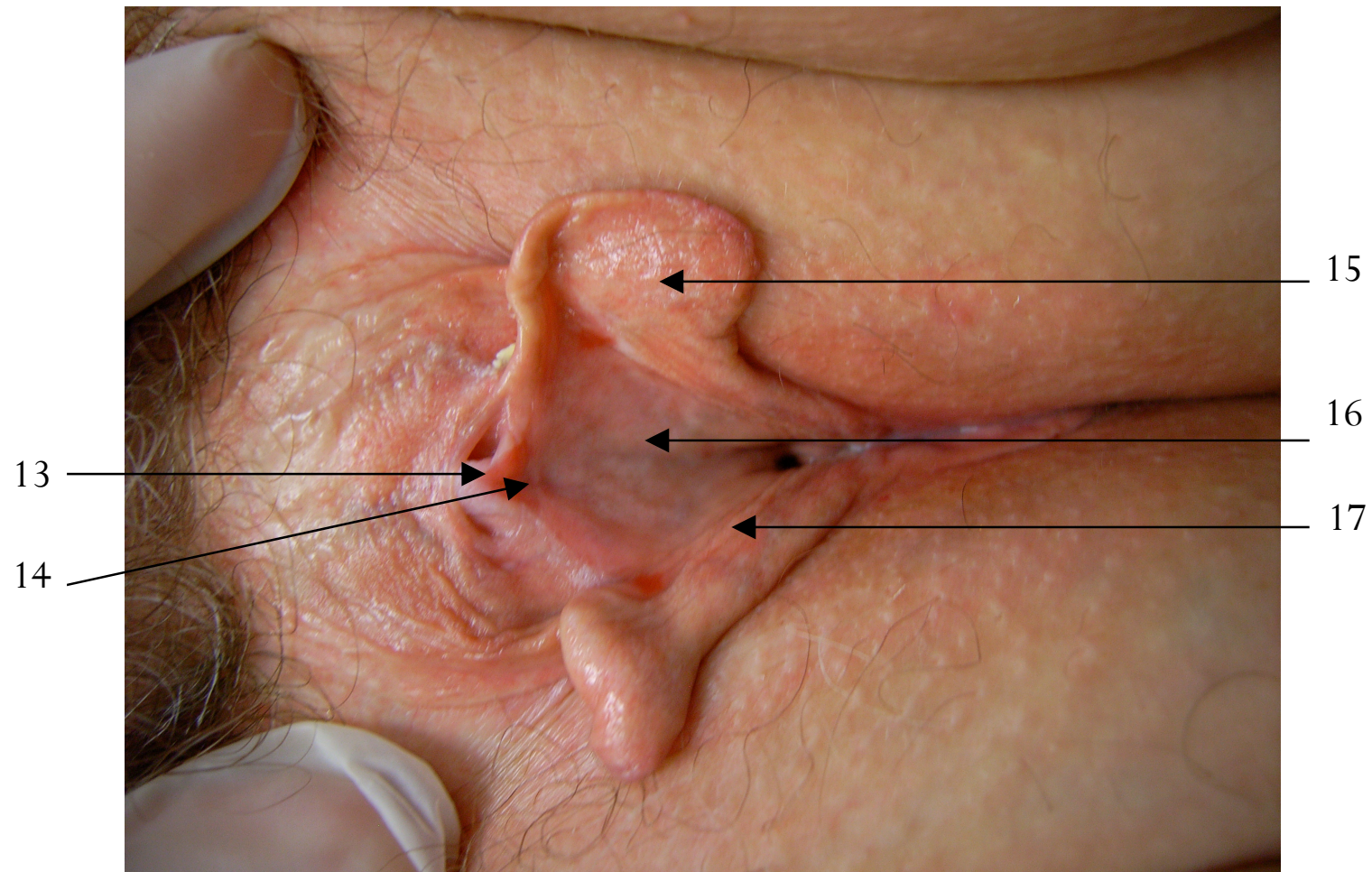
Anatomic structures

1. Mons pubis
2. Interlabial sulcus
3. Clitoral hood / prepuce
4. Vaginal introitus
5. Posterior commissure
6. Perineum
7. (Anus)
8. Anterior commissures
9. Labia majora
10. Labia minora (nymphae)
11. Genitofemoral / genitocrural fold
12. Posterior fourchette
13. Glans
14. Frenulum
15. Fordyce spots
16. Vestibule
17. Hart's line
18. Urethral meatus
19. Fossanavicularis
20. Vestibular papillae / papillomatosis

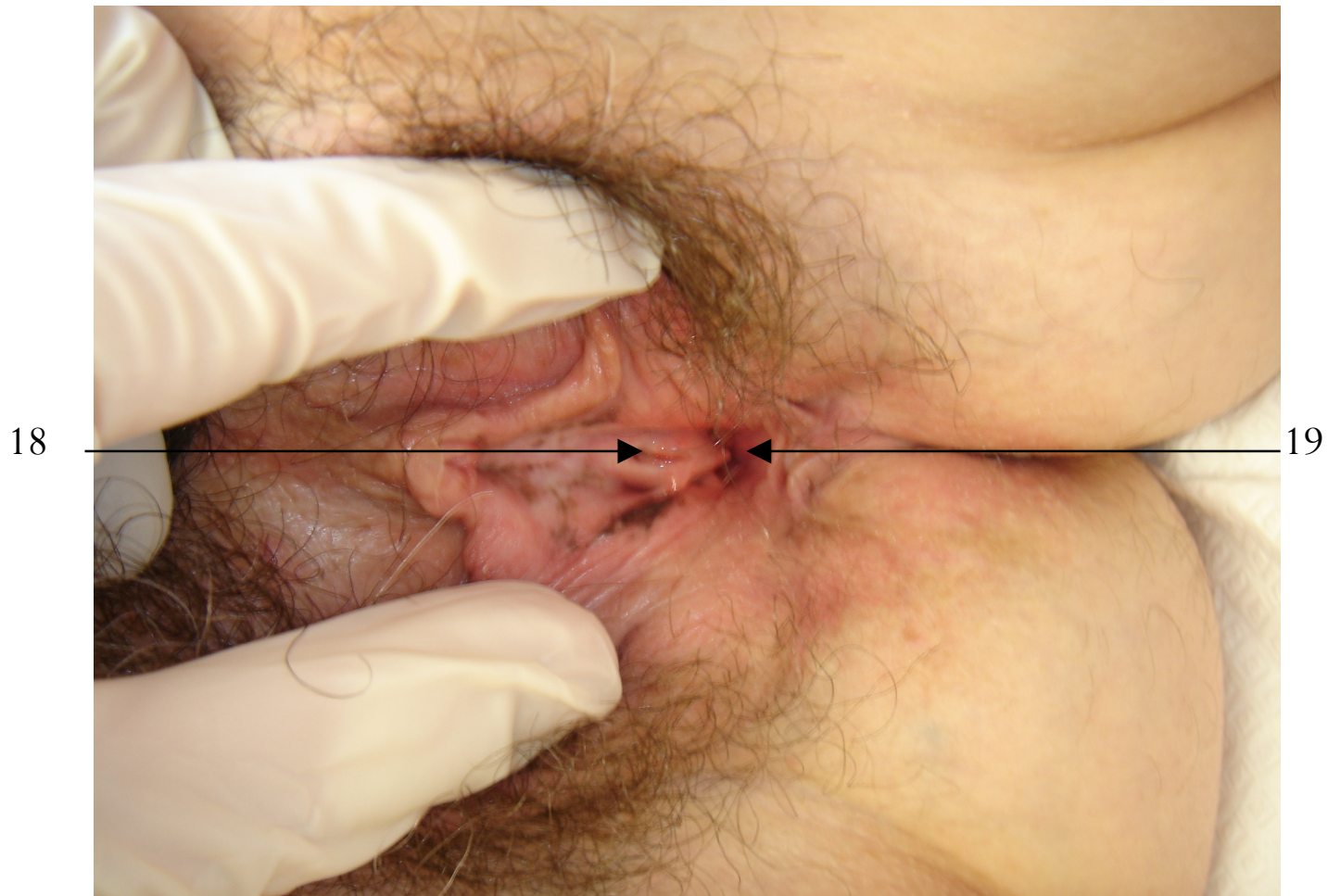
Vulval anatomy



Vulval anatomy



Vulval anatomy



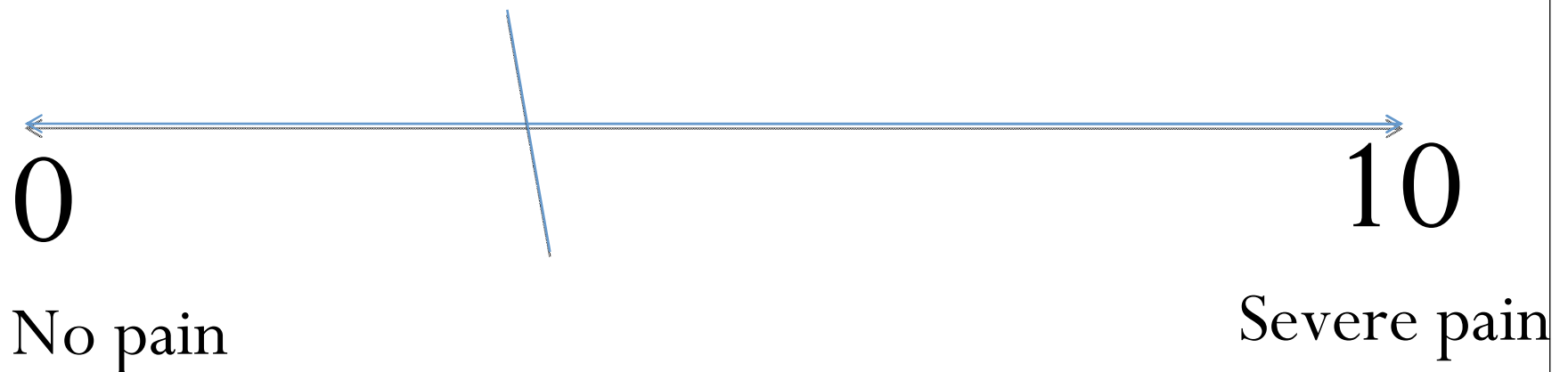
Pruritus

- Maybe unremitting and extremely severe
- Scratching may cause lichenification, whatever the cause
- *Candida* / eczema / lichen sclerosus
- Assess using linear analogue scale



Soreness

- Soreness associated with inflammatory skin disease may be mild tenderness to very severe pain
- Pain may only be associated with intercourse (dyspareunia), and can prevent it (apareunia)
- Lichen sclerosis / erosive lichen planus



Vulvodynia

- Burning discomfort, in the absence of an inflammatory skin disease

Pruritus: *Candida albicans* infection

- Symptoms tend to be centred on vagina
- Often more severe premenstrually
- Requires oestrogen: vagina full of glycogen, so arises in young women, or a systemic condition (e.g. diabetes)
- Yeasts found in vagina, occasionally mucosal vulva
- Irritant contact dermatitis due to candida metabolites
- Irritant dermatitis may spread to labia majora
- Scratching may lead to chronic lichen simplex

Candida albicans– oral thrush



Candida albicans– vaginal thrush



Candida albicans – vulvovaginal thrush



Candida albicans – irritant dermatitis



Candida albicans - lichen simplex



Candida albicans: treatment

- Confirm with high vaginal swab
- Topical therapy vs. oral azole
 - Topical: inexpensive but irritant
 - Oral: convenient but expensive, drug interactions
- Hydrocortisone to treat the dermatitis

Pruritus: eczema

- Cutaneous not mucosal, so affects labia majora
- Affects all ages including prepubertal girls
- Irritant contact dermatitis from soap, overwashing, urine
- Allergic contact dermatitis is much less common
- Lichen simplex is due to scratching and is very itchy
- Mostly improves with mild topical steroid short term
- Severe lichen simplex may require ultrapotent topical steroid for months

Irritant contact dermatitis



- Burn-like
- Glazed erythema
- Superficial erosions
- Oedema

Faecal and urinary incontinence

Irritant contact dermatitis



- Erythema
- Oedema
- Fissuring

Soap, water, rubbing

Irritant contact dermatitis: treatment

- Reduce contact with irritants
 - Napkins, hygiene, nursing care
 - Barrier creams, soap substitutes
 - Avoid unnecessary washing, wiping
- Hydrocortisone cream / ointment
 - Stronger topical steroids are not particularly effective
- Soothing emollients frequently

Lichen simplex



- Perineum - ?*Candida*
- Skin thickening
- Fissuring
- Skin coloured
- Dyspigmentation common

Lichen simplex



- Unilateral, labium major
- Due to rubbing
- Well-demarcated
- Skin thickening
- Increased skin markings
- Skin coloured
- Dyspigmentation common

Lichen simplex



- May be bilateral
- Thickened skin
- Prominent skin markings
- Broken off hairs

Lichen simplex



- Bilateral – ?psoriasis
- Skin thickening
- Increased skin markings
- Skin coloured
- Dyspigmentation common

Lichen simplex: treatment

- Explain the role of scratching / rubbing / irritants
- Consider underlying disease
 - *Candida*, psoriasis, lichen sclerosus
- Frequent cooling emollients
- Potent topical steroid cream until lichenification has gone
 - Several weeks to months
 - Follow with pulse weekend applications
- Hydrocortisone cream ad libitum

Pruritus: lichen sclerosus

- Mainly labia minora, clitoral hood, perineum
- Perianal in 50%
- Extragenital in 15%
- Most are 50 years or older but children may be affected
- May have associated autoimmune conditions
 - 25% thyroid antibodies; 20% hypothyroid
 - 17% psoriasis
 - 5% morphoea, vitiligo, lichen planus

Lichen sclerosis

- Itch: nil to very severe
- Soreness: nil to very severe
- Dyspareunia / apareunia
 - Itch
 - Soreness
 - Fissuring due to sclerosis
 - Introital narrowing due to agglutination / bridging

Lichen sclerosus



- Distribution: figure of 8 with wings
- Colour – white, crinkly ('cigarette-paper')
- Sclerosis - firm
- Anatomic change - destructive

Lichen sclerosus



- Labia minora / perineum
- Spotty white

Lichen sclerosus



- Labia minora / perineum / perianal / labial sulci
- White spots

Lichen sclerosus



- White, like vitiligo
- Anatomic change
 - Loss of labia minora

Lichen sclerosus



- LS + vitiligo
- Anatomic change
 - Loss of labia minora

Lichen sclerosus

- Perianal / perineum
- White spots



Lichen sclerosus



- Distorted clitoral hood / labia minora
- Postinflammatory white areas
- Postinflammatory brown pigmentation

Lichen sclerosus



- Resorption of labia minora
- Fissures in labial sulci, perineum

Lichen sclerosus



- May involve crural creases
- White plaques
- Fissures in sulci

Lichen sclerosus



- Flattening of clitoral hood
- Resorption of labia minora
- Hemorrhages

Lichen sclerosus



- Flattening of clitoral hood
- Resorption of labia minora

Lichen sclerosus



- Distortion of labia minora
- Ulceration
- Haemorrhage

Lichen sclerosus



- Bridging / fusion / agglutination of left and right labia

Lichen sclerosus



- Bridging / fusion / agglutination of left and right labia
- Ulceration

Lichen sclerosus



- Severe introital narrowing
- Splitting of posterior forchette

Lichen sclerosus



- Haemorrhage in posterior forchette

Lichen sclerosus



- Haemorrhage right labia minora

Lichen sclerosus



- Ulcer right labia minora

Lichen sclerosus



- Hyperkeratosis
 - Consider biopsy
 - DD VIN

Lichen sclerosus



- Hyperkeratosis
 - Consider biopsy
 - DD VIN

Lichen sclerosus: extragenital



Lichen sclerosus: treatment

- Ultrapotent topical steroid, usually clobetasol ointment
- Once daily until clear (one to three months)
- Then alternate days for the same duration
- Then once or twice per week longterm
- Adjust frequency if necessary
- Explain to patient precisely where to apply the cream
- Clinical photographs are useful for follow-up

Lichen sclerosis – follow-up

- Review after 3 months
- Then after 6 months
- Then annually if symptomatic or requiring treatment
 - To ensure adequately treated
 - To ensure not over-treated
 - To assess for VIN, LP and invasive SCC

Lichen sclerosis: other treatment

- Treatments that sometimes work
 - Milder topical steroids
 - Calcineurin inhibitors (expensive)
 - Hydroxychloroquine 200mg bdx 6 months
 - Acitretin 25mg daily x 3 to 6 months
 - Prednisone 40mg daily
- Treatments that don't usually work
 - Calcipotriol cream
 - Methotrexate

Pruritus: lichen planus

- Various clinical presentations
 - Cutaneous
 - Mucosal
 - Mixed
- Most are adults
- May also have lichen sclerosis
- Absent to very severe symptoms
- Treatment-resistant

Lichen planus: cutaneous type



- Violaceous plaques
- Wickham's striae
- Labia majora
- Irregular distribution
- Typical LP elsewhere

Lichen planus: cutaneous type



- Violaceous plaques
- Postinflammatory pigmentation may be marked
- Irregular distribution

Lichen planus: mucosal type

- May be very itchy
- More often very painful
- Exquisitely tender
- Destructive – loss of clitoral hood, labia minora
- May cause introital narrowing
- More likely to involve vagina (unlike lichen sclerosis)
- May also affect oral mucosa
 - Buccal mucosa, inner lips, tongue
 - Desquamative gingivitis

Oral lichen planus



Lichen planusL gingivitis

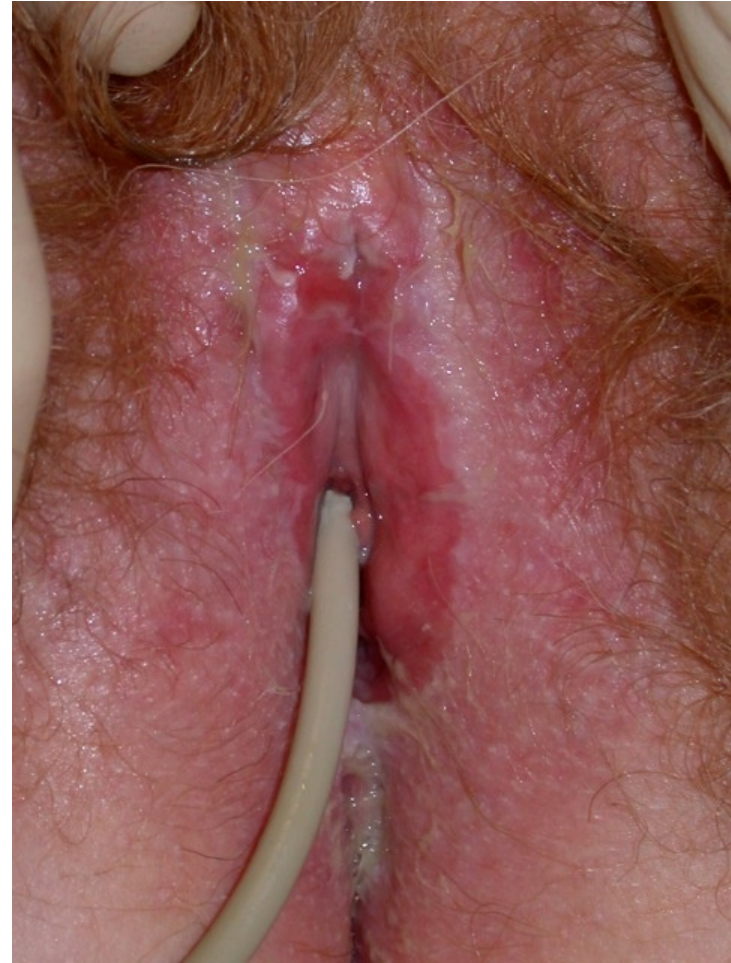


Erosive lichen planus



- Red glistening painful mucosal patch
- Loss of labia minora

Erosive lichen planus



Erosive lichen planus



- Mucosal disease may result in postinflammatory pigmentation

Erosive lichen planus



- Bridging may be due to concomitant lichen sclerosis or to the lichen planus

Erosive lichen planus



- Destruction of tissue
- Introital narrowing
- Eaten-away appearance

Erosive lichen planus



- Mixed picture
- Eaten away right introitus
- Eroded left introitus

Erosive lichen planus: treatment

- Topical clobetasol daily
 - May be required longterm
- Topical calcineurin inhibitors, e.g., pimecrolimus cream
 - Sometimes extremely effective
- Oral prednisone
 - 40mg daily for 2-3 months then reduce over 2-3 months
- Methotrexate
 - 15mg once weekly for 3 months, may be able to discontinue

Erosive lichen planus: other oral tx

- Hydroxychloroquine
- Itraconazole
- Griseofulvin
- Ciprofloxacin
- Metronidazole

Plasma cell vulvitis

- Rare histological diagnosis when you think patient has LP
- Mild to moderate symptoms
- Introital / vaginal haemorrhagic red patches
- Non-destructive

Plasma cell vulvitis



- Deeper red than LP
- More internal
- More patchy

Plasma cell vulvitis



- May extend periurethrally
- Contact bleeding

Plasma cell vulvitis: treatment

- Topical antibiotic
 - Clindamycin cream
 - Metronidazole cream
- Hydrocortisone cream

Psoriasis

- Vulval psoriasis often seen in association with psoriasis affecting other sites
- Well-demarcated erythematous plaques
- External aspects of vulva and flexures
- May or may not be scaly
- Mild or absent symptoms

Vulval psoriasis



Vulval psoriasis



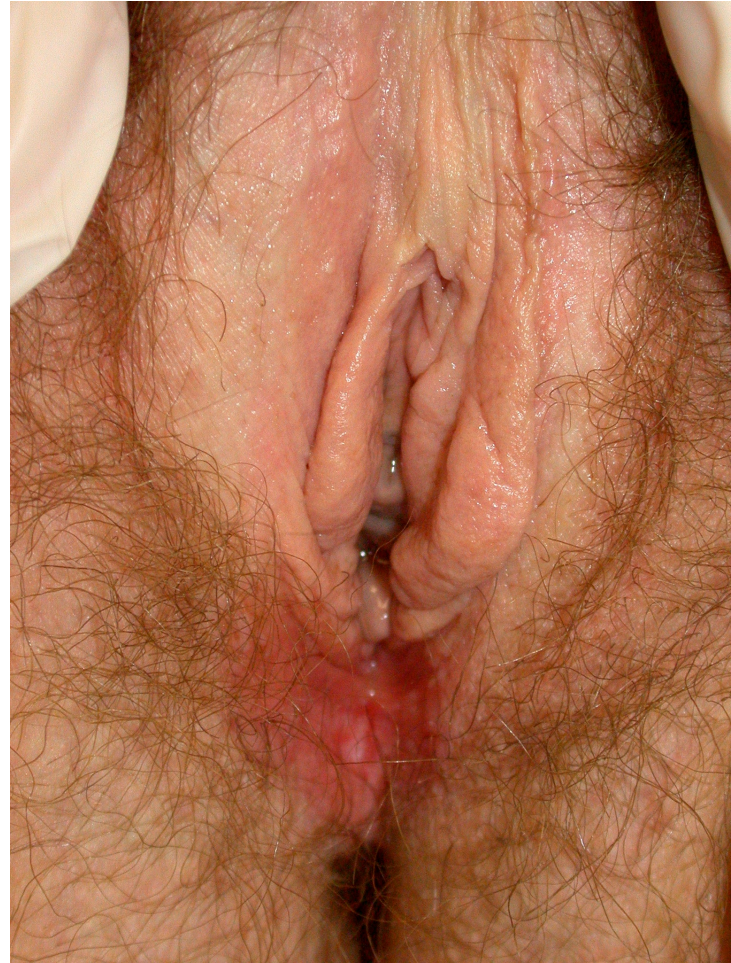
Malignant lesions in the vulva

- Vulval intraepithelial neoplasia
- Invasive squamous cell carcinoma
- Extramammary Paget's disease
- Melanoma

Vulval intraepithelial neoplasia

- Slowly enlarging irregular white, red, pigmented plaques
- Mild or no symptoms
- Sometimes, known history of genital warts / HPV
 - VIN (usual type) - warty, basaloid or mixed
- At other times, lichen sclerosus or lichen planus
 - VIN (differentiated type) – associated with aggressive SCC
- New histological classification

VIN



VIN



+ Lichen sclerosus



Invasive squamous cell carcinoma

- Arises within VIN or within a chronic inflammatory skin disease, especially lichen sclerosus (6%)
- Enlarging irregular, hard nodule, often ulcerated
- May be multifocal
- Often aggressive

Invasive squamous cell carcinoma



Invasive squamous cell carcinoma



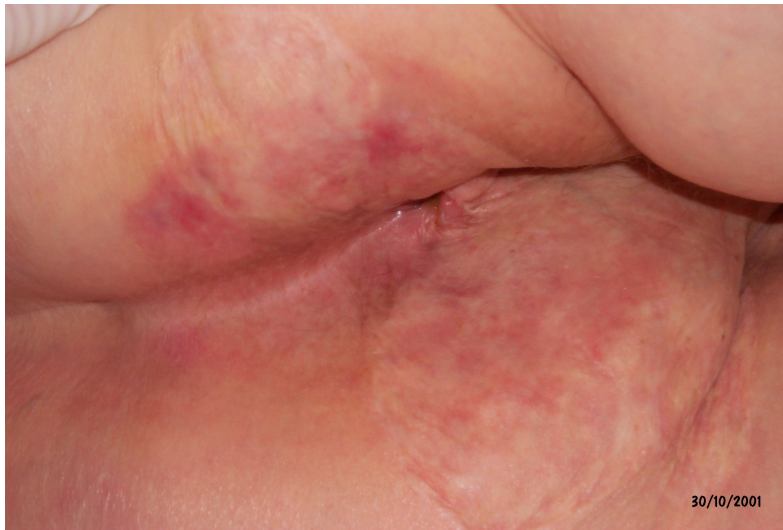
Extramammary Paget's disease

- Slowly growing discrete plaques
- Border may appear defined yet recurrence is usual after wide excision

Paget's disease



Paget's disease: recurrence



Melanoma

- Rare
- Mucosal melanoma similar to acral melanoma
- Not related to skin colour or sun exposure
- Mild or no symptoms

Vulval melanoma



Neoplastic lesions of the vulva

- Surgical management

Adverse effects of topical steroids

- Atrophy
- Striaeatrophicans
- Telangiectasia
- Periorificial dermatitis
 - Burning discomfort
 - Bright red skin
 - Often: spotty margin or erythematous streaks
- Usually prescribed for minor reason, or applied to incorrect site more frequently than necessary
- Rare when used properly (once daily, thin smear, to affected area only)

Steroid atrophy



Periorificial dermatitis – cf perioral



Lichen sclerosus



Psychosocial factors

- Culture, ethnicity, religion
 - Age, education, beliefs and fears
 - Marital status, family support
 - Severity of symptoms
-
- Most women present late, and in distress, fearing the worst

Vulvology

- An important role for the dermatologist
 - Don't leave it to the gynaecologist or sexual health physician
- Make the diagnosis, confirm by biopsy if necessary, and explain management
- Follow-up is important
 - Few women default if their vulvologist is caring, knowledgeable and puts them at ease