Vulval Disease

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Vulval Disease

- Learning Objectives

- How to recognise most common conditions
- How to investigate if required
- What questions to ask
- How to assess
- How to initiate treatment
- When to refer

Vulval Disease

- Important
- History
- Itchy
- Painful
- Soreness
- Intermittent/ Constant
- ‘Rash’
- Red/ White
- Scaly
- Previous treatments
- Associated conditions

Vulval Disease

- Examination – Important to examine
- Evidence of lesions
- Where are the areas of concern?
- Labia Majora
- Labia Minora – size
- Vestibule
- Red/ White/ Scaly
- Single/ Multiple lesions
- Pain versus not
- Creases versus not
- Lymph Nodes

Vulval Disease

- Vulvo-vaginal problems are among 10 leading disorders encountered by primary care clinicians.

- Benign lesions of the vulva are mentioned in **three** categories :
  1. Epithelial conditions.
  2. Benign neoplastic disorders.
  3. Dermatologic disorders.
- VIN
- Cancer vulva

Vulval Disease

- Epithelial Conditions
- Lichen simplex .
- Contact Dermatitis
- Lichen sclerosis.
- Lichen planus,
- Erosive lichen planus.

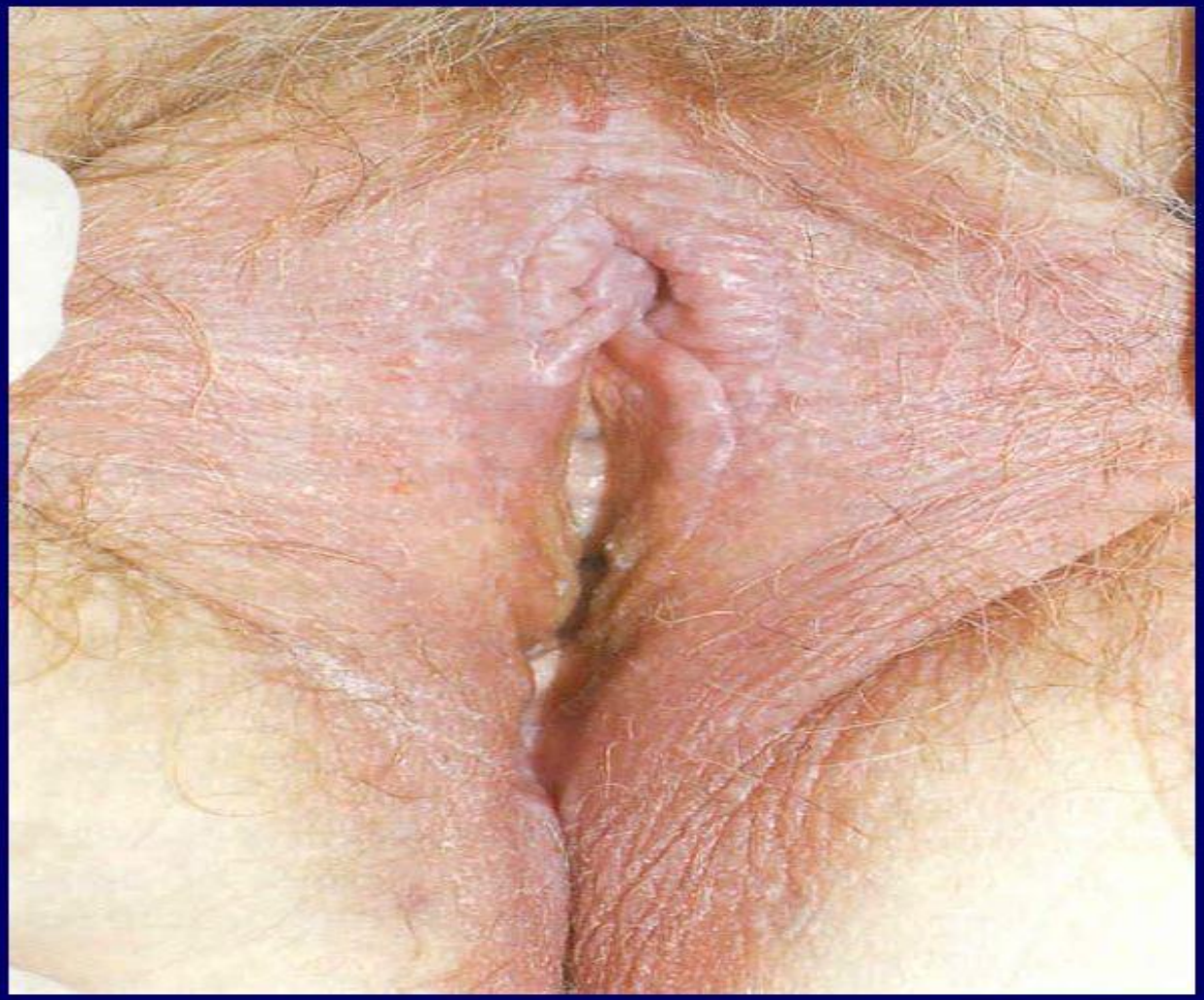
Vulval Disease – Lichen Simplex

Lichen simplex chronicus

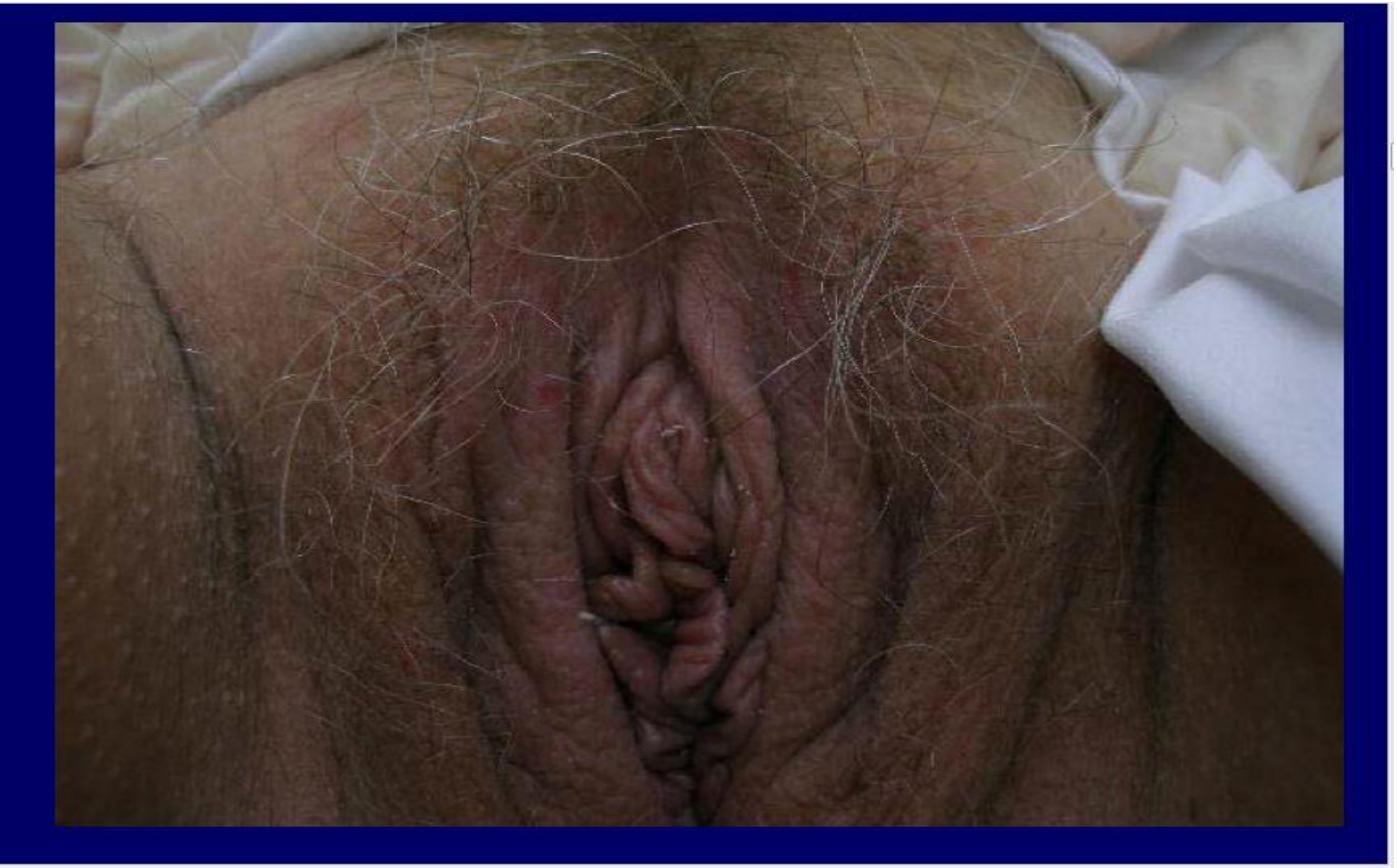
"The scratch that itches"

- Chronic eczematous inflammation that results in thickened skin often associated with excoriation and fissures.
- Itch - Scratch - Itch Cycle
  - Often leads to difficulty sleeping
  - Scratching may lead to secondary infection

Vulval Disease – Lichen Simplex



Vulval Disease – Lichen Simplex



Vulval Disease – Lichen Simplex Chronicus

Differential Diagnosis LSC

- Diseases that cause itching
  - Psoriasis
    - More sharply demarcated
    - May include extra-genital locations
  - Candidiasis
    - Wet mount or yeast culture
  - Lichen sclerosus
    - No scaly appearance
    - May be concurrent condition
  - Infestations (lice & scabies)

Vulval Disease – Lichen Simplex Chronicus

Clinical Findings LSC

PHYSICAL EXAMINATION:

Thick, lichenified skin so labia are enlarged, rugose +/- edematous

Bilateral or unilateral

Localized to generalized

Color - variably pink, red, violaceous to ruddy brown; often a white appearance will be present when there is a thick keratin layer deposited on the surface of the epithelium

Secondary changes - erosions, ulcer, oozing, fissuring, honey-colored or serosanguineous crusting

Vulval Disease – Lichen Simplex Chronicus

Etiology LSC

LSC develops in several itchy skin conditions:

- Atopic dermatitis (eczema)
- Contact dermatitis
- Lichen sclerosus

Contact dermatitis can start this condition or be the main long-term promoting factor

Vulval Disease – Lichen Simplex and Contact Dermatitis

Etiology LSC

LSC develops in several itchy skin conditions:

- Atopic dermatitis (eczema)
- Contact dermatitis
- Lichen sclerosus

Contact dermatitis can start this condition or be the main long-term promoting factor

Vulval Disease – Lichen Simplex Chronicus - Treatment

Lichen Simplex – nonspecific measures

- Improve Skin Barrier Function
- Stop Excessive Hygiene
  - Avoid irritants (perfumes, dyes)
- Tepid Soaks
  - Soothing, promotes circulation, cleans
- Daily Skin Protection
  - "Band-aid" effect (zinc, Vaseline, veg. oil)
- Identify and treat co-existing conditions

Vulval Disease – Lichen Simplex Chronicus - Treatment

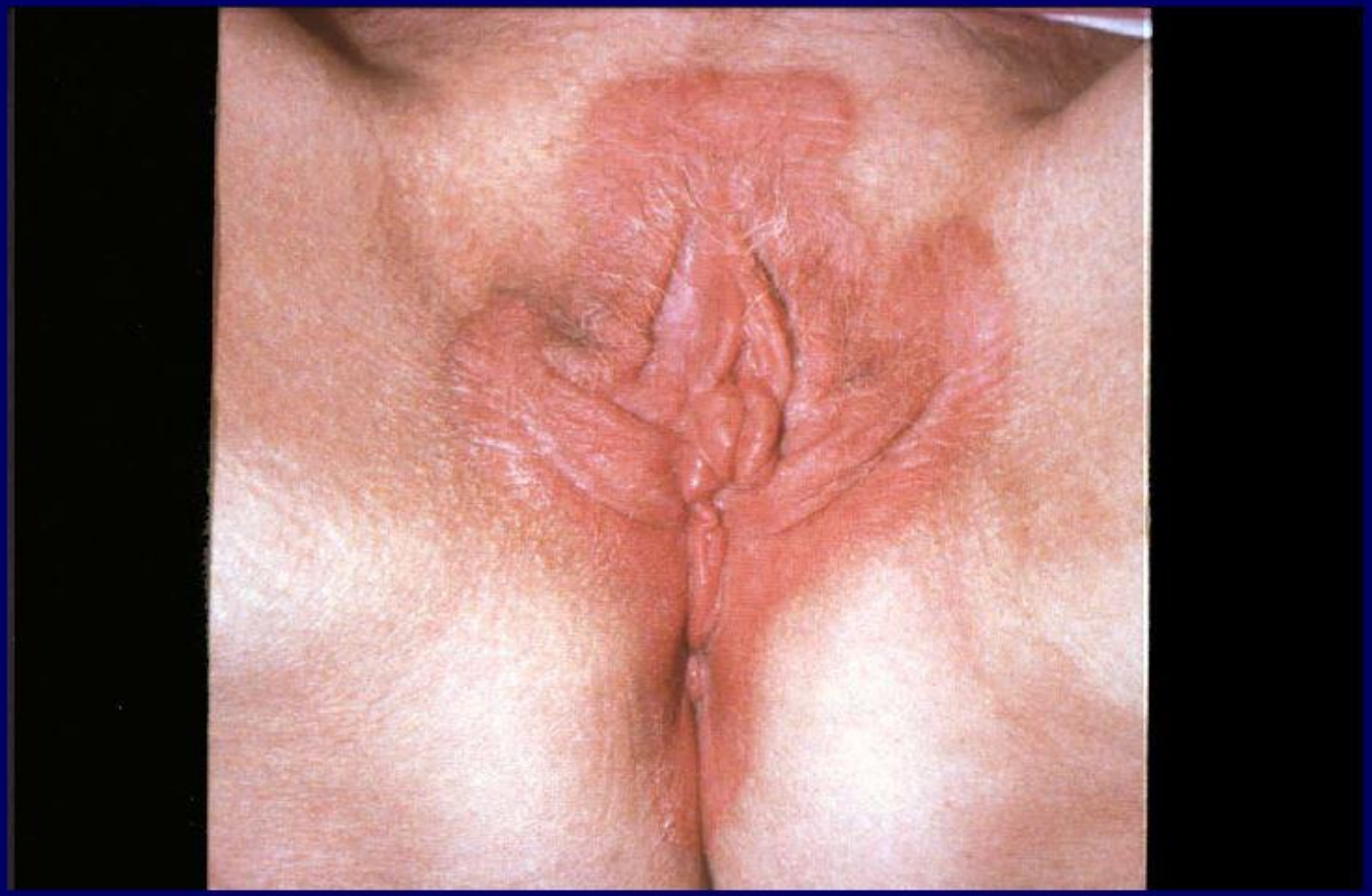
- Remove irritant/ contact allergen if possible
- Break itch-scratch cycle
- Look at various aspects of cleaning etc.
- Bland emollients
- Cetraben
- Steroid ointment –
- Most potent – Clobatesol (Dermovate) –
- Topically sparingly b.d. for 8 weeks, then in decreasing dosage
- Dermol washes

Vulval Disease – Contact Dermatitis

Common Vulvar Contactants

ALLERGENS	IRRITANTS
Benzocaine (Vagisil)	Soaps/cleansers
Preservatives	Sweat, urine, feces
Neomycin	Creams (alcohol)
Latex condoms	Douches
Chlorhexadine (K-Y)	Medications – TCA, 5FU
Lanolin (A&D ointment)	Spermicides
Perfume	Panty liners
Nail Polish	

Vulval Disease – Contact Dermatitis



Vulval Disease – Contact Dermatitis

Tips: Vulvar Contact Dermatitis

Irritation is the commonest cause

History of contact difficult to elicit

Loss of epidermal barrier
from - rash or loss of estrogen increases susceptibility to contact dermatitis

Suspect: allergic if sudden onset of rash
irritant if persistent rash

Vulval Disease – Contact Dermatitis

- Treatment/ Management
- Remove irritant/ allergen if known
- Bland emollients
- Dermol washes
- Steroids if required
- Barrier creams

Vulval Disease – Lichen Sclerosis

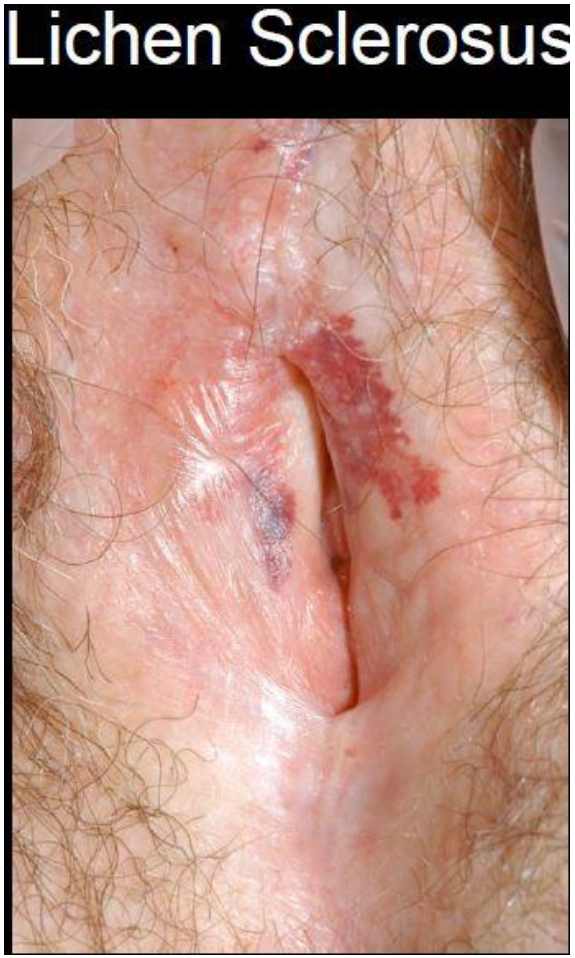
Lichen Sclerosus Introduction

- Common chronic vulvar disease
- Inflammation present
- Age range from childhood to elderly (bimodal distribution)
- Previously termed et atrophicus, now called lichen sclerosus

Vulval Disease – Lichen Sclerosis

- Chronic progressive disease which constrict and destroy the normal genital anatomy.
- Longer term, labia minora are lost ,labia majora flatten, clitoris can become inverted .
- Found on the vulva of postmenopausal women
- Can involve all the genital area from mons to anal area.
- Risk of malignant progression – 6%

Vulval Disease – Lichen Sclerosis



Vulval Disease – Lichen Sclerosis

Location of Lichen Sclerosus on the Vulva and Adjacent Areas

Labia	100%
Clitoris	70.4%
Perineum	67.9%
Buttocks	32.3%
Perianus	32.1%
Crural area	8.6%
Urethra	3.7%

Lorenz B. Kaufman RH. Kutzner SK. Lichen sclerosus. Therapy with clobetasol propionate. Journal of Reproductive Medicine. 43(9):790-4, 1998

Vulval Disease – Lichen Sclerosis

- Can be asymptomatic
- Most common symptom is prirutis
- Can be severe/ intolerable/ interfere with sleep
- Pruritis ani
- **OTHER SYMPTOMS**
- Burning
- Soreness
- Dyspareunia
- Pain with defaecation
- Constipation

Vulval Disease – Lichen Sclerosis

Signs

- Hypopigmentation
- Ivory white papules or plaques
- Cigarette paper appearance
- Cellophane-like sheen to surface
- Hour glass-figure of eight appearance
- Patchy or generalized
  - Vulva, perineum, perianal
  - No vaginal involvement

Vulval Disease – Lichen Sclerosis

Signs
Secondary Changes

- Fusion of labia minora
- Scratching yields open areas causing erosions
- Urinary retention
- Tearing

Vulval Disease – Lichen Sclerosis

Vulval Disease – Lichen Sclerosis

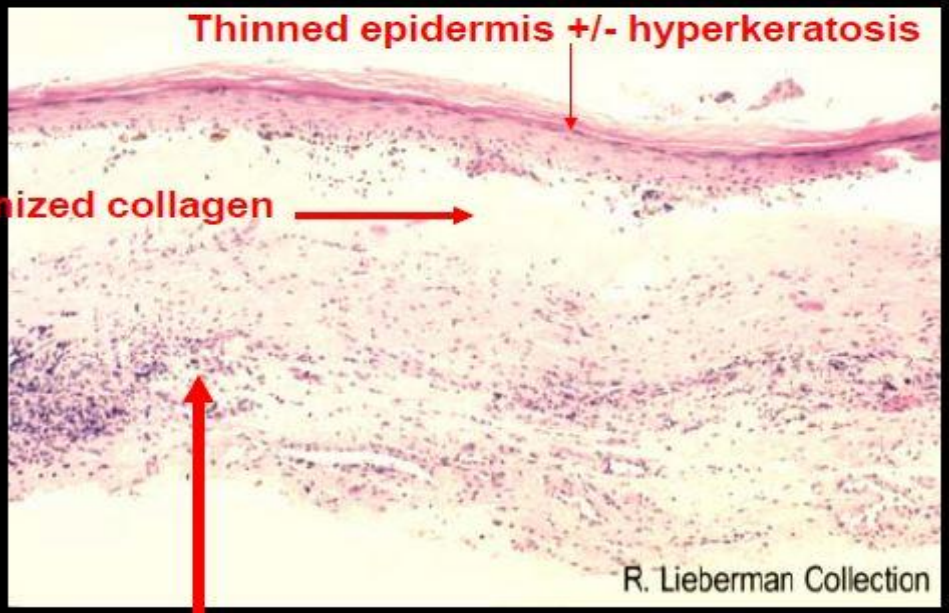
Vulval Disease – Lichen Sclerosis

Differential Diagnosis

- Vitiligo
- Lichen planus
- Lichen simplex chronicus
- Vulvar intraepithelial neoplasia (squamous and nonsquamous)
- Psoriasis
- Scarring from laser surgery
- Graft versus host disease
- Tinea

Vulval Disease – Lichen Sclerosis

Histopathology



Lymphocytic infiltrate under the band

Vulval Disease – Lichen Sclerosis

- **TREATMENT**
- Thorough assessment
- Biopsy if any concerns about VIN/ cancer
- Treat any superimposed infection – i.e. yeasts
- General Care measures –
- Avoid tight occlusive clothing
- Bland emollients
- 100% cotton underwear
- No soaps on vulva – can use Dermol washes

Vulval Disease – Lichen Sclerosis

Treatment of Lichen Sclerosus

- Superpotent steroid ointment (clobetasol propionate 0.05%)
  - Twice daily in a thin, invisible film for 1 month then daily for two months

Vulval Disease – Lichen Sclerosis

- **Maintenance**
- May require Clobatesol twice a week
- Decrease to Clobatesone? Or less potent steroid

- Aim of treatment to treat symptoms and to halt destruction of labia/ vulva by lichen sclerosis

- ??Decrease risk of transformation to VIN

Vulval Disease – Lichen Sclerosis

- **Other treatments**
- Topical Tacrolimus
- Oral Steroids – rarely required
- Side effects
- Intralesional injections - steroids

- Retinoids
- Anti-malarial agents – chloroquine
- Oral in intralesional

Vulval Disease – Lichen Sclerosis

- Surgical Treatment

- Limited role
- Healing
- Recurrence

- Surgical Division of mucosal adhesions helpful in clitoral phimosis

- Surgical division of introital narrowing or labia if unable to pass urine

Vulval Disease – Lichen Planus

Lichen Planus

- T-cell mediated autoimmune response to an unknown antigen
- Histology and morphology resemble other hyperimmune conditions
- May be found with ulcerative colitis, alopecia areata, vitiligo, morphea, dermatomyositis, LS, and myasthenia gravis

Vulval disease – Lichen Planus

Vulval Disease – Lichen Planus

Lichen Planus Disease Course

- Erosive mucosal LP typically chronic course with waxing and waning
- Progression to vulvovaginalgingival scarring is common
- SCC is recognized risk but rare
  - Estimated to be between 1% and 2%
- Nonresponsive lesions should be biopsied.

Vulval Disease – Lichen Planus

- Lichen planus can affect oral/ vaginal area
- Importance of examining oral cavity in patient's suspected of lichen planus

- Goal of treatment is relieve symptoms and prevent disease progression and complications

- Remission induction and maintenance

- Can usually be treated with topical agents
- Oral agents for severe cases

Vulval Disease – Lichen Planus

- Topical Steroids – potent first line therapy

- Clobatesol 0.05 ointment
- Initially topically twice a day

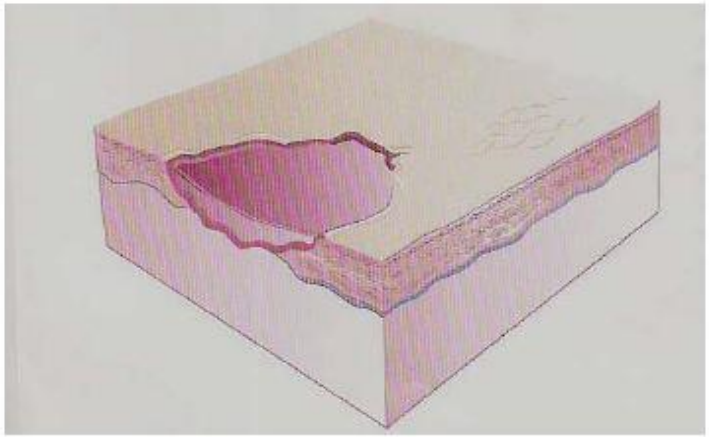
- May require maintenance

- Can require topical tacrolimus

- Intravaginal steroid

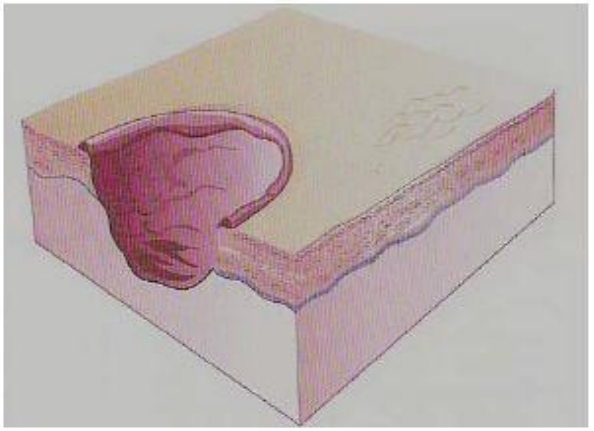
Vulval Disease – Erosions and Ulcers

Erosion



- Superficial or deep epidermis
- Not past basal layer

Ulcer



- Always secondary
- Full thickness extending into dermis or deeper

Vulval Disease – Erosions

Erosions

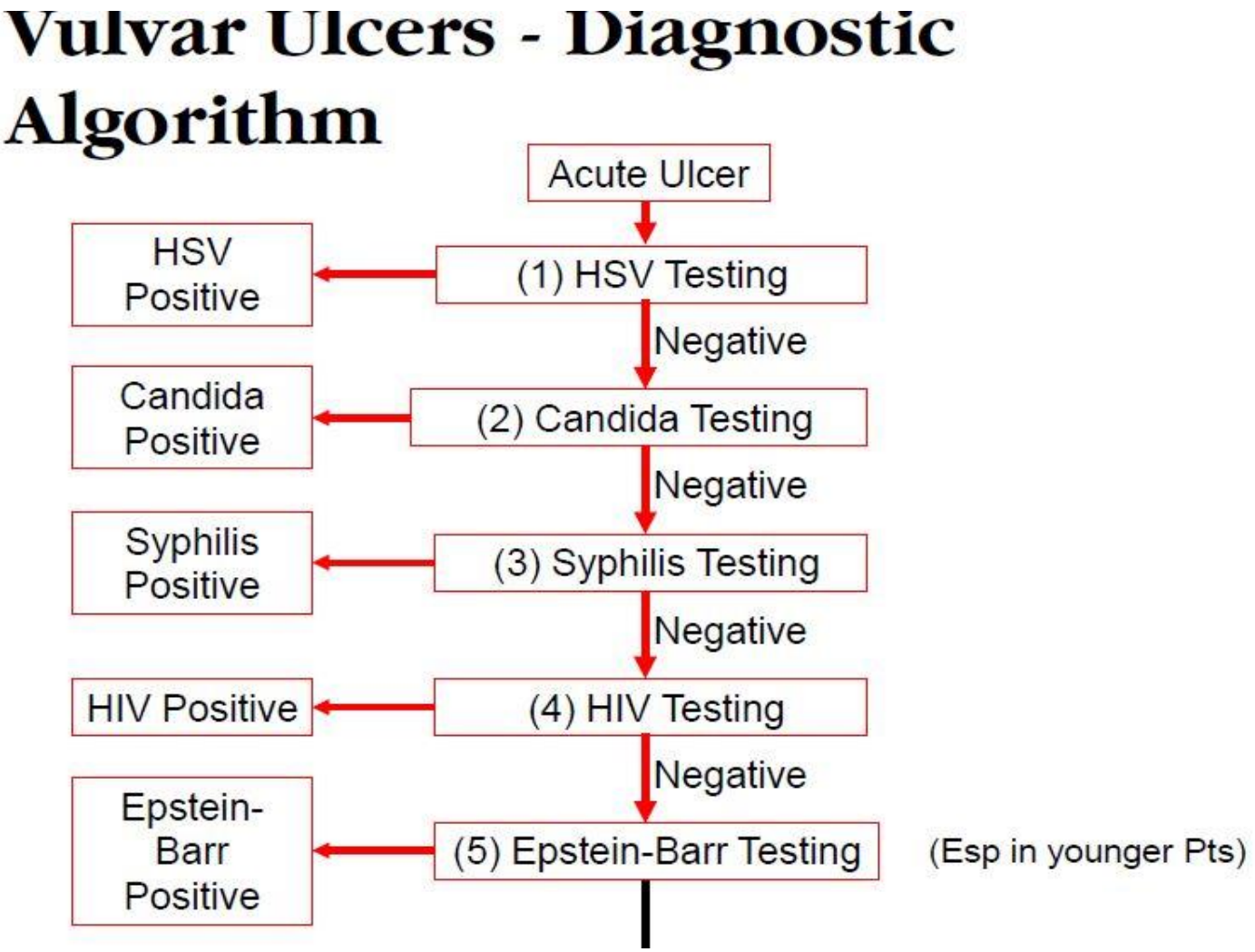
Common	Uncommon	Rare
• Excoriation • Fissures • Lichen simplex • Herpes simplex • Candidiasis • Erosive lichen sclerosus • Erosive lichen planus • Impetigo • Irritant contact dermatitis	• Allergic contact dermatitis • Post-coital fissures • Herpes zoster • Stevens Johnson syndrome • Pemphigus • Pemphigoid • Cicatricial pemphigoid • Pemphigoid gestations • Eroded malignancy • Plasma cell vulvitis.	• Hailey-Hailey disease • Extramammary Paget dis. • Toxic epidermal Necrolysis

Vulval Disease - Ulcers

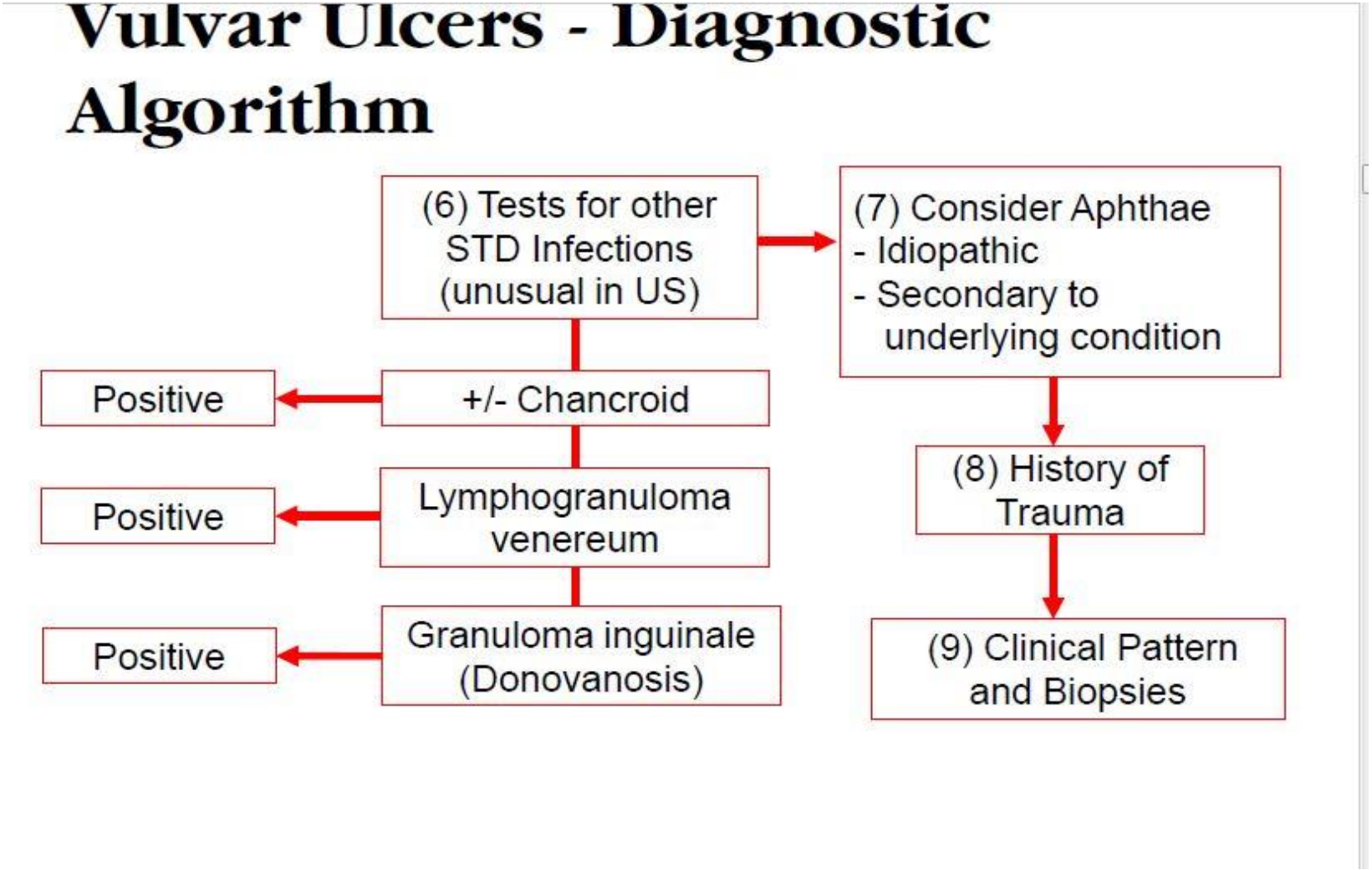
Ulcers

Common	Uncommon	Rare
• Aphthous ulcers, idiopathic • Aphthous ulcers 2° to infection or inflammatory disease • Crohn's disease • HSV in immunosuppressed	• Primary syphilis (chancre) • Trauma, accidental and self- induced • Ulcerated malignancy • Pyoderma gangrenosum	• Granuloma inguinale • Chancroid • Behçet's disease

Vulval Disease - Ulcers



Vulval Disease - Ulcers



Vulval Disease – VIN and Cancer

Two Types of VIN

VIN (usual) (Warty-Basaloid)

- HPV associated
  - HPV 16 most common
- Often multifocal
  - Associated with CIN, VaIN, AIN
- Mean age 30
- Increasing incidence
- 3-10% develop SCC

VIN (Differentiated) (Simplex)

- Not HPV associated
- Often unifocal
- Associated with longstanding LS
- Average age 67
- 2-10% of VIN
- Most diagnosed adjacent to or in follow-up of SCC

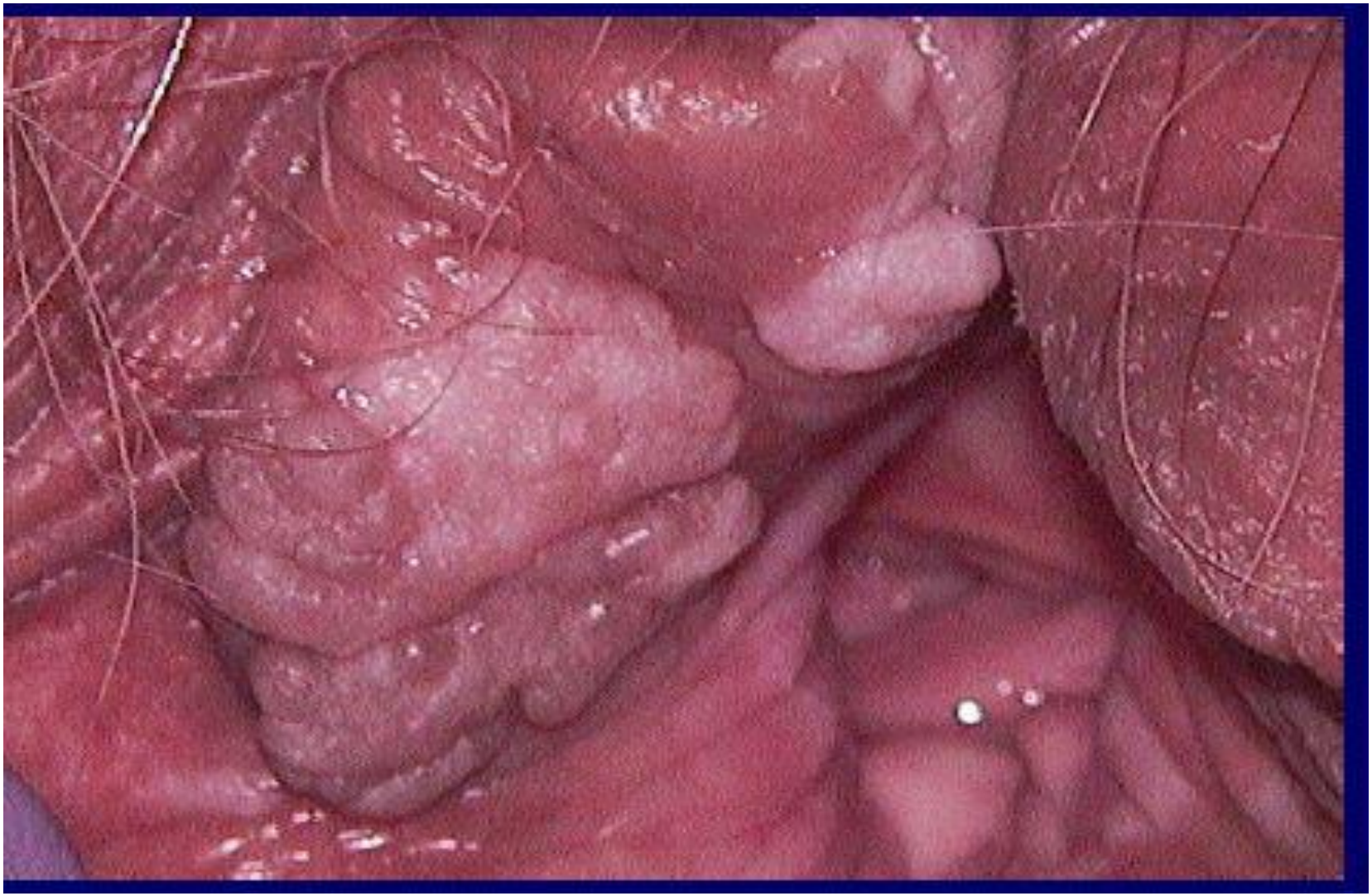
Vulval Disease – VIN and Cancer

VIN: Clinical Presentation

van Seters et. al. Gynecol Oncol 2005;97:645-651

- Based on review of 3322 published patients
- Symptomatic: 64%
  - Vulvar pruritis, burning, superficial discomfort, dyspareunia
- Dx made on finding visible lesion: 30%
- Multifocal: 49%
- Multicentric: 32%
- Cigarette Smoking: 84% in New Zealand Study

Vulval Disease – VIN and Cancer



Vulval Disease – VIN and Cancer

- Treatment of VIN
- Surgical excision for solitary lesions

- Wide local excision with 0.5 – 1cm margin
- /Skinning vulvectomy for large areas or multifocal disease

- Laser excision and Plasmajet excision

- Topical Imiquimod – as primary treatment vs adjunctive

Vulval Disease – VIN and Cancer

Outcomes of 63 women with Warty-Basaloid VIN followed without treatment

Jones RW et al. Obstet. Gynecol. 2005;106:1319-26

Course variable

- 47 (75%) regressed before tx
- 10 (16 %) progressed to invasive ca before tx
- 6 (10 %) persistent disease
  - 5 young women being followed with tx
  - 1 refused tx >20 years

Vulval Disease – VIN and Cancer

Treatment outcomes: Warty-Basaloid VIN

Jones RW et al. Obstet. Gynecol. 2005;106:1319-26

- Recurrent/persistent disease after treatment
  - Laser vaporization: (39%)
  - Excision: (34%)
    - Margins positive: 50%
    - Margins negative: 15%
- Recurrent / persistent disease after treatment
  - 50% if followed up to 13.7 years
    - Most required 2nd tx within 5 years of initial tx
  - Age a factor in need for repeat treatment
    - Age • • 24%
    - Age >30 36%

Vulval Disease – VIN and Cancer

Treatment of VIN with 5% Imiquimod Cream

- Immune response modifier
  - Stimulates secretion of cytokines including interferon - and and IL-12
  - Effective in treatment of condylomata accuminata
- 10 Studies / 99 Patients with VIN
  - Complete response: 48.5%
  - Partial response (≥ 50% reduction): 28.3%
  - No cases of invasive cancer during tx or f/u
- Toxicity
  - Local Itching, burning; less common, induration, excoriation, HA, myalgia, flu-like syndrome
- *Off label use*

Le T, Menard C, Hicks-Boucher W. et.al. Gynecol Oncol 2007;106:579-584

Vulval Disease – VIN and Cancer

Invasive Ca after Treatment for Warty-Basaloid VIN

Jones RW et al. Obstet. Gynecol. 2005;106:1319-26

- 17 of 312 women treated for VIN developed invasive cancer at least one year after treatment
  - 9 at site of initial tx
    - Median 2.4 years (range 1.1-7 years)
  - 8 at different site
    - Median 13.5 years (range 3-16 years)

Vulval Disease – VIN and Cancer – Imperial Audit

- Review 2002 – 2012
- Obtained from Histology database
- Reviewec with regards to diagnosis, outcomes, ‘other ‘ conditions

Results

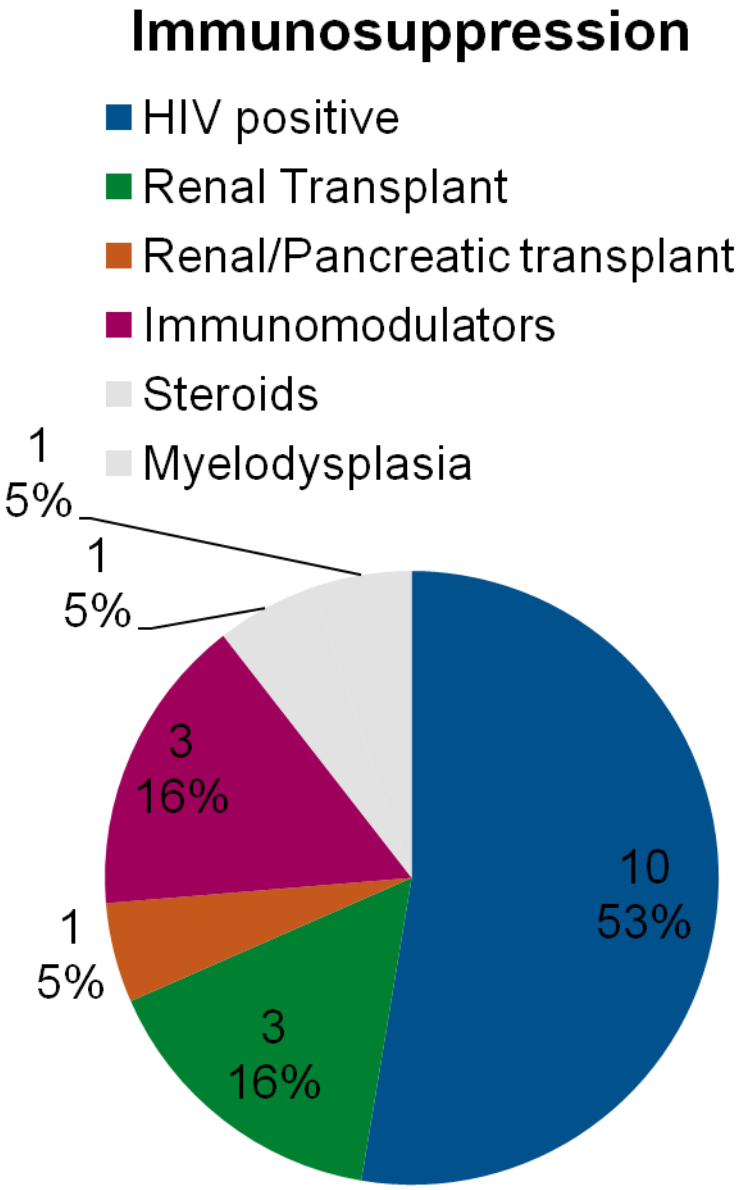
- 66 patients underwent investigation biopsy/excision
- 8 cases of benign HPV change
- 58 patients had high-grade dysplasia (VIN)
  - 10 (17%) cases were differentiated type VIN
  - 48 (83%) had usual type VIN
  - 27 (47%) had recurrent disease
  - 3 (5%) went onto develop invasive carcinoma
  - 40 (69%) had multicentric dysplasia

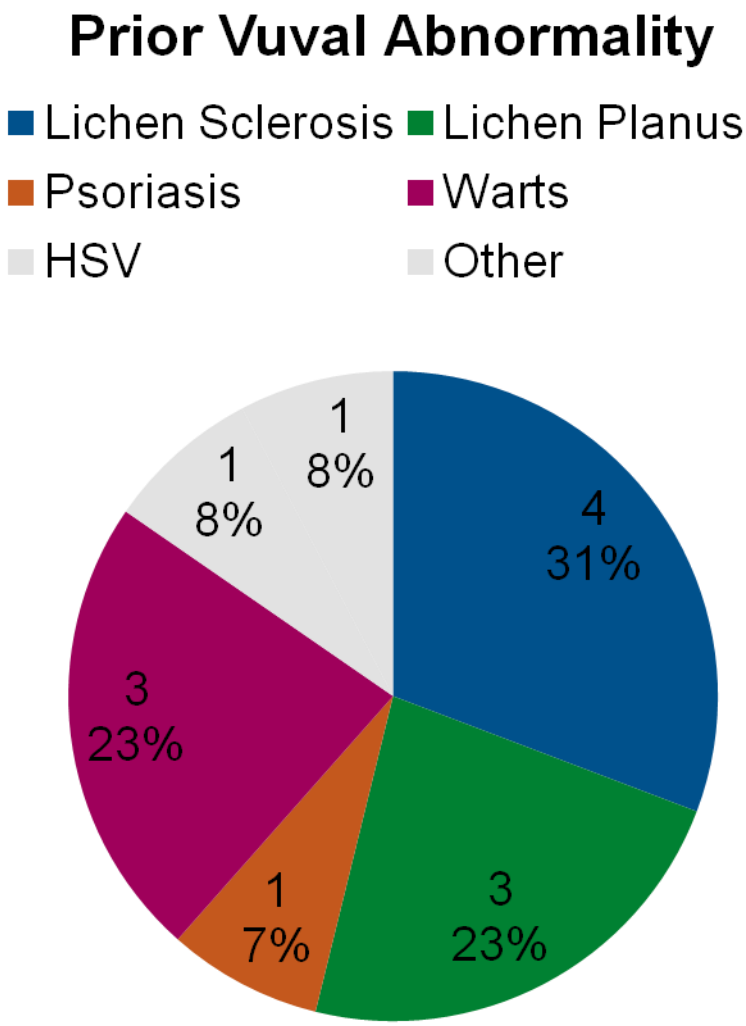
Demographics

Characteristics	n	%
Age (years) mean range	 44 21-86	
Nulliparous	27	59%
Multiparous	18	40%
Smoking	*18*	*28%*
Alcohol	*10*	*15%*
Immunosuppression	*19*	*29%*
Preexisting vulval abnormalities	*13*	*20%*

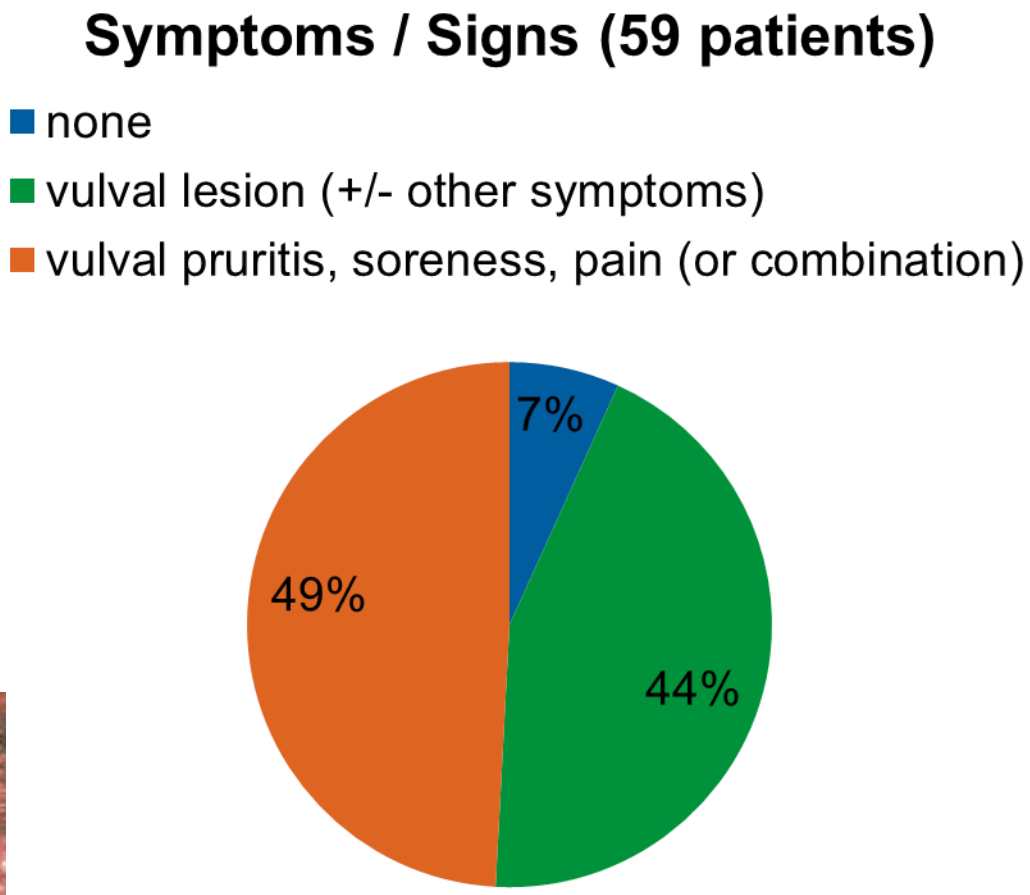
Van Seters *et al* – quotes mean of 46

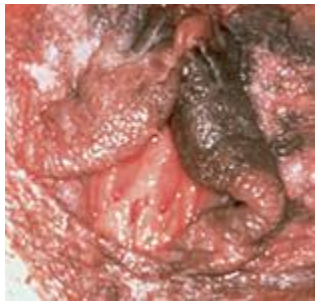
Demographics

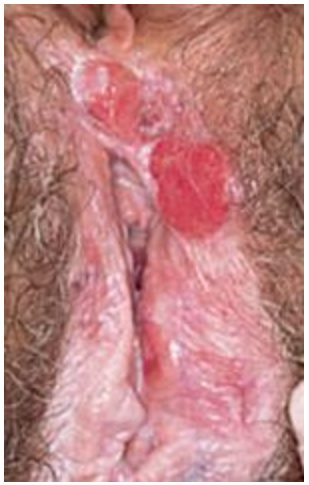




Referral



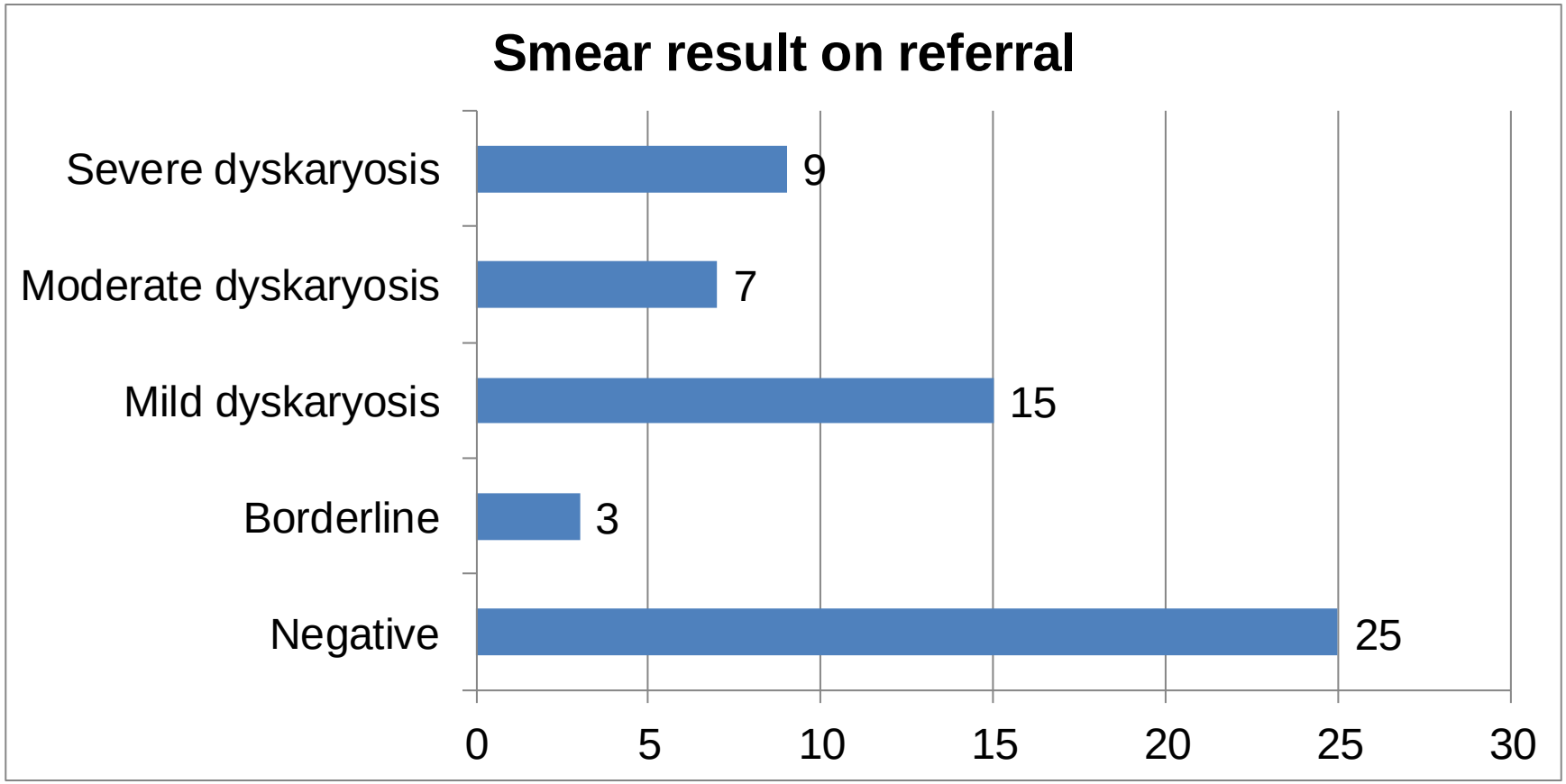




93% had symptoms OR a lesion or BOTH

Referral

- 35 (63%) had abnormal smears on referral

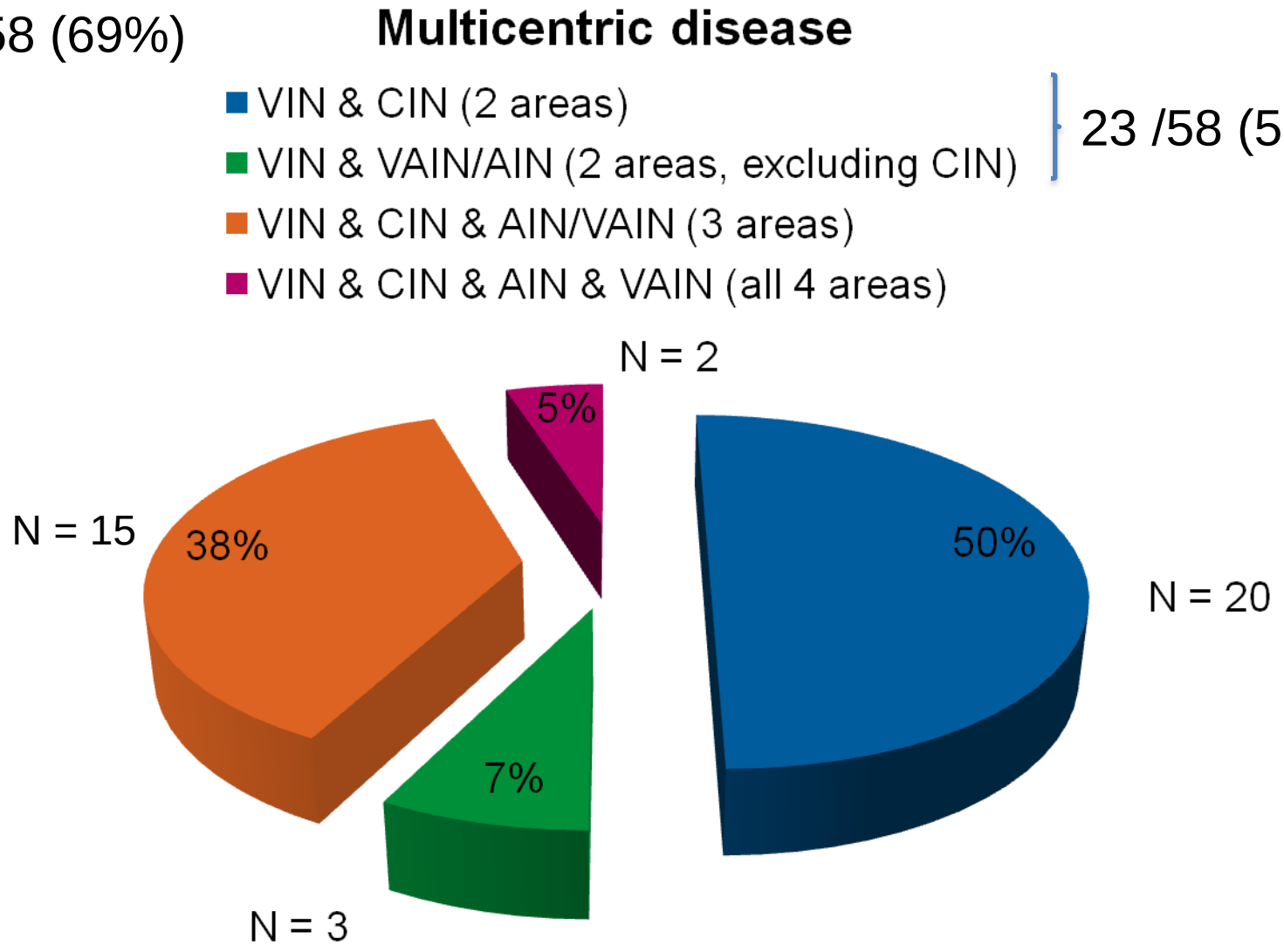


Differentiated VIN

- 10/58 (17%)
  - Average age 56 (range 35-86)
  - No smokers
  - Half were immunosuppressed (4 HIV+, 1 HSV)
    - All 5 patients had multicentric disease
  - 6 had multifocal disease
  - 4 had recurrent disease
    - Intervals of 1, 6, 16 years to recurrence
  - Range of histology :
    - 6 VIN3
    - 1 VIN2, 1 melanocytic change with atypia
    - 1 microinvasive
    - 1 invasive carcinoma at referral– lichen sclerosis
  - Treatment
    - 7 had excisional treatments (average #1.6 (range 1-3))
    - 2 patients had imiquimod and 1 vulvectomy

Multicentricity

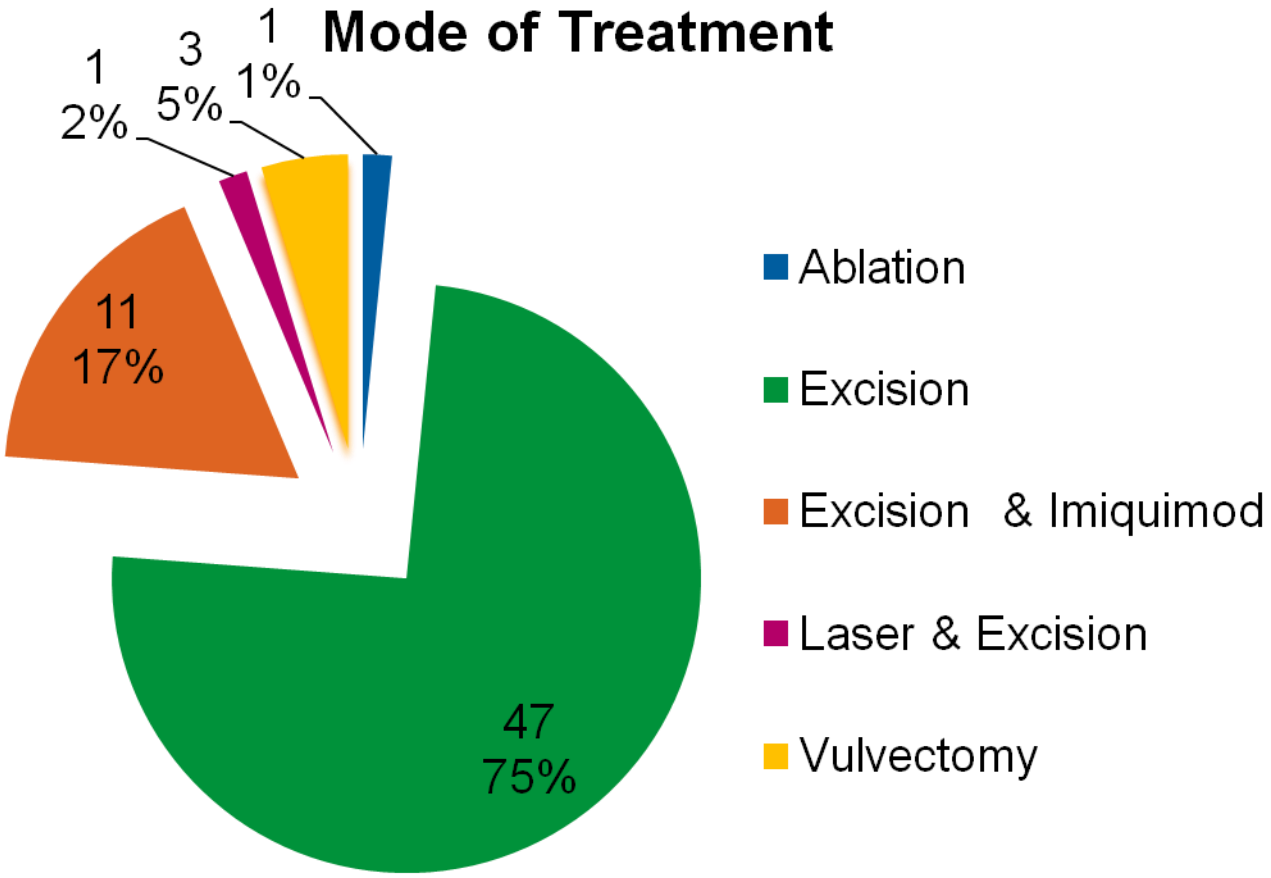
40 /58 (69%)



23 /58 (57%)

Treatment Method

- 63 patients received some form of treatment or required excisional biopsy



Recurrent VIN

- 27 (47%) + 2 incomplete excisions
  - Range of number of treatments 2-7 (mode 2)
  - Average time to recurrence or development of multicentric disease was 3.2 years (range <1-20 years)
- 5 microinvasive (9%)
  - 2 had VIN2/3 following prior excision for microinvasion

3 (5%) had invasive disease

- Average age 54 (range 33-86)
  - Included 2/4 of lichen sclerosis patients (differentiated type)
  - Had multiple VIN diagnoses, including VIN3
  - All had 1-5 previous excisions, 1 failed IMQ
  - All invasive disease was treated with vulvectomy

Vulval Disease

- Thank you for your attention
- What questions would you like to ask