VULVAR DISEASE UPDATE

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Conflicts of interest
Honorarium from Taro
Little evidence based treatment
Too few studies done in vulvar disease.
Most treatments discussed are “off-label”

Why Do Vulvar Disease ?
VULVAR CARE IS COMMONLY UNAVAILABLE
For women this is devastating
- suffer with undiagnosed symptoms
- waste millions of dollars on anti-yeasts
- hide and scratch
- endure vulvar pain and dyspareunia
- are desperate for help

VULVA !
What is that?
Down there?

Vulvar Education
Let's eliminate the
“Down there” generation
Use diagrams and handouts
See www.issvd.org - patient education
**BASIC PREMISES**

- Patient anxiety and depression
- Normal anatomy can be confusing
- Look carefully for subtle abnormalities

**Punch Biopsy**

**Pre-anesthesia** -
- use a topical anesthetic
  - 2.5% prilocaine / lidocaine cr
  - 5% lidocaine oint
  - 15 min under plastic wrap
- Use a punch, shave or scissor biopsy
- Stop bleeding:
  - Monsel’s or ferric chloride
  - 5 or 6-0 Vicryl suture

**Herpes Simplex Virus (HSV)**

- Commonest cause of vulvar erosions / ulcers
- Primary HSV is uncommon
- HSV I increasing
- Reversal of HSV I/I ratio - for new cases 70-80% HSV I
  - Usually spread from an asymptomatic partner
- Women are unaware of their infection and most often present with recurrent HSV with no primary HSV history

**Candidiasis**

- Candidiasis is the commonest genital disease
- Causes:
  - Candida albicans 75%
  - Candida tropicalis, etc. 25%
- NO TELEPHONE DIAGNOSIS
- Candida can complicate all vulvar problems
  - e.g. LP, LSC, LS, Contact

**Herpes Simplex Virus (HSV)**

- Think of HSV in a patient with a non-healing genital very painful ulcer
- Onset as erosions that deepen and extend with punched out ulcers or ulcerated nodules.
- Can be treatment resistant in HIV

**Diagnosis** -
- Tsank smear, PCR, culture, Type-specific serology, biopsy

**Biopsy**

- Site ○
- Number 1-3
- size 4 mm
- H&E + IF

Any question - do more than 1 biopsy
### Treatment Candidiasis

**Topical** imidazole cream or vag tabs - 1, 3, 7d  
nystatin cream or vag tabs  
**Oral** imidazole - fluconazole 150 mg on day 1, 3, 7  
**Suppression**  
clotrimazole 500g vag tab weekly or 200 mg twice a wk  
fluconazole 150 - 200 mg orally weekly  
ketoconazole 100 mg orally daily  
itraconazole 100 mg orally daily

**Resistant Candida**  
boric acid vag suppositories 600mg X 14 d

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### Vulvar Patients are desperate! with Itch, burn, pain

They try to:  
“Wash it away” and “Clean up” the dirty area using Soaps, cleansers, Anesthetics, Yeast Rx

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### Vulvar Contact Dermatitis

**Primary irritant:**  
Prolonged or repeated exposure to caustic or physically irritating agent  
This is a “chemical burn”  
Very common with ALL vulvar problems

**Causes:**  
Hygiene habits - soap, wipes, pads  
Moisture - urine, feces, sweat  
Topicals - lotions, antifungals

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### Treatment Vulvar Contact Dermatitis

**Stop Contact** - Irritant or Allergen  
- Stop irritants - Educate patient  
- Stop scratching - Treat infection - yeast, bacteria  
- Patch Test as indicated  

**Control Inflammation**  
- triamcinolone 0.1% oint twice a day for 7-10 d  
- If severe, systemic corticosteroids

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### Vulvar Contact Dermatitis

**Frequent**  
Complicates all vulvar conditions

Irritant contact most common  
Skin barrier lost from soaps, urine, feces

BEWARE THE “DIRTY” VULVA
**Psoriasis**

In genital area - often atypical
moist, thin red patches in skin folds, gluteal cleft
In hairy areas - red, scaly papules, plaques
Secondary changes with infection, common
- fissuring, pustules

**Often missed or hidden**

Please Ask

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**Psoriasis Treatment**

Stop irritants

**Treat infection** - yeast and bacteria

Stop inflammation
- topical steroids - triamcinolone 0.1%
  ointment am pm 5-7d then
- tacrolimus 0.1% ointment or
  pimecrolimus 1% cream
- if severe, systemic medications

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**Lichen Simplex Chronicus (LSC)**

**End stage of the cycle**

Itch ↔ Scratch ↔ Itch

Worse with heat, humidity, stress and irritants

Associations: atopic dermatitis, psoriasis, contact dermatitis

**Scratching feels so good**

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**Characteristics of Lichen Simplex Chronicus**

Relentless pruritus
dyspigmentation
“nothing helps”
excoriations, crusts
“years of itch”
lichenification
uni or bilateral
hair loss

The diagnosis is clinical

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**Treatment LSC**

**Optimize Epithelial Barrier function:**

Control infection (cefadroxil, fluconazole)
Reducing heat, sweat, irritation
Stop irritants - Stop excessive hygiene

Immediate therapy:
Tap water soaks in tepid water
Use cool packs or compresses to deaden nerves
No hot water - No ice packs
Seal in moisture with ointments long-term

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**Confirm diagnosis - biopsy ?**

- Educate patient
- Patch Test as indicated

**Stop Itch-Scratch-Itch cycle**

- Cool sitz baths/gel packs
- Sedate - doxepin or hydroxyzine PM, citalopram AM
- Topical superpotent steroids
clobetasol 0.05% oint. bid x 2 wks, OD x2 wks, MWF x 2 wks
- Severe - prednisone taper or IM triamcinolone
  1mg/kg up to 80 mg/dose

Look for more than one cause
### Treatment Tips LSC

If water stings raw skin:
- Normal saline Sitz baths:
  - 1 tsp salt in 3 cups of water
  - Soak for 5-7 min 2-3 times a day

For recurrent infection:
- Swab skin folds and nose for C&S - R/O MRSA, Candida
- Bleach Baths - 3 times a week
  - ½ cup bleach in 10” water for 5-7 min
  - 1 ¼ teaspoons of bleach per gallon (4 liters) of water

### Lichen Sclerosus

A chronic, autoimmune, mucocutaneous disease affecting the genital skin causing whiteness, tissue thinning and scarring affecting perimenopausal women 40-50 yrs

**Commonest cause of chronic vulvar disease**
- Prevalence 1:300 - 1:1000
- 670 cases in Boston - 6,600 in metropolitan area
- 15% cases children
- 15% have extragenital disease

**3-5% SCC**


### Lichen Sclerosus Treatment

**Confirm diagnosis - biopsy**

- Stop irritants
- Educate patient

**Control inflammation**

- clobetasol or halobetasol 0.05% oint once or twice a day for 3 months then maintenance 1-3 times a week
- If very thick, consider intralesional triamcinolone

Not responding?

- Reassess, rebiopsy, R/O SCC, contact
- Follow forever

### Lichen Sclerosus Alternate Treatments

**Calcineurin inhibitors:**
- pimecrolimus 1% cream bid
- tacrolimus 0.1% ointment bid

**Antimalarials** - hydroxychloroquine 200mg bid
- Methotrexate 10 -15 mg/ week po or sc + folate 1 mg/d
- Cyclosporine 4-5 mg/kg/d

**Compliance issues children and teens - 2.5% HC oint daily**

### Lichen Sclerosus

For very thick poorly responding LS

- Prednisone for short 2-3 week course

- Intralesional triamcinolone
  - 3.3-10 mg/ml after topical anesthetic
  - Monthly for up to 3 months

**Sites of Injections of T/C**

- 2.5 to 10 mg/ml

**Before RX**

- 2.5% lidocaine + prilocaine for 20-30 min

**MANAGEMENT PRINCIPLES**

- Explanation of the disease process, treatments, expectations
- Handouts
- Photographs
- Treat all factors

**Anticipate and minimize iatrogenic disease** (yeast, irritant contact dermatitis)

- Avoid cream vehicles on painful, inflamed, or estrogen-deficient vulvar skin
- Avoid topical therapy in general and use oral medications, except for corticosteroids
- The vulva is relative steroid resistant - use ultrapotents

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**Lichen Planus**

An autoimmune, mucocutaneous disorder of altered cell mediated immunity in older women - 50 - 60 years

- **Affects:** Skin, scalp, nails, mucous membranes - oral, genital, anus, esophageal, urinary tract
- Responds to immunosuppressive therapy
  - 2-5% SCC

**Always Examine the Vulva & Vagina & Mouth**

**Lichen Planus Vulvar Patterns**

- **60% itch**
- **70-75% pain**

**Classic**
- White, lacy, or fernlike topped papules

**Hypertrophic**
- Extensive white, thick scarring, destruction
- Very itchy
- Looks like LS

**Erosive**
- Red plaques with whitish to lacy edges
- Glazed erythema, erosions, ulcers, scarring

**Erosive Vulvovaginal LP**

- **Pain plus burning**
- Deep red erosions, glazed erythema with thin gray edge
- Fern-like or lacy white pattern
- Variable scarring / loss of architecture

**Always examine vagina**

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**Vaginal Lichen Planus**

In the vagina: may be asymptomatic

**LP causes Desquamative Inflammatory Vaginitis** DIV with:

- Pain - Symeclia - Stenosis/Scarring
- Erosions - Dyspareunia - Discharge
- Mucopurulent vaginal discharge - yellow, green, bloody

**Erosive Vulvovaginal LP**

- Pain plus burning
- Deep red erosions, glazed erythema with thin gray edge
- Fern-like or lacy white pattern
- Variable scarring / loss of architecture

**Always examine vagina**

**Examine:**
- Vulva + Vagina + Anus + Mouth + Skin
Lichen Planus Treatment

**Confirm diagnosis - biopsy**
- Stop irritants
- Educate patient
- Stop scratching
- Control infection

**Control inflammation**
clobetasol or halobetasol 0.05% oint
intralesional, vaginal or systemic corticosteroids
topical tacrolimus (Protopic) 0.03%, 0.1% oint - burns
2 mg suppositories qhs

Corticosteroids for Lichen Planus

**Systemic Corticosteroids:** Use for severe LP
Triamcinolone 1 mg/kg up to 80 mg/dose IM
(Kenalog 40®) q4wks x 3
Prednisone 40-60 mg PO OD, decreasing dose

**Intravaginal Hydrocortisone acetate QHS:**
- suppository 25mg (available) or 100 mg (compounded)
- 10% compounded vaginal cream, 4 - 5g per dose
  (400-500mg/dose)
Use nightly for 14 days then Mon Wed Fri use dilators
Note: adrenal suppression and candidiasis

Lichen Planus Treatment

**SEVERE**
Systemic corticosteroids - Triamcinolone 1mg/kg IM every 4 weeks for 3-4 shots

- Methotrexate 5-10mg/week PO or SC
- Cyclosporine 4-5 mg/kg/d -3-4months
- Mycophenolate mofetil 500mg -1.5 g bid
- Etanercept 50 mg 1-2 doses / week ?
- Hydroxychloroquine 200 mg bid
- Acitretin (1 mg/kg)

The Lichens

<table>
<thead>
<tr>
<th>LS</th>
<th>LP</th>
<th>LSC</th>
</tr>
</thead>
<tbody>
<tr>
<td>Itch or burn</td>
<td>Itch or burn</td>
<td>++++ Itch</td>
</tr>
<tr>
<td>Scars</td>
<td>Scars</td>
<td>No Scar</td>
</tr>
<tr>
<td>Not In Vagina</td>
<td>In Vagina And Mouth</td>
<td>Not In Vagina</td>
</tr>
</tbody>
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Causes of Treatment Failure

1) Noncompliance
   - poor education
   - Fear of topical steroids
   - Limited mobility
2) Incorrect diagnosis - was a biopsy done ?
3) Associated problems
   LS, LP, contact, Candida, HSV, estrogen loss, SCC

Mucous Membrane Pemphigoid

Rare mucocutaneous scarring & blistering disease
Autoimmune - auto-antibodies to epidermal basement membrane
Sites:
- mouth 85%
- eyes 65%
- vulva 50%
- skin 25%
Non-healing erosions vulva LP , LS, Pemphigus
- desquamative vaginitis
- scarring
Diagnosis : histopathology and immunofluorescence
Vulvar Squamous Cell Carcinoma

Commonest vulvar malignancy
85-90% of all vulvar cancers
Classification - Intraepithelial
- Invasive

Vulvar SCC

30-40% vulvar SCC occur in lichen sclerosus
Lichen sclerosus and lichen planus can develop SCC in 3-4% cases

Vulvar Aphthous Ulcers

“Canker Sores” on the Vulva

- Average age is 14 (9-19) yrs
- Sudden onset
- Usually multiple, painful, well demarcated punched-out ulcers
- Size: most <1cm; can be 1-3 cm
- Prodrome - flu-like with mild fever, headache, malaise
- Duration 1-3 weeks, can last months
- One episode, less common recurrent
- Past history of oral aphthae – canker sores
- Not Behcet’s

VIN and Vulvar SSC

Intraepithelial
Vulvar intraepithelial neoplasia (VIN 2-3, high grade VIN)
VIN, usual type - multifocal
- HPV 16, 18
VIN, warty type
VIN, basaloid type
VIN, mixed (warty/basaloid) type
VIN differentiated type - solitary
- In older women, associated with LS

Invasive SCC
- in women > 65 yr.
- up to 40% - lichen sclerosus

Causes of Vulvar Ulcers in North America

Infections

- Venereal
  - Herpes simplex
  - Syphilis
  - HIV
- Non-Venereal
  - EBV
  - M. pneumoniae

Dermatoses - Non-bullous

- Aphthosis
  - Lichen planus
  - Lichen sclerosus
  - Crohn’s disease
  - Contact
  - Drug

- Tumors
  - Squamous cell carcinoma

APHTHAES - Acute vs Chronic

Acute (more common)
- usually a prodrome - fever, headache, malaise, GI upset
- EBV, Mycoplasma pneumoniae, viral upper respiratory infection or gastroenteritis, influenza, Strep

Recurrent / Complex (recurrent oral and genital aphthae)

- Inflammatory Bowel disease - Crohn’s, Ulcerative colitis, Celiac disease
- Behcet’s disease
- Medications - cytotoxic, NSAIDs
- Myeloproliferative disease, cyclic neutropenia, lymphopenia
- HIV

Hupper et al. J Ped Adolesc Gyn 2006;195-204
Farhi et al. Arch Dermatol 2009;145;38-45

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Farhi et al. Arch Dermatol 2009;145;38-45
### Vulvar Aphthae

#### Diagnosis

- Diagnosis of exclusion
- Look for associated conditions
- Cultures negative, biopsies non-specific and blood work non-contributory
- DDx: HSV, EBV, drug, Crohn’s, Behcet’s, Syphilis, HIV

#### Vulvar Aphthae - Therapy

- **Pain control** - topical, systemic
- **Prednisone** 40 - 60 mg each morning until pain resolves (3-5 days, then ½ dose 3-5 days)
- Ultrapotent corticosteroid
- **Educate** - Most often a one-time event, can recur
  - If not controlled:
    - Intralesional triamcinolone (Kenalog 10) 5-10 mg/ml
    - Colchicine 0.6 mg bid-tid if tolerated
    - Dapsone 50-150 mg per day
    - Dapsone + Colchicine
    - Pentoxyfylline 400 mg tid
    - Thalidomide 100-150 mg per day

### Epstein-Barr Virus (EBV)

- Lymphotropic herpes virus
- Age - teens to early 20s
- Clinical - acute onset
- Fever, malaise
- Sore throat - lasts 2 to 3 weeks
- Punched out painful ulcer(s)
- Recent hx of mono in family member

- Serology - EBV IgM anti-VCA antibody

### Vulvar Behcet’s Disease

- Rare
- Triad - oral ulcers, genital ulcers, uveitis
- Clinical -
  - Painful, punched-out vulvar ulcers
  - Last 2-3 wks
  - Heal with scars, sinuses
  - Erythema nodosum
  - Minor to no eye problems
- Good prognosis

### Vulvar Crohn’s Disease

- Chronic inflammatory bowel disease
- Rare on vulva - 2% women have vulvar lesions
- Contiguous
  - Direct fistulae from bowel to skin
- Non-contiguous/metastatic
  - Painful labial edema +/- ulcers
  - “Knife cut” ulcers
- Non-specific
  - Aphthae - oral and vulvar

### Drug Eruptions

- Drugs can cause 100 different skin reactions
- Many can be seen on the vulva
- Erosive/ulcerative types -
  - Erythema multiforme
  - Aphthous ulcers
  - Pemphigoid
- Fixed eruptions
  - LP and LE eruptions,
**Topical Treatment Instructions**

Give specific instructions for topicals

**Amount to use**
- show her, in the office, how much to use
- squeeze onto your finger, and demonstrate

**Application site**
- some women have never seen their vulva
- use a mirror, and show the exact place(s)
- give her a diagram with correct areas indicated

www.issvd.org Patient Education

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**How much to apply?**

“PEA SIZED” AMOUNT?

“TOOTH PICK” Dermatology

The amount of ointment or cream on the large end of a toothpick will cover the vulva

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**Commonest Missed Concurrent Vulvar Diseases**

- Candidiasis
- Contact Dermatitis
- HSV
- Atrophy
- Cancer

LOOK FOR MORE THAN ONE PROBLEM

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**Treat Vulvar Disease**

Listen - review history
Biopsy and RE-BIOPSY
Look for
- infection - Candida, HSV, bacteria
- trauma from aggressive hygiene or other practices
- contact dermatitis
- squamous cell carcinoma
Educate
Support - counseling as needed
Assess compliance

LOOK FOR MORE THAN ONE PROBLEM

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**Vulvar Disease Treatment Handout**

Handout: Your Diagnosis Is? Test Your Knowledge of Vulvovaginal Disorders
Available at:
http://www.med.umich.edu/obgyn/cvd/ref_phys.htm
Click on Summary of Vulvovaginal Diseases

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issvd.org
Vulvar Disease References


See Dermatologic Therapy Diagnosis and treatment of vulvar and Vaginal Disorders Vol 23 Oct 2010
Dermatologic Clinics Vulvovaginal Dermatology Vol 28 Oct 2010