Common Vulvar Complaints and Management
What’s Your Diagnosis?

Hope K. Haefner, M.D.
Professor of Obstetrics and Gynecology
Co-Director, The University of Michigan Center for Vulvar Diseases
The University of Michigan Hospitals

Lynette J. Margesson, M.D.
Assistant Professor of Obstetrics and Gynecology and Medicine (Dermatology)
Dartmouth Medical School

This handout is available at

http://obgyn.med.umich.edu/patient-care/womens-health-library/vulvar-diseases

or go to Google and type in University of Michigan Center for Vulvar Diseases

click on Information on Vulvar Diseases

Disclosures:

Hope Haefner, MD is on the advisory board of Merck Co. Inc.

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Learning Objectives

At the end of this course, the participant should be able to:

- Identify the clinical features of various vulvovaginal conditions
- Recognize the gross features of non-neoplastic epithelial disorders of the vulva
- Identify the various ulcerative conditions of the vulva and their treatments
- Become familiar with a variety of treatments for skin diseases

A variety of dermatologic conditions affect the vulva and the vagina. It is important to become familiar with the appearances and treatments of the numerous vulvovaginal conditions that you may see in your patients.

Nonneoplastic Epithelial Disorders

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Lichen sclerosis

Lichen Sclerosis – is chronic, autoimmune disease affecting the genital skin causing whiteness, tissue thinning and scarring. It is the most common chronic vulvar condition

Histology - blunting or loss of rete ridges, hyperkeratosis and loss of melanocytes are seen with a zone of pallor and often a dense interstitial lymphocytic infiltrate.

Pathophysiology: Unknown. Various genetic, autoimmune, infectious and local factors are implicated. The cause is probably multifactorial with a genetic, environmental and possibly infectious input. Often associated with other autoimmune diseases. Thyroid disease is the most common. Familial cases have been reported.

Age of onset - middle age (about 40 years) but range is from less than one year to > 80 years
Symptoms - Pruritus is most common and can be severe and intolerable
Scratching causes secondary changes and open areas that cause dysuria, burning and dyspareunia
Scarring leads to dyspareunia, even apareunia
May be asymptomatic - common cause of asymptomatic vulvar scarring.

Physical exam – Scattered or confluent papules forming plaques of ivory white with cellophane-like sheen on the surface. Found anywhere on the vulva from the clitoris and periclitorally to the gluteal cleft. The involvement may be patchy or generalized in various patterns. Classically a “figure-of-eight”. It can involve any cutaneous surface but most commonly is found on the vulva in women. Extragential disease occurs in 10-20%. LS typically does not involve the vagina.

Secondary changes - excoriations, purpura, erosions, thickening (lichenification) crusting, and scarring, ranging from loss of labia or burying of the clitoris to loss of all normal vulvar structures.

Differential diagnosis - sexual abuse in children, vitiligo, lichen simplex chronicus, lichen planus, cicatricial pemphigoid.

Cancer risk - about 4% develop associated SCC

Treatment:
- Biopsy to confirm diagnosis
- Educate the patient
- Stop irritants
- Recommend cool, ventilated clothing
- Topical superpotent steroids (various regimens exist)

  Clobetasol propionate or halobetasol 0.05% ointment qd
  for 12 weeks, then M-W-F or 1-2 times a week and follow up at 6-12
  weeks then regularly at 6-12 month intervals
  versus
  Clobetasol propionate 0.05% bid x 1 month, then q d x 2 months. Decrease use of
clobetasol to 3 times down to once a week. In some cases decrease to a class 4 steroid
(see steroid table at the end of the handout), then gradually decrease frequency of
application to once a week. (There is debate regarding whether or not long term steroids
are required.)

- Treat associated Candida or secondary bacterial infection
- Stop scratching as this keeps LS active. Give 10 mg of hydroxyzine or doxepin at 6 to 7
PM to stop nightly scratching. (See Lichen Simplex Chronicus below)
- For thick lichen sclerosus consider intralimsional steroid (triamcinolone 3.3 to 10 mg/ml).
The dose is dependent on the location and thickness of the skin that is being injected.
This can be repeated monthly for 2-3 months. Do not inject high steroid doses into thin
skin or in small areas because the tissue can slough.
- If constantly scratching use IM triamcinolone 1 mg/kg up to 80 mg/dose. Never give
over 80 mg of triamcinolone acetonide IM per month. This can be repeated once a month
for 3 months with a maximum of 4 doses a year.
Tacrolimus 0.1% ointment and pimecrolimus 1% cream have been used for the treatment of vulvar lichen sclerosus. Burning may occur with these medications.

Tazorac 0.1% gel (can also use 0.05% or 0.1% cream for lower strength) may be used for lichen sclerosus when the skin is very thick or unresponsive to topical steroids. Apply to skin qhs with gradual decrease to two to three times a week.

Acitretin (Soriatane) is a retinoid that may be used for lichen sclerosus unresponsive to topical steroids (and in some cases lichen planus). It is most beneficial for thickened skin. Take 10 mg every 1-2 days for a dose of 30-70 mg per week. It must be taken with fatty food. The patients must not become pregnant as it is tearatogenic like isotretinoin. (expensive, but less costly in Canada).

Surgery is done on occasion to improve function or for scarring

In all patients with lichen sclerosus:
- Arrange follow-up always – indefinitely.
- Regular follow-up is needed because there is an increased risk of developing squamous cell carcinoma (SCC) (<5% in women). If not responding to treatment Look for concurrent conditions and biopsy and rebiopsy, as needed.

Note – LS involves the vulva not the vagina unless prolapse. Scarring is not reversible by any medical therapy.

**LICHEN SIMPLEX CHRONICUS (LSC)**

**Synonyms:** Squamous cell hyperplasia, neurodermatitis, pruritus vulvae, hyperplastic dystrophy

“LSC” – The end stage of the itch – scratch – itch cycle. It is usually part of the atopic dermatitis (eczema) spectrum. It can be associated with underlying, secondarily scratched and thickened psoriasis or contact dermatitis or the end stage of several itchy vulvar conditions (e.g. LS). Scratching “feels good” especially for patients with atopic dermatitis (patients with a background of allergies, eczema, hay fever or asthma). Stress makes all of this worse.

**Causes of LSC:**

**Infection:** Candida and dermatophytosis

**Dermatoses:**
- Atopic dermatitis
- Lichen Sclerosus
- Lichen Planus
- Psoriasis
- Contact Dermatitis

**Metabolic:** Diabetes and iron deficiency anemia

**Neoplasia:** Vulvar intraepithelial neoplasia

The most important causes are atopic dermatitis, contact dermatitis or both.
Less common causes – psoriasis, LS
Pathophysiology – in this condition there is an altered skin barrier with varying combination of allergens, irritants and skin pathogens that result in a changed immunoregulatory process. Stress further alters the skin barrier function, making all of this worse. This condition is defined by relentless pruritus. These patients scratch in their sleep ruining the effectiveness of their daytime treatments. The chronic scratching causes the skin to thicken and feel firm.

Clinical Presentation:
- Relentless pruritus
- Chronic – years of “chronic itch”
- Worse with heat, stress, menstruation
- “Nothing helps”
- Marked lichenification
- Pigmentation changes
- Unilateral or bilateral
- Hair loss from scratching
- Excoriations + crusts
- Diagnosis – clinical biopsy may be needed

Note: Scratching makes erosions with serosanguineous crusts; repeated rubbing causes skin thickening (lichenification). In LSC, you can see both erosions and lichenification.

Treatment:
- Rule out other conditions
- Stop all irritants
- Consider Patch testing looking for an allergen
- Stop itch/scratch/itch cycles
- Topical superpotent steroids, halobetasol or clobetasol 0.05% ointment, bid for two weeks, qhs for two weeks, then M-W-F for two weeks.
  (For severe disease, a longer duration of a mid dose topical steroid may be required.)
- Oral steroids may be required for a short duration (dose varies dependent on disease severity; consider prednisone 40 mg po q am x 5, then 20 mg po q am x 10, however a longer taper may be required)
- IM triamcinolone 1 mg/kg (up to 80 mg total) can be used instead of prednisone for severe, itchy or extensive LSC. Repeat is seldom necessary. If repeat is necessary, it can be repeated monthly x 3 total doses.
- Intralesimal triamcinolone can be used to thin the thick / lichenified skin as for LS above.
- Treat infections, bacterial and yeast
  - Cefadroxil 500 mg bid for 7 days
  - Fluconazole 150 mg po q week x 2
  - Doxepin or hydroxyzine 10 to 75 mg qhs for nighttime itching
  - Citalopram or fluoxetine or sertraline in the morning for daytime itching
  - Amitriptyline is also used at times for sedation (25 mg po qhs; can increase to 50 mg po qhs) in patients with severe itch scratch cycle. It puts the patient in a deeper sleep cycle than the other sedation agents listed above. Do not combine amitriptyline with the other sedation agents above. Give early in evening so not sleepy in morning (6-8PM). Caution for use in the elderly population. Check for other drug interactions.
- Sitz baths or cold soaks
- White cotton gloves at night

Note: If skin is very raw the topical steroids will burn. Start with plain Vaseline,
oral antibiotics, anti-yeast medication and nighttime sedation for 2-3 days, then start the topicals.
LSC reoccurs due to sensitive skin in the area so it will need repeated management.

LOOK FOR MORE THAN ONE CAUSE OR A COMBINATION OF CAUSES as it is not uncommon to have psoriasis, contact dermatitis and lichen simplex chronicus in the same patient.

**LICHEN PLANUS (LP)**
Lichen planus is an autoimmune, mucocutaneous disorder of altered cell mediated immunity in older women affecting the skin and mucous membranes.

Etiology: It is a disorder of altered cell mediated immunity with exogenous antigens targeting the epidermis.

The diagnosis is often missed on the vulva and in the vagina.
It tends to occur in menopausal women (age 40-60 years).
It affects skin and mucous membrane – mouth, vulva, vagina, nails, scalp, esophagus, nose, conjunctiva of the eye, ears and bladder.
Painful LP is usually erosive; patient can have LP plus chronic vulvar pain.

Clinical Presentation:
1. Papulosquamous – typical papules and plaques with white lacy pattern on the vulvar trigone and periclitoral area. It may be part of generalized LP. This can be itchy. It tends to respond to topical steroids.
2. Hypertrophic – least common with extensive white scarring and destruction (looks like LS)
   – can be very itchy. Treatment tends to be resistant.
3. Erosive (vulvovaginal gingival syndrome) – destructive, scarred lichen planus on the mucous membranes and vulva with a desquamative vaginitis, variable erosions plus atrophy, usually pain, burning and irritation rather than itch. The skin of the vulva often has a glazed erythema. Treatment tends to be resistant.

Note – LP involves the vulva and vagina, It may only be in the vagina.

Erosive LP (vulvovaginal gingival syndrome)
Symptoms:
- Severe pain and burning
- Dysuria
- Depression + anger
- Dyspareunia / apareunia

Signs – painful, glossy red erosions (glazed erythema) and scarring are seen around the labia minora and vestibule. The borders may be white to smudgy or smoky gray. The scarring causes flattening of the vulva and loss of the labia minora.
- May see desquamative inflammatory vaginitis
  - Vaginitis with vaginal erosions, atrophy, purulent malodorous discharge, vaginal synechiae and scarring. The vagina may be obliterated.

Note: up to 70% of women with vulvar LP have vaginal involvement.
This can be a chronic, destructive, debilitating and difficult condition.
The vagina may be involved alone.

Diagnosis:  Look at mouth and skin for evidence of LP
Consider biopsy for H&E and immunofluorescence
Biopsies may be nonspecific

Differential diagnosis:  Lichen sclerosus, drug eruption, cicatricial pemphigoid, graft vs.
host disease

Treatment:
Stop irritants                           Pain control
Bland therapy for ulcers               Sedation
Superpotent steroid ointment (clobetasol) topically once to twice a day.
Intralesional steroid – triamcinolone 3.3 up to 10 mg/ml q 3-4 wks x 3 (do not give high
dose in small area-erosions and ulcers may occur)

Intravaginal steroid – hydrocortisone acetate foam 40-80 mg qhs
or 25 to 200 mg compounded suppository qhs (if using high dose steroids, use for
short term use, then gradually decrease the dose).
If severe – hydrocortisone acetate 10% compounded in a Replens like base –3 to 5
grams (300 mg to 500mg/dose) nightly for 14 days then 3 nights a week and
continue to decrease dose as per response. (Some prefer to use every other night
initially, and then gradually decrease the dose)
Note: adrenal suppression and risk of candidiasis

IM Triamcinolone (Kenalog 40) 1 mg/kg every 4 weeks for 3 doses. (Dose up to a
maximum of 80 mg total per dose) Repeat monthly for up to 3 months. Max 4 doses per
year
Prednisone 30-60 mg a day with taper
Methotrexate 7.5-15 mg po or subcutaneously in abdomen or thigh, once a week with
folic 1 mg daily
Mycophenolate mofetil 250 mg/day building up to 3gm/day (pregnancy must be
prevented)
Acitretin 10 mg 3-7 days a week with fatty food for erosive disease. Counsel on no
pregnancy as this is a teratogen. (see above for lichen sclerosus)
Cyclosporine 3-4 mg / kg per day

Patient education and support needed
Dilators
Surgery for scarring followed by intravaginal treatment

Other Treatments:
- Clobetasol propionate 0.05% ointment virginally using 1-2 grams nightly via a
  “Premarin type applicator”
- Clobetasol propionate 0.05% ointment/Nystatin 100,000 units/gram/3% oxy-tetracycline
  in cream base
- pimecrolimus (Elidel) 1% cream bid for mild LP
- Topical tacrolimus (Protopic) 0.03 or 0.1% ointment (burns) as a steroid sparer
- Hydroxychloroquine, etanercept (see below)

Course: uncertain - often very chronic-10% resolve, 50% asymptomatic and 15% do poorly
What are the various treatments for Lichen Planus?
Papular lichen planus tends to respond to topical corticosteroids. Triamcinolone acetonide 0.1% ointment for mild disease and clobetasol propionate 0.05% ointment for severe disease.

For erosive disease the following table contains many medications that have been tried for LP treatment. It is important to note that many of these medications are formulated for off label use.

<table>
<thead>
<tr>
<th>Agent</th>
<th>Discussion</th>
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</table>
| **Long term Anti-inflammatory antibiotics** | This treatment works best for early erosive lichen planus  
Doxycycline or clindamycin used long-term. Consider adding weekly fluconazole to prevent yeast infection. |
| **Steroids are often used for lichen planus** | Vaginal LP  
Anusol HC 25 mg vaginal suppositories are used in the following manner:  
1/2 of a Anusol HC suppository per vagina twice daily for 2 months, then daily for 2 months, then maintenance treatment at 1 to 3 times per week. However, many patients do not experience significant long-term response to intravaginal steroids. The vaginal vault tends to continue to scar. To keep the vault open and prevent adhesions it often will be necessary to use vaginal dilators. The dilator may be lubricated with a hydrocortisone cream.  
At times a stronger steroid may be required for vulvar LP (see text).  
Topical- Clobetasol propionate (Temovate®) 0.05% ointment  
Intralesional- triamcinolone acetonide 5-10 mg/ml  
As above, for stronger treatment:  
– hydrocortisone acetate foam 40-80 mg qhs  
or 25 to 200 mg suppository qhs (if using high dose steroids, use for short term use, then gradually decrease the dose).  
If severe – hydrocortisone acetate 10% compounded in a Replens like base – 3 to 5 grams (300 mg to 500mg/dose) nightly for 14 days then 3 nights a week and continue to decrease dose as per response. (Some prefer to use every other night initially, then gradually decrease the dose)  
Oral- Oral prednisone may be required until healing has occurred. 30-40 mg qam with food for 3 weeks then slowly taper. As the skin heals, topical corticosteroids may be added as the prednisone is tapered.  
IM steroids (place into muscle in anterior thigh). Used for moderate disease. Dose 1 mg/kg (not to exceed 80 mg) every 4 weeks to every 8 weeks for up to 3 or 4 months.  
For Oral LP- Apply Clobetasol propionate (Temovate®) gel or ointment 0.05% to affected area up to qid  
Apply on a cotton ball in mouth for 5 min. Best to use in a dental tray for 15-30 min bid for gums. Some providers use dental molds to hold in medications in patients with gingival LP |
| Tacrolimus and Pimecrolimus | Tacrolimus (Protopic) 0.1% ointment bid to qid.  
Apply on a cotton ball in mouth for 5 min  
Vaginal medication (made by compounding pharmacy)  
tacrolimus vaginal suppositories  
Insert one suppository per vagina (2 mg tacrolimus per 2 gram supp) qhs  
Disp 50  
Or 0.1% vaginal cream (compounded in a vaginal cream / Replens like base) 2-5 g = 2 - 5 mg/dose for 2 weeks then Mon-Wed-Fri for 2 weeks and slowly decrease Disp 100 grams  
Vulvar medication Apply to skin bid  
Tacrolimus 0.1% ointment Available in 30 or 60 gram tubes |
|-----------------------------|---------------------------------------------------------------------------------|
| **Calcineurin inhibitors (steroid sparing)** | pimecrolimus (Elidel) 1% cream bid for mild LP  
topical tacrolimus (Protopic) 0.03%, 0.1% oint |
| Note – can burn especially on raw areas  
Long term safety unknown |
<table>
<thead>
<tr>
<th>Less frequently used medications</th>
<th></th>
</tr>
</thead>
<tbody>
<tr>
<td><strong>Hydroxychloroquine</strong> <em>(Plaquenil)</em></td>
<td>Occasionally used. Dose is 200 mg po bid.</td>
</tr>
<tr>
<td><strong>Retinoids</strong></td>
<td>There is no documented successful use of retinoids for vulvovaginal lichen planus. There is only personal experience with Acitretin <em>(Soriatane)</em> . It can work well in low dose 30-70 mg/week. <em>(Isotretinoin has been used to treat oral lichen planus; however, discontinuation of the medication results in recurrence of the oral lesions.)</em> Long-term use of retinoids may result in liver dysfunction, but not in the small doses recommended here. Liver function tests, cholesterol, triglycerides and complete blood cell counts should be monitored since laboratory changes are associated with the use of oral retinoids. Patients should be counseled concerning teratogenicity and need for optimal contraception. Acitretin is a strong teratogen that remains in the body for at least three months after the last dose. Topical retinoids <em>(Tazarotene (tazarac)</em> are often too irritating for this vulvar condition but have been used.</td>
</tr>
<tr>
<td><strong>Cyclosporine</strong></td>
<td>Used topically and systemically. Topical cyclosporine provides a safe and often effective but very expensive alternative for mucous membrane disease. Pelisse et al. described the use of the oral or injectable form of the medication in 100 mg amounts directly to the affected skin four times a day initially. If several mucous membranes were affected for example, 100 mg was applied to the vulva, 100 mg inserted into the vagina, and 100 mg held in the mouth for as long as tolerated before spitting. As disease is controlled, the frequency of application can be tapered. Systemically it is dosed at 4-5 mg/kg/day for 3 months <em>(used in severe disease)</em>. Occasionally, in patients with debilitating and painful disease not adequately treated by therapies discussed above, oral cyclosporine may be used. This medication should be used only by health care providers experienced in its use.</td>
</tr>
<tr>
<td><strong>Cyclophosphamide</strong></td>
<td>Systemic antimetabolite</td>
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<tr>
<td><strong>Azathioprine</strong></td>
<td>Systemic antimetabolite</td>
</tr>
<tr>
<td><strong>Etanercept <em>(Enbrel)</em></strong></td>
<td>This is used SQ <em>(50 mg sq 2x/week until symptoms improve, then 25 mg sq 2x/week)</em></td>
</tr>
<tr>
<td><strong>Mycophenolate mofetil <em>(CellCept)</em></strong></td>
<td>Oral use 250mg - 3 g/d in divided dose</td>
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<tr>
<td><strong>Methotrexate</strong></td>
<td>Oral or subcutaneous injection weekly. 7.5 to 15 mg oral or subcutaneously weekly using a 27 or 30 gauge needle. Need to give folate with this medication- 1 mg/d</td>
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Lichen Planus and Surgery

For scarred LP of the vagina - post surgery information

I. For dilation:

Dilation is vital to keep the vagina open in patients with vaginal lichen planus. Patients need specific instructions on size of dilator and how to use dilators. They may need a set of dilators and can to buy the dilator set from www.vaginismus.com. Start with the largest size that will fit, determined by surgery. Leave the dilator in once or twice a day for 15-20 minutes. For lubricating the dilator use either Vaseline or mineral oil. Hydrocortisone acetate cream or Estrace 0.01% vaginal cream can be used later.

II. To stop inflammation:

If not too severe 2-3 days preoperatively use prednisone 15-30 mg/d AM, with food, plus topical steroid. Keep on prednisone for 1 week post operatively then taper slowly at 5 mg/week. Use with the topical steroid (see below).

For more severe disease consider using a dose of intramuscular triamcinolone 1mg/kg up to a total of 80mg/dose to be given two days after surgery and repeat this monthly for up to three months. Follow and assess her to see if she is going to need other long-term systemic medication, cyclosporine, mycophenolate, methotrexate, etc. Once she is healed she may need a systemic anti-inflammatory. The medication will depend on the case. These medications can be used with intermittent doses of IM triamcinolone, also depending on the case.

A. For the vagina

Two days after surgery, when the stent is removed, the patient needs to start dilating with Vaseline on the dilator twice a day. Dilators must be used nightly. In 1 to 2 weeks if healing then consider 10% hydrocortisone acetate in a vaginal cream 300mg (3g) to 500 mg (5gms) nightly for a week then gradually decrease weekly to 1-3gram Mon-Wed-Fri depending on response. (The compounded prescription is 10% hydrocortisone acetate in vaginal cream base 100 g with 2 refills). As a steroid sparer consider tacrolimus 2 mg compounded suppository nightly, or 0.1% tacrolimus compounded vaginal cream 2 grams/dose. Note – tacrolimus can cause a burning sensation. Use fluconazole 150 mg weekly to prevent yeast as needed.

B. For the vulva - to start two days after surgery, if not very eroded, topical clobetasol 0.05% ointment in a thin film PM, If eroded use plain Vaseline for 2 weeks and then restart clobetasol. If tolerated consider using tacrolimus 0.1% ointment twice a day as a steroid sparer note – as above, it can cause a burning sensation.

III Follow up- patient needs to be seen often for support and to adjust treatment. Avoid sexual intercourse until well healed with adequate size.
**Atrophic Vulvovaginitis**

Postmenopausal women not on estrogen replacement experience thinning of the vulvar and vaginal epithelium. They may also have thinning of the pubic hair and smoothness and thinning of the vulvar skin. The labia minora and majora lose substance and become more wrinkled; complete resorption of the labia minora occurs in some and may mimic the end stage of lichen sclerosus. Patients may be asymptomatic, but many are aware of a sensation of dryness that sometimes makes intercourse uncomfortable. Some patients complain of dysuria, urgency, and frequency as a result of atrophic urethritis. The diagnosis of atrophic vulvovaginitis is by clinical examination and a history of estrogen deficiency. Vulvovaginal atrophy from lack of estrogen can be seen with use of BCP, Depoprovera, nursing etc. Atrophic vaginitis is suspected when parabasal cells and inflammatory cells are seen on wet prep in a symptomatic patient. Atrophic vulvovaginitis complicates all vulvovaginal conditions. Without estrogen the barrier functions are weaker and the tissues more susceptible to irritation from day to day hygiene practices, sexual activity etc. This can be further compounded by an already disrupted barrier with lichen sclerosus, lichen planus, even VIN. Estrogen topically and, if appropriate, systemically can make a big difference.

**CONTACT DERMATITIS**

Contact dermatitis is an inflammation of the skin resulting from an external agent that acts as an irritant or allergen. This reaction may be acute, subacute or chronic.

**Primary irritant contact** dermatitis results from prolonged or repeated exposure to a caustic or physically irritating agent. (e.g. urine, feces, soap residue) Anyone exposed to such a product often enough will have a reaction. This is a non-immunologic reaction. The skin is directly damaged. Top three causes –

1. Over-washing (some patients become obsessed with cleanliness and wash the area with soap and water multiple times each day, causing irritation. Some may become fixated on symptoms and even use harsh cleansers. Patients may remain secretive and not report these habits.)
2. Use of creams with drying bases
3. Wetness (urine, feces, menstruation)

**Allergic contact dermatitis** results from a frank allergic reaction, to a low dose of a substance (e.g. poison ivy, neomycin or benzocaine). This is a type IV delayed hypersensitivity reaction. Top three causes – Neomycin, benzocaine and preservatives.

**Note:** Irritant contact dermatitis is immediate; allergy takes 1-2 days.

**Clinical Presentation:** The same for both types of reactions

Varying degree of itch, burning and irritation; can be acute or chronic. With an irritant there is a history of repeated exposure, e.g. repeated use of soaps, cleansers, chronic incontinence. Allergic contact dermatitis can be more acute with sudden onset of symptoms of itching and burning that can be more intense. On physical exam there can be an acute blistered erosive eruption but most of the time there are subacute or chronic changes with evidence of excoriating, honey colored crusting (with or without secondary infection) or just dryness, scaling and erythema. There may be altered pigmentation.
Diagnosis: Morphology of rash plus history of an irritant substance or an allergen. Biopsy may be needed to sort this out. To define allergic etiology, patch testing must be set up by a dermatologist or allergist.

TIPS ON VULVAR CONTACT DERMATITIS
1. Irritant contact dermatitis of the vulva is common. Factors that promote vulvar irritation with disruption of barrier function are:
   a. Lack of estrogen that causes the epidermal barrier to be weakened/thinned and less moist and pliable. The result if cracking/fissuring, etc.
   b. Overzealous hygiene with excessive washing with a washcloth or sponge using caustic soaps results in dry cracked and burned skin. Beware of the “dirty” vulva. Women are convinced that the area is dirty and needs to be scrubbed.
   c. Excess maceration of the area from:
      Sweat, urine, wet pads of any type results in irritation
      Incontinence is a hidden epidemic
      Note – urine and feces burn enzymatically and/or chemically
   d. Existing dermatoses, infection or tumors, e.g. lichen sclerosus, lichen planus, candidiasis are susceptible to irritants.
2. History of contactants may be difficult to elicit
3. Always stop all unnecessary vulvar contactants
4. Suspect allergic contact dermatitis with a sudden onset of intense itching and/or vesiculation and weeping
5. Always set up patch testing to rule out possible common allergens for patients with chronic or recurrent, poorly responsive vulvar dermatoses. Work with a dermatologist or allergist who can do the patch testing. The best screen is the North American Patch Test series (about 60 or more allergens) not the True Test Series as it may test for too few allergens – 25 to 35.
6. Reassess you vulvar patients for contact dermatitis as women commonly self treat themselves to “wash away” or “clean up” their itchy or burning vulva. Contact dermatitis can complicate all vulvar conditions.

Treatment:
- Stop the irritant or allergen exposure
- Topical corticosteroids – clobetasol 0.05% or halobetasol 0.05% ointment bid x 5-7 days, then daily x 5-7 days (avoid long term use)
- Bland emollients such as petrolatum or mineral oil and nighttime use sedation for sleeping
- Antibiotics are needed for secondary infection – see lichen simplex chronicus above
- If very severe, prednisone 1 mg/kg decreasing over 14 – 21 days or 1 dose of triamcinolone acetonide IM 1 mg/kg (anterior thigh) (do not exceed 80 mg total IM)
Caution in patients with diabetes- high dose steroids can interfere with their glucose control.

Common Vulvar Irritants:
- Soaps/cleansers
- Medications -Trichloroacetic acid, 5FU
- Sweat, urine, feces
- Douches
- Spermicides
- Panty liners
Common Vulvar Allergens:
- Benzocaine (Vagisil)
- Neomycin (Neosporin)
- Chlorhexidine (KY jelly)
- Perfume
- Some wipes and paper products contain the preservative methylchloroisothiazolinone/methylisothiazolinone and this can cause an allergic contact dermatitis.

Crohn’s Disease Disease
Crohn’s disease is a chronic inflammatory bowel disease. It is an autoimmune disorder affecting the gastrointestinal tract from mouth to the anus. It affects over 300,000 women in North America. The onset for women is between 15 and 30 years of age. It is diagnosed by biopsy of the skin or the GI tract that shows a diffuse lymphohistiocytic infiltration with loose non-caseating granulomas. These granulomas are considered the hallmark of Crohn’s disease but they are found in only 20-60% of biopsies of Crohn’s patients, whether in bowel or skin.

The most common symptoms of Crohn’s disease are abdominal pain, cramping and diarrhea, often following a meal. There can be rectal bleeding, weight loss, joint pains and fever. Anemia is not uncommon. Patients often develop sores in the anal area and sometimes fistulae.

It is reported to be rare in the vulva though the prevalence is not known. There have been 101 reports on vulvar involvement since 1965 and recently these were summarized by Barrett et al, in Crohn’s Disease of the Vulva, J Crohn’s Colitis (2013). Vulvar Crohn’s disease is still considered rare with the common features of labial swelling, vulvar ulcers and hypertrophic lesions.

Patterns of Crohn’s disease on the vulva:

Specific:
1. Contiguous – fistulae, abscesses and ulcers, usually perianal fistulae, rarely rectovaginal fistulae.
2. Metastatic Crohn’s disease (MCD) on the vulva causes 90% of vulvar lesions. This represents a granulomatous inflammation of the vulvar skin with swelling and induration of the labia unilateral or bilateral and can have any of the following:
   a. Classic “knife cut ulcers”, linear ulcers, that can be fissures with linear ulcers in any of the creases of the perineum or perianal area such as inguinal sulci, periclitorally, perineum, and/or gluteal cleft. These can be associated with scattered ulcers, edema of the skin, drainage and pain (note only 38% show granulomas on biopsy)
   b. Swelling, and edema of labia majora, labia minor that is unilateral or bilateral-this can be associated with lymphangiectasia and there can be frank lymphangiectatic cobbling of the skin
   c. Perianal skin tags – these are often the harbinger of Crohn’s disease in 40-70% of cases. They can look often like hemorrhoids. These are classic and found in most cases of Crohn’s disease.

Reactive:
1. Aphthae- these ulcers can be genital and/or oral, single or multiple. These can be associated with “knife cut ulcers”. These can be totally asymptomatic or tender.
2. Suppurative lesions- hidradenitis suppurativa (HS) is associated with Crohn’s disease in about 17% of HS patients.

3. Extra-intestinal manifestation of Crohn’s disease include
   - Arthritis - Spondyloarthropathies
   - Ocular—conjunctivitis, uveitis, episcleritis
   - Hepatobiliary- primary sclerosing cholangitis
   - Skin – Erythema nodosum
   - Pyoderma gangrenosum – in 1% CD
   - Cheilitis, oral swelling- oral disease can be found in 8% of patients with cheilitis, cobblestoning of the buccal mucosa,
   - Lip swelling
   - Psoriasis
   - Vasculitis usually on lower legs
   - Epidermolysis bullosa acquisita

The anal area is often involved with:
   - Perianal abscesses and fistulae
   - Fissures in 25-35%
   - Fistulae 6 -35%
   - Ulcers 5%- these can be very large
   - Skin tags – these anal tags are due to lymphedema. They can be:
     a. Large hard tag-like lesions that develop in healed anal fistula ulcers
     b. The “elephant ear” type which are described as broad, soft and sometimes can resolve

Note: 25% of patients present with vulvar manifestations of Crohn’s disease before developing GI disease. The GI disease may not show up for many years.

Think of possible Crohn’s disease with the following vulvar lesions:
1) Vulvar swelling/edema, lymphedema with lymphangiectasia. The labia may be hypertrophic and pseudocondylomata can be very dramatic. This is due to granulomatous infiltration and impaired lymphatic drainage due to chronic inflammation from the Crohn’s disease. Recurrent cellulitis results in lymphatic vessel destruction and obstruction and more swelling
2) Ulcerations- knife cut ulcers, aphthous ulcers
3) Suppuration with hidradenitis suppurativa-type lesions
4) Perianal disease with swelling, fissures, anal and perineal tags

Diagnostic Workup
1) Biopsy – skin, bowel (GI workup needed always)
2) Consider differential diagnosis ruling out infectious diseases - Candida albicans, bacterial vaginosis and trichomoniasis. Note: Crohn’s disease can be associated with a desquamative inflammatory vaginitis (personal observation).
3) Rule out other
   a. Infections: lymphogranulomatosis, tuberculosis, syphilis, HSV in an immunosuppressed patient, HIV, rare causes of infectious ulcer on section on vulvar ulcers.
   b. Inflammatory conditions such as sarcoidosis, hidradenitis suppurativa, foreign body reaction, contact dermatitis. Rule out infiltrative conditions e.g. squamous cell Ca
c. Causes of chronic lymphedema: See section on lymphedema.
d. Causes of vulvar ulcers: See section on vulvar ulcers. Note: Granuloma inguinale and Langerhans cell histiocytosis both cause linear ulcers. Specific investigations will depend on screening for appropriate conditions.

Treatment
Most important aim is to control the bowel disease
1) First line treatment usually is systemic corticosteroids. Corticosteroids are the cornerstone of treatment but are not always well tolerated. Prednisone may be combined with metronidazole or azathioprine. Intralesional triamcinolone 10 mg/mL can be helpful (personal experience).
2) Further treatment can include azathioprine, methotrexate or mercaptopurine.
3) In more severe disease the usual treatment is with infliximab or adalimumab or, less commonly, certolizumab pegol or ustekinumab. Combination therapy is common.
4) In some patients surgery is necessary to debulk the significant edema and lymphangiectasia but this is not curative. Surgery should be considered especially if there are strctural complications or difficult draining lesions.
5) For local treatment for limited disease - superpotent steroids with clobetasol 0.05% ointment or halobetasol 0.05% ointment can be used for short periods of time for two weeks. Patients can be switched to the calcineurin inhibitor tacrolimus (Protopic®) 0.1% ointment twice a day if there is no burning. For thick perianal tags triamcinolone 5 to 10 mg/mL can be injected every three to four weeks. (See section on edema below).

HIDRADENITIS SUPPURATIVA
Definition – Hidradenitis suppurativa is a chronic follicular occlusive disease, characterized by recurrent painful, deep-seated nodules and abscesses located primarily in the axillae, groins, perianal, perineal and inframammary regions. The Second International HS Research Symposium (San Francisco March 2009) adopted the following consensus definition. “HS is a chronic, inflammatory, recurrent, debilitating, skin follicular disease that usually presents after puberty with painful deep seated, inflamed lesions in the apocrine gland-bearing areas of the body, most commonly the axilla, inguinal and anogenital region”. HS is frequently misdiagnosed as “boils”. This results in delayed diagnosis, fragmented care, and progression to a chronic, disabling condition that has a profoundly negative impact on quality of life.

The prevalence of hidradenitis suppurativa (HS) is 1 to 4%. Women are more commonly affected than men. Some studies have described a predilection in patients of afro-carib descent, but this has not been confirmed in all. 25% of patients present between the ages of 15 and 20 and 53% are aged 21 to 30. Female to male ratios range from 2:1 to 5:1. Prepubertal cases are rare, but occasional onset in neonates and infants has been described.

HS/AI has been erroneously linked to the apocrine sweat glands. The first pathogenic change is in the follicular portion of the folliculopilosebaceous unit (FPSU).
HS/AI is characterized by recurrent inflamed deep-seated acneform nodules that result in abscesses and chronic draining sinus tract formation leading to scarring, disfigurement and life-altering disability. The lesions classically occur in areas of the skin that host folliculopilosebaceous units. HS/AI is frequently misdiagnosed as “boils”, resulting in delayed diagnosis, fragmented care, and progression to a chronic, disabling condition.

**Diagnosis**—Relies on the following diagnostic criteria:

1. Typical lesions: either deep-seated painful nodules (blind boils) in early primary lesions or abscesses, draining sinuses, bridged scars and “tombstone” open comedones in secondary lesions.
2. Typical topography: axillae, groin, genitals, perineal and perianal region, buttocks, infra- and inter-mammary folds.
3. Chronicity and recurrences.

These three criteria must be met to establish the diagnosis.

Multiple skin abscesses occur, with draining subcutaneous sinus tracts. Scarring and deformity are present in many individuals. Although biopsy is not absolutely required for diagnosis of HS, if you send tissue to pathology and tell them that the clinical picture is consistent with HS, they will likely look for the characteristic findings of follicular hyperkeratosis, active folliculitis or abscess, sinus tract formation, fibrosis, granuloma formation, apocrine and eccrine stasis and inflammation, fibrosis, fat necrosis, inflammation of the subcutis.

The basic problem is that people with HS have genetically ‘weak pores’ that rupture easily. New histologic findings show that the connective tissue wrap around the follicular tube is weak to nonexistent at the point where the sebaceous glands attach to the follicle.

This defect leads to the following sequence of events:

1. The problem starts with innate and exogenous androgens acting on the follicle duct lining cells so that they build up and occlude the ducts. It is hypothesized that dietary factors that elevate insulin and insulin-like growth factor-1 sensitize the FPSU’s androgen receptors, creating the increase in end organ responsiveness that also leads to follicular occlusion.
2. The follicular duct content expands as keratinocytes accumulate and the wall of the follicle eventually ruptures due to the weakness in the follicle support. A number of genetic defects may play a role here.
3. Follicular rupture results in the release of numerous inflammatory stimuli and antigens, including keratin fragments, that trigger even more numerous elements of the innate and adaptive immune systems, leading to the development of an acute inflammatory response in the surrounding tissue. Extensive research has been done on the acute and chronic phase cellular and cytokine reactants in an effort to focus treatment appropriately for more effective therapy.
4. Attempted healing creates chronic inflammation and results in chronic tissue destruction through a foreign body-like reaction and subsequent resolution by scarring.
5. Mechanical factors can be important because any friction or shearing forces, from tight clothing to pinching the area can make it worse. Obesity with resulting sweating, maceration and friction can make things worse. Exogenous androgens such as progestins and drugs like lithium can also make things worse. Smoking is strongly associated with HS. It promotes follicular plugging in HS as it does in acne. High glycemic load diets, milk and milk products contribute to androgen sensitivity.

6. When the pores rupture, follicular stem cells can be released into the subcutis where they appear to trigger the formation of cysts and sinuses. An invasive proliferative gelatinous mass (IPGM) is produced in most cases, consisting of a gel in which are embedded both inflammatory cells and, it is postulated, the precursors of the epithelialized elements described above. Continuous growth of these hormonally stimulated remnants beneath the surface perpetuates the communicating sinuses and inflammatory mass and provides increasing volumes of invading material. The inflammation in the dermis and subcutis will not settle until this material is eliminated.

In summary - genetically weak-walled pores, distended under the influence of hormones and subject to friction and pressure, rupture and create painful inflammatory subcutaneous nodules.

Etiology
The development of HS/AI depends upon a combination of factors.

Genetic factors
A 35-40% positive family history may reflect inadequate family reporting. An autosomal dominant inheritance pattern has been noted, but no specifically genetic defect has been found. Von der Werth suggests that HS/AI is most likely a heterogeneous disease, probably with several genes involved.

Infection
Bacteria have long been considered in the pathogenesis of HS/AI. It is generally agreed that bacteria do not have a major direct role in the etiology of HS/AI but, as secondary invaders, may share in the pathogenesis of the chronic relapsing lesions causing some of the destructive processes that are seen. Septicemia and systemic illness in this disorder are exceptionally rare.

Hormonal factors
A strong relationship exists between sex hormones and HS/AI. The female preponderance suggests a greater sensitivity of females to androgens. There are no elevations in serum androgens in the vast majority of HS/AI patients. End organ sensitivity is likely responsible. Increased access to the androgen receptor is mediated by insulin and insulin-like growth factor-1 (IGF-1), both chronically raised by dietary factors.

In women, HS/AI onsets around menarche, flares premenstrually and following exposure to androgenic progestins like medroxyprogesterone acetate or levonorgestrel, but improves with pregnancy and fades after menopause.
Anti-androgen therapy helps HS/AI patients of both sexes. Finasteride, a selective inhibitor of the type II isomer of 5α-reductase, reduces levels of 5α-DHT. It was used to improve six of seven adults with HS/AI and three children, one with premature adrenarche and one with polycystic ovarian syndrome.

**Immune factors**

The disease does not usually produce acute systemic inflammatory effects. There is no fever, rare lymphadenopathy, no septicemia, occasional local cellulitis, cultures are usually sterile and, if the offending material beneath the surface is removed, the disease heals without further difficulty and without antibiotics. This is strongly suggestive of inflammation mediated on the local level by the innate immune system. Consider a simple ingrown hair. Flick out the ingrown hair and the inflammation fades.

The immune systems accelerate the disorder. Pathologic examination of excised early lesions demonstrates a wide variety of immune responses involving the innate and acquired (adaptive) immune systems. A vast catalogue of T-lymphocytes and cytokines are assembled. Unfortunately, cooling the inflammation does not cure the disease.

**Mechanical Factors**

Weakness in the support structure of the follicular portion of the FPSU likely predisposes to follicular rupture caused by local trauma. Patients worsen their lesions by pinching them. Obesity contributes to these increases in pressure and shear forces, but more important is the relationship of obesity to dietary habits that raise plasma glucose and insulin levels. This sensitizes the androgen receptors, increases the plugging of pores, causes insulin resistance and enhances obesity. HS/AI affects thin people but overweight patients have more severe disease.

**Smoking**

Smoking is strongly associated with HS/AI; smokers are generally more severely affected than non-smokers. Nicotine promotes follicular plugging.

**Diet**

The androgen receptors that control growth are normally closed to circulating androgens. Elevated insulin (from the combination of high glycemic carbohydrate load and dairy whey) and IGF-1 (induced by casein in milk) open these receptors and expose them to circulating androgens. Androgens from any source can then access previously inaccessible androgen receptors. Stimulation of follicular androgen receptors results in ductal keratinocyte overproduction and retention hyperkeratosis. Androgen sources include the adrenals, ovaries and testes, molecular precursors in dairy products, the androgenic progestins in birth control pills, the levonorgestrel-containing IUD, intramuscular medroxyprogesterone acetate (MPA) injections and contraceptive implants.

**Drugs**

Hidradenitis suppurativa can be triggered or flared by lithium and androgens in BCPs, even IUDs.
**Differential diagnosis** – Multiple conditions are to be considered in the differential diagnosis of hidradenitis suppurativa.

**Infections**
- Bacterial - Carbuncles, furuncles, abscesses, ischiorectal/perirectal abscess, Bartholin’s duct abscess
  - Mycobacteria – TB
  - STI– granuloma inguinale, lymphogranuloma venereum, syphilis
  - Deep fungi – blastomyces, nocardia

**Tumors**
- Cysts – epidermoid, Bartholin’s, pilonidal

**Miscellaneous**
- Crohn’s, anal or vulvovaginal fistulae

**Clinical features** – Early/primary lesions are a single, painful, deep-seated nodule 0.5-2cm, round, no “pointing” that may resolve, persist as a “silent” nodule that can recur, or abscess and drain and recur even if surgically drained. With time these can go on to chronic, recurrent lesions at same site, coalescing with fibrosis and sinus formation. Lesions persist for months with pain and drainage with foul odor. These can result in tertiary lesions with hypertrophic fibrous scarring with “bridged scars” forming rope-like bands with active, painful, inflammatory nodules and sinus tracts forming thick plaques over an area. Thick scarred areas can result in decreased mobility and lymphedema.

  - Lesion course – most form an abscess, rupture and drain purulent material then may resolve and/or recur, form a chronic sinus that can drain with a seropurulent and/or bloody discharge, ulcerate, burrow and rupture into nearby lesions.
  - Mean age of onset is 22 years old and it lasts on average 19 years but can remit or partially remit with pregnancy and breast feeding. This all can be variable. Each new painful lesion lasts 10-30 days. Flaring with menses is common.

**TREATMENT PRINCIPLES**

**Therapy and prognosis** – Planning treatment follows severity grading. The first two stages respond to medical treatment whereas the third stage requires biologics and surgery. All patients will need thorough education and constant reassurance and support.

**Treatment**
- Define the frequency of the flares and the intensity of the pain when deciding upon treatment.
- A permanent cure is achieved only with wide, thorough, surgical excision
- Combine medical and surgical treatment
Goals of treatment of hidradenitis:
1. To reduce the extent and progression of the disease to bring it to a milder stage
2. To heal existing lesions and prevent new ones from forming
3. To allow regression of scars and sinuses in cases of extensive hidradenitis suppurativa

Hurley’s criteria for Hidradenitis Suppurativa Staging
Hurley’s criteria for Hidradenitis Suppurativa Staging – used to assess severity

Treatment principles – choose treatment to fit disease severity staging

Stage I: Abscess formation, single or multiple without sinus tracts and cicatrisation/scarring.
Stage II: Recurrent abscesses with sinus tracts and scarring. Single or multiple widely separated lesions
Stage III: Diffuse or almost diffuse involvement or multiple interconnected tracts and abscess

70% stay in Stage I
28% progress to Stage II
4% progress to Stage III

General Hidradenitis Suppurativa Treatment

There is no single effective treatment or cure for HS/AI. The only permanent cure has been reported with wide surgery for very severe HS/AI (Hurley's III). Patients require metabolic, medical and surgical strategies and lifelong gentle atraumatic care.

Education, diet and support
Improve environment:
Reduce all trauma, friction in the area, heat, sweating and obesity
Loose clothing, boxer-type underwear
Tampon use if appropriate / avoid pads
Antiseptic washes are optional
Consider anti-androgen treatment
Stop smoking
Zero dairy diet with low glycemic load diet
At all stages – especially if weight an issue – consider use of metformin to improve sensitivity to insulin in patients on high glycemic load diets. Lowering chronic hyperglycemia reduces insulinemia and so decreases the impact on androgen receptors with a positive outcome.

Treatment - Hurley’s Stage I
Abscess formation, single or multiple without sinus tracts and cicatrisation/scarring.

This is the most limited form of disease and it is amenable to medical therapy.
The majority of patients with Stage I have a few flares a year, however they can be well controlled.

Medical Treatment for Stage 1 hidradenitis suppurativa
Topical antibiotics
  - Clindamycin 1% lotion bid

Intralesional
  - Triamcinolone acetonide 10 mg/mL, 0.5 to 1 ml injected with a 30g needle into individual, painful, early papules / small nodules to suppress inflammation. Inject right into the center of the lesion

Systemic Antibiotics (for 7-10 days) - wide choice
  - Tetracycline 250-500mg po qid or doxycycline 100 mg po bid or clindamycin 300 mg po bid, or amoxicillin/ clavulanic acid 500mg-1gm po q 8h
  - Caution in patients with diabetes- high dose steroids can interfere with their glucose control.

Adjunct preventive therapy
  - Zinc gluconate 50 mg with copper 2mg po bid and vitamin C 500 mg tid

Anti-androgens
  - Yasmin – consider extended regimen (daily x 84 – 126 days)
  - Yasmin plus spironolactone
  - Spironolactone 100-200mg/d
  - Finasteride 5 mg/d (Use of finasteride 5 mg per day in women and young girls as an antiandrogen for both therapy and long-term prevention)

Surgical Treatment – not usually needed for Hurley’s Stage I

General Care
  - Avoid irritants
  - Loose clothing
  - Stop smoking
  - Weight loss

Maintenance
  - Continue above as needed
Treatment - Hurley’s Stage II
Recurrent abscesses with sinus tract formation and scarring, either single or multiple widely separated lesions

The aim is to clear these patients or at least reduce them to stage I disease. If there are sinus tracts and scarring this will require combined medical and surgical therapy. For those with little scarring and much inflammation use antibiotics such as rifampin and/or clindamycin for 3 months and then decrease to maintenance on tetracyclines and/or high dose zinc and/or dapsone.

General care and intrallesional treatment is the same as for stage I. Antibiotics for at least three months are usual, with a decreased dose for maintenance. Systemic antibiotics include tetracycline, as above or, for more extensive disease, clindamycin 300 mg twice a day often combined with rifampin 300 mg twice a day for three months. (See below for prescribing details) Dapsone 100 mg per day can be used. (See below for prescribing details) Long-term maintenance is with a tetracycline etc. (as below) is often recommended. The same adjunctive therapy with diet, no nicotine and zinc gluconate and anti-androgens - see above.

A. Medical Treatment for Stage II
Topical antibiotics
   Clindamycin 1% lotion twice a day
Systemic Antibiotics
   Amoxicillin and clavulanic acid 3g loading then 1g po q8h for 5-7 days for acute painful lesions or
   Clindamycin 300 mg po bid with / without Rifampin 300 mg po bid or Dapsone 50 mg po and then 100 mg po with the appropriate blood work (See below for prescribing details).

   Maintenance – Tetracycline 250-500 mg qid, doxycycline or minocycline 100 mg bid

Adjunct preventive therapy
Zinc gluconate 50 mg with copper 2 mg po bid and Vitamin C 500 mg tid
Anti-androgens
   Yasmin – consider extended regimen (daily x 84 – 126 days)
   Yasmin plus spironolactone
   Finasteride 5 mg/d
Intrallesional triamcinolone as in Stage I
B. Surgical Treatment – If there are persistent chronic sinus tracts or cysts then obsessive surgical wide unroofing is necessary. Incision and drainage (I and D) should be avoided. Only do this for a tense abscess that is too painful to bear. Acute painful lesions sometimes develop into severely painful abscesses that need to be drained for pain relief only. This is not a curative procedure and needs concurrent antibiotics in full dose. Amoxicillin and clavulanic acid 3g in a single dose, then one gram po tid for 5-7 days is recommended. The lesion must be incised. Packing the wound for a few days may be needed to prevent premature superficial closure while the wound fills in from below.

C. and D. General Care and Maintenance- as for Stage I

Treatment - Hurley’s Stage III

Diffuse or almost diffuse involvement or multiple interconnected tracts and abscess

This stage is a surgical disease and supportive concurrent medical treatment is both prophylactic and essential. This requires a staged medical – surgical team approach

A. Medical Treatment

Pre-Op - These patients will need the anti-inflammatory effects of medical treatment to prepare them for surgical treatment.

- Corticosteroids 0.5 – 0.7 mg/kg/d methylprednisolone or prednisone (oral)
- Cyclosporine 4 mg/kg/d po
- Methotrexate 15 mg oral or subcutaneously weekly
- TNF-α inhibitors
  - Infliximab 5 mg/kg I.V Q6 weeks – use with the help of a knowledgeable health care provider
  - Adalimumab 40 mg every other week and ustekinumab also have been used

Biologics decrease swelling, inflammation and discharge pre-operatively, simplifying unroofing and excisional surgery, but affect neither the epithelialized sinus tracts nor the invasive proliferative gelatinous mass that is so resistant to therapy. Biologics are not a cure; improvement is rarely permanent.

- Clindamycin 300 mg po bid with Rifampin 300 mg po bid

Note – Medical treatment at this stage is only palliative and temporary. They should avoid nicotine after surgery in order to prevent new lesions and follow the dietary recommendations. Antiandrogens may still be needed.

B. Surgical Treatment
Wide surgical unroofing and debriding of all cysts and sinuses and fistulous tissue by a knowledgeable surgeon. Healing can be by secondary intent or it may be accelerated with mesh grafting. Primary closure is avoided in active disease. At times skin flaps are required.

Local Unroofing Surgery

Unroofing is simple surgery, an old technique that has been ignored for years. Recently revived, it deserves wide use. It is practical for lesions from the early hot nodules of Stage I to the advancing, branching lesions of Hurley Stage III. Removing early lesions and taking the tops off the deep epithelialized subcutaneous sinus tracts of HS/AI is invaluable. It requires nothing more than sturdy scissors, blades held parallel to the skin surface. Alternatively, laser has been used. It is far more effective than prolonged antibiotics and anti-inflammatory therapy.

Unroofing is not technically difficult, can be performed in the office setting under local anesthesia, and so is easily adapted to the Emergency Room.

This is the technique that we recommend replace “I&D” of fluctuant masses and other manifestations of HS/AI. Every opportunity to perform I&D should be converted into an opportunity to unroof the lesion. It provides superior drainage and pain control, eliminates the risk of inadequate ‘wound toilet’ that leaves behind the invasive proliferative gelatinous mass (IPGM) and fragments of the exploded FPSU. These are the sources of recurrences.

I&D is a temporary ‘solution’; unroofing is almost always permanent. It requires very simple post-operative dressings and post-operative pain is remarkably easy to manage.

Lidocaine 1-2% anesthesia with epinephrine is used. Controlled volumes are injected peripherally, avoiding leakage through sinuses. Time for vasoconstriction reduces pain and blood loss.

A single inflamed follicular unit requires only urgent mini-unroofing (not I&D). A biopsy punch of appropriate diameter (5-8mm) is centered over the involved FPSU and a twisting incision removes the central damaged material. This is then debrided with digital pressure, grattage with gauze wrapped around a cotton applicator, then ferric chloride hemostasis is applied with a cotton-tipped applicator.

Fluctuant masses are best initially incised and drained to reduce pressure. The central linear incision is extended to the edge of the loose tissue over the fluctuant area and the incision is extended through 360 degrees at the edge of the ‘roof’, beveling the edges with scissors. The base of the wound is then scrubbed with coarse gauze. Curettage with a spoon or bone curette may be needed to remove the IPGM. Excision of fat at the base of the wound is unnecessary and counterproductive. All depths and margins are explored digitally, visually, and with scissors tips. Any linear fibrous tissue is suspect as a possible sinus track and is best removed. Communicating sinuses once detected are unroofed. They can be surprisingly extensive and must be totally unroofed. Remove all tissue that is involved with active disease, devitalized or, if left behind, would interfere with healing. The wound base and small bleeders are dried and sealed with ferric chloride solution. Electrodesiccation or electrocautery are rarely needed. Scars are normally soft, contract to a much smaller area than that unroofed, and are quite acceptable to the patients.

Post-operatively, the wound is dressed with a thick coat of simple petrolatum. Running water only, no anti-bacterial soaps and no washcloths are used. Thick layers of petrolatum on cotton or soft gauze are re-applied once or twice daily or as needed. Patients (and wound care staff) must
avoid debriding the wound. Healing by secondary intention and epithelialization will proceed only if the fresh epidermis is allowed to cover the wound and is not debrided away.

HS/AI is not an infection; the inflammation is caused by the material removed by this procedure, so antibiotics are rarely necessary and are best avoided to minimize overgrowth of yeast and resistant bacteria.

Unroofing also eliminates the risk and costs of hospital or ambulatory surgical center care, laser, general anesthesia, graft donor sites, dehiscence, infection, the burying of residual inflammatory foci, post-operative antibiotics, time lost from work, and the need for travel to major centers. When performed correctly it stops forever the progression of the lesion treated.

**Pre-operative Clinic: Reminders for Hidradenitis Patients**

1. Consider *Nutrition consult* - screening tool per nutrition: albumin and prealbumin with preop labs
2. Encourage tobacco cessation; discuss impact on wound healing, need for avoidance of nicotine replacement products post-operatively.
3. Give instructions for extensive bowel prep, use Golytely prep if h/o kidney or heart disease. The patient must be clear prior to OR.
4. Correct anemia prior to OR.
5. If not on OC’s, try to schedule surgery in luteal phase to avoid menses in post-operative time frame.
6. Counselling re extent of excision, possibility of recurrence, prolonged hospitalization (at bed rest) and healing time.
7. Counselling re clear liquid long term diet in hospital with TPN and rectal tube (Bard Dignicare).
8. Administer DLQI, *Beck depression inventory, sexual health and function questionnaire*, etc. if not recently done.
9. Psychological needs to be addressed prior to OR
10. Discuss possible transfusion (need adequate HCT for adequate healing)
11. Arrange PIC line on POD 0 or 1
12. Arrange for a Clinitron specialty bed
13. Neurontin the day of OR (1200 mg)
14. Rule out Crohn disease *serologic markers for Crohn’s pANCA, ASCA, OmpC and CBir1 Flagelin markers as well as consider upper GI evaluation as well as colonoscopy.*
15. Consent for 3 procedures, plus numerous wound vac changes
   1. Radical vulvectomy, excision of buttock and thighs and wound vac(s) placements
   2. Wound vac removals and replacements, Split thickness skin graft after wound cleaning and wound vac(s) placements
   3. Removal of wound vats and staples
   4. Additional wound vac placements and removals

Will require 2 OR tables for extensive disease (rotate from prone to lithotomy)

**Consents for procedures**
Consent for radical vulvectomy for first procedure. Consent for skin grafts with possible skin flaps for second major procedure. Consent for removal of wound vats for other procedures and skin care for other procedure.
A bowel preparation prior to surgery is important if the anal area is involved and a wound VAC over that area is anticipated. It is a good idea anyways if a major area of the vulva is involved. The patients should be evaluated for malnutrition prior to surgery.

OR 1
Intra-operative: Have Available for **OR#1** (Radical Vulvectomy)

Bard Dignicare rectal tube  PUT IN RECTAL TUBE  
PRIOR TO PREP

Instructions for Use
1. Preparation of catheter and collection bag
   a. Attach the 60 ml syringe to the inflation port and draw all air from the retention cuff.
   b. After the cuff has been deflated, fill the syringe with 45 ml of water and set aside.
   c. Attach the collection bag to the catheter by inserting the ball valve connector of the catheter into the hub socket on the collection bag and turning clockwise until the connector snaps into place.

2. Insertion of Device
   a. Unfold the length of the catheter to lay flat on the bed towards the foot of the bed
   b. Attach the 60 ml syringe filled with 45 ml of water to the inflation port.

Insert the inflation cuff using a four-step process:
1. Squeeze the inflation cuff to ensure all air has been removed and hold the cuff flat in order to fold for insertion.

2. Holding the left point of the cuff between the thumb and index finger, fold the top right point of the cuff down and to the left in a 45 degree angle to create a conical shape with a leading edge for easy insertion.

3. Coat the cuff end with lubricating jelly.

4. Gently insert the cuff end through the anal sphincter until the cuff is beyond the external orifice and well inside the rectal vault.

Inflate the cuff with 45 ml of water by slowly depressing the syringe plunger. As the cuff inflates, the pilot balloon also inflates. The inflation port needs to remain parallel to the catheter.

Remove syringe from inflation port and gently pull on the silicone catheter to check that the cuff is securely in the rectum.

Position tubing on the inner leg.

Insert bag plug

Bulb needs to be filled with 45 cc water whenever in bed except for when deflated as follows:

Deflate for 5 minutes q 6 hours in 15 degrees Trendelenburg.

It can be used for 29 days. Periodically milk the catheter to facilitate flow. Change the collection bag before it becomes too full (between 600 and 800 ml). If the catheter becomes blocked with solid particles it can be rinsed with water.

Flush both tubes q 6 hours with fluid (water) tube flushes should be alternated with bulb deflation so that there is tube manipulation q3 hours.

Supplies

For prone, need gel pads, pillow and elbow protectors

Stryker irrigator with X-Ray bag
Yellow fin stirrups

Set Coag at 40/40 Blend

Supplies for aerobic and anaerobic culture of wound bed
Ligasure Cautery Hand (one large one for abdomen and mons pubis and one small one for vulva)

VAC foams (Silver (granu foam) for post-vulvectomy OR 1

VAC machine, canister and dressings  OF NOTE: Wound VAC must be on anterior vulvar aspect near mons. Make sure nothing is covering the holes on the wound VAC tube insertion point. It should be set for OR 1 continuous at 150. Consider 2 wound vats if large area involved. One at superior aspect and one at mons level versus on buttock.

To prevent further leaks around the Foley and the rectal tube use a Hollister urostomy wafer cushion over the initial wound vac plastic covering, stock number 7806. They keep them with the urology supplies. Cut a slit to the center and use the smaller one for the Foley, and the larger, one around the rectal tube (need to trim this one).

For large buttock resections start on prone (For prone, need gel pads, pillow and elbow protectors.) Cover the edge of the first part of the area excised with mastisol (need at least 4 of them) on edge. Then cover the excised area with Styrofoam and wound vac, leaving a portion of the wound vac approaching the perineum unsealed. Cover this with sterile towels, and then roll to lithotomy position. This way, the buttock can be sealed easily.

To prep the area for wound vac:

1) Make sure everything is dry, especially under the buttocks. Apply sticky plastic sheeting using ~1 inch strips around graft site in window pane fashion. This helps protect the skin and create a better seal.
2) Cut foam to fit Vulvectomy site. Silver-impregnated foam for OR 1 and first wound VAC change to improve antibacterial properties. Slits/holes are needed for the foley and rectal tube.
3) Apply Hollister wafer cushions around foley and rectal tube after foam is covered with intial plastic sheeting for wound VAC. For the rectal tube, cut a slit in the Hollister wafer to open it, then enlarge the hole a bit. Apply it around the tube and overlap to create a better seal.
4) Use window paning technique around Hollister wafers to get better seal.
5) Apply Mastisol (need at least 4) to the skin -- this can even go over the window pane plastic. It's especially important over the buttocks.
6) Put plastic sheeting/Tegaderms/etc. over foam to get good seals everywhere.
7) When ready to attach wound vac, cut a quarter-sized hole in plastic and apply the wound vac connector.
8) When starting suction, first deflate foam and remove as much air as possible using surgical suction (wall suction) canisters, and compress foam with hands to get as much air out as possible and get a better seal. Once the foam is essentially completely deflated, connect tubing to wound vac device.
9) Connect wound vac as above.

Make sure the Foley is draining at the correct angle
Make sure the rectal tube is at the correct angle. Irrigate with 60 cc water through Catheter irrigation port to make sure draining correctly
Take back from OR on Sport specialty bed.
NEED PICC line placed after OR on floor. Optimal placement at cavoatrial junction. OK if in distal 1/3 of SVC to cavoatrial junction. Once radiologist pages the nurse that placed the PICC, they page the resident to place the order to use the PICC.

**OR 2 (Wound Debridement and Wound Vac Changes)**. Ok to take to OR on Sport specialty bed for transfer back to floor to minimize risk of disrupting wound vac seals.

Intra-operative
Have Available for OR #2 (Wound Debridement and Wound Vac Change)

**Set Coag at 40/40 Blend**

Yellow fin stirrups
Stryker irrigator with X-Ray bag
Vac machine(s), canister(s) and dressings.
Change rectal tube prior to prep

**Bard Dignicare Rectal Tube**
Fill bulb with 45 cc fluid water. Deflate the bulb with patient in 15 degrees Trendelenburg qd for 5 mins. every 6 hours. Periodically milk the catheter to facilitate flow. Change the collection bag before it becomes too full (between 600 and 800 ml). If the catheter becomes blocked with solid particles it can be rinsed with water. Flush both tubes q6 hours with saline or water. The tube flushes should be alternated with the bulb deflations such that rectal tube manipulations occur q3 hours. The patient does not need to be put into Trendelenburg position for tube flushes.

If buttocks are involved, start in prone position (For prone, need gel pads, pillow and elbow protectors.)
Consider tissue cultures. Debride buttock wound and replace wound vac, including sealing plastic sheeting around area. Make sure rectal tube is at proper angle to allow for drainage. Replace rectal tube at beginning of procedure prior to prep.

Once in lithotomy position, debride wound and replace remainder of wound vac.

OF NOTE: Wound VAC must be on anterior vulvar aspect near mons. Make sure nothing is covering the holes on the wound VAC tube insertion point. Set at continuous at 150 mm Hg if large area (less if small area).
When placing wound vac, around the Foley and rectal tube, cut into smaller pieces to form a star around the wound tubes. Set wound vac at 150 mm Hg continuous if large area involved Can cover flaps with wound vac too.
Make sure everything is dry, especially under the buttocks. Apply sticky plastic sheeting using ~1 inch strips around graft site in window pane fashion. This helps protect the skin and create a better seal.

**OR 3 (Skin Grafts)** (Consider Flap with this surgery if needed. If a flap is done, the edges of the flaps need to be excised to healthy tissue.)

Stop Heparin 12 hours before OR.
Take to OR on specialty bed, then transfer to OR bed.

If flaps (especially muscle flaps with vessels reattached) performed, keep Hct above 30% to ensure wound healing.

Have Available for OR#3 (Split Thickness Skin Graft)

**Set Coag at 40/40 Blend**

Yellow fin stirrups
Stryker irrigator with X-Ray bag
VAC machine, canister and dressings  OF NOTE: May require two wound VACS (abdomen and Mons/buttock). Make openings quarter size. Make sure nothing is covering the holes on the wound VAC tube insertion point. Set at continuous at 150 mm Hg if large area (less if small area).

For skin grafting procedure:
- Have available large curette used by plastic surgery for debridement.
- Dermatome setting: 12 to 17/1000 inch  (15 ideal)
- 3 inch guard.
- Meshed 1.5/1. NEED TO CLEAN OUT MESHER and relubricate it after 4 passes
- Need extra carriers.
- Have an assistant to gently lift up the skin graft if it piles up on the guard.
- Change blade every 3 to 4  passes.
- Consider if you will need to prep both thighs; wipe off thighs with water or saline prior to putting on mineral oil prior to doing skin graft.

When doing the skin graft use a 45 degree angle. Can use towel clips, or just push down on the skin.

Intra-operative
- Start in prone position if both sides are being done. For prone, need gel pads, pillow and elbow protectors.
- Remove and replace rectal tube prior to prep.

**Bard Dignicare Rectal Tube**

Fill bulb with 45 cc fluid water. Deflate the bulb with patient in 15 degrees Trendelenburg qd for 5 mins. every 6 hours. Periodically milk the catheter to facilitate flow. Change the collection bag before it becomes too full (between 600 and 800 ml). If the catheter becomes blocked with solid particles it can be rinsed with water. Flush both tubes q6 hours with saline or water. The tube flushes should be alternated with the bulb deflations such that rectal tube manipulations occur q3 hours. The patient does not need to be put into Trendelenburg position for tube flushes.

Obtain grafts. After taking the graft, cover the thigh with epinephrine 1:1,000
(If small area can use 1% lidocaine with 1:200,000 epinephrine on raytec )
1) On buttock, need to apply a narrow skin graft in buttock groove vertically a, then cut two additional skin pieces to go on either side of central piece that indents some. Make sure everything is dry, especially under the buttocks. Cover over with Adaptic (Curity Non Adhering Dressing 5 x 9).

Ensure that there is overhang of the Adaptic (Curity Non Adhering Dressing 5 x 9) over the edge of the incision sites so that if things bunch, the skin is still protected.

2) Apply Mastisol (need 4) to the skin -- this can even go over the window pane plastic. It's especially important over the buttocks.

3) Cut black foam to fit buttock graft site. Slits/holes are needed for the foley and rectal tube. The foam pieces can be stapled together to keep the shape and location as desired. Need to apply a narrow sponge in buttock vertically over Adaptic (Curity Non Adhering Dressing 5 x 9) covering skin graft, then cut two additional sponges and staple them to the buttock central sponge.

4) Apply Hollister wafer cushions around foley and rectal tube after initial covering with plastic sheeting. For the rectal tube, cut a slit in the Hollister waffer to open it, then enlarge the hole a bit. Apply it around the tube and overlap to create a better seal.

5) Use window paning technique around Hollister wafers to get better seal.

6) Put plastic sheeting/Tegaderms/etc. over foam

For buttock flaps cover the buttock with towels, then roll to lithotomy position. This way, the buttock can be sealed easily later on. To prevent further leaks around the Foley and the rectal tube use a Hollister urostomy wafer, stock number 7806. They keep them with the urology supplies. Cut a slit to the center and use the inner, smaller part for the Foley, and the larger, outer part around the rectal tube, after plastic sheeting has been applied.

7) When ready to attach wound vac, cut a quarter-sized hole in plastic and apply the wound vac connector.

Rotate to lithotomy position.

Once in lithotomy position, drape as follows: Green towels wrapped around leg with long axis of towels perpendicular to long axis of leg. Wrap the towels around the leg just proximal to the knee to cover the knees and upper portion of the yellow-fin stirrups. Pull leg drapes over legs and attach to green towels, leaving thighs free. Clean Green towels can then be placed over the thighs to keep the donor sites clean. Place ¼ drapes under abdomen and thighs to keep undersurface clean. Drape remainder of patient (abdomen, legs, etc.) in usual fashion using standard laparotomy drape.

Obtain grafts. After taking the graft, cover the thigh with epinephrine 1:1,000 (If small area can use 1% lidocaine with 1:200,000 epinephrine on raytec)

Use 4’0’ monocryl on prepuce and labia if desired, however staples are fine too.

For lithotomy, need to apply a narrow skin graft in each lateral aspect of abdomen vertically and staple to the central piece to decrease tension on the sides of the abdomen. Then, cover graft over with Adaptic (Curity Non Adhering Dressing 5 x 9), then cut sponges in similar fashion and cover Adaptic (Curity Non Adhering Dressing 5 x 9) with sponges. Complete wound VAC in lithotomy.

When placing wound vac, around the Foley and rectal tube, to prevent further leaks around the Foley and the rectal tube use a Hollister urostomy wafer cushion over the initial wound vac plastic covering, stock number 7806. They keep them with the urology supplies. Cut a slit to the center and use the smaller one for the Foley, and the larger, one around the rectal tube (need to trim this one). Cut into smaller pieces to form a star around the wound tubes. Set wound vac at 150 mm Hg continuous if large area involved Can cover flaps with wound vac too.
Xeroform gauze to cover skin graft sites; Staple at corners. Place ABD over Xeroform, then Kerlex wrap and Ace bandage.(Remove Kerlex and ABD on POD 1 from thighs). Another option for wrapping the leg which worked nicely was to use xeroform gauze covered with ABD, then Kerlex, then cover with Bandnet 10” pack (precut Bandnet wrap). It is brought up over the heel and pulled up to the thigh.

Consider 2 or more wound vacs (vulva and buttocks) if large area involved.
To prep the area for wound vac:

When starting suction, compress foam with hands to get as much air out as possible and get a better seal.
Apply sticky plastic sheeting using ~1 inch strips around graft site in window pane fashion. This helps protect the skin and create a better seal. Cover with sponges and staple them to the buttock central sponge.

10) For graft donor site, the entire site can be covered with Xerofoam. Staple at corners and remove staples on legs at time wound vac removed POD 5 after split thickness skin graft. Wrap with Kerlex, then cover with ABD. Remove ABD and Kerlex one day after the grafts from the donor site.

Leave Xeroform to dry and trim away dry areas the come off of the skin.

Use heat lamp to thigh after ABD removed. If using this technique, after staples on thigh removed at time of wound vac removal, gradually cut off xeroform.

If doing flaps use 3’0’ vicryl buried stitches to reapproximate the skin through the dermis. Then close the skin with 3’0’ Nylon. The Nylon stitches should be removed in 3 weeks.

Remove Wound VAC after 5 days in on floor with SWAT team or in operating room, and take out staples from vulva and buttock POD 14

POD #5
Remove Wound VACS after on the floor with SWAT (use liquid bandage remover) or in operating room.
Irrigate wound VACS using a 60 cc syringe and (may need a catheter adapter (Christmas tree adapter) (blue one) Consider removing rectal tube and Foley versus leaving in for a day or two more.

On skin grafts, place Xeroform gauze (double layer) with Bacitracin touching the graft and areas that may not have taken and cover with Kerlex, followed by ABD, and stretchy underwear. If too wet, leave to air. Change the kerlex and ABD tid.
Cut edges of Xeroform as it dries.
Cotton flushes
Burn net panties for compression
OR on POD # 12-14
Remove staples

Post-operative Considerations

1. Check wound cultures, check if bacteria resistant to present antibiotic. If sterile culture, consider discontinuing antibiotics.
2. Continue TPN
3. Sips and chips

Post-Op - They will need ongoing medical treatment for their hidradenitis after surgery. Rectal tube can be left in short term if needed.

Orders OR 1 Vulvectomy Post-op Orders

Immediate Post-op
Admit to 8B
Service:
Attending:
Diagnosis: S/P Complete Radical Vulvectomy
Condition: Stable
Allergies:
Activity: Complete bedrest, do not elevate head of bed more than 20 degrees
VS: q 1 hour X 2, q 2 hour X 2, then q 4 hours
I/O’s q 4 hours
Diet-sips and chips
Hyperal
Start sliding scale

IV: D5NS with 20 meq/L KCl at 125 cc/hour, change to D5/0.45 NS with 20 meq/L KCL on POD#1, 80 cc/hr, decrease to KVO when tolerating po well
SCD’s on and functioning at all times
Incentive spirometry X 10 q 1 hour while awake
Instruct patient in cough and deep breathing, q 1 hour while awake
Physical therapy consult: supportive care while at bedrest, post-bedrest rehabilitation
Occupational therapy consult: activities for bedrest
Social work consult: home nursing needs, support

VAC Therapy Order: VAC machines, canisters and dressings to be placed at patient’s Bedside
Goal: Formation of granulation tissue in wound bed
VAC to be applied to vulva
Pressure setting: 150 mm Hg continuous if large area involved (if small area, 125 mm Hg)
Never leave subatmospheric pressure off or more than 2 hours per 24 hour period
Dressing will be changed POD 7 in the operating room
Bard Dignicare bowel system to closed drainage. Every 6 hours, place patient in 15 degrees Trendelenburg
and deflate the ballon (withdraw 45 cc from Balloon Inflation Port; wait 5 minutes, then place back 45 cc
sterile water). The nurse should not disconnect the syringe from the bulb inflation/deflation port, in order to
minimize the risk of introducing too much fluid into the bulb. After this, take out of Trendelenburg.
Every 6 hours, the rectal tube should be flushed at both tube ports (labeled “IRRIG” and “FLUSH”) with 45
cc each sterile water or saline. The flushes should be alternated with the bulb deflations, such that rectal
tube manipulations occur every 3 hours. The patient does not need to be in the Trendelenburg position
during tube flushes.
At other times, patient to be rotated from left lateral position to right lateral position every 2 hours. When
buttock involved, do not have patient lying on back.

Foley catheter to gravity drainage, do not remove

Labs: CBCDP, Basic, ical, Mg, Phos in am POD #1
(Consider labs in PACU depending on EBL/PRBC’s/pre-op Hct)
Medications:
  PCA: Start/Managed per Anesthesia, encourage epidural per anesthesia
  Toradol 30 mg IV X 24 hours, (use 15 mg if > 65 yrs or <50 kg,) change to PO Ibuprofen when
tolerating PO well
  Neurontin
  Tylenol
  Ancef: 1 gram IV q 8 hours (May need revision when wound culture results
available.)
  Diflucan 150 mg PO q week
  Heparin 5000 units SQ q 8 hours; D/C heparin 12 hours prior to OR 1 week later, and 12 hours prior
to removal of wound vac 5 days after second surgery

FeSO4 325 mg PO daily
Tylenol 325-650 mg PO every 4-6 hours PRN mild pain/ headache  (Not to
Exceed 3000 mg/24 hours)
Benadryl 12.5- 25 mg PO/IV q 6 hours PRN itching
Ambien 5-10 mg PO qhs PRN sleep
Phenergan 12.5-25 mg IV q 6 hours PRN nausea
Zantac 150 mg PO twice daily
Lomotil- Start on Lomotil up to qid a day before going for skin graft

OC’s:  continue if patient on preoperatively, consider other menstrual suppression
Tobacco service consult as indicated (No Nicotine containing products!)
  [Encourage tobacco cessation preop]
(Review home medications and resume those indicated)
Notify H.O. (pager 0005): temp > 100.4, SBP > 180 or < 80, DBP>95 or <50, HR >110 or < 60, UOP <120
cc/4 hours, dysfunction of VAC or rectal pouch, any sudden, rapid increase in bright, red blood in the tubing
or canister of the VAC.
Make sure they have a specialty bed – page ostomy nurses day before surgery ideally in order to allow bed to be ordered and delivered to the OR.

**Orders OR 2 Post-op Wound Vac Removal**

(Same as above for OR 1 Post-op)

Admit to 8B  
Service:  
Attending:  
Diagnosis: S/P Complete Radical Vulvectomy  
Condition: Stable  
Allergies:  
Activity: Complete bedrest, do not elevate head of bed more than 20 degrees  
VS: q 1 hour X 2, q 2 hour X 2, then q 4 hours  
I/O’s q 4 hours  
Diet-sips and chips  
Hyperal  
Start sliding scale  

IV: D5NS with 20 meq/L KCl at 125 cc/hour, change to D5/0.45 NS with 20 meq/L KCL on POD#1, 80 cc/hr, decrease to KVO when tolerating po well  
SCD’s on and functioning at all times  
Incentive spirometry X 10 q 1 hour while awake  
Instruct patient in cough and deep breathing, q 1 hour while awake  
Physical therapy consult: supportive care while at bedrest, post-bedrest rehabilitation  
Occupational therapy consult: activities for bedrest  
Social work consult: home nursing needs, support  

VAC Therapy Order: VAC machines, canisters and dressings to be placed at patient’s Bedside  
Goal: Formation of granulation tissue in wound bed  
VAC to be applied to vulva  
Pressure setting: 150 mm Hg continuous if large area involved (if small area, 125 mm Hg)  
Never leave subatmospheric pressure off or more than 2 hours per 24 hour period  
Dressing will be changed POD 7 in the operating room  
Bard Dignicare bowel system to closed drainage. Every 6 hours, place patient in 15 degrees Trendelenburg and deflate the ballon (withdraw 45 cc from Balloon Inflation Port; wait 5 minutes, then place back 45 cc sterile water). The nurse should not disconnect the syringe from the bulb inflation/deflation port, in order to minimize the risk of introducing too much fluid into the bulb. After this, take out of Trendelenburg. Every 6 hours, the rectal tube should be flushed at both tube ports (labeled “IRRIG” and “FLUSH”) with 45 cc each sterile water or saline. The flushes should be alternated with the bulb deflations, such that rectal tube manipulations occur every 3 hours. The patient does not need to be in the Trendelenburg position during tube flushes.  
At other times, patient to be rotated from left lateral position to right lateral position every 2 hours. When buttock involved, do not have patient lying on back.
Foley catheter to gravity drainage, do not remove

Labs: CBCDP, Basic, iCal, Mg, Phos in am POD #1
(Consider labs in PACU depending on EBL/PRBC’s/pre-op Hct)

Medications:
- PCA: Start/Managed per Anesthesia, encourage epidural per anesthesia
- Toradol 30 mg IV X 24 hours, (use 15 mg if > 65 yrs or <50 kg,) change to PO Ibuprofen when tolerating PO well
- Neurontin
- Tylenol
- Ancef: 1 gram IV q 8 hours (May need revision when wound culture results available.)
- Diflucan 150 mg PO q week
- Heparin 5000 units SQ q 8 hours; D/C heparin 12 hours prior to OR 1 week later, and 12 hours prior to removal of wound vac 5 days after second surgery

- FeSO4 325 mg PO daily
- Tylenol 325-650 mg PO every 4-6 hours PRN mild pain/ head ache (Not to Exceed 3000 mg/24 hours)
- Benadryl 12.5-25 mg PO/IV q 6 hours PRN itching
- Ambien 5-10 mg PO qhs PRN sleep
- Phenergan 12.5-25 mg IV q 6 hours PRN nausea
- Zantac 150 mg PO twice daily
- Lomotil- Start on Lomotil up to qid a day before going for skin graft

OC’s: continue if patient on preoperatively, consider other menstrual suppression
Tobacco service consult as indicated (No Nicotine containing products!)
[Encourage tobacco cessation preop]
(Review home medications and resume those indicated)

Notify H.O. (pager 0005): temp > 100.4, SBP > 180 or < 80, DBP>95 or <50, HR >110 or < 60, UOP <120 cc/4 hours, dysfunction of VAC or rectal pouch, any sudden, rapid increase in bright, red blood in the tubing or canister of the VAC.

Make sure they have a specialty bed – page ostomy nurses day before surgery ideally in order to allow bed to be ordered and delivered to the OR.

Orders OR 3 Post-op Skin Graft
Admit to 8B
Service:
Attending:
Diagnosis: S/P Vulvar skin graft
Condition: Stable
Allergies:
Activity: Complete bedrest, do not elevate head of bed more than 20 degrees
Patient to be rotated from left lateral position to right lateral position every 2 hours.
VS: q 1 hour X 2, q 2 hour X 2, then q 4 hours
I/O’s q 4 hours
Diet: sips and chips
Hyperal
Start sliding scale
IV: D5NS with 20 meq/L KCl at 125 cc/hour, change to D5/0.45 NS with 20 meq/L KCL on POD#1, 80 cc/hr, decrease to KVO when tolerating po well
SCD’s on and functioning at all times
Incentive spirometry q 1 hour while awake
Instruct patient in cough and deep breathing, q 1 hour while awake
VAC Therapy Order: VAC machine, canister and dressings to be placed at patient’s bedside
Goal: Formation of granulation tissue in wound bed
VAC to be applied to vulva
Pressure setting: 150 mm Hg continuous if large area involved (if small area 125 mm Hg)
Never leave subatmospheric pressure off or more than 2 hours per 24 hour period
Dressing will be changed POD 5 under conscious sedation or in operating room
Bard Dignicare bowel system to closed drainage. Every 6 hours, place patient in 15 degrees Trendelenburg and deflate the ballon (withdraw 45 cc from Balloon Inflation Port; wait 5 minutes, then place back 45 cc sterile water or saline).
After this, take out of Trendelenburg.
At other times, patient to be rotated from left lateral position to right lateral position every 2 hours. When buttock involved, do not have patient lying on back.

Abductor pillows
Foley catheter to gravity drainage, do not remove
Labs: CBCDP, Basic, iCal, Mg, Phos in am
(Consider labs in PACU depending on EBL/PRBC’s/pre-op Hct)
Medications: (Circle medications desired)
   PCA: Start/Managed per Anesthesia, encourage epidural per anesthesia
Toradol 30 mg IV X 24 hours, (use 15 mg if > 65 yrs or <50 kg,) change to PO Ibuprofen when tolerating PO well
   Ancef: 1 grams IV q 8 hours X 48 hours
   Diflucan 150 mg PO q week
   Heparin 5000 units SQ q 8 hours
   Lomotil –i po qid (NOT PRN), can decrease to tid, bid if needed.
   Neurontin 300 at bedtime
   FeSO4 325 mg PO daily
   Tylenol 325-650 mg PO every 4-6 hours PRN mild pain/ headache. (Not to exceed 3000 mg/24 hours)
   Benadryl 12.5- 25 mg PO/IV q 6 hours PRN itching
   Ambien 5-10 mg PO qHS PRN sleep
   Phenergan 12.5-25 mg IV q 6 hours PRN nausea
   Zantac 150 mg PO twice daily
OC’s: continue if patient on preoperatively, consider other menstrual suppression
Tobacco service consult as indicated (No Nicotine containing products!)
Start to wean TPN one day prior to removal of WOUND VAC

Wound care for donor site (If wound vac not applied to donor sites, remove Kerlex and ABD 24 hours after surgery; leave on Xeroform – cut edges as they dry):

After the outer dressing has been removed from the thigh, apply a heat lamp (100 W bulb, not closer than 18’) to the donor site for 15 minutes 3 times a day until dry (usually 1-2 days). If it starts burning, turn the lamp off or move it further away.

Notify H.O. (pager 0005): temp > 100.4, SBP > 180 or < 80, DBP > 95 or < 50, HR > 110 or < 60, UOP < 120 cc/4 hours, dysfunction of VAC or rectal pouch, any sudden, rapid increase in bright, red blood in the tubing or canister of the VAC.

Stop Heparin 12 hours before OR 3
Bring down to OR on specialty bed

Orders Removal of WOUND VAC

Turn off wound vac 30 minutes before removal planned. Need to order a Christmas tree to put on tube of wound vac. Use 30 cc syringe and inject saline about 30 minutes before removal planned. Can also inject with 1% lidocaine vial and allow that to soak to decrease pain.

There is a liquid adhesive remover that can be used to assist in removing the wound vac.

Cover graft with Adaptic (Curity Non Adhering Dressing 5 x 9), then ABD then stretchy underwear. Cover thigh with Adaptic (Curity Non Adhering Dressing 5 x 9) and Kerlex. The following day, remove the Adaptic (Curity Non Adhering Dressing 5 x 9) from the graft and leave to air to dry. Can cover the graft with Kerlex if needed. Leave Adaptic (Curity Non Adhering Dressing 5 x 9) on the thigh to dry and cut it as it dries.

New orders:
Consider leaving in rectal tube for a few more days, while TPN is being weaned. Start them on clear liquids to full liquid diet while rectal tube in during this time (the first 2 surgeries, keep NPO x chips and occasional sip)
D/C lomotil when rectal tube out
Advance diet
When rectal tube out- Milk of magnesia 30 cc po q 6 hours, when stools start, prn
Dressing changes-use saline to take off xeroform if needed. Do daily. Reapply xeroform with bacitracin daily, then cover with Kerlex, then an ABD.

Patient to remain in bed for 4 days. If flap, will have gradual increase in sitting as follows:

The standard sitting protocol for these pts:
1) No weight bearing on buttocks for 3 weeks
2) Begin sitting protocol 15 mins TID for 2 days
3) Advance to 30 mins TID for 2 days
4) Advance to 45 mins TID for 2 days
5) Advance to 60 mins TID for 2 days
6) Continue with this advancement until she reached 120 mins TID and then she can sit without restrictions.  
Rotate from left lateral position to right lateral position every 2 hours.

After each sitting time period, the buttocks is checked to make sure that the flaps are tolerating the sitting (erythema, venous congestion, stress at suture line, early wound separation).

Number of dressing changes per day: 2

D/C PICC line prior to home
Send home on Stage 1-2 hidradenitis regimen (antibiotics, OCPs, or spironolactone dependent on age).
Arrange for visiting nurse.

After the yellow Xeroform gauze has dried, lightly lubricate it daily with Vaseline.  
Cocoa butter to thighs once the xeroform comes off
FOR HOME DRESSING CHANGES
DESCRIBE the dressing change process including number of each type of dressing product:  
Using Toumy syringe and NS, irrigate all wounds. Apply ___# of xeroform gauze (5 x 9) impregnated with bacitracin to all wounds. Apply a middle layer of 4 inch kerlix (total of ___# of rolls) moistened with NS.  
Cover with ___# of Abd pads and hold in place with mesh panties.

Products needed to provide dressing changes as ordered for 1 month:
180 4 inch kerlix #6715
180 Abd pads 8 x 10 #6715
10 mesh panties #SBXL100
10 boxes of 50 xeroform gauze #433605
60 blue pads
1 tube bacitracin #001116
1 box tongue depressors #WOD3005
1 Toumy syringe #30962

Prognosis – The majority of patients are in stage 1 and can be controlled well. Stage 2 can be more difficult and Stage 3 is very difficult and requires a multi-disciplinary treatment approach. Average duration of disease is 20 years. Squamous cell carcinoma may occur in patients with HS. It tends to be seen in patients who have suffered from HS for ten years or more, will often be advanced in stage at diagnosis.

Specific Drug Information for Medications Used in the Treatment of Hidradenitis Suppurativa

CLINDAMYCIN
In hidradenitis, clindamycin is used as an anti-inflammatory medication.  
– helps settle down the redness, swelling, etc.  
It is also a very effective medication for bacterial infections.

Side effects
Bowel inflammation can occur due to an overgrowth in the bowel of bacteria (C. difficile) that release a toxin. This can occur in a few patients. If there is any problem with diarrhea, stop the medication. Other side effects include upset stomach, vomiting, and skin rashes. Clindamycin can be taken with the rifampin or used separately.

**Dose** – 150 - 300 mg po twice a day - to be taken with food. Use for 3-6 months.

**Interactions** – can interact with birth control pills

**AMOXICILLIN / CLAVULANATE**

Used as an anti-inflammatory

**Dose** – For acute nodules and incised abscessed lesions - amoxicillin and clavulanic acid 3g loading then 1g po q 8h for 5-7 days (taken with food). For indolent nodules, 500 mg po tid for 1-2 weeks.

**Side effects** – allergy, GI upset, nausea, diarrhea, yeast, rashes

**Contraindications** – hypersensitivity

**Indications** – For acute nodular flares.

**ZINC GLUCONATE**

Zinc gluconate is anti-inflammatory and helps in wound healing.

**Dose** is 50 mg po bid or 30 mg po tid . This is suppressive rather than curative

**Side effects** are occasional GI upset with nausea and / or diarrhea.

Zinc in high doses can affect iron in the body with resulting anemia and drop in white count.

Do not increase the dose of zinc.

**RIFAMPIN**

Rifampin 150 and 300 mg tablets – this is an antibacterial agent that is used for bacterial infections, both common ones and mycobacteria including tuberculosis. This medication is used in hidradenitis suppurativa as an anti-inflammatory and is usually combined with other medications.

**Dose** - 150 – 300 mg po twice a day. Take on an empty stomach. It is occasionally given as 600 mg in one dose. It can be given with other medication such as clindamycin taken in two doses daily or may be given as a single dose with a large glass of water at 4 AM to prevent any interaction with the other medicines.

**Monitoring blood tests for Rifampin** - baseline CBC, renal and liver function tests should be taken. Caution should be taken if there is pre-existing liver disease or liver function abnormalities. Repeat blood tests at 2-4 week intervals as needed.

**Drug interactions – many may occur**

Birth control pills – decreases effect of BCP

- Blood thinning drugs – increases INR / clotting time
- Heart drugs – digoxin, quinidine
- Beta blockers – verapamil
- Anti-convulsants – phenobarbital, phenytoin
- Anti-fungal drugs – ketoconazole
- Bronchodilators – theophylline
- Immunosuppressant drugs – cyclosporine
- Corticosteroids
- Sulfonylurea and other hypoglycemic medications
- Miscellaneous – acetaminophen, dapsone.

Enalapril can result in an increase in blood pressure.
Side effects

Urine discoloration – orange red
Permanent staining of soft contact lenses

Allergic reactions

Flu-like syndrome with fever, chills, headache, dizziness & rashes
Skin rashes – itching, hives, pimply reactions, and blisters, rarely erythema multiforme or toxic epidermal necrolysis
Dizziness, headache and fatigue can occur
Rarely anemia and hepatitis

DAPSONE
This is used as an anti-inflammatory. It reduces PMN/WBCs in tissue

Dose – 50 - 100 mg po per day. Start at 50 mg/day for first 2-4 weeks

Caution – the glucose-6 phosphate dehydrogenase should be measured. If this is low there is a higher risk of blood problems such as anemia.

This can be more of a problem for some African Americans and Asians resulting in a more toxic reaction from the dapsone. Dapsone affects red blood cells so that they do not “live as long”. Usually red blood cells last for 120 days but when a patient is on dapsone this can decrease to 80 days causing the hemoglobin, to drop. This can be a problem in patients with heart, liver and kidney disease. A thorough history and physical with attention to the heart, liver and renal function is important.

Patients must be checked to be sure there is no anemia.

Contraindications to the use of dapsone include prior hypersensitivity and agranulocytosis. Patient with severe allergy (hypersensitivity) to sulfonamides may be allergic to dapsone. If a mild allergy to sulfonamides, this is less likely.

Relative contraindication would be significant cardiopulmonary disease, G-6PD deficiency, and severe sulfonamide allergy.

Monitoring blood tests for patients for dapsone
1. G-6PD level must be assessed.
2. CBC with differential, liver function tests, BUN, creatinine and urinalysis.
3. Repeat blood work - CBC with differential, WBC and reticulocyte count every week for 4 weeks and then every 2 weeks for 8 weeks and then about every 3-4 months. Check reticulocyte count to assess response to Dapsone hemolysis.
4. Liver function and renal function tests every 4 months for maintenance.

Drug interactions
1. Dapsone levels are increased with trimethoprim, probenecid
2. Dapsone levels decreased with rifampin
3. Dapsone, if combined with hydroxychloroquine and sulfonamides, yields more red blood cell toxicity

Cross Reactions
Other sulfonamide type drugs - patients with severe allergic reactions to sulfonamide medications may be allergic to Dapsone. This is very rare.

Adverse Effects
1. Hemolytic anemia, methemoglobinemia – symptoms headache, lethargy
2. Hepatotoxicity – mono-like syndrome
3. Peripheral neuropathy
4. Allergy – rashes etc.
5. GI upset

http://www.hs-foundation.org/

OR Scheduling

Preop Psych, dietician, R/o Crohns

OR 1 Radical Vulvectomy Wound VAC

OR 2  Day 3  Wound VAC Change

OR 3  Day 7  Skin grafts, possible flaps, Wound VAC change

OR 4  Day 12  Wound VAC off with SWAT on floor

OR 5  DAY 21 Staple removal  can be done at EAST ANN ARBOR

Notes to print out on OR BOARD

Inform if prone to lithotomy (For prone, need gel pads, pillow and elbow protectors.)

Epi 1/1000

Ligasure

Wound VAC 150 continuous

Hollister urostomy wafer

DigniCare Rectal tube

Stryker irrigator

Xray bag
Mastisol (need 4)
Adaptic (Curity Non Adhering Dressing 5 x 9)
Wound Vac on thighs
12-17/1000

Mesh 1.5 inch/1

Lorezepam may be needed if patient having problems with position changes
Tilt bed at time of taking patient off OR table…easier to roll

Behçet's Disease
Is a very rare condition. It is uncommon in North America but not in the Middle East. Behçet’s disease was first described in 1937 by Hulusi Behçet, a Turkish dermatologist. It is defined by a triad, classically of oral ulcers, genital ulcers and uveitis. Oral ulceration is the most common cutaneous finding in Behçet's disease. The most common sites of involvement are the buccal mucosa, gums, tongue, lips, and pharynx. In order to make a diagnosis of Behçet's, a patient must experience oral ulceration occurring at least three times in one year and fulfill the other criteria discussed below. The lesions tend to be painful, shallow to deep, and have erythematous borders with yellow, fibrinous bases. Ten percent of patients, however, develop major aphthous ulcerations, which are lesions that are larger, more persistent, and may heal with scarring. Vulvar lesions are quite common. Involvement of the vagina and/or cervix may also occur. Pathergy is one of the diagnostic criterions for Behçet's and consists of development of a small pustule within 24 to 48 hours after the skin has been pricked by a blunt sterile needle. Although helpful if positive, its sensitivity is debatable with some studies finding it as low as 10 percent (Davies PG, Fordham JN, Kirwan JR, et al. The pathergy test and Behçet's syndrome in Britain. Ann Rheum Dis 1984;43:70-3).

International study group criteria for the diagnosis of Behçet's disease

<table>
<thead>
<tr>
<th>Major criteria (need 1)</th>
<th>Recurrent oral ulceration</th>
<th>Minor aphthous, major aphthous, or herpetiform ulceration observed by health care provider or patient that recurred at least three times over a 12-month period</th>
</tr>
</thead>
<tbody>
<tr>
<td>Minor criteria (need 2)</td>
<td>Recurrent genital ulceration</td>
<td>Aphthous ulceration/scarring observed by health care provider or patient</td>
</tr>
<tr>
<td>Eye lesions</td>
<td>Anterior or posterior uveitis or cells in vitreous on slit lamp examination; or retinal vasculitis observed by ophthalmologist</td>
<td></td>
</tr>
<tr>
<td>Skin lesions</td>
<td>Erythema nodosum observed by health care provider or patient, pseudofolliculitis or papulopustular lesions; or acneiform nodules observed by the health care provider in a postadolescent patient who is not receiving corticosteroid treatment</td>
<td></td>
</tr>
<tr>
<td>Positive pathergy test</td>
<td>As interpreted by health care provider at 24 to 48 hours</td>
<td></td>
</tr>
</tbody>
</table>

Treatments that have been utilized in the treatment of Behçet Disease

<table>
<thead>
<tr>
<th>Class 1 or II topical steroids</th>
<th>Dapsone</th>
<th>Cyclosporine</th>
</tr>
</thead>
<tbody>
<tr>
<td>Intral(esional) triamcinolone acetonide</td>
<td>Systemic steroids</td>
<td>Cyclophosphamide</td>
</tr>
<tr>
<td>Topical anesthetics</td>
<td>Methotrexate</td>
<td>Thalidomide</td>
</tr>
<tr>
<td>Colchicine</td>
<td>Azathioprine</td>
<td>Interferon alfa-2a</td>
</tr>
</tbody>
</table>

DIFFERENTIAL DIAGNOSIS OF VULVAR EDEMA

Swelling can be due to any of these conditions or combinations of inflammation, infiltration and lymphatic disruption or obliteration

Inflammatory Edema

I Allergic/Immune
   1. Allergic Reaction
      a. Angioedema with or without urticaria
      b. Allergic Contact Dermatitis
   2. Granulomatous Inflammation
      a. Crohn’s Disease
      b. Melkersson-Rosenthal Syndrome
      c. Sarcoidosis

II Infection – edema secondary to local infection
   1. Cellulitis – streptococcal
   2. Abscess – Bartholin’s duct
   3. Candidiasis
   4. Rare – Tuberculosis, actinomycosis, Granuloma Inguinale, Amebiasis, Blastomycosis, Schistosomiasis

III Other
   1. Direct Trauma
   2. Hidradenitis suppurativa (HS)
   3. Amyloidosis
   4. Infiltrative neoplasm – inflammatory breast CA
Note – all infections, Crohn’s and HS can cause inflammatory edema the scarring and secondary obstructive lymphedema.

**Obstructive Lymphedema**

I Congenital

1. Milroy’s disease (congenital lymphedema)
2. Lymphangioma

II Infection with secondary lymphatic damage

1. Recurrent cellulitis – streptococcal
2. Lymphogranuloma venereum
3. Filariasis

III Physical lymphatic obstruction with mass, tumor, or destructive process

1. Pregnancy
2. Pelvic or local trauma
3. Pelvic tumor
4. Post-radiation scarring
5. Congestive heart failure

IV Metabolic

1. Obesity
2. Renal failure
3. Hepatic failure

Note – Chronic obstructive lymphedema can result in lymphangieactasia/ acquired lymphangiomas.

**PROTOCOL FOR LYMPHANGIECTASIA AND CHRONIC LYMPHEDEMA OF THE VULVA**

A. First control infection. Usually it is Strep and occasionally Staph.
   1. Gently cleanse with Cetaphil or another triclosan-containing antibacterial cleanser morning and night, pat dry.
   2. Bleach baths can be very useful in reducing re-infection. Do 2-3 times a week. Add one half cup of household bleach (125 mL) to a 10 inch (25 cm) deep tub of comfortably warm bath water. Mix well. Soak for 5-7 minutes, ensuring penetration of the solution into all cracks and genital / buttock / skin folds, using a plastic cup and bare hands to spread over all involved areas. For sitz bath mix 1 ¼ tsp bleach in 1 gallon of water.
   3. Antibiotic ointment (mupirocin ointment twice a day) and if skin is crusty, debride loose matter only. Do not rub.
   4. Penicillin VK 500 mg qid for 2-4 weeks and then tid for 2-3 months, bid at least 6 months or more. Any flares, go back to four a day. If patient is doing very well, decrease to one or two a day indefinitely, for the next one or two years. Cephalexin 500 mg with the same dose may also be used. (For intermittent flares, bump up the dose to 500 mg qid.)

B. For the edema:
1. A brief course of prednisone or prednisolone starting at 20-30 mg in the morning for 2-3 weeks and then decreased gradually. Length of use of Prednisone depends on the response. Patients who flare acutely may require 30 mg per day for 1-2 weeks, then 15 mg per day for 1-2 weeks.
   Chronic edema may require 20-30 mg per day for 2-3 weeks and a slow stretched-out course over 2-3 months, dropping 2.5 mg every 1-2 weeks.

2. If the edema is very indurated and woody use intraleisional triamcinolone acetonide 10 mg/mL (Kenalog 10") instead of the oral steroid to soften or get rid of fibrosis.
   a. Anesthetize the keratinized skin for one hour with topical EMLA or equivalent under occlusion.
   b. If it is somewhat woody / indurated start with 10 mg and if quite woody use up to 40 mg total dose monthly (over large surface area). Inject with a 25-26g needle and use about a 1 cm grid. Inject into the subcutaneous tissue just enough to blanch the area. To soften this chronic lymphedema you can utilize it once a month.

3. Lymphatic massage:
   This may be helpful for the vulva and for the lower legs. Some physiotherapists are trained to teach the patient how to do this at home, depending on the complexity of the problem.

4. For lymphangiomas that remain open and draining:
   A. For extensive involvement excisional surgery to debulk the area may be necessary.
   B. For more localized involvement or those for whom surgery is not an option -
      a. Once you have the infection down and controlled then you can safely use the local anesthesia as above – 2.5% lidocaine 2.5% prilocaine in a cream base (EMLA) apply every ½ hour for 1 to 2 hours under occlusion, then local anesthesia 2% lidocaine with epinephrine.
      b. To cleanse the area do not use alcohol.
      c. Electrodesiccate on a high setting and put the needle into each one of the small lymphangiectatic "fish eggs" and cauterize them until they bubble, turn black and crust.
      d. Post-operatively:
         Soak in tepid water 1-2 times a day
         Mupirocin ointment 2 times a day to involved areas.
         Keflex 500 mg qid for 2 weeks then chronic penicillin VK
         Tylenol #2 for pain (acetaminophen and codeine)
         Loose ventilated clothing.
         Consider fluconazole suppression.
         Repeat the destruction when and if needed.

ULCERS OF THE VULVA

Ulcers of the vulvar are diagnostically challenging. It is often very difficult to differentiate them from erosions. Erosions involve loss of the epidermis only, not the dermis, and they appear as deep red, often weeping, patches. Ulcers are deeper, extending into the dermis with a white or yellowish fibrinous base. Most diseases produce either erosions or ulcerations but often these overlap. Erosions can be transformed into ulcers by secondary infection, irritant contact dermatitis, rubbing and other trauma.
The best example is severe herpes simplex virus (HSV) infection. The primary lesion of HSV is an intraepidermal vesicle that becomes a pustule that ruptures, creating an erosion. When severe, these erosions can ulcerate. An ulcer is characterized by loss of both epidermis and dermis.

A diagnosis of a vulvar ulcer based on morphology alone is erroneous 40% of the time. Laboratory testing is usually required.

**DIFFERENTIAL DIAGNOSIS**

In sorting out these conditions, try to identify the primary process. Is it a pustule within the epidermis as in candidiasis or herpes simplex, an intraepidermal vesicle in acute eczema (contact dermatitis), or a frank bulla (intraepidermal or sub-epidermal) as in the bullous diseases or drug eruptions. All these rupture, resulting in erosions and/or ulcerative disease.

All of these can look much alike and it can be very difficult to differentiate them clinically, especially if there are secondary changes with crusting and bleeding, etc.

A good history is important, as is the understanding that the history may be inaccurate. Many women have problems with discussing the genital area.

Note the following factors:

| Age | Immune status |
| Epidemicology and demographics of their community | Systemic disease |
| Travel and sexual exposure | History of abuse |
| Pattern of recurrence | Previous sexually transmitted diseases |
| Previous/present treatment | 

Note the following factors specific for vulvar ulcers:

| Pain | Systemic symptoms |
| Induration | Lymphadenopathy |
| Friability | Fever |
| Number of lesions (single or multiple) | Malaise |
| Acute or chronic | Headache |
| Speed of onset | Extragenital changes |

Tests for all ulcers:  
- HSV culture  
- Candida cultures  
- CBC  
- RPR (syphilis screen)  
- HIV screen  
- Serology as indicated (for Epstein Barr virus (EBV)) - antiviral capsid antigen – IgM for EBV and Serology for Mycoplasma Pneumoniae

- Biopsy for H&E +/- immunofluorescence

Consider more extensive workup depending on the case, e.g. cultures, smears and serology.

**Biopsies** are very important. Always biopsy the edge of the lesion – not the necrotic center. A wedge excision of the edge often gives the best information for the pathologist but may be impractical. Two smaller punch biopsies may be more appropriate.
Why biopsy? Because it is impossible to guess the cause of most ulcerative erosive conditions – biopsy gives the most information, especially for chronic ulcers. Although it is an uncomfortable procedure it can be made almost painless. One is adding an extra open area to an already tender area but your patient is already very stressed and wants to know the answer.

Most common causes of primary vulvar ulcers (not erosions):

INFECTION

<table>
<thead>
<tr>
<th>Venereal</th>
<th>Non Venereal</th>
</tr>
</thead>
<tbody>
<tr>
<td>Herpes simplex (HSV)</td>
<td>EBV</td>
</tr>
<tr>
<td>Immunosuppressed</td>
<td>Mycoplasma pneumoniae</td>
</tr>
</tbody>
</table>

Chancroid
Granuloma inguinale
Lymphogranuloma venereum
Syphilis
Human immunodeficiency virus

NON-INFECTION

| Aphthous ulcers | Basal cell carcinoma |
| Behçet’s disease | Squamous cell carcinoma |
| Crohn’s disease | |
| Factorial disease | |
| Fissures | |

The infectious ulcers are classically due to the STIs. The most common cause of genital ulcers in the world is herpes simplex. HSV in any Immunosuppressed patient can present with ulcers. These can be chronic, severe, punched out, and widespread. These are typically seen in a HIV positive individual. The other conditions are syphilis, Chancroid, granuloma inguinale and rarely Lymphogranuloma venereum. These conditions are all quite uncommon in North America. Much more common are the non-infectious ulcers, particularly aphthae, which classically present as punched out, painful ulcers. They are mostly idiopathic but they can be associated with underlying conditions, see below. Aphthous ulcers are also seen in Behçet’s disease, Crohn’s disease and HIV. Crohn’s disease may present with the deep classic “knife-cut” type ulcers. Pyoderma gangrenosum can cause ulcers. Last in this group are the factitial ulcerations. Tumors, classically squamous cell carcinoma, also ulcerate.

The limitation to this classification is the possibility of missing the less common conditions that could cause vulvar ulcers and erosions such as drugs, irritant contact dermatitis, secondary infected bullous diseases etc.

2. Etiologic classification vulvar ulcers and erosions:

INFECTION

<table>
<thead>
<tr>
<th>Venereal</th>
<th>Non-venereal</th>
</tr>
</thead>
<tbody>
<tr>
<td>Herpes simplex</td>
<td>Candida</td>
</tr>
<tr>
<td>Chancroid</td>
<td>Herpes zoster</td>
</tr>
<tr>
<td>Granuloma Inguinale</td>
<td>Varicella</td>
</tr>
<tr>
<td>Lymphogranuloma venereum</td>
<td>Hand Foot Mouth disease</td>
</tr>
<tr>
<td></td>
<td>Tuberculosis</td>
</tr>
<tr>
<td>Non-Bullous Dermatoses</td>
<td>Bullous Dermatoses</td>
</tr>
<tr>
<td>------------------------</td>
<td>------------------</td>
</tr>
<tr>
<td>Irritant contact dermatitis</td>
<td>a) Autoimmune BMM Pemphigoid P. vulgaris Bullous pemphigoid Linear IgA Disease EB Acquisita b) Non-autoimmune TEN / EM Contact Dermatitis Hailey-Hailey EB Inherited</td>
</tr>
</tbody>
</table>

*12 meds for known to cause a drug reaction

Antibiotics
- Sulfa
- PCN (not as much trouble as before (no polymers attached) cephalosporins

Cardiovascular
- HCTZ
- Lasix
- Beta blockers
- Ace inhibitors
- Dilantin

Miscellaneous
- Allopurinol
Vaccines
New biologicals
NSAIDs

Of all this list, the most important causes of ulcers and erosions are, in North America are:

**Infections**

<table>
<thead>
<tr>
<th>Venereal</th>
<th>Non Venereal</th>
</tr>
</thead>
<tbody>
<tr>
<td>HSV</td>
<td>Candida</td>
</tr>
<tr>
<td>Syphilis</td>
<td>EBV</td>
</tr>
<tr>
<td>HIV</td>
<td>M Pneumoniae</td>
</tr>
</tbody>
</table>

**Dermatoses**

<table>
<thead>
<tr>
<th>Bullous</th>
<th>Non-Bullous</th>
</tr>
</thead>
<tbody>
<tr>
<td>Contact dermatitis</td>
<td>Aphthosis</td>
</tr>
<tr>
<td></td>
<td>LS</td>
</tr>
<tr>
<td></td>
<td>LP</td>
</tr>
<tr>
<td></td>
<td>Drug</td>
</tr>
<tr>
<td></td>
<td>Contact</td>
</tr>
<tr>
<td></td>
<td>Crohn’s</td>
</tr>
</tbody>
</table>

**Tumors** Squamous Cell Carcinoma

**APHTHAE** (aphthous ulcers)
- Canker sores on the vulva
- Very common in the mouth and not uncommon on the vulva
- Acute painful ulcer or ulcers of sudden onset
- Can be recurrent or chronic
- Minor or major in size, single or multiple

Painful, non-sexually transmitted ulcers in young girls or women are referred to by may terms and there is no consensus on best term. See list below:
- Ulcus vulvae acutum
- Lipschütz ulcers
- Nonsexually acquired genital ulceration (NSAGU)
- Complex Aphthosis or aphthae
- Vulvar aphthous ulcers
- Acute vulvar ulcers

Clinical:
- Average age is 14 (9-19) yrs, but patients can be older
- 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
  - There is not always a prodrome especially with recurrent cases in older patients
- Duration 1-3 weeks, can last months
One episode, less common recurrent
Often past history of oral aphthae – canker sores
Not Behçet’s
  Associated with oral aphthae – complex aphthae

The following associations have been made:
**Acute (more common)** – these can recur
  Usually with a prodrome - fever, headache, malaise, GI upset
  These have been reported in the literature associated with:
  EBV, Mycoplasma pneumoniae, viral upper respiratory infection
  (parvovirus, influenza, paramyxovirus) or gastroenteritis, Strep, CMV,
  Mumps, salmonella, toxoplasma gondii

**Chronic or recurrent aphthae:**
  No prodrome.
  Associations:
  Bowel disease - Crohn disease, Ulcerative colitis, Celiac disease
  Infections – HIV
  Behçet’s disease
  Medications – cytotoxic, NSAIDs
  Myeloproliferative disease, cyclic neutropenia, lymphopenia

**Syndromes with Genital Aphthous Ulcers:** rare
  Sweet’s syndrome
  Mouth and Genital Ulcers Inflamed Cartilage - MAGIC Syndrome
  Periodic Fever, Aphthae, Pharyngitis, Adenitis - PFAPA Syndrome

Note Acute aphthae are probably immune complex related and can be precipitated by infection such as a viral illness. e.g. viral gastroenteritis or upper respiratory tract infection, influenza, CMV. Epstein Barr virus (EBV) could directly infect the skin or cause an immune complex reaction. Mycoplasma pneumoniae can do the same. Streptococcal infection has been found. Most common cause of acute onset aphthae in a 12-20 year old is probably an infection.
For recurrent aphthae and complex aphthosis look for inflammatory bowel disease or, less likely, a lymphoproliferative problem.

**Diagnosis of exclusion**
  Cultures negative, biopsies non-specific and
  blood work non-contributory

Differential diagnosis:
  - HSV, Syphilis, HIV, Chancroid, LGV, Granuloma Inguinale
  - pyoderma gangrenosum
  - trauma
  - contact dermatitis
Evaluation of Vulvar Aphthae:
Thorough history and physical – eye, oral, genital
Only testing for HSV may be necessary

**Biopsy rarely needed**
Lab tests **that could be considered**–
- CBC, diff
  - Serology for HSV, HIV, EBV, syphilis, CMV, *Mycoplasma pneumoniae*
  - Influenza – swab PCR
  - **HSV - swab for PCR – always rule out HSV**
    - For strep -throat swab and antistreptolysin O titer
    - Tests as indicated for – paratyphoid and typhoid (stool, blood culture), TB enterocolitis, Yersinia

GI investigations –
  - for inflammatory bowel disease and celiac disease
Note – in HIV + patients with genital ulcers - 60% of genital ulcers
  - are due to aphthae and 40% to HSV

Treatment: depends on the severity. If mild comfort measures may be all that is needed
Local therapy  AGNO3 sticks
Pain control – topical – 5% lidocaine ointment
  - systemic – mild, moderate pain – NSAID  severe - opioids
Immunosuppression -
  - Prednisone 40 – 60 mg each morning until pain resolves (3-7 days, then ½ dose 3-7 days) with food
  - Methylprednisolone (Medrol) 4-8mg bid-tid 3-7 days then ½ dose 3-7days) with food
  - Clobetasol or halobetasol 0.05% ointment AM & PM
  - If not sure if HSV use antiviral meds until HSV test report available.
Educate -Most often a one-time event, can recur

For persistent or chronic aphthae:
Oral corticosteroid for initial control - prednisone or methylprednisolone
Intralesional triamcinolone (Kenalog 10) 5-10 mg/ml
doxycline 50 - 100 mg od
colchicine 0.6 mg bid-tid if tolerated
dapsone 50-150 mg per day
dapsone + colchicine
cyclosporine  100 mg up to tid decreasing to 100 mg 2-3 doses/week
pentoxyfylline 400 mg tid
thalidomide 100-150 mg per day (Concern for teratogenesis)
TNF alpha inhibitors- infliximab, adalimumab, etanercept

Prognosis:
Most often a one-time event
Scarring can occur
Occasionally recurrent
**Desquamative Inflammatory Vaginitis (DIV)**

Desquamative inflammatory vaginitis (DIV) is an erosive vulvovaginitis characterized by dyspareunia, and a profuse purulent vaginal discharge. There is significant vaginal cell exfoliation. Numerous parabasal cells are seen in vaginal smears, as well as large numbers of neutrophils (neutrophils/epithelium > 1:1 in at least 4 HPFs on wet smear). The pH is increased (> 4.5). Lactobacilli are decreased or absent, and there is often increased gram positive cocci and gram negative bacilli.

When the speculum is inserted, fine red “dots” may be present in the vagina. Vaginal lichen planus can present with this appearance, as can atrophy. Can be seen commonly in Crohn’s disease. Rarely it can be seen with the chronic bullous diseases – cicatricial or classic Pemphigus.

**Treatment**

The treatment varies among providers. Some prefer intravaginal clindamycin, while others prefer intravaginal steroids such as hydrocortisone in 25 mg doses. Some providers combine the clindamycin and hydrocortisone per vagina.

Below is a treatment regimen that you might consider:

- Clindamycin 2% cream; 1 applicator per vagina, qhs x 14 days as initial therapy

If that fails, try using clindamycin 2% per vagina (1 applicator) combined with a 25 mg hydrocortisone suppository per vagina every other night x 14 doses.

When the patient does not respond to the above treatments consider:

- Hydrocortisone 100 mg/gram in clindamycin 2% emollient cream base
- Insert 5 gram (applicator full) per vagina q.o.d. (at night) x 14 doses
- If recurrent, when controlled, decrease to 3 times a week and slowly decrease and stop

**HERPES SIMPLEX VIRUS (HSV) (adapted from CDC STD Treatment Guidelines 2010)**

This is a common sexually transmitted disease worldwide and it is the most common cause of vulvar ulcers. A history of HSV is unreliable. Primary HSV is uncommon. The majority of patients present with non-primary recurrent disease. Etiology: 80% HSV 2 (Genital HSV 1 increasing with more “oral sex” up to 35%)

- HSV 1 recurs in 25% of cases
- HSV 2 recurs in 89% of cases

Infection is usually from sexual contact. Most transmission occurs during periods of asymptomatic viral shedding. Most persons infected with HSV-2 have not been diagnosed with genital herpes. Many people have mild or unrecognized infections but they shed the virus intermittently in the genital tract. Thus, the majority of genital herpes infections are transmitted by persons unaware that they have the infection or who are asymptomatic when transmission occurs.
Symptoms:
Primary HSV - Paresthesia for 2-3 days, followed by fever, malaise, headache and myalgia
There can be pain, moderate to severe (“deep boring pain” reflecting nerve involvement)
Recurrent infection - there is more tingling, itching and burning before the onset of vesiculation

Physical Examination
Can be seen anywhere on the vulva, vagina, over cervix, anus, buttocks and thighs.
Primary – red swollen vulva with extensive vesiculation, rapidly becoming pustular with open tender erosions lasting two weeks.
Recurrent infection – lesions are less extensive and are clear in 5-7 days with only mild swelling.

Note – 91% of HSV 2 carriers are unaware of their infection yet 80% have symptoms.
Women think they have: Vaginitis, GU infection, clothing irritation or hemorrhoids
Symptoms can occur with no rash and no blistering in HSV sine eruption – herpes simplex without visible eruption

Immunosuppressed HSV – chronic ulcers that gradually extend at the periphery. There may be varying degrees of necrosis. These are painful and indolent.

Diagnosis: Cultures can be unreliable. Keep viral media refrigerated, on hand, and up to date. Failure to detect HSV by culture or PCR does not indicate an absence of HSV infection, because viral shedding is intermittent.
Scrape the base of an early lesion with 15 blade to get adequate material. The sensitivity of viral culture is low, especially for recurrent lesions, and declines rapidly as lesions begin to heal.

Virologic Tests
Cell culture and PCR are the preferred HSV tests
PCR assays for HSV DNA are more sensitive and are increasingly used in many settings
Viral culture isolates should be typed to determine which type of HSV is causing the infection.
The use of cytologic detection of cellular changes of HSV infection is an insensitive and nonspecific method of diagnosis, both for genital lesions (i.e., Tzanck preparation) and for cervical Pap smears and therefore should not be relied upon.

Type-Specific Serologic Tests
Both laboratory-based assays and point-of-care tests that provide results for HSV-2 antibodies from capillary blood or serum during a clinic visit are available. The sensitivities of these glycoprotein G type-specific tests for the detection of HSV-2 antibody vary from 80%–98%, and false-negative results might be more frequent at early stages of infection. The specificities of these assays are $\geq 96\%$. False-positive results can occur, especially in patients with a low likelihood of HSV infection. Repeat or confirmatory testing might be indicated in some settings, especially if recent acquisition of genital herpes is suspected.

IgM testing for HSV is not useful
Since nearly all HSV-2 infections are sexually acquired, the presence of type-specific HSV-2 antibody implies anogenital infection. The presence of HSV-1 antibody alone is more difficult to interpret. Most persons with HSV-1 antibody have oral HSV infection acquired during childhood, which might be asymptomatic. However, acquisition of genital HSV-1 appears to be increasing, and genital HSV-1 also can be asymptomatic. Lack of symptoms in an HSV-1 seropositive person does not distinguish anogenital from orolabial or cutaneous infection, and regardless of site of infection, these persons remain at risk for acquiring HSV-2.

Type-specific HSV serologic assays might be useful in the following scenarios: 1) recurrent genital symptoms or atypical symptoms with negative HSV cultures; 2) a clinical diagnosis of genital herpes without laboratory confirmation; or 3) a partner with genital herpes. HSV serologic testing should be considered for persons presenting for an STD evaluation (especially for those persons with multiple sex partners), persons with HIV infection, and MSM at increased risk for HIV acquisition.

Differential diagnosis:
- Syphilis, chancroid, aphthous ulcers, Herpes zoster, HIV

Note – patients with HIV can have vulvar ulcers. 60% are due to aphthous ulcers alone. The other 40% are due to HSV. Always look for multiple or atypical infections in these patients.

Non-specific treatment for pain, discomfort etc. R/O other STD’s
Treatments for the relief of discomfort
The following non-specific treatments can alleviate the pain and discomfort of genital sores.

- **SALT BATHS** (1 teaspoon of salt in 600 ml of water or a handful in a shallow bath) can be used to wash, soothe and dry the sores.
- **PAIN RELIEVERS**
- **LOOSE UNDERCLOTHES**, preferably cotton (not nylon), can help minimize discomfort and allow healing.
  For anyone experiencing extreme pain when urinating, the process can be less painful when done in a cool bath. Encourage plenty of fluids to dilute the urine.

NEW CDC STD TREATMENT GUIDELINES WERE RELEASED IN WINTER 2010

http://www.cdc.gov/std/treatment/2010/

**First Clinical Episode of Genital Herpes**

The initial genital herpes episode can last for more than 20 days. Symptoms include tingling, itching, burning or pain. Patients may experience a range of generalized symptoms, such as fever, aches and pains, swollen lymph nodes, as well as specific vulvar symptoms. For others, the initial infection can be mild with minimal symptoms. They may be totally unaware that they have had a herpes outbreak. The severity of symptoms for genital herpes varies in the population. The initial episode can be so mild as to pass unnoticed and a first recurrence may take place many years after the first infection.

Many patients with primary herpes present with mild clinical manifestations but later develop severe or prolonged symptoms. Newly acquired genital herpes can cause a prolonged clinical illness with severe
genital ulcerations and neurologic involvement. Even persons with first-episode herpes who have mild clinical manifestations initially can develop severe or prolonged symptoms. Therefore, all patients with first episodes of genital herpes should receive antiviral therapy.

First Clinical Episode of Genital Herpes  Recommended Regimens

**Acyclovir** 400 mg orally three times a day for 7-10 days,

*OR*

**Acyclovir** 200 mg orally five times a day for 7 -10 days,

*OR*

**Famciclovir** 250 mg orally three times a day for 7-10 days,

*OR*

**Valacyclovir** 1 g orally twice a day for 7-10 days.

**NOTE:** Treatment may be extended if healing is incomplete after 10 days of therapy.

**Recurrent Episodes of HSV Disease** Most patients with symptomatic, first-episode genital HSV-2 infection subsequently experience recurrent episodes of genital lesions; recurrences are much less frequent following initial genital HSV-1 infection. Antiviral therapy for recurrent genital herpes can be administered either continuously as suppressive therapy to reduce the frequency of recurrences or episodically, to ameliorate or shorten the duration of lesions.

**Suppressive Therapy for Recurrent Genital Herpes**

Suppressive therapy reduces the frequency of genital herpes recurrences by 70% to 80% among patients who have frequent recurrences. Safety and efficacy have been documented among patients receiving daily therapy with acyclovir for as long as 6 years and with valacyclovir or famciclovir for 1 year.

**Recommended Regimens for Continuous Suppressive Therapy**

**Acyclovir** 400 mg orally twice a day,

*OR*

**Famciclovir** 250 mg orally twice a day,

*OR*

**Valacyclovir** 500 mg orally once a day,

*OR*

**Valacyclovir** 1.0 gram orally once a day. (Valacyclovir 500 mg once a day might be less effective than other valacyclovir or acyclovir dosing regimens in patients who have very frequent recurrences (i.e., ≥10 episodes per year).

**Episodic Therapy for Recurrent Genital Herpes** Effective episodic treatment of recurrent herpes requires initiation of therapy within 1 day of lesion onset, or during the prodrome that precedes some outbreaks. The patient should be provided with a supply of drug or a prescription for the medication with instructions to self-initiate treatment immediately when symptoms begin. There is a new single-day

**Episodic Therapy for Recurrent Genital Herpes Recommended Regimens**
Acyclovir 400 mg orally three times a day for 5 days
OR
Acyclovir 800 mg orally twice a day for 5 days
OR
Acyclovir 800 mg orally three times a day for 2 days
OR
Famciclovir 125 mg orally twice daily for 5 days
OR
Famciclovir 1000 mg orally twice daily for 1 day
OR
Famciclovir 500 mg once, followed by 250 mg twice daily for 2 days
OR
Valacyclovir 500 mg orally twice a day for 3 days
OR
Valacyclovir 1 g orally once a day for 5 days

Few comparative studies of valacyclovir or famciclovir with acyclovir have been conducted. The results of these studies suggest that valacyclovir and famciclovir are comparable to acyclovir in clinical outcome. Ease of administration and cost also are important considerations for prolonged treatment.

**Severe Disease**  Intravenous acyclovir therapy should be provided for patients who have severe disease or complications that necessitate hospitalization, such as disseminated infection, pneumonitis, hepatitis, or complications of the central nervous system (e.g., meningitis or encephalitis).

**Treatment Immunosuppressed Patients With HSV**

Topical imiquimod 5% cream
Usual treatment:
   - acyclovir, famciclovir, valacyclovir
   - foscarnet
cidofovir
   - imiquimod 5% cream- 3 times a week for 2-8 weeks
Molluscum contagiosum
Molluscum contagiosum is caused by a DNA poxvirus. The disease is more prevalent in children (lesions involve the face, trunk and extremities). Adults tend to have lesions most often near the genital areas. The incidence of molluscum has increased over the last 30 years. There are four main subtypes of molluscum contagiosum virus (MCV), MCV I, MCV II, MCV III and MCV IV. The disease is transmitted by direct skin contact. It presents clinically with a papular eruption of multiple umbilicated lesions. The central depression contains a white waxy curd-like core. The size of the papule ranges from 2-6 mm. The clinical appearance of molluscum contagiosum is the general diagnostic method, though it can be examined histologically (curetted or biopsied lesion). Large brick shaped inclusion bodies are seen. In-situ hybridization for MCV DNA has also been performed.

Treatment of molluscum contagiosum
Molluscum contagiosum is a self-limited disease, which will generally resolve in immunocompetent hosts. However, the time to resolution can be quite long. Treatment of molluscum contagiosum is advisable in healthy individuals to prevent autoinoculation or transmission.

Common treatments for molluscum
- Cryosurgery (liquid nitrogen, dry ice)
- Evisceration (scalpel, IV needle)
- Curettage
- Tape stripping
- Podoflox
- Imiquimod 5% cream
- TCA

Condyloma accuminata
Genital warts are caused by the human papillomavirus (HPV), of which more than 200 subtypes exist, over 30 that are found on the genital area. The diagnosis is usually based on clinical appearance. They are soft in texture, nonpigmented and usually asymptomatic. At times they cause itching, bleeding and occasionally pain. They may involve the anus too. Of genital warts, 90% are caused by HPV 6 or 11.
Numerous treatments exist (2010 CDC STD Treatment Guidelines).

**Recommended Regimens for External Genital Warts**

*Patient-Applied:*

Podofilox 0.5% solution or gel (preferred in transplant patients)

OR

Imiquimod 5% cream

OR

Sinecatechins 15% ointment (preferred in transplant patients)

*Provider–Administered:*

Cryotherapy with liquid nitrogen or cryoprobe. Repeat applications every 1–2 weeks.

OR

Podophyllin resin 10%–25% in a compound tincture of benzoin

OR

Trichloroacetic acid (TCA) or Bichloroacetic acid (BCA) 80%–90%

OR

**Surgical removal** either by tangential scissor excision, tangential shave excision, curettage, or electrosurgery.

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**Vulvar Neoplasia**

**Benign Cysts and Tumors**

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<tr>
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<td>Cyst of canal of Nuck (hydrocele)</td>
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Syringomas
Syringomas are often associated with itching. There are a number of treatment options for itchy syringomas:

1. Atropine 1% aqueous solution (5 mL bottle) Apply 2-4 drops at a time (20 drops to 1 ml so that would last about 3 weeks).
2. Destruction which can either be electrodessication or laser CO laser destruction.
3. Tretinoin can be given as a 0.025 or 0.05% cream but it can be a bit irritating. Oral isotretinoin or Accutane has been reported to be helpful.
4. Steroids topically with antihistamines have been used but notoriously give poor results.
5. Tranilast (brand name Rizaben) is an anti allergenic drug used in Asia for bronchial asthma. It has been reported to be helpful. It seems to block macrophages. The dosage is 300 mg po daily (in a report out of Japan).
6. Topical glycopyrrolate 0.1% in a compounded topical cream has been used. This stops sweating and has been helpful in patients that sweat a great deal in the vulva area and that potentially might be helpful. It is used daily.

Intraepithelial Neoplasia
VULVAR INTRAEPITHELIAL NEOPLASIA (VIN) OLD TERMINOLOGY
1. Squamous type (with or w/o HPV change)
   a. VIN I
   b. VIN II
   c. VIN III (Squamous cell CIS, Bowen's disease, Erythroplasia of Queyrat, CIS simplex)
2. Nonsquamous type
   Paget’s disease
   Melanoma in Situ

VIN TERMINOLOGY
SQUAMOUS VIN TERMINOLOGY (ISSVD 2004)
VIN, usual type
   VIN, warty type
   VIN, basaloid type
   VIN, mixed (warty/basaloid) type
VIN, differentiated type
   Note: The occasional example of VIN that cannot be classified into either of the above VIN categories (usual type and differentiated type) may be classified as VIN, unclassified type (or VIN, NOS). The rare VIN of pagetoid type may be classified as such, or placed in this category.

2012 Lower Anogenital Squamous Terminology (LAST) Project
New terminology regarding the histopathologic nomenclature system that reflects current knowledge of HPV biology, optimally uses available biomarkers, and facilitates clear communication across different medical specialties was developed in 2012. The Lower Anogenital Squamous Terminology (LAST) Project was cosponsored by the College of American Pathologists and the American Society for Colposcopy and Cervical Pathology met and published the terminology to use across all lower genital tract sites, including
the vulva. A two tiered nomenclature was recommend, consisting of LSIL and HSIL. However, this does not generally refer to VIN differentiated (most often non HPV related), which must be considered, especially in patients with lichen sclerosus.

Before 1970, VIN was found most often in women in the fifth or sixth decade of life; currently about half of the patients are less than 40 years old. VIN in young women is frequently in multiple locations and is associated with HPV. Currently, approximately 80% of patients with VIN are HPV positive. Patients may be asymptomatic or complain of pruritus or burning.

Treatment: Biopsy before any therapeutic trial is initiated.
- Smoking cessation may be necessary for the methods below to succeed
- Wide local excision in hair bearing tissue is recommended

1. Local: Scalpel
   a. Standard procedure: an inked margin around the lesion is made providing gross clearance (0.5 cm to 1 cm) at resection. The depth of resection is to the subcutaneous fat but not deeper. Closure depends on the size of resection but is often by primary approximation. Smaller resections may not require closure and larger lesions may require local advancement skin flaps or grafts.

   Special points: Vulvar skin thickness varies considerably by location. Particular care must be taken in the clitoral, urethral, anal and labia minora locations as the squamous epithelium is very thin. Resections in this area don’t require deep dissection and every effort should be made to minimize trauma.

2. CO2 laser- in non hair bearing areas
   Confidence that no invasive disease exists is important to patient selection.

3. LEEP: difficult to control depth of dissection

4. Medical
   - Imiquimod (Aldara®) has reported to be effective for VIN 3 (same dosage as for condyloma)(Off label use)

It is important to screen these patients with Anal Pap smears. Use a moistened Dacron swab or Cytobrush. Insert into the canal approximately 5-6 cm above the anal verge to the rectum. Rotate, applying pressure to the walls of the canal while removing the sampling device.

Anal Cytology

Place the cytology sampling device (Dacron swab or Cytobrush moistened with water) into the anal canal until resistance is met (approximately 4 cm) Rotate/apply pressure to walls of canal while removing sampling device slowly (count to 10) Place in liquid media.
Notify Pathology (Cytology) Department before you start these, so that they are prepared for them. HPV testing is not required on these specimens.

Anal colposcopy
After the cytology has been obtained, place an anoscope (use a clear plastic anoscope) with a small amount of lubricant into the anus. Then place an opened 4 x 4 soaked in 3 to 5% acetic acid over a cotton swab
through the anoscope. Remove the anoscope, leaving in the 4 x 4 and cotton swab. Place a 4 x 4 with 3 to 5% acetic acid around the outer anus. Leave these on for about 3 minutes. Then, remove the 4 x 4’s and cotton swab and reinsert a lubricated anoscope. The anus is visualized sequentially, with a colposcope, keeping in mind the location of the dentate line.

Paget’s Disease of the Vulva

Primary extramammary Paget’s Disease – an epidermotropic carcinoma arising within the epidermis or epidermal appendages (may arise in Toker cells) – no underlying carcinoma (most common)

Secondary extramammary Paget’s disease – is a visceral carcinoma (anorectal, bladder or urethra) that is epidermotropic to the skin.

Clinical Presentation:

Itching “rash” on perineum with eczematous, soft velvety papules slowly growing into crusty scaly plaques that do not respond to topical steroid

Paget's disease of the skin is generally confined to the integument along the mid line. It occurs most commonly on the nipple and areola, where its presence signifies an underlying adenocarcinoma of the breast. Extramammary lesions have been described in the genital, perianal, and axillary regions as well as the ear canal, all of which contain abundant apocrine glands.

Vulvar Paget's disease appears as a red velvety area with white islands of hyperkeratosis and at times may be pinkish and eczematoid. It primarily occurs on the labia majora. Pruritus is present in over half of the patients. The mean age for Paget’s disease of the vulva is 65 years. Almost all of the patients are Caucasian.

Signs
- Red and white vulva - ulceration and hyperkeratosis
- Well demarcated
- Eczematoid

Symptoms
- Pruritus in over 50%
- Soreness
- Bleeding or discharge

When present on the vulva, it is most commonly an intraepithelial disease that tends to recur locally and has a minimal propensity to invade. Usually it is a slowly progressive, indolent, superficial process. It is rarely associated with an underlying skin appendage carcinoma such as a primary carcinoma of the rectum, urethra, or bladder

Only about 25% of vulvar Extramammary Paget’s are associated with an underlying adenocarcinoma of an adnexal tissue or a Bartholin gland. Less commonly it is associated with a distant carcinoma of breast, GI, GU or the genital tract. Perianal Extramammary Paget’s is associated with underlying colorectal adenocarcinoma in 80% of cases. In view of the possible coexistence of sweat gland carcinoma of the vulva or another adjacent internal carcinoma, the overall prognosis for Paget's disease is less favorable than for VIN III. Clinical diagnosis based on gross appearance may be erroneous. Biopsy confirmation of the diagnosis is mandatory. Large, irregular Paget's cells containing clear, vacuolated pale cytoplasm are seen on histologic evaluation. Nuclei are vesicular. Mitotic figures are uncommon. Paget cells are most numerous in the tips and sides of the rete pegs and deep in the epithelium. They may be scattered throughout the outer keratinized layer. Paget cells, as well as the cells and secretions of normal eccrine and apocrine glands are rich in CEA.
Markers
The immunoprofile of vulvar Paget's disease includes cells that are typically positive for cytokeratin 7, keratin CAM5.2, EMA, CEA and GCDFP; mucin stains are also positive in a subset of the neoplastic cells (less cost).

Work up to detect associated adenocarcinoma (location dependent)
- H+P
- Pap
- Mammogram
- Hemoccult
- Cystoscopy
- Flex sigmoidoscopy vs BE vs colonoscopy

Treatment
Paget’s disease of the vulva is generally treated with a wide local excision of the circumscribed lesions. It is important to remove the full thickness of the skin to the subcutaneous fat to be certain that all of the skin adnexal structures are excised. Even if resection margins are free of Paget's disease at the time of surgical excision, local recurrence remains a major risk. Laser therapy has been used on Paget’s disease (particularly recurrent Paget’s). On rare occasions, radiation therapy has been used to treat Paget’s disease. 5% imiquimod cream 1 to 5 times a week (frequency of application depends on tolerance) has been used for superficial involvement and when surgery would be poorly tolerated. Duration of treatment depends on response and this can be months.

Atypical junctional melanocytic hyperplasia
This is a preinvasive condition. If margins are not clear, a repeat resection should be performed.

Melanoma in situ
Clear margins should be obtained.

Malignant Tumors
Vulvar Cancer
Most vulvar cancers are found in patients age 60 to 70 years. The risk for vulvar cancer continues to increase with age. The diagnosis is often delayed (mean = 1 year). It is usually unifocal. Most vulvar cancers are squamous cell carcinomas.
- Squamous carcinoma –87%
- Melanoma-6%
- Bartholin's Adenocarcinoma-4%
- Basal Cell carcinoma <2%
- Sarcoma <2%
2009 FIGO Staging System for Vulvar Cancer

**Carcinoma of the vulva.**

Stage I Tumor confined to the vulva
- IA Lesions ≤ 2 cm in size, confined to the vulva or perineum and with stromal invasion ≤ 1.0 mm, no nodal metastasis
- IB Lesions > 2 cm in size or with stromal invasion > 1.0 mm, confined to the Vulva or perineum, with negative nodes

Stage II Tumor of any size with extension to adjacent perineal structures (1/3 lower urethra, 1/3 lower vagina, anus) with negative nodes

Stage III Tumor of any size with or without extension to adjacent perineal structures (lower urethra, lower vagina, anus) with positive inguino-femoral lymph nodes
- IIIA (i) With 1 lymph node metastasis (≥ 5 mm), or
  (ii) 1–2 lymph node metastases(es) (< 5 mm)
- IIIB (i) With 2 or more lymph node metastases (≥ 5 mm), or
  (ii) 3 or more lymph node metastases (< 5 mm)
- IIIC With positive nodes with extracapsular spread

Stage IV Tumor invades other regional (2/3 upper urethra, 2/3 upper vagina) bladder mucosa, rectal mucosa, or distant structures
- IVA Tumor invades any of the following:
  (i) upper urethral and/or upper vaginal mucosa, bladder mucosa, rectal mucosa; or fixed to pelvic bone
  or
  (ii) fixed or ulcerated inguino-femoral lymph nodes
- IVB Any distant metastasis including pelvic lymph nodes

a. The depth of invasion is defined as the measurement of the tumor from the epitheliostromal junction of the adjacent most superficial dermal papilla to the deepest point of invasion.

**Melanoma**

**Clinical Features**

Lesions suspicious for melanoma are often characterized by the ABCDs. They are asymmetrical (A), have irregular or scalloped border (B), often black in color (C) or variegated with shades of red, white, or blue, and
may have a diameter (D) greater than 6 millimeters. However, a melanoma with all or many of these characteristics is often more advanced. For melanoma, the earliest signs are change in size, shape, and color of a lesion. The earliest symptom is persistent pruritus in a lesion. Late features include bleeding, ulceration, pain, and tenderness. The most common presenting signs/symptoms for vulvar melanoma are bleeding, mass, discharge, pruritus, burning, pain, ulceration, foul odor, local discoloration, and dysuria. Vulvar melanoma is usually detected later than cutaneous melanoma simply due to location, resulting in more advanced lesions at presentation with a poorer prognosis. Melanoma may occur in any vulvar location and has been reported as multifocal in up to 20% of cases, arising in the scattered background of malignant and premalignant melanocytic cells.

**Biopsy**

The first step for evaluating any lesion suspicious for melanoma is biopsy. Excisional biopsy is optimal, with narrow margins of 1-2 mm to provide the pathologist a complete specimen for accurate diagnosis and microstaging. Wide excision for diagnosis is not recommended in the event that sentinel lymph node biopsy (SLNB) is indicated. Sentinel node biopsy may not be as accurately performed following a wide local excision. The biopsy may be performed as an elliptical excision, punch, or saucerization (a modified shave technique extending to the adipose tissue. A superficial shave biopsy is never recommended due to the risk of transecting the lesion, which precludes accurate measurement of tumor thickness, the most important factor that determines treatment. If the lesion is too large or too close to vital structures to excise completely, an incisional biopsy may be performed. Several studies have documented that an incisional biopsy for melanoma does not increase the risk of tumor seeding, metastasis, or decrease survival. If the incisional biopsy reveals a melanocytic lesion with melanoma still in the differential diagnosis, complete excision of the lesion is still indicated due to possibility of biopsy sampling error. The biopsy should be interpreted by a pathologist with experience in the interpretation of pigmented lesions and melanoma. Results from large melanoma centers show that up to 20% of patients referred to a specialized melanoma center have significant alterations in diagnosis and/or microstaging parameters following review by a pigmented lesion pathology expert. The two most important factors in the cutaneous melanoma pathology report are tumor thickness measured in millimeters (Breslow depth) and ulceration status. Other potentially important factors include mitotic rate, microsatellitosis, angiolymphatic involvement, Clark level (anatomic measure of thickness), neurotropism, and extensive regression. Additional factors routinely documented in the pathology report include a gross and microscopic description of the specimen, histologic pattern subtype, tumor infiltrating lymphocytes, growth phase, presence of preexisting nevus, age, gender, anatomic site, and margin involvement. The most common histologic pattern subtype reported for vulvar melanoma is acral mucosal lentiginous; for cutaneous melanoma superficial spreading is the most common. The histologic subtype, in general, does not correlate with prognosis or survival after correction for tumor thickness, ulceration, and other prognostic parameters.


**Summary**

An overview of the different types of vulvovaginal conditions has been given. Many vulvar conditions must be considered when a patient complains of discharge and itching. It is important to remember that

**IF TREATMENT IS NOT WORKING, RECONSIDER THE DIAGNOSIS.**
Prescriptions for Vulvar Disease

**Pain Medications**

**Xylocaine® (lidocaine)**
- 5% Xylocaine® (lidocaine) ointment
- sig: apply to vulva prn
- Disp: 35 grams

**Elavil® (amitriptyline)**
- Start low and increase dose slowly.
- Initial amitriptyline prescription:
  - amitriptyline 10-25 mg
  - Sig: 1 po qhs x 1 week; If sx s persist, 2 po qhs x 1 wk; if sx s persist, 3 po qhs x 1 wk; if sx s persist, 4 po qhs. Maintain nightly dose that relieves symptoms (Generally not to exceed 4 po qhs) Do not stop suddenly
- Start at 10 mg in patients age 60 or older; increase by 10 mg weekly

- Future amitriptyline prescriptions
  - Amitriptyline ____mg
  - Sig: i po qhs (comes in 10 mg, 25 mg, 50 mg, 75 mg, 100 mg and 150 mg tablets)

**Neurontin®**
- Neurontin® (gabapentin)
- Sig: 300 mg po qd x 3 days; if sx s persist, 300 mg po bid x 3 days; if symptoms persist, 300 mg po tid. Stay on this dose for a month and increase gradually if needed.
- It comes in 100, 300, 400, 600 and 800 mg doses
- Do not exceed 2700 to 3600 mg total dose per day
- Do not give more than 1200 mg in a single dose

- Gabapentin ointment 3% or 6%
- Sig: apply to affected area bid-tid
- Disp: 3 month supply

**Lyrica®**
- Lyrica® (pregabalin)
- 50 mg po qd x 4 days, if sx s persist, 50 mg po bid x 4 days, if sx s persist, 50 mg po tid
- Can gradually increase up to 100 mg po tid; doses up to 300 mg po bid have been used for pain control

**Blocks**
- Bupivacaine (0.25% or 0.5%) and Kenalog®
- Draw up Kenalog® first (40 mg /cc) (can use up to 40 mg steroid in single dose per month)
- Combine with Bupivacaine (large area use 0.25%; small area use 0.5%) Inject into specific area or use as a pudendal block
- Can be repeated monthly
- Do not use high doses on thin skin.
**Medications for localized pain or itching**

- **Zonalon® (doxepin) 5 % cream**
  Sig: apply to skin q d with gradual increase not to exceed qid
  Disp: 30 g

- **Topical Elavil® (amitriptyline) 2% with Baclofen 2% in water washable base (WWB)- squirt ½ cc from syringe onto finger and apply to affected area qd to tid**
  Disp: 30 day supply

- **Gabapentin 6% with Ketamine 5% WWB – 30mL apply ½-1 mL to Vulvar Vestibule twice daily for pain**

- **Amitriptyline 2% with Baclofen 2% WWB and Lidocaine 5% mg – 30mL Apply ½-1 mL to Vulvar Vestibule twice daily for pain**

- **Estradiol 0.1mg with Lidocaine 5% ointment – Disp 30g Apply thin layer over Vulva twice daily for pain**

**Yeast medications**

- **Fluconazole 150 mg**
  Sig: 1 po q 3 days x 3, then 1 po q week for up to 6 months (If using for longer than 6 months, check LFT’s) Do not use with active liver disease.

- **5 flucystosine 500 mg/5 grams compounded in a hydrophilic cream base**
  - Insert 5 gram applicator (500 mg of active drug) full of mixture per vagina qhs x 14 days

- **Boric acid- fill 0-gel capsule halfway (600 mg)**
  To treat active yeast infection - Insert **per vagina** nightly for 14 days
  For prevention of yeast - Insert **per vagina** twice weekly. Keep out of reach of children. Warn patients not to receive oral sex while on the boric acid. There is an herbal product called Yeast Arrest. It contains 600 mg boric acid, Oregon Grape Root and Calendula flowers.

- **Gentian violet- 0.25% or 0.5% aqueous solution applied at home daily or it may be given in the office as a 1.0% solution (once weekly for up to three times). Warn patients that if they have oral sex, their partner’s teeth and lips could stain.**

**Medications for Lichen planus**

- **Anusol HC suppository**
  1/2 of a 25 mg suppository per vagina bid x 2 months
  Decrease to qd x 2 months
  Maintenance therapy of 1 - 3 x per week

- **Hydrocortisone acetate 100 mg compounded suppository used QHS**
  Use for 2-4 weeks then use Mon Wed Fri for 2-4 weeks and change to milder 25 mg suppository as needed
Hydrocortisone acetate 10% compounded Vaginal cream used QHS –
4-5 gram q d (400 to 500 mg dose). For severe vaginal Lichen Planus
Use for 2-4 weeks then use Mon Wed Fri for 2-4 weeks and change to milder
25 mg suppository as needed

**Tacrolimus**

<table>
<thead>
<tr>
<th>For oral Lichen planus:</th>
<th>For vaginal lichen planus</th>
<th>For vulvar Lichen planus</th>
</tr>
</thead>
<tbody>
<tr>
<td>Tacrolimus 0.1% in Orabase</td>
<td>Tacrolimus vaginal suppositories</td>
<td>Tacrolimus 0.1% Ointment</td>
</tr>
<tr>
<td>Sig: apply to mouth bid</td>
<td>Insert one supp per vagina (2 mg tacrolimus per 2 gm supp) qhs</td>
<td>Sig: apply to skin bid</td>
</tr>
<tr>
<td>Disp 50 g</td>
<td>Disp 50</td>
<td>Available in 30 or 60 gram tubes</td>
</tr>
</tbody>
</table>

**Folliculitis** (swab for culture to r/o MRSA)
- Emgel® 2% topical gel (erythromycin) or 1% clindamycin lotion
- Sig: apply to skin bid
- Available in 27 or 50 gram bottles

Other topical antibiotics include bacitracin, neomycin, mupirocin

**For fungal folliculitis**
- Topical clotrimazole, miconazole
- Oral terbinafine, itraconazole, griseofulvin

**Furunculosis- very responsive to antibiotics** (swab for culture to r/o MRSA)
- Topical antibiotics (bacitracin, neomycin, mupirocin)
- Oral antibiotics (dicloxacillin, cephalxin)
- Dial soap or Phisohex
- If wrinkled, I and D useful

**For Recurrent Impetigo Staphlococcus +/- Steptococcus**
- Take bacterial culture from site of infection, nose and gluteal cleft to find any hidden source of infection.
- Do bleach baths to reducing re-infection 2-3 times a week. Add one half cup of household bleach (125 mL) to a 10 inch (25 cm) deep tub of comfortably warm bath water. **Mix well.** Soak for 5-7 minutes, ensuring penetration of the solution into all cracks and genital / buttock / skin folds, using a plastic cup and bare hands to spread over all involved areas. For sitz bath mix 1 ¼ tsp bleach in 1 gallon of water.
- Treat with oral antibiotics as indicated by culture results.
- Use an antibiotic ointment (mupirocin ointment twice a day) bid for nose or gluteal cleft. If MRSA use retapamulin 1% ointment (Altabax) bid for 5 days.

**Desquamative inflammatory vaginitis**
- Can utilize clindamycin 2% per vagina qhs x 14 days as initial therapy

If that fails, try using clindamycin 2% per vagina combined with a 25 mg Anusol HC suppository per vagina every other night x 14 doses.

Another treatment for DIV that is used when the patient does not respond to the above treatments is:
- Hydrocortisone 100 mg/gram in clindamycin 2% emollient cream base
- Insert 5 gram (applicator full) per vagina q.o.d. (at night) x 14 doses
Steroid medications

<table>
<thead>
<tr>
<th>Clobetasol propionate ointment (Temovate®) 0.05%</th>
<th>Triamcinolone acetonide ointment 0.1%</th>
</tr>
</thead>
<tbody>
<tr>
<td>Sig: apply to vulva bid x 1 month, then qd x 2 months</td>
<td>Sig: apply to vulva qd to bid</td>
</tr>
<tr>
<td>Disp: 30 g</td>
<td>Disp: 80 g</td>
</tr>
<tr>
<td>Consider decreasing gradually to Triamcinolone acetonide ointment 0.025% qd to bid</td>
<td></td>
</tr>
</tbody>
</table>

**TOPICAL CORTICOSTEROIDS**

Learn three to four ointments of different strengths, making appropriate selections as needed

- Ointments are stronger than creams
- Ointments stay on longer than creams (creams are diluted and washed away with body fluids)
- Ointments are less irritating and have fewer allergens than other bases

Patients may find one base more irritating than another. Be flexible.

Do not use steroids for dysesthetic vulvodynia - steroids work by reducing inflammation, not pain

**Note:** **Topical steroids are not a cure.** Use the steroid potency that will do the job in the quickest period of time and then decrease to a lower potency. Either stop or maintain with the lowest potency or use intermittently as necessary.

**Tips:**

- When considering topical corticosteroids, especially the superpotent types, consider:
  - There are more available than you need
  - Use them in an educated way
  - Limit the amount prescribed to 15g to 30 grams for high dose topical steroids
  - Show the patient exactly how to use it – a tiny dab spread in a thin film just to the involved area is all that is necessary
  - Vulvar mucous membrane (vulvar trigone and inner labia minora) is remarkably steroid resistant. The outside of the labia minora and the labiocrural fold and the thighs will thin easily and develop striae.
  - When the patient improves, decrease the frequency of topical steroid or manage with a low potency product.
  - Use under close supervision.
  - At any suggestion of secondary yeast infection, add a topical or oral anti-fungal.

For example, for thick itchy dermatoses like lichen simplex chronicus – use name brand clobetasol or halobetasol 0.05% ointment bid for 1-2 weeks, once a week for 1-2 weeks and then M-W-F for 1-2 weeks and for long term maintenance either infrequent and intermittent usage each week of the same or switch to intermittent use of a mild ointment such as 1% -2.5% hydrocortisone in petrolatum or a 1% hydrocortisone / 1% pramoxine cream mix.

**Effects of corticosteroids:**

- Vasoconstriction – decrease erythema and swelling
- Decreasing fibroblastic proliferation thins out thickened dermal lesions
- Decreasing rapidly turning over keratinocytes thins out thickened epidermal lesions

Corticosteroid responsive vulvar dermatoses include:
Thick and scaly (lichen sclerosus, lichen simplex chronicus, psoriasis, contact dermatitis)
Blistering erosive disease
Bullous diseases

Corticosteroid potency depends on:
- Cortisone molecule
- Concentration of steroid in vehicle
- Partition co-efficient of steroid vehicle system
- Application frequency and length of time used

Caution: steroids can be associated with irregular menses, increased BP, worsening of diabetes control, infection and glaucoma.
<table>
<thead>
<tr>
<th>Class</th>
<th>U.S. Brand Name</th>
<th>Generic name</th>
</tr>
</thead>
<tbody>
<tr>
<td>Super-high Potency</td>
<td>Temovate® Cream, 0.05%</td>
<td>clobetasol propionate</td>
</tr>
<tr>
<td></td>
<td>Temovate® Ointment, 0.05%</td>
<td>clobetasol propionate</td>
</tr>
<tr>
<td></td>
<td>Temovate® E, 0.05%</td>
<td>clobetasol propionate</td>
</tr>
<tr>
<td></td>
<td>Diprolene® Cream, 0.05%</td>
<td>betamethasone dipropionate</td>
</tr>
<tr>
<td></td>
<td>Diprolene® Ointment, 0.05%</td>
<td>betamethasone dipropionate</td>
</tr>
<tr>
<td></td>
<td>Diprolene® AF Cream, 0.05%</td>
<td>betamethasone dipropionate</td>
</tr>
<tr>
<td></td>
<td>Psorcon® Ointment, 0.05%</td>
<td>diflorasone diacetate</td>
</tr>
<tr>
<td></td>
<td>Ultravate® Cream, 0.05%</td>
<td>halobetasol propionate</td>
</tr>
<tr>
<td></td>
<td>Ultravate® Ointment, 0.05%</td>
<td>halobetasol propionate</td>
</tr>
<tr>
<td>II</td>
<td>Cyclocort® Cream, 0.1%</td>
<td>Amcinonide</td>
</tr>
<tr>
<td></td>
<td>Cyclocort® Ointment, 0.1%</td>
<td>amcinonide</td>
</tr>
<tr>
<td></td>
<td>Diprosone® Ointment, 0.05%</td>
<td>betamethasone dipropionate</td>
</tr>
<tr>
<td></td>
<td>Florone® Ointment, 0.05%</td>
<td>diflorasone diacetate</td>
</tr>
<tr>
<td></td>
<td>Lidex® Cream, 0.05%</td>
<td>flucinonide</td>
</tr>
<tr>
<td></td>
<td>Lidex® Ointment, 0.05%</td>
<td>flucinonide</td>
</tr>
<tr>
<td></td>
<td>Lidex-E® Cream, 0.05%</td>
<td>flucinonide</td>
</tr>
<tr>
<td></td>
<td>Maxiflor® Ointment, 0.05%</td>
<td>diflorasone diacetate</td>
</tr>
<tr>
<td></td>
<td>Maxivate®, Ointment, 0.05%</td>
<td>betamethasone dipropionate</td>
</tr>
<tr>
<td></td>
<td>Topicort® Cream, 0.25%</td>
<td>betamethasone valerate</td>
</tr>
<tr>
<td></td>
<td>Topicort® Ointment, 0.25%</td>
<td>betamethasone valerate</td>
</tr>
<tr>
<td>III</td>
<td>Aristocort A® Cream 0.5%</td>
<td>triamcinolone acetonide</td>
</tr>
<tr>
<td></td>
<td>Cutivate® Ointment, 0.05%</td>
<td>fluticasone propionate</td>
</tr>
<tr>
<td></td>
<td>Diprosone® Cream, 0.05%</td>
<td>betamethasone dipropionate</td>
</tr>
<tr>
<td></td>
<td>Elocon® Ointment, 0.1%</td>
<td>mometasone furoate</td>
</tr>
<tr>
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<td>Florone® Cream, 0.05%</td>
<td>diflorasone diacetate</td>
</tr>
<tr>
<td></td>
<td>Maxiflor® Cream, 0.05%</td>
<td>diflorasone diacetate</td>
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<td>Maxivate® Cream, 0.05%</td>
<td>betamethasone dipropionate</td>
</tr>
<tr>
<td></td>
<td>Valisone® Ointment, 0.1%</td>
<td>betamethasone valerate</td>
</tr>
<tr>
<td>IV</td>
<td>Aristocort® Ointment, 0.1%</td>
<td>triamcinolone acetonide</td>
</tr>
<tr>
<td></td>
<td>Cordran® Ointment, 0.05%</td>
<td>flurandrenolide</td>
</tr>
<tr>
<td></td>
<td>Elocon® Cream, 0.05%</td>
<td>mometasone furoate</td>
</tr>
<tr>
<td></td>
<td>Kenalog® Ointment, 0.1%</td>
<td>triamcinolone acetonide</td>
</tr>
<tr>
<td></td>
<td>Synalar® Ointment, 0.025%</td>
<td>fluocinolone acetonide</td>
</tr>
<tr>
<td></td>
<td>Topiort LP® Cream, 0.05%</td>
<td>desoximetasone</td>
</tr>
<tr>
<td>V</td>
<td>Aristocort® Cream, 0.1%</td>
<td>triamcinolone acetonide</td>
</tr>
<tr>
<td></td>
<td>Cordran® Cream, 0.05%</td>
<td>flurandrenolide</td>
</tr>
<tr>
<td></td>
<td>Cutivate® Cream, 0.05%</td>
<td>mometasone furoate</td>
</tr>
<tr>
<td></td>
<td>Dermatop® Emollient cream, 0.05%</td>
<td>triamcinolone acetonide</td>
</tr>
<tr>
<td></td>
<td>Kenalog® Cream, 0.1%</td>
<td>triamcinolone acetonide</td>
</tr>
<tr>
<td></td>
<td>Kenalog ointment, 0.025%</td>
<td>hydrocortisone butyrate</td>
</tr>
<tr>
<td></td>
<td>Locoid® Cream, 0.1%</td>
<td>fluocinolone acetonide</td>
</tr>
<tr>
<td></td>
<td>Synalar® Cream, 0.025%</td>
<td>betamethasone valerate</td>
</tr>
<tr>
<td></td>
<td>Valisone® Cream, 0.1%</td>
<td>hydrocortisone valerate</td>
</tr>
<tr>
<td></td>
<td>Utitcort® Cream 0.025%</td>
<td>hydrocortisone valerate</td>
</tr>
<tr>
<td></td>
<td>Westcort® Cream, 0.2%</td>
<td>hydrocortisone valerate</td>
</tr>
<tr>
<td>VI</td>
<td>Aclovate® Cream, 0.05%</td>
<td>alclometasone dipropionate</td>
</tr>
<tr>
<td></td>
<td>Aclovate® Ointment, 0.05%</td>
<td>alclometasone dipropionate</td>
</tr>
<tr>
<td></td>
<td>Tridesilon® Cream, 0.05%</td>
<td>desonide</td>
</tr>
<tr>
<td>VII</td>
<td>Numerous preparations exist</td>
<td>Dexamethasone, flumethalone,</td>
</tr>
<tr>
<td>Low Potency</td>
<td></td>
<td>hydrocortisone, Methylprednisolone, prednisolone</td>
</tr>
</tbody>
</table>
ALTERNATIVES TO CORTICOSTEROIDS

Alternative topicals to corticosteroids are the Calcineurin inhibitors. Calcineurin inhibitors:
- Pimecrolimus 1% cream (Elidel)
- Tacrolimus 0.03 and 0.1% ointment (Protopic) or compounded 0.1% vaginal cream or a 2g suppository.

These are non-steroidal
Does not cause atrophy
May sting or burn initially when used topically
Equivalent to mild to moderate topical steroids — Pimecrolimus to a mild topical steroid and tacrolimus equivalent to a moderate to strong topical steroid.

These are topical immunosuppressants usually for maintenance of steroid responsive dermatoses

Note: there is a black box warning on these medications. This is because of reports of skin cancers and lymphoma with systemic Calcineurin inhibitors used in organ transplant patients. This warning was also imposed because of one manufacturer’s failure to conduct safety studies.

Note: Skin application results in minimal systemic exposure.
Vaginal use can result in systemic absorption.

Side effects of Calcineurin inhibitors:
- Burn, sting
- Infection — worsening of HSV, HPV, tinea, molluscum contagiosum

Safety with regard to lichen sclerosus and squamous cell carcinoma? There are a number of studies showing good results with this medication in lichen sclerosus in adults and children. There are three reports of genital squamous cell carcinoma with patients who have used tacrolimus and one with squamous cell carcinoma on pimecrolimus.

Treatment of choice for lichen sclerosus is still superpotent topical steroids

For lichen planus that is difficult to treat with only partial control of topical steroids consider using tacrolimus and pimecrolimus. The response reported is between 55 and 94%.

Summary of Calcineurin inhibitors:
- For lichen planus start with topical steroids and consider alternating with Calcineurin inhibitors.
- For lichen sclerosus with atrophy or reaction to topical steroids, consider usage, discuss the risks and follow carefully. No refills without follow-up vulvar exams.
- Consider for use in the following: vulvar dermatoses, psoriasis, Crohn’s, pemphigoid, etc.

Systemic corticosteroids can be useful at times. A full discussion is beyond this lecture. IM triamcinolone acetonide (Kenalog 40) 1 mg per kg for an acute dermatosis (e.g. contact dermatitis or severe lichen simplex chronicus). This can be repeated in 3-4 weeks once or twice to get a severe condition under control. See appropriate monograph for all side effects of all corticosteroids and calcineurin inhibitors.

Caution in patients with diabetes- high dose steroids can interfere with their glucose control.
TO DO FOR ALL VULVAR RASHES
   Educate
   Support
   Stop: irritation, contact dermatitis, scratching
   Treat: infection – Candida, bacteria, atrophy, and inflammation
   Poor response: biopsy

CAUSES OF TREATMENT FAILURE
   Non-compliance
      Poor education
      Fear of topical steroids
      Limited mobility

INCORRECT DIAGNOSIS
   Associated problems
      LS plus SCC or contact dermatitis
      Scarring

MOST COMMON ASYMPTOMATIC VULVAR DISEASES
   Lichen sclerosus, Lichen planus, Malignancy – compounded by
      Ignorance
      Denial

CAUSES OF POOR COMPLIANCE
   Fear of steroids
   Vulvar ignorance
   Miscommunication
   Physical impairment
   Secondary gain – no sex
   Phobic about touching “down there”
List of Lubricants

This does not attempt to be a complete list, but rather describes commonly used lubricants. We do not officially recommend use of any one of these products, nor do we recommend any one product over any other products.

Slippery Stuff a silken gel that does not leave a sticky residue. It is hygienic, water-based and water-soluble, odorless, long lasting and latex compatible.

Astroglide: A long lasting, light lubrication that is odorless and flavorless. It is water soluble. Many like it because it is a long lasting lubricant that does not become "stringy"

Femigel Natural product from tea trees. For vaginal dryness.

Jo- water based, silicone based or a combination of both

K-Y Jelly: Generally considered an all-purpose lubricant that many people have found helpful with a "medium" degree of thickness. Some report it comes out too fast and gets "gummy."

Lubrin: A suppository. Many post-menopausal women find this a helpful lubricant because, since it is inserted into the vagina, it lasts longer. They indicate that it needs some time to melt inside the vagina because it is a suppository. For some women, they indicate that it is almost "too much" lubrication.

Moist Again Natural

Replens: A lubricant that is inserted by applicator into the vagina. It comes in a package of 12 single-use applications. This vaginal gel is considered to have medium thickness and properties similar to Ortho Personal Lubricant. Women note that, like Lubrin, it does not dissolve too quickly. Must be used several times weekly.


Surgilube: Many consider this to be thicker than K-Y Jelly

Alboline - Most drug stores sell it in the cosmetic section. Is actually intended to remove make up and provide moisture to a the face.

Vitamin E oil: Available in health food stores, preferred by some women for natural, non-irritating qualities.

Vegetable oil (like olive oil) can also be used.

Egg whites have been used for lubrication.
Saliva has been used for lubrication

Pre-Seed is a vaginal lubricant that does not appear to cause significant damage to sperm

**Agents for sexual enhancement**

Viafem – Aminophylline 30mg/mL 15mL Apply ½-1 mL to clitoris before intercourse
Ergoloid Mesylates 0.5 mg/mL
Nitroglycerine 1mg/mL
L-Arginine 60mg/mL
Pentoxyphylline 50mg/mL

Trimix FM – Papaverine 30mg/mL 5mL Apply 0.5mL to clitoris one hour before intercourse
Phentolamine 1mg/mL
PGE\_1 20mcg/mL

Testosterone 0.5mg/mL 30mL Apply 1/2mL to labia and 1/2mL to inner arm or Thigh q AM.
References

General


**Nonneoplastic Epithelial Conditions/Lichen sclerosus**


Fite C, Plantier F, Dupin N, Avril MF, Moyal-Barracco M. Vulvar verruciform xanthoma: Ten cases associated with lichen sclerosus, lichen planus, or other conditions. Archives of Dermatology. 2011;147(9): 1087-92.


van de Nieuwenhof HP, Bulten J, Hollema H, Dommerholt RG, Massuger LF, van der Zee AG, et al. Differentiated vulvar intraepithelial neoplasia is often found in lesions, previously diagnosed as lichen sclerosus, which have progressed to vulvar squamous cell carcinoma. Modern Pathology : An Official Journal of the United States and Canadian Academy of Pathology, Inc 2011;24(2):297-305.


**Lichen Planus**


Fite C, Plantier F, Dupin N, Avril MF, Moyal-Barracco M. Vulvar verruciform xanthoma: Ten cases associated with lichen sclerosus, lichen planus, or other conditions. Archives of Dermatology 2011;147(9):1087-92.


**Crohn’s Disease**


**Hidradenitis suppurativa**


Stewart EG, Marjesson LJ, Danby FW. Hidradenitis suppurativa. Uptodate.com 2008


Preliminary findings suggest hidradenitis suppurativa may be due to defective follicular support. Danby FW, Jemec GB, Marsch WCh, von Laffert M. Br J Dermatol. 2013 May;168(5):1034-9.


Contact Dermatitis

O'Gorman SM, Torgerson RR. Allergic contact dermatitis of the vulva. Dermatitis. 2013 Mar-Apr;24(2):64-72

**Lymphedema**


**Ulcers (including aphthous ulcers)**


**Behçet’s Disease**


**Desquamative Inflammatory Vaginitis**


Infectious Diseases

General


Sheeley A. Sorting out common causes of abnormal vaginal discharge. JAAPA. 2004;17(10):15-6, 18-20, 22.

Bacterial Vaginosis


Trichomonas


Candidiasis


Herpes


Molluscum contagiosum

Pruritus ani

Vulvodynia


**Persistent Genital Arousal Disorder**


**Langerhan’s Cell Histiocytosis**


**Preinvasive and Invasive Diseases of the Vulva**


TM, Barr E, Haupt R. Four year efficacy of prophylactic human papillomavirus quadrivalent vaccine against low grade cervical, vulvar, and vaginal intraepithelial neoplasia and anogenital warts: randomised controlled trial. BMJ. 2010;341:c3493.


www.cdc.gov/cancer/knowledge/ Vaginal and Vulvar cancers Inside Knowledge