

#### **Vulvar Terminology**

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# Vulvar Pain: A New Terminology

Thanks to Colleen Stockdale, MD and Hope Haefner, MD for contributing slides used in this talk.

I have no commercial relationships related to this talk.



# 58 y.o. G0 c/o 20 years of pelvic and introital pain

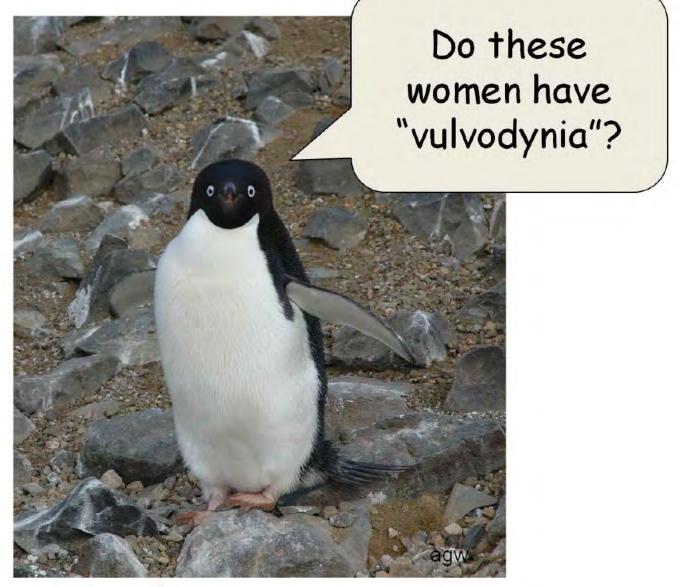
- 20 year history of vulvar burning. Constant but significantly exacerbated with touch. Worse in the past 6 months.
  - Worse when sitting radiates down her left leg
  - Relieved somewhat by elevating her legs
    - When travels by car, sits in the back seat so she can stretch her legs horizontally
    - at work she stands or sits on a rubber donut
  - Unable to have intercourse because of pain
  - Vulvar biopsy c/o lichen planus
- PMH Endometriosis, Irritable Bowel Syndrome
- On exam,
  - Atrophic external genitalia, superficial erosion across fourchette, second superficial erosion in interlabial fold on left
  - Q-tip test positive both in fourchette and in are of interlabial erosion



## A 27 y.o. G0 female complains of introital dyspareunia.

- Intercourse has been impossible for the past 6 months.
  - She finds it very painful even to use a tampon.
- She also has lower abdominal pain and urinary urgency – repeated urine cultures are negative.
- Examination is negative except for slight erythema around the minor vestibular glands.
- Light touch with a cotton applicator elicits severe pain at multiple points around the forchette.
- Search for vulvar and vaginal candida is negative.







#### Educational Objectives

- Introduce the 2015 Consensus Terminology and Classification of Vulvar Pain
- Contrast the 2015 Terminology with the 2003 ISSVD Terminology.
- List potential factors associated with vulvodynia



#### Vulvar pain – first described late 1800s

- "...excessive sensibility of the nerves supplying the mucous membrane of some portion of the vulva..." –T.G.Thomas, 1874
- Disease "characterized by a supersensitiveness of the vulva...When, however the examining finger comes in contact with the hyperaesthetic part, the patient complains of pain, which is sometimes so great as to cause her to cry out." A.J.C. Skene, 1889
- "exquisitely sensitive deep-red spots in the mucosa of the hymeneal ring are a fruitful source of dyspareunia- tender enough at times to make a vaginal exam impossible." Howard Kelly -1928



Haefner. J lower genital tract dis. 2006;11:48-49

#### Vulvodynia, other terms

- "Burning vulva syndrome" ISSVD, 1976
- "Vestibular adenitis" Edward Friedrich, 1983
- "Vulvar vestibulitis syndroms" Friedrich, 1987
- Other terms
  - Vulvar dysesthesia
  - Dysesthetic vulvodynia
  - Essential vulvodynia



Haefner. J lower genital tract dis. 2006;11:48-49

#### **Vulvodynia**

- Vulvar discomfort most often described as burning, stinging, rawness, soreness, aching occurring in the absence of relevant visible findings or a specific, clinically identifiable neurologic disorder
- Generalized
  - Involvement of the whole vulva or migratory
- Localized
  - Involvement of a portion of the vulva
    - e.g. vestibulodynia, clitorodynia



Moyal-Barracco, Lynch; J reproductive med. 2003 49:772-777

"Generalized" / "Localized" each further categorized

- Provoked
  - Triggered by physical contact- sexual, nonsexual or both
    - e.g. intercourse, insertion of tampon, clothing pressure, cotton tip applicator pressure, fingertip pressure, etc.
- Unprovoked
  - Occurs spontaneously without specific physical trigger
- Mixed
  - Provoked and unprovoked



Moyal-Barracco, Lynch; J reproductive med. 2003 49:772-777

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- Considerations
  - Periductal erythema usually a normal finding and not relevant
    - Bilatral, symmetrical around orifices of Bartholin's ducts and minor vestibular ducts
  - The term "vestibulitis" is discouraged
    - "-itis" implies inflammation, not a part of vulvodynia
    - Replaced with "Provoked vestibulodynia"
  - Other vulvar diseases present but not a cause of vulvar pain are not relevant
    - e.g. genital warts, nevi, cysts



#### A. Vulvar pain related to a specific disorder

- Infectious
  - Candidiasis, Herpes, etc.
- Inflammatory
  - Lichen Planus, Immunobullous disorders, etc.
- Neoplastic
  - Paget disease, squamous cell carcinoma, etc.
- Neurologic
  - Herpes neuralgia, spinal compression, etc.



Moyal-Barracco, Lynch; J reproductive med. 2003 49:772-777

#### B. Vulvodynia

- "Vulvar discomfort most often described as burning pain occurring in the absence of relevant visible findings or a specific, clinically identifiable neurologic disorder"
- i.e. The implication is that vulvodynia is idiopathic.



Moyal-Barracco, Lynch; J reproductive med. 2003 49:772-777





#### Why change the terminology?

- Since 2003, research as expanded the category of "identifiable causes" as well as identifying and elaborating on factors potentially associated with vulvodynia.
  - Vulvodynia is in all likelihood not one disease, but a constellation of symptoms of sometimes overlapping disease processes.



Bornstein et al 2016

- Collaborative effort of several organizations
- Consensus Conference April 8-9, 2015
  - ISSVD
    - International Society for the Study of Vulvovaginal Disease
  - ISSWSH
    - International Society for the Study of Women's Sexual Health
  - IPPS
    - International Pelvic Pain Society
  - ASCCP
  - ACOG



#### Pain caused by a specific disorder

- Infectious (e.g. recurrent candidiasis, herpes)
- Inflammatory (e.g. lichen sclerosis, lichen planus, immunobullous disorders)
- Neoplastic (e.g. Paget disease, SCC)
- Neurologic (e.g. post-herpetic neuralgia, nerve compression or injury, neuroma)
- \*Trauma (e.g. female genital cutting, obstetrical)
- \*latrogenic (e.g. post-operative, chemotherapy, radiation)
- \*Hormone deficiencies (e.g. Genito-Urinary Syndrome of Menopause [vulvo-vaginal atrophy])
- Women may have both a specific disorder and vulvodynia



#### Vulvodynia:

- Vulvar pain of at least 3 months duration without clear identifiable cause, which may have potential associated factors
  - Three months most common duration specified in literature
  - "Potential associated factors" may be clinically prominent and may help direct further evaluation and/or treatment
    - Factors may co-occur and overlap helping direct treatment targets
    - Level of evidence for potential associated factors is 2 except for "structural defects" which is level 3.



#### **Descriptors of Vulvodynia:**

- Generalized / Localized (e.g. vestibulodynia, clitorodynia) / Mixed
- Provoked (e.g. insertional, contact) / Spontaneous / Mixed
- \*Onset
  - Primary or secondary
- \*Temporal pattern
  - Intermittent/ persistent / constant / immediate / delayed
  - \*new



#### Potential factors associated with vulvodynia

- Co-morbidities and other pain syndromes
  - Painful bladder syndrome (Interstitial cystitis)
  - Fibromyalgia
  - Irritable Bowel Syndrome
  - Orofacial pain (Temporomandibular syndrome)
- Often more than one comorbid condition present.



#### Chronic Comorbid Pain Disorders

Reed BD et al Obstet Gynecol 2012;120:145-51

- N= 1890 women In SE Michigan
  - Used validated questionnaires for chronic pain conditionsbaseline and 6 month follow-up
  - Mean age 50.4
    - 76.8% white; 16.2% black; 2.4% Hispanic; 4.5% other
- 27.1% (512 women) screened positive for at least one of four chronic pain conditions



#### Chronic Comorbid Pain Disorders

Reed BD et al Obstet Gynecol 2012;120:145-51

#### Prevalence of chronic pain

- Interstitial cystitis
- Vulvodynia
- Irritable Bowel syndrome
- Fibromyalgia
- Negative for chronic pain

7.5% (95% CI 6.2-9.0)

8.7% (95% CI 7.3-10.4)

9.4% (95% CI 8.1-11.0)

11.8% (95% CI 10.1-13.7)

**72.9%** (95% CI 70.2-75.4)



#### Chronic Comorbid Pain Disorders

Reed BD et al Obstet Gynecol 2012;120:145-51

#### Odds of Additional Comorbidity in Women with One Pain Condition

Comorbidity	Vulvodynia	Fibromyalgia	IC	IBS
Vulvodynia		3.4 (2.2-5.3)	2.3 (1.3-4.0)	3.0 (1.9-4.7)
Fibromyalgia	3.3 (2.1-5.2)		5.1 (3.2-8.1)	3.0 (2.0-4.5)
IC	2.3 (1.3-4.0)	5.1 (3.3-8.1)		6.2 (4.0-9.5)
IBS	3.0 (1.9-4.8)	2.9 (1.9-4.5)	6.1 (4.0-9.4)	

Odds ratios controlled for age, marital status, ethnicity, education. All sig. P<.001

#### Odds of Having 2 or more Comorbidities in Addition to Index Condition

Vulvodynia	Fibromyalgia	IC	IBS
4.9 (2.6-9.2)	6.7 (4.0-11.4)	11.8 (6.2- 22.3)	8.0 (4.4-14.5)

Referent condition is no additional comorbidities.



#### Potential factors associated with vulvodynia

- Genetics
  - Some women with provoked vestibulodynia have genetic predisposition
    - Genetic polymorphisms increase risk of cancida and other infections
    - Genetic mediated prolonged / exaggerated inflammatory reponses
    - Increase susceptibility to hormonal changes with OCPs
- Hormonal (e.g. pharmacologically induced)
  - OCP associated increased risk of provoked vestibulodynia

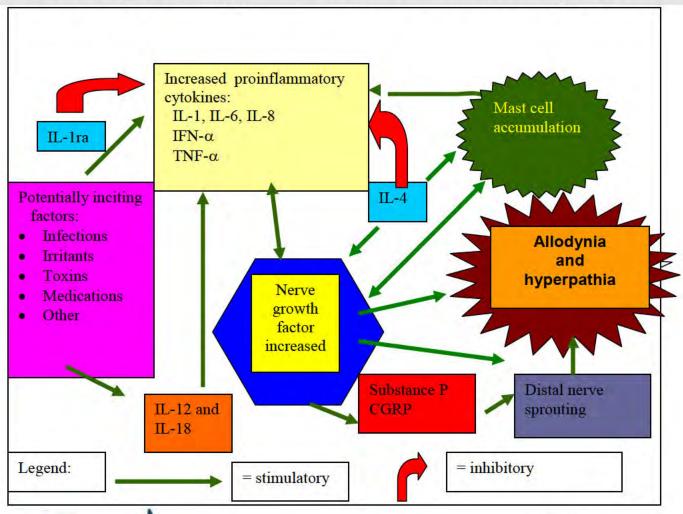


#### Potential factors associated with vulvodynia

- Musculoskeletal
  - Pelvic muscle hypertonicity and other pelvic floor myofascial dysregulation in provoked vestibulodynia
- Inflammation
  - Increased inflammatory cells in painful regions of vestibule
  - Increased mast cells, degranulinated mast cells and subepithelial heparinase activity associated with hyperinnervation in provoked vestibulodynia



### Proposed inflammatory mechanism of the allodynia/hyperpathia of vulvodynia



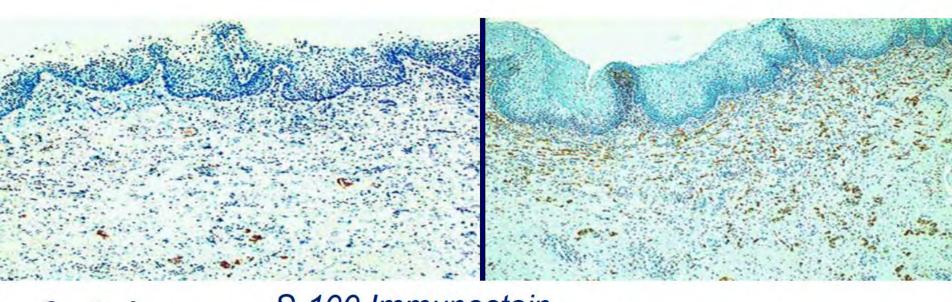
Slide thanks to Hope Haefner, MD



#### Potential factors associated with vulvodynia

- Neurological
  - Central (Spine, brain)
    - Women with provoked vestibulodynia more sensitive to stimulation in non-genital areas than pain free women.
    - Brain imaging studies have shown changes in structure, function and resting state in women with provoked vestibulodynia
  - Peripheral (Neuroproliferation)
    - Increased density of nociceptor nerve endings in vestibule with provoked vestibulodynia

# Biopsy of introitus shows increased cutaneous nerve fibers



Control

S-100 Immunostain

Patient with vestibulodynia

Few nerve fibers

**Nerve fiber proliferation** 

Slide thanks to Hope Haefner, MD

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#### Potential factors associated with vulvodynia

- Psychosocial
  - Increased anxiety, depression, childhood victimization, PTSD in population studies
  - OR =4 for antecedent mood or anxiety disorders
  - Pain catastrophizing, fear of pain, hypervigilance of pain, avoidance, anxiety, depression.
- Structural defects (e.g. perineal descent)
  - 2 small case series showed resolution of vulvodynia and pelvic pain after surgery for POP.



- The changes in the 2015 nomenclature are minor with the exception of the addition of "Potential Associated Factors"
  - Suggests multifactorial nature of vulvodynia
  - Promotes individualization of treatment
  - Factors considered "associated" not necessarily "causal"
    - May change pending more research

