Sept 2024

Missouri Cancer Registry and Research Center
Show-Me-Tips

Coding...Abstracting...Education…

Ed 24:03
Vulva

Vulvar Cancer and HPV

Vulvar Cancer

Vulvar cancer is a rare cancer that starts in the tissues of the vulva. It is a type of cancer that occurs on the outer surface of the female genitalia. The vulva is the area of skin that surrounds the urethra and vagina, including the clitoris and labia. Having vulvar intraepithelial neoplasia (VIN) or HPV (human papillomavirus) infection can increase the risk of vulvar cancer. An area of VIN may look different from normal vulvar skin. It is often thicker and lighter than the normal skin around it. However, an area of VIN can also appear red, pink, or darker than the surrounding skin.

https://www.mayoclinic.org/diseases-conditions/vulvar-cancer/symptoms-causes/syc-20368051

https://www.cancer.org/cancer/types/vulvar-cancer/detection-diagnosis-staging/signs-symptoms.html

Invasive squamous cell cancer of the vulva

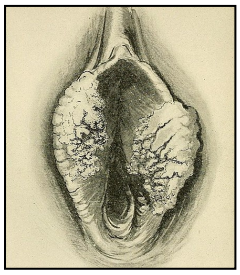
Almost all women with invasive vulvar cancers will have symptoms.

These can include:

- An area on the vulva that looks different from normal – it could be lighter or darker than the normal skin around it, or look red or pink.
- A bump or lump, which can be red, pink, or white and can have a wart-like or raw surface or feel rough or thick
- Thickening of the skin of the vulva
- Itching
- Pain or burning
- Bleeding or discharge not related to the normal menstrual period
- An open sore (especially if it lasts for a month or more)

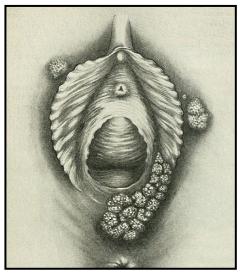
Verrucous carcinoma, a subtype of invasive squamous cell vulvar cancer, looks like cauliflower-like growths similar to genital warts.

Vulvar Cancer



https://commons.wikimedia.org/wiki/File:The_Principles_and_practice_of_gynecology_-_for_students_and_practitioners_(1904)_(14581300578).jpg

Genital Warts



https://commons.wikimedia.org/wiki/File:A_manual_of_gyn%C3%A6colo-gy_and_pelvic_surgery,_for_students_and_practitioners_(1916)_(14581183677).jpg

Vulvar melanoma

Vulvar melanoma can have many of the same symptoms as other vulvar cancers, such as:

- A lump
- Itching
- Pain
- Bleeding or discharge

Most vulvar melanomas are black or dark brown, but they can be white, pink, red, or other colors. They can be found throughout the vulva, but most are in the area around the clitoris or on the labia majora or minora.

Vulvar melanomas can sometimes start in a mole, so a change in a mole that has been present for years can also indicate melanoma. The **ABCDE** rule can be used to help tell a normal mole from one that could be melanoma.

Asymmetry: One-half of the mole does not match the other.
Border irregularity: The edges of the mole are ragged or notched.
Color: The color over the mole is not the same. There may be differing shades of tan, brown, or black and some times patches of red, blue, or white.
Diameter: The mole is wider than 6 mm (about 1/4 inch).
Evolving: The mole is changing in size, shape, or color. The most important sign of melanoma is a change in size, shape, or color of a mole. Still, not all melanomas fit the ABCDE rule.

This project was supported in part by a cooperative agreement between the Centers for Disease Control and Prevention (CDC) and the Missouri Department of Health and Senior Services (DHSS) (NU58DP007130-03) and a Surveillance Contract between DHSS and the University of Missouri.

HPV (human papillomavirus)

HPV is a group of more than 200 related viruses, some of which are spread through vaginal, anal, or oral sex. Sexually transmitted HPV types fall into two groups: low risk and high risk.

- High-risk HPVs can cause several types of cancer. There are 12 high-risk HPV types: HPV 16, 18, 31, 33, 35, 39, 45, 51, 52, 56, 58, and 59. Two of these, HPV 16 and HPV 18, are responsible for most HPV-related cancers.
- Low-risk HPV types rarely cause cancer, although a few low-risk HPV types can cause warts on or around the genitals, anus, mouth, or throat.

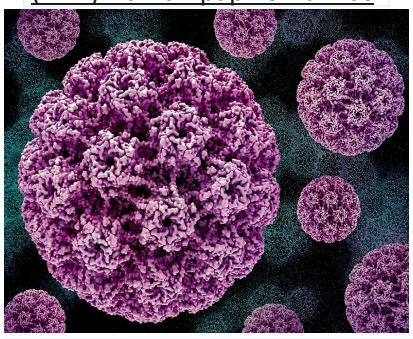
HPV infection is common: Nearly all sexually active people, regardless of their sex, gender identity, or sexual orientation, are infected with HPV within months to a few years of becoming sexually active. Around half of these infections are with a high-risk HPV type.

High-risk HPV infections that persist can cause cancer: Sometimes HPV infections are not successfully controlled by your immune system. When a high-risk HPV infection persists for many years, it can lead to cell changes that, if untreated, may get worse over time and become precancerous and then cancerous.

HPV can cause six types of cancer: These include anal cancer, cervical cancer, oropharyngeal cancer, penile cancer, vaginal cancer, and vulvar cancer.

HPV vaccination can prevent cancer: HPV vaccines can prevent infection with disease-causing HPV types, preventing many HPV-related cancers and cases of genital warts.

(HPV) Human papillomavirus



https://commons.wikimedia.org/wiki/File:Human_Papillomavirus_(HPV).jpg

Long-lasting infections with high-risk HPVs can cause cancer in parts of the body where HPV infects cells. HPV infects the thin, flat cells (squamous cells) that line the inner surfaces of these organs. Most HPV-related cancers are called squamous cell carcinomas. Some cervical cancers come from HPV infection of glandular cells in the cervix and are called adenocarcinomas.

Cervical cancer: Virtually all cervical cancer is caused by HPV
Vaginal cancer: Most vaginal cancer (75%) is caused by HPV
Vulvar cancer: Most vulvar cancer (69%) is caused by HPV
Anal cancer: Over 90% of anal cancer is caused by HPV

Infection with high-risk HPV: Infection with high-risk HPV does not cause symptoms. However, depending on the site of infection, the precancers and cancers caused by a high-risk HPV infection that persists for many years may cause symptoms such as lumps, bleeding, and pain.

Normal cells may become cancer cells. Before cancer cells form in tissues of the body, the cells go through abnormal changes called hyperplasia and dysplasia. Hyperplasia and dysplasia may or may not become cancer.

Once high-risk HPV infects cervical cells, it interferes with the ways in which these cells replicate, divide, and communicate with one another, causing infected cells to multiply in an uncontrolled manner. These infected cells are usually recognized and controlled by the immune system. However, sometimes the infected cells remain and continue to grow, eventually forming an area of precancerous cells that, if not treated, can become cancer. Although research has focused on how high-risk HPV causes cancer in the cervix, HPV-caused cancers at other sites are likely to arise through similar mechanisms.

Research has found that it can take 5 to 10 years for HPV-infected cervical cells to develop into precancers and about 20 years to develop into cancer.

https://www.cancer.gov/about-cancer/causes-prevention/risk/infectious-agents/hpv-and-cancer#:~:text=High%2Drisk%20HPVs%20can%20cause,for%20most%20HPV%2Drelated%20cancers

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ICD-O-3.2 Update Effective January 1, 2022 Updated 9/21/2021

ICD-O Code	Term	Remarks
8085/3	Squamous cell carcinoma, HPV-associated	New term for uterine cervix (2021+) vagina, and vulva beginning 1/1/2022
8086/3	Squamous cell carcinoma, HPV-independent	New term for uterine cervix (2021+) vagina, and vulva beginning 1/1/2022

https://cancerbulletin.facs.org/forums/node/118647#post134971

Version 9 Cervix Cases

The 2021+ cervix histology codes and p16 SSDI was not available to update/complete until the software release in 2022.

8483 and 8484 adenocarcinoma (HPV positive/negative) for any site (including cervix) will trigger an edit for cases diagnosed prior to 2022.

Solid Tumor Rules - Other Sites (Vulva Histologies) (2024 update)

Vulva Coding Note: p16 is a valid test to determine HPV status and can be used to code HPV associated and HPV independent histologies https://seer.cancer.gov/tools/solidtumor/

Specific and NOS Terms and Code	Synonyms	Subtypes/Vaients
Squamous cell carcinoma, NOS **8070**		Squamous cell carcinoma, HPV-associated **8085** Squamous cell carcinoma, HPV-independent **8086**

Site-Specific Data Item (SSDI): Vulva primary added to p16 (V9:2024+)

Description: The p16 biomarker is overexpressed (produced) in response to HPV. It is therefore a surrogate marker for HPV disease.

Rationale: Patients with HPV have a different survival or outcome, so it is important to be able to distinguish this by documenting the p16 results. Testing is performed by immune histochemistry (IHC) which is inexpensive and has near universal availability. It has an easily standardized interpretation. HPV testing is usually performed through DNA testing which is more expensive and less widely available. HPV testing also has technically more variability with the interpretation.

Definition: p16 is a tumor suppressor protein also known as cyclin-dependent kinase inhibitor 2A.

Coding Instructions and Codes:

Note 1: This SSDI is effective for diagnosis years 2024+

- For cases diagnosed 2018-2023, leave this SSDI blank

Note 2: Code 0 for p16 expression of weak intensity or limited distribution.

Note 3: Data item must be based on testing results for p16 overexpression

- Testing for HPV by DNA, mRNA, antibody, or other methods should not be coded in this data item
- Do not confuse p16 with HPV, which is a specific strain of virus. A statement of a patient being HPV positive or negative is not enough to code this data item.

https://apps.naaccr.org/ssdi/list/

Code	Description
0	p16 Negative; Nonreactive
1	p16 Positive; Diffuse, Strong reactivity
8	Not applicable; Information not collected for this case
9	Not tested for p16; Unknown
blank	N/A - Diagnosis year is prior to 2024

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FIGO Stage: Vulva (2018+)

Note 1: There must be a statement about FIGO stage from the managing physician in order to code this data item

- Do **not** code FIGO stage based on the pathology report
- Do **not** code FIGO stage based only on T, N, M
- If "FIGO" is not included with a stated stage, then do **not** assume it is a FIGO stage

Note 2: FIGO stage is not the same thing as FIGO grade. Only code FIGO stage in this field, do not code FIGO grade

- Code FIGO grade in the grade fields

Note 3: If there is more than one FIGO stage provided from the clinical and pathological work up, code the most extensive FIGO stage.

Note 4: The FIGO stage definitions do not include Stage 0 (Tis). Code 97 for any non-invasive neoplasm (behavior /2)

https://apps.naaccr.org/ssdi/list/

HPV Vaccine:

Most of the time, the body can find and clear out HPV. But if the virus stays in the body for a long time, it can cause cancer. Getting vaccinated against HPV helps prevent cancer. The HPV vaccine protects against genital warts and most cases of cervical cancer. It protects against cancer of the vagina, vulva, penis or anus caused by HPV. The HPV vaccine also protects against mouth, throat, head and neck cancers caused by HPV.

The vaccine gives the body a safe way to build immune system awareness of some HPV strains. This means the body has an easier time clearing out those strains of the virus if a person catches them later.

The Gardasil 9 vaccine is approved by the U.S. Food and Drug Administration (FDA). It can be given to people age 9 and older. This vaccine can be given at the same time as other vaccines.

https://www.mayoclinic.org/diseases-conditions/hpv-infection/in-depth/hpv-vaccine/art-20047292

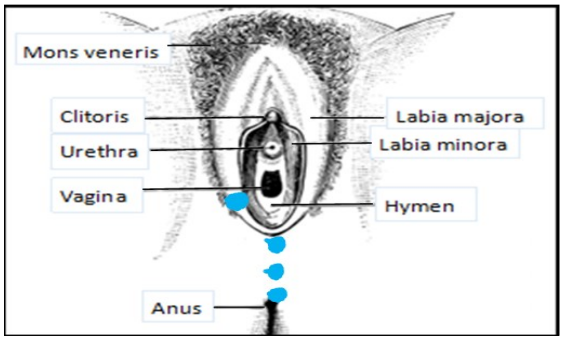
Lymph Node Status: Pelvic - Vulva (2018+)

Question: Should LN Status: Pelvic SSDI be coded as 0 or 9 when the patient has a localized vulvar carcinoma treated with partial vulvectomy with a negative inguinal lymph node dissection, but NO imaging of the pelvis was performed?

Answer: For vulva, pelvic lymph nodes are distant (see Note 2). An inguinal lymph node dissection would not be enough to determine if the pelvic lymph nodes are involved or not.

Code pelvic lymph nodes to 9 for unknown

https://cancerbulletin.facs.org/forums/node/137063#post137113



Vulvar vs Perineal vs Perianal lesions

AJCC TNM Staging Manual 8th edition. Vulva Chapter Fig. 50.1

Vulva and perineum lesions, from top to bottom:
The lesion at the top is vulvar, the middle two lesions are perineal, and the lesion at the bottom is considered perianal.

https://commons.wikimedia.org/wiki/File:Haultain_and_Ferguson_-_external_female_genital_organs.svg

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