Vulvar Cancer Causes, Risk Factors, and Prevention

Risk Factors

A risk factor is anything that affects your chance of getting a disease such as cancer. Learn more about the risk factors for vulvar cancer.

- What Are the Risk Factors for Vulvar Cancer?
- Do We Know What Causes Vulvar Cancer?

Prevention

There is no way to completely prevent cancer. But there are things you can do that might lower your risk. Learn more.

- Can Vulvar Cancer Be Prevented?

What Are the Risk Factors for Vulvar Cancer?

A risk factor is anything that changes a person's chance of getting a disease such as cancer. Different cancers have different risk factors. For example, exposing skin to strong sunlight is a risk factor for skin cancer. Smoking is a risk factor for many cancers.

There are different kinds of risk factors. Some, such as your age or race, can't be changed. Others may be related to personal choices such as smoking, drinking, or diet. Some factors influence risk more than others. But risk factors don't tell us everything. Having a risk factor, or even several, does not mean that a person will get the disease. Also, not having any risk factors doesn't mean that you won't get it, either.
Although several risk factors increase the odds of developing vulvar cancer, most women with these risks do not develop it. And some women who don’t have any apparent risk factors develop vulvar cancer. When a woman develops vulvar cancer, it is usually not possible to say with certainty that a particular risk factor was the cause.

**Age**

The risk of vulvar cancer goes up as women age. Less than 20% of cases are in women younger than age 50, and more than half occur in women over age 70. The average age of women diagnosed with invasive vulvar cancer is 70, whereas women diagnosed with non-invasive vulvar cancer average about 20 years younger.

**Human papilloma virus**

Human papilloma virus (HPV) is a group of more than 150 types of viruses. They are called papilloma viruses because some of them cause a type of growth called a papilloma. Papillomas — more commonly known as warts — are not cancers. Different HPV types can cause different types of warts in different parts of the body. Some types cause common warts on the hands and feet. Other types tend to cause warts on the lips or tongue.

Certain HPV types can infect the outer female and male genital organs and the anal area, causing raised, bumpy warts. These warts may barely be visible or they may be several inches across. The medical term for genital warts is *condyloma acuminatum*. 2 types of HPV, HPV 6 and HPV 11, cause most cases of genital warts. These 2 types are seldom linked to cancer, and so are called *low-risk* types of HPV.

Other HPV types have been linked with cancers of the cervix, vagina, and vulva in women, cancer of the penis in men, and cancers of the anus and throat (in men and women). These are known as *high-risk* types of HPV and include HPV 16, HPV 18, HPV 31, as well as others. Infection with a high-risk HPV may produce no visible signs until pre-cancerous changes or cancer develops.

HPV can be passed from one person to another during skin-to-skin contact. One way HPV is spread is through sex, including vaginal and anal intercourse and even oral sex.

More than half of all vulvar cancers are linked to infection with the high-risk HPV types. HPV linked vulvar cancer is more common in younger women and is seen less often in older women. Some doctors think there are 2 kinds of vulvar cancer. One kind is associated with HPV infection and tends to occur in younger women. The other is not
associated with HPV infection, is more often found in older women, and may develop from a precursor lesion called differentiated vulvar intraepithelial neoplasia (discussed below).

Vaccines have been developed to help prevent infection with some types of HPV. Right now, 2 different HPV vaccines have been approved for use in the United States by the Food and Drug Administration (FDA): Gardasil® and Cervarix®.

For more about HPV, see HPV (Human Papilloma Virus).

**Smoking**

Smoking exposes people to many cancer-causing chemicals that affect more than their lungs. These harmful substances can be absorbed into the lining of the lungs and spread throughout the body. Smoking increases the risk of developing vulvar cancer. Among women who have a history of HPV infection, smoking further increases the risk of developing vulvar cancer. If women are infected with a high-risk HPV, they have a much higher risk of developing vulvar cancer if they smoke.

**HIV infection**

Human immunodeficiency virus (HIV) causes acquired immunodeficiency syndrome (AIDS). Because this virus damages the immune system, it makes women more likely to get and to stay infected with HPV. This could increase the risk of vulvar pre-cancer and cancer. Scientists also believe that the immune system plays a role in destroying cancer cells and slowing their growth and spread.

**Vulvar intraepithelial neoplasia (VIN)**

Squamous cell carcinoma of the vulva usually forms slowly over many years. Precancerous changes often occur first and can last for several years. The medical term most often used for this pre-cancerous condition is vulvar intraepithelial neoplasia (VIN). "Intraepithelial" means that the abnormal cells are only found in the surface layer of the vulvar skin (epithelium).

VIN is typed by how the lesions and cells look: usual-type VIN and differentiated-type VIN. It is sometimes graded VIN2 and VIN3, with the number 3 indicating furthest progression toward a true cancer. However, many doctors use only one grade of VIN.
• Usual-type VIN occurs in younger women and is caused by HPV infection. When usual-type VIN changes into invasive squamous cell cancer, it becomes the basaloid or warty subtypes.
• Differentiated-type VIN tends to occur in older women and is not linked to HPV infection. It can progress to the keratinizing subtype of invasive squamous cell cancer.

In the past, the term dysplasia was used instead of VIN, but this term is used much less often now. When talking about dysplasia, there is also a range of increasing progress toward cancer -- first, mild dysplasia; next, moderate dysplasia; then severe dysplasia; and, finally, carcinoma in situ.

Although women with VIN have an increased risk of developing invasive vulvar cancer, most cases of VIN never progress to cancer. Still, since it is not possible to tell which cases will become cancers, treatment or close medical follow-up is needed.

The risk of progression to cancer seems to be highest with VIN 3 and lower with VIN2. This risk can be altered with treatment. In one study, 88% of untreated VIN3 progressed to cancer, but of the women who were treated, only 4% developed vulvar cancer.

In the past, cases of VIN were included in the broad category of disorders known as vulvar dystrophy. Since this category included a wide variety of other diseases, most of which are not pre-cancerous, most doctors no longer use this term.

**Lichen sclerosus**

This disorder, also called lichen sclerosus et atrophicus (LSA), causes the vulvar skin to become very thin and itchy. The risk of vulvar cancer appears to be slightly increased by LSA, with about 4% of women having LSA later developing vulvar cancer.

**Other genital cancers**

Women with cervical cancer also have a higher risk of vulvar cancer. This is probably because these cancers share certain risk factors. The same HPV types that are linked to cervical cancer are also linked to vulvar cancer. Smoking is also linked to a higher risk of both cervical and vulvar cancers.

**Melanoma or atypical moles**
Women who have had melanoma or dysplastic nevi (atypical moles) in other places have an increased risk of developing a melanoma on the vulva. A family history of melanoma also leads to an increased risk.

- References

See all references for Vulvar Cancer

Do We Know What Causes Vulvar Cancer?

Several risk factors for cancer of the vulva have been identified, and we are beginning to understand how these factors can cause cells in the vulva to become cancerous.

Researchers have made a lot of progress in understanding how certain changes in DNA can cause normal cells to become cancerous. DNA is the chemical that carries the instructions for nearly everything our cells do. We usually look like our parents because they are the source of our DNA. However, DNA affects more than our outward appearance. Some genes (parts of our DNA) contain instructions for controlling when our cells grow and divide.

Certain genes that promote cell division are called oncogenes. Others that slow down cell division or cause cells to die at the right time are called tumor suppressor genes. Cancers can be caused by DNA mutations (defects) that turn on oncogenes or turn off tumor suppressor genes. Usually DNA mutations related to cancers of the vulva occur during life rather than having been inherited before birth. Acquired mutations may result from cancer-causing chemicals in tobacco smoke. Sometimes they occur for no apparent reason. For more on genes and cancer, see Oncogenes and Tumor Suppressor Genes.

Studies suggest that squamous cell cancer of the vulva (the most common type) can develop in at least 2 ways. In up to half of cases, human papilloma virus (HPV) infection appears to have an important role. Vulvar cancers associated with HPV infection (the
basaloid and warty subtypes) seem to have certain distinctive features. They are often found along with several other areas of vulvar intraepithelial neoplasia (VIN). The women who have these cancers tend to be younger and are often smokers.

The second process by which vulvar cancers develop does not involve HPV infection. Vulvar cancers not linked to HPV infection (the keratinizing subtype) are usually diagnosed in older women (over age 55). These women may have lichen sclerosis and may also have the differentiated type of VIN. DNA tests from vulvar cancers in older women rarely show HPV infection, but often show mutations of the \textit{p53} tumor suppressor gene. The \textit{p53} gene is important in preventing cells from becoming cancerous. When this gene has undergone mutation, it is easier for cancer to develop. Younger vulvar cancer patients with HPV infection rarely have \textit{p53} mutations.

These discoveries have not yet affected treatment. But they may help in finding ways to prevent cancer of the vulva and at some point might lead to changes in treatment.

Because vulvar melanomas and adenocarcinomas are so rare, much less is known about how they develop.

- References

  See all references for Vulvar Cancer

Can Vulvar Cancer Be Prevented?

The risk of vulvar cancer can be lowered by avoiding certain risk factors and by having pre-cancerous conditions treated before an invasive cancer develops. Taking these steps cannot guarantee that all vulvar cancers are prevented, but can greatly reduce your chances of developing vulvar cancer.

Avoid HPV infection

Infection with human papilloma virus (HPV) is a risk factor for vulvar cancer. In women,
genital HPV infections occur mainly at younger ages and are less common in women over 30. The reason for this is not clear.

HPV is passed from one person to another during skin-to-skin contact with an infected area of the body. HPV can be spread during sex -- including vaginal intercourse, anal intercourse, and oral sex -- but sex doesn't have to occur for the infection to spread. All that is needed is skin-to-skin contact with an area of the body infected with HPV. The virus can be spread through genital-to-genital contact. It is even possible for a genital infection to spread through hand-to-genital contact.

An HPV infection also seems to be able to be spread from one part of the body to another. This means that an infection may start in the cervix and then spread to the vagina and vulva.

It can be very hard to avoid being exposed to HPV. It might be possible to prevent genital HPV infection by not letting others have contact with your anal or genital area, but even then there could be other ways to become infected that aren't yet clear. For example, a recent study found HPV on the surface of sex toys, so sharing sex toys might spread HPV.

Infection with HPV is common, and in most cases your body is able to clear the infection on its own. But in some cases, the infection does not go away and becomes chronic. Chronic infection, especially with high-risk HPV types, can eventually cause certain cancers, including vulvar cancer.

Certain types of sexual behavior increase a woman's risk of getting a genital HPV infection, such as having sex at an early age and having many sexual partners. Although women who have had many sexual partners are more likely to get infected with HPV, a woman who has had only one sexual partner can still get infected. This is more likely if she has a partner who has had many sex partners or if her partner is an uncircumcised male.

Delaying sex until you are older can help you avoid HPV. It also helps to limit your number of sexual partners and to avoid having sex with someone who has had many other sexual partners.

A person can be infected with HPV for years and not have any symptoms, so the absence of visible warts cannot be used to tell if someone has HPV. Even when someone doesn't have warts (or any other symptom), he (or she) can still be infected with HPV and pass the virus to somebody else.

**HPV in men**
The 2 main factors influencing the risk of genital HPV infection in men are circumcision and the number of sexual partners. Men who are circumcised (have had the foreskin of the penis removed) have a lower chance of becoming and staying infected with HPV. Men who have not been circumcised are more likely to be infected with HPV and pass it on to their partners. The reasons for this are unclear. It may be that after circumcision the skin on the glans (of the penis) goes through changes that make it more resistant to HPV infection. Another theory is that the surface of the foreskin (which is removed by circumcision) is more easily infected by HPV. Still, circumcision does not completely protect against HPV infection -- men who are circumcised can still get HPV and pass it on to their partners.

The risk of a man being infected with HPV is also strongly linked to having many sexual partners (over a man's lifetime).

**Condoms and HPV**

Condoms ("rubbers") provide some protection against HPV, but they do not completely prevent infection. Men who use condoms are less likely to be infected with HPV and pass it on to their female partners. One study found that when condoms are used correctly every time sex occurs, they can lower the HPV infection rate in women by about 70%. Condoms cannot protect completely because they don't cover every possible HPV-infected area of the body, such as the skin on the genital or anal area. Still, condoms do provide some protection against HPV, and they also protect against HIV and some other sexually transmitted diseases. Condoms (when used by the male partner) also seem to help genital HPV infections clear (go away) faster in both women and men.

**Get vaccinated against HPV**

Vaccines are available that protect against certain HPV infections. All of them protect against infection with HPV subtypes 16 and 18. Some can also protect against infections with other HPV subtypes, including some types that cause anal and genital warts.

These vaccines can only be used to prevent HPV infection --they do not help treat an existing infection. To be most effective, the vaccine should be given before a person becomes exposed to HPV (such as through sexual activity).

All of these vaccines can help prevent cervical cancer and pre-cancer. At this time though, only 2 of the HPV vaccines available, Gardasil and Gardasil 9, are approved to
prevent vulvar cancers and pre-cancers. They are also approved to help prevent anal and genital warts, as well as other cancers.

More HPV vaccines are being developed and tested.

For more information, see HPV (Human Papilloma Virus).

Don’t smoke

Not smoking is another way to lower the risk for vulvar cancer. Women who don’t smoke are also less likely to develop a number of other cancers, like those of the lungs, mouth, throat, bladder, kidneys, and several other organs.

Find and treat pre-cancerous conditions

Pre-cancerous vulvar conditions that are not causing any symptoms can be found by regular gynecologic checkups. It is also important to see your health care provider if any problems come up between checkups. Symptoms such as vulvar itching, rashes, moles, or lumps that don't go away could be caused by vulvar pre-cancer and should be checked out. If vulvar intraepithelial neoplasia (VIN) is found, treating it might help prevent invasive squamous cell vulvar cancer. Also, some vulvar melanomas can be prevented by removing atypical moles.

The vulva is examined at the same time a woman has a pelvic examination. Cervical cancer screening with a Pap test (sometimes combined with a HPV test) is often done at the same time. Neither the Pap test nor the HPV test is used to screen for vulvar cancer. The purpose of these tests is to find cervical cancers and pre-cancers early. For more information about these tests and the American Cancer Society guidelines for the early detection of cervical cancer, see Cervical Cancer.

How Pap tests and pelvic examinations are done

First, the skin of the outer lips (labia majora) and inner lips (labia minora) is examined for any visible abnormalities. The health care professional then places a speculum inside the vagina. A speculum is a metal or plastic instrument that keeps the vagina open so that the cervix can be seen clearly. Next, using a small spatula, a sample of cells and mucus is lightly scraped from the exocervix (the surface of the cervix that is closest to the vagina). A small brush or a cotton-tipped swab is then inserted into the cervical opening to take a sample from the endocervix (the inside part of the cervix that
is closest to the body of the uterus). Then, the speculum is removed. The doctor then will check the organs of the pelvis by inserting 1 or 2 gloved fingers of one hand into the vagina while he or she palpates (feels) the lower abdomen, just above the pubic bone, with the other. The doctor may do a rectal exam at this time also. It is very important to know that a Pap test is not always done when a pelvic exam is done, so if you are uncertain you should ask if one was done.

**Self-exam of the vulva**

For most women, the best way to find VIN and vulvar cancer is to report any signs and symptoms to their health care provider and have a yearly well-woman exam. If you have an increased risk of vulvar cancer, you may also want to check your vulva regularly to look for any of the signs of vulvar cancer. This is known as self-examination. Some women choose to examine themselves monthly using a mirror. This can allow you to become aware of any changes in the skin of your vulva. If you do this, look for any areas that are white, darkly pigmented, or red and irritated. You should also note any new growths, nodules, bumps, or ulcers (open sores). Report any of these to a doctor, since they could indicate a vulvar cancer or pre-cancer.

- References
  See all references for Vulvar Cancer

Last Medical Review: July 2, 2014 Last Revised: February 16, 2016

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