



VULVAR CANCER

What is cancer?

Cancer develops when cells in a part of the body begin to grow out of control. Although there are many kinds of cancer, they all start because of out-of-control growth of abnormal cells.

Normal body cells grow, divide, and die in an orderly fashion. During the early years of a person's life, normal cells divide more rapidly until the person becomes an adult. After that, cells in most parts of the body divide only to replace worn-out or dying cells and to repair injuries.

Because cancer cells continue to grow and divide, they are different from normal cells. Instead of dying, they outlive normal cells and continue to form new abnormal cells.

Cancer cells develop because of damage to DNA. This substance is in every cell and directs all its activities. Most of the time when DNA becomes damaged the body is able to repair it. In cancer cells, the damaged DNA is not repaired. People can inherit damaged DNA, which accounts for inherited cancers. Many times though, a person's DNA becomes damaged by exposure to something in the environment, like smoking.

Cancer usually forms as a tumor. Some cancers, like leukemia, do not form tumors. Instead, these cancer cells involve the blood and blood-forming organs and circulate through other tissues where they grow.

Often, cancer cells travel to other parts of the body, where they begin to grow and replace normal tissue. This process is called metastasis. Regardless of where a cancer may spread, however, it is always named for the place it began. For instance, breast cancer that spreads to the liver is still called breast cancer, not liver cancer.

Not all tumors are cancerous. Benign (non-cancerous) tumors do not spread (metastasize) to other parts of the body and, with very rare exceptions, are not life threatening.

Different types of cancer can behave very differently. For example, lung cancer and breast cancer are very different diseases. They grow at different rates and respond to different treatments. That is why people with cancer need treatment that is aimed at their particular kind of cancer.

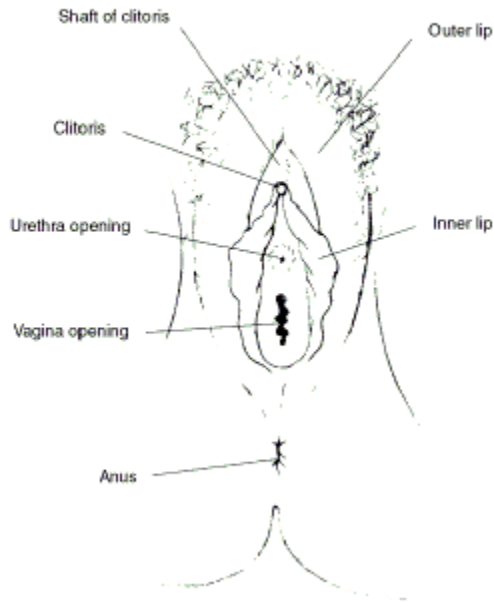
Cancer is the second leading cause of death in the United States. Nearly half of all men and a little over one third of all women in the United States will develop cancer during their lifetimes. Today, millions of people are living with cancer or have had cancer. The risk of developing most types of cancer can be reduced by changes in a person's lifestyle, for example, by quitting smoking and eating a better diet. The sooner a cancer is found and treatment begins, the better are the chances for living for many years.

What is vulvar cancer?

The vulva is the outer part of the female genitals. The vulva includes the opening of the vagina (sometimes called the vestibule), the labia majora (outer lips), the labia minora (inner lips), and the clitoris.

Around the opening of the vagina, there are 2 sets of skin folds. The inner set, called the *labia minora*, are small and hairless. The outer set, the *labia majora*, are larger, with hair on the outer surface. These inner and outer labia (Latin for lips) meet, protecting the vaginal opening and, just above it, the opening of the urethra (the short tube that carries urine from the bladder). The *Bartholin glands* are found just inside the opening of the vagina -- one on each side. These glands produce a mucus-like fluid that acts as a lubricant during sex.

At the front of the vagina, the labia minora meet to form a fold or small hood of skin called the *prepuce*. The *clitoris* is beneath the prepuce. The clitoris is an approximately $\frac{3}{4}$ -inch structure of highly sensitive tissue that becomes swollen with blood during sexual stimulation. The labia minora also meet at a place just beneath the vaginal opening, at the *fourchette*. Beyond the fourchette is the anus, the opening to the rectum. The space between the vagina and the anus is called the *perineum*.



Cancer of the vulva (also known as vulvar cancer) most often affects the inner edges of the labia majora or the labia minora. Less often, cancer occurs on the clitoris or in the Bartholin glands.

Types of vulvar cancer

Squamous cell carcinomas

Most cancers of the vulva are *squamous cell carcinomas*. This type of cancer begins in squamous cells, the main type of skin cells. *Verrucous carcinoma* is a slow-growing subtype of squamous cell carcinoma. This cancer looks like a large wart and a biopsy is needed to determine it is not a benign (non-cancerous) growth. This form of vulvar cancer tends to have a good prognosis (outlook).

Adenocarcinoma

About 8% of vulvar cancers are *adenocarcinomas*, the type of cancer that develops in gland cells. Vulvar adenocarcinomas most often start in cells of the Bartholin glands. These glands are found just inside the opening of the vagina. A Bartholin gland cancer is easily mistaken for a cyst (accumulation of fluid in the gland), so a delay in accurate diagnosis is common.

Most Bartholin gland cancers are adenocarcinomas. Adenocarcinomas can also form in the sweat glands of the vulvar skin.

Paget disease of the vulva is a condition in which adenocarcinoma cells are found in the top layer of the vulvar skin. Up to 25% of patients with vulvar Paget disease also have an invasive vulvar adenocarcinoma (in a Bartholin gland or sweat gland). In the remaining patients, the cancer cells are found only in the skin's top layer and have not grown into the tissues below.

Melanoma

Another type of vulvar cancer is *melanoma*. Melanomas develop from the pigment-producing cells that give skin color. About 5% to 8% of melanomas in women occur on the vulva, usually on the labia minora and clitoris. More information about melanoma can be found in our document *Melanoma Skin Cancer*.

Sarcoma

Less than 2% of vulvar cancers are *sarcomas*, tumors of the connective tissues under the skin that tend to grow rapidly. Unlike other cancers of the vulva, vulvar sarcomas can occur in females at any age, including in childhood.

Basal cell carcinoma

Basal cell carcinoma, the most common type of skin cancer, is more often found on sun-exposed areas of the skin. It occurs very rarely on the vulva. It is discussed further in our document *Basal and Squamous Cell Skin Cancer*.

What are the key statistics about vulvar cancer?

In the United States, vulvar cancer accounts for about 4% of cancers of the female reproductive organs and 0.6% of all cancers in women. In the United States, women have a 1 in 406 chance of developing vulvar cancer at some point during their life. The American Cancer Society estimates that in 2009, about 3,580 cancers of the vulva will be diagnosed in the United States, and about 900 women will die of this cancer.

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. But risk factors don't tell us everything. Cancers often occur in patients without risk factors. Also, having a risk factor, or even several, doesn't mean that you will get the disease.

Although several risk factors increase the odds of developing vulvar cancer, most women with these risks do not develop it. And some women without 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 with 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 100 types of viruses. They are called papilloma viruses because some of them cause a type of growth called a papilloma. Papillomas are not cancers, and are more commonly called warts. HPV can be passed from one person to another during skin-to-skin contact. One way that HPV can be spread is through sex - including vaginal intercourse, anal intercourse, and even during oral sex.

Different HPVs 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. HPV infection is thought to be responsible for up to half of vulvar cancers overall, and most cases that occur in younger women.

Certain HPV types can infect the outer female and male genital organs and the anal area, causing raised, bumpy warts. These warts may be barely 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. However, other HPV types have been linked with genital cancer and so are known as *high-risk* types of HPV. These 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.

In general, vulvar cancer in younger women tends to be associated with infection with the high-risk HPV types. In older women HPV is less likely a risk factor. Some doctors think there are 2 kinds of vulvar cancer. One is associated with HPV infection and tends to occur in

younger women. The other kind is not associated with HPV infection, and more often is found in older women.

Smoking

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

Human immunodeficiency virus

Human immunodeficiency virus (HIV) is the virus that causes the acquired immunodeficiency syndrome (AIDS). Because this virus damages the body's immune system, it makes women more susceptible to persistent HPV infections. This may, in turn, 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

Squamous cell carcinoma of the vulva usually forms slowly over many years. Often, it is preceded by pre-cancerous changes that may 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 often divided into 3 categories -- VIN1, VIN2, and VIN3, with higher numbers indicating furthest progression toward a true cancer. Most cases of VIN are caused by HPV infection.

In the past, the term *dysplasia* had been used instead of VIN. This term is used much less 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.

In the past, cases of VIN were included under 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 likely 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) elsewhere on the body have an increased risk of developing a melanoma on the vulva. A family history of melanoma also leads to an increased risk.

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 great 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.

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 seem to have certain

distinctive features. They are often found along with several other areas of vulvar intraepithelial neoplasia (VIN). The women with 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 usually are diagnosed in older women (over age 55). These women often have lichen sclerosis but rarely have VIN. DNA tests from vulvar cancers in older women rarely show HPV infection, but often show mutations of the p53 tumor suppressor gene. The 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 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 may lead to changes in treatment.

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

Can vulvar cancer be prevented?

The risk of vulvar cancer can be reduced by avoiding certain risk factors and by treating pre-cancerous conditions before an invasive cancer develops. These steps cannot guarantee prevention but can greatly reduce your chances of developing vulvar cancer.

Avoiding risk factors

HPV infection

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

Certain types of sexual behavior increase a woman's risk of getting HPV infection, such as:

- having sex at an early age
- having many sexual partners
- having a partner who has had many sex partners
- having sex with uncircumcised males

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. Men who have not been circumcised are more likely to be infected with HPV and

pass it on to their partners. This may be because the surface of the foreskin (which is removed by circumcision) is more easily infected by HPV. Remember that HPV can be present for years with no symptoms -- it does not always cause warts or any other symptoms. Someone can have the virus and pass it on without knowing it.

Condoms provide some protection against HPV. One study found that when condoms are used correctly they can lower the HPV infection rate by about 70% -- if they are used every time sex occurs. Condoms cannot protect completely because they don't cover every possible HPV-infected area of the body, such as the skin of 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.

Vaccines have been developed to help prevent infection with some types of HPV. Right now, there is an HPV vaccine that has been approved for use in the U.S. by the Food and Drug Administration (FDA). This vaccine is called Gardasil®, and it protects against HPV types 6, 11, 16, and 18. It is currently recommended for use in young females before they become sexually active. This vaccine was designed to lower the risk of cervical cancers and pre-cancers, but it also can prevent vulvar cancers and pre-cancers caused by HPV 16 and 18. More HPV vaccines are being developed and tested.

Smoking

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

Detecting pre-cancerous conditions

Pre-cancerous vulvar conditions that are not causing any symptoms can be found through 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 VIN is found, treating it may help prevent invasive squamous cell vulvar cancer. Also, some vulvar melanomas can be prevented by removing atypical moles.

Examination of the vulva is done at the same time a woman has a Pap test and pelvic examination. The Pap test is not used to screen for vulvar cancer. The purpose of the Pap test is to find cervical cancers and pre-cancers early. The American Cancer Society recommends these guidelines for the early detection of cervical cancer:

- All women should begin cervical cancer screening about 3 years after they start having sex (vaginal intercourse). A woman who waits until she is over 18 to have sex should start screening no later than age 21. Screening should be done every year with the regular Pap test. If the newer liquid-based Pap test is used, testing can be done every 2 years.

- Beginning at age 30, women who have had 3 normal Pap test results in a row may be screened less often - every 2 to 3 years. Testing can be with either the conventional (regular) or liquid-based Pap test. Some women should continue getting tested yearly -- such as those exposed to diethylstilbestrol (DES) before birth, and those with a weakened immune system (from HIV infection, an organ transplant, chemotherapy, or chronic steroid use).
- Another reasonable option for women over 30 is to get screened every 3 years (but not more frequently) with either the conventional or liquid-based Pap test, *plus* the HPV DNA test.
- Women 70 years of age or older who have had 3 or more normal Pap tests in a row and no abnormal Pap test results in the last 10 years may choose to stop having cervical cancer screening. Women with a history of cervical cancer, DES exposure before birth, HIV infection, or a weakened immune system should continue to have screening as long as they are in good health.
- Women who have had a total hysterectomy (removal of the uterus and cervix) may also choose to stop having cervical cancer screening, unless the surgery was done as a treatment for cervical cancer or pre-cancer. Women who have had a hysterectomy without removal of the cervix need to continue cervical cancer screening, and should continue to follow the guidelines above.

See the American Cancer Society documents *Cervical Cancer*, and *Cervical Cancer: Prevention and Early Detection* for more information about finding female reproductive system cancers early.

Self-examination of the vulva is also a way to find vulvar cancer early. You can become aware of any changes in the skin of your vulva by examining yourself monthly using a mirror. 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 vulvar cancers or pre-cancerous conditions.

Can vulvar cancer be found early?

Having pelvic exams and knowing any signs and symptoms of vulvar cancer greatly improve the chances of early detection and successful treatment. If you have any of the problems discussed in the next section, you should see a doctor. If the doctor finds anything abnormal during a pelvic examination, you may need more tests to figure out what is wrong. This may mean referral to a gynecologist (specialist in problems of the female genital system).

There is no standard screening for this disease other than routine physical examinations.

How is vulvar cancer diagnosed?

Signs and symptoms of vulvar cancers

Vulvar intraepithelial neoplasia

Most women with vulvar intraepithelial neoplasia (VIN) have no symptoms at all. When a woman with VIN does have a symptom, it most often is itching that does not go away or get better. An area of VIN may look different than 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.

Because these changes are often caused by other conditions that are not pre-cancerous, some women don't realize that they might have a serious condition. Some try to treat the problem themselves with over-the-counter remedies. Sometimes even doctors may not recognize the condition at first.

Invasive Squamous Cell Cancer of the Vulva

Almost all women with invasive vulvar cancers will have symptoms. The symptoms of early invasive vulvar cancer are similar to those that can be seen with VIN. As the cancer grows, a distinct tumor may be seen. The cancer may appear as a red, pink, or white bump (or bumps) with a wart-like or raw surface. The area could also appear white and feel rough.

About half of the women with vulvar cancer complain of persistent itching and a growth. Some also complain of pain, burning, painful urination, bleeding, and discharge not associated with the normal menstrual period. An open sore (ulcer) that persists for more than a month is another sign.

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

Vulvar melanoma

Melanoma can appear as a darkly pigmented growth. A change in a mole that has been present for years can also indicate melanoma. The *ABCD* rule can 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 sometimes patches of red, blue, or white.

Diameter: The mole is wider than 6 mm (about 1/4 inch).

The most important sign of melanoma is a change in size, shape, or color of a mole. Still, not all melanomas fit the ABCD rule.

Bartholin gland cancer

A distinct mass (lump) on either side of the opening to the vagina can be the sign of a Bartholin gland carcinoma. More often, however, a lump in this area is from a Bartholin gland cyst, which is much more common.

Paget disease

Soreness and a red, scaly area are symptoms of Paget disease of the vulva.

Knowing what to look for can sometimes help with early detection, but it is even better not to wait until you notice symptoms. Have a regular Pap test and pelvic examination.

Medical history and physical exam

The first step is for the doctor to take a complete medical history to check for risk factors and symptoms. Then your doctor will give you a complete physical exam, including a pelvic exam. He or she will feel your uterus, ovaries, cervix, and vagina for anything irregular. Your doctor will also look at your vagina and cervix using a speculum and will take a Pap smear.

Biopsy

Although certain signs and symptoms may strongly suggest vulvar cancer, many of them can be caused by conditions that aren't cancer. The only way to be certain that a vulvar cancer is present is to do a biopsy. In this procedure, a small piece of tissue from the suspicious area is removed and examined under the microscope. A pathologist (a doctor specializing in diagnosing diseases by laboratory tests) will look at the tissue sample under a microscope to see if cancer or a pre-cancerous condition is present and, if so, what type it is.

Rarely, to find all areas of abnormal vulvar skin and to select the best areas to take a biopsy sample from, the doctor may paint the vulva with toluidine blue dye. This dye causes skin with certain diseases to turn blue, including vulvar intraepithelial neoplasia (VIN) and vulvar cancer.

The doctor may use a colposcope, an instrument with binocular magnifying lenses, or a hand held magnifying lens, to select areas to biopsy. The skin is treated with a dilute solution of

acetic acid (which is also the main ingredient in vinegar) that causes areas of VIN and cancer to turn white, making them easier to see through the colposcope.

Once the abnormal areas are found, local anesthetic is injected into the skin to make it numb. If the abnormal area is small, it may be completely removed by an excisional biopsy. For this procedure, the doctor uses a scalpel to remove a small ellipse of skin and sews the skin edges together with surgical thread.

If the abnormal area is larger, a punch biopsy is used to take a small sample. The instrument used looks like a tiny apple corer and removes a small, cylindrical piece of skin 4 mm (about 1/6 inch) across. No stitches are needed after the punch biopsy. Depending on the results of the punch biopsy, additional surgery may be necessary.

Further testing

If you have cancer, tests will be done to see how far it has spread. The results of your physical examination and certain diagnostic tests will be used to determine the size of the tumor, how deeply it has invaded tissues at the site of origin, the extent of any invasion into surrounding organs, and the extent of metastasis (spread to lymph nodes or distant organs). This is called staging (see below). The stage of your cancer is the most important factor in selecting the right treatment plan.

If your biopsy shows that you have vulvar cancer, your health care professional will refer you to a *gynecologic oncologist*, a specialist in female reproductive system cancers. The specialist will also look at your complete personal and family medical history to learn about related risk factors and symptoms of vulvar cancer.

The doctor will perform a complete physical examination to evaluate your general state of health. In addition, he or she will pay special attention to the lymph nodes, particularly those in the groin region, to check for signs of cancer spread. Depending on the biopsy results, several more tests may be done to determine if the vulvar cancer has spread to other areas.

Cystoscopy

This is an examination using a lighted tube to check the inside surface of the bladder. Some advanced cases of vulvar cancer can spread to the bladder, so any suspicious areas noted by this exam are removed for biopsy. This procedure can be done using a local anesthetic, but some patients may require general anesthesia. Your doctor will let you know what to expect before and after the procedure. This procedure was used more often in the past, but is no longer a standard part of the work-up of a woman with vulvar cancer.

Proctoscopy

This is a visual inspection of the rectum using a lighted tube. Some advanced cases of vulvar cancer can spread to the rectum. Any suspicious areas are biopsied. This test was used more

often in the past, but is no longer a standard part of the work-up of a woman with vulvar cancer.

Examination of the pelvis under anesthesia

This permits a more thorough examination that can better evaluate how much the cancer has spread to internal organs of the pelvis.

Imaging tests

Chest x-ray

A plain x-ray of your chest may be done to check for other health problems that might make certain treatments difficult to tolerate. This x-ray can be done in any outpatient setting.

Computed tomography (CT)

The CT scan is an x-ray procedure that produces detailed cross-sectional images of your body. Instead of taking one picture, as does a conventional x-ray, a CT scanner takes many pictures as it rotates around you. A computer then combines these pictures into an image of a slice of your body (think of a loaf of sliced bread). The machine will take pictures of multiple slices of the part of your body that is being studied.

Before any pictures are taken, you may be asked to drink 1 to 2 pints of a liquid called *oral contrast*. This helps outline your intestine so that certain areas are not mistaken for tumors. You may also receive an IV (intravenous) line through which a different kind of contrast dye (IV contrast) is injected. This helps better outline structures in your body.

The injection can cause some flushing (redness and warm feeling that may last hours to days). A few people are allergic to the dye and get hives. Rarely, more serious reactions like trouble breathing and low blood pressure can occur. Medicine can be given to prevent and treat allergic reactions. Be sure to tell the doctor if you have ever had a reaction to any contrast material used for x-rays.

CT scans take longer than regular x-rays and you will need to lie still on a table while they are being done. Also, you might feel a bit confined by the ring-like equipment you're in when the pictures are being taken.

A CT scan can provide information about the size, shape, and position of a tumor, and can be helpful to see if the cancer has spread to other organs. It can also help find enlarged lymph nodes that might contain cancer.

Magnetic resonance imaging (MRI)

MRI scans use radio waves and strong magnets instead of x-rays. The energy from the radio waves is absorbed and then released in a pattern formed by the type of tissue and by certain diseases. A computer translates the pattern of radio waves given off by the tissues into a very detailed image of parts of the body. Not only does this produce cross sectional slices of the body like a CT scanner, it can also produce slices that are parallel with the length of your body.

Sometimes a contrast material is injected into a vein -- just as with CT scans. The contrast used for MRI is different from the one used for CT, so being allergic to CT dye doesn't mean that you are allergic to MRI contrast. MRI scans are a little more uncomfortable than CT scans. First, they take longer -- often up to an hour. Also, you have to be placed inside a tube-like piece of equipment, which is confining and can upset people with claustrophobia (a fear of close spaces). Newer, "open MRI" machines can help people with this fear. The machine also makes a thumping noise that many people find annoying. Some places will provide headphones with music to block this noise out.

MRI images are particularly useful in examining pelvic tumors. They may often detect enlarged lymph nodes in the groin. They are also helpful in detecting cancer that has spread to the brain or spinal cord.

Positron emission tomography (PET)

Positron emission tomography uses glucose (a form of sugar) that contains a radioactive atom. Because cancers use glucose (sugar) at a higher rate than normal tissues, the radioactivity tends to concentrate in the cancer. A special camera is used to detect the radioactivity. This test can be helpful for spotting small collections of cancer cells, and can be useful in seeing if the cancer has spread to lymph nodes. PET scans are also useful when your doctor thinks the cancer has spread, but doesn't know where. PET scans can be used instead of several different x-rays because they scan your whole body. Newer devices combine a CT scan and a PET scan to even better pinpoint the tumor.

How is vulvar cancer staged?

The FIGO/AJCC system for staging vulvar cancer

The stage of most types of vulvar cancer is most often described using the *FIGO* (International Federation of Gynecology and Obstetrics) System of Staging combined with the American Joint Committee on Cancer TNM system. This system classifies the diseases in Stages 0 through IV depending on the extent of the tumor (T), whether the cancer has spread to lymph nodes (N) and whether it has spread to distant sites. This system is not used to stage vulvar melanoma. It is staged like melanoma of the skin. Information about melanoma staging can be found in our document, *Melanoma Skin Cancer*.

Tumor extent (T)

Tis: The cancer is not growing into the underlying tissues.

T1: The cancer is growing only in the vulva or perineum and is smaller than 2 cm. (about 0.8 inches).

T1a: The cancer has grown no more than 1 mm into underlying tissue.

T1b: The cancer has grown more than 1 mm into underlying tissue.

T2: The cancer is growing only in the vulva or perineum and is larger than 2 cm. (about 0.8 inches).

T3: The cancer is growing into the anus, vagina, or lower urethra (the tube that drains urine from the bladder).

T4: The cancer is growing into the upper urethra, bladder or rectum or into the pubic bone.

Lymph node spread of cancer (N)

N0: No lymph node spread

N1: The cancer has spread to lymph nodes on the same side as the tumor

N2: The cancer has spread to lymph nodes in both groin regions

Distant spread of cancer (M)

M0: No distant spread

M1: The cancer has spread to distant sites (includes spread to pelvic lymph nodes)

Stage grouping

The grouping of T, N, and M determines the stage:

Stage 0 (Tis, N0, M0)

This is a very early cancer found in the surface of the skin of the vulva only. It is also known as carcinoma in situ. and as Bowen disease.

Stage I (T1, N0, M0)

The cancer is in the vulva or the perineum (the space between the rectum and the vagina) or both. The tumor is 2 cm or less (about 3/4 inch) in diameter and has not spread to lymph nodes or distant sites.

Stage IA: T1a: These are stage I cancers with invasion no deeper than 1 mm (about 1/25 inch).

Stage IB: T1b: These are stage I cancers that have invaded deeper than 1 mm.

Stage II (T2, N0, M0)

The cancer is in the vulva or perineum or both, and the tumor is larger than 2 cm. It has not spread to lymph nodes or distant sites.

Stage III (T3, N0, N1, M0, or T1 to T2, N1, M0)

The cancer is growing into the anus, vagina, or lower urethra. It may have spread to nearby lymph nodes on one side of the groin. It has not spread to distant sites (T3, N0, N1, M0).

OR

Cancer is found in the vulva or perineum or both and has spread to nearby lymph nodes on 1 side of the groin. It has not spread to distant sites. (T1-T2, N1, M0)

Stage IVA (T1 to T3, N2, M0, or T4, any N, M0)

Cancer has spread to lymph nodes on both sides of the groin (N2) or it has spread beyond nearby tissues to the upper part of the urethra, bladder, rectum, or pelvic bone (T4). It has not spread to distant sites.

Stage IVB (any T, any N, M0)

Cancer has spread to distant organs or lymph nodes. This is the most advanced stage of cancer.

Recurrent

The cancer has come back after treatment.

Survival by stage

The 5-year survival rate refers to the percentage of patients who live *at least 5 years* after their cancer is diagnosed. Five-year survival rates are used to produce a standard way of discussing prognosis. Of course, many people live much longer than 5 years.

Five-year relative survival rates assume that people will die of other causes and compare the observed survival with that expected for people without vulvar cancer. That means that relative survival only talks about deaths from vulvar cancer. Similarly, 10-year survival rates refer to the percentage of patients who live at least 10 years after diagnosis.

Keep in mind that 5-year survival rates are based on patients diagnosed and initially treated more than 5 years ago. Improvements in treatment often result in a more favorable outlook for women more recently diagnosed with vulvar cancer

The numbers below come from the National Cancer Institute. They are based on patients diagnosed from 1998 to 2001.

Survival rates for squamous cell carcinoma of the vulva, by stage

Stage	Relative 5-Year Survival Rate	Relative 10-Year Survival Rate
I	93%	87%
II	79%	69%
III	53%	46%
IV	29%	16%

Survival rates for adenocarcinoma of the vulva, by stage

Stage	Relative 5-Year Survival Rate	Relative 10-Year Survival Rate
I	100%	89%
II	92%	72%
III	74%	71%
IV	Not available	Not available

Survival rates for vulvar melanoma, by stage

Stage	Relative 5-Year Survival Rate	Relative 10-Year Survival Rate
I	83%	71%
II	64%	57%
III	35%	21%
IV	Not available	Not available

How is vulvar cancer treated?

This information represents the views of the doctors and nurses serving on the American Cancer Society's Cancer Information Database Editorial Board. These views are based on their interpretation of studies published in medical journals, as well as their own professional experience.

The treatment information in this document is not official policy of the Society and is not intended as medical advice to replace the expertise and judgment of your cancer care team. It is intended to help you and your family make informed decisions, together with your doctor.

Your doctor may have reasons for suggesting a treatment plan different from these general treatment options. Don't hesitate to ask him or her questions about your treatment options.

After the stage of your vulvar cancer has been established, your cancer care team will recommend a treatment strategy. Think about your options without feeling rushed. If there is anything you do not understand, ask to have it explained again.

The choice of treatment depends largely on the stage of the disease at the time of diagnosis, but other factors can play a part in choosing the best treatment plan, such as your age, your general health, your individual circumstances, and your preferences. Be sure you understand all the risks and side effects of the various therapies before making a decision.

You may want to get a second opinion. This can provide more information and help you feel confident about the treatment plan you choose. Some insurance companies require a second opinion before they will pay for treatments. The 3 main types of treatment used for patients with vulvar cancer are surgery, radiation therapy, and chemotherapy.

Surgery

Choosing the best surgical treatment for each woman means balancing the importance of maintaining sexual functioning with the need to remove all the cancer. In the past, surgeons removing a vulvar cancer also took out a large amount of surrounding normal tissue and possibly local lymph nodes, regardless of the stage of the cancer, because they wanted to be sure that no undetected cancer cells remained. Such extensive surgery resulted in a good chance of cure, but it was deforming and impaired the woman's sexual function if the clitoris were removed. The removal of all the lymph nodes in the groin often led to disabling swelling of the leg on that side.

Today, the importance of sexuality to a woman's quality of life is well recognized. It has also been established that, when cancer is detected early, it is not necessary to remove so much surrounding healthy tissue to achieve a cure. In addition, the sentinel node biopsy procedure avoids removing lymph nodes if the cancer has not spread. However, the use of sentinel lymph node biopsy has not been conclusively shown to be as effective as standard groin dissections. Studies are on-going to evaluate their role (see below). When cancer is more advanced, an extensive procedure may be necessary. Radiation can be combined with chemotherapy and surgery to kill more cancer cells in advanced cases.

The following types of surgery are listed in order of how much tissue is removed (from least to most):

Laser surgery

A focused laser beam vaporizes (burns off) the layer of vulvar skin containing abnormal cells. Laser surgery is used as a treatment for VIN (pre-invasive vulvar cancer). It is not used to treat invasive cancer.

Excision

The cancer and a margin of normal-appearing skin (usually about ½ inch) around it are excised (cut out). This is sometimes called wide local excision. If extensive, it may be called a simple partial vulvectomy.

Vulvectomy

In this type of operation, all or part of the vulva is removed.

- A *skinning vulvectomy* means only the top layer of skin affected by the cancer is removed. Although this is an option for treating extensive VIN3, this operation is rarely done.
- In a *simple vulvectomy*, the entire vulva is removed.
- A *radical vulvectomy* can be complete or partial. When part of the vulva, including the deep tissue, is removed, the operation is called a *partial vulvectomy*. In a *complete radical vulvectomy*, the entire vulva and deep tissues, including the clitoris, are removed.

Sometimes these procedures remove a large area of skin from the vulva, requiring skin grafts from other parts of the body to cover the wound. However, most of the time the surgical wounds resulting from these procedures can be closed without grafts and still provide a very satisfactory appearance. If a graft is required, the gynecologic oncologist may perform the surgery and consult with a plastic/reconstructive surgeon.

Reconstructive surgery is available for women who have had more extensive surgery. A reconstructive surgeon will take a piece of skin and underlying fatty tissue and sew it into the area where the cancer was removed. Several sites in the body can be used, but it is complicated by the fact that the blood supply to the transplanted tissue needs to be kept intact. This is where a skillful surgeon is needed because the tissue must be moved without damaging the blood supply. If you are having this procedure, ask the surgeon to explain how this will be done in your case, because there is no set way of doing it.

Pelvic exenteration

Pelvic exenteration is an extensive operation that includes vulvectomy and removal of the pelvic lymph nodes, as well as removal of one or more of the following structures: the lower colon, rectum, bladder, uterus, cervix, and vagina. How much has to be removed depends on how far the cancer has spread.

If the bladder is removed, a new way to store and eliminate urine is needed. Usually a short segment of intestine is used to function as a new bladder. This may be connected to the abdominal wall so that urine is drained periodically when the woman places a catheter into a small opening (called a *urostomy*). Or urine may drain continuously into a small plastic bag attached to the front of the abdomen over the opening.

If the rectum and part of the colon are removed, a new way to eliminate solid waste will be needed. This is made by attaching the remaining intestine to the abdominal wall so that fecal material can pass through a small opening (called a colostomy) into a small plastic bag worn on the front of the abdomen. Sometimes it's possible to remove a piece of the colon and then reconnect it. In that case, the woman will not need bags or external appliances.

Inguinal lymph node dissection

Because vulvar cancer often spreads to lymph nodes in the groin, these must be removed. This procedure is called an inguinal lymph node dissection. Usually only lymph nodes on the same side as the cancer are removed. If the cancer is in the middle, then both sides may have to be done.

In the past, the incision that was used to remove the cancer was made larger to remove the lymph nodes. Now, doctors prefer to remove the lymph nodes through a separate incision located about 1 cm below and parallel to the groin crease. The incision is made fairly deep, down through membranes that cover the major inguinal vein and artery. This will expose most of the lymph nodes, which are then removed. A major vein, the saphenous, may or may not be closed off by the surgeon. Some surgeons will try and save the saphenous vein in an effort to prevent leg swelling, which is often a problem with this surgery. After the surgery, a suction drain is placed into the incision for a few days, and the wound is closed.

Sentinel lymph node biopsy

This is a newer procedure that can help some women avoid having a full inguinal node dissection. This involves finding and removing the lymph nodes that drain the area where the cancer is. These lymph nodes are known as sentinel lymph nodes because cancer would be expected to spread to them first. The lymph nodes that are removed are then looked at under the microscope to see if they contain cancer cells. If they do, then the remaining lymph nodes in this area need to be removed. If the sentinel nodes do not contain cancer cells, further lymph node surgery is not needed. The sentinel lymph node biopsy procedure is still being studied to see if it finds cancer spread as well as a full inguinal lymph node dissection. This approach is not regarded as standard treatment at this time.

To find the sentinel lymph node(s), a small amount of radioactive material and/or blue dye is injected into the tumor site on the day before surgery. The groin is scanned to identify the side (left or right) that picks up the radioactive material. This is the side that will be operated on. During the surgery to remove the cancer, blue dye will be injected into the tumor site. This allows the surgeon to find the sentinel node by its blue color and then remove it. Sometimes 2 or more lymph nodes turn blue and are removed.

If a lymph node near a vulvar cancer is abnormally large, a sentinel lymph node biopsy is usually not done. Instead, a fine needle aspiration (FNA) biopsy or surgical biopsy of that lymph node is done.

Complications and side effects of vulvar surgery

After vulvar surgery, women often feel discomfort if they wear tight slacks or jeans because the "padding" around the urethral opening and vaginal entrance is gone. The area around the vagina also looks very different.

Removal of wide areas of vulvar skin may result in problems with wound healing, wound infections, or failure of the skin graft to take.. The more tissue removed, the greater the risk of these complications.

Other complications of vulvar and groin node surgery include formation of fluid-filled cysts near the surgical wounds, blood clots that may travel to the lungs, urinary infections, and reduction of sexual desire or pleasure.

Lymphedema: Removal of lymph nodes during a radical vulvectomy with radical lymphadectomy can result in poor fluid drainage from the legs. This causes the fluid to build up leading to leg swelling that is severe and doesn't go down at night. This is called lymphedema. Support stockings or special compression devices may help reduce swelling. Women with lymphedema need to be very careful to avoid infection in the affected leg or legs. They can do this by taking these precautions:

- protect the leg and foot from sharp objects and care for any cuts, scratches, or burns right away
- avoid sunburn of the affected leg(s) and avoid cutting or tearing the cuticles of the toenails
- report any redness, swelling, or other signs of infection to the nurse or doctor without delay

More information about lymphedema can be found in our document, *Understanding Lymphedema (for cancers other than breast cancer)*.

Sexual impact of vulvectomy: Women often fear their partners will feel turned off by the scarring and loss of the outer genitals, especially if they enjoy orally stimulating the woman as part of lovemaking. Some women may be able to have surgery to rebuild the outer and inner lips of the genitals.

It may be difficult for women who have had a vulvectomy to reach orgasm. The outer genitals, especially the clitoris, are important in a woman's sexual pleasure. For many women, the vagina is just not as sensitive. Women may also notice numbness in their genital area after a radical vulvectomy. The feeling may return over the next few months.

When touching the area around the vagina, and especially the urethra, a light caress and the use of a lubricant can help prevent painful irritation. If scar tissue narrows the entrance to the vagina, penetration may be painful. Vaginal dilators can sometimes help stretch the opening. When scarring is severe, the surgeon can use skin grafts to widen the entrance.

Lymphedema resulting from removal of lymph nodes in the groin area can cause pain and fatigue. This also can be a problem during sex. A couple will need to use good communication to cope with such problems.

Radiation therapy

Radiation therapy uses high-energy rays (such as gamma rays or x-rays) and particles (such as electrons, protons, or neutrons) to kill cancer cells. In treating vulvar cancers, radiation is delivered from outside the body in a procedure that is much like having a diagnostic x-ray. This is called external beam radiation therapy. It is sometimes used along with chemotherapy to treat more advanced cancers to shrink them so they can be removed with surgery. Radiation alone may be used to treat lymph nodes in the groin and pelvis.

Common side effects of radiation therapy include tiredness, upset stomach, or loose bowels. Serious fatigue, which may not occur until about 2 weeks after treatment begins, may also occur. When radiation is given to the pelvis, diarrhea is common, but can usually be controlled with over-the-counter medicines. Nausea and vomiting may also occur, but can be treated with medicines. These side effects tend to be worse when chemotherapy is given with radiation. Radiation to the pelvis can also irritate the bladder, and cause problems with urination. Irritation to the bladder is called radiation cystitis, and can result in discomfort and an urge to urinate often. Pelvic radiation can also lead to premature menopause.

Skin changes are also common. As the radiation passes through the skin to the cancer, it may damage the skin cells. This can cause irritation ranging from mild, temporary redness to permanent discoloration. The skin may release fluid, which can lead to infection, so the area exposed to radiation must be carefully cleaned and protected.

Radiation can also lead to low blood counts, causing anemia (low red blood cells) and leukopenia (low white blood cells). The blood counts usually return to normal after radiation is stopped.

Women who receive radiation to the inguinal (groin) area after a lymph node dissection may have problems with the surgical wound site. It may open up or have trouble healing.

If you have side effects from radiation, discuss them with your cancer care team. There are often methods to relieve these symptoms.

Chemotherapy

Chemotherapy (chemo) uses anti-cancer drugs that are usually given intravenously (into a vein), by mouth, or applied to the skin in an ointment. Drugs taken by mouth or injected into a vein, called systemic chemotherapy, enter the bloodstream to reach throughout the body, making this treatment potentially useful for cancer that has spread to distant sites.

Drugs most often used in treating vulvar cancer include cisplatin, mitomycin, and fluorouracil (5-FU). Sometimes a combination of drugs is given as this is often more effective than using just a single drug alone.

The role of chemotherapy in treating vulvar cancer remains to be determined. In more advanced disease, chemotherapy may be given with radiation therapy before surgery. This combined treatment may shrink the tumor, making it easier to remove it with surgery. So far, the results of treating vulvar cancers that have spread to other organs with chemo have been disappointing.

Many of the drugs used in cancer chemotherapy work by attacking cells that are rapidly dividing. This is helpful in killing cancer cells, but these drugs can also affect normal cells, leading to side effects. Side effects of chemotherapy depend on the type of drugs, the amount taken, and the length of time you are treated. Common side effects include:

- nausea and vomiting
- loss or increase of appetite
- temporary loss of hair
- mouth or vaginal sores
- increased chance of infections (due to low white blood cell count)
- increased chance of bleeding and bruising (due to low blood platelet count)
- tiredness (due to anemia, that is, low red blood cell count)
- changes in the menstrual cycle, premature menopause, and infertility (inability to become pregnant). Note: Most women with vulvar cancer are postmenopausal.

Other side effects can occur depending on what drug is used. Most side effects are temporary and stop when the treatment is over, but chemo drugs can have some long-lasting or even permanent effects. For example, cisplatin can cause nerve damage (called neuropathy). This

can lead to numbness, tingling, or even pain in the hands and feet. Ask your cancer care team about the chemo drugs you will receive and what side effects you can expect. Also be sure to talk with them about any side effects you do have so that they can be treated. For example, you can be given medicine to reduce or prevent nausea and vomiting.

Sometimes fluorouracil (5-FU) is applied as a cream directly to the skin. This is called topical chemotherapy. It is rarely used to treat VIN and is not a treatment for invasive cancer of the vulva. Chemotherapy applied directly to the skin as an ointment will cause local irritation and peeling. This is normal and is part of the local destruction of cancer cells. Medicated ointments suggested by the health care team can help relieve the discomfort of this treatment. Topical chemotherapy for VIN is less effective than laser treatment or surgery.

Clinical trials

You may have had to make a lot of decisions since you've been told you have cancer. One of the most important decisions you will make is choosing which treatment is best for you. You may have heard about clinical trials being done for your type of cancer. Or maybe someone on your health care team has mentioned a clinical trial to you.

Clinical trials are carefully controlled research studies that are done with patients who volunteer for them. They are done to get a closer look at promising new treatments or procedures.

If you would like to take part in a clinical trial, you should start by asking your doctor if your clinic or hospital conducts clinical trials. You can also call our clinical trials matching service for a list of clinical trials that meet your medical needs. You can reach this service at 1-800-303-5691 or on our Web site at <http://clinicaltrials.cancer.org>. You can also get a list of current clinical trials by calling the National Cancer Institute's Cancer Information Service toll-free at 1-800-4-CANCER (1-800-422-6237) or by visiting the NCI clinical trials Web site at www.cancer.gov/clinicaltrials.

There are requirements you must meet to take part in any clinical trial. If you do qualify for a clinical trial, it is up to you whether or not to enter (enroll in) it.

Clinical trials are one way to get state-of-the-art cancer treatment. They are the only way for doctors to learn better methods to treat cancer. Still, they are not right for everyone.

You can get a lot more information on clinical trials in our document called *Clinical Trials: What You Need to Know*. You can read it on our Web site or call our toll-free number (1-800-ACS-2345) and have it sent to you.

Complementary and alternative therapies

When you have cancer you are likely to hear about ways to treat your cancer or relieve symptoms that your doctor hasn't mentioned. Everyone from friends and family to Internet groups and Web sites offer ideas for what might help you. These methods can include vitamins, herbs, and special diets, or other methods such as acupuncture or massage, to name a few.

What exactly are complementary and alternative therapies?

Not everyone uses these terms the same way, and they are used to refer to many different methods, so it can be confusing. We use *complementary* to refer to treatments that are used *along with* your regular medical care. *Alternative* treatments are used *instead of* a doctor's medical treatment.

Complementary methods: Most complementary treatment methods are not offered as cures for cancer. Mainly, they are used to help you feel better. Some methods that are used along with regular treatment are meditation to reduce stress, acupuncture to help relieve pain, or peppermint tea to relieve nausea. Some complementary methods are known to help, while others have not been tested. Some have been proven not to be helpful, and a few have even been found harmful.

Alternative treatments: Alternative treatments may be offered as cancer cures. These treatments have not been proven safe and effective in clinical trials. Some of these methods may pose danger, or have life-threatening side effects. But the biggest danger in most cases is that you may lose the chance to be helped by standard medical treatment. Delays or interruptions in your medical treatments may give the cancer more time to grow and make it less likely that treatment will help.

Finding out more

It is easy to see why people with cancer think about alternative methods. You want to do all you can to fight the cancer, and the idea of a treatment with no side effects sounds great. Sometimes medical treatments like chemotherapy can be hard to take, or they may no longer be working. But the truth is that most of these alternative methods have not been tested and proven to work in treating cancer.

As you consider your options, here are 3 important steps you can take:

Look for "red flags" that suggest fraud. Does the method promise to cure all or most cancers? Are you told not to have regular medical treatments? Is the treatment a "secret" that requires you to visit certain providers or travel to another country?

Talk to your doctor or nurse about any method you are thinking about using.

Contact us at 1-800-ACS-2345 to learn more about complementary and alternative methods in general and to find out about the specific methods you are looking at.

The choice is yours

Decisions about how to treat or manage your cancer are always yours to make. If you want to use a non-standard treatment, learn all you can about the method and talk to your doctor about it. With good

information and the support of your health care team, you may be able to safely use the methods that can help you while avoiding those that could be harmful.

Treatment options for squamous cell vulvar cancer by stage

The stage of a vulvar cancer is the most important factor in choosing treatment. However, other factors that affect this decision include the exact location of the cancer on the vulva, the type of cancer, your age, and your overall condition.

Stage 0 (carcinoma in situ)

Treatment options for carcinoma in situ and for less advanced pre-cancerous changes (vulvar intraepithelial neoplasia, or VIN) are the same. Laser surgery, wide local excision, or a skinning vulvectomy may be used, depending on the size and location of the cancer. Fluorouracil (5-FU) ointment may be prescribed. Stage 0 cancers may recur (come back) or new stage 0 cancers may form on other areas of the vulva. The 5-year survival rate is nearly 100%, similar to pre-invasive skin cancers in other body sites.

Stage I

Treatment options depend on the size and depth of the cancer and whether the patient also has VIN. If the depth of invasion is 1 mm or less (stage IA) and there are no other areas of cancer or VIN, the cancer is removed along with a 1-cm margin of the normal tissue around it.

For stage IB cancers, treatment includes a partial radical vulvectomy and inguinal lymph node dissection (removal of nearby groin lymph nodes). Sentinel lymph node biopsy may be done instead of the lymph node dissection (although this is not standard).

Another option that is rarely used for cancers that are larger and quite extensive is a complete radical vulvectomy and removal of the groin lymph nodes. More often, the doctor will perform a radical wide local excision and either a lymph node dissection or a sentinel node biopsy. If the lymph nodes are not removed because the patient is not healthy enough to withstand the surgery, radiation therapy to the groin areas may be used instead of the lymph node dissection.

Stage II

The treatment for most stage II vulvar cancers is partial radical vulvectomy and removal of the lymph nodes in the groin on both sides of the body or sentinel node biopsies. Radiation therapy to the area of surgery will be needed if cancer cells are present at or near the margins (edges of the tissue removed by surgery).

Stage III

Some of these bulky cancers can be cured by radical operations. A radical vulvectomy with removal of the lymph nodes in the groin (superficial and deep groin lymph nodes) may be successful in completely removing the tumor. However, a newer approach is to give chemotherapy along with radiation therapy followed by surgery. Clinical trials are underway to evaluate the use of radiation therapy and/or systemic chemotherapy with cisplatin with or without 5-FU, followed by surgery in cases with a good response to chemotherapy or radiation.

Stage IV

The extent of the surgery beyond a radical vulvectomy depends on what organs contain cancer cells. Pelvic exenteration is an option, although it is used rarely. This operation includes vulvectomy and removal of the pelvic lymph nodes plus removal of some of the following : the lower colon, rectum, bladder, uterus, cervix, and vagina. The standard approach is to combine surgery, radiation, and chemotherapy. Radiation therapy may be done before or after surgery. Chemotherapy may also be given before surgery. Radiation and possibly chemotherapy can also be given to women who cannot have surgery because of prior medical problems. Multimodality treatment (surgery, radiation and chemotherapy) is the standard approach in locally advanced vulvar cancer.

Recurrent vulvar cancer

When a cancer has come back after treatment, it is called recurrent. Treatment options will depend on how soon the cancer comes back and whether it is local (in the vulva), regional (in nearby lymph nodes), or distant (has spread through the bloodstream to organs such as the lungs or bone).

If the recurrence is local, it may still be possible to remove the cancer by surgery or by using combinations of chemotherapy, radiation therapy, and surgery. When the cancer comes back locally more than 2 years after the initial treatment, the prognosis is better than if the cancer had recurred sooner.

When the cancer is unresectable (has grown too large or spread too far to be surgically removed), chemotherapy and/or radiation therapy may be used to help relieve symptoms such as pain caused by the cancer, or to shrink the tumor so that surgery may become an option. If treatment is given only to relieve pain or bleeding, it is called palliative (symptom relief) therapy.

It's very important to understand that palliative treatment is not expected to cure a cancer. Women with stage IV vulvar cancer are encouraged to enter a clinical trial where they may receive new forms of therapy that may be beneficial but are as yet unproven.

Treatment of vulvar adenocarcinoma

If Paget disease is present and there is no associated invasive carcinoma, treatment is wide local excision or simple vulvectomy. If an invasive adenocarcinoma of a Bartholin gland or of vulvar skin sweat glands is present, a partial radical vulvectomy is recommended with removal of inguinal (groin) lymph nodes on one or both sides of the body, depending on the site of the primary tumor.

Treatment of vulvar melanoma

Treatment options depend on how deeply the melanoma has grown into the skin of the vulva. If the depth is less than 0.75 mm, partial vulvectomy with 2 cm (about ¾ inch) margins is the usual treatment. Radical vulvectomy may rarely be used when the lesion extensively involves the vulva. Lymph nodes in the groin are usually sampled or a sentinel node biopsy procedure is done to determine prognosis.

More treatment information

For more details on treatment options -- including some that may not be addressed in this document -- the National Comprehensive Cancer Network (NCCN) and the National Cancer Institute (NCI) are good sources of information.

The NCCN, made up of experts from 19 of the nation's leading cancer centers, develops cancer treatment guidelines for doctors to use when treating patients. Those are available on the NCCN Web site (www.nccn.org).

The NCI provides treatment guidelines via its telephone information center (1-800-4-CANCER) and its Web site (www.cancer.gov). Detailed guidelines intended for use by cancer care professionals are also available on www.cancer.gov.

What should you ask your doctor about vulvar cancer?

It is important for you to have honest, open discussions with your cancer care team. They want to answer all of your questions, no matter how trivial you might think they are. Here are some questions to consider:

- What type of vulvar cancer do I have?
- Has my cancer spread beyond the vulva?
- What is the stage of my cancer and what does that mean in my case?
- What treatments are appropriate for me? What do you recommend? Why?
- What should I do to be ready for treatment?
- What risks or side effects I should expect?
- Will I be able to have children after my treatment?
- Will I be able to enjoy normal sexual relations?
- What are the chances my cancer will recur (come back) with the treatment programs we have discussed?

- Should I follow a special diet?
- What is my expected prognosis, based on my cancer as you view it?
- What do I tell my children, husband, parents, and other family members?

In addition to these sample questions, be sure to write down some questions of your own. For instance, you might want specific information about anticipated recovery times so that you can plan your work schedule. Or you may want to ask about second opinions or about clinical trials for which you may qualify.

What will happen after treatment for vulvar cancer?

Completing treatment can be both stressful and exciting. You will be relieved to finish treatment, yet it is hard not to worry about cancer coming back. (When cancer returns, it is called recurrence.) This is a very common concern among those who have had cancer.

It may take a while before your confidence in your own recovery begins to feel real and your fears are somewhat relieved. Even with no recurrences, people who have had cancer learn to live with uncertainty.

Follow-up care

An important part of your treatment plan is a specific schedule of follow-up visits after surgery, radiation therapy, or chemotherapy to check whether any additional treatment is necessary.

After your treatment is over, it is very important to go to all follow-up appointments. During these visits, your doctors will ask about symptoms, do physical exams, and order blood tests or imaging studies such as CT scans or x-rays. Follow-up is needed to check for cancer recurrence or spread, as well as possible side effects of certain treatments. This is the time for you to ask your health care team any questions you need answered and to discuss any concerns you might have.

Almost any cancer treatment can have side effects. Some may last for a few weeks to several months, but others can be permanent. Don't hesitate to tell your cancer care team about any symptoms or side effects that bother you so they can help you manage them.

It is also important to keep your medical insurance. Even though no one wants to think of their cancer coming back, it is always a possibility. If it happens, the last thing you want is to have to worry about paying for treatment.

Follow-up may involve procedures such as x-rays, CT scans, ultrasound studies, or magnetic resonance imaging (MRI) scans. The doctor may also want you to have blood tests, biopsies (to get tissue samples for microscopic evaluation), or other tests and procedures.

Seeing a new doctor

At some point after your cancer diagnosis and treatment, you may find yourself in the office of a new doctor. Your original doctor may have moved or retired, or you may have moved or changed doctors for some reason. It is important that you be able to give your new doctor the exact details of your diagnosis and treatment. Make sure you have the following information handy:

- a copy of your pathology report from any biopsy or surgery
- if you had surgery, a copy of your operative report
- if you had radiation, a copy of your radiation treatment summary
- if you were hospitalized, a copy of the discharge summary that every doctor must prepare when patients are sent home from the hospital
- finally, since some drugs can have long-term side effects, a list of your drugs, drug doses, and when you took them

Lifestyle changes to consider during and after treatment

Having cancer and dealing with treatment can be time-consuming and emotionally draining, but it can also be a time to look at your life in new ways. Maybe you are thinking about how to improve your health over the long term. Some people even begin this process during cancer treatment.

Make healthier choices

Think about your life before you learned you had cancer. Were there things you did that might have made you less healthy? Maybe you drank too much alcohol, or ate more than you needed, or smoked, or didn't exercise very often. Emotionally, maybe you kept your feelings bottled up, or maybe you let stressful situations go on too long.

Now is not the time to feel guilty or to blame yourself. However, you can start making changes *today* that can have positive effects for the rest of your life. Not only will you feel better but you will also be healthier. What better time than now to take advantage of the motivation you have as a result of going through a life-changing experience like having cancer?

You can start by working on those things that you feel most concerned about. Get help with those that are harder for you. For instance, if you are thinking about quitting smoking and need help, call the American Cancer Society's Quitline® tobacco cessation program at 1-800-ACS-2345.

Diet and nutrition

Eating right can be a challenge for anyone, but it can get even tougher during and after cancer treatment. For instance, treatment often may change your sense of taste. Nausea can be a problem. You may lose your appetite for a while and lose weight when you don't want to. On the other hand, some people gain weight even without eating more. This can be frustrating, too.

If you are losing weight or have taste problems during treatment, do the best you can with eating and remember that these problems usually improve over time. You may want to ask your cancer team for a referral to a dietitian, an expert in nutrition who can give you ideas on how to fight some of the side effects of your treatment. You may also find it helps to eat small portions every 2 to 3 hours until you feel better and can go back to a more normal schedule.

One of the best things you can do after treatment is to put healthy eating habits into place. You will be surprised at the long-term benefits of some simple changes, like increasing the variety of healthy foods you eat. Try to eat 5 or more servings of vegetables and fruits each day. Choose whole grain foods instead of white flour and sugars. Try to limit meats that are high in fat. Cut back on processed meats like hot dogs, bologna, and bacon. Get rid of them altogether if you can. If you drink alcohol, limit yourself to 1 or 2 drinks a day at the most. And don't forget to get some type of regular exercise. The combination of a good diet and regular exercise will help you maintain a healthy weight and keep you feeling more energetic.

Rest, fatigue, work, and exercise

Fatigue is a very common symptom in people being treated for cancer. This is often not an ordinary type of tiredness but a "bone-weary" exhaustion that doesn't get better with rest. For some, this fatigue lasts a long time after treatment, and can discourage them from physical activity.

However, exercise can actually help you reduce fatigue. Studies have shown that patients who follow an exercise program tailored to their personal needs feel physically and emotionally improved and can cope better.

If you are ill and need to be on bed rest during treatment, it is normal to expect your fitness, endurance, and muscle strength to decline some. Physical therapy can help you maintain strength and range of motion in your muscles, which can help fight fatigue and the sense of depression that sometimes comes with feeling so tired.

Any program of physical activity should fit your own situation. An older person who has never exercised will not be able to take on the same amount of exercise as a 20-year-old who plays tennis 3 times a week. If you haven't exercised in a few years but can still get around, you may want to think about taking short walks.

Talk with your health care team before starting, and get their opinion about your exercise plans. Then, try to get an exercise buddy so that you're not doing it alone. Having family or friends involved when starting a new exercise program can give you that extra boost of support to keep you going when the push just isn't there.

If you are very tired, though, you will need to balance activity with rest. It is okay to rest when you need to. It is really hard for some people to allow themselves to do that when they are used to working all day or taking care of a household. (For more information about fatigue, please see the publication, "Cancer Related Fatigue and Anemia Treatment Guidelines for Patients."

Exercise can improve your physical and emotional health.

- It improves your cardiovascular (heart and circulation) fitness.
- It strengthens your muscles.
- It reduces fatigue.
- It lowers anxiety and depression.
- It makes you feel generally happier.
- It helps you feel better about yourself.

And long term, we know that exercise plays a role in preventing some cancers. The American Cancer Society, in its guidelines on physical activity for cancer prevention, recommends that adults take part in at least 1 physical activity for 30 minutes or more on 5 days or more of the week. Children and teens are encouraged to try for at least 60 minutes a day of energetic physical activity on at least 5 days a week.

How about your emotional health?

Once your treatment ends, you may find yourself overwhelmed by emotions. This happens to a lot of people. You may have been going through so much during treatment that you could only focus on getting through your treatment.

Now you may find that you think about the potential of your own death, or the effect of your cancer on your family, friends, and career. You may also begin to re-evaluate your relationship with your spouse or partner. Unexpected issues may also cause concern -- for instance, as you become healthier and have fewer doctor visits, you will see your health care team less often. That can be a source of anxiety for some.

This is an ideal time to seek out emotional and social support. You need people you can turn to for strength and comfort. Support can come in many forms: family, friends, cancer support groups, church or spiritual groups, online support communities, or individual counselors.

Almost everyone who has been through cancer can benefit from getting some type of support. What's best for you depends on your situation and personality. Some people feel safe in peer-support groups or education groups. Others would rather talk in an informal setting, such as church. Others may feel more at ease talking one-on-one with a trusted friend

or counselor. Whatever your source of strength or comfort, make sure you have a place to go with your concerns.

The cancer journey can feel very lonely. It is not necessary or realistic to go it all by yourself. And your friends and family may feel shut out if you decide not to include them. Let them in -- and let in anyone else who you feel may help. If you aren't sure who can help, call your American Cancer Society at 1-800-ACS-2345 and we can put you in touch with an appropriate group or resource.

You can't change the fact that you have had cancer. What you can change is how you live the rest of your life -- making healthy choices and feeling as well as possible, physically and emotionally.

What happens if treatment is no longer working?

If cancer continues to grow after one kind of treatment, or if it returns, it is often possible to try another treatment plan that might still cure the cancer, or at least shrink the tumors enough to help you live longer and feel better. On the other hand, when a person has received several different medical treatments and the cancer has not been cured, over time the cancer tends to become resistant to all treatment. At this time it's important to weigh the possible limited benefit of a new treatment against the possible downsides, including continued doctor visits and treatment side effects.

Everyone has his or her own way of looking at this. Some people may want to focus on remaining comfortable during their limited time left.

This is likely to be the most difficult time in your battle with cancer -- when you have tried everything medically within reason and it's just not working anymore. Although your doctor may offer you new treatment, you need to consider that at some point, continuing treatment is not likely to improve your health or change your prognosis or survival.

If you want to continue treatment to fight your cancer as long as you can, you still need to consider the odds of more treatment having any benefit. In many cases, your doctor can estimate the response rate for the treatment you are considering. Some people are tempted to try more chemotherapy or radiation, for example, even when their doctors say that the odds of benefit are less than 1%. In this situation, you need to think about and understand your reasons for choosing this plan.

No matter what you decide to do, it is important that you be as comfortable as possible. Make sure you are asking for and getting treatment for any symptoms you might have, such as pain. This type of treatment is called *palliative* treatment.

Palliative treatment helps relieve these symptoms, but is not expected to cure the disease; its main purpose is to improve your quality of life. Sometimes, the treatments you get to control your symptoms are similar to the treatments used to treat cancer. For example, radiation

therapy might be given to help relieve bone pain from bone metastasis. Or chemotherapy might be given to help shrink a tumor and keep it from causing a bowel obstruction. But this is not the same as receiving treatment to try to cure the cancer.

At some point, you may benefit from hospice care. Most of the time, this is given at home. Your cancer may be causing symptoms or problems that need attention, and hospice focuses on your comfort. You should know that receiving hospice care doesn't mean you can't have treatment for the problems caused by your cancer or other health conditions. It just means that the focus of your care is on living life as fully as possible and feeling as well as you can at this difficult stage of your cancer.

Remember also that maintaining hope is important. Your hope for a cure may not be as bright, but there is still hope for good times with family and friends -- times that are filled with happiness and meaning. In a way, pausing at this time in your cancer treatment is an opportunity to refocus on the most important things in your life. This is the time to do some things you've always wanted to do and to stop doing the things you no longer want to do.

What's new in vulvar cancer research and treatment?

Research is being done to find new ways to prevent and treat cancer of the vulva. There are some promising new developments.

Oncogenes and tumor suppressor genes

Scientists are learning more about how certain genes called oncogenes and tumor suppressor genes control cell growth and how changes in these genes cause normal vulvar cells to become cancerous. The ultimate goal of this research is gene therapy. Gene therapy involves replacing the damaged genes in cancer cells with normal genes in order to stop the abnormal behavior of these cells.

HPV vaccines

Vaccines for preventing and treating vulvar and cervical cancer are being developed and tested.

Some of these vaccines are meant to prevent infection with certain types of HPV by boosting the body's immunity to them. One HPV vaccine (Gardasil) is now available. Early studies of this vaccine have found that it is able to prevent infection with HPV types 16 and 18 (as well as 6 and 11) and prevent pre-cancerous changes in the cervix. More recent studies have shown that Gardasil also prevents vulvar and vaginal cancers caused by HPV types 16 and 18. Other preventive vaccines are also under study.

Some vaccines being studied are intended for women with established HPV infections, to help their immune systems destroy the virus and cure the infection before a cancer develops.

Still other vaccines are meant to help women who already have a cancer that has recurred or metastasized. These vaccines attempt to produce an immune reaction to the parts of the virus (E6 and E7 proteins) that specifically contribute to the abnormal growth of cancer cells. It is hoped that this immunity will kill the cancer cells or stop them from growing.

Other prevention methods

Topical imiquimod , an anticancer drug , is being studied for treatment of VIN.

Combining surgery, radiation therapy, and chemotherapy

Clinical trials are underway to determine the best way to combine surgery, radiation therapy, and chemotherapy. For example, these trials will provide information about whether certain groups of patients benefit from radiation after surgery and whether patients with cancer that has spread to lymph nodes benefit from chemotherapy or pelvic radiation therapy.

Additional resources

More information from your American Cancer Society

We have selected some related information that may also be helpful to you. These materials may be ordered from our toll-free number, 1-800-ACS-2345.

After Diagnosis: A Guide for Patients and Families (also available in Spanish)

Human Papilloma Virus (HPV), Cancer, and HPV Vaccines -- Frequently Asked Questions

Melanoma Skin Cancer

Sexuality and the Woman with Cancer (also available in Spanish)

Understanding Chemotherapy (also available in Spanish)

Understanding Radiation Therapy (also available in Spanish)

Understanding Lymphedema (for cancers other than breast cancer)

The following books are available from the American Cancer Society. Call us at 1-800-ACS-2345 to ask about costs or to place your order.

American Cancer Society's Guide to Pain Control

Cancer in the Family: Helping Children Cope with a Parent's Illness

Caregiving: A Step-By-Step Resource for Caring for the Person with Cancer at Home

National organizations and web sites*

In addition to the American Cancer Society, other sources of patient information and support include*:

Gynecologic Cancer Foundation

Toll-free number: 1-800-444-4441

Web site: www.thegcf.org

National Cancer Institute

Toll-free number: 1-800-4-CANCER (1-800-422-6237)

TTY: 1-800-332-8615

Web site: www.cancer.gov

National Coalition for Cancer Survivorship (NCCS)

Toll-free number: 1-888- 650-9127

1-877-622-7937(1-877-NCCS-YES) for some publications and Cancer Survivor Toolbox[®] orders

Web site: www.canceradvocacy.org

*Inclusion on this list does not imply endorsement by the American Cancer Society

No matter who you are, we can help. Contact us anytime day or night, for information and support. Call us at 1-800-ACS-2345 (1-800-227-2345) or visit our Web site at www.cancer.org

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1 · 800 · ACS-2345 or www.cancer.org