

VULVAR CANCER

factsheet



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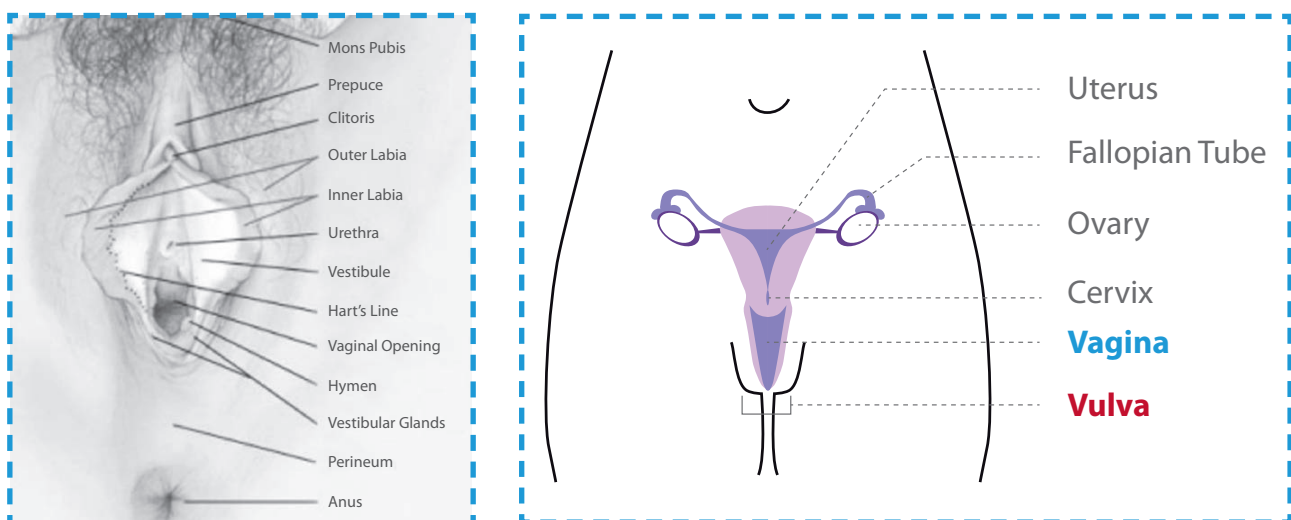
What are the vulva and vagina?

Women's genitalia can be divided into external and internal genital organs.

The **vulva** is the name of the female external genital organ. It consists of the labia, the prepuce, the clitoris and the vestibule. The **outer labia** (Latin for "lips") are two folds of skin and fatty tissue partially covered with hair which can vary greatly in size and shape. The **inner labia** are thinner folds of skin without fatty tissue which go from the clitoral hood to below the **vagina**. They can have small **glands** and can also vary in colour and shape. The **clitoris** is located below the point where the inner labia meet and can vary in size from a small pea to bigger than a fingertip. It becomes erect during sexual stimulation and can have varying levels of sensitivity. The **vestibule** is the inner area of the inner lips. It contains gland openings which produce secretions that increase with arousal. The **urethra** (which connects the bladder to the outside) opens into this area. In adults, the remnants of the **hymen** form a ring around the **vaginal opening**. The **mons pubis** is a hair-covered cushion of fat lying over the pubic bone.

The female internal genitalia consist of the vagina, the cervix, the uterus, the fallopian tubes and the ovaries.

Although the term "**vagina**" is colloquially used to refer to women's external genitalia, in medical terms the vagina is the canal that extends from the vestibule to the cervix and therefore acts as a passage from the uterus to the outside. It is a muscular-elastic organ which dilates during childbirth and through which blood flows out during menstruation. The vagina is about 7-10 cm long and coated with mucosa, similar to the inner coating of the mouth.



What is vulvar cancer?

Vulvar cancer is an abnormal growth of cells that can occur on any part of the **vulva**. Cancer of the **vulva** is not common. It accounts for 3-5% of all gynaecological cancers in the world. This puts it in fourth place among gynaecological malignancies. The first three places are occupied by cancer of the uterus, ovaries, and cervix. Every year about 27,000 women worldwide are diagnosed with vulvar cancer.⁽¹⁾

There are several types of vulvar cancer; they are classified according to which cells become abnormal.

- **Squamous cell carcinomas** are the most common type of vulvar cancer (over 90%). This cancer develops from squamous cells, which make up the outer layer of the skin. It occurs most often on the labia majora and minora, but can be found anywhere on the **vulva**.
- **Malignant melanoma** is the second most common type of vulvar cancer. Melanoma develops from the color producing skin cells called “melanocytes”, and is most often found on the labia minora.

Other types of vulvar cancer include:

- Adenocarcinomas (which develop from gland cells)
- Paget’s disease (a rare disease of the glandular cells)
- Sarcomas (which develop from muscle cells)
- Basal cell carcinomas (which develop from the cells which form the base layer of the skin)

The statistic incidence of squamous cell carcinomas (SCCs) has been calculated between 2 and 7 cases per 100,000 women/year. These incidence rates have increased since the 1970s. Historically viewed as a cancer affecting post-menopausal women, vulvar cancer has seen a shift in age of onset. Women over 65 years old remain the most vulnerable to developing vulvar cancer, but recent years have marked a decline in the mean age of diagnosis, largely due to rising rates of HPV (Human Papilloma Virus) infection and tobacco use.^(1,2) Indeed, in the case of women below the age of 50, cancer tends to be HPV-associated which implies a coincidence of about 20% for cervical and anal preinvasive or invasive disease.

What causes vulvar cancer?

Less than 30% of vulvar carcinomas are caused by HPV. HPV is a DNA virus capable of inducing a malignant transformation of epithelial cells and causing cervical, anal, vulvar, vaginal, penile and some oral cancers (so it can affect both women and men). While genital HPV infections are very common, the **persistence of the virus**, not opposed by the immune system, is a rare event. The persistence of high-risk HPV infections for years **can lead to cancer**.

Most of vulvar carcinomas are non-HPV related. They often develop from vulvar chronic inflammatory disease such as **lichen sclerosus or lichen planus**. HPV-related vulvar carcinomas generally affects younger women, while non HPV-related vulvar carcinomas are more frequent in older women.⁽²⁾

What are the symptoms of vulvar cancer?

Early diagnosis of vulvar cancer simplifies treatment and results in the best outcome both for disease free survival and quality of life. Examining your **vulva** regularly may help you discover abnormal changes, leading to an early diagnosis. Women should examine their **vulva** using a mirror, looking for changes in skin color, texture, or irritated skin. Any change should be reported to your health care provider.⁽³⁾

For more information of vulvar self-examination please consult our brochure “Vulvar self-examination: A guide for patients”

Signs and symptoms of vulvar cancer can include:

- A cut or sore on the vulva that does not heal (an ulcer)
- A lump, a warty growth, or a mass on the vulva
- Skin on the vulva that looks lighter, darker, or of a different colour than the surrounding skin, or that feels more firm
- Any change in colour or size of a mole that was already on the vulva
- Persistent vulvar itching or burning
- Painful urination (peeing)
- Bleeding from the vulva

It is important not to use creams or lotions that may cover up the symptoms and delay diagnosis. For example, creams that contain local anaesthetics that numb the area, or corticosteroids may relieve the symptoms. If you have a symptom that bothers you, get professional advice in order to get the right diagnosis and treatment.



How is vulvar cancer diagnosed?

When an abnormal change in the vulva is found, a **biopsy** (the removal of a small piece of involved tissue) is necessary to provide the correct diagnosis and decide on the best treatment. A biopsy is performed after injecting a small amount of local anaesthetic around the area to be sampled. A magnifying lens may be used to help to identify the best area to biopsy. More than one biopsy may be necessary for some conditions. The tissue will be sent to a laboratory for examination (**histology**).

Stages of vulvar cancer

Once vulvar cancer is diagnosed, it is important to check that it involves only the vulva and has not spread to other parts of the body (this is called staging). Correct staging of the disease is important in order to determine the correct treatment. Staging may include other testing procedures (PET scan, CT scan, MRI, etc.) before a final treatment plan is made.

How is vulvar cancer treated?

Treatment of vulvar cancer is directly related to the stage, type, and location of the disease. The patient's general state of health is also important in determining the appropriate treatment.

- **Surgery** is the most common treatment for vulvar cancer. The size of the lesion and its location on the **vulva** will determine what type of surgery is needed. Often, when the area is small, removal of the cancer lesion and a rim (about 1 cm) of normal tissue is sufficient. Larger lesions require removal of more tissue and structures. The most drastic intervention (for more advanced cases) is a complete vulvectomy including the clitoris.

Lymph nodes in the groin must be assessed for the presence of disease that affects prognosis. Different techniques are available from removal of single lymph node (**Sentinel node**) to a complete nodal removal (**Lymphadenectomy**), according to the extent of tumours.

In all cases the use of compression stockings and exercise are being recommended to prevent lymphedema, which is an accumulation of fluid usually drained through the lymphatic system. When lymph nodes are removed, this fluid may collect in the legs. However, even with these preventive measures, there is no guarantee to not develop lymphedema.

Other treatment options:

- **Radiation therapy** uses high energy (gamma rays, electrons, protons, neutrons) to kill cancer cells and shrink the tumor or the residual tumoral cells. Radiation can be given before or after surgery, and is sometimes used to treat the lymph nodes in the groin and pelvis.
- **Chemotherapy** uses medication to kill cancer cells. The medication may be given orally (by mouth) or intravenously (directly in the veins). ^(4,5)

How can vulvar cancer be prevented?

The causes of vulvar cancers are not fully known. However, certain factors may increase the risk of vulvar cancer. One strategy for cancer prevention is to avoid any of the modifiable risk factors.

Risk factors include:

- **Human papillomavirus (HPV) infection:** HPV types 16 is thought to be responsible for a part of the vulvar cancers in younger women. It can start as a **precancerous condition called vulvar high grade squamous intraepithelial lesion (VHSIL)**. Reducing the number of sexual partners, avoiding sex with someone with a history of multiple sexual partners, and avoiding sex at an early age can lessen the risk of HPV infection. The majority of preinvasive vulvar lesions are related to HPV infections. Identifying and treating them substantially reduce the progression to invasive cancer.
- **Chronic vulvar inflammation and skin conditions** - long term irritation and inflammation of the vulva can increase the risk of vulvar cancer. **Lichen sclerosus and lichen planus** are long term skin conditions that can affect the vulva. These conditions may increase the possibility of developing vulvar cancer with their chronic oxidative damage to vulvar skin. If you have one of these skin conditions, you must see your healthcare provider on a regular basis. If your provider notices any suspicious changes, a biopsy will be taken to rule out a preinvasive or an invasive vulvar disease.
- **Cigarette smoking:** smoking weakens the immune system. This makes women who smoke more likely to develop persistent infections, including HPV.
- **HIV/Immunosuppression:** an HIV (human immunodeficiency virus) infection, or any other kind of immunosuppression, decreases the body's ability to fight off an infection, and increases the chance of getting a variety of diseases, including HPV related vulvar cancer.⁽⁶⁾

A second preventive strategy is to **identify and treat any precancerous and preinvasive lesions early**. These strategies greatly reduce the chances of developing invasive cancer both HPV and non-HPV related as described above.

Finally, HPV related vulvar cancer could be prevented by using **HPV vaccine** before the onset of HPV infection.

What is the follow-up care?

Once a diagnosis of vulvar cancer has been made and treatment is complete, patients will need regular follow-up visits with their healthcare provider to assure that they remain cancer free.

- As with any cancer diagnosis, patients may need **continued psychological support** throughout their recovery process. What is inherent to vulvar cancer is that this affects one of the most private, intimate parts of a woman's body. Some women may experience the feeling of shame. In some cultures the stigma surrounding sexuality and sexually transmitted disease may lead to the reluctance to talk about the symptoms. Onco-sexologists and onco-psychologists are trained to tackle these issues and women should address them if they feel the need.

- **A sexual therapist** can assist with maintaining a sex life with your partner. Sexuality in a relationship is a complex task where physical and emotional wellbeing may play a crucial role. Some women may need more time to adapt to their changed body image and to overcome fear from pain or disappointment. Every healthy relationship should place respect to the other first. For tips on **Sexuality and intimacy following gynaecological cancer treatment** please consult the ENGAGe brochure of the same name. ⁽⁷⁾
- Cancer treatment and its physical as well as psychological burden may alter libido of some women. In and ENGAGe brochure entitled **Loss of libido after cancer** ⁽⁸⁾ there are many tips on intimacy and sexual closeness with your partner without penetration that can be useful to explore.

Is vulvar reconstruction possible?

The extent to which vulvar reconstruction is possible, depends on the size and extension of the tumour that needs to be removed. For small, localised tumours, reconstruction is not necessary, and the surgery only causes minor aesthetic changes. In the case of more extensive surgery, vulvar reconstruction aims to restore anatomy and the physiological function where possible. Skin from the thigh, the gluteus or the abdomen can be used to replace the missing tissue. Surgical reconstruction may not be perfect. The texture of the skin may have a different feel and may be less sensitive. However, many patients confirm that learning to accept one's scars was a valuable step for them to self-love and appreciation of their strength that enabled them to fight the disease.

Stigma in vulvar cancer?

Is there a stigma in society when it comes to cancer, especially when it comes to gynaecological cancers? Many patient stories confirm that it is still a sad reality although a lot has been done in communication to provide facts to de-stigmatise gynaecological cancers. Vulvar cancer is mostly associated with older women and, when diagnosis at mature age, women and their life partners may ask questions how this is possible in a long-term relationship.

It is crucial to communicate on the HPV virus behaviour in the body and its manifestation. **Women who have a long time persistent HPV infection may develop HPV-related cancers, such as vulvar cancer, over time and these cancers may only manifest at later age.** This may cause emotional issues with women and their lifetime partners, especially in older women who are not sexually active anymore, or who have only had one sexual partner for a long time.

Conclusion

The aim of this brochure was to provide facts, references and relief to women suffering from vulvar cancer and their entourage.

Early detection is crucial to survival and it is only possible if women share any doubts with the gynaecologists early on.

Once diagnosed, support from families and friends is very important and knowing the right facts is key to destigmatising all gynaecological cancers.

References

- (1) Faber MT, Sand FL, Albieri V, et al. Prevalence and type distribution of human papillomavirus in squamous cell carcinoma and intraepithelial neoplasia of the vulva. *Int J Cancer*. 2017 Sep 15;141(6):1161–9.
- (2) Mancini S, Bucchi L, et al. AIRTUM Working Group. Trends in Net Survival from Vulvar Squamous Cell Carcinoma in Italy (1990-2015). *J Clin Med*. 2023 Mar 10;12(6):2172. doi: 10.3390/jcm12062172. PMID: 36983173; PMCID: PMC10054662.
- (3) Preti M, Selk A, Stockdale C, et al. Knowledge of Vulvar Anatomy and Self-examination in a Sample of Italian Women. *J Low Genit Tract Dis*. 2021 Apr 1;25(2):166-171. doi: 10.1097/LGT.0000000000000585. PMID: 33470738.
- (4) Oonk MHM, Planchamp F, et al. European Society of Gynaecological Oncology Guidelines for the Management of Patients with Vulvar Cancer - Update 2023. I., *Int J Gynecol Cancer*. 2023 Jul 3;33(7):1023-1043. doi: 10.1136/ijgc-2023-004486. PMID: 37369376; PMCID: PMC10359596.
- (5) Merlo S. Modern treatment of vulvar cancer. *Radiol Oncol*. 2020 Sep 22;54(4):371-376. doi: 10.2478/raon-2020-0053. PMID: 32960779; PMCID: PMC7585347.
- (6) The Vulvar Cancer, International Society for the Study of Vulvovaginal Disease Patient Information Committee, 2024. HYPERLINK „<http://www.issvd.org/publications/patient-handouts>“www.issvd.org/publications/patient-handouts
- (7) <https://engage.esgo.org/brochures/sexuality-intimacy-following-gynaecological-cancer-treatment/>
- (8) engage.esgo.org/brochures/loss-libido-cancer/



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ENGAGe recommends contacting your local patient association!



