

Cancer Association of South Africa (CANSA)



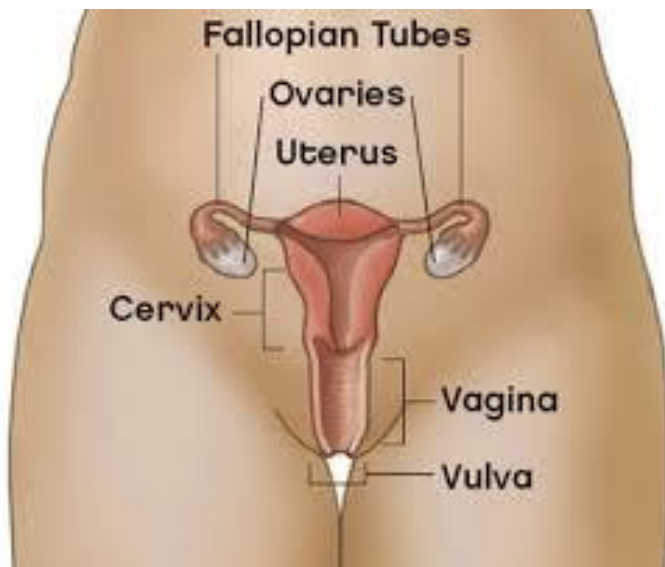
CANSA Fact Sheet on Cancer of the Vulva

Introduction

The vulva (from the Latin *vulva*) consists of the external genital organs in the female.

[Picture Credit: Vulva]

The vulva has many major and minor anatomical structures, including the labia majora, mons pubis, labia minora, clitoris, bulb of vestibule, vulval vestibule, greater and lesser vestibular glands, as well as the opening of the vagina. As the outer portal of the human uterus or womb, it protects its opening by a 'double door': the labia majora (large lips) and the labia minora (small lips). The vagina is a self-cleaning organ, sustaining healthy microbial flora that flow from the inside out.



The vulva has a sexual function - these external organs are richly innervated and provide pleasure when stimulated. The vulva also contains the opening of the female urethra, but apart from this has little relevance to the function of urination.

Cancer of the Vulva

Cancer is a disease in which cells in a particular part of the body grow out of control. Cancer is always named for the part of the body where it starts, even if it spreads to other body parts later.

It is not always clear what causes vulvar cancer. This cancer starts on the area of skin that surrounds the urethra and vagina. This area of skin is called the vulva.

Vulvar cancer is a rare cancer of the external female genitals, with symptoms including persistent itching, pain, changes in skin colour or thickening, a lump or sore, and bleeding. Key risk factors include Human Papillomavirus (HPV) infection and vulvar intraepithelial neoplasia (VIN), a precancerous condition.

Vulvar cancer treatment often starts with surgery to remove the cancer and a small amount of surrounding healthy tissue. Sometimes vulvar cancer surgery requires removing the entire vulva. The earlier vulvar cancer is diagnosed, the less likely an extensive surgery is needed for treatment.

[Picture Credit: Cancer of the Vulva]

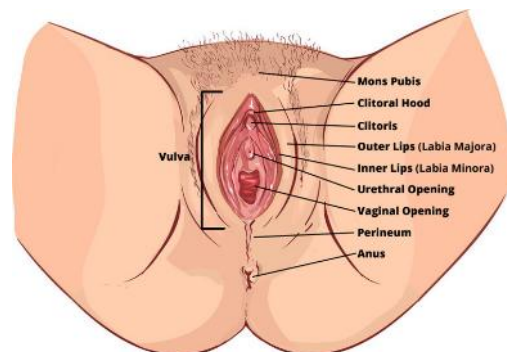


The Facts:

- Vulvar cancer is very rare, accounting for approximately 0.7 percent of all cancers in women. According to the National Cancer Registry (2023) vulvar cancer represents 2.23% of cancers among south African women.
- The HPV vaccine can prevent the strains of HPV responsible for most cervical, vaginal and vulvar cancers.
- Risk factors that may increase a woman's chances of developing vulvar cancer include age, infection with certain types of HPV, smoking and HIV infection.
- Symptoms of vulvar cancer include severe itching, burning and pain on the vulva.

Vulvar cancer is a rare cancer that forms in the tissues of the vulva – it develops slowly over several years. 'Vulva' is the collective name for all the female external sex organs, or genitals. it includes:

- Opening of the vagina
- Labia minora (inner lips)
- Labia majora (outer lips)
- The outer part of the clitoris
- Mons pubis
- Opening of the urethra
- Perineum



[Picture Credit: Vulvar Anatomy]

Vulvar cancer usually develops slowly over several years. Precancerous areas of tissue (lesions) typically develop first. Healthcare providers usually discover the abnormal growth in the outermost layer of your skin. These precancerous lesions are called vulvar intraepithelial neoplasia (VIN). (Mayo Clinic, 2025; Johns Hopkins Medicine, 2025; Foundation for Women's Cancer, 2025).

Islam, M.E., Debnath, K.C., Moniruzzaman, R., Okuyama, K., Islam, S. & Dongre, H.N. 2025.

"The present review aimed to elucidate the roles of extracellular matrix (ECM) components in the progression of vulvar squamous cell carcinoma (VSCC) and explore potential therapeutic avenues for this type of malignancy. This exploration holds promise for identifying precise molecular targets within the ECM milieu, thus facilitating the development of innovative therapeutic modalities tailored to disrupt these interactions and ultimately improve patient outcomes in VSCC. The dysregulated ECM serves as a potent driver of SCC tumor progression, orchestrating key processes such as angiogenesis, inflammation and stromal cell behavior. Yet, the exploration of ECM role in VSCC is still in its early stages. Recent research highlights the critical role of ECM organization and expression within the tumor microenvironment (TME) in influencing key aspects of VSCC, including tumor staging, grading, metastasis, invasion and patient survival. Cancer-associated fibroblasts play a pivotal role in this dynamic by engaging in reciprocal interactions with VSCC cells, leading to significant ECM alterations and creating an immune-suppressive TME. This hinders antitumor immunity and fosters therapeutic resistance in VSCC treatment. The dysregulated ECM in VSCC drives tumor progression, metastasis

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and affects patient survival. Targeting ECM, along with emerging therapies such as immune checkpoint blockade, offers promise for improved VSCC treatment outcomes.”

Conflict of interest statement

The authors declare that they have no competing interests.

Olawaiye, A.B., Cuello, M.A., Beriwal, S. & Rogers, L.J. 2025.

“Vulvar cancer is an uncommon gynecological malignancy primarily affecting postmenopausal women. No specific screening exists and the most effective strategy to reduce the incidence of vulvar cancer is the opportune treatment of predisposing and preneoplastic lesions associated with its development. Although vulvar cancer may be asymptomatic, most women present with vulvar pruritus or pain or have noticed a lump or ulcer. Therefore, any suspicious vulvar lesion should be biopsied to exclude invasion. Once established, the most common subtype is squamous cell carcinoma. The treatment of vulvar cancer depends primarily on histology, clinical, radiological, or surgical staging. Treatment is predominantly surgical and/or adjuvant radiation therapy, particularly for squamous cell carcinoma, although definitive concurrent chemoradiation is an effective alternative, particularly for advanced tumors. In those cases where vulvar cancer presents with distant metastases, systemic therapy is modeled after cervical cancer, a more common cancer where there is more robust data to guide treatment. Management should be individualized and carried out by a multidisciplinary team in a cancer center experienced in the treatment of these tumors.”

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The authors have no conflicts of interest.

Tumour Grade vs Tumour Stage

A cancer’s grade describes how abnormal the cancer cells and tissue look under a microscope when compared to healthy cells. Cancer cells that look and organise most like healthy cells and tissue are low grade tumours. Doctors describe these cancers as being well differentiated. Lower grade cancers are typically less aggressive and have a better prognosis.

While a grade describes the appearance of cancer cells and tissue, a cancer’s stage explains how large the primary tumour is and how far the cancer has spread in the patient’s body.

There are several different staging systems. Many of these have been created for specific kinds of cancers. Others can be used to describe several types of cancer.

One common system that many people are aware of puts cancer on a scale of 0 to IV.

- Stage 0 is for abnormal cells that haven’t spread and are not considered cancer, though they could become cancerous in the future. This stage is also called “in-situ.”
 - Stage I through Stage III are for cancers that haven’t spread beyond the primary tumour site or have only spread to nearby tissue. The higher the stage number, the larger the tumour and the more it has spread.
 - Stage IV cancer has spread to distant areas of the body.
- (University of Texas, MD Anderson Cancer Center. 2025).

The International Classification of Diseases 10th version (ICD-10)

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The International Classification of Diseases (ICD) is designed to promote international comparability in the collection, processing, classification, and presentation of mortality statistics. This includes providing a format for reporting causes of death on the death certificate.

ICD serves a broad range of uses globally and provides critical knowledge on the extent, causes and consequences of human disease and death worldwide via data that is reported and coded with the ICD. Clinical terms coded with ICD are the main basis for health recording and statistics on disease in primary, secondary and tertiary care, as well as on cause of death certificates. These data and statistics support payment systems, service planning, administration of quality and safety, and health services research. Diagnostic guidance linked to categories of ICD also standardizes data collection and enables large scale research.

For more than a century, the International Classification of Diseases (ICD) has been the basis for comparable statistics on causes of mortality and morbidity between places and over time. Originating in the 19th century, the latest version of the ICD, ICD-11, was adopted by the 72nd World Health Assembly in 2019 and came into effect on 1st January 2022.

The ICD-10 Code for malignant neoplasm of vulva, unspecified, is C51.9; moderate dysplasia of vulva N90.1; vulvar intraepithelial neoplasia II [VIN II] N90.1.
(World Health Organization, 2025).

Incidence of Cancer of the Vulva in South Africa

According to the latest edition of the National Cancer Registry (2023) the following number of cases of cancer of the vulva cases was histologically diagnosed in South Africa during 2023:

Group - Females 2023	Actual No of Cases	Estimated Lifetime Risk	Percentage of All Cancers
All females	1 031	1:345	2,23%
Asian females	18	1:572	1,23%
Black females	854	1:3299	3,95%
Coloured females	58	1:527	0,98%
White females	101	1:408	0,55%

The frequency of histologically diagnosed cases of cancer of the vulva in South Africa for 2023 was as follows (National Cancer Registry, 2023):

Group - Females 2023	0 – 19 Years	20 – 29 Years	30 – 39 Years	40 – 49 Years	50 – 59 Years	60 – 69 Years	70 – 79 Years	80+ Years
All females	2	28	247	391	171	100	63	29
Asian females	0	0	1	3	4	4	4	2
Black females	2	26	224	359	140	60	34	9
Coloured females	0	1	13	13	9	12	6	4
White females	0	1	9	16	18	24	19	14

N.B. In the event that the totals in any of the above tables do not tally, this may be the result of uncertainties as to the age, race or sex of the individual. The totals for 'all males' and 'all females', however, always reflect the correct totals.

Causes and Risk Factors for Cancer of the Vulva

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Several risk factors for cancer of the vulva have been identified. Researchers have made a lot of progress in understanding how certain changes in DNA can cause normal cells to become cancerous. DNA is the chemical that carries the instructions for nearly everything the body's cells do.

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 *tumour suppressor genes*. Cancers can be caused by DNA mutations (defects) that turn on oncogenes or turn off tumour 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.

The second process by which vulvar cancers develop does not involve HPV infection. Vulvar cancers not linked to HPV infection (the keratinising subtype) are usually diagnosed in older women (over age 55). These women often show mutations of the p53 tumour 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.

According to the IGCS, although the exact cause of vulvar cancer is not known, certain factors appear to increase the risk of the disease, including:

- Increasing age. The risk of vulvar cancer increases with age, though it can occur at any age. The average age at diagnosis is 65.
- Being exposed to human papillomavirus (HPV). HPV is a sexually transmitted infection that increases the risk of several cancers, including vulvar cancer and cervical cancer. Many young, sexually active people are exposed to HPV, but for most the infection goes away on its own. For some, the infection causes cell changes and increases the risk of cancer in the future.
- Smoking. Smoking increases the risk of vulvar cancer.
- Having a weakened immune system. People who take medications to suppress the immune system, such as those who've undergone organ transplant, and those with conditions that weaken the immune system, such as human immunodeficiency virus (HIV), have an increased risk of vulvar cancer.
- Having a history of precancerous conditions of the vulva. Vulvar intraepithelial neoplasia is a precancerous condition that increases the risk of vulvar cancer. Most instances of vulvar intraepithelial neoplasia will never develop into cancer, but a small number do go on to become invasive vulvar cancer. For this reason, your doctor may recommend treatment to remove the area of abnormal cells and periodic follow-up checks.
- Having a skin condition involving the vulva. Lichen sclerosus, which causes the vulvar skin to become thin and itchy, increases the risk of vulvar cancer.

(International Gynecologic Cancer Society (IGCS), 2024).

Signs and Symptoms of Cancer of the Vulva

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Signs and symptoms of cancer of the vulva may include:

- itching that does not go away
- pain and tenderness
- a lump
- bleeding that is not from menstruation
- skin changes, such as colour changes or thickening
- abnormal bleeding
- burning
- painful urination
- wart-like growths (similar to genital warts)
- change in the appearance of an existing mole (specific to vulvar melanoma)
- itching
- local pain

(NHS UK, 2025).

Diagnosis of Cancer of the Vulva

If someone suspects that they may have cancer of the vulva, they should visit a medical practitioner that specialises in women's cancers (gynaecological cancer specialist).

Apart from a close examination of the vulval area, the doctor will also do a general medical examination of the patient to determine the general condition of health of the person.

The doctor may use a bright light and a magnifier to examine the vulva, so that the skin can be seen more clearly. He/she may then take small samples of tissue (biopsies) from any areas that look unusual. This may be done under a local anaesthetic.

The doctor will also usually do an internal examination to check the vagina, cervix and the neck of the womb for any abnormality. The doctor will use a speculum (a plastic or metal instrument) to hold the vaginal walls open. He/she may also take a cervical smear test (a small sample of cells taken from the cervix). The doctor may also examine the back passage (anus).

(Mayo Clinic, 2025).

Treatment of Cancer of the Vulva

Treatment options for cancer of the vulva depend on the type and stage of the cancer as well as the person's overall health and preferences, and may include:

Surgery

Operations used to treat cancer of the vulva include:

- Removing the cancer and a margin of healthy tissue (excision).
- Removing a portion of the vulva (partial vulvectomy).
- Removing the entire vulva (radical vulvectomy).
- Extensive surgery for advanced cancer.
- Reconstructive surgery.
- Possible surgery to remove nearby lymph nodes.

Radiation therapy

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Radiation therapy uses high-powered energy beams, such as X-rays, to kill cancer cells. Radiation therapy for cancer of the vulva is usually administered by a machine that moves around one's body and directs radiation to precise points on the skin (external beam radiation).

Chemotherapy

Chemotherapy is a drug treatment that uses chemicals to kill cancer cells. Chemotherapy drugs are typically administered through a vein in the arm or by mouth. (Foundation for Women's Cancer, 2025).

Follow-up Tests after Treatment

After completing treatment for cancer of the vulva, the doctor may recommend periodic follow-up examinations to look for a cancer recurrence.

About Clinical Trials

Clinical trials are research studies that involve people. They are conducted under controlled conditions. Only about 10% of all drugs started in human clinical trials become an approved drug.

Clinical trials include:

- Trials to test effectiveness of new treatments
- Trials to test new ways of using current treatments
- Tests new interventions that may lower the risk of developing certain types of cancers
- Tests to find new ways of screening for cancer

The South African National Clinical Trials Register provides the public with updated information on clinical trials on human participants being conducted in South Africa. The Register provides information on the purpose of the clinical trial; who can participate, where the trial is located, and contact details.

For additional information, please visit: <https://pactr.samrc.ac.za/>

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