

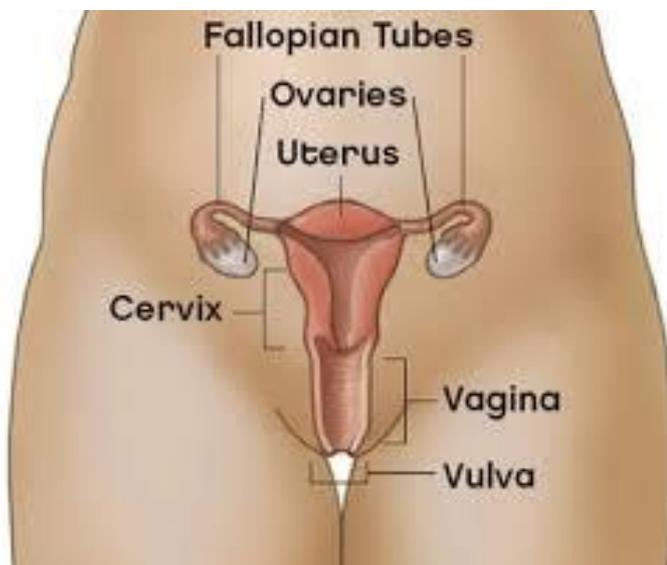
**Fact Sheet
on
Cancer of the Vulva**

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. Cancer of the vulva is a malignant (cancerous) invasive growth and multiplication of cells in the vulva and mainly affects women later in life.

Incidence of Cancer of the Vulva in South Africa

According to the National Cancer Registry (2014) the following number of cancer of the vulva cases was histologically diagnosed in South Africa during 2014:

Group - Females 2014	Actual No of Cases	Estimated Lifetime Risk	Percentage of All Cancers
All females	343	1:789	0,91%
Asian females	16	1:386	1,38%
Black females	259	1:833	1,61%
Coloured females	15	1:1 508	0,37%
White females	53	1:722	0,32%

The frequency of histologically diagnosed cases of cancer of the vulva in South Africa for 2014 was as follows (National Cancer Registry, 2014):

Group - Females 2014	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	0	15	97	71	70	35	28	15
Asian females	0	0	0	1	4	5	4	2
Black females	0	14	90	60	49	17	8	5
Coloured females	0	0	0	2	6	4	2	1
White females	0	1	4	7	10	9	13	7

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

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.

[Picture Credit: Cancer of Vulva]

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.

Lichen Sclerosus

Lichen sclerosus appears in:

- women (often after menopause)
- men (uncommon)
- children (rare)

[Picture Credit: Lichen Sclerosus]

Early in the disease, small white spots appear on the skin. The spots are usually shiny and smooth. Later, the spots grow into bigger patches. The skin on the patches becomes thin and crinkled. Then the skin tears easily, and bright red or purple bruises are common. Sometimes, the skin becomes scarred. If the disease is a mild case, there may be no symptoms.



Other symptoms are:

- itching (very common)
- discomfort or pain
- bleeding
- blisters

Signs and Symptoms of Cancer of the Vulva

Signs and symptoms of cancer of the vulva may include:

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

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- itching
- local pain

Diagnosis of Cancer of the Vulva

If someone suspects that she may have cancer of the vulva she 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 her general condition of health.

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).

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

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.

Rajaram, S. & Gupta, B. 2015.

“Vulvar cancer is an uncommon malignancy and accounts for around 5% of all gynaecologic cancers. Incidence rates have increased for young adults and may be linked to increasing HPV prevalence. Treatment of vulvar cancer has evolved from 'en-bloc' surgery with high morbidity to more conservative approaches without compromising oncological safety. In recent years sentinel node evaluation has been advocated in early stage cancers to reduce complications of inguino-femoral lymphadenectomy. Treatment decision is still a challenge as there is no standard recommended treatment strategy. Neoadjuvant chemoradiation is an effective modality for locally advanced vulvar cancer, as it reduces tumour size and renders the lesion operable. Primary chemoradiation without post treatment surgery has been used as an alternative treatment to avoid extensive radical surgery and complex reconstructive procedures.”

Micheletti, L. & Preti, M. 2014.

“The standard radical mutilating surgery for the treatment of invasive vulval carcinoma is, today, being replaced by a conservative and individualised approach. Surgical conservative modifications that are currently considered safe, regarding vulval lesion, are separate skin vulval-groin incisions, drawn according to the lesion diameter, and wide local radical excision or partial radical vulvectomy with 1-2 cm of clinically clear surgical margins. Regarding inguinofemoral lymph nodes management, surgical conservative modifications not compromising patient survival are omission of groin lymphadenectomy only when tumour stromal invasion is ≤ 1 mm, unilateral groin lymphadenectomy only in well-lateralised early lesions and total or radical inguinofemoral lymphadenectomy with preservation of femoral fascia when full groin resection is needed. Sentinel lymph node dissection is a promising technique but it should not be routinely employed outside referral centres. Pelvic nodes are better managed by radiation. Locally advanced vulval carcinoma can be managed by ultraradical surgery, exclusive radiotherapy or chemoradiation.”

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](http://www.sanctr.gov.za/) 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: www.sanctr.gov.za/

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Cancer of Vulva

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Lichen Sclerosus

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Vulva

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