

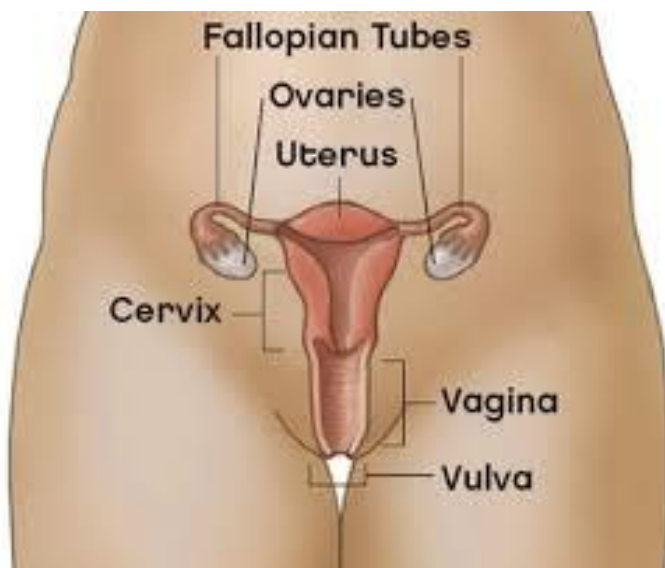
**Fact Sheet  
on  
Cancer of the Vulva**

**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.

**Staples, J.N. & Duska, L.R. 2019.**

“The Pap smear is the only proven screening intervention in the field of gynecologic oncology. Women should receive treatment for precancerous conditions of the cervix, vulva, vagina, and

endometrial lining. Women with inherited conditions should consider having a risk-reducing surgery once they have finished childbearing. The human papilloma virus vaccination should be offered to all girls and boys aged 11 to 12 years, and can also be given as early as age 9 and through 26 years of age.”

### Incidence of Cancer of the Vulva in South Africa

According to the outdated National Cancer Registry (2017), known for under reporting, the following number of cancer of the vulva cases was histologically diagnosed in South Africa during 2017:

Group - Females 2017	Actual No of Cases	Estimated Lifetime Risk	Percentage of All Cancers
All females	541	1:580	1,30%
Asian females	12	1:667	0,93%
Black females	439	1:573	2,35%
Coloured females	29	1:906	0,64%
White females	61	1:607	0,36%

The frequency of histologically diagnosed cases of cancer of the vulva in South Africa for 2017 was as follows (National Cancer Registry, 2017):

Group - Females 2017	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	5	34	186	150	63	57	28	18
Asian females	0	0	1	0	1	2	6	2
Black females	4	28	175	133	51	33	9	6
Coloured females	0	1	7	7	3	5	5	2
White females	1	5	3	11	8	17	8	8

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.

According to **Bruni, et al.**, (2019), the burden of cervical cancer for South Africa for 2018 is estimated as:

- Annual number of cases of cancer of the vulva 2 262
- Annual number of deaths due to cancer of the vulva 210



### 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.

[Picture Credit: Cancer of Vulva]

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.

### **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)
- 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).

**Michalski, B.M., Pfeifer, J.D., Mutch, D. & Council, M.L. 2020.**

**Background:** Vulvar cancers, although rare, are becoming an increasingly serious threat to women's health. Cancer of the vulva accounted for 0.3% of all new cancers in the United States in 2019, with 6,070 newly diagnosed cases. This review details the epidemiology, pathogenesis, diagnosis, staging, and treatment of vulvar malignancies.

**Objective:** To review cancer entities of the vulva, including vulvar intraepithelial neoplasms, squamous cell carcinoma (SCC), malignant melanoma, basal cell carcinoma, neuroendocrine tumors, and adenocarcinomas.

**Materials and methods:** Literature review using PubMed search for articles related to cancer of the vulva.

**Results:** Vulvar intraepithelial neoplasms represent premalignant precursors to SCC of the vulva. There are several different histopathologic subtypes of SCC, and treatment is dependent on characteristics of primary tumor and lymph node involvement. Melanoma is the second most common cancer to affect the vulva, and staging is based on tumor, node, and metastatic spread.

**Conclusion:** Vulvar malignancies are rare, and diagnosis is dependent on biopsy and pathologic evaluation. Treatment for vulvar malignancies depends on histopathologic diagnosis but ranges from wide local excision with or without lymph node biopsy or dissection to radiation therapy with chemo- or immunotherapy. Overall survival varies by diagnosis.

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**Tan, A., Bieber, A.K., Stein, J.A. & Pomeranz, M.K.** 2019.

“Vulvar malignancies represent a serious gynecologic health concern, especially given the increasing incidence over the past several decades. Squamous cell carcinoma and melanoma are common subtypes, although other neoplasms, such as basal cell carcinoma and Paget disease of the vulva, might be seen. Many vulvar cancers are initially misdiagnosed as inflammatory conditions, delaying diagnosis and worsening prognosis. It is essential that dermatologists are familiar with characteristic findings for each malignancy to ensure appropriate diagnosis and management. Herein, we review the unique epidemiologic and clinical characteristics of each major vulvar malignancy, as well as discuss their respective prognoses and current management recommendations.”

### **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.

**Michalski, B.M., Pfeifer, J.D., Mutch, D. & Council, M.L.** 2020.

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**Conclusion:** Vulvar malignancies are rare, and diagnosis is dependent on biopsy and pathologic evaluation. Treatment for vulvar malignancies depends on histopathologic diagnosis but ranges from wide local excision with or without lymph node biopsy or dissection to radiation therapy with chemo- or immunotherapy. Overall survival varies by diagnosis.

**Frøding, L.P., Zobbe, V., Fokdal, L.U., Niemann, I.C.S., Nøtrup, T.J., Kristensen, E., Christiansen, A.P., Loft, A. & Schnack, T.H.** 2019.

“This review summarises the current guidelines for vulva cancer in Denmark. Vulva cancer is a rare disease. The diagnosis is often delayed, which results in large tumours and regional spread. The most

important prognostic factor is inguinal lymph node metastases. Staging and treatment is centralised to two hospitals. Primary treatment is wide local excision combined with removal of either inguinal sentinel nodes or lymphadenectomy. Treatment is associated with considerable morbidity, and supportive care is often necessary. Local curable recurrences are common. Relapses in the groin are associated with a poor prognosis. Thus, long term follow-up is essential. \*) On behalf of Dansk Gynækologisk Cancer Gruppe for vulvacancer.”

### 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.

**Singareddy, R., Bajwa, H.K., Alluri, K.R., Raju, K.V.V.N., Rao, T.S. & Rajeev, G. 2019.**

**AIM:** The aim of this study was to evaluate the patterns of recurrence and outcomes in patients with vulvar cancer treated with surgery, with or without postoperative radiotherapy (RT) or RT alone.

**MATERIALS AND METHODS:** The hospital records were retrospectively analyzed from February 2007 to May 2016. All patients that had biopsy-proven carcinoma vulva were included for analysis. Surgery was done by simple or radical vulvectomy with or without lymph node dissection. Radical RT dose was 60 Gy (photons alone or with electron boost). Adjuvant RT was administered in case of node positivity or positive margins.

**RESULTS:** A total of 76 patients were diagnosed as squamous cell carcinoma of vulva. The median age was 59 years. About 59 patients were treated by surgery and 17 patients were treated by radical RT. The most common stage was IB. Adjuvant RT was administered in 10 patients. About 13 patients (22%) underwent inguinal lymphadenectomy. At a median follow-up of 35 months, 49 patients (64.4%) were alive, 27 died, and 6 patients were lost to follow-up. Locoregional recurrence rate was 24.5% in radical surgery group, 12% in surgery plus adjuvant RT group, and 47% in radical RT group. Distant metastasis rate was 4%. The progression-free survival at 3 years was 45.3% and 35.5% in the surgery group and radical RT group, respectively.

**CONCLUSION:** Surgery resulted in favorable local control rates in early-stage carcinoma vulva. Adjuvant RT is indicated in case of nodal positivity or positive margins. Radical RT can be considered as an alternative in patients with comorbidities.

**Nandwani, M., Barmon, D., Begum, D., Liegise, H. & Katak, A.C. 2019.** An overview of vulvar cancer: a single-center study from Northeast India. *J Obstet Gynaecol India*. 2019 Dec;69(6):541-545. doi: 10.1007/s13224-019-01261-z. Epub 2019 Aug 16.

**STUDY:** Carcinoma vulva is a rare cancer of the female genital tract. It mostly presents in postmenopausal women. The treatment of vulvar cancer is surgery, chemoradiation, radiotherapy or a combination of all modalities. Here, we present a study of 33 cases of carcinoma vulva over a period of 2 years at a Northeast India regional cancer institute describing its demographic features and treatment outcomes.

**METHODOLOGY:** A retrospective cohort study of vulvar cancer diagnosed at Northeast India regional cancer institute from January 2017 to December 2018.

**RESULTS:** A total of 33 cases of biopsy proven carcinoma (Ca) vulva were studied. Maximum number of cases belonged to the age group: 60-69 years (39.4%). 66.67% cases had palpable inguinal lymph nodes at presentation, and 100% had squamous cell carcinoma on histopathology. Maximum number of cases belonged to stage III (44.8%), and least number of cases belonged to stage IV (10.3%) of FIGO 2009 staging of Ca vulva. 87.9% cases underwent treatment, and 12.1% were lost to follow-up. Out of the cases who underwent treatment, 55.2% cases were taken up for primary surgery and 44.8% cases for primary radiotherapy. 75% cases who underwent surgery received adjuvant radiotherapy. No complication was seen in patients post-radiation. But, 6.25% patients post-surgery developed lymphocyst and 18.75% patients developed wound necrosis ( $p > 0.05$ ).

**CONCLUSION:** Vulvar cancer is not a common malignancy of the female genital tract that presents in sixth and seventh decades of life and often with palpable inguinal lymph nodes. Though early stages of Ca vulva are treated by surgery, the incidence of immediate postoperative complications in our study was more as compared to post-radiotherapy. Also, maximum patients in the present study post-surgery received adjuvant radiotherapy. Thus, radiotherapy can be considered as the primary treatment modality for patients with early as well as advanced vulvar carcinoma.

### **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: [www.sanctr.gov.za/](http://www.sanctr.gov.za/)

### **Medical Disclaimer**

This Fact Sheet is intended to provide general information only and, as such, should not be considered as a substitute for advice, medically or otherwise, covering any specific situation. Users should seek appropriate advice before taking or refraining from taking any action in reliance on any information contained in this Fact Sheet. So far as permissible by law, the Cancer Association of

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#### **Vulva**

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