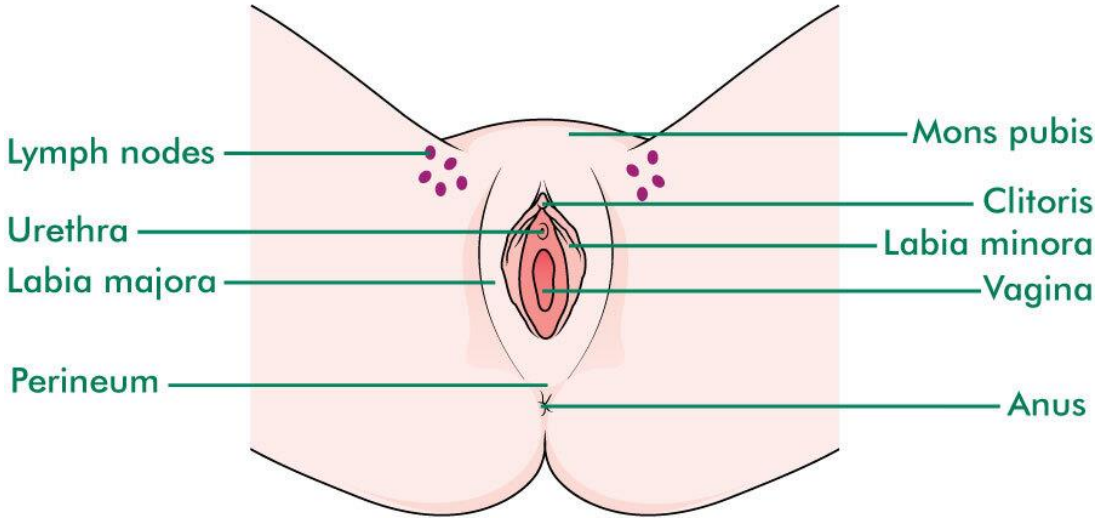
VULVAL CANCER

The Facts

Cancer of the vulva is rare with around 55 cases diagnosed each year in Ireland. It is more common in women over the age of 65, however, the number of younger women affected is increasing.

Vulval cancer is a type of cancer that occurs on the outer surface area of the female genitalia. The vulva is the area of skin that surrounds the opening of the vagina (sometimes called the vestibule), the labia majora (outer lips), the labia minora (inner lips), and the clitoris.



There are many benign (non-cancerous) causes for the symptoms below, but if you experience any of them, you should see your GP:

- ***itchiness, burning, pain/discomfort of the vulva***
- ***an ulcer, swelling or lump that does not go away***
- ***changes to the texture/feel of the skin***
- ***changes to the colour of the skin***
- ***bleeding from the vulva or vagina***

Cancer of the vulva may take years to develop and there is usually a pre-cancerous change. This is called Vulval Intraepithelial Neoplasia or VIN which can be often treated with surgical removal, topical creams or laser therapy.

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Risk Factors

Although the exact cause of vulvar cancer isn't known, certain factors appear to increase your 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 infection increases the risk of several cancers, including vulval 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 cigarettes increases the risk of vulval 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 vulval cancer.
- **Having a history of precancerous conditions of the vulva.** Vulval intraepithelial neoplasia (VIN) is a precancerous condition that increases the risk of vulval cancer. Most cases of VIN 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 vulval skin to become thin and itchy, increases the risk of vulval cancer.

Diagnosis

Your history of symptoms will be taken by your GP or Gynaecologist and an examination of the vulva is required. In order to diagnosis vulval cancer a small biopsy is required. This confirms whether cancer is present and also what type of vulval cancer it is. This is often able to be performed under local anaesthetic as will be small but sometimes it may be easier to perform this under an anaesthetic especially if a more thorough examination is required.

Scans that assesses if the cancer has spread include **MRI and CT** of the chest, abdomen and pelvis and also occasionally a scan called a **PET CT** which is very accurate at assessing the lymph nodes of the body (the drainage system).

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You case will be referred to the gynaecological oncology service and you will attend an appointment at our clinic. Here you will be reviewed by a gynaecological oncology surgeon and will be introduced to our clinical specialist nurse team.

We often ask permission to examine you again, discuss your results and also what the expected treatment plan will be. Every cancer case is discussed at our MDT (multi-disciplinary meeting) with all the members of our extended team. Therefore, each decision for treatment is made by an expert group and is personalised for each patient. Often, we may see you at clinic on the morning before this meeting. You will be given a treatment plan at that visit, however in 1 in 10 cases this plan will be changed at the MDT meeting and we may contact you after this to confirm the treatment plan.

Types of Vulval Cancer

There are different types of vulval cancer:

- **Squamous Cell Carcinoma**
  The most common type of vulval cancer with about 90% being this type and usually forms slowly over many years.
  Most start on the outer lips of the vulva (labia majora). But it can also start in the inner lips (labia minora), clitoris or perineum. Before it develops, there might be precancerous changes in the cells of the vulva. These can be there for several years.

- **Vulval Melanoma**
  The second most common type of vulval cancer with around 8% of cases. It is most often found in women older than 50. Melanomas develop from the skin cells that give the skin its colour by producing pigment.

- **Sarcoma**
  Rare with <1% of cases. Sarcomas are cancers that start in tissue such as muscle or fat under the skin. These cancers tend to grow quite quickly. There are several different types of sarcomas that can affect the vulva. They include: leiomyosarcomas, malignant fibrous histiosarcoma, rhabdomyosarcomas, angiosarcomas and neurofibrosarcomas.
  These are different types of sarcoma which develop from different types of body tissues. Leiomyosarcomas and rhabdomyosarcomas are both muscle tumours. Angiosarcomas begin in the cells of the blood vessels (veins, arteries or capillaries).

- **Paget's Disease**
  Rare type of skin cancer affecting the surface of the skin of the vulva and is slow growing. It is generally seen in women over the age of 50. Paget's disease of the vulva causes itching, and it may appear red and flaky. You usually have a biopsy to confirm the diagnosis. Treatment

includes: surgery with a wide local excision, imiquimod cream, radiotherapy or photodynamic therapy (PDT).
This is also known as extramammary Paget's disease as it is similar to Paget's disease of the breast

- **Adenocarcinoma**
  Very rare type which develops from glands in the vulval skin. An example of this is Bartholin's gland cancer. The Bartholin's glands are 2 small mucous producing glands at the opening of the vagina. They make a fluid which acts as a lubricant during sexual intercourse. This type of vulval cancer is extremely rare and usually treated with surgery.

- **Basal Cell Carcinoma**
  A small number of vulval cancers are basal cell carcinomas. This type of cancer develops from the deepest layer of skin cells called the basal cells.

- **Verrucous**
  This type of cancer is very rare. It looks like a large wart and is a slow growing type of squamous cell carcinoma.

Staging of Vulval Cancer

For Vulval Cancer we apply the International Federation of Gynaecology and Obstetrics (FIGO) staging system. There are 4 stages, numbered 1 to 4.

- **Stage 1** – the cancer is confined to the vulva
- **Stage 2** – the cancer has spread to other nearby parts of the body, such as the lower vagina, anus or lower urethra (the tube urine passes through out of the body), but the lymph nodes are unaffected
- **Stage 3** – the cancer has spread into nearby lymph nodes
- **Stage 4** – the cancer has spread to other parts of the body, including more distant lymph nodes. This may be to the bladder or bowel nearby (Stage 4A) or to distant organs such as the lungs (Stage 4B)

The Staging of the cancer, the type of cancer and your general health will all be considered when deciding the best treatment pathway.

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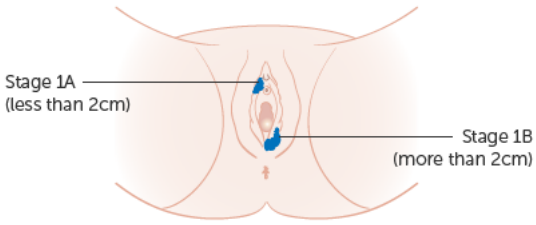
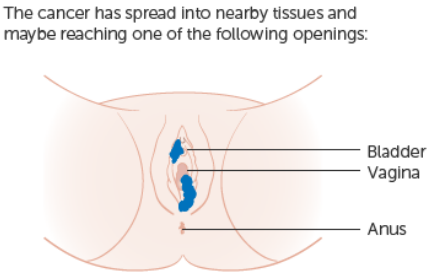
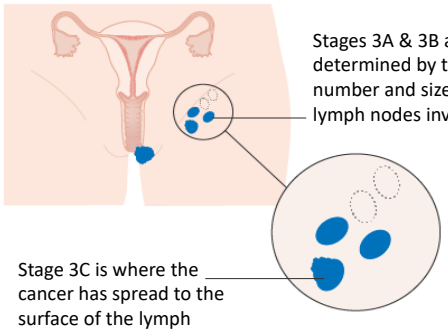


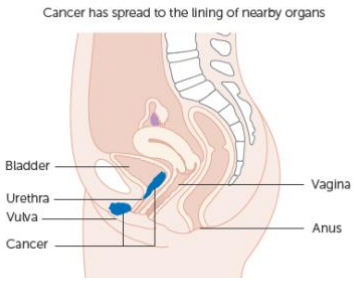
Stage 1



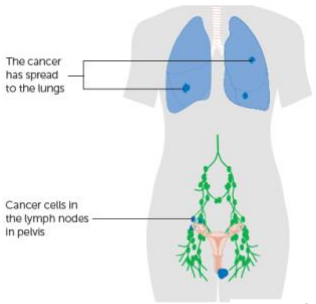
Stage 2



Stage 3



Stage 4A



Stage 4B

Adapted from Cancer Research UK

Treatment

The treatment for vulval cancer is usually surgery but can also be or include radiotherapy and chemotherapy.

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- **Surgery**

The aim of surgery is to remove all of the cancer with a margin of tissue around this whilst still preserving as much vulval skin and tissue as possible. The extent of the surgery therefore depends on where the cancer is located, the size and type of cancer.

The anatomy of the vulva helps to explain this as was shown in the diagram in the beginning.

Surgery can be in the forms of one or more of the following:

- **Wide local excision:** removal of the tumour/cancer area and a small area of tissue around it
- **Hemi-Vulvectomy:** removal of one side of the vulva including the labia minor and majora
- **Radical Vulvectomy:** removal of the whole of the vulva
- **Removal of the clitoris:** sensitive are of tissue just above the urethra (opening to the bladder)
- **Removal of the perineal body:** the skin & tissue situated between the vagina and the back passage (anus)
- **Removal of lymph nodes in the groin**

If your tumour is located on the vulva only and is <4cm size you may be offered a **sentinel lymph node removal** rather than the removal of all of the lymph nodes in the groin.

This is a safe and effective method where we use a specialised radioactive marker and a dye to identify the first lymph node the cancer drains to (the sentinel lymph node).

Women benefit from this technique if appropriate as it reduces the risk of developing lymphoedema. However, if the lymph node is positive we may need to return to remove all of the lymph nodes.

If the surgeon has to remove a large area of skin and tissue it is sometimes necessary to use plastic surgery techniques to reconstruct the area. We work closely with the plastic surgery team for this reason. There are several techniques possible using nearby areas of tissue and skin and relocating them to the area of surgery which give cosmetic and functional benefits to patients undergoing major surgery.

Major surgery in the form of a pelvic exenteration can be considered where the cancer is involving the urethra and/or rectum but there is no distant evidence of the disease. It can also be considered in recurrent disease. This is where the bladder, vagina, vulva and rectum can all be required to be removed. A new bladder in the form of an ileal conduit and an end colostomy may be required. This is major surgery so each case is carefully considered and every patient and their family will be counselled fully.

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- **Radiotherapy**

Radiotherapy uses radiation to kill cancer cells. It is usually given from outside the body using a machine that directs high-energy X-rays at the cancer. This is called external beam radiotherapy.

There are several reasons that radiotherapy may be used in vulval cancer:

- If your lymph nodes show signs of cancer, you are likely to have radiotherapy after your operation.
- If a patient is not well enough for an operation, radiotherapy may be offered instead of surgery.
- If it is not surgically possible to achieve a clear margin of normal tissue.
- Can also be used to help patients whose cancer cannot be cured. It can help relieve symptoms and prolong a good quality of life.

While it kills cancerous cells, radiotherapy can also affect healthy tissue and has a number of side effects, including:

- sore, red skin (like sunburn)
- pain while passing urine
- loss of taste & loss of appetite
- tiredness & nausea

- **Chemotherapy**

Chemotherapy can have several roles in the treatment of vulval cancer:

- It may be used to shrink your cancer before you have surgery. This is called neo adjuvant chemotherapy.
- May be recommended after surgery where the lymph nodes are found to be positive.
- If surgery is not appropriate another treatment pathway is chemotherapy with radiotherapy (chemoradiotherapy). The chemotherapy helps the radiotherapy to work. The drugs you are most likely to have are usually one or more of: cisplatin, 5FU, carboplatin, paciltaxel.
- To try to control vulval cancer that has spread. Carboplatin and paclitaxel are the drugs used most often.

It is difficult to say exactly how often you may have chemotherapy as there are different timetables for different chemotherapy drugs. The medical oncology team will explain the treatment plan to you in detail once it is decided. But you are most likely to have treatment for one to 5 days, with a break of 2 or 3 weeks between each treatment.

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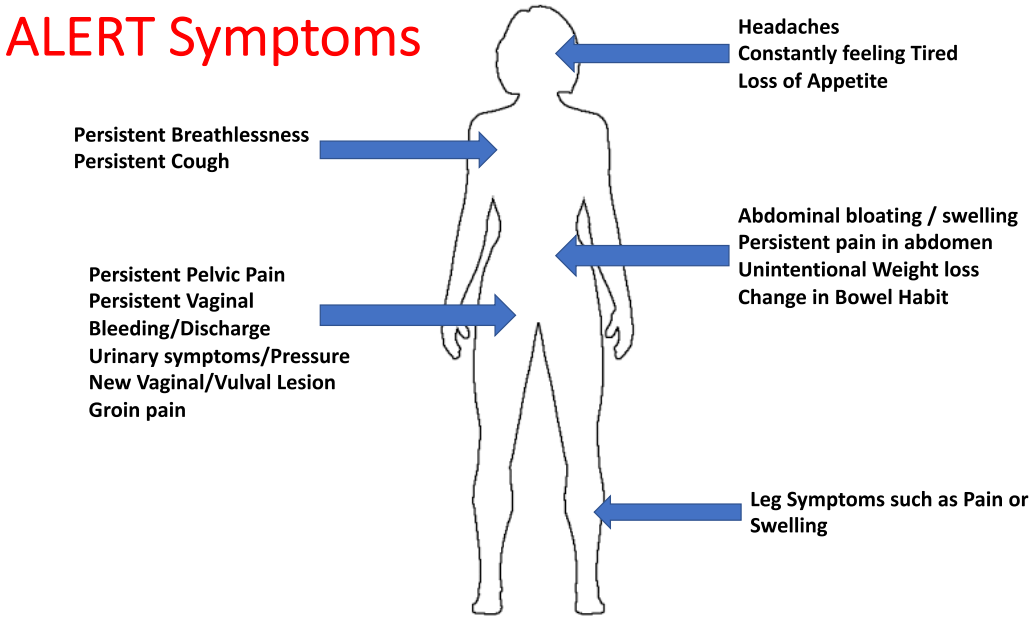
Research

Our team specialises in vulval cancer and we are involved in many research projects and clinical trials to ensure every effort is made to provide the best possible care and to try and develop new treatments. We will ask you to partake in our research program and this will be explained in detail along with asking for your consent.

Living With & After Vulval Cancer

Survival depends on many different factors. There are general statistics based on large groups of patients, but, they can't tell you what will happen in your individual case. Therefore, being aware of the symptoms of possible recurrence of your cancer (also known as ALERT symptoms) is important along with attending for follow up appointments. These appointments can be a combination of physical and virtual appointments and this will be explained to you.



The treatment of vulval cancer itself may result in symptoms and longer-term complications. We do everything possible to try and minimise the risk of these complications but also have developed services to identify and treat these as early as possible. Of importance is referral to our lymphoedema service and we do this at diagnosis. We aim to prevent and manage lymphoedema at an early stage where possible. Surgery and radiotherapy for vulval cancer can also affect aspects of your life including early menopause and your sex life. We aim to support your fully in these areas and can provide information and support during your cancer journey.

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Follow up after your treatment is completed is therefore extremely important both for monitoring for any sign the cancer may have returned and also to help support patients with any side effects of treatment.

Throughout your journey with our team we will support you fully in any decision you make. You can get emotional and practical support through our team, local hospice and GP practice. You can also get help and information from charities and support groups.

Further information and support for vulval cancer can be found at:

- Irish Cancer Society www.cancer.ie
- MacMillan Cancer Support www.macmillan.org.uk
- Arc Cancer Support www.arccancersupport.ie
- Cancer Research UK www.cancerresearchuk.org