

Vulvo-vaginal Mucosal Melanoma

Information for patients
and carers



A close-up photograph of a doctor in a white lab coat, wearing glasses and a gold ring, writing on a clipboard with a silver pen. The background is blurred, showing a clinical setting.

Introduction

The information in this leaflet relates specifically to melanomas of the vaginal and vulval mucous membranes. The leaflet summarises a guideline [melanomafocus.org/for-professionals/rare-melanoma-guidelines-and-consultations/mucosal-melanoma-guidelines/](https://www.melanomafocus.org/for-professionals/rare-melanoma-guidelines-and-consultations/mucosal-melanoma-guidelines/) developed by experts in the field to advise cancer specialists who treat patients with this condition and is based upon the best evidence available. Skin cancers can also develop in the same areas of the body, in the skin rather than in the mucous membranes. These are known as cutaneous melanomas and are not covered by this guideline. If you have been diagnosed with a skin (cutaneous) melanoma please refer to the NICE guideline [nice.org.uk/guidance/ng14](https://www.nice.org.uk/guidance/ng14) and the other organisations listed at the end of this leaflet.

The number of medical terms has been kept to a minimum in this leaflet. If you come across a term you don't understand, please see the definitions and abbreviations section at the end of this document or ask your consultant or nurse.

Vulvo-vaginal Mucosal Melanoma

What is it?

Melanoma develops if there is uncontrolled growth of melanocytes, the cells responsible for pigmenting (darkening) the skin. Mucosal melanoma is a kind of melanoma that occurs in mucous membranes. These are the moist surfaces that line cavities within the body. Mucosal melanomas can occur in the eyes, mouth, nasal passages and larynx. They can also develop in and around the anus, vagina, vulva or penis. Melanomas of the vulva that started on the outside of the vulva, which is skin, are cutaneous (skin) melanomas and are not covered by this leaflet. Melanomas of the vulva that started on the inside (i.e. in the mucous membranes) or in the vagina are mucosal melanomas which this leaflet covers.

Mucosal melanomas are very rare, in the Surveillance Epidemiology and End Results (SEER) database which registered melanomas over 22 years, only about 1 melanoma in 100 was a mucosal melanoma. The majority of melanomas grow within the skin. Mucosal melanomas can be more complicated to treat than skin melanomas as they are often diagnosed later. This is because they are in less visible places and because they are often not pigmented (darker), which makes them even more difficult to spot.

These melanomas are different in several ways from skin melanomas. For example, while the risk of getting skin melanoma is increased by too much exposure to the sun, there appears to be no link between sun and mucosal melanomas. No specific

causes or links with lifestyle have been found for mucosal melanoma and as far as we know there is nothing you can do to prevent it. It also appears that mucosal melanoma tumours are different from skin melanoma tumours. This means that some treatments for skin melanomas may not be as effective for mucosal melanoma and the outlook, or *prognosis* (or natural history), for mucosal melanomas is not as good as for skin melanomas. The prognosis will be different depending on whether the tumour can be fully removed surgically and whether it has spread. If you would like more information about your individual situation, you should discuss this with your clinical team.

Questions you may wish to ask about your visits

How often will I have appointments at the cancer centre?

What sort of support will I need at these appointments?

How long will I be at the centre each time?

What further signs and symptoms should I look out for?

What symptoms should I report urgently?

How and who should I contact if I need to do so urgently?

What are the symptoms?

The symptoms for vulval/vaginal mucosal melanomas are similar to those for other cancers of the vulva/vagina and other conditions that can affect the vulva and vagina. You may have a lump, a sore area and/or bleeding. You may have trouble urinating (passing water). As with skin melanomas, there may be an area which is pigmented (darker) but this is not always the case. If you have any of these symptoms and your GP cannot find another cause, you should be referred quickly (normally within about two weeks) to a specialist with experience of this condition, usually a Gynaecology-Oncology consultant surgeon, for tests and further evaluation. It is most important that the team you see regularly treats patients with this condition. As it is so rare, you may need to travel to a specialist centre rather than attend your local hospital.

Going to the hospital

Tests and examination

At the hospital the consultant or a senior doctor on the team will examine you. This may include an *examination under anaesthesia* (which means you will be given an anaesthetic to make you unconscious). If this is the case the consultant will use a cystoscope to look closely at any lumps or other abnormalities. The procedure is called a cystoscopy. You may also have an MRI scan of your pelvis. If the consultant identifies a suspicious lump, you will be offered an excision, incision or a punch biopsy depending on your symptoms and where the lesion is located. The consultant may also carry out a core biopsy and/or a fine-needle aspiration to send off to be examined under a microscope. The guideline does not recommend that sentinel node biopsy is done routinely but it may be



performed in some cases to decide on future treatments or entering into clinical trials.

If the consultant makes a diagnosis of vulval or vaginal mucosal melanoma, you would normally have a CT scan of your body to check whether the cancer has spread. There are several specific genes which have been identified that may be involved in some but not all mucosal melanoma tumours. The pathologist should test the tissue sample from the biopsy to see if one of these identified genes is involved in your cancer as this will help decide the best treatment for you now and in the future.

Diagnosis, staging and prognosis

The cancer team can estimate the possible outcomes – your prognosis – based on the results of your tests and medical examination. If you are diagnosed with a mucosal melanoma, your cancer will be evaluated according to a process known as *staging*. The stage indicates whether your cancer has spread in your body and, if so, where. This staging process will help both you and the cancer team make decisions about your future treatment.

For the staging, the American Joint Committee on Cancer **TNM** staging system is used. In addition to other information the team should record in particular:

- Location of the primary (original) tumour
- Primary **T**umour size and extent of tumours
- Lymph **N**ode involvement (whether or not the cancer has spread to the nearby lymph nodes (also called glands)
- Presence or absence of distant **M**etastasis (whether or not the cancer has spread to distant areas of the body).

The cancer team will make a prediction of the outcome of your cancer – your prognosis – based on the results of your tests and

examination and on what happened to others who have had similar cancers. If you and/or a relative or carer want to have a full discussion about your prognosis, make this clear to someone from the cancer team. It will mean that your appointments will be more informative and will enable you to be more involved in planning your treatment and other aspects of your life. On the other hand you may wish to take it 'one step at a time'. Whatever your decision, you will be fully supported by the cancer team.

Questions you may wish to ask about your diagnosis and prognosis

What are you looking for in the biopsies?

What is the difference between a CT scan, MRI scan and a PET scan – what do each show?

What stage is my cancer at and what is my prognosis?

Are any gene mutations involved in my cancer?

What is the best treatment available to me?

What sort of health problems might I have? And what are the plans I need to put in place to be prepared?

What supportive services are available to me and may I have their contact details for future reference?

Can you recommend leaflets or websites with information on my condition?

Are there any clinical trials available that I might be suitable for?

What to expect from your care

A specific specialist cancer team will look after you during your treatment. As this is a rare cancer which has similarities to both skin and gynaecological cancers, there will be cooperation between the skin and gynaecology/oncology experts. The guideline advises teams on specific ways in which they should work together.

When discussing your diagnosis and treatment you should be told:

- The name and contact details of your consultant and of your Cancer Nurse Specialist.
- Who your key worker is (this person is your first point of contact should you have questions or problems and is usually the Cancer Nurse Specialist; you need to make sure you are given the contact details of the person to contact if your keyworker isn't available).
- How to make an appointment with the consultant quickly should you run into problems.
- About referral to support services (for example, palliative care services) should you need them at any point in your treatment.

Treatment

If the cancer hasn't spread beyond the primary site the first line of treatment for a vulvo-vaginal mucosal melanoma is surgery. This should be done by a surgeon who is very experienced in complex vulvo-vaginal surgery and is based in a centre where there is support from an experienced skin cancer team.

In planning the operation, the consultant should discuss your current health and how the surgery may affect your quality of life and/or urinary continence (bladder control). If it is likely to make a difference, the consultant should discuss other treatment options with you to agree what is best for you. The aim of surgery is to remove a little of the surrounding healthy tissue to make sure that all of the cancer is removed. This is called having 'clear margins'.

If the cancer hasn't spread and the margins are clear, this may be the end of your treatment (see After Treatment, page 6) or, depending on the stage and tumour, you may be offered adjuvant therapy (see page 5).

If, after surgery, the margins of tissue aren't clear of cancer, the surgery may be repeated. If another operation isn't an option, the possibility of radiotherapy should be discussed with you.

If the tumour is large, or the cancer has spread, then a more complex operation may be performed, which may remove the affected lymph nodes. Once again, the consultant should discuss the risk that your quality of life or continence will be affected by the operation and whether surgery is the best option for you. You also may be offered further tests to help decide. If the cancer has spread (metastasised), you may be offered chemotherapy and/or radiotherapy instead of surgery (see page 7).

Questions you may wish to ask about your treatment

Why (or why not) is surgery the best option for me?

How should I expect to feel after my surgery?

What will the recovery time be?

Will I be able to carry on with life as normal?

What will be the effects of my surgery?

Will my sex life be affected?

What are the risks that I will experience incontinence?

What symptoms should I look out for, indicating whether the surgery has worked or not?

What support will I need after surgery?

Will I need adjuvant therapy? If so, what is the reason for this and what will it involve?

Adjuvant (Extra) therapy

Depending on the results of the surgery and the cancer stage, you may be offered adjuvant therapy. The aim of adjuvant therapy is to reduce the chances that the cancer will come back after it has been removed entirely by surgery (a 'belt and braces' approach). Recently there have been great advances in the treatment of skin melanomas with immunotherapy, which stimulates your own body to fight the cancer. By understanding more about the molecular make-up of different tumours, treatments such as immunotherapy and targeted therapies may can target unique aspects of the tumour. However, because mucosal melanomas are so rare, scientists are still uncertain whether these treatments work in the same way as with skin melanomas.

You may be asked if you would like to enter a clinical trial to help doctors answer this question. Or, as doctors learn more, you may be offered immunotherapy, particularly if the tumour contains one of the genes that can make such a treatment effective. (However keep in mind that some of these treatments are not currently funded by the NHS and you may have to pay for them yourself). Similarly, you may be offered adjuvant radiotherapy or be invited to enter a trial to learn more about the benefits of this type of treatment for people with mucosal melanoma.

After treatment

After your initial treatment, you should be offered regular follow-up appointments to check that no cancer has returned, either to the original site or elsewhere in your body.

You should be given information how to make an appointment quickly at any time after your treatment if you are concerned.

The schedule recommended in the guideline is for an appointment every three months during the first three years after your treatment. The procedure will be similar to the one you had when you first attended the hospital and should include a physical examination possibly with another examination under anaesthetic and cystoscopy to check everything is still clear at the original site.

You should be offered a CT scan two or three months after surgery to establish a baseline and then every six months during the first three years in order to check that the cancer hasn't spread to another site. Very occasionally the cancer spreads to the brain, the consultant should discuss the risks and benefits of a brain scan with you.

If you would like to read more about the advantages and disadvantages of having scans refer to the NICE option grid set out in skin cancer guideline NG14: [nice.org.uk/guidance/ng14/resources/followup-with-regular-ct-scans-yes-or-no-pdf-250598416](https://www.nice.org.uk/guidance/ng14/resources/followup-with-regular-ct-scans-yes-or-no-pdf-250598416).

The risk of the cancer returning reduces with time. Therefore from three to five years after treatment you should be offered an appointment every six months and a CT scan once a year. From six to ten years after treatment you should be offered an appointment once a year or alternatively rapid access to a clinic if you are concerned. After ten years you should not need to have appointments but can come back at any time if you are concerned.

If your cancer recurs or spreads (metastasis)

Surgery is again the first line of treatment if the tumour recurs. If the cancer has spread, or if surgery is not an option, a member of the cancer team should discuss the different treatments available. New treatments are being developed and your consultant should tell you about their success rates, as well as the risks and benefits in your particular case.

You may be offered immunotherapy treatment to slow or stop the growth of the tumour. Some vulvo-vaginal melanomas have unique aspects that may be targeted with specific drugs. In this case the cancer team should discuss the options with you and the availability of any clinical trials you could enter. There is little evidence that the routine use of chemotherapy or bio-chemotherapy is effective in the treatment of metastatic mucosal melanoma and it not currently recommended by the guideline. You can find the clinical trials which are currently running on the Melanoma Focus TrialFinder melanomafocus.org/trialfinder/.

Depending on numerous factors relating to your tumour, you may be offered palliative radiotherapy. Your consultant will discuss these types of treatment with you.

Following treatment for a recurrence or metastasis, you should be offered regular appointments at two- to three-monthly intervals, depending on the kind of treatment you have had. These should include CT scans and an MRI or CT scan of the brain. If you remain well and the disease does not progress, after two to three years, appointments can be every six months. After five years, the interval can be extended to yearly.

If the disease progresses, there are other options to make you more comfortable, which are used for skin melanomas. General guidance is available in NICE guidance CSG4: [nice.org.uk/guidance/csg4](https://www.nice.org.uk/guidance/csg4).

Questions you may wish to ask about your treatment for metastatic disease

Is there a chemotherapy, targeted therapy or radiotherapy available in my situation?

If so, what is the likelihood that it will reduce my symptoms and/or extend my life?

Is it available on the NHS?

Are there side-effects with this treatment and, if so, what are they?

What clinical trials are available?

What is the aim of the clinical trial in question?

What support will I need while on this treatment?

How will I be able to tell whether it's working or not?

If it is not working, how soon can I move on to something else?

Will this treatment prolong my life?

Will I have a reasonable quality of life while on this treatment?

Sources of information and support

Since mucosal melanoma is so rare, there are no UK websites dedicated to information about this disease.

Melanoma Focus (melanomafocus.org) is a charity that commissions and funds melanoma research, while providing support and information for patients, carers and healthcare professionals. It funded this leaflet and the associated guideline for health professionals, on which this leaflet is based. See melanomafocus.org/for-professionals/rare-melanoma-guidelines-and-consultations/mucosal-melanoma-guidelines/, where there are links to a Resources page giving access to the various guideline documents.

Macmillan (macmillan.org.uk) provides support for people who have cancer.

Although not specific to mucosal melanoma, **Cancer Research UK** has a great deal of information about vaginal and vulval cancer, including treatments and living with cancer, which is relevant to vulvo-vaginal mucosal melanoma. cancerresearchuk.org/about-cancer/vaginal-cancer and cancerresearchuk.org/about-cancer/vulval-cancer.

NHS Choices (nhs.uk) has information on vulval and vaginal cancer treatments and other aspects of care.

The **American Joint Committee on Cancer** (AJCC), which publishes the staging manual used by doctors, has a patient leaflet explaining staging here: facs.org/quality-programs/cancer-programs/american-joint-committee-on-cancer/cancer-staging-systems/.

Definitions and abbreviations

Adjuvant therapy

Adjuvant therapy is treatment in addition to that used to treat the initial cancer. The aim is to reduce the possibility that the cancer will come back after it has been removed entirely by surgery.

BIOPSY TECHNIQUES

A biopsy is when a piece of the abnormal tissue is removed so the cells can be examined under a microscope to determine whether it is cancer and if so what type.

There are many ways of taking a biopsy. The ones mentioned in this leaflet are listed below.

See NHS Choices, from which this information was taken, for more detail: nhs.uk/conditions/biopsy.

Core biopsy

For a core biopsy, the area is numbed with a local anaesthetic, a wide hollow needle is inserted and a sample of the cells taken.

Excision biopsy

An excision biopsy aims to remove the lesion or lump entirely. It will be examined to determine whether more treatment is needed.

Fine needle aspiration

A fine-needle aspiration (FNA) biopsy is similar to a core biopsy but isn't as deep and may not need a local anaesthetic. When the needle is in position, it will cut out a small sample of tissue.

Punch biopsy

For a punch biopsy, the area is usually numbed with a local anaesthetic, and a small hole is made in the skin and samples of the top layers of tissue are taken.

Sentinel node biopsy

Sentinel node biopsy is a test to determine whether there are cancer cells in the lymph nodes around your groin. The consultant will carry out a small operation to remove some tissue to be tested. Animated videos to explain sentinel lymph node biopsy can be found on the Melanoma Focus website [melanomafocus.org/patient-](http://melanomafocus.org/patient-guide/procedures-and-surgery/sentinel-lymph-node-biopsy/)

[guide/procedures-and-surgery/sentinel-lymph-node-biopsy/](http://melanomafocus.org/patient-guide/procedures-and-surgery/sentinel-lymph-node-biopsy/).

Cystoscopy

A procedure where a cystoscope, which is a fine tube with a camera, is inserted into the urethra (the passage that carries urine out of your bladder) to enable the doctor to look more closely.

More information is available here: cancerresearchuk.org/about-cancer/vulval-cancer/getting-diagnosed/tests-stage-cystoscopy.

Immunotherapy

Also called biologic therapy or bio-chemotherapy, stimulates the patient's own immune system to fight cancer. For more information and a video explaining immunotherapies see the Melanoma Focus website: melanomafocus.org/patient-guide/melanoma-treatment/treatment-for-advanced-melanoma/immunotherapy-treatment/.

Incontinence

Incontinence is the term used when a person has difficulty controlling their bladder (urinary incontinence) or bowels (faecal incontinence).

Lesion

An area of tissue that isn't normal. This term can be used when it is uncertain whether the area is cancer (malignant) or not (benign).

Lymph Nodes

Lymph nodes are often the first place the cancer spreads.

Fuller explanation of lymph nodes and cancer can be found here: cancer.org/cancer/cancer-basics/lymph-nodes-and-cancer.

Metastasis/Metastatic Cancer

Metastasis is when cancer cells break away from the original (primary) site, travel through the blood or lymph system, and form a new tumour in other organs or tissues of the body. The new, metastatic tumour is the same type of cancer as the primary tumour. For example, if mucosal melanoma spreads to the liver, the cancer cells in the liver are mucosal melanoma cancer cells, not liver cancer cells.

Palliative care support services

More information on what forms of home care are available can be found at: nhs.uk/conditions/social-care-and-support/home-care and cancerresearchuk.org/about-cancer/melanoma/advanced-melanoma/support-home-for-you-your-family.

Pigmentation

Pigmentation is colouring of the skin which can be different shades of brown. Cancerous cells may start producing more pigment, resulting in dark patches. However, some mucosal melanomas are not pigmented.

SCANS

For more information on the tests mentioned in this leaflet see the Cancer Research UK website link: www.cancerresearchuk.org/about-cancer/cancer-in-general/tests.

CT Scan

Computerised Tomography (CT) takes x-rays from different angles and the computer then puts them together as a three-dimensional picture. For more information see the Cancer Research UK website link above and nhs.uk/conditions/CT-scan.

MRI/MR

Magnetic Resonance Imaging (MRI or MR) takes pictures of the body using magnets and radio waves. These show up soft tissues such as the bowel, liver, lungs etc. better than CT scans. The scan can take up to 1½ hours. For more information see the Cancer Research UK website link above and nhs.uk/conditions/mri-scan.

PET-CT

This combines a CT Scan (see above) with a Positron Emission Tomography (PET) scan. A mildly radioactive substance is injected, which shows up parts of the body where cells are more active. That is, where a cancer might be growing. For more information see the Cancer Research UK website link above and nhs.uk/conditions/pet-scan.

Staging

Staging is an estimate of how much cancer there is and where it is located. Staging is used to plan treatment and future options. Further explanation can be found here melanomafocus.org/about-melanoma/types-of-melanoma/mucosal-melanoma/vaginal-and-vulval-mucosal-melanoma/.

Wide local excision

In a wide local excision (WLE), the surgeon cuts out the tumour (cancerous tissue) along with a small margin of around 1 millimetre of healthy tissue to increase the likelihood that all of the tumour has been removed.

ABBREVIATIONS

- AUG** Ano-uro-genital
- CNS** Clinical Nurse Specialist
- CT** Computed tomography
- MRI** Magnetic resonance imaging
- PET** Positron emission tomography
- SNB** Sentinel node biopsy
- WLE** Wide Local excision

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