

## **Uveal Melanoma**

Information for patients  
and carers



## Introduction

The information in this leaflet relates specifically to melanomas of the eye. The leaflet summarises a guideline [melanomafocus.org/for-professionals/rare-melanoma-guidelines-and-consultations/uveal-melanoma-guidelines/](https://www.melanomafocus.org/for-professionals/rare-melanoma-guidelines-and-consultations/uveal-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 (cutaneous) melanomas are not covered by this guideline. If you have been diagnosed with a skin melanoma, please refer to the NICE guideline [nice.org.uk/guidance/conditions-and-diseases/cancer/skin-cancer](https://www.nice.org.uk/guidance/conditions-and-diseases/cancer/skin-cancer). Melanoma Focus has produced extensive information on melanoma skin cancer.

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.

Melanoma Focus is most grateful to OcuMel UK for their donation towards the updated guidelines and corresponding patient information.

## Uveal Melanoma

### What is it?

Uveal melanoma (UM) is a cancer of the eye involving a tumour(s) of the iris, ciliary body or the choroid (known as the uvea). The tumours arise from the pigment cells in the eye called melanocytes.

These melanomas are different in many ways from skin melanomas. For example, the link between too much sun exposure and skin melanoma is strong, while there is no connection between sun exposure and most kinds of UM. Although UM is very rare with about 600-700 new cases in the year in the UK, there are other extremely rare melanomas of the eye and the umbrella term is ocular melanoma. Compare these numbers to melanoma skin cancer with about 17,000 new cases a year in the UK.

There are certain factors which make getting a UM more likely, these include being older, having lighter colour eyes and/or lighter skin. Very occasionally, there is a faulty gene inherited which may be linked to several types of cancer in the family. Because of these differences some treatments for skin melanomas may not be as effective for UM and the outlook, or prognosis (or natural history), for UMs may not be as good as for skin melanomas. The prognosis will be different depending on a number of factors which are discussed below. If you would like more information about your individual situation, you should discuss this with your clinical team.

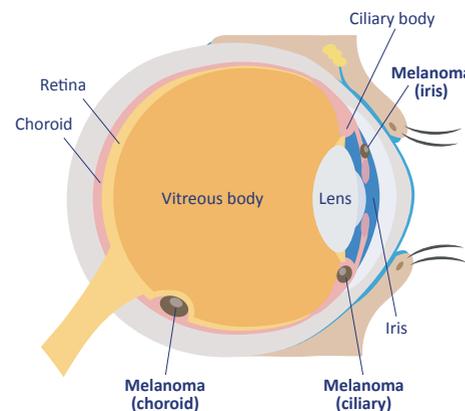
### What to expect from your care

A specific specialist cancer team will look after you during your treatment. The guideline [melanomafocus.org/for-professionals/rare-melanoma-guidelines-and-consultations/uveal-melanoma-guidelines/](https://www.melanomafocus.org/for-professionals/rare-melanoma-guidelines-and-consultations/uveal-melanoma-guidelines/) 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 (CNS).
- Who your key worker is (this person is your first point of contact should you have questions or problems and is usually the CNS). You should have the contact details including telephone and email address of your keyworker and who to contact should your keyworker not be available.

### Eye Cancer: Melanoma of the Eye



- How to make an appointment with the consultant quickly should you run into problems.
- About referral to support services (for example, palliative care services) and to other resources such as, for example, Melanoma Focus, OcuMel UK, CRUK, MacMillan, Maggie's should you need them at any point in your treatment. For more information on these organisations see the list at the end of this leaflet.

## What are the symptoms?

Unlike skin melanomas, you generally won't see a UM unless it is in the iris and there won't be symptoms at first. It may be noticed by the optometrist during a routine eye test. However, many are not discovered until there is pain or discomfort which triggers a trip to the GP's or A&E.

## What to expect next

### Tests and examination

If a UM is suspected, you will be referred to an ophthalmologist with experience of the condition. As it is rare, you may need to travel to a specialist centre, rather than attend your local hospital. UM is treated in three specialised centres in England: Liverpool, London and Sheffield and in Scotland in Edinburgh. These are called "supra-regional" centres. UM that has spread (called metastatic UM) requires the input of many different highly-specialised healthcare professionals and may be outside of the supra-regional centres.

At the hospital, the consultant\* will examine your eye with instruments to see the whole of the eye. A sample of the fluid in your eye may be taken.

\* This may be the consultant or another senior doctor on the 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 whom should I contact if I need to do so urgently?

Is there psychological support available should I need it?



## Diagnosis, staging and prognosis

The cancer team will make a diagnosis based on the results of your tests and medical examination. If you are diagnosed with a tumour which is UM, the consultant will be able to make a prognosis (estimate of the outcome) from the following factors:

- Your age
- Your sex
- The location of the tumour
- The size of the tumour
- Whether the ciliary body has been involved
- Whether the tumour has started to grow outside of the eye

As part of this process, you may also be offered a prognostic biopsy, where a small bit of the tumour in your eye is removed for analysis. There are both pros and cons to this procedure and the consultant should discuss these with you before a decision is made. If there is a biopsy, or the tumour has been removed by surgery, additional information can be provided by looking at the tumour through a microscope including:

- The type of cell
- The number of cells dividing
- The patterns seen within the tissue

If possible, a tissue sample should be analysed for its genetic make-up as new targeted therapies for tumours involving particular genes are continually being developed. This genetic information will inform current and future treatments.

The information will be evaluated in a process known as *staging*. The stage indicates if the cancer has spread and, if so, where. For the staging, the American Joint Committee on Cancer TNM staging system is used ([cancerresearchuk.org/about-cancer/eye-cancer/getting-diagnosed/tests-diagnose](http://cancerresearchuk.org/about-cancer/eye-cancer/getting-diagnosed/tests-diagnose)).

## Questions you may wish to ask about your diagnosis and prognosis

What are you looking for in the biopsies?

What are the risks of a prognostic biopsy?

What are the benefits of having a prognostic biopsy?

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

Are any gene mutations involved in my cancer and if so, is there a targeted treatment?

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?

The cancer team will then make a prediction of the outcome of the 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 medical 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.

## Treatment

There are a number of different methods to treating the primary tumour in the eye. First-line treatments include:

### Radiation therapy:

Plaque brachytherapy  
Proton beam radiotherapy  
Stereotactic radiosurgery

### Phototherapy:

Transpupillary thermotherapy  
Photodynamic therapy

### Surgery:

Exoresection  
Endoresection  
Enucleation

These are defined in the Glossary at the end of this document.

Generally, no one first-line treatment has been shown to be better than another. On the one hand, you may also prefer surgery where treatment is over quickly. On the other, you may prefer something less intrusive. But, depending on where the tumour is located and its size, the consultant may recommend one treatment over another. The options should be discussed fully with you and why a

particular treatment is being recommended.

If the UM hasn't spread, this may be the end of your treatment (see After Treatment). With many types of cancer adjuvant (extra) chemo or radiation therapy is offered following the primary treatment. However, there is, as yet, no proven benefit of adjuvant therapy for UM. Someone from the cancer team will discuss the options with you and may ask if you would like to enter a clinical trial to help doctors answer this question.

## Some questions you may wish to ask about your treatment

What size is my tumour?

What are my treatment options?

Are there alternative treatments offered at other centres?

Which treatment do you think is best for me and why?

How will my vision be affected?

How often will I have treatment?

How should I expect to feel after my treatment?

What will the recovery time be?

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

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

## After treatment

After your treatment, your consultant should have a discussion with you about the most appropriate follow-up schedule for you. The chances of your cancer spreading and the most likely places for it to appear in the body should be discussed with you.

Together you should decide what the goal of your future follow-ups will be and then develop an individual plan that takes into account the risks, benefits and consequences of your decision. If you are considered high-risk you should be offered a review every 6 months. As the UM spreads more frequently to the liver than other places, you should be offered scans of the liver at your follow-up visits. You should also have access to a nurse specialist for support.

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

## If the uveal melanoma recurs or spreads (metastasis)

If there is a suspicion that your cancer has spread, you should be looked after by a team of specialists which may be located away from your eye centre. The team should consist of the appropriate specialists and may include a clinical or medical oncologist, an interventional radiologist, a histopathologist, a liver surgeon and a clinical nurse specialist. The team should be experienced in treating UM and have direct links to the three eye specialist centres. They should have access to all the treatments and trials available nationally. You can find the clinical trials which are currently running on the Melanoma Focus TrialFinder [melanomafocus.org/trialfinder/](http://melanomafocus.org/trialfinder/) The recommendation that metastatic uveal patients should be treated by a specialist team is because of the rarity of the cancer and the benefits to patients of being seen by

those with experience. However, you are also able to choose to be treated nearer home if you prefer.

The specialist team should give you a variety of scans. Your chest, pelvis and abdomen should be scanned with contrast-enhanced CT or PET CT. Your liver should be scanned with contrast-enhanced MRI with diffusion weighting. In addition, if you have a pain in your bones, you should have a bone scan. You will not have a brain scan unless you have symptoms.

If the cancer has spread outside of the liver, you may be offered anticancer drugs. You should be told of any relevant clinical trials and these explained to you. If there are no clinical trials available or you do not wish to participate, you may be offered a systemic immune checkpoint inhibitor (ICI) if the tumour would respond to this. The risks and potential benefits of ICIs should be explained to you. A new drug called tebentafusp is showing promising results for people with UM tumours with a particular gene (HLA-A\*02:01).

If the cancer is only in the liver, surgery (resection) is the best treatment if you are a



suitable candidate. Before your operation you should be given a laparoscopy to check that surgery is possible. If you are not able to have a surgery, your specialists should consider other treatments directly to the liver.

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

## Questions you may wish to ask about your treatment and prognosis

What clinical trials are available?

What is the aim of the clinical trial in question?

What other treatments are available to me?

Is tebentafusp right for me and is it now available on the NHS or privately?

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

What are the side-effects with this treatment?

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

### **Melanoma Focus** ([melanomafocus.org](https://melanomafocus.org))

is a charity that provides support and information for patients, carers and healthcare professionals while commissioning and funding melanoma research. It funded this leaflet and the associated guideline for healthcare professionals with a much-appreciated donation from OcuMel UK.

**OcuMel UK** ([ocumeluk.org](https://ocumeluk.org)) is a registered charity representing those affected by ocular melanoma. You can contact their Helpline at [helpline@omuk.info](mailto:helpline@omuk.info).

**Macmillan** ([macmillan.org.uk](https://macmillan.org.uk)) provides support for people who have cancer.

Although not specific to UM, **Cancer Research UK** has a great deal of information about eye cancer, including treatments and living with cancer, which is relevant to UM: [cancerresearchuk.org/about-cancer/eye-cancer](https://cancerresearchuk.org/about-cancer/eye-cancer).

**Maggies** ([maggies.org](https://maggies.org)) offers support free to anyone with cancer and their families. They have centres alongside NHS hospitals as well as online support.

**NHS Choices** ([nhs.uk](https://www.nhs.uk)) has information on eye cancer treatments and other aspects of care: [nhs.uk/conditions/eye-cancer](https://www.nhs.uk/conditions/eye-cancer).

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/](https://www.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. It is used to boost the benefit of first-line treatment by attacking any possible remaining cancer cells circulating in the bloodstream or lymphatic system. The aim is to reduce the possibility that the cancer will come back after it has been removed entirely by surgery or travel to another part of the body.

### Biopsy techniques

A biopsy is when some 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 more details of biopsy on NHS Choices:

[nhs.uk/conditions/biopsy/](https://www.nhs.uk/conditions/biopsy/) and Cancer Research UK (CRUK): [cancerresearchuk.org/about-cancer/eye-cancer/getting-diagnosed/tests-diagnose/biops](https://www.cancerresearchuk.org/about-cancer/eye-cancer/getting-diagnosed/tests-diagnose/biops).

### Healthcare professionals you may encounter

#### Clinical nurse specialist

A nurse who has extra training in a particular speciality.

#### Clinical oncologist

A doctor with advanced training in cancer who is able to give radiation therapy.

#### Histopathologist

A pathologist who specialises in identifying tumours and their characteristics by looking at the cells under a microscope.

#### Interventional radiologist

A doctor who uses techniques which rely on the use of radiology to guide them, such as ultrasound-guided biopsy.

#### Medical oncologist

A doctor who gives chemotherapy but not radiation.

#### Ophthalmologist

A doctor specialising in diseases and injuries of the eye and performs surgery on the eye. The Ophthalmologist is likely to be the first consultant you see.

#### Pathologist

A doctor who uses laboratory tests to provide understanding about why a patient has fallen ill, what treatment could be useful and the success of treatment.

### Immunotherapy

Also called biologic therapy or bio-chemotherapy, stimulates the patient's own immune system to fight cancer.

#### Immune checkpoint inhibitors

Checkpoint inhibitors block the proteins in the tumour cells that stop the immune system from attacking the cancer cells. 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/](https://www.melanomafocus.org/patient-guide/melanoma-treatment/treatment-for-advanced-melanoma/immunotherapy-treatment/).

#### 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

Fuller explanation of lymph nodes and cancer is here: [cancer.org/cancer/cancer-basics/lymph-nodes-and-cancer](https://www.cancer.org/cancer/cancer-basics/lymph-nodes-and-cancer).

#### Melanomas of the Eye

For a diagram of the eye see: [nci-media.cancer.gov/pdq/media/images/543553.jpg](https://www.nci-media.cancer.gov/pdq/media/images/543553.jpg).

#### Choroidal melanoma

Uveal melanoma involving the back part of the eye (choroid). This is the most common type of uveal melanoma being about 90% of the total.

#### Ciliary body melanoma

UM involving the ciliary body which includes the ring of muscle on the inner surface of the front wall of the eye is responsible for providing the fluid that nourishes the lens and cornea of the eye.

#### Conjunctival melanoma

Melanoma that starts in the outer layer (conjunctiva) of the eye. It has more in common with skin melanoma and is not an uveal melanoma and not covered by this guideline.

#### Iris melanoma

Uveal melanoma involving the front, coloured part of the eye.

#### 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 uveal melanoma spreads

to the liver, the cancer cells in the liver are uveal 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](https://www.nhs.uk/conditions/social-care-and-support/home-care) and [cancerresearchuk.org/about-cancer/melanoma/advanced-melanoma/support-home-for-you-your-family](https://www.cancerresearchuk.org/about-cancer/melanoma/advanced-melanoma/support-home-for-you-your-family).

### 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](https://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. See the Cancer Research UK website link above and [nhs.uk/conditions/CT-scan/](https://www.nhs.uk/conditions/CT-scan/) for more information.

#### 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 1 ½ hours. See the Cancer Research UK website link above and [nhs.uk/conditions/mri-scan/](https://www.nhs.uk/conditions/mri-scan/) for more information.

#### 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. See the Cancer Research UK website link above and <https://www.nhs.uk/conditions/pet-scan/> for more information.

#### Staging

Staging is an estimate of how much cancer there is and where is located. Staging is used to plan treatment and future options. Further explanation can be found here: [melanomafocus.org/about-melanoma/types-of-melanoma/uveal-melanoma/](https://www.melanomafocus.org/about-melanoma/types-of-melanoma/uveal-melanoma/).

## Treatments for primary cancer

### RADIATION THERAPY

#### Plaque brachytherapy

Treatment using a radioactive plaque placed on the wall of the eye directly over the tumour in order.

#### Proton beam radiotherapy

Treatment using pinpoint radiation through the front of the eye targeted at the tumour.

#### Stereotactic radiosurgery

Treatment using radiation from several directions outside of the eye.

#### Phototherapy

#### Transpupillary thermotherapy

Laser treatment involving heating the tumour using an infrared laser beam.

#### Photodynamic therapy

A treatment using a dye injected into the arm. Infra-red laser is then directed at the tumour in the eye, which activates the dye and kills the tumour.

### Surgery

#### Endoresection

A method used to remove the eye tumour through a hole in the retina.

#### Enucleation

Removal of the entire eye.

#### Exoresection

A method used to remove the eye tumour through a trapdoor in the wall of the eye.

## ABBREVIATIONS

**CNS** Cancer Nurse Specialist sometimes called a Clinical Nurse Specialist

**CT** Computed tomography

**MRI** Magnetic resonance imaging

**mUM** Metastatic uveal melanoma

**ICI** Immune checkpoint inhibitors

**PET** Positron emission tomography

**SNB** Sentinel node biopsy

**UM** Uveal melanoma

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