# Uveal Melanoma National Guidelines
## Information for the Public

## About this information

### Does this information apply to me?

Yes, if you have uveal melanoma or are the family or carer of someone with uveal melanoma.

Uveal melanoma includes choroidal, ciliary body and iris melanomas.

If you have conjunctival melanoma, this information does not apply to you. Please see the forthcoming guidance from the British Association of Dermatologists.
http://www.bad.org.uk/healthcare-professionals/clinical-standards/clinical-guidelines

### How were the guidelines developed?

These guidelines were developed by a panel of experts, made up of clinicians and patients.

The group has been meeting from April 2012 under the leadership of Dr Paul Nathan to review the evidence and develop recommendations.

Funding was provided by the national charity Melanoma Focus.

### What do the recommendations cover?

The recommendations in the guideline cover the full range of care that should be available from the NHS to adults of all ages with uveal melanoma.

The guideline addresses four main topics:

1. How the tumour in the eye should be managed
2. How future risks should be managed
3. How patients should be looked after beyond their eye treatment
4. How cancer that has spread from the eye should be managed

The full guideline is available at http://melanomafocus.com/activities-2/um-guidelines-resources

# Your care

Your health care team should explain any treatment, care or support you should be offered so that you can decide together what is best for you.

For more information, please see the relevant information from NICE.
http://www.nice.org.uk/nhscare

If you think that the treatment or care you receive does not match the treatment or care described in the pages that follow, you should talk to your doctor or the health worker you have most contact with.

# Introduction to Uveal Melanoma

Uveal melanoma is a cancer of the eye involving the iris, ciliary body or choroid (known as the uvea).

Tumours arise from the pigment cells in the eye called melanocytes. This is the same as melanoma skin cancer, but uveal melanoma is different and is treated differently.

Uveal melanoma is treated in three specialised centres in England: Liverpool, London and Sheffield. These are called “supra-regional” centres.

Uveal melanoma that has spread (called metastatic uveal melanoma) requires the input of many different highly-specialised healthcare professionals. Patients are treated at centres with specialist cancer and liver teams.

The prognosis for uveal melanoma that has spread is usually very poor. There is no chemotherapy that has been found to be effective for metastatic uveal melanoma so patients have to rely on experimental treatments.

# Care recommended in the guideline

## Overview

Patient choice and shared decision-making is at the centre of this guidance. It is acknowledged as important that patients are given the information they need in order to fully participate in their care.

Quality of life is also given importance within the guideline; from selecting the right treatment for the cancer in the eye to agreeing on the most appropriate follow up schedule and deciding on the best treatment for cancer that has spread to other organs (metastatic disease).

The guideline advises that uveal melanoma patients should be dealt with under the national cancer guidelines and the appropriate timelines should be followed.

Collaboration between the various centres treating patients with uveal melanoma is encouraged in the guideline. Patients with metastatic disease should be treated by a specialist team.

## Diagnosis

If your ophthalmologist suspects you have uveal melanoma, he or she should notify one of the national specialist centres within 48 hours.

The relevant specialist centre should then see you within two weeks, as per the NHS Cancer Waiting Time targets.

According to the NHS Constitution, if the centre is unable to see you within two weeks, they should offer you “a range of suitable alternative providers” (in practice this would be one of the other two English centres).

Once you are at the centre, you should have a range of tests and examinations to determine whether you have uveal melanoma or not.

You should be offered your own clinical or medical oncologist who has experience and knowledge of uveal melanoma. You should also be told about any relevant clinical trials in which you may be able to take part.

## Initial treatment

In England, the ocular oncology service offers the full range of treatments for primary uveal melanoma (cancer inside the eye). These include, but are not limited to:

| | |
|---|---|
| Radiotherapy: | Plaque brachytherapy |
| | Proton beam radiotherapy |
| | Stereotactic radiosurgery |
| Phototherapy: | Transpupillary thermotherapy |
| | Photodynamic therapy |
| Surgery: | Exoresection |
| | Endoresection |
| | Enucleation |

Each treatment should be considered by your consultant, and the benefits and risks explained to you. Some treatments will not be appropriate for you, as they will depend on the tumour size and position.

Some treatments may not be available at all three centres, but you should be given the option to travel to an alternate centre if another treatment could be better – for example, if it could spare more of your vision.

Research shows that the choice of treatment makes no difference to how long people live overall. Occasionally some treatments may not initially succeed and a second eye treatment will be required.

As per the NHS Cancer Waiting Time targets, you should be treated within 31 days of your diagnosis.

## Thinking about future risks

Prediction of future risk is often very important to patients, and this is reflected within the guideline.

Whilst a positive outcome in initial treatment of the tumour is good, it does not mean your cancer has not spread. Uveal melanoma spreads through the blood and there is no way of testing for this at the present time.

When you visit your consultant at the eye centre, he or she will be able to get an idea of how dangerous your tumour is from the following factors:

- Your age
- Your gender
- 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

If the tumour is removed from the eye, either by surgery or biopsy, additional information can be provided by looking at the tumour through a microscope:

- The type of cell
- The number of cells dividing
- The patterns seen within the tissue
- The genetic profile of the tumour (requires further tests on the tumour)

The pathologists and clinicians should work together to estimate your overall risk of developing metastatic disease. You should be offered the choice as to how much information you would like to know.

The team at the eye centre should discuss the role of biopsy with you, including the benefits, risks and limitations of the procedure. You should consider the impact of any decision you make on your quality of life.

### What scans are appropriate

Your physician (most likely your clinical or medical oncologist) should have a discussion with you about the most appropriate screening schedule for you.

He or she should talk to you about the possibility of your cancer spreading and the most likely places in the body for it to appear.

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, where you should have scans of your liver and access to a nurse specialist for support.

CT scans of the liver should not be performed and blood tests alone are inadequate.

## What will happen if the cancer spreads

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 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 uveal melanoma and have direct links to the three eye specialist centres. They should have access to all the treatments and trials available nationally.

The recommendation that metastatic uveal patients should be treated by a specialist team reflects 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 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 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 resection, your specialists should consider regional treatments, that is, other treatments directly to the liver.

If the cancer is outside of the liver, or has also spread to other organs, you may be offered anti-cancer drugs. You may be offered one called Ipilimumab, an immunotherapy, which has been approved by NICE. Doctors are still uncertain how effective it is with uveal melanoma. You no longer have to have another treatment before Ipilimumab.

You should also be told about all the available clinical trials nationally that may appropriate for you.

# Questions you may want to ask your healthcare professionals

## Questions for your ocular oncologist

- 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?
- What is the best way of determining my risk of the cancer spreading?
- What tests will you do on my tumour if I have my eye removed or a biopsy?
- What follow-up will I receive after treatment?

## Questions for your clinical or medical oncologist

- Will I be able to work and continue my other usual activities?
- Am I entitled to free prescriptions and what other benefits am I entitled to?
- Is help available for my travel costs?
- Is there a clinical trial available to me?
- How does uveal melanoma spread?
- How will you determine if the cancer has spread?
- Which organs are most likely to be affected?
- What kind of scan will I need and how often?

# Your Feelings

## Diagnosis

Being diagnosed with eye cancer can be very frightening. It can feel like everything is happening very quickly and you may feel dazed as you attempt to make sense of the situation.

The staff at the eye centre should give you the time and support you need to absorb the news. You will usually have a clinical nurse specialist who will answer any additional questions you have and often they will give you their phone number so you can contact them once you have returned home.

Uveal melanoma is so rare you are unlikely to have heard of it before your diagnosis. There are local and national charities and support groups who can help put you in touch with other patients, which can help you feel less isolated.

## Predicting risk

Making a decision about whether to find out if you are at high risk of your cancer spreading can be very difficult. However, it is important to consider it carefully as once the tumour has been treated you may not be able to find out more.

You may find it helpful to talk to your family, other patients and/or healthcare workers (such as your clinical nurse specialist or your GP) about the following:

- If you were to discover you are high risk how do you think you will feel?
- Would you want to be able to take part in future trials that require genetic profiling of the eye tumour?
- How would you feel if you had a biopsy and you did not get a result?
- How would you feel if you had a biopsy and there was a complication?
- How would you feel if you did not have a biopsy and were consequently considered low risk but went on to develop metastatic disease?
- Would more information help you plan for your future?

## Choosing your follow-up

Making a decision about your follow-up scans will be done in collaboration with your consultant, however, you may find it helpful to talk to your family, other patients and/or healthcare workers (such as your clinical nurse specialist or your GP) about the following:

- If you decide to have scanning, how will you feel every six months in the run up to the scan and if there is a wait for the result?

- How will you feel if you are given a false negative or a false positive result? Which is the most appropriate type of scan to try and avoid this?

- If you decide against scanning how would you feel if your cancer spread and you didn't find out until a late stage?

- If a scan found something suspicious, what would happen next?

## Sources of information and support

**OcuMel UK**
A registered charity representing those affected by ocular melanoma.
www.ocumeluk.org
0300 790 0512

**Macmillian Cancer Support**
A registered charity supporting anyone affected by cancer.
www.macmillan.org.uk
0808 808 0000

**Cancer Research UK**
A registered charity providing high quality cancer information and support.
www.cancerresearchuk.org
0808 800 4040

**Rarer Cancers Foundation**
A registered charity campaigning for better services and outcomes for those with rarer cancers.
www.rarercancers.org
0800 334 5551

**The Eye Cancer Forum**
An independent online forum run by patients.
www.eyecancerforum.co.uk

# Explanation of medical words and terms

| | |
|---|---|
| Biopsy | A small piece of the eye tumour is removed using a needle and examined in the laboratory |
| Choroidal melanoma | Uveal melanoma involving the back part of the eye (choroid) |
| Clinical oncologist | A doctor with advanced training in cancer who is able to give radiation and chemotherapy |
| Clinician | A doctor, or other skilled healthcare professional, who has regular contact with patients |
| Conjunctival Melanoma | Melanoma that starts in the outer layer (conjunctiva) of the eye. It has more in common with skin melanoma |
| Consultant | A senior doctor who works in the hospital and has specialist training. They appear on the specialist register |
| CT (Computerised tomography) scan | A scan combining x-rays taken from many different angles. These are combined by computer to create a series of cross sectional images |
| . | |
| 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 |
| Histology | The study under the microscope of the anatomy of cells |
| Histopathologist | A pathologist who specialises in identifying tumours through histology |
| Interventional radiologist | A doctor who uses techniques which rely on the use of radiology to guide them, such as ultrasound-guided biopsy |
| Immunotherapy | Treatment that targets the body's immune system in order to stimulate or dampen a response to the cancer |
| Iris melanoma | Uveal melanoma involving the front, coloured part of the eye |

| | |
|---|---|
| Medical oncologist | A doctor who gives chemotherapy but not radiation. May have more of a research role than a clinical oncologist |
| Metastatic disease | Cancer that has spread from the place where it started to somewhere else in the body |
| MRI (Magnetic resonance imaging) scan | A scan that uses magnetic and radio waves to produce detailed pictures of the inside of the body |
| Ophthalmologist | A doctor specialising in diseases and injuries of the eye |
| Optometrist | A professional trained and licensed to detect visual difficulties and eye conditions and prescribe lenses for glasses (also known as optician) |
| 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 |
| PET (Positron emission tomography) scan | A scan that uses a radioactive tracer to detect disease in the body |
| 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 |
| Phototherapy | General term for treatment using lasers |
| Plaque brachytherapy | Treatment using a radioactive plaque placed on the wall of the eye directly over the tumour in order to kill it |
| Prognostication | The use of tests and other information to predict a patient's outcome, including life expectancy |
| Proton beam radiotherapy | Treatment using pinpoint radiation through the front of the eye in order to kill the tumour |
| Radiotherapy | General term for treatment using radiation |
| Resection | Surgery to remove a tumour or piece of an organ |
| Screening | The process of checking for cancer through the use of scanning and other tools, such as laboratory tests |
| Stereotactic radiosurgery | Treatment using radiation from several directions outside of the eye |

| | |
|---|---|
| Supra regional | Very specialised services provided in a small number of centres only |
| Surgery | Treatment using manual or instrumental means |
| Transpupillary thermotherapy | Laser treatment involving heating the tumour using an infrared laser beam |

## Other relevant guidelines and documents

Documents regarding the NHS Constitution can be downloaded from the UK government website.

https://www.gov.uk/government/publications/the-nhs-constitution-for-england

Alternatively you can order a copy of the NHS Constitution by phoning 0300 123 1002 and quoting the reference number 2900876.

NICE Guidance on Chemosaturation Therapy (PHP):

Chemosaturation via percutaneous hepatic artery perfusion and hepatic vein isolation for primary or metastatic liver cancer

http://www.nice.org.uk/guidance/IPG488

NICE Guidance on Ipilimumab:

http://www.nice.org.uk/Guidance/TA319