Shining a light on uveal melanoma

A Short Guide

This guide has been developed to provide information about uveal melanoma (UM). Use this guide to learn more about UM and explore important information about diagnosis and treatment options.

Advice and guidance are also provided on how to navigate challenges in daily life after diagnosis of UM and during treatment.

Contents

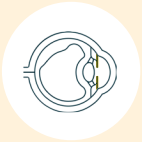
About UM

What is UM?

UM is a rare type of eye cancer found inside the middle layer of the eye wall – this is called the "uveal tract".[1] The different parts of the eye where UM can occur are shown in the images below.

UM develops if the cells in the uveal tract start to grow abnormally forming a cancerous tumour.[1]

The structure and function of the uveal tract



The iris is the coloured part of the eye
It controls the amount of light that can enter the eye,[2] and 4% of UM originates here[1]

The ciliary body is a ring of muscle found behind the iris
It makes fluid to improve eye function and helps the eye focus on objects that are nearby,[3] and 6% of UM originates here[1]

The choroid is a layer of tissue found at the back of the eye
It is filled with blood vessels that bring oxygen and nutrients to the eye,[4] and 90% of UM originates here[1]

UM is estimated to occur in more than 10 people per million in some parts of Europe each year, although the number of people diagnosed varies depending upon their location.[5]

More people are diagnosed with UM in Northern Europe (upwards of 10 people per million) compared to the rest of Europe (average of 6 people per million).[5]

What causes UM?

The cause of UM is not yet known,[6] but there are some risk factors that can increase the chance of it occurring.

These include:

Light coloured eyes: People with blue, grey, or green eyes are more likely to develop UM than people with darker eye colours[1]

Inability to tan: People who sunburn easily have a higher risk of developing UM than those who are able to tan well[7]

Fair skin: UM is more common in people with fair skin from White (Caucasian) ethnicity than in people from Hispanic, Asian, or Black ethnicity[1]

Older age: Whilst UM can be diagnosed at any age, it usually occurs in older adults aged between 50–70 years.[8] It is rare for it to occur in children and teenagers[7]

Inherited skin disorders: People who have skin conditions such as dysplastic nevus syndrome, ocular melanocytosis, or xeroderma pigmentosum, may have a higher risk of developing UM[1]

Genetic mutations: There are some mutations that can happen in a person's genes (part of their DNA) that may cause UM. These genes include *BAP1, GNAQ, GNA11, PLCB4* and *CYSLTR2*[1]

Environmental factors: Some research suggests that a high level of exposure to sunlight, ultraviolet (UV), or blue light may increase the risk of developing UM; however, this remains unclear[1]

What are the symptoms of UM?

Symptoms can vary between different people.[7] Some people experience a range of symptoms, but 30% of people with UM do not experience any symptoms at all.[7]

Blurry vision[7]

Loss of vision[7]

A visible dark patch in the iris that may grow overtime[9]

Eye pain or a feeling of pressure in the eye[9]

Flashes of light or floating specks in vision[7]

Change in the shape or colour of the eye[7]

Other symptoms may also occur which are not listed here[9]

Navigating UM

How is UM diagnosed?

UM is often found during a routine optician appointment.[6] This is partly because 30% of people do not experience symptoms.[7]

If an optician or doctor suspects UM, a referral will be made to a specialist eye doctor called an ophthalmologist.[10] The ophthalmologist will perform tests such as eye examinations including photographs, scans, and ultrasounds; other tests may also be given.[11] If a diagnosis of UM is suspected, treatment will be given in a national eye cancer centre, where a specialist cancer team will provide dedicated care.[10]

Prior to treatment the specialist team will first conduct tests to find out more about the size and position of the cancer and where it is located.[11,12] This is called "staging", and it helps doctors to plan the best treatment for each individual.[12]

Most people who receive a diagnosis of UM will have primary UM (also called "early UM") – this means that the cancer has not spread outside of the eye.[13]

However, there is a small risk (less than 2%) that the cancer may have begun to spread at the time of initial diagnosis.[1,13] If the cancer has spread to other parts of the body, this is called "metastatic UM" (or "mUM" for short).[10] Sometimes the doctors refer to this as "advanced UM".[10]

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What are the treatment options for UM?

The main goals of treatment for UM are to destroy or remove the tumour, preserve vision, and prevent the cancer from growing or returning.[1] The type of treatment provided will depend on many factors including:

- The location, size, and stage of the cancer – whether the cancer is primary UM or mUM[1]
- The level of vision in both eyes[1]
- Presence of other healthcare conditions that must be considered as part of the overall treatment[14]
- Personal preferences[14]

Types of treatment for primary UM may include different forms of radiotherapy, laser therapy, phototherapy, and/or surgical interventions.[1] Sometimes these therapies can be combined.[15] The most common treatment for primary UM is a type of radiotherapy known as brachytherapy.[16,17]

Side effects can occur with UM treatment, and the occurrence and type of side effects may differ between the different treatment options; some side effects are serious and can cause long-term issues.[1]

The doctor and healthcare team will discuss all treatment options for UM in detail, including how they work, potential side effects and complications, and why they are being offered.

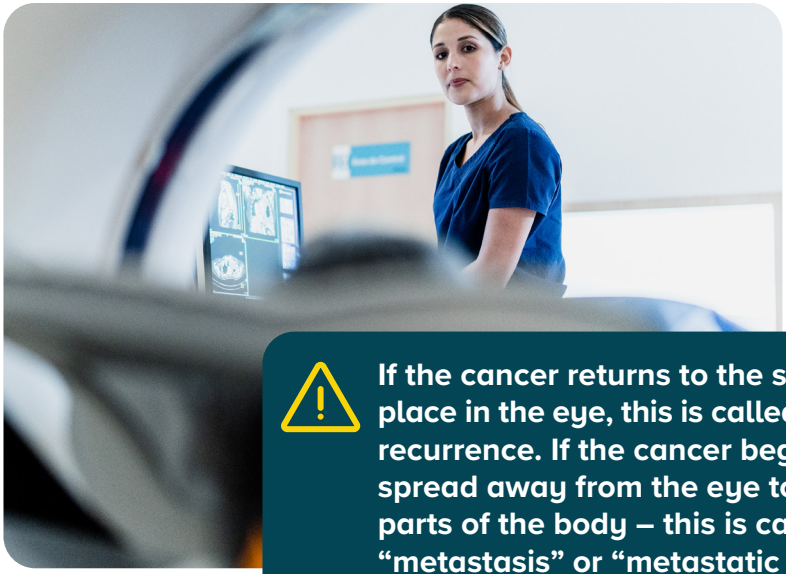
A decision will then be made to find the best option for each person.

What happens after the initial diagnosis and treatment of UM?

After initial treatment for UM, it is important for the doctor to understand how the cancer has responded to the treatment. As part of the follow-up care, each individual will undergo regular monitoring and tests by the doctor and healthcare team over time – this is called surveillance.[12]

As there is a risk that UM may return or worsen overtime, a doctor will perform tests to determine each person's risk of this happening, and a surveillance plan will be put together based on the results of these tests.[18] This will be discussed in-detail by the doctor and healthcare team.

The surveillance plan may include tests such as ultrasounds, X-rays, MRIs, and blood tests.[19] This will allow the doctor to detect any signs that the cancer may have returned to the same location in the eye or if the cancer has started to move to other parts of the body.[19]



If the cancer returns to the same place in the eye, this is called a local recurrence. If the cancer begins to spread away from the eye to other parts of the body – this is called "metastasis" or "metastatic UM" (mUM).[10]

What are the signs and symptoms of mUM?

Signs of mUM may be detected by regular surveillance tests.[20] Sometimes symptoms are noticed by an individual or their doctor that may suggest that the cancer has started to spread.[20]

Symptoms do not always occur and when they do they can differ between patients.[21] If signs and symptoms are experienced, they may include:

Fatigue[21]

Jaundice (yellowing of the skin)[22]

Weight loss[21]

Pain[21]

Abdominal swelling[22]

If any signs or symptoms are noticed it is important that they are reported to the doctor and healthcare team straight away so they can be investigated early and treated quickly.[23]

What are the treatment options for UM that has returned or spread?

If the cancer has returned to the eye (a local recurrence), surgical intervention is the most common treatment.[10] Radiotherapy may also be given.[24]

If the cancer has spread to other places in the body (mUM), the treatment given may be dependent upon where the cancer has spread.[21] Most UM cancers spread to the liver, but they may also spread to other places as well.[25]

Treatment options for UM that has spread to other parts of the body include:

- Types of therapies called "immunotherapy" which help the body fight against cancer cells[1,12]
- Surgery on the liver or other organs to remove the cancer[26,27]
- Treatment directly targeting the liver, including chemotherapy or heat treatment that kills cancer cells[1]

Each type of treatment may cause side effects and they may differ between treatments.[22] Your doctor and healthcare team will discuss and provide detailed information on any side effects or complications that may occur as part of these treatments.

After treatment, the healthcare team will provide a detailed plan for regular surveillance to monitor how the cancer responds to treatment.[18]

Living with UM

Receiving a diagnosis of UM can be a very difficult, challenging, and emotional experience. Hearing from others with UM, and joining a community of people going through a similar experience can help to build a support network, discover new reliable information and find encouragement when needed.

Visit the links below to discover more.

OcuMel UK
www.ocumeluk.org

Melanoma Focus
www.melanomafocus.org

More information on UM

Detailed information on UM diagnosis and treatment will be provided by the doctor and healthcare team. It may also help to ask questions about all aspects of UM from diagnosis to treatment.

More information about UM and a list of prepared questions that may be helpful to ask the healthcare team can be found at www.SpotlightUM.Care

Helpful tools and tips

Many people find it helpful to write down how they are feeling emotionally, physically, and mentally following a cancer diagnosis and whilst they are receiving treatment. This can help track overall health, symptoms, and wellbeing over time. It may also help to take journals and pre-prepared questions to healthcare appointments to get the most from the treatment.

Notes

Use this space to write notes, questions, and record information.

SpotlightUM

References

1. Branisteanu DC, *et al. Exp Ther Med* 2021;22:1428. **2.** Sturm RA, Larsson M. *Pigment Cell Melanoma Res* 2009;22:544-62. **3.** Delamere NA. *Adv Organ Biol* 2005;10:127-148. **4.** Nickla DL, Wallman J. *Prog Retin Eye Res* 2010;29:144-68. **5.** Wu M, *et al. Invest Ophthalmol Vis Sci* 2023;64:45. **6.** Amaro A, *et al. Cancer Metastasis Rev* 2017;36:109-140. **7.** Kaliki S, Shields CL. *Eye (Lond)* 2017;31:241-257. **8.** Lamas NJ, *et al. Cancers (Basel)* 2021;14.96. **9.** Ortega MA, *et al. Int J Oncol* 2020;57:1262-1279. **10.** Macmillan Cancer Support. Eye cancer (ocular melanoma). Available at: https://www.macmillan.org.uk/cancer-information-and-support/melanoma/eye-cancer. Accessed June 2024. **11.** Solnik M, *et al. Cancers (Basel)* 2022;14. **12.** Carvajal RD, *et al. Nat Rev Clin Oncol* 2023;20:99-115. **13.** Garg G, *et al. Br J Ophthalmol* 2022;106:510-517. **14.** Szeligo BM, *et al. Cancers (Basel)* 2021;13:3426. **15.** Bai H, *et al. Clin Exp Ophthalmol* 2023;51:484-494. **16.** Banou L, *et al. Curr Oncol* 2023;30:6374-6396. **17.** Brewington BY, *et al. Clin Ophthalmol* 2018;12:925-934. **18.** Nathan P, *et al. Eur J Cancer* 2015;51:2404-12. **19.** Francis JH, *et al. Am Soc Clin Oncol Educ Book* 2013:382-7. **20.** Lorenzo D, *et al. Jpn J Ophthalmol* 2019;63:197-209. **21.** Gonsalves CF, *et al. AJR Am J Roentgenol* 2015;205:429-33. **22.** Ng CA, *et al. Melanoma Res* 2024;34:248-257. **23.** Carvajal RD, *et al. Br J Ophthalmol* 2017;101:38-44. **24.** Seibel I, *et al. Am J Ophthalmol* 2015;160:628-36. **25.** Krantz BA, *et al. Clin Ophthalmol* 2017;11:279-289. **26.** Frenkel S, *et al. Br J Ophthalmol* 2009;93:1042-6. **27.** Rodriguez-Vidal C, *et al. Cancers (Basel)* 2020;12:2557.

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CM-GB—2400006 | November 2024