Advanced Ocular Melanoma

Patient information

What is ocular melanoma?

Ocular melanoma is a very rare form of cancer that affects the pigment-producing cells of the eye – known as melanocytes.

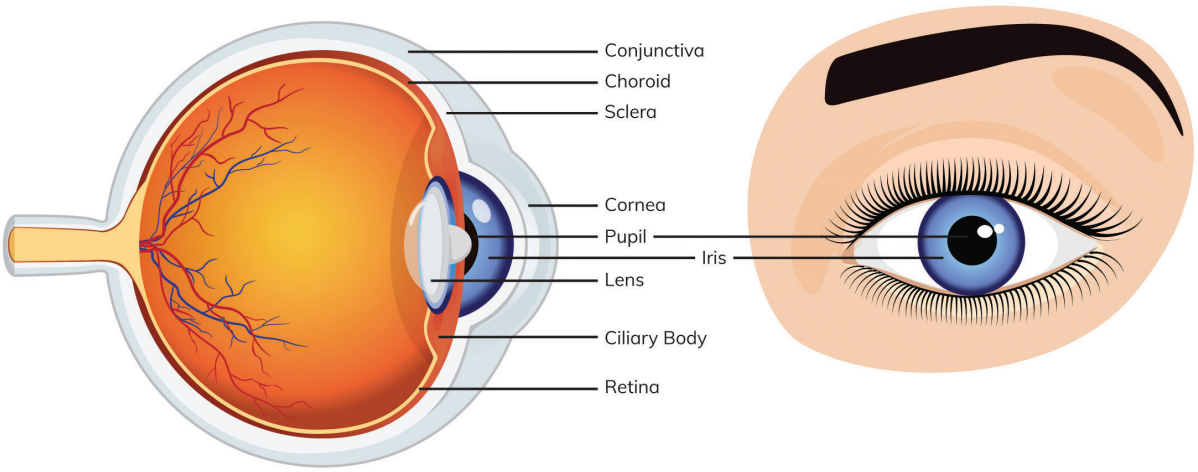
Ocular melanoma occurs when the DNA of the melanocytes in the eye develops errors, causing the cells to grow in an uncontrolled way forming a tumour.

Although they share a name, ocular melanoma and melanoma of the skin are different and distinct diseases.

Melanoma Institute Australia

Most ocular melanomas develop in the uvea – the middle layer of the eye. This is called uveal melanoma.

Ocular melanoma can also develop from the conjunctiva – the clear tissue that covers the white part of the eye. Conjunctival melanoma is rare and has more in common with melanoma of the skin than uveal melanoma.



The uvea consists of the:

- **Iris** – the coloured part of the eye.
- **Ciliary body** – the ring of muscle tissue that changes the size of the pupil and the shape of the lens.
- **Choroid** – the tissue layer at the back of the eye filled with blood vessels.

Uveal melanoma can develop in any of these areas.

Ocular melanoma can also develop from the conjunctiva – the clear tissue that covers the white part of the eye. Conjunctival melanoma is rare and has more in common with melanoma of the skin than uveal melanoma.

What is advanced ocular melanoma?

When ocular melanoma spreads beyond the eye to other parts of the body it is called **advanced ocular melanoma.**

Advanced ocular melanoma most often spreads to the liver, but it can also spread to the lungs, soft tissue and bones.

The risk of spread can depend on a number of factors including where the original tumour developed and your genetics.

Diagnosis

Ocular melanoma can spread beyond the eye before a primary tumour is first diagnosed and treated – although this is rare.

Most of the time, advanced ocular melanoma is diagnosed after a primary tumour has already been treated. About half of all patients treated for ocular melanoma will go on to develop advanced disease.

If you have had a primary ocular melanoma, you may be monitored regularly for disease recurrence. If your doctor suspects that your ocular melanoma has come back or spread to another area of your body, you will be referred for additional testing.

These tests will confirm where the ocular melanoma has spread, and how extensive that spread is.

BLOOD TESTS

- A blood test may be used to check how well the organs in your body are working (liver function tests are commonly used).

IMAGING OF THE BODY

- Computerised tomography (CT), magnetic resonance imaging (MRI) scans, positron emission tomography (PET) and ultrasounds can all be used to identify where ocular melanoma has spread throughout the body.
- An injection of dye into the veins may be used to enhance the clarity of the images.

The role for genetic testing

Genetic testing may be undertaken on a sample of cancerous tissue, taken from the primary ocular melanoma tumour or the site of spread. This testing looks for certain genetic mutations that are thought to 'drive' the growth of ocular melanoma.

Knowing your 'mutation status' may help to inform future treatment options.

These common genetic mutations are:

- **GNAQ and GNA11.** These are the most common mutations in uveal melanoma, found in about 80% of cases.
- **BAP1.** This mutation is found in about half of all uveal melanomas and is associated with a high-risk of cancer spread.
- **BRAF.** This mutation is common in melanoma of the skin but is not seen in uveal melanoma. It can occur in about 30% of conjunctival melanoma.

Treatment

Treatments used for advanced ocular melanoma can differ to the treatments used for melanoma of the skin. As such, it is important that you receive treatment at a specialised centre with doctors who are experienced in this disease area.

Systemic treatments

Systemic treatments are those that target the entire body, circulating through the bloodstream to kill cancer cells. They can include:

- **Immunotherapy** – a type of medicine that prompts your immune system to attack the cancer.
- **Chemotherapy** – a type of medicine that destroys rapidly dividing cells in your body, such as cancer cells.
- **Targeted therapy** – a type of medicine that targets specific genes or proteins involved in the growth and survival of cancer cells.

Currently, there is one systemic therapy for advanced ocular melanoma approved for use in Australia by the Therapeutic Goods Administration.

It is called tebentafusp and it works by drawing immune cells close enough to the ocular melanoma cells to attack them. In a clinical trial, tebentafusp was shown to improve the overall survival of patients with uveal melanoma.

Tebentafusp is only suitable for patients who are HLA-A2 positive (a molecule involved in immune system regulation). Approximately half of all white people – the race most affected by ocular melanoma – are positive for HLA-A2.

Treatments that target the liver

Treatments targeting the liver may be used in combination with systemic treatment.

Some treatments directly target cancerous tumours growing in the liver. These include:

- **Surgery.** Also called a resection, surgery may be used to remove a tumour in the liver when there is only a single tumour present.
- **Ablation.** Conducted through the skin or via surgery, ablation involves inserting small probes into a tumour and heating or freezing it to destroy it. Like surgery, it is typically only used when there is one tumour present in the liver.
- **Radiation.** Radiation therapy uses X-rays to destroy cancer cells. Targeted radiation therapies can be used to treat tumours in the liver, as well as other areas of the body such as the lungs, bone and brain.

Other liver-targeting therapies deliver medicine into the liver. These are called trans-arterial catheter-directed therapies and include:

- **Immunoembolisation.** Immunotherapy drugs are injected into the hepatic artery (the main blood vessel that supplies the liver). Blood supply to the tumours is also blocked.
- **Chemoembolisation (TACE).** Chemotherapy drugs are injected into the hepatic artery to block blood supply to the tumours.
- **Radioembolisation.** Small beads containing radiation are injected into the liver to destroy cancer cells.
- **Hepatic arterial chemoinfusion (HAI).** Chemotherapy drugs are infused into the liver over time. Direct infusion of chemotherapy into the liver results in fewer side effects and allows for higher doses to be delivered.
- **Isolated hepatic perfusion (IHP).** A catheter is placed into the arteries that supply blood to the liver, and into the vein that takes blood away from the liver. This temporarily cuts of the liver's blood supply from the rest of the body and allows for chemotherapy to be delivered directly to the liver only.

Clinical trials

Clinical trials are used to test new treatments to see if they are better than the currently available treatments. They are vital for improving outcomes for people with cancer.

Your doctor may suggest that you should take part in a clinical trial as part of your treatment for advanced ocular melanoma.

You can read more about clinical trials at **australiancancertrials.gov.au.**

Who makes up the treatment team?

Treatment for advanced ocular melanoma is provided by a multidisciplinary team (MDT) of healthcare professionals. This team may include:

- **an ophthalmologist** to diagnose ocular melanoma and conduct surgery to remove the primary tumour.
- **a radiation oncologist** to oversee any radiotherapy you may have to treat the ocular melanoma.
- **a medical oncologist** to prescribe any systemic therapies you may have to treat the ocular melanoma.
- **a palliative care specialist** to enhance your quality of life and reduce the impact of side effects and pain.
- **nurses** to care for your needs during and after treatment.
- **allied health professionals** such as a psychologist, counsellor, social worker, physiotherapist or occupational therapist.

More information and support

This brochure has been developed in partnership with the Australasian Ocular Melanoma Alliance (AOMA) – a group of medical, para-medical, nursing and consumer representatives aiming to enhance care and treatment of patients with ocular melanoma across the world. AOMA is a special interest group of Melanoma and Skin Cancer Trials (MASC Trials).

You can read more about AOMA and ocular melanoma at **aoma.org.au.**

Please note: The information in this brochure is of a general nature and should not replace the advice of healthcare professionals. All care has been taken to ensure the information presented here is accurate at the time of publishing (July 2022).

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