

Early Ocular Melanoma

Patient information

What is early 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.

Early ocular melanoma means that the cancer has not spread beyond the cells of the eye.

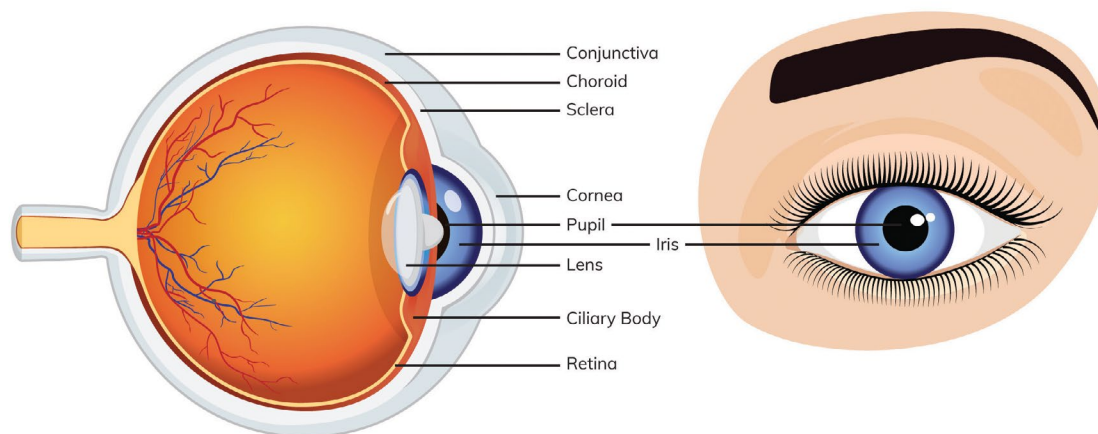


Although they share a name, ocular melanoma and melanoma of the skin are different and distinct diseases.

Where does early ocular melanoma occur?

The eye is made up of three layers. The outer layer is called the sclera, the inner layer is called the retina, and the middle layer is called the uvea.

Most ocular melanomas develop in the uvea. This is called 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.

Causes and risk factors

The underlying cause of ocular melanoma is unclear. However, several risk factors have been linked to an increased risk of developing the disease, including:

- light-coloured eyes
- fair skin
- skin that burns easily
- being older (the average age at diagnosis is 55 years old)
- having certain inherited skin conditions, such as dysplastic nevus syndrome (also known as atypical mole syndrome)
- having a mole or freckle on the surface of the eye, the iris or the choroid.

Although research has found an association with these factors, ocular melanoma can occur in any person, regardless of age, race, gender or eye colour.

Signs and symptoms

Early ocular melanoma may not cause any symptoms. As most ocular melanomas develop in a part of the eye that you cannot see, it can be difficult to recognise and diagnose.

When early ocular melanoma symptoms do occur, they may include:

- vision changes, including flashing, seeing floating spots or wavy lines
- loss of peripheral vision
- eye pain or redness
- visible dark or brown patches on the white of the eye
- a dark spot on the iris
- a change in the shape of the pupil.

These symptoms are common to a number of eye conditions. As such, examination by an eye specialist is needed to diagnose the presence of early ocular melanoma.



Diagnosis

A diagnosis of early ocular melanoma is usually made by a specialist eye doctor, called an ophthalmologist, using a variety of tests explained below.

OPHTHALMOSCOPY

- Also called fundoscopy.
- This examination allows the doctor to look inside the eye to check for abnormalities.
- Eye drops that dilate the pupil may be used to aid in the examination.
- A diagnosis of ocular melanoma can sometimes be made through this examination alone.

FUNDUS PHOTOGRAPHY

- Also called colour fundus photography.
- In this test, photographs are taken of the back of the eye to identify a tumour.
- Taking photographs before and after treatment can help the doctor to determine if treatment has been effective.

ULTRASOUND

- This examination uses sound waves to create an image of the inside of the eye.
- Anaesthetic eye drops are applied to numb the eye, and an ultrasound probe is placed on the surface of the eye.
- This can be helpful to determine the size, shape and location of a tumour.

TRANSILLUMINATION

- This examination may be performed before surgery to show exactly where a tumour is.
- In a darkened room, a doctor will shine a very bright light into the eye to identify the abnormal areas.

ANGIOGRAPHY

- This examination looks at blood vessels and blood flow inside the eye.
- A fluorescent dye is injected into a blood vessel in the arm. As the dye travels through blood vessels in the eye, a special camera takes pictures of the retina and choroid to detect any blockages or leakages.

BLOOD TESTS

- A blood test may be used to check how well organs in your body are working.
- Analysing liver function tests can help to determine whether a tumour has spread, but this is not very accurate.

BIOPSY

- Most of the time, a diagnosis of ocular melanoma can be made without the need for a biopsy. Sometimes, however, a biopsy will be performed.
- During a biopsy, a small sample of tissue is extracted from the suspicious area, usually using a fine needle. The tissue sample is then examined in a laboratory.

Treatment

Early ocular melanoma should be treated promptly to reduce the risk of the disease progressing and spreading to other areas of the body.

Treatment options for early ocular melanoma will depend on a number of factors, including:

- the location and size of the melanoma
- how close the melanoma is to other parts of the eye
- your age, fitness and general health
- your personal preferences.

Treatment options may include radiotherapy, surgery or a combination of the two.

Treatment for early ocular melanoma is not always recommended. For example, in elderly or chronically ill patients who do not have symptoms, or who have small sized tumours, close monitoring and 'watchful waiting' may be recommended.

Radiotherapy

Radiotherapy uses X-rays targeted at a tumour to kill the cancer cells. There are two main forms of radiation therapy used to treat early ocular melanoma – brachytherapy and external beam radiation therapy.

BRACHYTHERAPY

- Brachytherapy is the most common form of radiotherapy used for early ocular melanoma.
- The process requires an operation during which a plaque containing radioactive seeds is positioned on the eye.
- The plaque remains in place for up to a week and delivers targeted radiotherapy confined to the site of the tumour.
- It is generally used to treat smaller ocular melanomas.

EXTERNAL BEAM RADIATION THERAPY

- External beam radiation therapy (EBRT) uses a machine that delivers a beam of radiation through the front of the eye to the affected area.
- It is generally used to treat medium to large sized early ocular melanomas, including those that cannot be treated with brachytherapy due to their location or the obstruction caused by the optic nerve.

The **side effects** of brachytherapy and EBRT are similar and can include:

- damage to the blood vessels that supply the retina (radiation retinopathy)
- damage to the nerves that allow a person to see (optic neuropathy)
- increased risk of cataracts, poorer vision or bleeding in the eye.

Surgery

Surgery for ocular melanoma may involve removing the tumour and some healthy tissue surrounding it or removing the entire eye.

LOCAL TUMOUR REMOVAL

- Local tumour removal is surgery to remove the ocular melanoma tumour and some healthy tissue surrounding it.
- There are different types of local removal surgeries depending on the part of the eye that is removed:
 - **Iridectomy** – removal of part of the iris.
 - **Iridocyclectomy** – removal of part of the iris and ciliary body.
 - **Endoresection or transscleral resection** – removal of the tumour in the ciliary body or choroid.
- The surgery is conducted under general anaesthetic and will require a stay in hospital.

REMOVAL OF THE EYE (ENUCLEATION)

- Enucleation is removal of the entire eye.
- This type of surgery is performed when radiotherapy alone may not be enough to treat the tumour. For example, for larger tumours, or when the cancer has spread to nearby tissues (local spread).
- Following surgery, a prosthetic (artificial) eye can be placed in the eye socket.

The **side effects** of surgery can include:

- loss of vision
- damage to the retina and other nearby tissue
- changes in appearance
- blood clots or internal bleeding in the eye.

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 early ocular melanoma. You can read more about clinical trials at australiancancertrials.gov.au.

Follow up care and management

During treatment for ocular melanoma, you will attend regular follow-up visits with your doctors to ensure your treatment is working.

After treatment has finished, you will continue to attend follow-up visits to ensure the cancer has not returned or spread.

Follow-up visits may involve routine eye examinations, blood tests and/or imaging scans. An appointment with a dermatologist/skin physician may also be needed to assess your skin.

Who makes up the treatment team?

Treatment for early 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 tumour.
- **a radiation oncologist** to oversee any radiotherapy you may have to treat the ocular melanoma.
- **a medical oncologist** to prescribe any medical treatments you may have to treat the ocular melanoma.
- **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 2021).

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