

Understanding Eye Melanoma

A guide for people affected by eye cancer

This fact sheet is about a cancer of the eye called melanoma. It may also be called ocular melanoma or uveal melanoma. We hope this information helps you understand how this cancer is diagnosed and treated. We also include where to find support services.

About the eye

The eye is like a camera, using light to make pictures. It has layers of tissues and is filled with a clear jelly.

When we look at someone's eye, we see the clear front layer of the cornea, the dark centre called the pupil, and the coloured ring called the iris. All of these are protected by a layer called the conjunctiva.

Behind the white outer part of the eye (sclera) that you can see is a middle layer called the uvea. The uvea is

made up of 3 parts: the iris, the ciliary body and the choroid (see the diagram below). The front section (where the iris sits) is called the anterior uvea, and the back section is called the posterior uvea.

What is eye melanoma?

Inside the eye are cells called melanocytes which produce melanin – the pigment that also gives colour to your skin. In rare cases, these cells change and grow into melanoma, a type of cancer. A melanoma that starts in the eye may be called ocular melanoma. It may also be named by the part of the eye it starts in, for example, uveal melanoma starts in the uvea.

You can't see most types of eye melanomas when looking at the outside of the eye, so they can be hard to notice or check for. But these melanomas may sometimes be seen during an eye test or other scan.

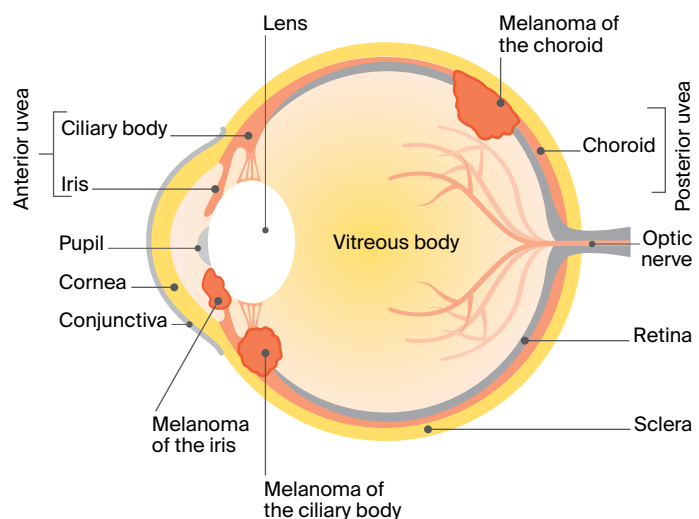
The eye: where melanoma starts

The choroid or posterior uvea – The layer between the retina and white outer part (sclera). It contains melanocytes and blood vessels that deliver oxygen and nutrients to the retina. About 90% of ocular melanomas start here.

The ciliary body – This part of the eye controls the shape of the lens. It makes the fluid the eye needs and maintains eye pressure. About 5–10% of ocular melanomas are found here.

The iris – This is the coloured part of the eye that helps regulate the amount of light entering the eye. Fewer than 5% of eye melanomas are found in the iris.

Conjunctival melanoma is a rarer type of eye melanoma that starts in the conjunctiva.



How common is eye melanoma?

Uveal melanoma is the most common type of cancer that starts inside the eye, but it is still rare. About 200 Australians are diagnosed with it each year, and it is more common in older people and in men. Conjunctival melanoma is very rare.

What are the risk factors?

It's unclear whether UV exposure (a risk factor for skin melanoma) is a risk factor for eye melanoma. Some studies have linked conjunctival melanoma to UV exposure (e.g. using tanning beds). Eye melanoma is less common in people with naturally brown eyes, and people with dark skin. While doctors don't know the exact cause, factors that may increase your ocular melanoma risk include:

- fair skin that burns easily
- blue or light eye colour
- fair or red hair
- older age (risk increases with age)
- a family history of melanoma
- certain skin conditions (e.g. dysplastic naevus syndrome)
- eye "freckles" or growths
- primary acquired melanosis (PAM), which causes brown or dark pigmentation on the conjunctiva, usually in one eye. People with PAM need regular check-ups so that any changes can be found early.

What are the symptoms?

Eye melanoma often has no symptoms, but if they happen, they may include:

- blurred or poor vision in one eye
- loss of peripheral vision – the outer edges of what you see
- dark patches on the white of the eye
- a dark spot on the iris
- floaters – dots or squiggles in your vision
- seeing flashing lights
- seeing shadows
- change in pupil shape.

These symptoms can also happen with other eye problems, but see a doctor if you notice changes.

Symptoms of conjunctival melanoma include a new or growing brown or dark patch on the white of the eye, or a raised skin-coloured bump on the eyeball.

Diagnosis

Your doctor or optometrist may check you for ocular melanoma or send you to an ophthalmologist, who also treats eye problems. Tests you may have include:

Ophthalmoscopy (fundoscopy) and slit lamp exam – These tests help doctors see inside your eye. A slit lamp is a microscope with a bright light that looks at the front section of the eye.

Eye drops may be used to make your pupil bigger so it's easier to look inside your eye. The drops can cause blurry vision or light sensitivity for a few hours, so take sunglasses to your test. You'll need to wait until your eyesight returns to normal before driving.

Colour fundus photography and ultra-wide-field fundus photography – These photography machines take detailed photos of large areas of the back of your eye (called the fundus). Drops may be used to widen (dilate) your pupil before the camera takes pictures. This test may be compared to previous imaging and used to help track changes before and after treatment.

Ultrasound – This scan uses sound waves to create pictures of the inside of the eye. It uses a small probe with ultrasound gel that is placed on the eye or eyelid and gently moved over the area. It can measure the size and shape of the tumour.

OCT (Optical Coherence Tomography) scan – This test uses light to create pictures of the retina, uvea and blood vessels. A rest for your chin and forehead helps to move you into position for the scans, which take a few minutes. The machine does not touch your eye.

Transillumination – This test may be done before surgery to show exactly where the melanoma is. The lights in the room are dimmed and a very bright light is shone into your eye to look for abnormal areas.

Angiogram of the eye – This may be done at an eye hospital or by a specialist ophthalmologist in the eye clinic. Drops are put into the eye and a dye is injected into a vein in your arm. The doctor then photographs the back of the eye to check the blood flow (circulation).

Biopsy – This removes a small sample of tissue with a needle to check for cancer cells. You will have either a local or general anaesthetic so you don't feel any pain. Doctors usually diagnose ocular melanoma using imaging tests, but a biopsy is sometimes also needed.

Genetic tests may be done on a biopsy sample to find out the risk of the cancer spreading to other parts of the body. The cancer may be classified as low, intermediate or high risk.

CT (computerised tomography) or MRI (magnetic resonance imaging) scans – These scans are used to check the rest of the body for signs of cancer. You lie on a table that slides into a scanner. Sometimes a dye is injected into a vein in your arm to make the images clearer.

Staging

This is the process of finding out how far the cancer has spread within the eye, and if it has travelled to other parts of the body such as the liver, lungs, lymph nodes or bones.

Early eye melanoma means that the cancer has not spread beyond the eye. If eye melanoma spreads to other parts of the body, it is called advanced or metastatic melanoma.

Treatment

You will be cared for by a multidisciplinary team (MDT) of health professionals. Because melanoma found in the eye is a rare cancer, you may also be referred to a specialist treatment centre or an eye hospital. The MDT may include an ophthalmologist, radiation oncologist (to prescribe and coordinate a course of radiation therapy), medical oncologist (to prescribe and coordinate drug or systemic treatments, such as immunotherapy), nurses, and allied health professionals (e.g. psychologist, counsellor, social worker, physiotherapist or occupational therapist).

Your doctor will talk to you about the best treatment available to you. Options will depend on tumour size and location, and whether it has spread. The aim of

treatment is to remove the cancer, and try to save the eye and as much vision as possible.

Treatment options may include surgery, radiation therapy, laser treatment, photodynamic therapy and immunotherapy. These may be given alone or in combination. There may be new treatments being investigated through clinical trials (including targeted therapy) and through the Australasian Ocular Melanoma Alliance (AOMA).

► See our *Understanding Clinical Trials and Research* booklet.

Certain small eye melanomas may not need immediate treatment. Instead, you may have regular checks until there is growth or an increased risk.

Surgery

Surgery may be done under a local anaesthetic with sedation or under a general anaesthetic. You may need to stay in hospital for a period of time.

► See our *Understanding Surgery* booklet.

Types of surgery for eye melanoma

iridectomy	Removal of part of the iris (the coloured part of the eye). May be done for a biopsy or to remove small melanomas.
iridocyclectomy	Removal of part of the iris and the ciliary body. Used for certain small to medium melanomas.
enucleation	Removal of the entire eyeball. This is done if the melanoma is large or if vision in the eye has already been lost. After surgery, an artificial eye is usually fitted to match the size and colour of your other eye.
orbital exenteration	Removal of the eyeball and some surrounding tissue. This may include some or all of the eyelid, fat and muscles. Only used for advanced cancers, or cancers that have grown into the area that surrounds the eyeball.

Radiation therapy

Radiation therapy uses high-energy rays to destroy cancer cells. It may be used:

- after surgery, to destroy any remaining cancer cells and stop the cancer coming back
- if the cancer can't be removed with surgery
- instead of removing the eye (enucleation)
- if the cancer has spread to other parts of the body.

Radiation therapy doesn't hurt and is given over a set time period. It's given in different ways.

Plaque brachytherapy – A small disc (a plaque) containing radioactive material is put over the tumour during surgery. You may have a general anaesthetic (so you are asleep) or a local anaesthetic with sedation (so you are awake but relaxed and feel no pain). You stay in hospital for 2–7 days with the disc in place. Then you have a short operation to remove the disc and go home.

- ▶ Visit The Royal Victorian Eye and Ear Hospital at eyeandear.org.au/patients-visitors/fact-sheets to read their fact sheet, *Plaque Radiotherapy*.

Stereotactic radiation therapy – This uses radiation in the form of x-ray beams. A large machine directs multiple small beams of radiation to precisely target the tumour in high doses. You usually need 5 sessions given over 10 days. This is planned during your first appointment. You will meet with a radiation oncologist and lie on an examination table for a CT scan (in the same position as during treatment). A mesh mask keeps your head still, but can make some people feel anxious. The specialist will plan the treatment area, type of radiation and how to deliver the right dose. Radiation therapists then deliver the treatment sessions over a number of days. A treatment session doesn't take very long and you can usually go home after each one.

Proton beam radiation therapy – This uses proton beams rather than x-ray beams. Protons release most of their radiation within the cancer, protecting nearby tissue. Treatment is given in high doses over several days. Proton therapy is most useful near very sensitive areas. It's not currently operating in Australia but may be available in the future. Talk to your radiation oncologist your options.

- ▶ See our *Understanding Radiation Therapy* booklet.

Should I join a clinical trial?

Your doctor may suggest you take part in a clinical trial. Clinical trials test new or modified treatments and ways of diagnosing disease to see if they are better than current methods. For example, if you join a randomised trial for a new treatment, you'll be chosen at random to receive either the best existing treatment or the modified new treatment. Over the years, trials have improved treatments and led to better outcomes for people diagnosed with cancer. If you decide to take part in a clinical trial, you can withdraw at any time. For more information, visit australiancancertrials.gov.au.

- ▶ See our *Understanding Clinical Trials and Research* booklet and website information.
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Laser treatment or photodynamic therapy

Laser treatment uses an infrared laser to heat and destroy cancer cells. Laser treatment is sometimes combined with photodynamic therapy, which uses a laser combined with a light-sensitive drug to destroy cancer cells. The drug is injected into your vein and makes the cells in your body more sensitive to light. The treatment is painless, but you will be sensitive to light for several days after treatment.

Immunotherapy

Immunotherapy is a drug treatment that uses your body's immune system to fight cancer. Immunotherapy for uveal melanoma that has spread is available for eligible patients (see eviQ.org.au). Other immunotherapy drugs may become available, and some are being investigated in clinical trials. Ask your doctor if these treatments are suitable for you.

Targeted therapy

Targeted therapy is a drug treatment that targets specific features of cancer cells to stop the cancer growing and spreading. Drugs are given as tablets or through a drip into a vein. They travel through the body like chemotherapy, but they work in a more focused way. A clinical trial (see above) is testing targeted therapy for eye melanoma. Your doctors may test the cancer cells (from a biopsy, see page 3) to see if this treatment is likely to work for you.

Managing side effects of treatment and changes to how you see

All treatments can cause side effects. The side effects you may have depend on the treatment you receive. Some people have very few side effects and others have more. Before your treatment begins, your specialist team will discuss all possible side effects, both short and long term (including those that have a late effect and may not start immediately).

Dry eye and eye discomfort



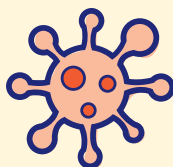
- Use lubricating eye drops (artificial tears) as recommended by your doctor.
- Avoid being around smoke, wind and air conditioning where possible.
- Wear wrap-around sunglasses outdoors to protect your eyes and reduce dryness.
- Take regular breaks from computer screens and remember to blink often.
- If you have pain or swelling after surgery or treatment, take pain medicines exactly as prescribed and tell your doctor if the pain is not controlled.
- Use cold compresses around the area if they have been recommended by your health care team – but never directly on the eye.
- Rest and avoid heavy lifting or strenuous activity until your doctor says it's safe.
- Contact your treatment team urgently or go to an emergency department if you have increased pain, redness, discharge or fever – these may be signs of infection.

Vision changes



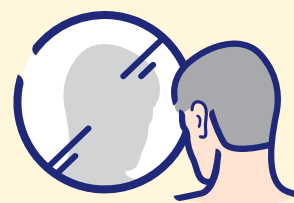
- You may have blurry or double vision, loss of peripheral vision or other loss of vision. Ask your eye specialist if these changes will be temporary or permanent, and what else to expect over time.
- Use good lighting at home and avoid glare where possible.
- Large-print books, magnifiers, screen-zoom settings and text-to-speech apps can make reading easier.
- If you have double vision, an eye patch or special glasses may help – ask your ophthalmologist or optometrist.
- An occupational therapist can suggest practical adjustments for daily tasks such as cooking, reading or using technology.

Risk of infection



- Wash your hands regularly and avoid close contact with people who are unwell.
- Tell your doctor straightaway if you develop fever, chills or flu-like symptoms (especially if you have had immunotherapy).
- Make sure to go to your follow-up appointments so that side effects can be monitored early.

Changes in appearance (including artificial eye)



- It can take time to adjust emotionally to changes in how you look – this is very common.
- If you have an artificial eye (prosthesis), you'll be shown how to care for and clean it, and who to contact if it feels uncomfortable.
- Specialist ocular prosthetists can adjust the artificial eye to improve comfort and appearance.
- Talking with a counsellor or connecting with others who've had similar treatment can be helpful.

When to seek help urgently



Contact your treatment team or go to an emergency department if you have any of the following:

- sudden or severe vision loss
- increasing eye pain or swelling
- signs of infection (fever, redness, discharge)
- severe headaches, chest pain, shortness of breath, or ongoing diarrhoea during immunotherapy.

Making treatment decisions



Understand your options

Learn about all available treatments (e.g. surgery, chemotherapy, radiation therapy, targeted therapy or immunotherapy), including what they involve.



Benefits vs side effects

Consider how each option may help control the cancer and what short- and long-term side effects might occur.



Your values matter

Think about what's most important to you – quality of life, length of treatment, recovery time, fertility, work or school, and family responsibilities.



Ask questions

It's okay to ask your doctor or health care team to repeat something or explain things in plain language



Second opinions

Getting another medical opinion is common and can help you feel more confident.



Time to decide

Some decisions are urgent, while others allow time to think. Ask how much time you have to think about it



Written information

Use fact sheets and reputable websites to review information at your own pace.



Shared decision-making

Decisions are usually made together with your health care team, respecting your preferences.



Travel and costs

You may need to consider whether to travel away from home to have treatment. You may be able to claim some of your travel costs via a government scheme. To find out more, call Cancer Council 13 11 20.

Looking after yourself

Having a range of emotions is common – from feeling shocked, overwhelmed, anxious or fearful, to feeling sad or lonely. Many people need emotional support before, during and after cancer. Getting used to scars, changes in how you look, or changes to your vision can be hard and may take time.

It can help to talk things through with a counsellor, psychologist, friend or family member. Ask your medical team or call Cancer Council 13 11 20 to find support services near you.

► See our *Emotions and Cancer* booklet.

Follow-up appointments

After treatment, you will have regular appointments to check that the cancer hasn't come back and to monitor any side effects. You may have scans, eye tests and physical examinations. You may have access to a care navigator who can assist with questions and appointments.

Some cancer centres will give you a survivorship care plan. This includes a summary of your treatment, sets out a schedule for follow-up care and lists any symptoms and long-term side effects to watch out for. It also tells you about any medical or emotional problems that you may experience and suggests ways to live a healthy lifestyle. If you don't have a survivorship care plan, ask your doctor for one and give a copy to your GP and your health team.

If the cancer comes back

For some people, eye melanoma does come back after treatment, which is called a recurrence. Treatment will depend on where the cancer is in your body, but may include surgery, radiation therapy, laser therapy or immunotherapy. A clinical trial may also be recommended (see page 4).

In some cases of advanced cancer, the main focus of treatment will be on managing any symptoms, such as pain, and improving quality of life. This is called palliative treatment and can be provided at home, in hospital, in a palliative care unit or hospice, or in a residential aged care facility.



Cancer in the eye is a rare type of cancer. Rare cancers can have their own challenges. You may need to travel for treatment or your doctor may not have treated this cancer. For more information, read our *Understanding Rare and Less Common Cancers* fact sheet.

Questions for your doctor

This checklist may be helpful when thinking about the questions you want to ask your doctor.

- What type of eye melanoma do I have?
- Has the cancer spread? Is it early or advanced?
- Will I lose my eyesight?
- What tests will I need to have?
- What are the treatment options for me? What do you recommend and why?
- What are the possible risks and side effects of my treatment? How will these be managed?
- Have you treated this type of cancer before?
- How complex is the surgery and how long does the operation take?
- How long will other treatments I may have take?
- Am I able to speak with someone who has had this type of cancer and treatment?
- Is this treatment covered by Medicare or private insurance? Will there be extra expenses?
- Are there any complementary therapies that might help me?
- If the cancer comes back, how will I know?
- If I am part of a clinical trial, what are the possible benefits and risks?

Where to get help and information

Call Cancer Council 13 11 20 for more information about eye melanoma. Our experienced health professionals can listen to your concerns, put you in touch with services and send you our free booklets. You can also visit your local Cancer Council website.

ACT	actcancer.org
NSW	cancercouncil.com.au
NT	cancer.org.au/nt
QLD	cancerqld.org.au
SA	cancersa.org.au
TAS	cancer.org.au/tas
VIC	cancervic.org.au
WA	cancerwa.asn.au
Australia	cancer.org.au

Other useful websites

Australasian Ocular Melanoma Alliance (AOMA)	aoma.org.au
Melanoma Patients Australia (MPA)	melanomapatients.org.au
Melanoma Institute Australia (MIA)	melanoma.org.au
Melanoma and Skin Cancer Trials Ltd (MASC Trials)	masc.org.au
Rare Cancers Australia	rarecancers.org.au
Vision Australia	visionaustralia.org

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Note to reader

Always consult your doctor about matters that affect your health. This fact sheet is intended as a general introduction and is not a substitute for professional medical, legal or financial advice.

Information about cancer is constantly being updated and revised by the medical and research communities. While all care is taken to ensure accuracy at the time of publication, Cancer Council Australia and its members exclude all liability for any injury, loss or damage incurred by use of or reliance on the information provided in this fact sheet.

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Cancer Council acknowledges Traditional Custodians of Country and recognises the continuing connection to lands, waters and communities. We pay our respects to Aboriginal and Torres Strait Islander cultures and to Elders past and present.

