Cancer Council

Cancer information fact sheet

Understanding Ocular (Uveal) Melanoma

A guide for people affected by eye cancer

This fact sheet has been prepared to help you understand more about ocular (uveal) melanoma.

Many people look for support after being diagnosed with a cancer that is rare or less common than other cancer types. This fact sheet includes information about how ocular melanoma is diagnosed and treated, as well as where to go for additional information and support services.

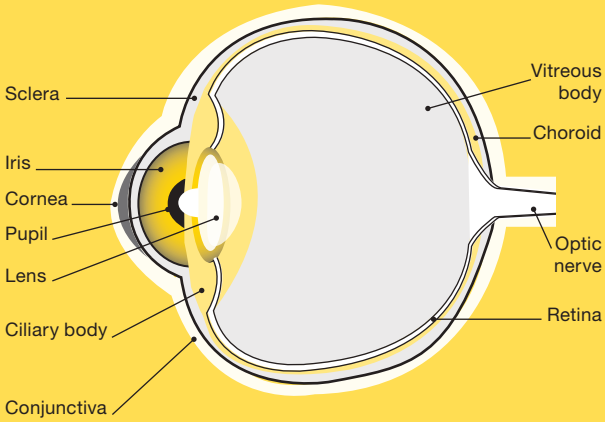
Many people feel shocked and upset when told they have cancer. You may experience strong emotions after a cancer diagnosis, especially if your cancer is rare or less common like ocular melanoma. A feeling of being alone is usual with rare cancers, and you might be worried about how much is known about your type of cancer and how to manage it. You may also be concerned about the cancer coming back after treatment. Contacting local support services (see last page) can help overcome feelings of isolation and will give you information that you may find useful.

About the eye

The human eye is a sense organ that reacts to light and allows us to see. Your eye works in a similar way to a camera. When you look at an object, light passes through the cornea (the clear front layer of the eye) and enters the eyes through the pupil (the black centre of the eye). The iris (the coloured part of the eye) controls how much light the pupil lets in.

Light then passes through the lens (the clear inner part of the eye) which, together with the cornea, focuses light onto the retina. When light hits the retina (layer of tissue at the back of the eye) special cells called photoreceptors convert the light into electrical impulses that travel through the optic nerve to the brain. The brain then turns these signals into the images that you see.

The human eye



What is ocular melanoma?

There are many different types of cancer that can affect the eye, but ocular melanoma is the most common. Melanoma is a type of cancer that develops in the cells of the body that produce melanin — the pigment that gives your skin its colour. Your eyes also have melanin-producing cells and can develop melanoma. Ocular melanoma is also known as uveal melanoma. The uvea is the middle layer of the eye beneath the white part and consists of the iris, ciliary body and choroid. Melanoma can occur any of these parts. It can also be named according to the part of the eye it started in.

Melanomas that develop on the skin usually occur on parts of the body that have been exposed to the sun. Some melanomas, however, can also start inside the eye or in a part of the body that has never been exposed to the sun.

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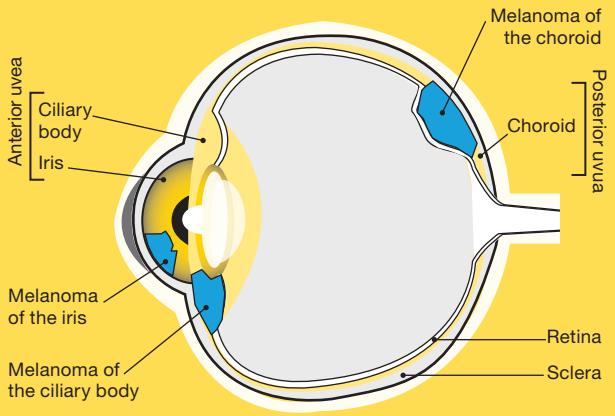
Ocular melanoma is much rarer than skin melanoma and behaves very differently. Normally, cells multiply and die in an orderly way, so that each new cell replaces one lost. Sometimes, however, cells become abnormal and keep growing. In cancers such as ocular melanoma, the abnormal cells form a mass called a tumour. Cancerous tumours, also known as malignant tumours, have the potential to spread to other parts of the body through the blood stream or lymph vessels and form another tumour at a new site. This is known as secondary cancer or metastasis.

Most ocular melanomas develop in the part of the eye that you can't see when looking in a mirror, so this makes ocular melanoma hard to diagnose. Ocular melanoma usually develops in any of the following three areas of the eye:

- the iris – the coloured part of the eye which helps regulate the amount of light entering the eye
- the ciliary body – the part of the eye that controls the shape of the lens and makes the fluid in the eye called aqueous humour, which provides nutrition and maintains pressure in the eye
- the choroid or posterior uvea – the vascular layer of the eye between the retina and the white outer layer (sclera).

These three areas are known as the uvea, hence the term uveal melanoma. Ocular (uveal) melanoma can occur in any of these areas, but it is more common in the choroid.

Where ocular (uveal) melanoma occurs



How common is ocular (uveal) melanoma?

Ocular (uveal) melanoma is rare. Each year, around 125–150 Australians are diagnosed with this type of cancer (about 5–6 cases per million people).[1] It is more likely to be diagnosed in men than women, and can occur at any age, but the risk increases with age.

What are the risk factors?

The cause of ocular melanoma is not known in most cases. However, there are several risk factors including:

- having pale or fair skin. People whose skin burns easily are most at risk.
- having a light eye colour. People with blue or green eyes have a greater risk than people with darker eyes.
- family history of melanoma. A very small number of people who have melanoma have inherited a faulty gene.
- having a growth on or in the eye. People with an "eye freckle" may be at risk.
- age. The risk increases with age.
- certain skin conditions and pigmentation. Some people have a skin disorder (dysplastic naevus syndrome) which causes moles to grow abnormally, and this can increase your risk.

What are the symptoms?

Ocular melanoma can be difficult to diagnose as it forms in the part of the eye that isn't visible to you or others. It doesn't typically cause any signs and symptoms and is usually detected by an optometrist during a routine eye test.

Symptoms that some people may experience include:

- poor or blurred vision in one eye
- loss of peripheral vision
- brown or dark patches on the white of the eye
- a dark spot on the iris
- small specks, wavy lines or 'floaters' in your vision
- flashes in your vision
- a change in the shape of the pupil.

These symptoms can be caused by other eye conditions, but if you experience any of these symptoms you need to discuss them with your doctor.

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Diagnosis

If your doctor or optometrist thinks that you may have ocular melanoma, they will carry out certain tests. If the results suggest that you may have ocular melanoma, your doctor will refer you to a specialist doctor called an ophthalmologist who specialises in treating eye disorders. The ophthalmologist will carry out more tests that may include:

- **Ophthalmoscopy (funduscopy) –** a test that allows your doctor to look at the inside of your eye to check for abnormalities. You may be asked to look into a large microscope that sits on a table (a slit lamp examination). The doctor may put eye drops in your eye to widen (dilate) your pupil. This will allow the doctor to see inside your eye, so they may not have to perform a biopsy (see opposite) to determine if a tumour is present. The eye drops make your eyesight blurry for a few hours and you might find bright light uncomfortable, so take sunglasses to your appointment. You cannot drive until your eyesight returns to normal.

- **Colour fundus photography –** in this test, photographs of the back of your eye (fundus) will be taken and can help show what the tumour looks like before and after treatment. The doctor will put eye drops in your eye to widen (dilate) your pupil and then use a special camera to take pictures of the fundus.

- **Ultrasound scan –** this test uses soundwaves to create pictures of the inside of your eye and surrounding areas. For this scan a gel will be spread over your closed eyelid and a small device called a transducer is moved over the area. The transducer sends out soundwaves that echo when they come across something dense, like an organ or tumour. The ultrasound images are then projected onto a computer screen. An ultrasound is painless, takes only a few minutes and accurately shows the size of the tumour.

- **Transillumination –** if you need surgery, this test may be done first to show exactly where the melanoma is. The lights in the room are turned down and the doctor shines a very bright light into your eye to look for abnormal areas.

- **CT (computerised tomography) or MRI (magnetic resonance imaging) scans –** special machines are used to scan and create pictures of the inside of your body and are used to find tumours or to check for any spread of disease. Before the scan you may have an injection of dye (called contrast) into one of your veins, which makes the pictures clearer. During the scan, you will need to lie still on an examination table. For a CT scan the table moves in and out of the scanner which is large and round like a doughnut; the scan itself takes about 10 minutes. For an MRI scan the table slides into a large metal tube that is open at both ends; the scan takes a little longer, about 30–90 minutes to perform and the machine is noisy so you will be given earplugs to wear. Both scans are painless.

- **Biopsy –** most of the time, the ophthalmologist can make a diagnosis from what they can see when they examine your eye, from photographs and ultrasound pictures. However, sometimes a biopsy is performed. In a biopsy, some tissue is removed from the affected area so it can be examined more closely under a microscope.

Finding a specialist

Rare Cancers Australia have a directory of health professionals and cancer services across Australia: rarecancers.org.au.

Treatment

You will be cared for by a multi-disciplinary team (MDT) of health professionals during your treatment for ocular melanoma. The team may include an ophthalmologist, radiation oncologist (to prescribe and coordinate a course of radiation therapy), medical oncologist (to prescribe and coordinate a course of systemic therapy which includes immunotherapy), nurses and allied health professionals such as a psychologist or counsellor, a social worker, physiotherapist and occupational therapist.

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Discussion with your doctor will help you decide on the best treatment for your cancer depending on:

- the site of the cancer you have (choroid, ciliary body or iris)
- size of the cancer
- how close the cancer is to other parts of the eye
- whether or not the cancer has spread
- your age, fitness and general health
- your preferences.

The main factors in deciding on what treatment you will have are the location and size of the tumour and wanting to save the sight of your eye. Preserving how your eye looks is also important. Treatments may include surgery, radiation therapy, laser treatment (transpupillary thermotherapy), photodynamic therapy and immunotherapy. These can be given alone or in combination.

Surgery

Surgery for ocular melanoma may involve removing just the tumour, removing part of the eye, or removing the entire eye (enucleation) if it has been severely damaged by the tumour. These operations are done while you are under a general anaesthetic and you will have to stay in hospital for one or two days.

› For a free copy of Cancer Council's booklet *Understanding Surgery and Research* visit your local Cancer Council website or call 13 11 20.

Surgical procedures for ocular melanoma	
Iridectomy	Removal of part of the iris (coloured part of the eye)
Iridocyclectomy	Removal of part of the iris and the ciliary body
Endoresection or transscleral resection	Removal of only the tumour in the ciliary body or choroid
Enucleation	Removal of the entire eye. This is performed for larger melanomas or if the vision in the eye has already been lost. An artificial eye matching your eye size and colour will usually be inserted after surgery to replace the eye
Orbital exenteration	Removal of the eye and some surrounding tissue

Radiation therapy

Radiation therapy (also known as radiotherapy) uses high energy rays to destroy cancer cells. It may be used for ocular melanoma:

- 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 (e.g. palliative radiation to control symptoms).

For ocular melanoma it is given the following ways:

- **Plaque radiation therapy (plaque brachytherapy) –** small seeds of radioactive material are placed in a small disc (called a plaque) and attached to the wall of the eye over the tumour during an operation. Radiation is then delivered to the tumour. The plaque is left in place until the right amount of radiation has been given. This is usually about four to five days and you will have to stay in hospital during this time. After this, you will have another short operation to remove the plaque.

- **Proton beam radiation therapy –** proton beams are aimed directly at the tumour and may cause less damage to the other tissues they pass through. Treatment is given in high doses over several days. This treatment is currently not available in Australia but check with your radiation oncologist.

- **Stereotactic radiation therapy –** multiple small beams of radiation are used to precisely target the tumour in high doses. You usually need five sessions given over ten days.

A course of stereotactic radiation therapy needs to be carefully planned. During your first appointment you will meet with a radiation oncologist. At this planning session you will lie on an examination table and have a CT scan in the same position you will be placed in for treatment. Specific equipment, such as a frame to immobilise your head and a light to focus your gaze on, will be used to ensure your eye does not move during treatment. The information from this session will be used by your specialist to work out the treatment area, the type of radiation and how to deliver the right dose. Radiation therapists will then deliver the course of radiation therapy as set out in the treatment plan.

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Radiation therapy does not hurt and is usually given over a period of time to minimise side effects.

If you need plaque brachytherapy the ophthalmologist will plan this treatment with you.

› For a free copy of Cancer Council's booklet *Understanding Radiation Therapy* visit your local Cancer Council website or call 13 11 20.

Other treatments

Other types of treatment for ocular melanoma are:

Laser treatment (transpupillary thermotherapy) or photodynamic therapy

This treatment uses an infrared laser to heat and destroy cancer cells. It 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

If the ocular melanoma has spread (metastasised) to other parts of the body, immunotherapy may be considered. This treatment has been very helpful in treating metastatic skin melanoma and uses drugs to stimulate your own immune system to recognise and attack cancer cells.

Clinical trials

Your doctor may suggest you take part in a clinical trial. Doctors run clinical trials to 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 will 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 with cancer. Talk to your specialist, clinical trials nurse or GP, or ask for a second opinion. If you decide to take part in a clinical trial, you can withdraw at any time.

For more information on clinical trials, visit australiancancertrials.gov.au.

› For a free copy of Cancer Council's booklet *Understanding Clinical Trials and Research* visit your local Cancer Council website or call 13 11 20.

Complementary therapies

Complementary therapies are designed to be used alongside conventional medical treatments (such as surgery and radiation therapy) and can increase your sense of control, decrease stress and anxiety, and improve your mood. Some Australian cancer centres have developed "integrative oncology" services where evidence-based complementary therapies are combined with conventional treatments to improve both wellbeing and clinical outcomes. Let your doctor know about any therapies you are using or thinking about trying, as some may not be safe or evidence-based.

› For a free copy of Cancer Council's booklet *Understanding Complementary Therapies* visit your local Cancer Council website or call 13 11 20.

Complementary therapy	Clinically proven benefits
acupuncture	reduces chemotherapy-induced nausea and vomiting; improves quality of life
aromatherapy	improves sleep and quality of life
art therapy, music therapy	reduce anxiety and stress; manage fatigue; aid expression of feelings
counselling, support groups	help reduce distress, anxiety and depression; improve quality of life
hypnotherapy	reduces pain, anxiety, nausea and vomiting
massage	improves quality of life; reduces anxiety, depression, pain and nausea
meditation, relaxation, mindfulness	reduce stress and anxiety; improve coping and quality of life
qi gong	reduces anxiety and fatigue; improves quality of life
spiritual practices	help reduce stress; instil peace; improve ability to manage challenges
tai chi	reduces anxiety and stress; improves strength, flexibility and quality of life
yoga	reduces anxiety and stress; improves general wellbeing and quality of life

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Nutrition and exercise

If you have been diagnosed with ocular melanoma, both the cancer and treatment will place extra demands on your body. Research suggests that eating well and exercising can benefit people during and after cancer treatment.

Eating well and being physically active can help you cope with some of the common side effects of cancer treatment, speed up recovery and improve quality of life by giving you more energy, keeping your muscles strong, helping you maintain a healthy weight and boosting your mood. You can discuss individual nutrition and exercise plans with health professionals such as dietitians, exercise physiologists and physiotherapists.

- For free copies of Cancer Council's booklets *Nutrition and Cancer* and *Exercise for People Living with Cancer* visit your local Cancer Council website or call 13 11 20.

Side effects of treatment

All treatments can have side effects. The type of side effects that you may have will depend on the type of treatment you have. Some people have very few side effects and others have more. 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), with you before your treatment begins.

Common side effects	
Surgery	Loss of vision, damage to nearby tissue, pain, bleeding, blood clots, infection after surgery, change in appearance
Radiation therapy	Blurry vision, dry eye, cataracts, glaucoma, loss of vision, eye discomfort, fatigue
Laser therapy	Loss of vision, eye discomfort, bleeding inside the eye
Immunotherapy	Infection, fatigue, skin reactions, headaches, inflammation of the heart, inflammation of the colon, inflammation of the liver, kidney problems

Making decisions about treatment

Making treatment decisions when you have been diagnosed with cancer can be overwhelming. Sometimes, it can be difficult to know which treatment is best for you. Take your time in talking with your doctors. Understanding the disease, the available treatments, possible side effects and any extra costs can help you weigh up the options and make well-informed decisions.

Some people prefer to seek several opinions before feeling confident to go ahead with the treatment. There is no need to feel awkward about seeking a second opinion and specialists will not be offended if you do so. If you are confused or want to check anything, ask your specialist questions. This will ensure you have all the information you need to make decisions about treatment and your future that you are comfortable with.

Check with your doctors how soon treatment should begin. Sometimes it won't affect the success of the treatment to wait a while. Ask them to explain your options and take as much time as you can before making a decision.

You may have to attend many appointments. It's difficult to remember everything your specialist is saying to you so it's a good idea to take someone with you. They will be able to listen, ask questions and remember what the doctor says.

It may help to take a list of questions with you, take notes (especially about anything you are unfamiliar with) or ask the doctor if you can record the discussion (many mobile phones have a recording function or you can use the CAN.recall app – visit rarecancers.org.au for more information). There are some suggestions for questions you could ask at the end of this sheet.

- For a free copy of Cancer Council's booklet *Cancer Care and Your Rights* visit your local Cancer Council website or call 13 11 20. Cancer Council's podcast on *Making Treatment Decisions* can be downloaded from your local Cancer Council website.

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Looking after yourself

There is no right way to feel if you have been diagnosed with a cancer such as ocular melanoma. Feeling a range of emotions is normal and you may feel overwhelmed, anxious, fearful, sad or lonely. Many people need emotional support before, during and after treatment. Adjusting to living with visible scars, changes to your physical appearance, changes to your lifestyle and bodily function can be hard and take time. If this is affecting you, or likely to affect you, it's important to seek help.

It can help to talk things through with a counsellor, psychologist, friend or family member. Talk to your medical team or call Cancer Council 13 11 20 about what support services are available.

- For a free copy of Cancer Council's booklet *Emotions and Cancer* visit your local Cancer Council website or call 13 11 20.

Practical and financial support

There are many services that can help you manage with practical or financial issues caused by having ocular melanoma. Benefits, pensions and hardship programs may be able to help pay for prescription medicines (for example the Australian Government's Pharmaceutical Benefits Scheme), transport costs or utility bills. Ask the hospital social worker which services are available and if you are eligible for them.

For additional income, you may be able to access your superannuation early or claim on insurance policies such as income protection, trauma, and total and permanent disability. Managing your ability to work or study during cancer treatment, is important to consider and will depend on your personal situation.

- For free copies of Cancer Council's booklets *Cancer and Your Finances* and *Cancer, Work & You* visit your local Cancer Council website or call 13 11 20.

If you need legal or financial advice, you should talk to a qualified professional about your situation. Cancer Council offers free legal and financial services in some states and territories for people who can't afford to pay – call **13 11 20** to ask if you are eligible.

Life after treatment

Once your treatment has finished, you will have regular check-ups to confirm that the cancer hasn't come back. Ongoing surveillance for ocular melanoma involves a schedule of tests and scans, eye tests and physical examinations. Let your doctor know of any health problems between visits.

Some cancer centres work with patients to develop a "survivorship care plan" which includes a summary of your treatment, sets out a schedule for follow-up care, lists any symptoms and long-term side effects to watch out for, identifies any medical or emotional problems that may develop and suggests ways to adopt a healthy lifestyle. Maintaining a healthy body weight, eating well and being active are all important. If you don't have a care plan, ask your specialist for one and make sure a copy is given to your GP and other health care providers.

- For a free copy of Cancer Council's booklet *Living Well After Cancer* visit your local Cancer Council website or call 13 11 20.

If the cancer comes back

For some people ocular melanoma does come back after treatment, which is known as a recurrence. If the cancer does come back, treatment will depend on where the cancer has returned in your body and may include a mix of surgery, radiation therapy, laser and immunotherapy. Enrolling in a clinical trial may also be recommended for you. In some cases of advanced cancer, treatment will focus on managing any symptoms, such as pain, and improving your quality of life without trying to cure the disease. This is called palliative treatment. Palliative care can be provided in the home, in a hospital, in a palliative care unit or hospice, or in a residential aged care facility.

When cancer is no longer responding to active treatment, it can be difficult to think about how and where you want to be cared for towards the end of life. But it's essential to talk about what you want with your family and health professionals, so they know what is important to you. Your palliative care team can support you in having these conversations.

- For free copies of Cancer Council's booklets *Understanding Palliative Care, Living with Advanced Cancer* and *Facing End of Life* visit your local Cancer Council website or call 13 11 20.

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Questions for your doctor

Asking your doctor questions will help you make an informed choice. You may want to include some of the questions below in your list.

- What type of ocular melanoma do I have?
- Has the cancer spread? What stage of cancer do I have?
- Will I lose my eyesight?
- 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 treatment take?
- Is this treatment covered by Medicare/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 you are thinking about taking part in a clinical trial, here are some questions you could ask:

- What are the possible benefits and risks to me?
- What is being tested and why?
- How many people will be involved in this research?

Reference

1. ME Aronow, AK Topham, AD Singh: Uveal Melanoma: 5-Year Update on Incidence, Treatment, and Survival (SEER 1973-2013). Ocul Oncol Pathol. 2018;4:145–151.

A web-based resource for Australians with less common cancers project is a Cancer Australia Supporting people with cancer Grant initiative, funded by the Australian Government. Website: canceraustralia.gov.au

Acknowledgements

This information is based on clinical practice guidelines and has been developed with help from health professionals and people affected by ocular melanoma. It was reviewed by: A/Prof William Glasson, Ophthalmologist, Queensland Ocular Oncology Service, Queensland; Dr Lindsay McGrath, Ophthalmic Surgeon, Queensland Ocular Oncology Service, Queensland; Caitriona Nienaber, 13 11 20 Consultant, Cancer Council WA; Jane Palmer, Senior Oncology Nurse and Researcher, Oncogenomics Laboratory, QIMR Berghofer Medical Research Institute, Queensland Ocular Oncology Service, Queensland; Katrina Potter, Consumer; Susan Vine, OcuMel Australia; Ann Marie Weber, Consumer; Dr Wenchang Wong, Senior Radiation Oncologist, Prince of Wales Hospital, Sydney, Conjoint Senior Lecturer, University of NSW

Support services

- Australasian Ocular Melanoma Alliance (AOMA): offering support to patients with ocular melanoma across Australia and New Zealand. Visit **aoma.org.au**
- Cancer Council: visit your local Cancer Council website (see below for details) or call **13 11 20**
- Melanoma Institute Australia: **melanoma.org.au** or call **(02) 9911 7200**
- Ocular Melanoma Awareness Support Group Australia: Facebook support group.
- Rare Cancers Australia: **rarecancers.org.au** or call **1800 257 600**
- Smart Patients Ocular Melanoma Support Group: **smartpatients.com/forums/ocular-melanoma**
- Talk to a nurse, social worker or Cancer Council **13 11 20** about what is available in your area.

Where to get help and information

Call Cancer Council **13 11 20** for more information about ocular melanoma. Trained health professionals can listen to your concerns, provide information and put you in touch with local services and support groups. Ask for free copies of booklets that may be relevant to you, or download digital versions from 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..... **cancertas.org.au**
VIC..... **cancervic.org.au**
WA..... **cancerwa.asn.au**
Australia..... **cancer.org.au**

Cancer Council 13 11 20

For information and support on cancer-related issues, call Cancer Council **13 11 20**. This is a confidential service.