

Understanding Adenoid Cystic Carcinoma (ACC)

A guide for people affected by cancer

This fact sheet has been prepared to help you understand more about adenoid cystic carcinoma (ACC).

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 ACC 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 ACC. 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.

What is ACC?

Adenoid cystic carcinoma (ACC) is a rare type of cancer that forms in glandular tissues most commonly in the head and neck, but it can also begin in other areas.

ACC often starts in the salivary glands in the neck, mouth or throat, which make saliva. Saliva keeps the mouth moist, helps you swallow food and protects the mouth against infections.

There are three pairs of major salivary glands and hundreds of minor salivary glands throughout the lining of the mouth and throat. The major salivary glands include:

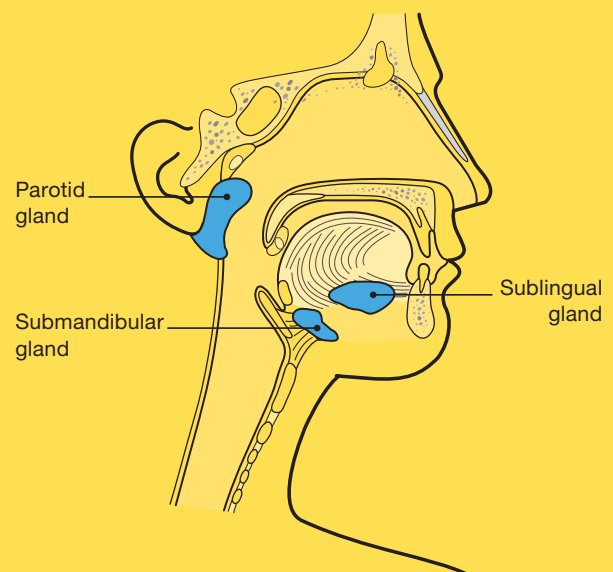
- **parotid glands** – in front of the ears
- **sublingual glands** – under the tongue
- **submandibular glands** – under the jawbone.

Malignant (cancerous) 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 new tumour is known as secondary cancer or metastasis.

More information on salivary gland cancer is available from Beyond Five: beyondfive.org.au and Rare Cancers Australia: rarecancers.org.au.

- ▶ For a free copy of Cancer Council's booklet *Understanding Head and Neck Cancers* visit your local Cancer Council website or call 13 11 20.

Salivary glands



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How common is ACC?

ACC is rare and may account for up to a quarter of salivary gland malignancies. Around 330 Australians are diagnosed with a salivary gland cancer each year (this is 1.2 cases per 100,000 people).¹

Estimates of 0.3 to 0.5 cases per 100,000 people each year have been reported for ACC in other countries.

While ACC can develop at any age, it is more common in people 40 to 60 years old. Slightly more females are diagnosed than males.

What are the risk factors?

The cause of ACC is not known. It might develop from genetic changes that happen during a person's lifetime, rather than inheriting a faulty gene. ACC is not considered to be hereditary.

What are the symptoms?

ACC generally develops slowly, sometimes over several years, and may not cause symptoms. Most often it is diagnosed as a single tumour but may have spread to nearby lymph nodes by the time it is diagnosed in a small number of cases. Rarely, ACC can spread along nerves or metastasise to other parts of the body (usually the lungs, liver or bone) and cause problems. It can also be unpredictable, growing slowly for a period of time and then suddenly growing quickly. ACC may behave differently in different people.

If you do have symptoms it will depend on where in the body the tumour is located and its size.

Symptoms may include:

- **Salivary gland (produces saliva)** – painless lump in the mouth, face or neck; numbness in the face; weakness in facial muscles (drooping in the face); problems swallowing or opening mouth
- **Lacrimal gland (produces tears)** – bulging eye; changes in vision
- **Larynx (voice box) and trachea (windpipe)** – hoarseness; changes in speech; difficulty breathing
- **Skin** – pain; increased sensitivity; pus and/or blood discharge
- **Breast** – slow growing lump that may be tender or cause pain.

Diagnosis

If your doctor thinks that you may have ACC, they will take your medical history, perform a physical examination (including feeling for any lumps) and carry out certain tests depending on the location of the suspected tumour. If the results suggest that you may have ACC, your doctor will refer you to a specialist for more tests. These can include:

- **CT (computerised tomography) and/or MRI (magnetic resonance imaging) scans** – special machines are used to scan and create pictures of the inside of your body. 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 lie on an examination table. For a CT scan, the table moves in and out of the scanner which is round; the scan 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 about 30–90 minutes. Both scans are painless.
- **PET (positron emission tomography) scan** – before the scan you will be injected with a small amount of radioactive glucose (sugar) solution. Many cancer cells will show up brighter on the scan. You will be asked to sit for 30–90 minutes to allow the glucose to move around your body. The scan itself will take around 30 minutes to perform.
- **Ultrasound scan** – soundwaves are used to create pictures of the inside of your body. For this scan, you will lie down and a gel will be spread over the affected part of your body. A small device called a transducer is moved over the area. It sends out soundwaves that echo when they hit something dense, like an organ or tumour. The images are then projected onto a computer screen. The ultrasound is painless and takes about 15 minutes.
- **Biopsy** – removal of some tissue from the affected area for examination under a microscope. The biopsy may be done in different ways. In a core needle biopsy, a local anaesthetic is used to numb the area, then a thin needle is inserted into the tumour under ultrasound or CT guidance. A fine needle aspiration (FNA) may also be used, which collects cells using a smaller needle and usually doesn't require an anaesthetic. A surgical biopsy is done under general anaesthesia. The surgeon will cut through the skin and take a tissue sample.

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Finding a specialist

The Australian and New Zealand Head and Neck Cancer Society can be contacted for a directory of specialist teams in ACC care and treatment: anzhnsc.org/mdt

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

The Adenoid Cystic Carcinoma Research Foundation (ACCRF) have a global list of specialists in ACC treatment, including those in Australia: accrf.org

Treatment

You will be cared for by a multi-disciplinary team of health professionals during your treatment for ACC. The team may include a surgeon, radiation oncologist (to prescribe and coordinate a course of radiation therapy), medical oncologist (to prescribe and coordinate a course of systemic therapy including chemotherapy), nurse and allied health professionals such as a speech pathologist, dietitian, social worker, psychologist or counsellor and occupational therapist.

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

- where it is in your body
- whether or not the cancer has spread (stage of disease)
- your age, fitness and general health
- your preferences.

The main treatments include surgery, radiation therapy and chemotherapy. These can be given alone or in combination. Surgical removal of the cancer with follow-up radiation is the standard treatment for ACC where there is a primary tumour.

Surgery

Surgery is usually the most effective treatment for ACC if the cancer can be safely removed. Surgery usually involves removing the cancer and some healthy tissue around the cancer. This is called a wide local excision. The healthy tissue is removed to help reduce the risk of the cancer coming back in that area.

The extent of the operation depends on where the cancer is and how far it has spread. The surgeon will examine nearby nerves and lymph nodes and may remove them if they are involved. It can be a difficult decision to remove nerves, especially major nerves or those that control the face. It is important that your surgery is carried out by a surgeon with special expertise in this area.

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

Radiation therapy

Radiation therapy (also known as radiotherapy) uses high energy rays to destroy cancer cells. It may be used effectively for ACC after surgery, to destroy any remaining cancer cells and stop the cancer coming back. It might also be used alone if surgery is not possible, for example:

- if the cancer is in a place in the body that is too hard to reach using surgery
- if the cancer has spread to other parts of the body (e.g. palliative radiation for the management of pain).

A course of radiation therapy needs to be carefully planned. During your first consultation session you will meet with a radiation oncologist. At this session you will lie on an examination table and have a CT scan in the same position you will be placed in for treatment. The information from this session will be used by your specialist to work out the treatment area and how to deliver the right dose of radiation. Radiation therapists will then deliver the course of radiation therapy as set out in the treatment plan.

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

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

For an overview of what to expect during all stages of your cancer care, visit cancerpathways.org.au/optimal-care-pathways/head-and-neck-cancer. Also, visit targetingcancer.com.au/useful-resources/julies-story-videos for a patient's treatment journey for head and neck cancer.



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Chemotherapy

Chemotherapy (sometimes just called “chemo”) is the use of drugs to kill or slow the growth of cancer cells. You may have one chemotherapy drug, or a combination of drugs. This is because different drugs can destroy or shrink cancer cells in different ways.

Chemotherapy is not commonly used for AC. It may be considered when the cancer is advanced as part of palliative treatment or as part of a clinical trial. Your medical oncologist will discuss options with you.

Chemotherapy is given through a drip into a vein (intravenously) or as a tablet that is swallowed. It is given in treatment cycles which may be daily, weekly or monthly. For example, one cycle may last three weeks where you have the drug over a few hours, followed by a rest period before starting another cycle. The length of the cycle and number of cycles depends on the chemotherapy drugs being given.

► For a free copy of Cancer Council’s booklet *Understanding Chemotherapy* visit your local Cancer Council website or call 13 11 20.

Clinical trials

Your doctor or nurse 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 diagnosed with cancer.

You may find it helpful to talk to your specialist, GP, or clinical trials nurse. If you decide to take part in a clinical trial, you can withdraw at any time.

Visit australiancancertrials.gov.au for information or contact the Australian and New Zealand Head and Neck Cancer Society who have a special fund to support research into ACC: anzhnscs.org. Cancer Council’s booklet *Understanding Clinical Trials and Research* includes a list of Australian clinical trials websites and has information on types of clinical trials and how to join a study. For a free copy 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, radiation therapy and chemotherapy) 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.

► 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	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 stress; improves strength and flexibility
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 ACC, 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 on *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 and where in your body the cancer is. Some people have very few side effects while others can 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.

After surgery or radiation therapy to the head and neck area, you may need to adjust to significant changes. Everyone will respond differently – talk to your doctor about what to expect and try to see a speech pathologist and/or dietitian before treatment starts. Some long-term side effects of treatment to this area include:

- mouth problems, including dry mouth
 - heartburn or indigestion
 - taste and smell changes
 - swallowing difficulties
 - speech or voice changes
 - breathing changes
 - appearance changes.
- For a free copy of Cancer Council's booklet *Understanding Head and Neck Cancers* visit your local Cancer Council website or call 13 11 20.

Common side effects

Surgery	Bleeding, damage to nearby tissue and organs (including nerves), drug reactions, pain, infection after surgery, blood clots, weak muscles (atrophy), lymphoedema
Radiation therapy	Fatigue, loss of appetite, nausea, hair loss, dry mouth, skin problems, lymphoedema
Chemotherapy	Fatigue, loss of appetite, nausea, bowel issues such as constipation or diarrhoea, mouth sores, skin and nail problems, increased chance of infections, loss of fertility, early menopause

Making decisions about treatment

It can be difficult to know which treatment is best for you. It is important that you speak with a specialist team before making your decision. Ask them to give you a clear plan of your treatment options, including written information about side effects. Some people prefer to seek several opinions before feeling confident to go ahead with the treatment. If you are confused or want to check anything, ask your specialist questions. This will make sure you have all the information you need to make decisions about treatment and your future that you are comfortable with.

You may have to attend many appointments. 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 *Care and Your Rights* visit your local Cancer Council website or call 13 11 20. Cancer Council's podcast *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 ACC. Feeling a range of emotions is normal and you may feel overwhelmed, anxious, fearful, angry, 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 ACC. Benefits, pensions and hardship programs can help pay for prescription medicines (for example the Australian Government's Pharmaceutical Benefits Scheme [PBS]), transport costs or utility bills. Ask the hospital social worker which services are available in your local area and if you are eligible to receive them.

For additional income, you may be able to access your superannuation early in some circumstances, or claim on insurance policies such as income protection, trauma, and total and permanent disability. Managing your ability to work or study, particularly 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.

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 ACC involves a schedule of ongoing blood tests and scans. Let your doctor know immediately 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 ACC does come back after treatment, which is known as a recurrence. It may also spread to other parts of the body after a couple of years. 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 and chemotherapy. In some cases of advanced cancer, treatment will focus on managing 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 on *Understanding Palliative Care*, *Living with Advanced Cancer* and *Facing End of Life* 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.

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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.

- How far has the cancer spread? What stage of cancer do I have?
- Have you treated this type of cancer before? If so, how many times? Is there another doctor who has more experience treating this type of cancer?
- What treatment do you recommend and why?
- What are the possible risks and side effects of my treatment? How will these be managed?
- How long will treatment take? Will I have to stay in hospital?
- Is this treatment covered by Medicare/private insurance? Will there be extra expenses?
- Will I have a lot of pain with the treatment? What will be done about this?
- Will my face or neck have significant scarring or will I look different? What can be done to minimise this?
- What kind of rehabilitation will be available to me afterwards?
- Are there any complementary therapies that might help me?

If you are thinking of 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 trial?

Reference

1. Australian Institute of Health and Welfare (AIHW). Cancer in Australia 2019. Cancer series no.119. Cat. no. CAN 123. AIHW, Canberra, 2019.

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

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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.

Support services

Australia

- Cancer Council: visit your local Cancer Council website (see below for details) or call **13 11 20**
- Head & Neck Cancer Australia: headandneckcancer.org.au or call **1300 424 848**
- Australian & New Zealand Head & Neck Cancer Society: anzhnccs.org or call **02 9431 8670**
- Rare Cancers Australia: rarecancers.org.au or call **1800 257 600**
- Without a Ribbon – Fighting for rare cancer support: withoutaribbon.org

International

- Adenoid Cystic Carcinoma Organization International: accoi.org
- Adenoid Cystic Carcinoma Research Foundation: accrf.org
- Salivary Gland Cancer UK: salivaryglandcancer.uk

Where to get help and information

Call Cancer Council **13 11 20** for more information about ACC. 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



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