Cancer Council

CANCER TYPE
fact sheet

Understanding
Primary Bone Cancer

A guide for people affected by cancer

This fact sheet has been prepared to help you understand more about primary bone cancer, also known as bone sarcoma. In this fact sheet we include general information about how bone cancer is diagnosed and treated.[1]

The bones

A typical healthy person has over 200 bones, which:

- support and protect internal organs
- are attached to muscles to allow movement
- contain bone marrow, which makes and stores new blood cells
- store proteins, minerals and nutrients, such as calcium.

What is bone cancer?

Bone cancer can start as a primary or secondary cancer. The 2 types are different and this fact sheet is only about primary bone cancer. We have a separate fact sheet on secondary bone cancer.

Primary bone cancer – This means that the cancer starts in a bone. It may develop on the surface, in the outer layer or from the centre of the bone. As a tumour grows, cancer cells multiply and destroy the bone. If left untreated, primary bone cancer can spread to other parts of the body.

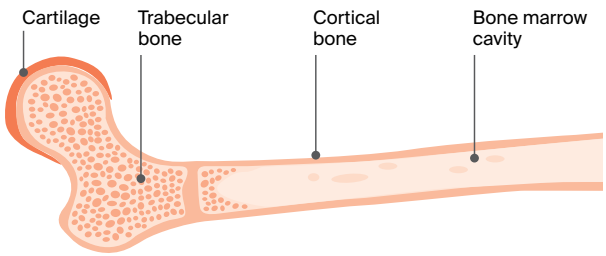
Secondary (metastatic) bone cancer – This means that the cancer started in another part of the body (e.g. breast, lung) and has spread to the bone.

How common is bone cancer?

Primary bone cancer is rare. About 300 Australians are diagnosed with primary bone cancer each year.[2] It affects people of all ages, but most often occurs in people aged 10–25 and over 50.

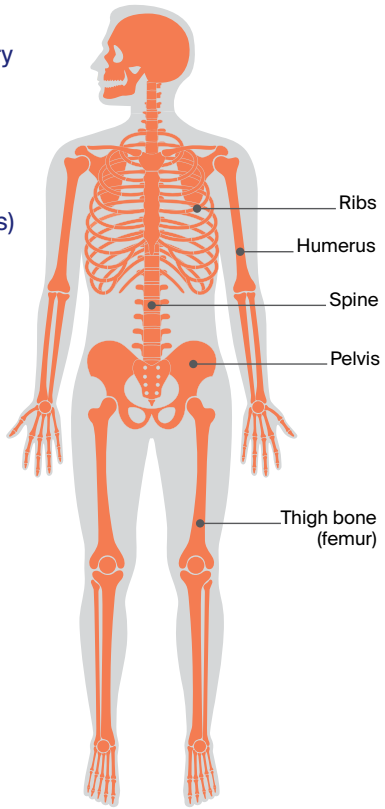
Bone structure

Bones are made up of different parts, including a hard outer layer (known as cortical bone) and an inner core (known as trabecular bone). The bone marrow is found in this inner core. Cartilage is the tough material at the end of each bone that allows one bone to move against another at a joint.



Bones of the body

Bones commonly affected by primary bone cancer include the spine, ribs, pelvis and upper bones of the arms (humerus) and legs (femur).



Types of primary bone cancer

There are more than 30 types of primary bone cancer. The most common types are:

osteosarcoma (about 35% of bone cancers)	• starts in cells that grow bone tissue • often affects the arms, legs or pelvis, but may occur in any bone • occurs in children and young adults with growing bones, and older people in their 70s and 80s • most are high-grade tumours
chondrosarcoma (about 30% of bone cancers)	• starts in cells that grow cartilage • often affects the bones in the upper arms and legs, pelvis, ribs or shoulder blades • most often occurs in adults aged over 40 • slow growing; rare types can spread to other parts of the body • most are low-grade tumours
Ewing sarcoma (about 15% of bone cancers)	• affects cells in the bone or soft tissue that multiply rapidly • often affects the pelvis, legs, ribs, spine or upper arms • most common in children and young adults • are high-grade tumours

Cancer that affects the soft tissues around the bones is known as soft tissue sarcoma, and may be treated differently. For an overview of what to expect at every stage of your care for bone and soft tissue sarcomas, visit cancer.org.au/cancercareguides/sarcoma. This is a short guide to what is recommended, from diagnosis to treatment and beyond.

What are the risk factors?

The causes of most bone cancers are unknown, but factors that may increase the risk include:

Previous radiation exposure including radiation therapy (radiotherapy) – Radiation therapy to treat cancer increases the risk of developing bone cancer. The risk is higher when high doses of radiation therapy have been given in childhood. Most people who have radiation therapy will not develop bone cancer.

Chemotherapy for another cancer – Some drugs may increase the risk of osteosarcoma.

Other bone conditions – Some people who have Paget disease of the bone, or other benign bone conditions, are at higher risk.

Genetic factors – Some inherited conditions, such as Li-Fraumeni syndrome, increase the risk of bone cancer. People with a strong family history of certain types of cancer are also at risk. Ask for a referral to a family cancer clinic for more information. Some people develop bone cancer because of gene changes that happen during their lifetime, rather than inheriting a faulty gene.

What are the symptoms?

The most common symptom is pain in the affected bone or joint that doesn't improve with mild pain medicines such as paracetamol. You might have this pain most of the time, and it may be worse at night or during activity.

Other symptoms can include:

- swelling over the affected part of the bone
- stiffness or tenderness in the bone
- problems moving around, e.g. walking with a limp
- loss of feeling in the affected arm or leg (limb)
- bone that breaks for no reason.

These symptoms do not mean you have primary bone cancer. If you have symptoms for more than 2 weeks, you should see your general practitioner (GP).

Diagnosis

If you have symptoms, your doctor will ask about your medical history and do a physical examination. It is likely that you will have some of the following tests:

- **x-rays** – can show bone damage or whether new bone is growing
- **blood tests** – help check your overall health
- **CT and/or MRI scans** – create pictures to highlight any bone abnormality
- **specialised scans** – a small amount of radioactive solution is injected before a scan to highlight any cancerous areas in the body, e.g. PET, thallium or technetium scans

- **bone biopsy** – collects some cells and tissues from the outer part of the affected bone. This is best done in a specialised sarcoma treatment centre. The biopsy may be done in one of two ways. In a core biopsy, a local anaesthetic is used to numb the area, then a sample is taken using a needle. A CT or ultrasound scan is used to guide the needle into place. In an open biopsy, under general anaesthesia the surgeon makes a cut in the skin to remove a piece of bone. The sample is also checked for genetic changes
- **bone marrow biopsy** – a thin needle is used to remove a sample of marrow from the hip bone.

Staging

The test results will help show where the cancer is and if it has spread. This is called staging. Knowing the stage helps your doctors plan your treatment.

Grades of primary bone cancer	
Grading describes how quickly a cancer might grow. In general, the lower the grade, the better the prognosis.	
low grade	The cancer cells look like normal cells. They usually grow slowly and are less likely to spread.
high grade	The cancer cells look very abnormal. They grow quickly and are more likely to spread.
Stages of primary bone cancer	
Many cancers are staged using a system that divides them into 4 stages. But bone cancer is different. It is usually divided into localised or advanced. Ask your doctor to explain the stage of cancer to you.	
localised	The cancer contains low-grade cells; found in the bone in which it started. It can be removed by surgery (resectable) or not removed completely (non-resectable).
advanced (metastatic)	The cancer is any grade and has spread to other parts of the body (e.g. the lungs).

Selecting the bone site to biopsy

A bone biopsy is a specialised test. It is best to have the biopsy at the specialist treatment centre (see below) where you would be treated if it is cancer. The specialists will usually work together to decide the best site to place the needle. The site to biopsy must be carefully chosen so it doesn't cause problems if further surgery is needed. This also helps ensure the sample is useful and reduces the risk of the cancer spreading.

Treatment

The treatment of bone cancer is complex and requires specialist care. Research shows that having treatment at a specialist treatment centre (see below) means better recovery and longer survival. Treatment will depend on:

- the type of primary bone cancer
- the location and size of the tumour
- whether or not the cancer has spread
- your age, fitness and general health.

Treatment for primary bone cancer usually involves one or more treatments, including surgery, chemotherapy and radiation therapy. The aim is to control the cancer and maintain the use of the affected area of the body. Many people who are treated for bone cancer go into complete remission (when there is no evidence of active cancer).

Specialist sarcoma treatment centres

You can find specialised sarcoma treatment centres at certain hospitals and cancer centres in major cities throughout Australia.

These specialist centres have multidisciplinary teams (MDTs) who regularly manage this cancer. The team will include surgeons, medical oncologists, radiation oncologists, pathologists, radiologists and clinical nurse consultants. It will also include allied health professionals such as physiotherapists, occupational therapists and social workers. Some centres also have oncologists with experience in treating children and young people with bone cancer.

To find a specialised sarcoma team in your state or territory, visit sarcoma.org.au. You might have to travel for treatment.

Preparing for treatment

Ask about fertility

Treatment may affect your ability to conceive a child (fertility). Before treatment starts, you may be able to store sperm, eggs, embryos, or ovarian tissue.

▸ See our *Fertility and Cancer* booklet.

Avoiding fractures

If your doctor thinks you may be at risk of a bone fracture, they may recommend you wear a splint to support the bone or use crutches.

Checking heart and kidneys

Your doctor may recommend you have some tests to check how well your heart and kidneys are working, as some types of chemotherapy and radiation therapy may affect these organs.

Surgery

The type of operation you have will depend on where the cancer is in the body.

Limb-sparing surgery

Surgery to remove the cancer but save (salvage) the arm or leg (limb) can be done in most people. You will have a general anaesthetic, and the surgeon will remove the affected part of the bone. The surgeon will also take out some surrounding normal-looking bone and muscle with a layer of surrounding normal tissue. This is called a wide local excision, and it reduces the chance of the cancer coming back. A pathologist checks the tissue to see whether the edges are clear of cancer cells.

The bone that is removed is usually replaced with a metal implant or a bone graft. A graft uses healthy bone from another part of your body or from a "bone bank". A bone bank is a facility that collects tissue for research and surgery. In some cases, the removed bone is treated with radiation therapy to destroy the cancer cells, then used to reconstruct the limb.

After surgery, you will be given medicine to manage pain and reduce the chance of getting an infection in the bone or metal implant. There are likely to be some changes in the way the limb looks, feels or works.

A physiotherapist can show you exercises to help you regain strength and improve how the limb works.

Surgery to remove the limb (amputation)

In cases when it is not possible to remove the cancer without affecting the arm or leg too much, the limb is removed (amputation). For about 1 in 10 people, this is the only way to remove the cancer completely. This procedure is less common now because techniques used for limb-sparing surgery have improved.

After surgery, you will be given medicine to manage the pain and taught how to care for the stump that remains (residual limb). After the area has healed, you may be fitted for an artificial limb (prosthesis).

If you have an arm removed, an occupational therapist will teach you how to eat and dress yourself using one arm. If you receive a prosthetic arm, the occupational therapist will teach you exercises and techniques to control and use the prosthesis.

If you have a leg removed and receive a prosthesis, a physiotherapist will show you exercises and techniques to improve how you walk and move with your new limb. Some people choose to use a wheelchair instead of a prosthetic leg.

Surgery in other parts of the body

- **Pelvis** – When possible, the cancer is removed along with some healthy tissue around it (wide local excision). Some people may need to have a bone graft or a metal implant to rebuild the bone.
- **Jaw or cheek bone (mandible or maxilla)** – The surgeon will remove the affected bone. Bone from other parts of the body may be used to replace the affected bone. As the face is a delicate area, it may be difficult to remove the cancer with surgery and some people may need to have chemotherapy or radiation therapy (see page 5).
- **Spine or skull** – If surgery isn't possible, a combination of radiation therapy and chemotherapy may be used. If you need one of these treatments, your doctor will explain what will happen.

For more information, see our *Understanding Surgery*, *Understanding Chemotherapy* and *Understanding Radiation Therapy* booklets.

Chemotherapy

This treatment uses drugs to destroy or slow the growth of cancer cells, while causing the least possible damage to healthy cells. It may be given for high-grade osteosarcoma and Ewing sarcoma:

- before surgery, to shrink the size of the tumour and make it easier to remove
- after surgery or radiation therapy, to kill any cancer cells possibly left behind
- as palliative treatment, to help stop the growth of an advanced cancer or control the symptoms.

Chemotherapy drugs are often injected into a vein. This may be as a day patient, or during a hospital stay. You will need scans (MRI, CT or PET–CT) during treatment to see how well the cancer is responding to the chemotherapy.

Side effects – These depend on the drugs that are given and where the cancer is in your body. Common side effects include fatigue (tiredness), nausea, vomiting and diarrhoea, appetite loss, hair loss, constipation, numbness or tingling in the hands and feet, effects on hearing and increased risk of infection. Talk to your treatment team about ways to manage side effects. If your red blood cell count drops too low, you may need a transfusion to build them up again.

Radiation therapy

This treatment uses targeted radiation to kill or damage cancer cells. The radiation is usually in the form of x-ray beams. Radiation therapy may be used for Ewing sarcoma:

- after surgery or chemotherapy, to kill any cancer cells possibly left behind
- as an alternative treatment to surgery if a wide local excision is not possible
- as palliative treatment, to help stop the growth of an advanced cancer or control the symptoms.

Radiation therapy is usually given every weekday, with a rest over the weekend, for several weeks. Your specialist will provide details about your specific treatment plan.

Side effects – These will depend on the area being treated and the strength of the dose you have. Common side effects include fatigue (tiredness), skin redness or soreness, and hair loss in the treatment area. Ask your treatment team for advice about dealing with any side effects.

Coping with primary bone cancer

Being diagnosed with a rare cancer can be frightening. The physical changes after treatment for bone cancer can affect how you feel about yourself (self-esteem) and make you feel self-conscious. It will take time to get used to the differences in how you look and what you can do.

Limb-sparing surgery is a major operation that can leave a scar and make the skin feel tight. If you have an amputation or a lot of bone is removed, you may feel grief and loss. Many people find it helps to talk to a counsellor, psychologist, friend or family member. Ask your treating team or call Cancer Council 13 11 20 to find out about support services in your area.

▸ See our *Emotions and Cancer* booklet and *Understanding Rare and Less Common Cancers* booklet.

Follow-up appointments

After treatment, you will need regular check-ups for several years to confirm that the cancer hasn't come back and to help you manage any treatment side effects.

How often you will need to see your doctor will vary depending on the type of bone cancer. Check-ups will become less frequent if you have no further problems. Let your doctor know immediately of any health concerns between appointments.

Should I join a clinical trial?

Over the years, clinical trials have improved treatments and led to better outcomes for people diagnosed with cancer. Talk with your doctor about the latest clinical trials and whether you're a suitable candidate. For more information, visit australiancancertrials.gov.au.

▸ See our booklet on clinical trials and research.

If the cancer comes back

For some people, bone cancer does come back (recur) after treatment. The risk that bone cancer will recur is greater within the first 3 years after treatment has finished. Treatment options may include surgery, chemotherapy and radiation therapy.

In some cases of advanced primary bone cancer, treatment will focus on managing your symptoms and improving your quality of life without trying to cure the disease. Palliative treatment can relieve pain and help to manage other symptoms.

Question checklist

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

- What type of bone cancer do I have?
- What treatment do you recommend and why?
- How can I find a specialist treatment centre?
- What is the prognosis?
- If I have surgery, what are the side effects?
- Do I need an amputation?
- If I have to travel for treatment, is there any government funding available to help with the cost?
- Are there any clinical trials I could join?
- If the cancer has spread outside the bone, what treatment options are available for me?
- How often will I need check-ups after treatment?
- If the cancer comes back, how will I know?

Where to get help and information

Call Cancer Council 13 11 20 for more information about primary bone cancer. 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

You can find many useful resources online, but not all websites are reliable. These websites are good sources of support and information.

Australia and New Zealand Sarcoma Association	sarcoma.org.au
Canteen	canteen.org.au
Rare Cancers Australia	rarecancers.org.au

References

1. Cancer Council Australia Sarcoma Guidelines Working Party, *Clinical practice guidelines for the management of adult onset sarcoma*, Cancer Council Australia, Sydney, viewed 11 August 2022, available from wiki.cancer.org.au/ australia/Guidelines:Sarcoma.
2. Australian Institute of Health and Welfare (AIHW), *Cancer Data in Australia 2022*, AIHW, Canberra, viewed 1 August 2022, available from aihw.gov.au/ reports/cancer/cancer-data-in-Australia.

Acknowledgements

This information was reviewed by: Prof Peter Choong AO, Orthopaedic Surgeon, and Sir Hugh Devine Professor, St Vincent's Hospital, and Head of Department of Surgery, The University of Melbourne, VIC; Catherine Chapman, Adolescent and Young Adult and Sarcoma Cancer Specialist Nurse, Division of Cancer and Ambulatory Support, Canberra Hospital, ACT; A/Prof Paul Craft AM, Medical Oncologist, Canberra Hospital and Australian National University, ACT; Belinda Fowlie, Bone Tumour Nurse Practitioner Candidate, SA Bone and Soft Tissue Tumour Unit, Flinders Medical Centre, SA; Prof Angela Hong, Radiation Oncologist, Chris O'Brien Lifehouse, and Clinical Professor, The University of Sydney, NSW; Vicki Moss, Nurse Practitioner, SA Bone and Soft Tissue Tumour Unit, Flinders Medical Centre, SA; A/Prof and Dr Marianne Phillips, Paediatric and Adolescent Oncologist and Palliative Care Physician, Perth Children's Hospital, WA; Chris Sibthorpe, 13 11 20 Consultant, Cancer Council Queensland; Stephanie Webster, Consumer. We would also like to thank the health professionals and consumers who have worked on previous versions of this information.

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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