Cancer Council

Cancer information 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 use the term bone cancer, and include general information about how bone cancer is diagnosed and treated.

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 produces and stores new blood cells
- store proteins, minerals and nutrients, such as calcium.

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

What is bone cancer?

Bone cancer can develop as either a primary or secondary cancer. The two types are different and this fact sheet is only about primary bone cancer.

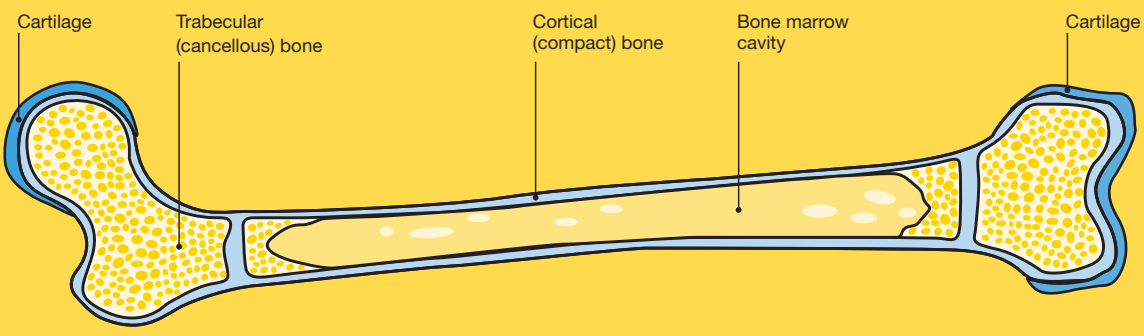
- **Primary bone cancer –** means that the cancer starts in a bone. It may develop on the surface, in the outer layer or from the centre of the bone. As a tumour grows, cancer cells multiply and destroy the bone. If left untreated, primary bone cancer can spread to other parts of the body.

- **Secondary (metastatic) bone cancer –** means that the cancer started in another part of the body (e.g. breast or lung) and has spread to the bones. See our fact sheet on secondary bone cancer.

How common is bone cancer?

Bone cancer is rare. About 250 Australians are diagnosed with primary bone cancer each year.[1] It affects people of all ages, but is most often seen in people aged 10–25 and over 50. If it develops later in life, it may be linked to another bone condition (see page 2).

Bone structure



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Types of 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 and 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 and shoulder blade • most often occurs in middle-aged and older people • slow-growing form of cancer that rarely spreads to other parts of the body • most are low-grade tumours
Ewing's sarcoma (about 15% of bone cancers)	• affects cells in the bone or soft tissue that multiply rapidly and is often associated with a large lump • often affects the pelvis, legs, ribs, spine, upper arms • common in children and young adults • all are high-grade tumours

Some types of cancer affect the soft tissues around the bones. These are known as soft tissue sarcomas, and may be treated differently. For more details, talk to your doctor or call Cancer Council **13 11 20**.

What are the risk factors?

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

Previous radiation therapy (radiotherapy) – Radiation therapy to treat cancer increases the risk of developing bone cancer. The risk is higher for people who have high doses of radiation therapy at a young age. Most people who have radiation therapy will not develop bone cancer.

Other bone conditions – Some people who have had Paget's disease of the bone, fibrous dysplasia or multiple enchondromas are at higher risk of developing bone cancer. Some studies also suggest that people who have had a soft tissue sarcoma have an increased risk of developing bone cancer.

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. Talk to a family cancer clinic for more information. Some people develop bone cancer due to genetic changes that happen during their lifetime, rather than inheriting a faulty gene. Most bone cancers are not hereditary.

What are the symptoms?

The most common symptom is strong pain in the affected bone or joint. The pain gradually becomes constant and doesn't improve with mild pain relievers such as paracetamol. 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 with moving around, for example, an unexplained limp
- loss of feeling in the affected limb
- unexplained fractured bone
- unexplained weight loss
- tiredness.

Most people who have these symptoms do not have bone cancer. If you have symptoms for more than two weeks, you should see your general practitioner (GP).

Diagnosis

If you have symptoms that could be caused by bone cancer, your doctor will take your medical history and perform a physical examination. Bone cancer can be difficult to diagnose, and it is likely that you will have some of the following tests:

- **x-rays –** can reveal bone damage or the creation of new bone
- **blood tests –** help check your overall health
- **CT or MRI scans –** a special computer is used to create pictures to highlight any bone abnormality

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- **PET scan –** you will be injected with a small amount of radioactive glucose solution to highlight any cancerous areas on the scan
- **bone biopsy –** removes some cells and tissues from the outer part of the affected bone for examination under a microscope. 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 thin needle is inserted into the bone under CT guidance to take a sample. In an open or surgical biopsy, the surgeon will cut through the skin under general anaesthesia to remove a piece of bone.

If Ewing's sarcoma is suspected, you will have a procedure to examine cells from the inner part of the affected bone. A thin needle is used to remove a sample of fluid (aspirate) from the bone marrow.

Staging

The test results will help show how far the cancer has spread in the body. This is called staging. Knowing the stage helps your health care team plan the most suitable treatment for you.

Grading bone cancer

Grading describes how quickly a cancer might grow.

low grade	The cancer cells look similar to normal cells. They are usually slow-growing and less likely to spread.
high grade	The cancer cells look very abnormal. They grow quickly and are more likely to spread.

Stages of bone cancer

There are different staging systems used for bone cancer. Ask your doctor to explain your stage to you.

stage 1 (localised)	The cancer contains low-grade cells; there is no spread beyond the bone.
stage 2 (localised)	The cancer contains high-grade cells; there is no spread beyond the bone.
stage 3 (localised)	There are several high-grade tumours in the same bone; there is no spread beyond the bone.
stage 4 (advanced)	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 highly specialised test. It is best that the biopsy is done at a specialist treatment centre (see below), preferably where you would be treated if it is cancer. The site to biopsy must be carefully chosen so it doesn't cause problems if further surgery is needed. It is important that a bone biopsy is performed by a doctor who is an expert in bone cancer. This also helps ensure the sample is useful and reduces the risk of the cancer spreading.

Treatment

The treatment of bone cancers is complex and requires specialist care. Treatment will depend on:

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

Treatment for primary bone cancer usually includes surgery, chemotherapy and radiation therapy, or a combination of these treatments. 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 remission (when the symptoms of bone cancer decrease or disappear).

Understanding your treatment options and possible side effects can help you weigh up the pros and cons of different treatments. You may want to get a second opinion from another specialist to confirm or clarify your first doctor's recommendations.

Specialist treatment centres

Located in major cities throughout Australia, these centres have expert multidisciplinary teams (MDTs) who regularly manage this cancer. The team will include surgeons, medical oncologists, radiation oncologists, pathologists, radiologists and clinical nurse consultants, as well as allied health professionals such as physiotherapists, occupational therapists and social workers. Some centres also have oncologists with experience in treating young people with bone cancer.

To find a specialist treatment centre, visit sarcoma.org.au.

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Preparing for treatment

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

Treatment may affect your fertility (your ability to conceive a child). If you are interested in having children in the future, discuss this with your doctor before treatment starts. You may be able to store sperm, eggs or embryos. For more information, read our *Fertility and Cancer* booklet.

Bones affected by cancer are weaker than normal bone. If your doctor suspects you are at risk of fracturing a bone because of the cancer, they may suggest you wear a splint to support the bone or use crutches.

Surgery

There are different types of operations depending on the location of the cancer.

Limb-sparing surgery

Surgery to remove the cancer but keep (spare) the limb is done in about nine out of 10 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 to remove as much of the cancer as possible, and to reduce the chance of the cancer coming back. This is called a wide local excision. A pathologist will check the tissue to see whether the edges are clear of cancer cells.

The bone that is removed is replaced with a metal implant (prosthesis) or a bone graft. A graft uses a piece of 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, it may be possible to treat the removed bone with radiation therapy to destroy the cancer cells, then use the treated bone to reconstruct the limb.

The risks of surgery will be explained to you. After surgery, you will be given medicine to manage any pain and reduce the chance of getting an infection in the bone or prosthesis. There will be some changes in the way the limb looks, feels or works.

A physiotherapist can plan an exercise program to help you regain strength and function in your limb.

Surgery to remove the limb (amputation)

In some cases when it is not possible to remove all of the cancer without affecting the arm or leg too much, the limb is removed (amputation). For about one in 10 people, this is the only effective treatment. This procedure has become less common as limb-sparing surgery has improved.

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

If you have a leg removed (amputated) and receive a prosthesis, a physiotherapist will teach you exercises and techniques to improve walking and other movement. Some people find using a prosthetic leg too difficult and prefer to use a wheelchair.

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 better control and use the prosthesis.

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 bone grafts to rebuild the bone.

- **Jaw or cheek bone (mandible or maxilla) –** The surgeon will remove the affected bone. Once healed, bones from other parts of the body may be used to replace the affected bone. As the face is a delicate area, it can be difficult to remove the cancer with surgery and some people may need to have other treatments (see page 5).

- **Spine or skull –** If surgery isn’t possible, a combination of treatments may be used. This may include radiation therapy, chemotherapy, cryotherapy (freezing method) or curettage (scooping out the cancer). If you need one of these specialised types of treatment, your doctor will discuss the details with you.

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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's 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 or control the symptoms of an advanced cancer.

Chemotherapy drugs are usually injected into a vein. Most people have chemotherapy as a day patient, but some types of drugs require a hospital stay.

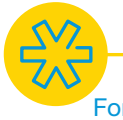
You may have additional imaging (MRI, CT or PET scans) during treatment to see how well the disease is responding to the chemotherapy.

The side effects of chemotherapy will depend on the drugs you receive and where the cancer is located in your body. Common side effects include tiredness, nausea, vomiting and diarrhoea, appetite loss, hair loss, mouth ulcers, 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. Some people may need to have blood transfusions to replace destroyed blood cells.

Radiation therapy

This treatment uses high-energy x-rays to destroy cancer cells. It may be used for Ewing's sarcoma:

- before surgery, to shrink the size of the tumour
- after surgery or chemotherapy, to kill any remaining cancer cells
- to help control the cancer if it's not possible to remove the tumour surgically
- to help control pain or other symptoms.



For more information on treatments and managing side effects, read Cancer Council's *Understanding Surgery*, *Understanding Chemotherapy* and *Understanding Radiation Therapy* booklets.

New treatments

Clinical trials test new treatments to see if they're better than current methods. Talk to your doctor about whether you may be able to access new treatments through a clinical trial.

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

Side effects will depend on the area being treated and the strength of the dose you have. Not everyone experiences side effects to the same degree. Common side effects include fatigue (tiredness), skin redness or soreness, and hair loss within 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 overwhelming. The physical changes that occur after treatment for bone cancer can affect your body image and self-esteem. It is natural to focus on the part of your body that has changed. Give yourself time to adapt to any changes in your appearance.

Limb-sparing surgery is a major operation that can leave a visible scar and skin tightness. If you have a limb amputated, it can take several months to feel comfortable with the prosthesis. You may avoid socialising with other people because of anxiety about how you look or because you find it difficult to move around. Physiotherapy can help you regain flexibility and improve movement.

Most people need emotional support before and after treatment, particularly if they have an amputation or a lot of bone is removed. You may feel emotions of grief and loss. Many people find it helps to talk things through with a counsellor, psychologist, friend or family member. Talk to your treating team or call Cancer Council 13 11 20 about support services available in your area.

For more information on coping with the emotional impact of bone cancer, read our *Emotions and Cancer* booklet.

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Follow-up appointments

After treatment, you will need check-ups every 3–12 months for several years to confirm that the cancer hasn't come back and to help you manage any treatment side effects. You will have a physical examination, and may have further imaging scans.

How often you will need to see your doctor will vary depending on the type of bone cancer. Appointments will become less frequent if you have no further problems.

Let your doctor know immediately of any health problems between appointments. Your doctor will tell you about things to look for and what to do if you think the bone cancer has come back.

If the cancer comes back

For some people, bone cancer does come back after treatment, which is known as a recurrence. The risk that bone cancer will recur is greater within the first five years after treatment. If the cancer does come back, treatment is likely to include a mix of surgery, chemotherapy and radiation therapy.

In some cases of advanced 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.

References

1. Australian Institute of Health and Welfare (AIHW), *Australian Cancer Incidence and Mortality (ACIM) books: bone cancer*, AIHW, Canberra, 2018.
2. Cancer Council Australia Sarcoma Guidelines Working Party, *Clinical practice guidelines for the management of adult onset sarcoma*, Cancer Council Australia, Sydney. [Cited 3 May 2019.] Available from: wiki.cancer.org.au/australia/Guidelines:Sarcoma.

Question checklist

- 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?
- Will I have to stay in hospital?
- 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

- **Vist rarecancers.org.au –** information from Rare Cancers Australia.
- **Visit canteen.org.au –** information and support for young people aged 12–25.
- **Call Cancer Council 13 11 20 –** health professionals can listen to your concerns and link you to services and support groups. You can also find information on your local website:

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

Acknowledgements

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Cancer Council 13 11 20

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

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