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've used the term bone cancer. It includes basic information about how primary bone cancer is diagnosed and treated.

The bones

A typical healthy adult 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.

The 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). 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 be either primary or secondary bone cancer. The two types are quite different and this fact sheet is only about primary bone cancer.

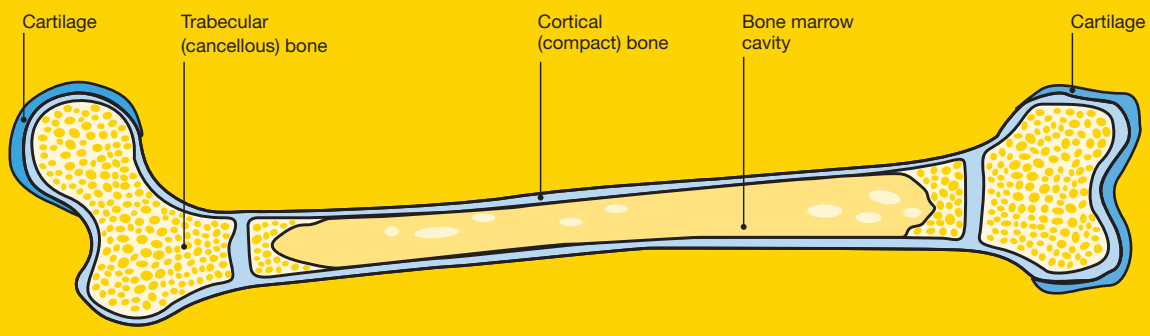
- **Primary bone cancer –** means that the cancer starts in the bones. It may develop on the surface of the bone, in the outer layer or from the centre of the bone. As a tumour grows, cancer cells multiply and destroy the bone. If left untreated, primary bone cancer can spread to other parts of the body.

- **Secondary (metastatic) bone cancer –** means that the cancer started in another part of the body, such as the breast or lung, and has spread to the bones.

How common is bone cancer?

Bone cancer is rare. About 200 Australians are diagnosed with primary bone cancer each year.[1] It affects people of all ages and is slightly more common in males than females. 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 include:

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 often have a large lump associated with them • often affects the pelvis (hips), legs, ribs, spine, upper arms • common in children and young adults • are all 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 some factors that increase the risk include:

Previous radiotherapy – Radiotherapy to treat cancer increases the risk of developing bone cancer. The risk is higher for people who have high doses of radiotherapy at a young age. Most people who have radiotherapy 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 bone cancer. Some studies also suggest that people who have had a soft tissue sarcoma are at 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 other 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 of bone cancer is strong pain in the bones and joints. The pain gradually becomes constant and does not improve with mild pain-relieving medicines 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 movement, e.g. an unexplained limp
- loss of feeling in the affected limb
- a 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 are experiencing 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 –** painless scans of the bones, which can reveal bone damage or the creation of new bone
- **blood tests –** including a full blood count
- **CT or MRI scans –** a special computer is used to scan and create pictures to highlight any bone abnormality; it takes about an hour to perform

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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; it may take around 90 minutes to perform
- **bone biopsy –** removal of some cells and tissues from the affected bone for examination under a microscope. The biopsy may be done in one of two ways. In a core needle biopsy, a local anaesthetic is used to numb the area, then a thin needle is inserted into the bone under CT guidance. An open or surgical biopsy is done under general anaesthesia. The surgeon will cut through the skin to expose the bone and take a tissue sample.

Staging

The test results will help your doctors assign a stage to describe the bone cancer. You may also undergo some other diagnostic tests (such as a bone scan, bone marrow biopsy, PET scan or chest CT) to assess whether the bone cancer has spread from the original site to other parts of the body. Staging describes how far the cancer has spread and helps your health care team plan the most suitable treatment for you.

Grading

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).

Selection of bone biopsy site

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 specialises in treating 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. For a better outcome, it is recommended you are referred before the biopsy to a specialist sarcoma service.

Your doctor will recommend the best treatment for you, depending 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 and general health
- your preferences.

Treatment for primary bone cancer usually includes surgery, chemotherapy and radiotherapy, or a combination of these treatments, with the aim of controlling the cancer and maintaining 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 the available treatments 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 the doctor's recommendations.

Specialist treatment centres

Diagnosis and treatment decisions can be very difficult. If your GP suspects that you have bone cancer, they'll refer you to one of the specialist centres throughout Australia that have expert multidisciplinary teams (MDTs) who manage this cancer on a regular basis. See australiansarcomagroup.org for a list of specialist sarcoma services.

The team will include a range of specialist doctors, nurses and allied health professionals who will be responsible for different aspects of your treatment.

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Preparing for treatment

- Some types of chemotherapy and radiotherapy can affect your heart and kidneys. Your doctor may recommend you have some tests to check your heart and kidney functions.
- 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, download the *Fertility and Cancer* booklet from your local Cancer Council website.
- If your doctor suspects you risk fracturing a bone because of the cancer, you may need to 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 make sure they 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 surgeon replaces the bone that is removed with an implant (prosthesis) or a bone graft. A graft involves using 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 use during surgery. In some cases, it may be possible to treat the removed bone with radiotherapy to destroy the cancer cells, then use the sterilised bone to reconstruct your limb.

After surgery, the remaining soft tissue and skin will heal. You will be given medicines to help you manage any pain. There will be some changes in the way the remaining limb looks, feels or works. A physiotherapist can plan an exercise program to help you regain strength and function in your limb.

Your doctor will talk to you about the risks of surgery. It is likely that you will be given antibiotics to reduce the risk of getting an infection in the bone or prosthesis.

Surgery to remove the limb (amputation)

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

After surgery, any remaining tissue (called the residual limb) will be swollen and painful. You will be given medicine to manage the pain and taught how to care for the 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 your function, such as walking. In some cases, using a prosthetic leg may be too difficult and you may 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 (a 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 surgically 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 radiotherapy, 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

Chemotherapy uses drugs to destroy or slow the growth of cancer cells, while causing the least possible damage to healthy cells. For certain types of bone cancer, such as high-grade osteosarcoma and Ewing's sarcoma, chemotherapy may be given:

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

Drugs are usually injected into a vein over several hours. Most people have several treatment cycles. The number and length of chemotherapy cycles you have depends on the type of bone cancer.

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

The side effects of chemotherapy will depend on the drugs you receive and where the cancer is located in your body. Some people have few side effects; others have more. Common side effects include tiredness, nausea, vomiting and diarrhoea, appetite loss, hair loss, and increased risk of infection. Most can be managed with medicines or other techniques.

Radiotherapy

Radiotherapy uses high-energy x-rays to destroy cancer cells. It may be used for certain types of bone cancer, such as 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.

New treatments

Clinical trials test new treatments to see if they're better than current methods. Accessing new treatments is an important consideration in your care. Talk to your doctor about the latest developments and whether you're a suitable candidate.

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

Radiotherapy is usually given every weekday, with a rest over the weekend. How long your treatment takes will depend on the type and size of the cancer, but it may take a few weeks. 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 will experience 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.

Emotional wellbeing

The physical changes associated with treatment for bone cancer can affect your self-esteem and body image. 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 noticeable scar and tightness. If you have a limb amputated, it can take several months to feel comfortable with the prosthesis. You may restrict your interactions 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 ease of movement.

Most people need emotional support before and after treatment, particularly if they have an amputation or a lot of bone is removed. 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 Cancer Council's *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 you have. 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 advise 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 recur, treatment is likely to include a mix of surgery, chemotherapy and radiotherapy.

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 any pain and help to manage other symptoms.

Reference

1. Australian Institute of Health and Welfare (AIHW), *Australian Cancer Incidence and Mortality (ACIM) books: Bone cancer*, AIHW, Canberra, 2017.

Questions checklist

- What type of bone cancer do I have?
- What treatment do you recommend and why?
- What is the prognosis?
- How long will treatment take?
- Will I have to stay in hospital?
- If I have surgery, what are the side effects? Do I need an amputation?
- Are the latest tests and treatments for this type of bone cancer available in this hospital?
- 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. 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..... **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

The information is based on the clinical practice guidelines for bone sarcoma diagnosed in adults. This information was reviewed by: Dr Richard Boyle, Orthopaedic Oncological and Reconstructive Surgeon, Royal Prince Alfred Hospital, North Shore Private Hospital and Chris O'Brien Lifehouse, NSW; Dr Jayesh Desai, Medical Oncologist, Peter MacCallum Cancer Centre, VIC; Sandeep Gupta, Head of Physiotherapy, Balmain Hospital, NSW; Prof Angela Hong, Radiation Oncologist, Chris O'Brien Lifehouse and University of Sydney, NSW; James Hyett, Consumer; Caitriona Nienaber, 13 11 20 Consultant, Cancer Council WA.

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.

Cancer Council 13 11 20

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