

Understanding Primary Bone Cancer

Information 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 (cortical bone) and a spongy inner core (trabecular bone). Cartilage is the tough material at the end of each bone that allows one bone to move against another. These are called joints.

What is bone cancer?

Bone cancer can be either primary bone cancer or secondary bone cancer. The two types of bone cancer are quite different. This fact sheet is only about primary bone cancer (bone sarcoma).

Primary bone cancer is cancer that 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 a cancer that started in another part of the body, such as the breasts, lungs or prostate, and has spread to the bones.

How common is bone cancer?

Bone cancer is rare. About 195 Australians are diagnosed with primary bone cancer each year.

Bone cancer affects people of all ages, and it is slightly more common in males than females. If it develops later in life, it may be linked to another bone disease, such as Paget's disease, which causes enlarged and deformed bones.

Secondary bone cancer is more common than primary bone cancer, and is treated differently.

What types are there?

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

Osteosarcoma	<ul style="list-style-type: none"> • affects cells that grow bone tissue • often occurs in younger people with growing bones
Chondrosarcoma	<ul style="list-style-type: none"> • begins in the cartilage • often affects the pelvis, thigh bone, ribs and shoulder blade • most often occurs in middle-aged people
Ewing's sarcoma	<ul style="list-style-type: none"> • affects cells in the bone or soft tissue that multiply rapidly and often have a large lump associated with them • is usually found in children and young adults

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**.

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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 in the future. The risk is higher for people who have had high doses of radiotherapy at a young age. Most people who have had 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 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 centre (also known as Clinical Genetics or Familial Cancer Service) for more information.

Some people develop bone cancer due to genetic changes that happen during their lifetime, rather than inheriting a faulty gene.

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 medication 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, such as an unexplained limp
- unexplained weight loss
- loss of feeling in the affected limb
- tiredness.

Most people who have these symptoms do not have bone cancer. However, 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 arrange some tests, including:

- **x-rays** – painless scans of the bones
- **CT or MRI scans** – a special computer is used to scan and create pictures of your body, sometimes in 3D, that will help to highlight any bone abnormality. It may take about an hour to perform
- **biopsy** – a small tissue sample is taken. A local anaesthetic is used to numb the area, then a thin needle is inserted into the bone. The sample is examined under a microscope. A bone biopsy may also be done using general anaesthesia.

Grading

Grading describes how quickly a cancer might grow.

Low grade	the cancer cells look similar to normal bone cells, are usually slow-growing and are less likely to spread
High grade	the cancer cells look very abnormal and grow quickly, and are more likely to spread

Staging

Staging describes how far the cancer has spread. Knowing the stage helps doctors plan the most suitable treatment for you. The stages of bone cancer are also based on the grade of the cancer.

Stage 1	the cancer contains low-grade cells, and there is no spread beyond the bone
Stage 2	the cancer contains high-grade cells, and there is no spread beyond the bone
Stage 3	the cancer is any grade, and has spread to other parts of the body

Making decisions about treatment

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.

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Treatment

Your doctor will recommend the best treatment for you, depending on:

- the type of primary bone cancer
- 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.

Specialist treatment centres

Although bone cancer is rare and treatment is often complex, there are specialist centres throughout Australia that have expert teams who manage this cancer on a regular basis.

Surgery

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

Surgery to remove the cancer but spare the limb (limb-sparing surgery)

Limb-sparing surgery 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 where the cancer is growing. Often the surgeon takes out some surrounding bone and muscle to be sure they are removing as much of the cancer as possible, and to reduce the chance of the cancer coming back. This is called a wide local excision.

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 the body or a bone transplant from a 'bone bank'. A bone bank is a facility that collects tissue for research and use during surgery.

After surgery, the remaining soft tissue and skin will heal. 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.

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 out of 10 people, the only effective treatment is to remove the limb. This procedure is becoming less common as limb-sparing surgery has improved in the past few decades.

After surgery, any remaining tissue (called the stump) will be swollen and painful. You will be given medication to manage the pain. After the area has healed, you will be measured for a replacement limb (prosthesis). A physiotherapist will teach you exercises and techniques to regain movement using a prosthetic arm or leg.

Surgery for bone cancer in other parts of the body

Other types of surgery are used for bone cancers that don't affect the limbs.

- **Pelvis** – When possible, the cancer is removed along with some healthy tissue around it. This is called 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 bone that was removed. As the face is a delicate area, it can be difficult to remove the cancer surgically and other treatment may be required.
- **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.

Most people need emotional support before and after surgery, particularly if they have an amputation or a lot of bone is removed. A counsellor or psychologist can talk to you about how you are feeling and help you adjust to the changes.



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Chemotherapy

Chemotherapy drugs are given to damage or destroy cancer cells. For certain types of bone cancer, such as high-grade osteosarcoma, chemotherapy is given in combination with surgery:

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

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

The side effects of chemotherapy will depend on the drugs you receive and where the cancer is in your body. Some people have few side effects and others have more. Most side effects can be managed with medication or other techniques.

Radiotherapy

Radiotherapy uses high-energy rays to damage or destroy cancer cells. If it is used, it may be given before or after surgery. Talk to your specialist if radiotherapy is recommended for you.

Treatment is usually given in the hospital radiotherapy department, 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.

Side effects will depend on the area being treated and the strength of the dose you have.

Question checklist

You may find this checklist helpful when thinking about the questions you want to ask your doctor about primary bone cancer and treatment. If your doctor gives you answers that you don't understand, ask for clarification.

- What type of primary bone cancer do I have?
- How far has the cancer spread? How fast is it growing?
- What treatment do you recommend and why?
- 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 cancer available in this hospital?
- Are there any clinical trials or research studies I could join?
- If my cancer has spread outside the bone, what treatment options are available for me?
- How often will I need check-ups? Which specialists will I see?
- 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 additional information, and put you in touch with local services. Ask for a free copy of *Understanding Surgery*, *Understanding Chemotherapy* or *Understanding Radiotherapy*.

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.



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