

Soft tissue sarcoma

For people with cancer, their families and friends

This information sheet is for people with soft tissue sarcoma who would like to know more about their cancer and its treatment. It describes the types, causes and symptoms of soft tissue sarcoma as well as treatments and their possible side effects.

This does not replace talking to your doctors or nurses. However, we hope that it will help you make decisions with your doctor about your treatment.

What is soft tissue sarcoma?

Soft tissue is the name for all the supporting tissues in the body, apart from the bones. They include fat, muscle, nerves, deep skin tissue, blood vessels and the tissue that surrounds joints (synovial tissue).

These tissues support and connect all the organs and structures of the body. They are known as connective tissue. A soft tissue sarcoma is a rare type of cancer that forms as a painless lump (tumour) in any one of these soft tissues. They most commonly develop in the thigh, shoulder and pelvis. Sometimes they can grow in the abdomen or chest (trunk).

Types of soft tissue sarcoma

There are over 70 types. They are named after the abnormal cells that make up the sarcoma.

The most common type is malignant fibrous histiocytoma (MFH), which is made up of millions of abnormal spindle-shaped cells.

Liposarcoma is the next most common type of soft tissue sarcoma. It is made up of millions of abnormal fat cells.

Other types of soft tissue sarcoma include:

- leiomyosarcoma and rhabdomyosarcoma, from muscle tissues
- angiosarcoma, from blood vessels
- Ewing sarcoma and primitive neuroectodermal tumour (PNET)
- malignant peripheral nerve sheath tumour (MPNST or PNST)
- gastrointestinal stromal sarcoma (GIST) (this is treated differently from other types of soft tissue sarcoma)
- stromal sarcoma from supporting tissues
- Kaposi sarcoma of the skin
- synovial sarcoma.

How common is soft tissue sarcoma?

Soft tissue sarcoma is not common. Around 226 Victorians develop one of these cancers each year.

Soft tissue sarcoma can develop at any age. Although it is more likely to happen in people over the age of 55, young adults may also be affected. There is almost an equal risk for males and females.

Causes

The causes of most sarcomas are not known. There are several risk factors.

Radiotherapy

There is a very small risk for people who have had radiotherapy. The risk is higher for people who had high doses of radiotherapy at a very young age. Most people who have had radiotherapy in the past will not develop a sarcoma.

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Genetic factors

Some rare, inherited conditions can put people at more risk of soft tissue sarcoma. These are:

- Von Recklinghausen disease (neurofibromatosis): non-cancerous lumps that develop in the nerves under the skin and over time can turn into sarcoma
- Li–Fraumeni syndrome: a genetic condition that increases the risk of many cancers in one family, including soft tissue sarcoma
- Retinoblastoma: a rare type of eye cancer mainly found in children.

Most people know if one of these very rare conditions runs in their family, and if so, that they may pass it to their children.

A small number of people develop sarcoma due to genetic changes that happen during their lifetime, rather than inheriting a faulty gene.

Symptoms

Sarcoma usually doesn't cause symptoms in its early stages. As it grows over a period of months, you may develop a painless lump. You may begin to have pain as the lump grows and presses on nerves and muscles. Other symptoms will depend on where in the body the sarcoma is.

Sometimes a soft tissue sarcoma may be mistaken for a benign fatty lump (lipoma) or bruise (haematoma). This can delay tests that would help make the right diagnosis.

Most people who develop a painless lump do not have a sarcoma. It is much more likely to be a less serious condition. However, if you have symptoms that stay for more than two weeks you should see your doctor.

Diagnosis

If your doctor thinks that you may have a soft tissue sarcoma they will carry out blood tests, x-rays and scans. These tests are needed to rule out other conditions. If the results suggest that you may have sarcoma, your doctor will refer you to a specialist centre.

You may need to have a biopsy. This is the only sure way to diagnose a soft tissue sarcoma. A biopsy means taking a sample of cells from the lump and examining them under a microscope. Your doctor will use a fine needle to do this. You will have local anaesthetic to numb the area before the needle is put into the lump.

A tissue biopsy should only be carried out by a specialist who does them regularly. It is important that the biopsy is done properly in order to minimise the chance of the cancer spreading. It could also affect the type of treatment that you may be able to have.

Treatment

Your doctor will help you decide on the best treatment for your sarcoma depending on:

- the type of sarcoma you have
- where the sarcoma is in your body
- whether or not the cancer has spread (its stage)
- your age, fitness and general health
- what you want.

Treatment may include surgery, chemotherapy and radiotherapy. They can be given alone or in combination.

Surgery

Surgery is the main treatment for most types of soft tissue sarcoma. This usually means removing the cancer as well as some healthy tissue around the sarcoma.

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Doctors call this a wide local excision. The healthy tissue is taken away to help decrease the risk of the cancer coming back in that area.

The type of operation you have will depend on where the sarcoma is. Most sarcomas are in the arm or leg, which usually means having limb sparing surgery, or, less commonly, amputation. But sarcoma can develop in other parts of the body, for example the chest or abdomen. Surgery to these areas will depend on the exact position and size of the cancer. Your surgeon will discuss with you the type of operation you may need.

The most important thing is that you have your surgery done by an experienced surgeon working in a specialist centre for sarcoma.

Plastic surgeons are sometimes asked to work with surgeons to help restore parts of the body affected by surgery. This would be part of your main operation. The type of surgery you have will depend on the location of the sarcoma and how much tissue needs to be removed.

Before you make any decision about treatment, your surgeon will discuss all your options in detail.

Surgery for sarcoma that has spread to the lungs

Sometimes sarcoma spreads into the lungs. Unlike some other types of cancer, sarcoma lung cancers can sometimes be taken out using surgery. This depends on the size and number of cancers. You will see a specialist lung surgeon for an opinion. Alternatively, or as well as surgery, you may have chemotherapy to help stop the cancer coming back.

Limb sparing surgery

This means taking out the cancer as well as a margin of healthy tissue while still keeping your limb. If necessary, bones may be rebuilt using a metal implant (prosthesis) or a bone graft. A graft means taking a piece of healthy bone from somewhere else in your body or from a 'bone bank'. Any other gaps in the tissue are usually reconstructed using plastic surgery.

After this type of surgery a physiotherapist will work with you to plan an exercise program that will help you gain strength and function back in your limb. There will be some changes in the way the limb looks, feels or functions after limb sparing surgery. The aim is for your limb to return to as normal a state as possible.

This complex surgery needs to be done by a surgeon with a lot of experience in treating sarcoma. This may mean that you have to travel to a treatment centre that has a team of experts. Surgery done well may decrease the risk of the cancer cells spreading to nearby areas or other parts of the body.

Amputation

Sometimes it is not possible to remove all of the cancer without badly affecting the arm or leg. The doctor may advise that the only effective treatment will be to remove the limb (amputation).

Amputation can be very distressing emotionally and physically. Most people will need a lot of support at this time. It could have a huge affect on you and the people who share your life.

After surgery you will have a carefully planned rehabilitation program. This will include seeing a specialist who makes false (prosthetic) limbs. A physiotherapist will work with you to help you become as independent and mobile as possible.

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Other types of support are also useful. You may find it helps to speak with a social worker or counsellor or even someone else who has had the same operation. They may be able to offer advice on ways to cope and what to expect.

Radiotherapy

Radiotherapy uses high-energy rays to destroy the cancer cells. Sarcoma is known to be very sensitive to radiotherapy. It may be used for several reasons, including:

- if the cancer is too big to remove with surgery
- if the cancer has spread to other parts of the body
- after surgery, to destroy any remaining cancer cells and stop the cancer coming back
- if the cancer is in a place in the body that is too difficult to get to using surgery
- before surgery, to destroy the cancer cells and create a 'rind' around the cancer, which helps the surgeon to perform good surgery.

Radiotherapy can shrink the cancer down to a smaller size. This may help to make surgery easier and safer. Sometimes chemotherapy is given as well as radiotherapy to help make the treatment more effective. This depends on the type of sarcoma.

Radiotherapy is usually given in small doses over a period of time to minimise side effects. For example, you may have a single dose five days a week for five or six weeks. Each dose only takes a couple of minutes, but some preparation time is needed as you have to be treated in the same position each time. Treatment given after surgery will usually begin within six weeks after your surgery.

A course of radiotherapy needs to be carefully planned. During your first visit you will need to lie under a large machine called a simulator. Your specialist will use this machine to calculate the correct dose and exact angles on your body for the radiation to target. This process ensures that the cancer receives the highest doses of radiation and the healthy tissue around it gets as little as possible.

Radiotherapy does not hurt, but you need to lie very still during the treatment. You can have radiotherapy as an outpatient. Some people are able to continue working throughout their treatment while others become tired. Some people may have a skin reaction. Your treating team will discuss possible side effects with you before you start your treatment.

The Cancer Council's booklet *Coping with Radiotherapy* discusses ways of managing side effects. Visit www.cancervic.org.au or telephone **13 11 20** for a copy.

Chemotherapy

Chemotherapy drugs are given to destroy cancer cells.

Chemotherapy is most commonly given to treat Ewing sarcoma and sarcoma in children. Sometimes it is given to help control symptoms or slow down the growth of sarcoma that has spread. This is known as palliative chemotherapy. It may also be given to help stop a sarcoma coming back after surgery. Your doctor will discuss your options with you.

You may also be asked whether you would like to join a clinical trial.

Your treatment will depend on your situation and the type of chemotherapy you have. Your doctor will discuss these with you before you begin treatment.

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Chemotherapy is given as a course of several treatment cycles. You may have one or a combination of drugs. For example, one cycle commonly lasts three weeks. Depending on the drugs you are having, you will have them over a few hours or days. You will then have a rest for three weeks before starting another cycle. You may have between three and five cycles before surgery and then more after your surgery.

The doctor may recommend a 'central line' (portacath, Hickman line) in your chest or 'PICC line' in your arm. These are put in under local or general anaesthetic and stay in throughout your treatment. Their main advantage is that the chemotherapy can be given safely through the line: you don't have to have lots of needles. You can also have your blood taken from this line without having a needle.

If you have a central line, you will need to take good care of it. It needs flushing and cleaning regularly to prevent any infection or blockage. A nurse will teach you before you go home or you may have a district nurse to help you take care of it. The line is taken out when treatment is finished.

The Cancer Council's booklet *Coping with Chemotherapy* discusses ways of managing side effects.

Visit www.cancervic.org.au or telephone **13 11 20** for a copy.

Side effects of treatment

All treatments can have side effects. Many are short term and will disappear once the treatment is over. For example, chemotherapy can cause a drop in your blood cell counts, putting you more at risk of infection, anaemia and bleeding problems. These usually return to normal once your treatment is over.

Some side effects are long term. This may mean a change in the way a part of your body looks, feels or functions.

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 and others have more. Your medical team will discuss all possible side effects (long and short term) before your treatment begins. For further information about treatment side effects call the Cancer Council Helpline on **13 11 20**.

Making decisions about treatment

It can be difficult to know which treatment is best for you. It is very important that you speak with a sarcoma specialist team before making your decision. Ask them to give you a clear plan of your treatment options including information about long and short-term side effects. Some people prefer to seek several opinions before feeling confident to go ahead with the treatment.

You may have to attend many appointments. It often helps to take someone with you. They will be able to listen, ask questions and remember what the doctor says. You can discuss your treatment options later. It may also help to take a list of questions. See the list on the next page.

Complementary and alternative treatments

It's common for people with cancer to seek out complementary and alternative treatments. Many people feel that it gives them a greater sense of control over their illness, that it's 'natural' and low-risk, or that they just want to try everything that seems promising.

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Complementary therapies include massage, meditation and other relaxation methods that are used along with medical treatments. Many people find using one or more types of complementary therapies very beneficial.

'During my treatment I did a daily meditation to help keep my emotions under control. I also had weekly massages, which made me feel more relaxed.' (Jane, 59)

Alternative therapies are unproven and include some herbal and dietary remedies that are used instead of medical treatment. Some of these have been tested scientifically and found to be not effective or even harmful.

Some complementary therapies are useful in helping people to cope with the challenges of having cancer and cancer treatment. However, some alternative therapies are harmful, especially if:

- you use them instead of medical treatment
- you use herbs or other remedies that make your medical treatment less effective
- you spend a lot of time and money on alternative remedies that simply don't work.

Be aware that a lot of unproven remedies are advertised on the Internet and elsewhere without any control or regulation. Before choosing an alternative remedy, discuss it with your doctor or a cancer nurse at the Cancer Council Helpline on **13 11 20**.

For more information we strongly recommend you read the following Cancer Council Victoria resources:

- *Complementary and Alternative Cancer Therapies* (booklet)
- *Complementary and Alternative Medicine: Making Informed Decisions* (information sheet).

These resources aim to help you recognise which therapies may be helpful, and recognise false claims about 'cures'. Call the Cancer Council Helpline or visit www.cancervic.org.au for copies of these resources.

You can find information on the Memorial Sloan-Kettering website (www.mskcc.org/mskcc/html/11570.cfm). The US National Center for Complementary and Alternative Medicines (NCCAM) (nccam.nih.gov) and Quackwatch (www.quackwatch.com) are also reliable websites.

Questions for your doctor

If you have a soft tissue sarcoma there are many questions that you will need to ask your specialist. Getting the answers will help you make the best decision about which treatment to choose. Here are a few suggestions.

- How will I know if I have a soft tissue sarcoma?
- What has caused my soft tissue sarcoma?
- Is my family more at risk of soft tissue sarcoma?
- If I need to have a biopsy, will I have it at a specialist centre?
- If I need to have treatment, where is the best centre for this type of cancer?
- Is it possible to have surgery to try to cure my soft tissue sarcoma?
- If I can have surgery, which type will I need and why?

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- What are the risks and long-term side effects of each type of surgery?
- What type of rehabilitation will I need after my surgery? How long will recovery take?
- Will I need to have any other type of treatment, such as radiotherapy or chemotherapy?
- How effective is radiotherapy and chemotherapy for my type of sarcoma?
- If my cancer has spread outside the area it began, what treatment options are there for me?

Getting support

For more information contact the Cancer Council Helpline on **13 11 20** (cost of a local call). This is a confidential service staffed by cancer nurses. Information is available in languages other than English.

The cancer nurse can supply you or your family with up-to-date information about your cancer and treatment. They can also link you in with another person who has had your type of cancer, through Cancer Connect. There is also Family Cancer Connect for your partner or carer. If you prefer you may also be linked in with your nearest Cancer Support Group.

The Cancer Council Helpline operates Monday to Friday from 8.30 am to 6 pm. All services are free.

If you are an adolescent or young adult you may want to contact onTrac@PeterMac (www.petermac.org/ontrac), a support service for young people with cancer.

Caring for someone with cancer

A carer is anyone, whether family or friend, who is helping to look after someone with cancer.

Caring for someone with cancer can be very stressful, particularly when it is someone you care about very much. The person with cancer may be distressed about their diagnosis, side effects from treatment and mood changes from the effects of drugs.

Look after yourself during this time. Give yourself some time out, enjoy a cup of coffee with a friend, and share your worries and concerns with someone not involved. Make a list of 10 things you like to do and make sure that you do one each day.

Cancer Support Groups are usually open to patients and carers. A support group can offer the chance to share experiences and ways of coping.

A range of support services, such as home help, meals on wheels and visiting nurses, can help you cope with treatment at home. These are provided by local councils, the Royal District Nursing Service and the palliative care services.

Call the Cancer Council Helpline on **13 11 20** to:

- be linked with another carer by telephone
- speak with a cancer nurse for further support and information
- be sent a carers kit so that you can find out about financial assistance and other resources.

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