Soft tissue sarcomas are a group of cancers that develop from cells of connective tissue. These include cells that make up our muscles, fat, lining of joints and blood vessels. Soft tissue sarcomas can therefore develop almost anywhere in the body.

The most common type of soft tissue sarcoma is called a rhabdomyosarcoma, which forms from early cell types (called rhabdomyoblasts) that make up skeletal muscles. Rhabdomyosarcomas are more common in younger children (under 10 years of age), although they can also develop in teenagers and adults.

Other types of soft tissue sarcomas include:

- Ewing sarcoma of soft tissue
- liposarcoma, which forms in fat
- synovial sarcoma, which can form near a joint such as the elbow or knee, or in other places in the body
- leiomyosarcoma, which forms in smooth muscle.

Risk factors

A risk factor is anything that increases a person's chance of developing a certain condition or disease, such as cancer. In adults, lifestyle and environmental factors (such as smoking or exposure to certain chemicals) can be significant risk factors for developing certain types of cancer. In children, very few risk factors have been identified that increase the chance of developing cancer. For most children with cancer, the underlying cause is unknown.

Even if your child has a risk factor, it does not mean they will develop cancer. Many children with a risk factor will never develop cancer, while others with cancer may have had no known risk factors. Even if a child with a risk factor develops cancer, it is usually hard to know how much that risk factor contributed to the development of their disease.

The causes of soft tissue sarcomas are not well understood, but factors associated with a higher chance of developing soft tissue sarcomas include the following.

Genetic conditions

Certain genetic disorders are associated with a higher chance of developing soft tissue sarcomas. These include:

- Beckwith–Weideman syndrome
- Costello syndrome
- Li–Fraumeni syndrome
- neurofibromatosis type 1
- Noonan syndrome

- Werner syndrome
- familial adenomatous polyposis
- changes in the *Rb* (retinoblastoma) gene.

If your child is diagnosed with one of these genetic conditions, they will need specific follow-up. The health care team will advise which ongoing tests your child will need.

Cancers in children that are linked to genetic conditions may also affect the risk for other family members. Speak to your child's treatment team to see whether genetic counselling is recommended for you or your family.

For more information about genetic conditions, see the children's cancer glossary or the Centre for Genetics Education.

Exposure to radiation

If your child has had treatment with radiation therapy in the past, they may have a higher chance of developing soft tissue sarcoma.

Certain virus infections

Children who have HIV and Epstein–Barr virus infections at the same time have a higher chance of developing soft tissue sarcomas.

Symptoms

Symptoms of soft tissue sarcomas may include a painless lump under the skin. If the tumour grows and presses on nearby organs, tissue, nerves or blood vessels, other symptoms can appear, such as pain or weakness. These symptoms vary, depending on where in the body the tumour has formed.

Note about symptoms

Many conditions – including common childhood infections – can cause these symptoms, not just soft tissue sarcomas. If your child has any of these symptoms and you are concerned, talk to your child's doctor.

Diagnosis

Your child will have a number of tests to investigate their symptoms and confirm a diagnosis of soft tissue sarcoma, including:

- medical history and physical examination
- medical imaging, which may include:
    - X-ray
    - ultrasound
    - computed tomography (CT) scan
    - magnetic resonance imaging (MRI)
    - bone scan
    - positron emission tomography (PET) scan
- biopsy – where a small sample of the cancer is removed to be examined under a microscope. The sample can also be tested for genetic changes that can help determine the best type of treatment for your child
- lumbar puncture (spinal tap) – where a sample of cerebrospinal fluid is taken to be examined under a microscope
- bone marrow aspiration and biopsy – where a sample of bone marrow is taken with a small piece of bone to be examined under a microscope.

These tests are explained in more detail in How is cancer diagnosed?.

Staging

If your child is diagnosed with soft tissue sarcoma, some of the diagnostic tests will also help to stage the tumour. Staging determines where the tumour is, how big it is, which nearby organs are involved and whether the cancer has spread to other parts of the body. This is important to determine the outlook (prognosis) for your child, and to decide on the best options for treatment.

There are different ways to assess the stage or extent of disease. One of the most common ways to describe stages for soft tissue sarcoma is as follows:

- Stage IA – the tumour is low grade (likely to grow and spread slowly) and less than 5 cm across
- Stage IB – the tumour is low grade (likely to grow and spread slowly) and more than 5 cm across
- Stage IIA – the tumour is mid grade (somewhat likely to grow and spread quickly) or high grade (likely to grow and spread quickly) and less than 5 cm across
- Stage IIB – the tumour is mid grade (somewhat likely to grow and spread quickly) and more than 5 cm across
- Stage III – the tumour is high grade (likely to grow and spread quickly) and more than 5 cm across, or has spread to nearby lymph nodes
- Stage IV – the tumour has spread to distant parts of the body, such as the lungs.

More information about staging for soft tissue sarcomas can be found from the National Cancer Institute (United States).

Treatment

Treatment and care of children with cancer is usually provided by a team of health professionals called a multidisciplinary team. Members of this team are specialists in children's cancers – they understand the differences between children's cancer and adult cancer, and each team member brings different skills in managing care to meet the needs of both you and your child.

The team will be led by a childhood cancer specialist (paediatric oncologist). Other members of the team depend on the age of your child and their type of disease, and may change over time as your child's needs change. A list of team members who might make up the multidisciplinary team can be found in The treatment team.

Treatment for soft tissue sarcomas depends on the age of your child, the stage of the disease, the biological features of the cancer and other factors identified during diagnosis. Treatment will be tailored to your child's particular situation, and may involve one or more of the following (see How is cancer treated for more detail).

Surgery

Your child is likely to have surgery to remove all or part of the tumour.

Chemotherapy

Chemotherapy uses anti-cancer medicines to destroy cancer cells. It is often given as a combination of medicines to try to prevent the cancer cells from becoming resistant to just one or two medicines.

Chemotherapy medicines are given together in courses, often over a few days. Once the body has recovered from the side effects, the next course is given. Most children receive multiple courses of chemotherapy.

Chemotherapy may be used:

- before surgery (to shrink the tumour and make it easier to remove)
- after surgery (to destroy any remaining cancer cells)
- as the main treatment (if the cancer has spread to other parts of the body, or if surgery is not a good option).

Most children with rhabdomyosarcoma will receive chemotherapy, but other types of soft tissue sarcomas do not respond to chemotherapy.

Radiation therapy

Radiation therapy (also called radiotherapy) uses high-energy X-rays or other types of radiation to destroy cancer cells or stop them from growing. It can be used for soft tissue sarcomas:

- before surgery (to shrink the tumour and make it easier to remove)
- after surgery (to destroy any remaining cancer cells)
- as the main treatment (if removing the tumour with surgery would damage important organs or cause disfigurement, or if the tumour is likely to grow and spread quickly).

Radiation therapy may be used in combination with chemotherapy.

Radiation therapy can have long-term side effects in children. If radiation therapy is included in your child's treatment, special care will be taken to reduce these risks.

Targeted therapy

Some medicines can target the specific changes in cancer cells that make them different from normal cells. This means that they work differently from standard chemotherapy, and they usually have fewer side effects, or the side effects are not as severe.

Drugs called tyrosine kinase inhibitors may be used to treat certain types of soft tissue sarcomas.

Other treatments

Some types of soft tissue sarcomas can be treated with hormone therapy or nonsteroidal anti-inflammatory drugs (which can stop the tumour growing).

Careful observation

For a few carefully selected patients, specific treatment may not be needed. If your child has a tumour that is not growing or spreading, they might be monitored closely but not given any treatment until they develop symptoms, or until their symptoms change .

Support

Diagnosis of cancer in a child is a very difficult time for the child, their family and their friends. You might feel overwhelmed, scared, anxious or angry. These are all normal feelings. It is very important to seek support from family, friends, health professionals or other services to help you, your child and your family cope with cancer.

Talk to your child's treatment team if you are having difficulties coping.

Living with children's cancer has information about physical, emotional and practical issues during and after diagnosis and treatment. There is also a page with helpful links on where to find support.

The Cancer Council in your state or territory can give you general information about cancer, as well as information on resources and support groups in your local area. Call the Cancer Council Helpline from anywhere in Australia for the cost of a local call on **13 11 20** .

For additional specific information about childhood cancer, contact any of the major children's hospitals and networks in your state or territory.

Chance of cure

Many children with cancer are cured of the disease. Children's bodies have great capacity for healing. Also, huge improvements have been made in the treatment of childhood cancer in the past few decades. In the 1980s, around 65% of children diagnosed with cancer were alive more than 5 years after their diagnosis. Today, around 83% of children are successfully treated and become long-term survivors.

Long-term survival (also called the outlook or prognosis) and treatment options depend on a range of factors, including:

- age of your child at diagnosis
- extent or stage of the cancer
- appearance of the cancer cells under the microscope (the shape, function and structure of the cells)
- how the cancer responds to treatment
- cancer or tumour biology, which includes
    - the patterns of the cancer cells
    - how different the cancer cells are from normal cells
    - how fast the cancer cells are growing.

Talk to your child's doctor about your child's individual disease, treatment options and outlook.

Clinical trials

Researchers are trialling new ways to diagnose and treat different types of cancer. Your child may be invited to be part of a clinical trial to test new ways of treating soft tissue sarcomas.

New treatments have to go through very strict regulation and approval processes before they can be used in a clinical trial. Your child's doctor will explain everything about the trial and give you detailed written information. You will need to give special permission for your child to be part of the trial.

Participating in a clinical trial may or may not directly benefit your child, but the results of clinical trials today will help children with cancer in the future.

See Clinical trials and research for more information, including whether there are any clinical trials your child can join.

More information

For more information about soft tissue sarcomas, see:

- Rhabdomyosarcoma, from the American Cancer Society
- Treatment PDQs® from the National Cancer Institute (United States), including
  - Childhood soft tissue sarcoma
  - Ewing sarcoma
  - Childhood rhabdomyosarcoma.