

## What is sarcoma?

A sarcoma is a rare type of cancer that begins in the connective tissues of the body such as fat, muscle, blood vessels, nerves, bone and cartilage. Sarcoma occurs when abnormal cells in these types of tissue grow in an uncontrolled way.

Sarcomas can develop anywhere in the body. Although sarcomas are rare across all age groups, they are among the more common types of solid tumour in childhood.

## Types of sarcoma

### Soft tissue sarcoma

Soft tissue sarcomas develop in soft tissues such as fat, muscle, blood vessels, lymphatic vessels, nerves, tendons and cartilage.

There are more than 50 types of soft tissue sarcomas. In adults, the most common types are:

- ▶ **Undifferentiated pleomorphic sarcoma (UPS):** an aggressive type of soft tissue sarcoma with high incidence of local recurrence and metastasis, most common in the age group of 50-70 years.
- ▶ **Leiomyosarcoma:** a malignant smooth muscle tumour that arises most commonly in the limbs, abdomen and uterus.
- ▶ **Liposarcoma:** a malignant tumour that arises from fat cells, most commonly in the trunk, limbs and abdomen.
- ▶ **Angiosarcoma:** a malignant tumour that arises from blood vessels or lymphatic vessels. Prior radiotherapy is a risk factor for angiosarcoma, often with a median latency period of 10 years.
- ▶ **Malignant peripheral nerve sheath tumour:** a malignant tumour that arises in the lining of nerves, often in the deep tissue of the arms, legs and trunk..
- ▶ **Fibroblastic sarcoma (fibrosarcoma):** a malignant tumour that develops in the fibrous tissues of the body, most commonly in the limbs, skin and trunk.
- ▶ **Gastrointestinal stromal tumour:** a common type of soft tissue sarcoma that starts in the digestive tract. These tumours are most common in people aged 50–80.
- ▶ **Kaposi's sarcoma:** a malignant tumour, which is caused by a virus that can cause the disease in people with a compromised immune system, such as people with AIDS, most commonly affecting the skin, mouth and internal organs.

In children, the most common types are:

- ▶ **Rhabdomyosarcoma:** an aggressive type of soft tissue sarcoma that which arises from skeletal muscles. Most common in children aged less than 10, although they can also develop in teenagers and adults.
- ▶ **Synovial sarcoma:** a malignant tumour that which develops in cells around joints and tendons, more commonly in children and young adults, but can occur in older people.

### Primary bone sarcoma

Primary bone sarcoma starts in the bone. This is different from bone metastases, which have spread to the bones from somewhere else, such as the breast or lung, when the cancer in these other organs is at an advanced stage. Bone sarcomas are more common in children and teenagers than in older adults. They include:

- ▶ **Osteosarcoma:** most common primary bone cancer, usually occurring at the ends of the long bones, especially around the knees
- ▶ **Ewing sarcoma:** common sites are the pelvis, the chest wall and the middle of the long bones in the legs; it may also form in soft tissue.

## What are the risk factors for sarcoma?

A risk factor is any factor that is associated with an increased chance of developing a particular health condition, such as sarcoma. There are different types of risk factors, some of which can be modified and some that cannot.

It should be noted that having one or more risk factors does not mean a person will develop sarcoma. Many people have at least one risk factor but will never develop sarcoma, while others with sarcoma may have had no known risk factors. Even if a person with sarcoma has a risk factor, it is usually hard to know how much that risk factor contributed to the development of their disease.

### **Risk factors for soft tissue sarcoma include:**

- ▶ certain inherited disorders – retinoblastoma, neurofibromatosis (von Recklinghausen disease), tuberous sclerosis (Bourneville disease), familial adenomatous polyposis (Gardner syndrome), Li–Fraumeni syndrome, Werner syndrome, nevoid basal cell carcinoma syndrome (Gorlin syndrome)
- ▶ previous radiation therapy for other cancers, such as breast cancer or lymphoma
- ▶ exposure to some chemicals (thorium dioxide, vinyl chloride, arsenic)
- ▶ having lymphoedema (swelling) in the arms or legs for a long time – lymphoedema can occur when lymph nodes have been removed or damaged by radiation therapy.

### **Risk factors for osteosarcoma include:**

- ▶ certain inherited disorders – Bloom syndrome, Diamond–Blackfan anaemia, retinoblastoma, Li–Fraumeni syndrome, Paget disease, Rothmund–Thomson syndrome, Werner syndrome, hereditary multiple osteochondromas (benign bone tumours)
- ▶ previous radiation therapy
- ▶ previous treatment with anticancer medicines called alkylating agents
- ▶ age and height.

Studies of children with Ewing tumours have not found any links to radiation, chemicals, other environmental exposures or inherited disorders.

Most gastrointestinal stromal tumours (GISTs) have no clear cause and are not inherited. However, inherited disorders increase the risk in some families. These disorders include primary familial GIST syndrome, neurofibromatosis (von Recklinghausen disease) and Carney–Stratakis syndrome.

Lifestyle factors such as smoking, diet and exercise are not risk factors for sarcoma.

## What are the symptoms of sarcoma?

### **The most common symptoms of soft tissue sarcoma are:**

- ▶ a growing lump under the skin, often on an arm or a leg – this may be either painless or painful
- ▶ pain
- ▶ trouble breathing.

### **Symptoms of sarcoma in the bone include:**

- ▶ for osteosarcoma, swelling over a bone, or pain in a bone
- ▶ for Ewing sarcoma, pain and/or swelling or a lump in the area of the tumour, such as the arms, legs, chest, back or pelvis – the lump is often soft and feels warm
- ▶ for Ewing sarcoma, fever for no known reason
- ▶ a bone that breaks for no known reason.



Gastrointestinal stromal tumours may not cause any symptoms until they reach a certain size. Very small tumours are common, and some of these will not grow or spread. Symptoms of gastrointestinal stromal tumour can include:

- ▶ blood in the stools or vomit
- ▶ black, tarry stools
- ▶ abdominal pain
- ▶ swelling in the abdomen
- ▶ nausea and vomiting
- ▶ loss of appetite, and feeling full after only a little food has been eaten
- ▶ weight loss
- ▶ feeling very tired
- ▶ trouble swallowing (if the tumour is in the oesophagus).

There are a number of conditions that may cause these symptoms, not just sarcoma. If any of these symptoms are experienced, it is important that they are discussed with a doctor.

## How is sarcoma diagnosed?

A number of tests may be performed to investigate symptoms of sarcoma and confirm a diagnosis. Some of the more common tests include:

- ▶ a physical examination
- ▶ imaging of the area with the lump, which may include X-ray, computed tomography (CT) scans, magnetic resonance imaging (MRI), ultrasound, positron emission tomography (PET) scans, bone scans, and endoscopy (use of a thin tube with a light and camera to look at the lining of the gastrointestinal tract – for gastrointestinal stromal tumours)
- ▶ taking a sample of tissue (biopsy) for examination under a microscope.

After sarcoma has been diagnosed, other tests may be done to find out whether the cancer cells have spread within the body and, if so, how far. This is called staging of the tumour, and is an important step in planning treatment. Tests that might be used as part of staging include the ones listed above, as well as testing of blood samples and bone marrow samples (for bone sarcomas).

## Treatment options

Treatment and care of people with cancer is usually provided by a team of health professionals – called a multidisciplinary team. Treatment for sarcoma depends on:

- ▶ the type of sarcoma
- ▶ where it is in the body
- ▶ whether and how much the cancer has spread
- ▶ your general health and wishes.

Treatment options can include surgery, chemotherapy, radiotherapy and targeted therapy. Research is ongoing to find new ways to diagnose and treat different types of cancer. You may be invited to participate in a clinical trial to test new ways of treating sarcoma.

### Finding support

You might feel overwhelmed, scared, anxious or upset if you have been diagnosed with cancer – these are all normal feelings. It's very important to have support from family, friends, health professionals or other services to help you cope with cancer. *Living with cancer* has information about physical, emotional and practical issues during and after diagnosis and treatment.

Cancer Australia's resource *Cancer – How are you travelling?* provides information to help you understand the emotional and social impact of cancer. Order or download a copy.

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**.

Other cancer support organisations can also help you and your loved ones deal with the challenges of cancer.

Australia Sarcoma Study Group website provides a resource for patients, families, carers, clinicians and health professionals who seek information about sarcoma and current research or wish to contact a specialist or to find out how to make a difference.



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