Ewing’s sarcoma in children

This fact sheet is about Ewing’s sarcoma in children. You may find it helpful to read it alongside the Macmillan/CCLG booklet A parent’s guide to children’s cancers, which contains more information about cancers in children, their diagnosis and treatment, and the support services available.

We hope this fact sheet answers your questions. If you have any further questions, you can ask a nurse or doctor involved in your child’s treatment.

If you'd like to discuss this information with our cancer support specialists, call the Macmillan Support Line free on 0808 808 00 00, Monday–Friday, 9am–8pm. If you're hard of hearing you can use textphone 0808 808 0121, or Text Relay. For non-English speakers, interpreters are available. Alternatively, visit macmillan.org.uk

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Sarcomas

Sarcomas are rare types of cancer that develop in the supporting tissues of the body. There are two main types: bone sarcomas and soft tissue sarcomas.

Bone sarcomas can develop in any of the bones of the skeleton, but may also develop in the soft tissue near bones.

Soft tissue sarcomas can develop in muscle, fat, blood vessels or any of the other tissues that support, surround and protect the organs of the body.
Ewing’s sarcoma

Ewing’s sarcoma is named after Dr James Ewing, who described the tumour in the 1920s. It’s a cancer that can develop anywhere in the body, although it most often starts in the bone. Any bone can be affected, but the pelvis, thigh bone (femur) and shin bone (tibia) are the most common places.

Fewer than 30 children in the UK develop Ewing’s sarcoma each year. It usually occurs in the teenage years, and more commonly affects boys than girls.

Although Ewing’s sarcoma is a type of bone cancer, it can also occur very rarely in the soft tissues rather than starting in the bone. This is called extraosseous Ewing’s sarcoma. Sometimes these cancers are called primitive neuroectodermal tumours (PNET).

Causes of Ewing’s sarcoma

The exact causes of primary bone cancer are unknown. The development of Ewing’s sarcoma may be related in some way to times of rapid bone growth, which may explain why more cases of Ewing’s sarcoma are seen in teenagers.

Like other cancers, it is not infectious and cannot be passed on to other people.

Signs and symptoms

Pain is the most common symptom of bone cancer. However, symptoms may vary depending on the position in the body and the size of the cancer. There may be some swelling in the affected area and it may become tender to touch. Bone cancer is sometimes discovered when a bone that has been weakened by cancer breaks after the child has a minor fall or accident.

How it is diagnosed

Usually you begin by seeing your family doctor (GP), who will examine your child and may arrange tests or x-rays. If a sarcoma is suspected, your GP will probably refer your child directly to a specialist hospital or bone tumour centre for further investigations. Many of the specific tests for diagnosing bone tumours, such as biopsies, need to be done by an experienced team using specialist techniques.

A variety of tests and investigations may be needed to diagnose Ewing’s sarcoma. An x-ray of the painful part of the bone is usually able to identify a tumour, although sometimes it can be difficult to see. Other tests are done to check whether the cancer has spread elsewhere. These include a chest x-ray, bone scan, bone marrow biopsy, and an MRI or CT scan.

Any tests and investigations that your child needs will be explained to you. The Macmillan/CCLG booklet A parent’s guide to children’s cancers gives details of what the tests and scans involve.

Treatment

A combination of treatments is used to treat Ewing’s sarcoma. These include chemotherapy, surgery and radiotherapy. Treatment will depend on a number of factors, including the size and position of the tumour.

Chemotherapy

Chemotherapy is the use of anti-cancer (cytotoxic) drugs to destroy cancer cells. This is an important treatment for most children with Ewing’s sarcoma and has greatly improved the results of surgical treatment. It’s often given before surgery. The course of chemotherapy will then continue after surgery in order to destroy any remaining cancer cells and prevent the sarcoma from spreading outside the bone. Chemotherapy given in this way is called adjuvant chemotherapy.

Surgery

If surgery is needed, it may be carried out at a specialist orthopaedic centre. Often, surgery can remove the tumour without causing too much damage. If the tumour
is in one of the main bones of the arm or leg, however, it may be necessary to remove the whole limb (amputation) or part of the affected bone. If only part of the affected bone is removed, this is known as limb-sparing surgery.

**Amputation** Sometimes amputation of the limb is unavoidable if the cancer has affected the surrounding blood vessels and nerves. After amputation, a false limb will be fitted, and this will be regularly adjusted as the child grows. False limbs can work very well. It should be possible for the child to join in with normal activities and even sport.

**Limb-sparing surgery** There are several ways in which limb-sparing surgery may be done:

- replacing the bone with a prosthesis (a specially designed artificial part)
- replacing the bone with bone taken from another part of the body (a bone graft).

After limb-sparing surgery, the child is usually able to use the limb almost normally. However, it’s best not to take part in any contact sports, because any damage to the bone graft or prosthesis may require another major operation to repair or replace it. If the child is growing, the limb prosthesis will need to be lengthened from time to time as the bone grows. This may mean further short stays in hospital, although some prostheses can be lengthened as an outpatient procedure.

**Radiotherapy**

Radiotherapy treats cancer by using high-energy rays that destroy cancer cells while doing as little harm as possible to normal cells. Ewing’s sarcoma responds very well to radiotherapy, and this treatment is often used after chemotherapy and before or after surgery. If surgery is not possible, for example if the tumour is in the spine, radiotherapy may be given instead of surgery.

**Side effects of treatment**

Treatment often causes side effects, and your child’s doctor will discuss this with you before the treatment starts. Any possible side effects will depend on the actual treatment being used and the part of the body that’s being treated. Side effects can include:

- feeling sick (nausea) and being sick (vomiting)
- hair loss
- increased risk of infection
- bruising and bleeding
- tiredness
- diarrhoea.

**Late side effects**

A small number of children may develop side effects, sometimes many years later. These include a possible reduction in normal bone growth, infertility, a change in the way the heart and lungs work, and a small increase in the risk of developing a second cancer later in life. Your child’s doctor or nurse will explain any possible late side effects.

More detailed information about these late side effects is available in the Macmillan/CCLG booklet, *A parent’s guide to children’s cancers*.

**Clinical trials**

Many children have their treatment as part of a clinical research trial. Trials aim to improve our understanding of the best way to treat an illness, usually by comparing the standard treatment with a new or modified version. Specialist doctors carry out trials for children’s cancer.

If appropriate, your child’s medical team will talk to you about taking part in a clinical trial and will answer any questions you may have. Written information is provided to help explain things.
Currently a large research study called the Euro-Ewing 99 trial is being carried out for children, young people and adults up to the age of 50 affected by Ewing's sarcoma. It's looking at the effects of surgery, chemotherapy and radiotherapy.

Before any trial is allowed to take place, it must have been approved by an ethics committee, which protects the interests of patients taking part. Your doctor or a research nurse must discuss the treatment with you so that you have a full understanding of the trial and what it means for your child to take part. You may decide not to take part or withdraw from a trial at any stage. Your child will then receive the best standard treatment available.

Taking part in a research trial is completely voluntary and you'll be given plenty of time to decide if it's right for your child. Your child's doctor or specialist nurse can tell you more about this.

**Follow-up**

Your child will have regular follow-up appointments, with x-rays or scans as necessary.

Many children with Ewing's sarcoma can be cured. Even if the tumour comes back, further treatment may be given successfully. Surgery and radiotherapy can stop bones growing and may make muscles and tendons shorter. This may mean that further operations will be necessary later in life. However, most children who are successfully treated for Ewing's sarcoma grow up without major disability.

If you have specific concerns about your child's condition and treatment, it's best to discuss them with your child's doctor, who knows the situation in detail.

**Your feelings**

As a parent, the fact that your child has cancer is one of the worst situations you can be faced with. You may have many emotions, such as fear, guilt, sadness, anger and uncertainty. These are all normal reactions and are part of the process that many parents go through at such a difficult time.

It's not possible to address in this fact sheet all of the feelings you may have. However, the Macmillan/CCLG booklet *A parent’s guide to children’s cancers* talks about the emotional impact of caring for a child with cancer and suggests sources of help and support.

Your child may have a variety of powerful emotions throughout their experience of cancer. The parent's guide discusses this further and talks about how you can support your child.

Our booklet *Peppermint Ward* is a storybook for younger children with cancer. It looks at the issues that they and their family may face and helps them explore their feelings.

**Useful organisations**

**Children's Cancer and Leukaemia Group (CCLG)**
University of Leicester, 3rd Floor, Hearts of Oak House, 9 Princess Road West, Leicester LE1 6TH
Tel 0116 249 4460
Email info@cclg.org.uk
www.cclg.org.uk
Coordinates research and care for children and their parents. There are 21 CCLG specialist centres for the treatment of childhood cancer and leukaemia, covering all areas of the UK and Ireland (there's a map of the centres on the website).
Has information about the CCLG, childhood cancer and leukaemia.

**CLIC Sargent**
Griffin House, 161 Hammersmith Road, London W6 8SG
Tel 0300 330 0803
Email info@clicsargent.org.uk
www.clicsargent.org.uk
Offers practical support to children and...
young people with cancer or leukaemia, and to their families.

**Related Macmillan information**

- A parent's guide to children's cancers
- Katie's Garden (a storybook for primary school-age children about a girl's experience of cancer)
- Peppermint Ward – A story about children with cancer
- Understanding cancer research trials
- Understanding chemotherapy
- Understanding radiotherapy

For copies of this related information call free on **0808 808 00 00**, or see it online at [macmillan.org.uk](http://macmillan.org.uk)

Our website [click4tic.org.uk](http://click4tic.org.uk) has cancer information written specifically for young people. Get advice and support, and connect with other young people affected by cancer.

This fact sheet has been written, revised and edited by Macmillan Cancer Support's Cancer Information Development team. It has been approved by our medical editor, Dr Terry Priestman, Consultant Clinical Oncologist.

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This fact sheet has been compiled using information from a number of reliable sources, including:


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