Ewing sarcoma is a type of bone cancer in children and young people. This information describes Ewing sarcoma, its symptoms, diagnosis and possible treatments.

More children than ever are surviving childhood cancer. There are now new and better drugs and treatments, and we can now also work to reduce the after-effects of having had cancer in the past.

It is devastating to hear that your child has cancer, and at times it can feel overwhelming but there are many healthcare professionals and support organisations to help you through this difficult time.

Understanding more about the cancer your child has, and the treatments that may be used, can often help parents to cope. We hope you find the information here helpful. Your child’s specialist will give you more detailed information and if you have any questions it is important to ask the specialist doctor or nurse who knows your child’s individual situation.

This factsheet is published in conjunction with CCLG’s booklet entitled: ‘Children and Young People with Cancer: A Parent’s Guide’.

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 sarcoma

Ewing sarcoma is the second most common primary sarcoma in children and young people. This type of cancer is a bone cancer and most commonly occurs in the long bones, ribs, pelvis and spine (vertebral column).

Fewer than 30 children in the UK develop Ewing sarcoma each year. It usually occurs in the teenage years, and is more common in boys.

Although Ewing sarcoma is a type of bone cancer, it can also occur very rarely in the soft tissues. This is called extraosseous Ewing sarcoma. Another type of Ewing sarcoma is a primitive neuroectodermal tumour (PNET). These can be found in either the bone or soft tissue.

Causes

The exact causes of primary bone cancer are unknown. The development of Ewing sarcoma may be related in some way to times of rapid bone growth, which may explain why more cases are seen in teenagers. Like other cancers, it’s not infectious and cannot be passed on to other people.

Signs and symptoms

Pain is the most common symptom of bone cancer. It is frequently worse at night. However, symptoms may vary depending on the position and size of the cancer. There may be some swelling in the affected area if the tumour is close to the surface of the body and it may become tender to touch. This may cause a limp if in the leg or pelvis. Bone cancer is sometimes discovered when a bone that has been weakened by cancer breaks after the child has a minor fall or accident. Occasionally, there may be fever or weight loss.
How Ewing sarcoma is diagnosed

Usually you begin by seeing your GP, who will examine your child and may arrange tests or x-rays. If a sarcoma is suspected, your GP should refer your child directly to a specialist hospital or bone tumour centre. A variety of tests and investigations are needed to diagnose Ewing sarcoma, including an x-ray of the painful part of the bone, a chest x-ray and a blood test. A specialist doctor will remove a small piece of the tumour to look at under a microscope (biopsy). Other tests may be done, such as a bone scan, PET scan, a bone marrow biopsy; an MRI or CT scan may also be done.

Any tests and investigations that your child needs will be explained to you.

Treatment

A combination of various treatments is used to treat Ewing 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 a very important component of treatment for the majority of children with Ewing sarcoma and can help make surgery more straightforward. A combination of different chemotherapy drugs are given before surgery and continued afterwards in order to destroy any remaining cancer cells and prevent the sarcoma from spreading. Chemotherapy given in this way is called adjuvant chemotherapy.

Surgery

If surgery is needed, it should be carried out at a specialist orthopaedic bone tumour centre. The aim of surgery is to 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 your child grows. False limbs work very well. It should be possible for your 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. It may involve:

- 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 often able to use the limb almost normally. However, it is 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 during 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 sarcoma responds very well to radiotherapy. It is often used after chemotherapy and before or after surgery. If the tumour is impossible to remove surgically, it is a good option.

Side effects of treatment for Ewing sarcoma

Treatment often causes side effects. 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

Radiotherapy can cause irritation or soreness of the skin in the area being treated and general tiredness. If your child is having surgery, the surgeon will explain the possible complications of the surgery that your child is having.
Late side effects

A small number of children may develop other side effects, sometimes many years later. These include a reduction in normal bone growth, reduced fertility, a change in heart function, and a small increase in the risk of developing a second cancer later in life. Your child’s doctor or nurse will explain all of this to you and will monitor your child carefully for any potential late side effects.

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 cancers. If appropriate, your child’s medical team will talk to you about taking part in a clinical trial, and will answer any questions you have. Written information will be provided to help explain things.

Treatment guidelines

Sometimes, clinical trials are not available for your child’s tumour. This may be because a recent trial has just finished, or because the tumour is very rare. In these cases, you can expect your doctors and nurses to offer treatment which is agreed to be the most appropriate, using guidelines which have been prepared by experts across the country. The Children’s Cancer and Leukaemia Group (CCLG) is an important organisation which helps to produce these guidelines.

Follow-up care

Your child will have regular follow-up appointments, with x-rays or scans as necessary. Many children with Ewing sarcoma can be cured. Even if the tumour comes back, further treatment may be given successfully. Your child will be checked up on every few months for the first three years after treatment and then every six months for another two years.

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 is not possible to address in this factsheet all of the feelings you may have. However, the CCLG booklet ‘Children & Young People’s Cancer; A Parent’s Guide’, 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 these further and talks about how you can support your child.
Useful organisations

Children's Cancer and Leukaemia Group (CCLG)
www.cclg.org.uk
CCLG supports the 1,700 children who develop cancer each year in the UK. It gives support to healthcare professionals involved in caring for children with cancer and is key to the development of high standards of care.

CLIC Sargent
www.clicsargent.org.uk
CLIC Sargent offers practical support to children and young people with cancer or leukaemia, and to their families.

Macmillan Cancer Support
www.macmillan.org
Offers support and advice to those affected by cancer.

Bone Cancer Research Trust
www.bcrt.org.uk
Information and support for patients and their families.

References

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

- Grimer et al. UK Guidelines for the Management of Bone Sarcoma. Sarcoma 2010