Children's Cancer and Leukaemia Group | the EXPERTS in CHILDHOOD CANCER

Ewing sarcomas in children

An information guide for parents, carers and families

The purpose of this guide is to give information about Ewing sarcoma to help you understand more about the type of cancer your child has.

Information in this guide should be used to support professional advice specific to your child's diagnosis. If you have any questions, it is important to ask your child's medical team.

What are sarcomas?

Sarcomas are rare tumours that develop in the supporting tissues of the body, such as bone, muscle, or cartilage.

There are two main types of sarcomas:

- **bone sarcomas** can develop in any of the bones of the skeleton
- **soft tissue sarcomas** can develop in muscle, fat, blood vessels, or in any of the other tissues that support, surround, and protect the organs of the body

What are Ewing sarcomas?

Ewing sarcoma is the second most common primary bone sarcoma in children and young people. Approximately, 30 children in the UK develop Ewing sarcoma each year. It usually occurs in the teenage years and is more common in boys.

Ewing sarcoma usually occurs in the long bones such as legs and arms, ribs, pelvis and spine. Very rarely, it can also occur in the soft tissues. This is called extraosseous Ewing sarcoma.

The cells in a Ewing sarcoma have a specific genetic change within them that confirms the diagnosis. There are other Ewing sarcoma-like tumours that occur in bone or soft tissue, including primitive neuroectodermal tumour (PNET), BCOR or CIC-DUX rearranged tumours.

Causes of Ewing sarcoma

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.

It is important to remember that nothing you have done has caused the cancer.

Signs and symptoms

Pain is the most common symptom of bone cancer. It is often worse at night. 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 found when a bone that has been weakened by cancer breaks after a minor fall or accident. Occasionally, there may be fever or weight loss.

How it is diagnosed

A variety of tests and investigations are needed to diagnose Ewing sarcoma, including an x-ray and MRI scan of the painful part of the bone, a chest x-ray or CT scan and blood tests. 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 or a bone marrow biopsy. 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 drugs to destroy cancer cells. This is a very important component of treatment for Ewing sarcoma and may 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.

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.

Surgery

If surgery is needed, it will 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. Limb-sparing surgery will be performed if at all possible.

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 can work very well. It should be possible for your child to join in with normal activities and even sport.

Limb-sparing surgery

There are two 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 this type of surgery, children are often able to use their limbs almost normally. However, it is best not to participate in any contact sports. This is because any damage to the bone graft or prosthesis may require another operation to repair or replace it.

If your child is growing, the limb prosthesis will need to be lengthened as the bone grows. This may mean there are short stays in hospital, although some prostheses can be lengthened as an outpatient procedure.

Side effects during treatment

Treatment for Ewing sarcoma can cause side effects, and your child's doctor will discuss these with you before the treatment starts. Any side effects will depend on the part of the body that's being treated and what treatment is being used.

Most side effects are short-term and gradually disappear once treatment stops. Side effects can include:

- feeling sick (nausea) and being sick (vomiting)
- temporary hair loss
- tiredness
- low blood count leading to an increased risk of infection and bruising and bleeding (sometimes blood and/or platelet transfusions are needed)
- a sore mouth and tummy
- diarrhoea
- pins and needles in fingers and toes

Radiotherapy can make your child feel tired, and the skin in the area that's being treated may go red or get darker.

Relapse

Many children with Ewing sarcoma are treated successfully. If the cancer comes back further treatment can be given, although a cure will be more challenging.

Your child may be offered the opportunity to take part in a clinical trial of a new drug. Your child's doctor will discuss all the options with you.

Clinical trials

Many children have their treatment as part of a clinical research trial or study. Clinical trials are carried out to try to improve our understanding of the best way to treat an illness, usually by comparing the standard treatment with a new or modified version. Clinical trials mean there are now better results for curing children's cancers compared with just a few years ago.

Your child's medical team will talk to you about taking part in a clinical trial and will answer any questions you have. 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. You may decide not to take part, or you can withdraw from a trial at any stage. You will still receive the best treatment available.

National treatment guidelines

Sometimes, clinical trials are not available for your child's tumour. In this case, your doctors will still offer the most appropriate treatment, using guidelines which have been agreed by experts across the UK. Children's Cancer and Leukaemia Group (CCLG) is an important organisation which helps to produce these guidelines.

Donating to a tissue bank

Ewing sarcoma is a rare disease and more research is needed to help doctors develop better treatment for the future. Your child's hospital team will offer you the opportunity to anonymously donate tissue left over from tests carried out, for example, a biopsy or surgery, to the tissue bank. This sample of tissue can then be used by scientists to learn more about Ewing sarcoma and how best to treat it. This is voluntary, and you will have plenty of time to decide if you wish to take part.

Late side effects

Months or years later some children may develop late side effects from the treatment they have had. These may include a reduction in bone growth, a change in the way the heart, lungs and kidneys work, a risk of infertility and a small increase in the risk of developing another cancer in later life. For more information please visit www.cclg.org.uk/life-after-childhood-cancer.

Your child's doctor or nurse will talk to you about any possible late side effects and will keep a close eye on possible long-term side effects in follow-up clinics.

Follow-up care

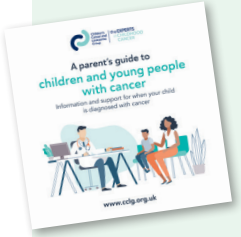
Once treatment has finished, the doctors will monitor your child closely with regular appointments and x-rays or scans where necessary to be sure that the cancer has not come back and there are no complications. After a while, you will not need to visit the clinic so often.

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

It's devastating to hear that your child has cancer and you may feel overwhelmed, but there are many professionals and organisations to help you through this difficult time. You may have many feelings, such as fear, guilt, sadness, anger, and uncertainty. These are all normal reactions and are part of the process that many parents go through.

It's not possible to address in this guide all of the feelings you may have. However, the CCLG booklet 'A parent's guide to children and young people with cancer', 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.



The CCLG booklet 'A parent's guide to children and young people with cancer' is available FREE of charge from your child's hospital

All CCLG booklets and leaflets can be downloaded or ordered from our website:

www.cclg.org.uk/publications

USEFUL ORGANISATIONS

Children's Cancer and Leukaemia Group (CCLG) publishes a variety of free resources to order or download.
www.cclg.org.uk

Young Lives vs Cancer offers practical support to children and young people with cancer and to their families
www.younglivesvscancer.org.uk

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

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

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Children's Cancer and Leukaemia Group (CCLG) is a leading national charity and expert voice for all childhood cancers.

Each week in the UK and Ireland, more than 30 children are diagnosed with cancer. Our network of dedicated professional members work together in treatment, care and research to help shape a future where all children with cancer survive and live happy, healthy and independent lives.

We fund and support innovative world-class research and collaborate, both nationally and internationally,to drive forward improvements in childhood cancer. Our award-winning information resources help lessen the anxiety, stress and loneliness commonly felt by families, giving support throughout the cancer journey.

Our work is funded by donations. If you would like to help, text '**CCLG**' to **70300** to donate **£3**. You may be charged for one text message at your network's standard or charity rate. CCLG (registered charity numbers 1182637 and SC049948) will receive 100% of your donation.

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