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

Osteosarcomas in children

An information guide for parents, carers and families

The purpose of this guide is to give information about osteosarcoma 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 osteosarcomas?

Osteosarcoma is a type of bone cancer. About 35 children in the UK develop osteosarcomas each year.

These tumours occur more commonly in older children and teenagers and are very rarely seen in children under five.

Osteosarcoma often starts at the end of the long bones, where new bone tissue forms as a young person grows. Any bone in the body can be affected, but the most common sites are in the arms and legs, particularly around the knee and shoulder joints.

There are several different types of osteosarcoma. Most occur in the centre of the bone. There are also rare subtypes, such as parosteal, periosteal, telangiectatic, and small cell osteosarcoma.

Causes of osteosarcoma

The exact cause of osteosarcoma is not known. However, there are some factors which are associated with osteosarcoma. Children who have hereditary retinoblastoma (a rare tumour of the eye) have an increased risk of developing osteosarcoma. Children who have previously had radiotherapy and chemotherapy also have an increased risk of developing osteosarcoma. It is not caused by injuries or damage to the bone, although an injury may draw attention to a bone tumour.

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

Signs and symptoms

Pain in the affected bone is the most common symptom. This pain may initially come and go, and then gradually become more severe and constant, especially at night. There may also be swelling around the affected bone.

Osteosarcoma is sometimes discovered when a bone that has been weakened by cancer breaks after the person has had a minor fall or accident. Some patients may develop a limp if the tumour is in the leg or pelvis.

How it is diagnosed

A variety of tests and investigations may be needed to diagnose an osteosarcoma. An x-ray of the painful part of the bone will usually identify a tumour, although sometimes they can be difficult to see.

A small piece of the tumour will be removed and looked at under a microscope. This is called a biopsy. It's a small

operation, performed under a general anaesthetic. Other tests are done to check whether the cancer has spread to other parts of the body. These include a chest x-ray, blood tests, a bone scan and an MRI or CT scan.

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

Grading

Grading refers to the appearance of the cancer cells under the microscope, and gives an idea of how quickly the cancer may develop. Low-grade cancer cells are usually slow-growing and less likely to spread. In high-grade tumours the cells are likely to grow quickly and are more likely to spread. Most osteosarcomas are high-grade, but a type known as parosteal osteosarcoma is usually low-grade.

Staging

The 'stage' of a cancer is a term used to describe its size and whether it has spread beyond its original site. Knowing the particular type and stage of the cancer helps the doctors to decide on the most appropriate treatment. Most patients are grouped depending on whether cancer is found in only one part of the body (localised disease), or whether the cancer has spread from one part of the body to another (metastatic disease).

The staging system commonly used for osteosarcoma is described below:

- **Stage 1A** - the tumour is low-grade and is only found within the hard coating of the bone.
- **Stage 1B** - the tumour is low-grade, extending outside the bone and into the soft tissue spaces that contain nerves and blood vessels.
- **Stage 2A** - the tumour is high-grade and is completely contained within the hard coating of the bone.
- **Stage 2B** - the tumour is high-grade and has spread outside the bone and into surrounding soft tissue spaces that contain nerves and blood vessels. Most osteosarcomas are stage 2B.
- **Stage 3** - the tumour can be low or high-grade and is either found within the bone or extends outside the bone. The cancer has spread to other parts of the body, or to other bones not directly connected to the bone where the tumour started.

Treatment

Treatment will depend on a number of factors including the size, position, and stage of the tumour.

Chemotherapy

Chemotherapy is a very important part of treatment for osteosarcoma. Chemotherapy uses anti-cancer (cytotoxic) drugs to destroy cancer cells, and is usually given to shrink the main tumour before surgery. It's also given after the tumour has been removed by surgery, to help reduce the risk of the cancer coming back. It's common for a combination of drugs to be used.

Radiotherapy

Radiotherapy may occasionally be given. This treats cancer by using high energy rays to destroy the cancer cells, while doing as little harm as possible to normal cells.

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 is affecting the surrounding blood vessels and nerves.

After amputation, a false limb will be fitted and 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 sports.

Limb-sparing surgery

There are two ways that limb-sparing surgery may be done:

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

After this type of surgery, children are often able to use their limbs almost normally. However, they are advised 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.

Biological therapy

Mifamurtide, also called Mepact®, is a new type of biological therapy treatment. It may be offered to your child if they have had an operation to remove the osteosarcoma and the tumour has not spread. It will be given along with chemotherapy after surgery to try to reduce the risk of the osteosarcoma coming back.

Mifamurtide is a relatively new drug and so more research needs to be done to fully understand its side effects and how to use it best.

Side effects during treatment

Treatment for osteosarcoma 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

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

Clinical trials

Many children have their treatment as part of a clinical research trial. Clinical trials and studies are carried out to try to improve our understanding of the best way to treat an illness. There are better results for curing children's cancers compared with just a few years ago because of clinical trials.

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 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. Your child 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

Osteosarcoma 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 osteosarcoma 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.

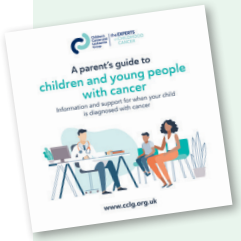
Follow-up care

Once treatment has finished, the doctors will monitor your child closely with regular appointments 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

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

Children's Cancer and Leukaemia Group
Century House, 24 De Montfort Street
Leicester LE1 7GB

0333 050 7654
info@cclg.org.uk | www.cclg.org.uk

ChildrensCLG CCLG_UK

Registered charity in England and Wales (1182637) and Scotland (SC049948).

This edition: August 2022
Next review date: August 2025

Trusted Information Creator
Patient Information Forum

With thanks to Dr Rachael Windsor, Consultant Paediatric and Adolescent Oncologist at University College London Hospitals NHS Foundation Trust and CCLG member, who reviewed this factsheet on behalf of the CCLG Information Advisory Group, comprising multi-professional experts in the field of children's cancer.

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.

We are grateful to all those who have contributed to this publication. We make every effort to ensure that this information is accurate and up to date at the time of printing. CCLG does not accept any responsibility for information provided by third parties including those referred to or signposted to in this publication. Information in this publication should be used to supplement appropriate professional or other advice specific to your circumstances.

If you have any comments on this factsheet, please contact us at publications@cclg.org.uk