Osteosarcoma

Osteosarcoma is a type of bone cancer. About 30 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.

More children than ever are surviving childhood cancer. There are 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. If you have any questions it's important to ask the specialist doctor or nurse who knows your child's individual situation.

This factsheet has been published in conjunction with CCLG's booklet entitled: 'Children and Young People with Cancer: A Parent's Guide'.

Osteosarcoma

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

There has been a lot of research into the causes of osteosarcoma but, like most childhood cancers, a definite cause is unknown. There are a few risk factors that have been 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.

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. These symptoms can be caused by many things other than cancer. However, any persistent bone pain should be checked by your child's doctor. Symptoms are often attributed to a sporting injury.

How osteosarcoma is diagnosed

Usually you begin by seeing your GP, who will examine your child and may arrange tests or x-rays. Many of the specific tests for diagnosing bone tumours require experience and specialist techniques so if a bone tumour is suspected, the doctor will refer your child directly to a specialist hospital or bone tumour centre.
for further tests. The doctor at the hospital will take your child’s full medical history. They will then do a physical examination. This will include an examination of the painful bone to check for any swelling or tenderness. Your child will probably have a blood test to check their general health.

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). A staging system commonly used for osteosarcomas is described below:

<table>
<thead>
<tr>
<th>Stage</th>
<th>Description</th>
</tr>
</thead>
<tbody>
<tr>
<td>1A</td>
<td>The cancer is low-grade and is only found within the hard coating of the bone.</td>
</tr>
<tr>
<td>1B</td>
<td>The cancer is low-grade, extending outside the bone and into the soft tissue spaces that contain nerves and blood vessels.</td>
</tr>
<tr>
<td>2A</td>
<td>The cancer is high-grade and is completely contained within the hard coating of the bone.</td>
</tr>
<tr>
<td>2B</td>
<td>The cancer 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.</td>
</tr>
<tr>
<td>3</td>
<td>The cancer 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.</td>
</tr>
</tbody>
</table>

If the cancer comes back after initial treatment, this is known as recurrent or relapsed cancer.

**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 (recurring). 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**

The type and extent of surgery depends on the position and size of the tumour in the body. This surgery will need to be carried out at a specialist orthopaedic centre, and your child should be referred to one. Surgery may include removing the whole limb (amputation) or part of the affected bone, which is then replaced by some form of false limb (prosthesis). If only part of the affected bone is removed, this is known as limb-sparing surgery. Most children will be considered for limb-sparing surgery.

**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 will usually be 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 major 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 extra 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 a surgical resection to remove the osteosarcoma and the tumour has not spread. It will be given along with chemotherapy after surgery to try and stop 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 of treatment
Treatment often causes side effects, and your child’s doctor will discuss these with you before the treatment starts. Any possible side effects will depend upon the treatment being given and the part of the body that’s being treated.

Chemotherapy side effects
Some general side effects of chemotherapy can include:
- feeling sick (nausea) and being sick (vomiting)
- hair loss
- increased risk of infection bruising and bleeding tiredness
- diarrhoea

Radiotherapy side effects
Radiotherapy can cause irritation or soreness of the skin in the area being treated and general tiredness.

Surgery side effects
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 late side effects, sometimes many years later. These include a reduction in bone growth, reduced fertility, a change in heart function, and a slight increase in the risk of developing another cancer in later life.

Your child’s doctor or nurse will talk to you and monitor your child for any potential late effects.

Clinical trials
Many children have their treatment as part of a clinical research trial, if there is a suitable one open. 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 have. Written information will be provided to help explain things.

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 can tell you if there are any trials available that might be suitable for your child.

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
Many children with osteosarcoma are cured. However, your child may need to have surgery to lengthen the affected limb as they grow. Your child will have regular check-ups and x-rays in the paediatric or adolescent oncology clinic, and at the orthopaedic centre. If you have specific concerns about your child’s condition and treatment, it is best to discuss them with your child’s doctor, who knows the situation in detail.

USEFUL ORGANISATIONS

Children’s Cancer and Leukaemia Group (CCLG)  www.cclg.org.uk

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.uk
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 factsheet has been compiled using information from a number of reliable sources, including:

- National Institute for Health and Clinical Excellence (NICE). Improving Outcomes for People with Sarcoma. 2006