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

Rhabdomyosarcoma in children

An information guide for parents, carers and families

The purpose of this guide is to give information about rhabdomyosarcoma 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:

- 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
- bone sarcomas can develop in any of the bones of the skeleton

What is rhabdomyosarcoma?

Rhabdomyosarcoma is the most common soft tissue sarcoma in children. Less than 60 children are diagnosed in the UK each year. Most of them are younger than 10 years old. It is more common in boys than in girls.

Rhabdomyosarcoma has muscle-like features and can grow in any part of the body. The most common areas of the body to be affected are around the head and neck, bladder, testes, uterus, or vagina.

Sometimes rhabdomyosarcoma is found in a limb, in the chest or in the abdominal wall. If the tumour is in the head or neck area, it can occasionally spread into the brain.

In children, about 20% of rhabdomyosarcomas have a gene rearrangement involving the PAX and FOXO1 genes (fusion positive), and the remaining tumours do not have the gene fusion (fusion negative).

The genetic information of the tumours is important to help doctors to decide on the best treatment for your child.

Causes of rhabdomyosarcoma

We don't know what causes rhabdomyosarcoma but there is research going on all the time to try to find out.

Children with certain rare genetic disorders, such as Li-Fraumeni syndrome, have a higher risk of developing rhabdomyosarcoma.

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

Signs and symptoms

The most common sign of rhabdomyosarcoma is a lump or swelling. Other symptoms will depend on the part of the body that's affected by the rhabdomyosarcoma:

- a tumour in the head or neck area can sometimes cause a blockage (obstruction) and discharge from the nose or throat. Occasionally, an eye may appear swollen and protruding.
- a tumour in the tummy (abdomen) can cause pain or discomfort in the tummy and difficulty going to the toilet (constipation).
- a tumour in the bladder may cause symptoms such as blood in the urine and difficulty being able to wee.

How is it diagnosed?

Different tests are usually needed to diagnose a rhabdomyosarcoma. Your child may need a small operation to remove a sample from the tumour (a biopsy) so that it can be examined under a microscope. This is usually done under a general anaesthetic.

Tests may be done to check the exact size of the tumour, to check whether the tumour is fusion positive or fusion negative and to find out if it has spread to any other part of the body. These may include:

- a chest x-ray to check the lungs
- an ultrasound scan
- CT, PET/CT or MRI scans
- blood and bone marrow tests
- molecular gene testing

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

Staging

The stage of a cancer is a term used to describe its size and whether it has spread from where it first started. Knowing the stage helps the doctors decide on the most effective treatment for your child.

The staging system for rhabdomyosarcoma is based on:

- where in the body the tumour started
- if it is in only one part of the body (localised disease), or if it has spread to another part of the body (metastatic disease).

There are different ways of staging rhabdomyosarcoma, and your child's doctor will explain more about the system they are using.

Treatment

Treatment will depend on the size of the tumour, the type of rhabdomyosarcoma, its position in the body and whether it has spread.

The three main types of treatment for soft tissue sarcomas are chemotherapy, surgery, and radiotherapy. Your child may have a combination of treatments.

Chemotherapy

Chemotherapy is the use of anti-cancer drugs to destroy cancer cells.

It can be given:

- to shrink the tumour before surgery
- after surgery to reduce the risk of the cancer returning

The drugs used and the length of treatment depends on the type and stage of the rhabdomyosarcoma.

Surgery

If it is possible, your child will have an operation to remove all or as much as possible of the tumour, without damaging surrounding tissue or organs. The operation will depend on the size of the tumour and where it is in your child's body. The surgeon will explain what is involved.

Chemotherapy is usually given before surgery to shrink the tumour and make it easier to remove. If an operation isn't possible, both chemotherapy and radiotherapy are given.

Radiotherapy

Radiotherapy treats cancer by using high energy rays, which destroy the cancer cells while doing as little harm as possible to normal cells. It may be given after surgery to the area where the rhabdomyosarcoma started.

Depending on the site of the rhabdomyosarcoma, there are different radiotherapy techniques, and this may mean travelling to a different treatment centre. Examples include brachytherapy and proton beam.

Sometimes patients are offered radiotherapy before surgery, rather than after surgery.

Side effects of treatment

Treatment for rhabdomyosarcoma 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

treatment being given and the part of the body that is being treated.

Most side effects are short-term and gradually disappear once treatment stops.

Chemotherapy may cause side effects such as:

- feeling sick (nausea) and being sick (vomiting)
- temporary hair loss
- tiredness
- a 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.

Relapse

Unfortunately, when rhabdomyosarcoma relapses it can be difficult to treat. However, there are treatments available, including chemotherapy and sometimes radiotherapy.

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. 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 then receive the best standard treatment available.

National treatment guidelines

Sometimes, clinical trials are not available for your child's tumour. In these cases, your doctors will 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

Rhabdomyosarcoma 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 bone marrow test, to the tissue bank. This sample of tissue can then be used by scientists to learn more about rhabdomyosarcoma 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. It is important to understand that not all late effects will happen to all patients. 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

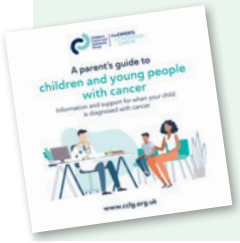
Once treatment has finished, the doctors will monitor your child closely with regular appointments to be sure that the cancer has not come back and there are no complications. After a while, you will not need to visit the clinic so often.

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 their 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

European Paediatric Soft Tissues Sarcoma Study Group (EpSSG) is an international organisation for professionals involved with the care and treatment of children and young people with soft tissue sarcomas.
www.epssgassociation.it

References

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

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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. This will cost £3 plus a standard rate message.

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